The Craniovertebral Junction: Diagnosis, Pathology, Surgical Techniques

Atul Goel, MCh
Professor and Head, Department of Neurosurgery
Seth G.S. Medical College and King Edward Memorial Hospital
Parel, Mumbai, India
Consultant Neurosurgeon
Tata Memorial Hospital and Lilavati Hospital and Research Center
Mumbai, India

Francesco Cacciola, MD
Consultant Neurosurgeon
U.O.C. Neurochirurgia 1
Azienda Ospedaliera Universitaria Senese
Policlinico Santa Maria alle Scotte
Siena, Italy

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Dedicated

To my parents

—Atul Goel
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Contributors

Max Aebi, MD, DHC, FRCSC
Chief of Staff, Department of Orthopaedic Surgery
Hirslanden-Salem Spital
Bern, Switzerland

Kuniyoshi Abumi, MD, DrMedSci
Professor of Spinal Reconstruction
Hokkaido University Graduate School of Medicine
Sapporo, Japan

Antonio Barbieri, MD
E. Morelli Hospital
Sondalo, Italy

Sanjay Behari, MCh, DNB
Department of Neurosurgery
Sanjay Gandhi Postgraduate Institute of Medical Sciences
Lucknow, India

Edward C. Benzel, MD
Chairman, Department of Neurosurgery
Center for Spine Health
Neurological Institute
Cleveland Clinic
Cleveland, Ohio

Paolo Bolognese, MD
The Chiari Institute
Northshore-Long Island Jewish Health System
Great Neck, New York

Nishigandha Burute, MBBS, MD
Radiologist
Piramal Diagnostics-Jankharia Imaging
Mumbai, India

Francesco Cacciola, MD
Consultant Neurosurgeon
U.O.C. Neurochirurgia 1
Azienda Ospedaliera Universitaria Senese
Policlinico Santa Maria alle Scotte
Siena, Italy

Alexandre Carpentier
Department of Neurosurgery
Hôpital La Salpetrière
Paris, France

Ricardo L. Carrau, MD
Professor of Otolaryngology, Head and Neck Surgery, and Neurosurgery
Director of the Maxillofacial Trauma Service
University of Pittsburgh Medical Center
Pittsburgh, Pennsylvania

Ondrej Choutka
Department of Neurosurgery
University of Cincinnati
Cincinnati, Ohio

Neil R. Crawford, PhD
Associate Staff Scientist
Spinal Biomechanics Laboratory
Division of Neurological Surgery
Barrow Neurological Institute
Phoenix, Arizona

Nitin Dange, MCh
Assistant Professor of Neurosurgery
Seth G.S. Medical College and K.E.M Hospital
Parel, Mumbai, India

Nicola Desogus, MD
Department of Neurosurgery
Livorno City Hospital
Livorno, Italy

Prakash Dhande, MVSc, PhD
Professor of Veterinary Anatomy, Histology and Embryology
Bombay Veterinary College
Parel, Mumbai, India

Nicola Di Lorenzo, MD, PhD, FACS
Professor and Head of Department
Clinica Neurochirurgica Università di Firenze
Firenze, Italy

Michael G. Fehlings, MD, PhD
Division of Neurosurgery
Toronto Western Hospital
Toronto, Ontario, Canada

Richard G. Fessler, MD, PhD
University of Chicago Hospitals
Chicago, Illinois
Manu Kothari, MS  
Professor Emeritus of Anatomy  
Seth G.S. Medical College and K.E.M Hospital  
Parel, Mumbai, India

Craig A. Kuhns, MD  
Assistant Professor of Orthopaedic Surgery  
University of Missouri-Columbia  
Missouri Spine Center  
Columbia, Missouri

Vinod Laheri, MS  
Professor Emeritus of Orthopedics  
Seth G.S. Medical College and K.E.M Hospital  
Parel, Mumbai, India

Giuseppe Lanzino, MD  
Department of Neurologic Surgery  
Mayo Clinic  
Rochester, Minnesota

Ronald A. Lehman Jr., MD  
Chief, Pediatric and Adult Spine  
Chief, Division of Orthopaedics and Rehabilitation, USUHS  
Walter Reed Army Medical Center  
Bethesda, Maryland

Gordon Li, MD  
Department of Neurosurgery  
Stanford University Medical Center  
Stanford, California

Shraddha Maheshwari, MD  
Chief Resident of Neurosurgery  
Lilavati Hospital and Research Center  
Bandra, Mumbai, India

Arnold H. Menezes, MD  
Professor and Vice Chairman  
Department of Neurosurgery  
University of Iowa Hospitals and Clinics  
Iowa City, Iowa

Thomas H. Milhorat, MD  
The Chiari Institute  
Northshore-Long Island Jewish Health System  
Great Neck, New York

Ranjith K. Moorthy, MD  
Department of Neurological Sciences  
Christian Medical College  
Vellore, India

Nobuhito Morota, MD  
Chief, Division of Neurosurgery  
National Medical Center for Children and Mothers  
National Center for Child Health and Development  
Tokyo, Japan

Praveen V. Mummaneni, MD  
Associate Professor of Neurosurgery  
Co-director, UCSF Spine Center  
University of California, San Francisco  
San Francisco, California

Dattatraya Muzumdar, MCh  
Associate Professor of Neurosurgery  
Seth G.S. Medical College and K.E.M Hospital  
Parel, Mumbai, India

Trimurti Nadkarni, MCh  
Professor of Neurosurgery  
K.E.M Hospital and Seth G.S. Medical College  
Parel, Mumbai, India

Suresh Nayak, MCh  
Department of Neurosurgery  
Sanjay Gandhi Postgraduate Institute of Medical Sciences  
Lucknow, India

Alfred T. Ogden, MD  
Assistant Professor of Neurological Surgery  
Columbia University  
New York, New York

Kenji Ohata, MD  
Professor and Chairman  
Department of Neurosurgery  
Osaka City University Graduate School of Medicine  
Osaka, Japan

Umesh Phalke, MCh  
Professor  
Dr. D.Y Patil Medical College  
Pimpri, Pune, India

Daniel M. Prevedello, MD  
Assistant Professor of Neurological Surgery  
Training Program Director  
Center for Skull Base Surgery  
University of Pittsburgh School of Medicine  
Pittsburgh, Pennsylvania

Sanjay Rajan, MCh  
Assistant Professor of Neurosurgery  
V. M. Medical College and Safdarjung Hospital  
New Delhi, India

Vedantam Rajshekhar, MD  
Professor of Neurosurgery  
Department of Neurological Sciences  
Christian Medical College  
Vellore, India

Stylianos K. Rammos, MD  
Arkansas Neuroscience Institute  
Little Rock, Arkansas
K. Daniel Riew, MD
Mildred B. Simon Distinguished Professor of Orthopaedic Surgery
Chief, Cervical Spine Surgery
Washington University Orthopedics
Professor, Neurological Surgery
Washington University School of Medicine
St. Louis, Missouri

Chan Roonprapunt, MD, PhD
Assistant Professor of Neurosurgery
Albert Einstein College of Medicine
Roosevelt and Beth Israel Medical Centers
New York, New York

Stephen Ryu, MD
Department of Neurosurgery
Palo Alto Medical Foundation
Palo Alto, California
Department of Electrical Engineering
Stanford University
Stanford, California

Darshana Sanghvi, MD
Department of Neuroradiology
Kokilaben Dhirubhai Ambani Hospital and Medical Research Institute
Mumbai, India

Chandranath Sen, MD
Department of Neurosurgery
St. Luke’s Roosevelt Hospital Center
New York, New York

Abhidha Shah, MCh
Tutor in Neurosurgery
Seth G.S. Medical College and K.E.M Hospital
Parel, Mumbai, India

Raj K. Shrivastava, MD
Department of Neurosurgery
St. Luke’s Roosevelt Hospital Center
New York, New York

Namit Singhal, MCh
Department of Neurosurgery
Sanjay Gandhi Postgraduate Institute of Medical Sciences
Lucknow, India

Carl H. Snyderman, MD
UPMC Minimally Invasive EndoNeurosurgical Center
University of Pittsburgh Medical Center
Pittsburgh, Pennsylvania

Volker K. H. Sonntag, MD
Vice Chairman Emeritus
Barrow Neurological Institute
Phoenix, Arizona
Professor of Clinical Surgery
University of Arizona Medical School
Tucson, Arizona

Petr Suchomel, MD, PhD
Associate Professor, Department of Neurosurgery
Head of Neurocenter
Regional Hospital
Liberec, Czech Republic

Meher Ursekar, MBBS, DMRD, MD
Consultant Neuroradiologist
Piramal Diagnostics-Jankharia Imaging
Mumbai, India

K. Michael Webb, MD
Executive Medical Director
Director of Spinal Disorder Program
NeuroTexas Institute
Austin, Texas

Christopher E. Wolff, MD
Associate Professor of Neurosurgery
Medical College of Wisconsin
Milwaukee, Wisconsin

Narayan Yoganandan, PhD
Professor and Chair, Biomed Engineering
Professor of Orthopedic Surgery and Neurosurgery
Department of Neurosurgery
Medical College of Wisconsin
Milwaukee, Wisconsin
Sir William Osler’s famous aphorism, “As is our pathology, so is our practice,” can be extrapolated to the practice of surgery: *As is our anatomy, so is our surgery*. Anatomy, by itself, is no longer a catalogue of structures, but is pregnant with an increasing appreciation and understanding of the complexity of function. Philosophy, rightly called *Scientia scientiarum*, or the science of all sciences, must govern all our surgical concepts, constructs, and procedures.

The term craniovertebral junction (CVJ), seemingly self-evident anatomically, appears to have been hurriedly conceived. The junctionality is suspect on many grounds. If the head is the expanded part of the neck, pray, where and what is the junction? Phylogeny and ontogeny obey the universal principle of a single, one-piece *homeobox*, wherein (during development, not joining, but selectively planned) multiple cavitations (coelomizations) occur to create a descriptive illusion of a joint. Moreover the appellation *junction* fails to convey the sense of symphony that orchestrates the machinery and movements at the so-called CVJ.

The game of football drives home the biological lesson that the so-called human head is a limb that can and does kick a ball as deftly as the foot—and this is not a metaphor. Vertebrate ontogenesis sees the neck and head as the fifth limb: the neck being the leg and the head being the expanded foot, as it were. The axioatlantic joint is akin to the supratalar ankle joint and the atlantooccipital joint the subtalar complex, the two acting corporately to produce a symphonic dance. An ace footballer pushes the ball literally with the skull (foot) encasing the brain. At such times the vital contents of the skull are taken care of by the neuraqua (CSF) to allow speed with security. The sheer symphonic nature of the so-called CVJ merits reverential elaboration.

To fashion a free pivotal joint and to harmonize it with a quasi ball-and-socket joint between the occiput and atlas, nature has exercised the pervasive principle: *In Nature functional necessity is the mother of structural innovation*. So the atlas centrum turns into a pivot and the atlas lateral masses turn broad and strong to match the equally strong occipital condyles. The wide periosteal cuffs, called capsule/ligaments, serve as beds for rich proprioception to feed the cord with a flood of sensory inputs that gets translated into precise yet powerful control and movements of the neck. Long, short, straight, and oblique muscles provide enormous mobility with amazing stability.

Above all, the craniospinal fraternity needs tremendous humility (bordering on reverence) for yet one more marvel of animal body: the CVJ. The other limbs can be handled or even injured with relative impunity, but not so for the CVJ—because of its contents and the complex web of nerves and blood vessels, it needs to be handled expertly, deliberately, with eyes wide open, using the knife, scissors, and the retractor in guided restraint so as not to precipitate iatrogenic disasters. Here, an error of omission is preferable to that of commission. *When in doubt, don’t*. That means that the slur of “abnormality” should not be lightly heaped on a patient to provide the surgeon the right to interfere and hurt. CVJ anomalies are far too common than the patients they produce. Ergo, this means that an obvious CVJ anomaly clear on a scan may need no interference, for it is compatible with lifelong symptom-free existence.

This being said, surgery in this region is highly result-oriented and gratifying. Most of the anomalies in the region are benign in nature and the patient usually presents with disabling neurological deficits. Because the disease in question is not integral to the person, the patient can live their full, normal life. This provides the caregiver a unique opportunity to set right the incongruity and, in the bargain, reinstate the patient and their family to a comfortable existence.

The present anthology is a global collaborative effort to understand CVJ and the problems associated therein. Each chapter is but a journey and nowhere a destination. We must reflect more, discuss more, create less controversies, and find more consensus to alleviate the load of disease on our fellow beings called the patients.

Atul Goel, MCh  
Francesco Cacciola, MD  
Manu L. Kothari, MS
Acknowledgments

My wife Naina and my daughter Aimee have been the pillars of my strength. Professor V. J. Laheri has been my long-term companion in the journey exploring the craniovertebral junction. Professors Edward Benzel and Volker Sonntag have offered me useful insights now and again. Professor Vijay Goel's clarity on the biomechanics of craniovertebral junction has been summed up in the chapter he authored. I am thankful to my teacher Professor Manu Kothari for helping me tread uncharted paths. I acknowledge with gratitude the constant support of our Dean Professor Sanjay Oak. All the contributors to this volume are friends to whom I am academically and personally obliged. It is my honor to be associated with the top scientific publisher of our times, Thieme Medical Publishers.

My father, recently deceased, fashioned my character and styled my craft. I miss him.

—Atul Goel, MCh

This book sees the light almost exactly 7 years after I first met Professor Atul Goel.

The providential value I would like to attribute to this number is not only the expression of a mystical quirk or randomness (for which I beg the scientific purist for forgiveness), but it is instead a characterization of the contribution this friendship has made to my own growth, not only as a neurosurgeon, and undoubtedly has made and will make to the growth of all those who have and will come in contact with Atul Goel’s “philosophy.” I like to define him as philosopher first and then as a brilliant surgeon—he is capable of making that paradigm shift that takes us out of the picture we have been looking at forever and making us see, in that same picture, something completely different or unexpected.

I had the chance to witness firsthand the development of a truly revolutionizing technique which is the C1–C2 distraction-fixation, to learn it directly from Professor Goel in Mumbai, and successfully apply it in my own practice. Just like it was with the technique of C1–C2 lateral mass fixation—which was pioneered by him in the 1980s and began its full diffusion in the spinal surgical community only two decades later—I suppose it will also take a while for the distraction technique to make its way into the spinal surgeon’s armamentarium. This delay will not be down to the fact that this technique is particularly difficult to grasp or perform by a thoughtful and well-trained surgeon, but it requires that “a-ha moment” after which the thinker and his or her practice will never be the same again, having progressed a step further on the endless journey of enriching and improving our practice.

I would like to express a very sincerely felt and huge “thank you” to Thieme Stuttgart and Clift Bergman and his great team. Working with them has been a real pleasure and they have believed in the project from the beginning and continued to do so with the same friendly enthusiasm as we moved on, even though we surely stretched all the imaginable limits during all stages of the production process.

A sincere “thank you” obviously also to all the great thinkers and surgeons, all authorities in their fields, who have contributed their chapters and chosen to join Professor Goel in this project. Amongst them I am nothing but a humble scholar and, having been able to see how all the pieces of those prestigious contributions joined together to form what I feel is a fantastically sound and harmonic end product, this has been a true privilege and honor.

May this book serve as a manual, guiding the reader in the diagnosis and management of craniovertebral junction pathology, but may it also be a source of inspiration and many “a-ha moments” as we are permitted to join in the thought processes and witness the experience of great minds who have dedicated a conspicuous part of their activity to this exciting entity which is the craniovertebral junction.

—Francesco Cacciola, MD
Comparative Anatomy

Chapter 1
Comparative Quantitative Analysis of Osseous Anatomy of the Craniovertebral Junction of the Tiger, Horse, Deer, Bird, and Human
The anatomy of craniovertebral junction bones of the tiger, horse, deer, and bird was analyzed and compared with human bones (Figs. 1.1, 1.2, 1.3, and 1.4). The evolutionary changes and structural alterations that have occurred due to the functional variations are clearly seen in the comparison. Understanding the anatomy of these animals clarifies the function of the various components of the bones of the craniovertebral junction. The evolutionary changes in the shapes and architectural design of the craniovertebral junction bones in each of these animals have been perfected to suit the job at hand. Despite the wide variations in the size of the bones, the basic patterns of structure, vascular and neural relationship, and joint alignments have remarkable similarities and a definite pattern of differences.

Apart from the sizes and weights, there are several structural variations in the bones of these animals that depend on their functional needs. The more remarkable difference in joint morphology is noticed in the occipitoatlantal joint. The occipitoatlantal articulation is remarkably large and deep, resembling a hinge joint in all the four animals studied. The odontoid process is C shaped in the deer, horse, and bird and is denslike in the tiger and humans. The transverse processes of the atlas are in the form of large wings in animals having heavy heads and thicker neck muscles like the horse, deer, and tiger. The arches of the atlas are large and flat, but the traverse of the vertebral artery resembles, to an extent, that of the human vertebral artery. The rotatory movements of the head at the craniovertebral junction are wider ranged in the horse and deer when compared with that of the tiger and humans. In the bird, the craniovertebral joints are designed to provide a circumferential 360° degree movement of the head. The bones of the craniovertebral junction of all the three mammals are adapted to the remarkable thickness and strength of the extensor muscles of the nape of the neck.

Text continues on page 8
Fig. 1.2a–k  Images of the craniovertebral junction bones of a horse.

a  Inferior view of the posterior aspect of the skull showing the large occipital condyles (1).
b  Superior (anterior) surface of the atlas, as seen from the superior and anterior perspective, showing the deep cup-shaped articular surface (1) for the articulation with occipital condyles.
c  Inferior surface showing the atlas articulated with the occipital bone. Note the acute flexed position of the head in relation to the atlas.
d  Posterior view showing the occipitoatlantal articulation.
e  Ventral view of the C1 vertebra showing the (1) ventral arch, (2) ventral tubercle, (3) transverse process, (4) transverse foramen, (5) atlantal fossa, and (6) superior articular facets.
Fig. 1.2f–i

f Dorsal view of the C1 vertebra showing the (1) dorsal arch, (2) inferior articular facets, (3) transverse foramen, (4) alar foramen, and (5) lateral vertebral foramen.

g Superior view of the axis vertebra of the horse showing the C-shaped configuration of the (1) odontoid process and the deep impressions for the (2) longitudinal ligament. The superior articular facets of the axis are seen in relation to the odontoid process.

h Anterior view of the C1-C2 articulation. The atlantoaxial joints are relatively flat when compared with the deep concavity of the superior facets of the atlas in relation to the occipital bone.

i Posterior view of the C1-C2 articulation.
Lateral view of the C1-C2 bones. The notch for the C2 spinal nerve is converted into the lateral vertebral foramen following ossification of the ligament.

Inferior view of the head of a horse.
Fig. 1.3c–f

c  Anterior view of the axis vertebra. The articular surface of the C-shaped (1) odontoid process can be seen in continuity with the superior articular facets forming a saddle-shaped joint.

d  Superior view of the axis vertebra of the deer showing the C-shaped (1) odontoid process and the confluent (2) superior articular surfaces. The saddle-shaped articular surface can be vividly seen.

e  Anterior view of the C1-C2 articulation.

f  Posterior view of the C1-C2 articulation.

Fig. 1.3 g–h
Fig. 1.3g, h

**g** Posterior view of the craniovertebral junction.
**h** Lateral view of the head of the deer. Note the location of the occipital condyles and the prominence of the occipital crest.

---

**Fig. 1.4a–f** Images of the bones of tiger.

**a** Ventral view of the C1 vertebra showing the (1) ventral arch, (2) transverse process, (3) superior articular facets, and (4) alar foramen. Note the wings of the transverse process and the depth of the superior facets.

**b** Dorsal view of the C1 vertebra showing the (1) dorsal arch and (2) transverse process.
Fig. 1.4c–f

c Lateral view of the axis vertebra showing the denslike odontoid process (1) and the characteristic spinous process (2). Note the shape of the odontoid process and the articular surface of the facet that resemble the human C2 vertebra.

d Anterior view of the C1-C2 articulation.

e Posterior view of the C1-C2 articulation.

f Lateral view of the skull of the tiger showing the large occipital crest.

Comparative Anatomy of the Bones of the Horse, Deer, Tiger, Bird, and Human

Tables 1.1 and 1.2 compare the C1 and C2 vertebrae, respectively, of humans to the horse, deer, tiger, and bird.

In quadrupeds, the cervical spine is a vertical part of the entire vertebral column, and the thoracic spine is more or less horizontally oriented. The head of the tiger, deer, and horse protrudes anteriorly in such a fashion that it is in the maximally flexed position at the occipitoatlantal joint articulation (Fig. 1.2). In contrast, the cervicothoracic junction is aligned in the maximally extended position. This asymmetric placement of the vertebrae in quadrupeds ensures an energy-saving balance of the head when the animal is in the resting position. While in the resting position, the movements permitted at the occipitoatlantal articulation are primarily of extension (flexion being a gravity-assisted passive movement); accordingly, the posterior cervical neck musculature is markedly strong, providing a site for muscular attachment. The occipital crest is most remarkably thick in the tiger, as seen in
In humans, the entire spine assumes a general vertical orientation and its curvatures are much less pronounced when compared with the quadrupeds studied. The vertical stance of the human being places the head directly over the neck in line of the weight bearing of the rest of the spine. The muscles of the nape of the neck and the occipital crest in humans are significantly smaller by comparison. The atlantoaxial bone and joint complex of the tiger have much more remarkable resemblance to humans than the bones of herbivorous animals like the horse and deer.

**Platybasia**

There is a platybasia in all three animals studied. The clivus and the anterior skull base are in the same horizontal plane. The maxilla and the upper jaw protrude anteriorly from the cranial base. The brain size is relatively small, and the olfactory nerves are well developed and long, reaching to a length of ~1 foot (~30 cm) in the horse. The cerebellum is proportionately large in animals as compared with the cerebral hemispheres. In humans, the angulation of the anterior skull base in relationship to the clivus is probably related to the relatively large size of the cerebral hemispheres.

### Occipitoatlantal Articulation

The superior articular surface of the atlas (referred to as the anterior articular facet in quadrupeds) and the occipital condyles are much larger, thicker, and stronger in all the three animals studied when compared with the corresponding human bones. The large occipital condyles of these animals lie deep in the cup-shaped anterior (superior) surface.

### Table 1.1 Analysis of C1 vertebrae

<table>
<thead>
<tr>
<th>Definition of Parameters</th>
<th>Humans (cm)</th>
<th>Horse (cm)</th>
<th>Deer (cm)</th>
<th>Tiger (cm)</th>
<th>Bird (cm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anteroposterior diameter of superior facet of C1</td>
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<td>4.0</td>
<td>1.0</td>
<td>3.0</td>
<td>0.3</td>
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<td>Transverse diameter of superior facet of C1</td>
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<td>3.0</td>
<td>1.5</td>
<td>2.0</td>
<td>0.5</td>
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<tr>
<td>Anteroposterior diameter of inferior facet of C1</td>
<td>1.3</td>
<td>4.0</td>
<td>1.2</td>
<td>2.6</td>
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</tr>
<tr>
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<td>4.0</td>
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<td>2.0</td>
<td>0.4*</td>
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</tr>
<tr>
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<td>5.0</td>
<td>1.9</td>
<td>3.0</td>
<td>0.8</td>
</tr>
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<td>3.0</td>
<td>1.4</td>
<td>2.7</td>
<td>–</td>
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<tr>
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<td>9.7</td>
<td>3.8</td>
<td>5.0</td>
<td>–</td>
</tr>
<tr>
<td>Width of transverse process of C1</td>
<td>1.8</td>
<td>4.7</td>
<td>1.5</td>
<td>3.5</td>
<td>–</td>
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*In birds, the anterior arch is replaced by a vertebral body that articulates superiorly with the occipital condyle and inferiorly with the body of C2.

### Table 1.2 Analysis of C2 vertebrae

<table>
<thead>
<tr>
<th>Definition of Parameters</th>
<th>Humans (cm)</th>
<th>Horse (cm)</th>
<th>Deer (cm)</th>
<th>Tiger (cm)</th>
<th>Bird (cm)</th>
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<td>0.9</td>
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</tr>
<tr>
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<td>1.7</td>
<td>0.5</td>
</tr>
<tr>
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<td>0.8</td>
<td>1.8</td>
<td>0.4</td>
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<td>4.9</td>
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<td>1.3</td>
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<td>1.8</td>
<td>1.2</td>
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<td>0.4</td>
</tr>
<tr>
<td>Transverse diameter of spinal canal at C2</td>
<td>2.1</td>
<td>2.5</td>
<td>1.3</td>
<td>2.2</td>
<td>0.6</td>
</tr>
</tbody>
</table>
articular facets of the atlas (Figs. 1.2, 1.3, and 1.4) and form a joint that resembles a ginglymus or a hinge joint, providing an opportunity for extra stability and enhanced mobility when compared with the human occipitotantal joint. The superior facets of the atlas are much deeper in all the three animals when compared with the human superior facet, which is almost flat. The types of movement at the occipitotantal articulation are chiefly of extension and flexion, with a small amount of lateral oblique movements. The total range of motion at the occipitotantal articulation varies between species. It is 90 to 105° in quadruped mammals and only 11 to 13° in humans. The cervical part of the vertebral column is most mobile in horses; the mouth may be brought around to reach the flank on full lateral flexion of the neck and ventrally to reach the pasture on ventral flexion.

### Atlas

The atlas bone in all three animals studied has ring-shaped anterior and posterior arches and wide lateral platelike projections or wings of the transverse processes (Figs. 1.2, 1.3, and 1.4). The transverse process in the human atlas is reduced to only the vertebral artery canal. The ventral (anterior) and dorsal (posterior) arches of the atlas are much thicker in the tiger, horse, and deer when compared with human arches. The dorsal arch presents a median tubercle. It is perforated on either side near its cranial margin by the lateral vertebral foramen (Fig. 1.2). In these animals, the term lateral vertebral foramen is used instead of intervertebral foramen in the case of the atlas and axis, because this foramen does not lie between the two vertebrae, as the prefix inter indicates, but rather on the dorsal arch of the atlas. The ventral arch is thicker, narrower, and less curved than the dorsal arch. On the ventral surface is the ventral tubercle, which is more prominent in the horse and deer as compared with humans and the tiger.

Although the transverse processes are large in the horse and deer when compared with humans, they are proportionately smaller when compared with those of a tiger. The ventral surfaces of the transverse process of the atlas in the horse and deer have a greater depth than that of a tiger, in which they are shallower but wider. Between the ventral aspect of the transverse process and the lateral mass is a depression called the atlantal fossa. In the horse, each wing is perforated by two foramina; the cranial one is the alar foramen, which connects with the lateral vertebral foramen by a short groove, and the caudal one is the transverse foramen (Fig. 1.2). In the tiger, there is a lateral vertebral foramen for the first cervical nerve close to the cranial border of the dorsal arch. The alar foramen is replaced by a notch in the cranial border of the wing, which transmits the ventral branch of the first cervical nerve. The base of the wing is perforated by the transverse foramen. Although the human transverse process is horizontally oriented (transverse), it is vertically aligned in the animals studied.

The articular surface of the superior facet of the atlas accounts for ~75% of the entire ring of the atlas when compared with ~< 40% in the atlas of humans. The joint surfaces are separated by a wide notch dorsally and a narrow one ventrally. The atlantodental joint is remarkably prominent in the horse and deer, providing articulation to the large and C-shaped odontoid process in these herbivorous animals (Figs. 1.2 and 1.3). The inferior articular surfaces (referred to as the posterior articular surface in quadrupeds) of the lateral mass of the atlas are confluent anteriorly with the joint surface on the posterior arch of the atlas to form a saddle-shaped articular surface.

### Axis

In the horse and deer, the axis is the longest of all vertebrae. It measures 16 cm in the horse and 4.9 cm in the deer in its vertical length.

The odontoid process of the tiger resembles the odontoid process of a human. The odontoid process is denslike in human and tiger bones, whereas it is a C-shaped, relatively thin and flat ring that has a wide area of joint formation with the posterior surface of the anterior arch of the atlas (Figs. 1.2, 1.3, and 1.4). This wide area of the atlantodental joint is seen uniformly in herbivorous animals as opposed to the denslike odontoid process in carnivorous animals. The human odontoid process resembles more closely that of the carnivorous animals. The anterior surface of the odontoid process forms a well-defined joint with the posterior surface of the anterior arch of the atlas. The joint is much larger in the horse and deer when compared with the joints of the tiger and humans. The rotatory movements of the neck at the craniovertebral junction are superior in the horse and deer when compared with the tiger and humans. The limitations of the rotatory movements at the craniovertebral junction and the placement of eyeballs in a more anterior perspective of the head in the tiger and humans when compared with the horse and deer are adaptations that suit their lifestyle and preying, hunting, and survival needs. The dorsal surface of the odontoid process has two deep impressions on either side of the midline in the horse and deer for the attachment of the thick, fan-shaped longitudinal ligament (Fig. 1.2). This ligament extends from the rough concave dorsal surface of the dens, widens cranially, and is attached to the transverse rough area on the inner surface of the ventral arch of the atlas. These impressions are not prominent in the odontoid process of the tiger and humans. The atlantoaxial joints are relatively similar in their inclination and depth in humans and in all the three animals studied.

In the horse, the lamina or the arch of the axis has a notch on each side of its cranial border, which is converted into a lateral vertebral foramen (intervertebral foramen) by a ligament that ossifies later (Fig. 1.2). A groove extends ventrally and caudally from this foramen and houses the ventral branch of the second cervical spinal nerve. In the deer and tiger, the cranial border has a deep
notch that is not converted into a foramen. In humans, there is no such notch or groove. The inferior articular processes of the three animals are vertically oriented when compared with their more horizontal orientation in humans. The transverse process in the deer and horse is small, single, and projects caudally. It has an obliquely oriented foramen for the vertebral artery. The transverse process and the vertebral artery foramen in the axis of the tiger are similar to that in humans.

The spinous processes of the axis of all the three animals studied are large, strong, and bifid (Figs. 1.1, 1.2, 1.3, and 1.4). The axis in the tiger is characterized by its length and enormous spinous process, which overhangs both the dorsal arch of the atlas and the laminae of the C3 vertebra. The cranial extent of the spinous process matches that of the dens (Fig. 1.4). The human C2 spinous process is short, stubby, and bifid and is smaller than that of the other three animals. The lateral surfaces are concave and rough for muscular attachments.

Vertebral Artery

The vertebral artery has a peculiar relationship to the transverse process of the horse. After exiting from the transverse foramen of the atlas, it crosses the capsule of the atlantoaxial joint and enters the transverse foramen of the atlas. After coursing through the atlantal fossa, it anastomoses with the occipital artery. It then runs dorsally through the alar foramen and enters the vertebral canal through the lateral vertebral foramen (intervertebral foramen).

In the tiger, the vertebral artery, after exiting from the transverse process of the axis, courses over the dorsal arch of the atlas and enters the transverse foramen, which is present in the base of the wing of the transverse process. It then runs an intrasosseous course in the wing for ~2.5 cm. It exits on the ventral aspect, loops posteriorly, and enters the lateral vertebral foramen to pursue its intracranial course.

C1 Spinal Nerve

The first cervical nerve emerges through the lateral vertebral foramen of the atlas and supplies several large muscles of the nape of the neck in all the three animals studied. This is unlike the situation in humans, where the C1 nerve root is rudimentary in nature and function. In the horse and deer, the dorsal branch of the nerve passes dorsolaterally and supplies the dorsal neck musculature. In the horse, the ventral branch descends through the alar foramen of the atlas and passes anteriorly into the neck. In the tiger, the C1 nerve exits from the lateral vertebral foramen, and its ventral branch exits through the notch in the cranial border of the atlas along the course of the vertebral artery.

C2 Spinal Nerve

The second cervical nerve is larger than the first in all three animals, as in humans. It emerges from the spinal canal through the lateral vertebral foramen on the cranial border of the lamina of the axis. This nerve also supplies several muscles and the skin over the neck. Its function is also significantly greater than that in humans.

Comparative Anatomy of the Bones of the Bird

The craniovertebral junction of a bird is architecturally designed to permit circumferential (360°) movement of the head. The most unique aspect of the anatomical variation is the presence of the body of the atlas vertebra and of a single occipital condyle (Fig. 1.5). The body of the atlas articulates superiorly with the occipital condyle, which is single, and is the only joint at the craniovertebral junction. The presence of a single joint instead

Fig. 1.5a–e  Images of bones of a kite.

a  Lateral view of the head of a kite. Note the large orbital cavity.

b  Inferior view showing platybasia. A single occipital condyle can be seen.
of two probably allows a wider range of neck movement. The laxity of the ligaments at the junction and the extra length of the spinal cord in the region allow 360° neck rotation. The body of the atlas articulates inferiorly with the body of the axis. The odontoid process, which resembles to some extent the odontoid process of the horse, fits snuggly into the posterior aspect of the body of the atlas bone and forms an atlantoodontoid articulation. The atlantoaxial facet joints are lateral and are not in direct communication with the atlanto-occipital and atlantoaxial articulations, as is seen in the saddle-shaped joint of the horse and deer.

Essentially, it appears that, in a bird, the occipitoatlan-
tal articulation favors movement over stability. The light weight of the bird’s head also permits quick and circumferential movements and does not pose excessive stress on the craniovertebral joints. The atlantoaxial joints, in contrast, are remarkably stable due to the presence of an extra joint between the body of the axis and the atlas. The architectural design of the atlantoaxial bones resembles bones of herbivorous animals, such as the horse and deer.

Fig. 1.5c–e

c. Inferior view of the posterior cranial fossa showing the single occipital condyle in the anterior aspect of the foramen magnum.
d. Image of the axis vertebra. Note the shapes of the odontoid process and the presence of the body of the axis.
e. Image of the articulated atlantoaxial bones. Note the presence of the body of the axis and its relationship with the odontoid process.

Conclusion

The craniocervical junction bones form a pillar of stability and mobility. The basic architecture and design have remarkable similarities in all of the animals studied. The variations in morphometry have a relationship with the quadruped stance, acute flexion position of the neck in the neutral position, and hunting and survival needs of the individual animal.

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Basic Concepts

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The skull and the vertebral column are two separate entities. A distinct description of these two entities is therefore more than justified in terms of anatomical classification and also with regard to their distinct functions.

The term craniovertebral junction (CVJ) refers to a region that includes part of the skull or cranium and part of the vertebral column and represents the area where these separate entities are joined together. It encompasses the part of the occipital bone that surrounds the foramen magnum of the skull and the atlas vertebra of the vertebral column.

Looking at the CVJ as a junction of two distinct entities, however, is misleading when it comes to both understanding the congenital anomalies that can occur in this area and comprehending the peculiar anatomical–radiological findings that occur during normal development. Anomalies such as atlas assimilation and occipital vertebrae and radiolucent lines, which are normal in a child but could represent a fracture in an adult, have their origin in the peculiar embryological and developmental mechanisms leading to the CVJ.

Up to a certain stage of embryological development, the CVJ is not at all the junction of two separate entities but rather a unique entity, with the “embryological” CVJ located more rostrally, between the developing occipital bone and the other cranial bones it articulates with, particularly the sphenoid bone. This concept of an embryological CVJ is derived from the fact that the occipital bone originates from the same process that leads to the formation of the vertebral column, whereas the other cranial bones are generated through different mechanisms.1,2

Most skull and facial bones develop by intramembranous ossification. Intramembranous ossification is a process that bypasses the intermediate cartilaginous stage characteristic of the occipital bone and the verteabrae. The formation of the occipital bone and the vertebrae is determined by the notochord and the somites.3

Understanding the role of the notochord, the somites, and their segmentation, as well as the three developmental stages of membranous, cartilaginous, and osseous formation, is crucial in understanding the CVJ.

### Formation of the Notochord, the Somites, and the Neural Tube

A detailed description of the peculiar embryological events that give rise to the skull and vertebral column would go beyond the scope of this chapter. Nevertheless, the most salient steps are synthesized in a simplified manner to get an approximate picture of the entire process.

During the first 4 days after fertilization, the human embryo forms a mass of ~32 cells, the blastocyst. The blastocyst is a cystic cavity with a wall formed by a layer of cells and an inner mural nodule consisting of a mass of cells, the embryonic cell proper. Attachment to the uterine wall occurs around the fifth day, and implantation in the uterus happens within the second week of gestation. The cyst grows and evolves to a formation that contains two cavities—the amnionic sac and the yolk sac—separated by a disk of two layers of cells. Once this two-layered cyst is implanted in the uterus, a cavity called the chorion forms around it. The cyst stays connected to the uterus by the stalk that will become the umbilical cord (Fig. 2.1).

Gastrulation, which occurs during the third week of gestation, is the process that transforms the two-layered embryonic disk into a three-layered structure, or germ layer, made of ectoderm, mesoderm, and endoderm.4

During the third week of gestation, a definite notochord forms in the mesoderm between the ectoderm and endoderm. It is represented by a longitudinal tube of cells...
with a virtual central canal, which is located in the center of the embryonic disk along its major axis. (The term disk is used for ease of understanding. In this discussion, it indicates the relatively flat, round structure of the initial germ layer, even though a major axis, and thus a rather oval shape and loss of the flatness develops with cranio-caudal differentiation and further development.)

On both sides of the notochord, the mesoderm differentiates into three main areas: the paraxial, intermediate, and lateral mesoderms. The paraxial mesoderm divides on either side of the notochord into 42 to 44 pairs of somites (specialized cell accumulations). Development of the somites occurs in a craniocaudal fashion, and the number of somites can be used to estimate embryonic age. The first somites, and thus the cranialmost, appear in the third week of gestation, and the most caudal and last ones will have formed by the end of the fifth week. The somites are the precursors of the occipital bone, the vertebrae, other bony structures of the thorax, and the associated musculature of these structures.

Each somite differentiates into two parts: a sclerotome, which is located medially, next to the notochord, and a dermomyotome, located laterally. The cells of the sclerotome are responsible for the formation of the bone, and the dermomyotomes form muscle cells and the overlying dermis of the skin.

In the meantime, between the second and third weeks of gestation, and parallel to the formation of the notochord and the somites, the ectoderm above the notochord thickens and differentiates into neural ectoderm. By the end of the third week of gestation, the neural tube will have formed from this neural ectoderm and will overlay the notochord, suspended in the mesodermal layer. This process is called neurulation. The ectoderm from which the neural tube derived will have re-formed a continuous layer above the neural tube.

The third week of gestation is a crucial period for the formation of the CVJ and vertebral column because three important processes take place contemporaneously: definite formation of the notochord, formation and closure of the neural tube lying right above it, and formation and progressive appearance of the paired somites on either side of the notochord and neural tube (Fig. 2.2).

Yet another event takes place in the third week of gestation that affects the somites as they progressively appear and differentiate into sclerotomes and dermomyotomes: segmentation, which characterizes the membranous stage of development and demarcates the division between the skull and vertebral column.5

### Segmentation

As already mentioned, the somites differentiate into a medial part, the sclerotomes, and a lateral part, the dermomyotomes. Subsequently, the sclerotomes migrate medially and surround the notochord and the neural tube, forming a continuous mesodermal sheath and giving rise to a membranous basioccipital bone cranially and a membranous vertebral column below. The dermomyotomes will develop into the cervical musculature and overlying skin in the region.

Sclerotomes 1 to 5 form the basioccipital elements of the skull. Sclerotomes 1 to 4 give rise to the basioccipital (midline segment that lies just ahead of the foramen magnum), exoccipital (lateral margins of the foramen magnum), and the paired somites on either side of the notochord and neural tube (Fig. 2.2).
magnum and superior part of the occipital condyles and jugular tubercles), and supraoccipital (posterior boundary of the foramen magnum) centers. Sclerotome 5 is highly specialized because it is the first one to form a motion segment and starts the process of segmentation.

Segmentation of a sclerotome is a process in which proliferation leads to a more loosely packed cellular area in the cranial half of the sclerotome and a more densely packed area in its caudal half. As segmentation progresses, the loose cranial part and the dense caudal part will eventually separate, and the dense caudal part of one sclerotome will fuse with the loose cranial part of the one below. The splitting, and thus segmentation, of a sclerotome occurs in the area in which the future spinal nerve will transit.

In subaxial spinal development, the intervertebral disk develops, around and including the notochord, in the gap created by the segmentation of a sclerotome into cranial and caudal parts. The process can therefore be described in the following way.

Division of a sclerotome into cranial and caudal parts at the passage point of the developing spinal nerve leads to formation of a disk in the gap created by fusion of the caudal half of the sclerotome with the cranial half of the following one, forming a vertebra. This cranial half of the following sclerotome was created through division from its inferior half. A disk will form in the gap, perpetuating this process caudally (Fig. 2.3).

It is important to focus on this process because it takes place in the same regular fashion at the CVJ, even though it leads to a very particular expression.6

As stated previously, sclerotome 5 plays a key role, because it starts segmentation.

Sclerotomes 1 to 4 do not undergo segmentation and, by simply fusing one with the other, give rise to the occipital bone. The fifth sclerotome, however, will divide. Its loose cranial half will fuse with sclerotome 4 and give rise to the most caudal part of the occipital condyles and foramen magnum. Its dense caudal half will give rise to the cranial part of the atlas and the tip of the axis odontoid process. It is exactly at this point and in this particular moment of embryological development that the CVJ is formed, by segmentation of sclerotome 5.

According to the previously described regular subaxial development of vertebrae and disks, it should be expected that a disk forms in the gap created by the segmentation of the fifth sclerotome into cranial and caudal parts. Obviously, there is no disk between the occiput and the atlas, but given the fact that the apical ligament has been found to contain notochordal tissue, it can be regarded as a rudimentary intervertebral disk, along with the rest of the ligamentous apparatus.7

In sclerotome 6, segmentation gives rise to a cranial half that forms the caudal part of the atlas and the odontoid process of the axis; the caudal half will form the cranial half of the body of the axis, and the cranial half of sclerotome 7 after segmentation will form the caudal half of the axis. The CVJ is thereby completed (Fig. 2.4).

It can thus be stated that segmentation gives rise in sclerotome 5 to the articulation between the occipital condyles and the atlas, in sclerotome 6 to the articulation between the atlas and the axis, and in sclerotome 7 to the

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**Fig. 2.3** Illustration of the segmentation process of the sclerotomes once they have migrated around the neural tube and the notochord. (From Larsen WJ. Human Embryology. 2nd ed. Edinburgh, Scotland: Churchill Livingstone; 1998. Reprinted with permission.)

**Fig. 2.4** Illustration of the segmentation process in an avian embryo, showing which structures of the occipital bone and cervical spine arise from sclerotomes 1 to 7. (From Huang R, Qixia Z, Patel K, Wilting J, Christ B. Contribution of single somites to the skeleton and muscles of the occipital and cervical regions in avian embryos. Anat Embryol 2000;202:375–383. Reprinted with permission.)
articulation between the axis and C3. Again, at the level of segmentation of sclerotome 6, an intervertebral disk could be expected, and indeed at birth a cartilaginous band between the odontoid process (deriving from the cranial half of sclerotome 6) and the cranial part of the axis body (deriving from the caudal half of sclerotome 6) represents a vestigial disk, referred to as a neural central (or subdental) synchondrosis. This line lies below the anatomical base of the odontoid and fuses through ossification between 3 and 6 years of age in most children, as discussed in the following section.

Three Stages of Vertebral Development: Membranous, Cartilaginous, and Osseous

The processes described so far—notochord formation, somite formation, and sclerotome segmentation—as well as migration of the sclerotomic cells around the notochord and neural tube, occur in the membranous stage, which is mainly in the third week of gestation. This is the first stage of vertebral column development and also of occipital bone development, which is also somite dependent: the membranous vertebral column stage.

Around the beginning of the fourth week, chondrification centers appear as two centers in each half of the vertebral body, on each side of the notochord, and a center for each half of the forming neural arch, on each side of the neural tube. As they are forming and joining, the notochordal cells from the vertebral bodies are squeezed into the disk spaces. These notochordal cells expand and become the nucleus pulposus of the disk at subaxial levels, whereas there is a differentiation of the notochord in the ligamentous complex between the occiput, the atlas, and the axis. At the level of the occipital bone, where no segmentation has occurred, the notochordal cells regress and do not give rise to any structure. During the later stage of cartilaginous formation of the spine, the four chondrification centers fuse into the cartilaginous vertebral body and arch.

Around the seventh to eighth week of gestational age, the stage of ossification begins in the midthoracic region, extending progressively both cranially and caudally.8,9

As far as the formation of the occipital bone is concerned, around the 10th to 12th week of gestation, heavy chondrification is observed in the basioccipital, exoccipital, and supraoccipital bones. Subsequently, one or two ossification centers appear in each of the regions. Fusion of the supraoccipital bone to the exoccipital bone occurs around 2 years of age, and not until 4 years of age will the basioccipital bone have fused with the exoccipital bone.

Ossification is an ongoing process until early childhood for the vertebrae, extending until early adulthood for subaxial vertebrae. The time of appearance of the ossification centers of the atlas and the axis is slightly different when compared with the subaxial vertebrae. Only the former will be discussed here.

Atlas

The atlas is usually ossified from three centers (Fig. 2.5). One of these appears in each lateral mass around the seventh week of fetal life and extends backward; at birth, these portions of bone are separated from one another by a narrow interval filled with cartilage. Between the third and fourth years, they unite either directly or through the medium of a separate center developed in the cartilage. At birth, the anterior arch consists of cartilage; a separate center appears in this around the end of the first year and joins the lateral masses between the sixth and eighth years—the lines of union extend across the anterior portions of the superior articular facets. Occasionally, there is no separate center, and the anterior arch is formed by the forward extension and ultimate junction of the two lateral masses. Sometimes this arch is ossified from two centers, one on either side of the midline.

Axis

The axis is ossified primarily from six centers (Fig. 2.6). The body and vertebral arch are ossified in the same
manner as the corresponding parts in the other vertebrae (i.e., one center for the body and two for the vertebral arch). The centers for the arch appear around the seventh or eighth week of fetal life and for the body about the fourth or fifth month. In the dens, which originally is a cartilaginous mass above the ossifying body, two centers make their appearance around the sixth month of fetal life; they are placed laterally and join before birth to form a conical bilobed mass deeply cleft above. The interval between the cleft and the summit of the dens at this stage continues to be formed by a wedge-shaped piece of cartilage. The base of the ossifying dens is separated from the body by a cartilaginous disk, which gradually becomes ossified at its circumference but remains cartilaginous in its center until advanced age (Fig. 2.7). This is called the subdental synchondrosis, as discussed previously. It appears as a lucent line slightly below the plane of the superior border of the body of the axis and usually ossifies by 3 to 6 years of age, even though it may persist as a lucent line into adulthood, as seen on radiographs (Fig. 2.8). The sixth center of ossification of the axis is responsible for the tip of the dens. It may appear as early as the second year of life but usually does not develop until 5 or 6 years of age. It fuses with the body of the dens by 10 to 12 years of age although, rarely, it may not fuse and instead persist as a small ossicle (ossiculum terminale).

**Clinical and Practical Implications**

Knowledge of the various lines and times of fusion between the bony structures that compose the CVJ carries a utility that is of immediate intuition (Fig. 2.7). Especially when dealing with the pediatric population, it proves very helpful to distinguish between normal and pathological. The widespread times of fusion of the single elements not only lead to peculiar lines of fusion within a specific vertebra, but also imply variations during the developmental process of the relationships between the vertebrae (Fig. 2.8). During postnatal maturation of the spine, certain measurements and pseudodisplacements in children differ in comparison with those in adults. As far as the CVJ is concerned, the atlantoaxial relationship between the posterior margin of the arch of C1 and the anterior margin of the odontoid process (C1–dens) usually measures

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**Fig. 2.6** The axis and its structures derive from six ossification centers.

**Fig. 2.7** Synoptic graph illustrating the segmentation and maturation processes leading from sclerotomes to fully developed vertebral structures.
between 2 and 4 mm but may be as great as 5 mm in young infants and children and can increase by 1 to 2 mm on lateral flexion films. Pseudosubluxations of up to 5 mm of C2 onto C3 or at lower levels may be considered as normal until 10 years of age.10 Understanding the process of segmentation and its very regular proceeding, even at the level of the CVJ with its atypical vertebrae, helps to create a solid framework on the basis of which the normal development and derangement can be learned and memorized.

The understanding of the process of segmentation, however, is not particularly helpful when it comes to ossification of the single elements, which occurs, as we have seen, with great time differences among certain elements. This is because osteoblasts have been shown not to possess a segmental identity.6 Although the formation of cartilage is segmental, and thus sclerotome dependent and driven, once the cartilaginous structure has been formed, the osteoblasts and the ossification centers still seem to appear according to segments. Their subsequent pattern of evolution, however, is driven by blood vessels or information from surrounding tissue. This phenomenon, which is still not completely understood, seems to be responsible for the fact that it is difficult to track down most of the congenital malformations of the CVJ in terms of the pathological process implied and most of all when trying to identify at what stage a derangement occurs that later on leads to the malformation.

When looking at the congenital anomalies that occur at the CVJ (Table 2.1) from an etiopathogenetic point of view, errors seem to occur in terms of both hypersegmentation and hyposegmentation. The former, for example, probably leads to clivus segmentations or occipital vertebrae in the form of proper bony remnants around the foramen magnum, such as a third occipital condyle or epitransverse processes (Fig. 2.9). Hyposegmentation, on the other hand, seems to be responsible for atlas assimilation or the rare cases of atlantoaxial assimilation.

Although the pathogenetic process in these cases might have its origin in the overexpression or underexpression of the genes responsible for segmentation, it is more difficult to find an explanation for malformations characterized by hypoplasia or aplasia of single elements. This is the case with basilar invagination, condylar hypoplasia, and odontoid process aplasia, for example.

The processes that regulate developmental steps in the complex region of the CVJ are surely multifactorial, with multiple links among them. This can be appreciated not only in the fact that an association of malformations can be observed in one patient (e.g., occipitalization of the atlas, odontoid malformations, or atlas arch abnormalities with basilar invagination), but also in the involvement of other organ system abnormalities, such as genitourinary tract or cardiovascular malformations (e.g., Klippel-Feil syndrome).11,12

![Fig. 2.8 Anteroposterior radiograph of a 30-week-old fetus showing the separate ossification centers of the body and dens of the axis. The ossification center of the dens tip has not yet appeared. (From Herkowitz HN, Garfin SR, Balderston RA, et al., eds. Rothman–Simeone, The Spine. 4th ed. Philadelphia, PA: WB Saunders; 1999. Reprinted with permission.)](image)

### Table 2.1 Congenital disorders of the craniovertebral junction resulting from either hyposegmentation or hypersegmentation and the various levels involved

<table>
<thead>
<tr>
<th>Occiput</th>
<th>Atlas</th>
<th>Axis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Basilar invagination</td>
<td>Hypoplasia or aplasia of the</td>
<td>Hypoplasia or aplasia of the</td>
</tr>
<tr>
<td>Condylar hypoplasia</td>
<td>arches or lateral masses</td>
<td>dens</td>
</tr>
<tr>
<td>Atlas assimilation</td>
<td>Atlas assimilation</td>
<td>Osisculum terminale</td>
</tr>
<tr>
<td>Occipital vertebrae (clivus</td>
<td>Atlantoaxial fusion</td>
<td>Atlantoaxial fusion</td>
</tr>
<tr>
<td>segmentation, third occipital</td>
<td></td>
<td></td>
</tr>
<tr>
<td>condyle, epitransverse processes</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Fig. 2.9 Lateral radiograph showing an occipital vertebra formed by the articulation of an epitransverse process of the atlas with a paracondylar mass from the occiput.

Ultimately, the importance of all these malformations lies in their dangerous potential to cause neurological compression. The available refined imaging technologies enable us to identify and pinpoint both bony malformation and neurological compromise at the CVJ with high accuracy. A thorough understanding of the developmental processes and derangements, however, is essential to selecting the most appropriate and at the same time physiological approach to CVJ pathology.

References

3 The Vertebral Artery in Relation to the C1–C2 Complex
Francesco Cacciola, Umesh Phalke, and Atul Goel

A three-dimensional understanding of the anatomy is crucial for any kind of surgery in the craniovertebral region. Within the C0–C1–C2 complex, which represents the craniovertebral junction (CVJ), the C1–C2 relationship merits special attention because of the unusual shape of these atypical vertebrae. The peculiar course of the vertebral artery, as well as the number of surgical interventions that involve this structure, makes it especially significant.1–5

The popularity of the transoral surgical route, lateral mass screw fixations, transarticular screw fixations, occipitocervical fusions, and lateral approaches to anterior foramen magnum lesions has made learning of the anatomy of the bony–vascular relationship in the CVJ more relevant. Inadvertent injury to the vertebral artery during surgery can lead to catastrophic intraoperative bleeding, and compromise to the blood flow can lead to unpredictable neurological deficits, depending on the adequacy of blood flow from the contralateral side.6,7

This chapter focuses on portraying the relationship of the vertebral artery to the C1–C2 complex and its bony landmarks.

Osseous Anatomy

Foramen Magnum and Occipital Condyles (C0)

The foramen magnum is round to oval shaped. The anteroposterior length measures on average ~35 mm, whereas the transverse length is ~30 mm.1 In its anterior half, the foramen magnum is flanked on both sides by the paired occipital condyles, which can be considered an integral part of it. Ovoid in structure, the occipital condyles have a major axis that is oriented anteromedially. Their articular surfaces face downward and slightly lateral and are convex anteroposteriorly. The condyles articulate with the superior facets of the atlas caudally. The average length of the condyles in their major axis is ~20 mm. Posterior to the condyles, a more pronounced depression represents the condylar fossae. In ~60% of our specimens, they are the site of a canal that transmits the posterior condylar emissary vein, connecting the vertebral venous plexus with the sigmoid sinus just proximal to the jugular bulb (Fig. 3.1).

Fig. 3.1 Craniovertebral junction viewed from behind in a specimen displaying both condylar canals and emissary veins (arrowheads). Note the exposed C1–C2 joint after section of the C2 ganglion (arrow).

The hypoglossal canals course within the occipital condyles, transmitting the respective nerves. They are directed anteriorly and laterally at an angle of ~45° with respect to the sagittal plane, the intracranial end being located ~10 mm from the posterior border of the condyle.8

Atlas (C1)

The superior facet of the atlas is oval with a groove on either side. Less frequently, it is kidney-shaped, with the groove present on only one side of the facet. This facet is longer in its anteroposterior dimension (mean 20 mm) than in its transverse dimension (mean 11 mm). Our study showed that none of the superior facets of the atlas were exactly symmetrical to those on the contralateral side.9 Both superior and inferior facets of the atlas face medially toward the spinal canal. The inferior facet of the atlas is almost circular in most of the vertebrae without any significant difference in the mean anteroposterior and transverse (mean 15 mm) dimensions. The thickness of the inferior facet under the posterior arch of the atlas is on average 3.5 mm (a in Fig. 3.2). The thickness of the posterior arch of the atlas separating the vertebral artery groove from the
The vertebral artery foramen is in the transverse process lateral to the lateral mass of the C1 vertebra. The groove for the vertebral artery on the superior surface of the posterior arch of the atlas is occasionally converted into a complete bony foramen. The distance from the midline to the medialmost edge of the vertebral artery groove on the outer cortex of the posterior arch is on average 18 mm (Figs. 3.2 and 3.3).

**Axis (C2)**

The superior facet of the C2 vertebra differs from the facets of all other vertebrae in two important characteristics, making this region prone to vertebral artery injury during screw fixation. First, the superior facet of C2 is closer to the body when compared with facets of the other vertebrae, which are located in proximity to the lamina (Fig. 3.4). Second, the vertebral artery foramen is present partially or completely in the inferior aspect of the superior facet of C2, whereas in the subaxial cervical vertebrae the vertebral artery foramen is located entirely within the transverse process and delimits the anterior border of the vertebral pedicle (Figs. 3.5 and 3.6). The size of the pedicle of the C2 vertebra varies according to its definition. We have adopted the definition as outlined in Fig. 3.4, making it thus very small in size and essentially being the zone of contact between the vertebral body and the superior articular surface.9,10 The course of the vertebral artery in relation to the inferior surface of the superior articular facet of C2 makes it susceptible to injury during transarticular and interarticular screw implantation techniques (Figs. 3.4 and 3.6).

The axis vertebral body has a thick, conical superior projection—the dens. The dens is flanked by two large superior facets, extending laterally onto the adjoining pars interarticularis and articulating with the inferior atlantal facets. Unlike superior facets of all other vertebrae, these do not form a vertical pillar with the inferior facets, because they are considerably anterior to the inferior facets (Fig. 3.7).
The shape of the superior articular surface of the axis varies from oval to circular. It is convex in the sagittal plane and is directed laterally to articulate with the inferior facet of C1. The average depth of the vertebral artery groove on the inferior surface of the superior facet is 4 mm. In the 20 specimens studied, the vertebral artery groove extended in the superior facet up to its medial third in 5 cases, middle third in 9 cases, and lateral third in 6 cases (Fig. 3.8). In only one specimen did the vertebral artery groove extend minimally into the body of the axis vertebra (Fig. 3.9).

In no case did the vertebral artery groove extend into the pars interarticularis. The transverse thickness or the width of the pars interarticularis is 8 mm on average. The total length of the pars interarticularis is ~15 mm.

The angle of its projection toward the superior facet is measured as the angle of the pars interarticularis from the vertical plane and ranges from 38 to 50° (average 45°). The intertransverse process height between C1 and C2 is ~18 mm.\textsuperscript{1}

### Anatomy of the Vertebral Artery

The anatomy of the vertebral artery in the region of the CVJ is significantly different from the relatively straightforward course in the transverse foramina of C6 to C3 vertebrae.

The vertebral artery has a serpentine course in relation to the craniovertebral region. The artery has multiple loops and an intimate relationship with the atlas and axis.
bones. We observed a wide variability of the course of the artery in our specimens. Of the 20 vertebral arteries studied, no two arteries matched exactly in their course, length, and size. Also, the shape, size, and location of the vertebral artery groove on the inferior aspect of the superior articular facet of C2 and over the posterior arch of the atlas had wide variations. Venous plexuses covered the entire course of the vertebral artery, making identification possible during the surgery.

After a relatively linear ascent of the vertebral artery in the foramen transversarium of C6 to C3, the artery makes a loop medially toward an anteriorly placed superior articular facet of the C2 vertebra, making a deep groove on its inferior surface. The degree of medial extension of the loop varied. The nearest distance of the artery from the midline of the vertebral body of C2 as observed during a transoral surgical procedure was 11.7 mm on average. The anterior surface of the body of C2 was continuous with the anterior surface of the superior facets on both sides, with no definite identification landmark. This suggests that during transoral surgery and drilling of the C2 body, exact identification of the midline is crucial, and the lateral limit of bone removal should take into account the location of the artery. The vertebral artery loops away from the midline underneath the superior articular facet of C2. This makes drilling safe, in regard to the vertebral artery, above the level of the C2 vertebral body and over the odontoid process.

During its entire course, the vertebral artery is covered with a large plexus of veins. The venous plexuses are the largest in the region lateral to the C1–C2 joint. In our study, the diameter of the vertebral artery ranged from 2.3 to 7.4 mm (average 4.2 mm).

To assist in the description, the vertebral artery was labeled as V1 in its course from the C3 transverse foramina to the C2 transverse foramina, as V2 from the C2 transverse foramina to the C1 transverse foramina, and as V3 in its course from the transverse foramen of C1 to the point of the artery’s dural entry (Figs. 3.10 and 3.11).

**V1 Segment**

After its exit from the transverse foramen of the C3 vertebra, the V1 segment of the artery courses posterosuperiorly and forms a loop within the vertebral artery groove on the inferior surface of the superior articular facet, then exits from the transverse foramina of the C2 vertebra. The total length of the segment ranges from 17.2 to 46.1 mm (average 23.4 mm). The distal part of the artery is intrasosseous and cannot be seen until its bony unroofing. The length of the artery prior to its entry into the osseous compartment is 6 to 14 mm (average 11 mm). The length of the osseous segment of the artery ranges from 8.5 to 36 mm (average 15 mm). The artery courses medially and takes a reverse loop inside the vertebral artery bony foramina on the inferior surface of the superior facet of the C2 vertebra. In our study, the angle of the loop ranged from 2 to 110° (average 86°). The body and the pars interarticularis of the C2 vertebra are free of the artery. The distance of the tip (or dome) of the vertebral artery loop from the midline of the C2 body, as seen from an anterior transoral surgery perspective, ranged from 6.1 to 16.2 mm (11.7 mm). The distance of the tip (or dome) of the loop from the articular surface of the superior facet ranged from 0.6 to 4.8 mm (average 2.5 mm). The distance of the dome of the vertebral artery groove from the dome of the artery varied from 0 to 4.2 mm (average 2.7 mm). The extra space was filled up with the venous plexus and periosteal tissue (Fig. 3.12).

In ~30% of specimens in our study, the vertebral artery occupied the entire volume of the vertebral artery groove in the inferior surface of the superior facet of C2. In these cases, the occupancy ratio of vertebral artery to bony confinements was considered to be 100%. In the remaining specimens, the oblique width of the bony groove, once unroofed, was measured and put in relation to the diameter of the vertebral artery in that point. The extent of the vertebral artery occupancy in the vertebral artery groove in C2 ranged from 34% to 100% (average 79%) (Fig. 3.13).
V2 Segment

The vertebral artery exits from the transverse process of the C2 vertebra, takes an initial lateral bend, then traverses superiorly. The artery courses anterior to the two roots of the C2 ganglion. The length of this segment of the artery is ~15 mm. The distance of the lateral edge of the ganglion from the vertebral artery ranges from 5 to 11 mm (average 7.5 mm). Two sets of branches arise from the vertebral artery in this segment: a relatively large muscular branch and a small artery traversing along the C2 ganglion into the spinal canal (Fig. 3.14).

V3 Segment

After exiting the transverse process foramen of C1, the vertebral artery takes a posterior bend of ~90° and turns medially to engage in the groove on the superior surface of the posterior arch of the atlas, where, turning around the superior facet of the atlas, it bends anteriorly to enter the spinal canal. The total length of this segment of the artery is ~35 mm. The C1 roots course posteroinferiorly to the artery.

In our study, the distance between the most medial extension of the vertebral artery and the medial edge of the vertebral artery groove on the outer cortex of the posterior arch of the atlas ranged from 2.1 to 5.2 mm (average 4.24 mm). The vertebral artery groove was on average 18.2 mm from the midline, and the vertebral artery in relationship with the groove was 22.1 mm from the midline.\(^1\)

The vertebral artery occupancy ratio was calculated by comparing the width of the bony groove to the width of the artery in that point. The width of the bony groove was measured from the posterior aspect of the lateral mass of C1 to a line connecting the most medial edges of the groove on the posterior arch. The vertebral artery occupancy on the posterior arch of the atlas ranged from 42% to 71% (average 57%) (Fig. 3.15).

A dynamic relationship exists between the vertebral artery and the bones in the CVJ. The serpiginous course of the vertebral artery relative to the axis and the presence of buffer space that is filled up with ve-

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The Vertebral Artery in Relation to the C1–C2 Complex

Microscopic sinuses permit safe and stress-free movements of the artery during rotation of the head and neck. During the turning of the neck, the vertebral artery on the ipsilateral side occupies and fills up the buffer space. In our study, we described the vertebral artery as “tight” when the buffer space in the facet of the axis for the vertebral artery was less defined, as opposed to “loose” when there was abundant buffer space. We speculate that the presence of a tight vertebral artery is a possible cause of vertigo.1

Muscular Anatomy

The muscles concerned with the C1–C2 complex on the anterior aspect are the recti capitis anterior and lateralis, both stretching from the anterior aspect of the transverse process of C1 to the inferior surface of the basilar occipital bone. The longus capitis extends from the inferior surface of the basilar occipital bone and clivus to its attachment on the transverse processes of the third to sixth cervical vertebrae. The longus colli is anterior to the vertebral column and is covered by the longus capitis in its superior aspect. It is divided into three parts and attaches to the transverse processes of the cervical and first thoracic vertebrae, as well as to the anterior aspect of the bodies of the first thoracic vertebrae (Fig. 3.16).

On the posterior aspect, the short, thick bifid spinous process of C2 gives attachment to three muscles: the semispinalis cervicis, the origin of which extends from the C2 to C5 and attaches to the transverse processes of the upper five or six thoracic vertebrae; the inferior oblique, which extends to the transverse process of C1 and forms the inferior limit of the suboccipital triangle; and the rectus capitis posterior major, which extends to the inferior nuchal line, forming the medial limit of the suboccipital triangle. The lateral limit of the triangle is formed by the superior oblique, which extends from the transverse process of C1 to the inferior nuchal line.

The rectus capitis posterior minor converging from the inferior nuchal line to the posterior tubercle on the arch of C1 is the only muscle that attaches to the posterior arch of the atlas. Laterally, the scalenus medius attaches to the transverse process of the axis, and the...
levator scapulae attaches to the transverse process of the atlas (Fig. 3.17).

**Conclusion**

Although the intricate osseous anatomy of the C1–C2 complex can be easily derived from the study of dry specimens or models, which is certainly highly recommended before embarking on surgery in the area, appreciation of the course of the vertebral artery—in particular, its variations—remains more difficult.

An average occupancy ratio of the vertebral artery of 79% in relation to C2 and of 57% in relation to C1 represent a buffer zone that can enhance confidence in dissection and safe reliance on bony landmarks during posterior approaches, especially in regard to C1. As far as C2 is concerned, the range of discrepancies between the bony and vascular anatomy can explain why copious bleeding during transarticular or C2 pedicle screw placement may be more likely to stem from a generous venous plexus filling the periaxial arterial space within the C2 foramen than from arterial injury. Furthermore, and probably more importantly so, in cases in which a screw fixation technique is indicated but a big vertebral artery foramen seen on computed tomography seems to make the procedure too risky, an additional angiographic computed tomographic sequence can reveal the actual presence of a buffer zone and thus enhance both safety and confidence in carrying out the procedure.

**References**

Both lateral mass fixation of the C1 and C2 vertebrae using the plate-and-screw method and transarticular screw fixation provide biomechanically stable unions in patients with atlantoaxial dislocation. Various groups, however, have indicated their concern about the safety of these techniques, considering the danger to the vertebral artery. To use these techniques optimally and safely, it is necessary for the surgeon to have exact anatomical information about the region in a three-dimensional perspective. Only a few reports in the literature describe the quantitative measurements of the craniovertebral region and the parameters appropriate for screw implantation. Most of these reports are based on radiological measurements and deal with the dens and canal diameters. There are only isolated reports detailing quantitative measurements of the lateral masses of C1 and C2.

This chapter considers the various dimensions of the lateral masses of the atlas and axis vertebrae and analyzes their relation with the vertebral artery foramen. It also outlines the safe sites of entry and trajectories for screw implantation. These findings are based on a cadaveric study by Gupta and Goel of 50 dried C1 and C2 vertebrae.

### Atlas

In the study by Gupta and Goel, the superior facet of the atlas was oval and had a groove on either side in the center of the facet in 76% of specimens; it was kidney-shaped in 24% of specimens, and in those cases, the groove was on only one side of the facet. The anteroposterior dimension (mean 19.73 mm) was more than the transverse dimension (mean 11.12 mm) in all superior facets. In no specimen was the facet found to be exactly symmetrical to that on the contralateral side. Both the su-
The inferior facet of the atlas was circular in most of the vertebrae, and the mean anteroposterior (15.76 mm) and transverse (15.22 mm) dimensions of this facet were similar. The vertebral artery foramen lies in the transverse process lateral to the lateral mass. The thickness of the facet under the lateral aspect of the posterior arch of the atlas was 3 to 7 mm (mean 4.16 mm) in the vertical plane and 8.5 to 15 mm (mean 10.65 mm) in the transverse plane. The thickness of the posterior arch of the atlas in relation to the facet of the atlas varied from 3.0 to 7.5 mm (mean 5.05 mm).

The safe angle for screw implantation in the inferior facet of the atlas was determined to be 10 to 20° (mean 15.7°) medial to the sagittal plane and 10 to 20° (mean 15°) superior to the axial plane. In the atlas, the safe site for entry of the screw is in the middle of the posterior surface of the inferior facet. Table 4.1 shows the results of measurements of C1 vertebrae.

### Axis

The shape of the superior articular facet of the axis was oval with a greater anteroposterior than transverse diameter in 54% of cases, oval with a greater transverse than anteroposterior dimension in 27% of cases, and circular in 19% of cases (Fig. 4.2). It is convex in the sagittal plane and is directed laterally to articulate with the inferior facet of C1. The superior facet of C2 characteristically projects from the anterior part of the pedicle and the body of axis in contrast to the inferior facet of C2 and facets of other vertebrae, which project from the junction of the pedicle and laminae. The mean external and internal heights of the superior facet were 8.46 mm (range 5.5–11.5 mm) and 4.62 mm (range 1.5–8.5 mm), respectively. The internal height (i.e., the thickness of the superior facet in the center) was < 2.5 mm in 16% of facets. The depth of the vertebral artery foramen of the axis in the superior facet was 4.36 mm (range 2.0–8.2 mm). The vertebral artery

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**Table 4.1 Analysis of atlas vertebrae**

<table>
<thead>
<tr>
<th>Definition of Parameters</th>
<th>Range (mm)</th>
<th>Mean (mm)</th>
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<tr>
<td>Maximum anteroposterior dimension of the articular surface of the inferior facet</td>
<td>13–21</td>
<td>15.76</td>
</tr>
<tr>
<td>Maximum transverse dimension of the articular surface of the inferior facet</td>
<td>13–20</td>
<td>15.22</td>
</tr>
<tr>
<td>Maximum anteroposterior dimension of the articular surface of the superior facet</td>
<td>15–26</td>
<td>19.73</td>
</tr>
<tr>
<td>Maximum transverse dimension of the articular surface of the superior facet</td>
<td>8–16</td>
<td>11.12</td>
</tr>
<tr>
<td>Thickness of the medial surface of the lateral mass</td>
<td>4–12</td>
<td>8.57</td>
</tr>
<tr>
<td>Thickness of the lateral surface of the lateral mass</td>
<td>15–25</td>
<td>19.11</td>
</tr>
<tr>
<td>Screwable thickness of the posterior surface of the inferior facet in the vertical plane</td>
<td>3–7</td>
<td>4.16</td>
</tr>
<tr>
<td>Screwable thickness of the posterior surface of the inferior facet in the transverse plane</td>
<td>8.5–15.0</td>
<td>10.65</td>
</tr>
<tr>
<td>Thickness of the posterior arch of the atlas near the facet</td>
<td>3.0–7.5</td>
<td>5.05</td>
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<tr>
<td>Best angle for screw implantation in relation to the sagittal plane</td>
<td>15–20°</td>
<td>15.07°</td>
</tr>
</tbody>
</table>

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![Fig. 4.2a–b](image1.png) **a** Photo of a dry C2 bone.

![Fig. 4.2c–d](image2.png) **b** Photo showing the anterior surface of the C2 bone.
Fig. 4.2c–d

C2 facet.
- Photo showing the inferior surface of the C2 vertebra. The location of the vertebral artery groove on the inferior surface of the C2 facets is seen.

Fig. 4.3a–d

Vertebral artery.
- Axial computed tomographic scan showing the large vertebral artery groove in the facet of the axis.
- Coronal T2-weighted magnetic resonance image (MRI) showing the vertebral artery traversing through a large groove in the C2 facet.
- Axial contrast-enhanced MRI showing the vertebral artery in the facet of C2.
- Magnetic resonance angiogram showing the medial loop of the vertebral artery.

Photo showing the superior surface of the C2 vertebra. The screw-able region of the pedicle is divided into nine compartments.
The foramen varies in its extent in the inferior surface of the superior facet. In 84% of cases, the foramen was present partially in the inferior surface of the superior facet and partially in the transverse process, and it was directed laterally and cranially. In 15% of cases, the foramen was present entirely within the inferior surface of the superior facet, and it was directed laterally and not cranially. In one facet, the foramen extended into the body of the axis vertebra.

Because of these anatomical variations, the chances of entering the vertebral artery foramen are significantly high if the screw is passed in the sagittal plane in the superior facet of the axis. The relatively safe screw trajectory was found to be 40° on the medial to sagittal plane and 20° on the superior to axial plane. The safe site of screw entry into the superior facet of the axis is through the superior and medial third compartment of the superior facet without entering into the vertebral artery foramen. The risk is < 5% if the screw is passed from the middle part of the superior facet in the proposed screw trajectory. Screw entry from the lateral third of the superior facet is not safe, with the probability of screw entry into the vertebral artery foramen being in excess of 15%. Medial to an arbitrary vertical line drawn in the sagittal plane at the medial end of the vertebral artery foramen of the axis, the quality and thickness of the cancellous bone in the superior facet of C2 were found to be good (Line A) (Figs. 4.3 and 4.4). Lateral to this line the superior facet is thin, < 2.5 mm in 16% of cases. Thus, not only is screw fixation in this part of the facet dangerous because of its relation of vertebral artery, but bone quality for the screw purchase is also not satisfactory.14 Table 4.2 shows the results of measurements of C2 vertebrae.

Gottlieb measured superior articular facets of 30 human atlas vertebrae and found that a classically described kidney-shaped facet is an infrequent finding and that none of the facets of the two sides were mirror images in their symmetry.13 Huggare and Houghton analyzed articular masses of atlastes in prehistoric Polynesian and Thai skeletons and observed that a right-sided articular mass was constantly and significantly higher than that on the left side.15 None of the facets in the series by Gupta and Goel had mirror symmetry, and the superior facets were kidney-shaped in 24%.14 Heggeness and Doherty studied the trabecular anatomy of the axis and found good cancellous bone quality in the lateral masses beneath the articular surface of the facets, which

![Fig. 4.4 Three-dimensional computed tomographic scan of the atlas and axis showing the location of the vertebral artery and an arterial branch traversing along the C2 root.](image)

**Table 4.2 Analysis of axis vertebrae**

<table>
<thead>
<tr>
<th>Definition of Parameters</th>
<th>Range (mm)</th>
<th>Mean (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maximum anteroposterior dimension of the articular surface of the inferior facet</td>
<td>7–15</td>
<td>10.59</td>
</tr>
<tr>
<td>Maximum transverse dimension of the articular surface of the inferior facet</td>
<td>6–15</td>
<td>9.58</td>
</tr>
<tr>
<td>Maximum anteroposterior dimension of the articular surface of the superior facet</td>
<td>13.0–20.5</td>
<td>16.52</td>
</tr>
<tr>
<td>Maximum transverse dimension of the articular surface of the superior facet</td>
<td>12.0–19.5</td>
<td>15.9</td>
</tr>
<tr>
<td>External height of the superior facet, from the midpoint of the articular surface to the lowest point on the inferior surface of the facet</td>
<td>5.5–11.5</td>
<td>8.46</td>
</tr>
<tr>
<td>Internal height of the superior facet, from the midpoint of the articular surface to the nearest point on the inferior surface of the vertebral artery groove</td>
<td>1.5–8.5</td>
<td>4.62</td>
</tr>
<tr>
<td>Depth of the vertebral artery foramen in the superior facet (i.e., difference between the external and internal heights)</td>
<td>2.0–8.2</td>
<td>4.36</td>
</tr>
<tr>
<td>Screwable thickness of the pedicle, from its internal surface to its external surface at the level of the transverse foramen</td>
<td>4.5–12.0</td>
<td>7.77</td>
</tr>
<tr>
<td>Height of the pedicle, from its superior surface to the inferior surface at the level of the transverse foramen</td>
<td>4.8–14.5</td>
<td>7.90</td>
</tr>
<tr>
<td>Distance of the medial border of the superior facet to line A</td>
<td>0–12</td>
<td>7.5</td>
</tr>
<tr>
<td>Distance of line A from the inferior end of the body</td>
<td>1.0–10.5</td>
<td>5.52</td>
</tr>
</tbody>
</table>
suggested that these are good areas for insertion of an internal fixation device.\(^{16}\)

### Conclusion

The vertebral artery foramen lies laterally in the transverse process of the atlas vertebra. Safe screw implantation is from the middle of the posterior surface of the inferior facet of the atlas, directed 15° on the medial to sagittal plane and 15° on the superior to axial plane. Because the screw passes through the center of the facet, the purchase of the screw is good.

The superior facet of the C2 vertebra has two differing characteristics that make the vertebral artery prone to injury. First, the superior facet of the axis is present in proximity to the body and the medial aspect of the pedicle of the axis. Second, the vertebral artery foramen is located partially or completely in the undersurface of the superior facet of the axis. This location of the vertebral artery foramen makes the vertebral artery prone to injury if the direction of the screw is straight anteriorly in the sagittal plane.\(^5\) The direction of screw implantation in the C2 superior facet joint should be sharply anteromedial to avoid the vertebral artery. If the screw is implanted from the superior and medial third of a screwable pedicle, and the trajectory of the screw is 40° on the medial to sagittal plane and 20° on the superior to axial plane, the probability of entering into the vertebral artery foramen is almost nil.

Table 4.3 shows the site of screw entry and the probability of entering into the vertebral artery foramen of the axis.

<table>
<thead>
<tr>
<th>Medial Third</th>
<th>Middle Third</th>
<th>Lateral Third</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior third</td>
<td>0%</td>
<td>3%</td>
</tr>
<tr>
<td>Middle third</td>
<td>0%</td>
<td>5%</td>
</tr>
<tr>
<td>Inferior third</td>
<td>2%</td>
<td>6%</td>
</tr>
</tbody>
</table>

### References

Stabilization of the craniocervical junction presents a unique biomechanical challenge. The combination of variable anatomy, complex pathologies, complicated biomechanics, and imperfect internal fixators makes in-depth understanding of this region mandatory for successful surgical management. This chapter focuses on biomechanical considerations for stabilization. The reader should be mindful, however, that optimal outcome also depends on other facets of treatment, such as indications, perioperative management, arthrodesis, and adequate decompression of neural elements.

### Relevant Anatomy

#### Occipital Bone

The occipital bone comprises the most caudal portion of the calvarium (Fig. 5.1). The broad, convex portion of the occipital bone below the superior nuchal line is referred to as the suboccipital region. Midway between the superior nuchal line and the foramen magnum is the inferior nuchal line, bisected by the external occipital crest. This region is most frequently used as a point of fixation for occipitocervical constructs. The center of the intracranial transverse sulcus, containing the transverse sinus, is a mean of only ~0.9 mm above the superior nuchal line.¹ The superior nuchal line therefore represents the uppermost landmark of the area potentially available for fixation. In addition, fixators placed at or above this area tend to be quite prominent and pose potential problems with wound healing and skin breakdown. Below the superior nuchal line, the suboccipital bone varies widely in thickness. In the midline, the presence of an internal occipital crest, corresponding to the external occipital crest superficially, contributes to a mean thickness of 13.8 mm at the external occipital protuberance and 8.3 mm at the level of the inferior nuchal line. More laterally, the occipital bone may be considerably thinner, with a mean thickness of only 5.7 mm halfway between the foramen magnum and inferior nuchal line.¹

Relevant to the present discussion, the occipital bone forms the occipital condyles. The paired occipital condyles articulate with the superior articular surfaces of the atlas. The mean anteroposterior length of the ovoid occipital condyle is 23.6 mm, and the mean width and height are 10.6 and 9.2 mm, respectively. The mean anterior and posterior intracondylar distances are 21.0 and 41.6 mm, respectively,² emphasizing the axial plane anteroposterior divergence of these structures. Variations in the shape of the occipital condyle are common. In 20% of specimens, the occipital condyles protrude into the foramen magnum. The intracranial orifice of the hypoglossal foramen is found at the junction of the second and third quarter of the occipital condyle in the majority of specimens.²

#### Atlas (C1)

The first cervical vertebra, or atlas, articulates with the occipital condyles above and the axis below via the odontoid process and the paired atlantoaxial joints. This unique, vaguely ring-shaped vertebra neither possesses a true vertebral body nor articulates with an intervertebral disk. The atlas has a mean anteroposterior and lateral diameter of 45.8 and 78.6 mm, respectively.³ The mean thicknesses of the anterior and posterior rings are 6.4 and 8.0 mm, respectively. Relevant to screw-based methods of fixation to the atlas, the width and height of the C1 lateral mass are 11.6 and 12.7 mm, respectively. The mean distance from the midline to the inner wall of the C1 lateral mass is 14.2 mm. The mean distance to the outer wall is 23.0 mm.⁴

#### Axis (C2)

The second cervical vertebra, or axis, articulates with the occipital condyles above and the third cervical vertebra below. It possesses a unique projection, the odontoid process, which
is critically important in constraining normal movements at the craniocervical junction. The axis has a mean total height of 39.9 mm and a mean odontoid height of 16.6 mm. At the tip, the odontoid has a mean diameter of 11.2 mm in the sagittal plane and 10.8 mm in the coronal plane. Both of these dimensions are decreased closer to the base, or isthmus. The odontoid is angled posteriorly a mean of 16.6°. Significant variation in this angle is seen from patient to patient. The C2 pedicle has a mean height of 9.4 mm and a mean width of 8.0 mm. Pedicles are medially angled, with a mean angulation of 39°. The axis–C3 articulation is similar to other articulations in the subaxial cervical spine.

- **Ligaments**
  - **Transverse Ligament**
    The transverse ligament attaches to the dorsal surface of the odontoid and the lateral masses of the atlas, restricting flexion as well as anterior displacement of the atlas. It is frequently involved in pathological processes involving the craniocervical junction. This ligament has a mean length of 21.9 mm.
  - **Alar Ligament**
    The alar ligament attaches to the ventral and lateral surfaces of the odontoid process and occipital condyles, frequently with a secondary attachment to the lateral masses of the atlas. It restrains axial rotation, side bending, and flexion/extension of the upper cervical spine. This ligament has a mean length of 10.3 mm and is oriented 70° from the sagittal plane. It is elliptical in cross section, 3.6 by 6 mm. Rupture of the alar ligaments appears to be less common than failure of the bony attachments.
    Failure of the alar ligament results in significant alterations in kinematics. Unilateral alar ligament transaction results in an increase in the neutral zone at the occipital–C1 articulation in lateral bending on the contralateral side, without an increase in the range of motion (ROM). In addition, transection of the alar ligament decreases occipital–C1 flexion but not extension. Unilateral and bilateral alar ligament transaction results in significant increases in flexion and extension at C1–C2.

- **Capsular Ligaments**
  The C1–C2 capsular ligaments function primarily in axial rotation. In the presence of intact transverse and alar ligaments, unilateral transection of the capsular ligament results in increased ROM in rotation on the opposite side. It has been theorized, however, that damage to one or both capsular ligaments is not sufficient to produce C1–C2 dislocation.

### Kinematic Biomechanics

#### Occiput–C1

The kinematics of the occiput–C1 segment are determined largely by bony elements. Sagittal plane rotation is the primary movement occurring at the occiput–C1 articulation. Approximately 23.0 to 24.5° of rotation is allowed, limited by impingement of the tip of the dens on the foramen magnum in flexion and tension on the tectorial membrane in extension.

Under normal circumstances, axial plane rotation and coronal plane bending are both <10° per side, both resisted by the occipital–C1 articulation and the alar ligament. Axial rotation at the occiput–C1 is associated with up to 20° of lateral bending in the same direction as axial rotation. Translational movements at the occipital–C1 articulation are reported to be minimal, though sagittal plane rotation is associated with sagittal plane translation, whereas axial plane rotation is associated with lateral translation. The instantaneous axis of rotation in axial rotation is located anterior of the foramen magnum.

#### C1–C2

The kinematics of the C1–C2 segment are determined largely by ligamentous elements. Axial plane rotation is the primary movement occurring at the C1–C2 articulation. Approximately 23.3 to 38.9° of rotation per side are allowed, limited by the C1–C2 articulation, ipsilateral transverse ligament, contralateral alar ligament, and capsular ligaments. Approximately 77% of cervical axial rotation occurs at the C1–C2 segment, with only 4% occurring at the occiput–C1. Axial rotation at C1–C2 is associated with up to 11° of lateral bending in the opposite direction. Interestingly, axial rotation at C1–C2 is also associated with opposite direction axial rotation (though of less magnitude) at the occiput–C1. The instantaneous axis of rotation in axial rotation is located in the central portion of the dens at C1–C2. Sagittal plane rotation is limited to 10.1 to 22.4° by the transverse ligament in flexion, tectorial membrane, and bony anatomy of the C1–C2 articulation. The instantaneous axis of x-plane rotation at C1–C2 has been observed to lie near the posterior cortex of the odontoid, midway between the base and tip. Lateral bending at C1–C2, other than that associated with axial rotation, is limited to 6.8°, primarily by the alar ligament.

Anteroposterior translation at C1–C2 is frequently found in pathological states. Most frequently, this is assessed by measurement of the atlantodens interval on plain radiographs. Although it is generally accepted that an atlantodens interval ≤ 3 mm is normal, a computed tomography study by Rojas et al. suggests that 95% of normal individuals have an atlantodens interval ≤ 2 mm. Anterior translation is resisted by the...
transverse ligament, with lesser contributions from the alar ligaments, accessory atlantoaxial ligaments, and capsular ligaments. Posterior translation is resisted by abutment of the dens on the arch of C1. Translation in other planes is minimal under normal circumstances.

Although overall cervical ROM tends to decrease with age, axial and sagittal plane rotation at C1–C2 tends to remain the same or increase slightly with age.

**Strength Biomechanics**

Ligaments are uniaxial structures that respond to direct tensile or distractive forces. Certain ligaments are capable of resisting tensile forces in a range of directions because of their anatomy. Because more than one type of ligament spans the neighboring vertebrae, the response is dependent on the nature of the load vector. Ligaments are most effective when distracted along the direction of the fibers. Depending on the severity, that is, the magnitude and application of the load vector, internal forces resisted by the various ligaments differ. Internal responses such as strain or stress depend on mechanical properties that are a function of the constituents.

Structural and material properties are obtained by subjecting the ligament to distraction forces. Experiments have been conducted with human cadaver spines: from intact subjects to isolated ligament tests and in situ bone–ligament–bone preparations. Removal of the ligament from the spinal column for testing often results in damage to the structure and may induce fixation failures. In situ preparations have the unique advantage that the ligament under testing is not isolated from its surroundings. In general, to perform these tests, all soft tissues are transected carefully, leaving the ligament under testing to be the only structure to resist the uniaxial distractive force. Anatomical features are given importance during transection procedures. Vertebrae above and below the ligament under testing are fixed, and a six-axis load cell is used to ensure the uniaxial nature of the distractive force; the load cell is placed at the distal end.

Static loading tests are generally divided into strength and stability categories. Strength tests are conducted by applying external loading using testing devices such as an electrohydraulic apparatus. Generally, the specimen is loaded at a constant velocity (millimeters per second) until failure, which is generally identified as the point on the force–deformation curve where an increasing deformation corresponds to a decrease in the force. From the force–deformation curves, peak load, peak deformation, energy, and stiffness characteristics are obtained. Energy is defined as the area of the force–deformation curve until failure. Stiffness is defined as the slope of the force–deformation curve in its most linear phase. Because the specimen is subjected to failure, repeated tests are not possible using strength tests. This procedure can identify the load-carrying capacity of the specimen; provide information regarding the ROM of the specimen under a specific load vector, thus indicating the allowable range of the external insult that can be applied for specimen failure; and delineate the mechanisms of injury, such as bony fractures versus ligament avulsions or rupture, which may assist in understanding the injury biomechanics leading to the instability of the cervical spine. These data can be used to develop mathematical analogues, such as the finite element model of the cervical spine, which can be used to predict the internal (e.g., stresses) and the external biomechanical responses of the cervical spine under normal, injured, and stabilized conditions.

A typical force–deformation response of a specimen (e.g., anterior longitudinal ligament) from a distraction strength test is illustrated in Fig. 5.3. Following the principles of structural mechanics, the nonlinear biomechanical response (in which force does not increase linearly with respect to deformation or vice versa), referred to as the sigmoidal curve, has been classified in terms of the ambient, physiological, traumatic, and failure or posttraumatic loading phases. The stiffness–deflection response of the structure has been used to derive these biomechanical classifications. This system has been used to design a schema to evaluate the onset of injury secondary to loading that can help to define the mechanism of spinal disorders. The ligament is used as an example of the spine structure in the following discussion. During
During this phase, the distractive force and stiffness increase in physiological range. These physiological, traumatic, and microlevel failures in the form of load in the physiological range without compromise structure is capable of effectively absorbing/transmitting load in the physiological range without compromise its integrity, and microlevel failures in the form of fiber tears begin to occur when it is loaded beyond this physiological range. These physiological, traumatic, and posttraumatic phases have been reclassified as the neutral, elastic, and plastic zones, respectively. The neutral zone represents the displacement beyond the neutral position due to the application of a small force, the elastic zone represents the displacement beyond the neutral zone and up to the physiological limit, and the plastic zone represents the deformations beyond the elastic zone until failure. The point of initiation of trauma as identified by the first drop in stiffness in the stiffness–deflection response is the transition region between the elastic and plastic zones. The physiological ROM is from the neutral zone to the end of the elastic zone. Trauma is identified on a biomechanical basis by the beginning of the plastic zone.

Using the above principles, biomechanical testing of the upper cervical ligaments has been conducted. Under in situ testing, the following ligaments at the occipitocervical junction were tested for determining the uniaxial tensile load-carrying capacity. The anterior longitudinal ligament was tested at the C1–C2 level, and the anterior atlanto-occipital membrane was tested at the occiput–C1 level. The ligamentum flavum was evaluated at the C1–C2 level, and its equivalent structure by location, the posterior atlanto-occipital membrane, was evaluated at the occiput–C1 level. The apical and alar ligaments were evaluated individually. The vertical portion of the cruciate ligament was tested. The joint capsules at both the occiput–C1 and C1–C2 levels were tested. The tectorial membrane attaching from C2 to the occiput was also tested. Axial distraction tests at a constant rate of 1 cm per second were applied using an electromechanical testing apparatus. The applied load and the resulting deformations were gathered to obtain the force–deformation response. Table 5.1 outlines the failure force, distraction, energy, and stiffness of human upper cervical spine ligaments.

Factors such as age, gender, and loading rate affect the responses of spine structures. Disease states affect spine biomechanical responses, as the mechanical integrity is often compromised by the abnormality; however, specific
Basic biomechanics of the craniovertebral junction were presented in this chapter. This included the anatomy and fundamental strength and kinematics of the complex. Although emphasis was placed on the adult, interactions between the various components of this complex are critical to maintain the normal functional relationships during development, physiological activities, aging, disease, and traumatic events.

### Table 5.1  Failure force, distraction, energy, and stiffness of human upper cervical spine ligaments

<table>
<thead>
<tr>
<th>Level</th>
<th>Type</th>
<th>Force (N)</th>
<th>Distraction (mm)</th>
<th>Energy (Nm)</th>
<th>Stiffness (N/mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Occipital–C1</td>
<td>Joint capsules</td>
<td>320 ± 129</td>
<td>9.9 ± 8.4</td>
<td>2.03 ± 1.00</td>
<td>32.6 ± 28.0</td>
</tr>
<tr>
<td>Occipital–C1</td>
<td>Anterior atlanto-occipital membrane</td>
<td>232 ± 23</td>
<td>18.9 ± 2.7</td>
<td>2.44 ± 0.63</td>
<td>16.9 ± 3.2</td>
</tr>
<tr>
<td>Occipital–C1</td>
<td>Posterior atlanto-occipital membrane</td>
<td>83 ± 17</td>
<td>18.1 ± 2.7</td>
<td>0.87 ± 0.21</td>
<td>5.7 ± 0.4</td>
</tr>
<tr>
<td>C1–C2</td>
<td>Anterior longitudinal ligament</td>
<td>263 ± 152</td>
<td>11.8 ± 7.0</td>
<td>2.07 ± 1.81</td>
<td>24.0 ± 11.7</td>
</tr>
<tr>
<td>C1–C2</td>
<td>Joint capsules</td>
<td>314 ± 143</td>
<td>9.3 ± 4.5</td>
<td>2.55 ± 2.55</td>
<td>32.3 ± 23.5</td>
</tr>
<tr>
<td>C1–C2</td>
<td>Ligamentum flavum</td>
<td>111 ± 85</td>
<td>9.6 ± 4.3</td>
<td>0.58 ± 0.47</td>
<td>11.6 ± 11.0</td>
</tr>
<tr>
<td>Occipital–C2</td>
<td>Tectorial membrane</td>
<td>76 ± 44</td>
<td>11.9 ± 2.5</td>
<td>0.53 ± 0.31</td>
<td>7.1 ± 2.3</td>
</tr>
<tr>
<td>Occipital–C2</td>
<td>Apical ligament</td>
<td>214 ± 115</td>
<td>8.0 ± 5.3</td>
<td>1.55 ± 1.53</td>
<td>28.6 ± 29.0</td>
</tr>
<tr>
<td>Occipital–C2</td>
<td>Alar ligament</td>
<td>357 ± 220</td>
<td>14.1 ± 7.2</td>
<td>2.72 ± 1.77</td>
<td>21.2 ± 15.7</td>
</tr>
<tr>
<td>Occipital–C2</td>
<td>Cruciate ligament (vertical portion)</td>
<td>436 ± 69</td>
<td>12.5 ± 4.9</td>
<td>4.29 ± 2.98</td>
<td>19.0 ± 0.2</td>
</tr>
</tbody>
</table>

### Acknowledgment
Support by VA Medical Research.

### References


6 Biomechanics of the Unstable Craniovertebral Junction

Vijay K. Goel

From a bioengineer’s perspective, biomechanics of the spine involves an understanding of the interaction among spinal components to provide the desired function in a normal person. Thereafter, one needs to analyze the role of these elements in producing instability. Abnormal motion may be because of external environmental factors the spine is subjected to during activities of daily living (e.g., impact, repetitive loading, and lifting), degeneration, infectious diseases, injury or trauma, disorders, and/or surgery. Furthermore, the field of spinal biomechanics encompasses a relationship among conservative treatments, surgical procedures, and spinal stabilization techniques. This chapter deals with the biomechanics of the unstable craniovertebral joint and the common fixation procedures.

● Role of Environmental Factors in Producing Instability/Injury

High-speed impact loads that may be imposed on the spine are one of the major causes of spinal instability in the cervical region, especially in the upper region. To quantify the likely injuries of the atlas, Oda and coworkers subjected upper cervical spine specimens to high-speed axial impact by dropping 3 to 6 kg weights from various heights. The load produced axial compression and flexion of the specimen. Both bony and soft tissue injuries, similar to Jefferson fractures, were observed. The bony fractures were six bursting fractures, one four-part fracture without a prominent bursting, and one posterior arch fracture. The major soft tissue injury involved the transverse ligament. There were five bony avulsions and three midsubstance tears. The study was extended to determine the three-dimensional load displacements of upper cervical spines (C0–C3) in flexion, extension, and lateral bending before and following the impact loading in the axial mode. The largest increase in flexibility due to the injury was in flexion-extension (~42%). In lateral bending, the increase was on the order of 24%; in axial rotation, it was minimal (~5%). These increases in motion are in concordance with the actual instabilities observed clinically. In patients with burst fractures of the atlas, Jefferson noted that the patients could not flex their heads, but they could easily rotate without pain.3

Heller et al.4 tested the transverse ligament attached to the C1 vertebra by holding the C1 vertebra and pushing the ligament in the middle along the anteroposterior (AP) direction. The specimens were loaded with an MTS Bionix (MTS Systems, Inc., Minneapolis, MN) testing device at various loading rates. Eleven specimens failed within the substance of the ligament, and two failed by bone avulsion. The mean load to failure was 692 N (range 220–1590 N). The displacement to failure ranged from 2 to 14 mm (mean 6.7 mm). This study, when compared with the work of Oda et al.,1,5 suggests that (1) AP translation of the transverse ligament with respect to the dens is essential to produce its fracture; (2) rate of loading affects the type of fracture (bony vs. ligamentous) but not the displacement at failure; and (3) even “axial” impact loads are capable of producing enough AP translation to produce a midsubstance tear of the ligament, as reported by Oda and coworkers.

The contribution to stabilization by the alar ligament of the upper cervical spine is of particular interest in evaluating the effects of trauma, especially in the axial rotation mode. Goel and associates6,7 in a study of occipitoatlantoaxial specimens, determined that the average values for axial rotation and torque at the point of maximum resistance were 68.1° and 13.6 Nm, respectively. They also observed that the value of axial rotation at which complete bilateral rotary dislocation occurred was approximately the point of maximal resistance. The types of injuries observed were related to the magnitude of axial rotation imposed on a specimen during testing. Soft tissue injuries, such as stretch/rupture of the capsular ligaments and subluxation of the C1–C2 facets, were confined to specimens rotated to or almost to the point of maximum resistance. Specimens that were rotated well beyond the point of maximum resistance also showed avulsion fractures of the bone at the points of attachment of the alar ligament or fractures of the odontoid process inferior to the level of alar ligament attachment. The alar ligament did not rupture in any of the specimens. Chang and associates6 extended this study to determine the effects of the rate of loading (dynamic loading) on the occipitoatlantoaxial complex. The specimens were divided into three groups and tested until failure at three different dynamic loading rates, 50°/second, 100°/second, and 400°/second, as compared with the quasistatic (4°/s) rate of loading used by Goel et al.7 The results showed that at the higher rates of loading, (1) the specimens became
fractures have been implicated as being the result of high-energy traumatic events. Indeed, there have been numerous accounts as to the events that lead to odontoid fracture. Schatzker et al. reported that 16 of the 37 cases they reviewed were due to motor vehicle accidents, and 15 cases were the result of high-energy falls. Clark and White reported that all type II (96 patients) and type III (48 patients) fractures they reviewed were attributable to either motor vehicle accidents (−70%) or falls. Alker et al. examined postmortem radiographs of 312 victims of fatal motor vehicle accidents. The cohort exhibited 98 injuries of the cervical spine, of which 70 were seen in the craniovertebral junction. The authors, although not quantifying the degree of dens fractures, hypothesized that odontoid fractures were probably due to hyperextension because of the posterior displacement of the fracture pieces.

There is considerable controversy as to the major load path that causes odontoid fractures. A review of the clinical and laboratory research literature fails to designate a consensus on this issue. Schatzker et al. reviewed clinical case presentations and concluded that odontoid fractures are not the result of simple tension, and that there must exist a complex combination of forces needed to produce these failures. Althoff performed a cadaver study in which he applied various combinations of compression and horizontal shear to the head via a pendulum. Before load onset, the head was placed in neutral, extension, or flexion. The position of the load and the angle of impact, determining the degree of compression with shear, were changed for each experiment. The loading cases in which odontoid fractures were obtained are given in Table 6.1. The results indicated that an impact in the sagittal plane (anterior or posterior) produced fractures that involved the C2 body (type III). As the force vector moved from anterior to lateral, the location of the fracture moved superiorly, with lateral loading producing type I fractures. This led the author to propose a new hypothesis: impact loading corresponding to combined horizontal shear and compression results in odontoid fractures. Althoff dismissed the contributions of sagittal rotation (flexion and extension) to the production of resultant odontoid fracture.

Table 6.1 Ranges of motion reported from in vivo and in vitro studies for the occipitoatlantal joint (occiput–C1)

<table>
<thead>
<tr>
<th>Study</th>
<th>Type of Study*</th>
<th>Total Flexion/ Extension</th>
<th>Unilateral Bending</th>
<th>Unilateral Axial Rotation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fick</td>
<td>in vivo</td>
<td>50</td>
<td>34–40</td>
<td>0</td>
</tr>
<tr>
<td>Poirier and Charpy</td>
<td>in vivo</td>
<td>50</td>
<td>14–40</td>
<td>0</td>
</tr>
<tr>
<td>Werne</td>
<td>in vivo</td>
<td>13</td>
<td>8</td>
<td>0</td>
</tr>
<tr>
<td>Penning</td>
<td>in vivo</td>
<td>30</td>
<td>10</td>
<td>0</td>
</tr>
<tr>
<td>Dvorak et al.</td>
<td>in vivo</td>
<td>–</td>
<td>–</td>
<td>5.2</td>
</tr>
<tr>
<td>Penning</td>
<td>in vivo</td>
<td>–</td>
<td>–</td>
<td>1.0</td>
</tr>
<tr>
<td>Dvorak et al.</td>
<td>in vitro</td>
<td>–</td>
<td>–</td>
<td>4.0</td>
</tr>
<tr>
<td>Panjabi et al.</td>
<td>in vitro</td>
<td>3.5/21.0</td>
<td>5.5</td>
<td>7.2</td>
</tr>
</tbody>
</table>

*The in vivo studies represent passive range-of-motion tests, whereas the in vitro studies represent motion at 1.5-Nm occipital moment loading.
Mouradian et al. reported on a cadaver and clinical model of odontoid fracture. In their opinion, “it seems reasonable to assume that shearing or bending forces are primarily involved.” The cadaver experimentation involved anterior or lateral translation of the occiput, as well as lateral translation of the atlantal ring. In forward loading, the odontoid was fractured in 9 of the 13 cases, with 8 type III fractures and 1 type II fracture. The lateral loading specimens evidenced similar patterns of odontoid fracture regardless of the point of load application (on the occiput or on the atlas). In 11 specimens, lateral loading resulted in 10 type II fractures and 1 type III fracture. The clinical model involved reviewing 25 cases of odontoid fracture. The authors reported that 80% of these cases resulted from flexion or flexion-rotation injuries. They pointed out that the clinical data do not reflect the lateral loading cadaver experimentation results. In fact, they state that “a pure lateral blow probably did not occur in any [clinical] case.” However, their clinical data indicated that the remaining 20% of the odontoid injuries could be ascribed to extension injuries. Technical difficulties precluded cadaver experimentation of this possible mechanism.

Experimental investigations dealing with the pathogenesis of odontoid fractures have failed to produce a consensus as to the etiology of these fractures. These findings may actually reflect the diversity of causal mechanisms, suggesting that various mechanical factors are coincident in producing these fractures. It is difficult to discern if this is the case or if this is due to the inhomogeneity of cadaver experiment methodology. That is, some of the boundary and loading conditions used by the surveyed studies are vastly different and have produced divergent results. In addition, the anatomical variants of the craniovertebral osteoligamentous structures could also be integral to the cadaver study outcomes. The purpose of the study undertaken by Puttlitz et al. was to use the finite element method, in which the loading and kinematic constraints can be exactly designated, for elucidating the true fracture etiology of the upper cervical spine (Fig. 6.1). Previous laboratory investigations of odontoid process failure have used cadaver models. However, shortcomings associated with this type of experimentation and the various loading and boundary conditions may have influenced the resulting data. Utilization of the finite element method for the study of odontoid process failure has eliminated confounding factors often seen with cadaveric testing, such as interspecimen anatomical variability and age-dependent degeneration. This has allowed the authors to isolate changes in complex loading conditions as the lone experimental variable for determining odontoid process failure.

There are many scenarios that are capable of producing fracture of the odontoid process. Force loading, in the absence of rotational components, can reach maximum von Mises stresses that far exceed 100 MPa. Most of these loads are lateral or compressive in nature. The maximum stress obtained was 177 MPa due to a force directed in the posteroinferior direction. The net effect of this load vector and its point of application, the posterior aspect of the occiput, is to produce a compression, posterior shear, and extension because of the load’s offset from the center of rotation. This seems to suggest that
extension and compression can play a significant role in the development of high stresses, and possibly failure, of the odontoid. The location of the maximum stress for this loading scenario was in the region of a type I fracture. The same result, with respect to lateral loading, was obtained by Althoff. However, he dismissed the contribution of sagittal plane rotation to development of odontoid failures. The results of this study disagree with that finding. Posteroinferior loading with extension produced a maximum von Mises stress in the axis of 226 MPa. As stated previously, the load vector for this case intensifies the degree of extension, probably producing hyperextension. The addition of the extension moment did not change the location of the maximum stress, still identifiable in the region of a type I fracture. Mouradian et al.’s clinical study suggested that almost 20% of the odontoid fracture cases they reviewed involved some component of extension. The involvement of extension in producing odontoid process failures can be explained by its position with respect to the atlantal ring and the occiput. As extension proceeds, the contact force produced at the atlanto-dental articulation increases, putting high bending loads on the odontoid process. The result could be failure of the odontoid. Increasing tension of the alar ligaments as the occiput extends could magnify these bending stresses via superimposition of the loads, resulting in avulsion failure of the bone (type I).

Although the finite element model predicted mostly higher stresses with the addition of an extension moment, it showed that, in most cases, flexion actually mitigates the osseous tissue stress response. This was especially true for compressive (inferior) force application. Flexion loading with posterior application of an inferior load vectorally decreases the overall effect of producing extension on the occiput. None of the studies surveyed for this investigation pinpointed flexion per se as a damage mechanism for odontoid failure. The findings of this study supported the lack of evidence of flexion as a causal mechanism for failure. In addition, the data suggested that flexion can act as a preventive mechanism against odontoid fracture.

Once again, the lateral bending results support the hypothesis of extension being a major injury vector in odontoid process failure. Inferior and posteroinferior loads with lateral rotation resulted in the highest maximal von Mises stress in the axis. Lateral loading also intensified the maximal stress in compression, suggesting rotations that incorporate a component of both lateral and extension motion may cause odontoid failures. Many of the lateral bending scenarios resulted in the maximum von Mises stress being located in the type II and type III fracture regions. In fact, the only scenarios that lead to the maximum stress in the type I area were when there was an inferior or posterior load applied with the lateral bending. This is, again, suggestive that the extension moment, produced by these vectors and their associated moment arms (measured from the center of rotation), can result in more superiorly located fractures.

Overall, this investigation has indicated that extension and the application of extension via force vector application cause the greatest risk of superior odontoid failure. The hypothesis of extension as a causal mechanism of odontoid fracture includes coupling of this motion to other rotations. Flexion seems to provide a protective mechanism against force application that would otherwise cause a higher risk of odontoid failure.

Changes in Motion Due to Degeneration/Trauma

The degenerative process can affect all of the spinal elements, and trauma can lead to partial or full destruction of the spinal elements. As such, the motion behavior of the segment will change. The rotation-limiting ability of the alar ligament was investigated by Dvorak et al. A mean increase of 10.8° or 30% (divided equally between the occipito-atlantal and atlanto-axial complexes) in axial rotation was observed in response to an alar lesion on the opposite side. Panjabi and coworkers determined the effects of alar ligament transections on the stability of the joint in flexion, extension, and lateral bending modes. Their main conclusion was that the motion changes occurred subsequent to alar ligament transection. The increases, however, were directional-dependent. Crisco and associates compared changes in threedimensional motion of C1 relative to C2 before and after the capsular ligament transections in axial rotation. Two groups of cadaveric specimens were used to study the effect of two different sequential ligamentous transections. In the first group (n = 4), transection of the left capsular ligament was followed by transection of the right capsular ligament. In the second group (n = 10), transection of the left capsular ligament preceded transection of left and right alar and transverse ligaments. The greatest changes in motion occurred in axial rotation to the side opposite the transection. In the first group, transection of left capsular ligaments resulted in a significant increase in axial rotation range of motion to the right of 1°. After the right capsular ligament was transected, there was a further significant increase of 1.8° to the left and of 1.0° to the right. Lateral bending to the left also increased significantly by 1.5° after both ligaments were cut. In the second group, with the nonfunctional alar and transverse ligaments, transection of the capsular ligament resulted in greater increases in range of motion: 3.3° to the right and 1.3° to the left. Lateral bending to the right also increased significantly by 4.2°. Although the matter is more complex than this, in general, these studies show that the major function of the alar ligament is to prevent axial rotation to the contralateral side. Transection of the ligament increases the contralateral axial rotation by ~15%.

The dens and the intact transverse ligament provide the major stability at the C1–C2 articulation. The articu-
lar capsules between C1 and C2 are loose, to allow a large amount of rotation and provide a small amount of stability. Although the C1–C2 segment is clinically unstable after failure of the transverse ligament, resistance against gross dislocation is probably provided by the tectorial membrane, the ala, and the apical ligaments. With transection of the tectorial membrane and the alar ligaments, there is an increased flexion of the units of the occipitoatlantoaxial complex and a subluxation of the occiput. It was also demonstrated that transection of the alar ligament on one side causes increased axial rotation to the opposite side by ~30%.

Fielding performed a biomechanical study investigating lesion development in rheumatoid arthritis. The study tested 20 cadaveric occipitoatlantoaxial specimens for transverse ligament strength by application of a posterior force to the atlantal ring and found atlantoaxial subluxation of 3 to 5 mm and increased atlas movement on the axis after rupture of the transverse ligament. From this study, Fielding was able to conclude that “transverse ligament represents a strong primary defense against anterior shift of the first cervical vertebra.” Puttlitz et al. developed an experimentally validated ligamentous, nonlinear, sliding contact three-dimensional finite element model of the occiput–C1–C2 complex generated from 0.5-mm-thick serial computed tomography scans (Fig. 6.1). The model was used to determine specific structure involvement during the progression of rheumatoid arthritis and to evaluate these structures in terms of their effect on clinically observed erosive changes associated with the disease by assessing changes in loading patterns and degree of anterior atlantoaxial subluxation (AAS) (Table 6.2). The role of specific ligament involvement during the development and advancement of AAS was evaluated by calculating the anterior atlantoaxial interval and posterior atlantoaxial interval after reductions in transverse, alar, and capsular ligament stiffness. (The stiffness of transverse, alar, and capsular ligaments was sequentially reduced by 50%, 75%, and 100%, respectively, of their intact values.) All models were subjected to flexion moments, replicating the clinical diagnosis of rheumatoid arthritis using full-flexion lateral plane radiographs. Stress profiles at the odontoid process–transverse ligament junction were monitored. Changes in loading profiles through the occiput–C1 and C1–C2 lateral articulations and their associated capsular ligaments were calculated. Posterior atlantoaxial interval values were calculated to correlate ligamentous destruction to advancement of AAS. As an isolated entity, the model predicted that the transverse ligament had the greatest effect on anterior atlantoaxial interval in the fully flexed posture. Without transverse ligament disruption, both alar and capsular ligament compromise did not contribute significantly to the development of AAS. Combinations of alar and capsular ligament disruptions were modeled with transverse ligament removal in an attempt to describe the interactive effect of ligament compromise, which may lead to advanced AAS. Alar ligament compromise with intact capsular ligaments markedly increased the level of AAS (Table 6.2). Subsequent capsular ligament stiffness loss (50%) with complete alar ligament removal led to an additional decrease in posterior atlantoaxial interval of 0.92 mm. Simultaneous resection of the transverse, alar, and capsular ligaments resulted in a highly unstable situation. The model predicted stresses at the posterior base of the odontoid process would be greatly reduced, with transverse ligament compromise beyond 75% (Fig. 6.2). Decreases through the lateral occiput–C1 and C1–C2 articulations

Table 6.2 Combinations of ligament stiffness reductions with the resultant degree of AAS, as indicated by the AADI and PADI values at full flexion (1.5-Nm moment)

<table>
<thead>
<tr>
<th>Reduction in Ligament Stiffness (%)</th>
<th>Criteria (mm)</th>
</tr>
</thead>
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<tr>
<td>Transverse Alar Capsular AADI PADI</td>
<td></td>
</tr>
<tr>
<td>0 0 0 2.92 15.28</td>
<td></td>
</tr>
<tr>
<td>100 0 50 5.77 12.43</td>
<td></td>
</tr>
<tr>
<td>100 0 75 6.21 11.99</td>
<td></td>
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<tr>
<td>100 50 0 7.42 10.79</td>
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</tr>
<tr>
<td>100 75 0 7.51 10.71</td>
<td></td>
</tr>
<tr>
<td>100 100 50 8.43 9.83</td>
<td></td>
</tr>
</tbody>
</table>

* Zero (0%) ligament stiffness values represent completely intact ligament stiffness, 100% corresponds to total ligament destruction (via removal).

Abbreviations: AADI, anterior atlantodental interval; AAS, anterior atlantoaxial subluxation; PADI, posterior atlantodental interval.

Fig. 6.2a–d Contours of periosteal von Mises equivalent stresses as compromise of the transverse ligament progresses.

a Fully intact (normal stiffness) case.
b After 50% stiffness reduction.
c After 75% stiffness reduction.
d After complete removal of the transverse ligament. The contours demonstrate gradual stress relief of the bone at the odontoid process–transverse ligament junction.
were compensated by their capsular ligaments. The data indicate that there may be a mechanical component (in addition to enzymatic degradation) associated with the osseous resorption seen during rheumatoid arthritis. Specifically, erosion of the base of the odontoid may involve Wolff’s law of unloading considerations. Changes through the lateral aspects of the atlas suggest that this same mechanism may be partially responsible for the erosive changes seen during progressive rheumatoid arthritis. PADI values indicate that complete destruction of the transverse ligament, coupled with alar and/or capsular ligament compromise, exists if advanced levels of AAS are present.

Biomechanics of Stabilization Procedures

Stabilization of the craniovertebral junction is common, and its importance for treating lesions, fractures, and tumors associated with rheumatoid arthritis cannot be overestimated. Currier et al. studied the degree of stability provided by a rod/plate-based instrumentation system. They compared this new device to the Ransford loop technique and a plate system using C2 pedicle screws, first described by Goel and Laheri in 1988. Transverse and alar ligament sectioning and odontoidectomy destabilized the specimen. All three fixation systems significantly reduced motion as compared with intact and injured spines in axial rotation and extension. The new device did not significantly reduce motion at C1–C2 in flexion, and none of the devices were able to produce significant motion reductions in C1–C2 lateral bending. The authors claimed, based on these findings, that the new system is equivalent or superior to the other two systems for obtaining occipitocervical stability. Oda et al. investigated the comparative stability afforded by five different fixation systems. Type II odontoid fractures were created to simulate instability. The results indicate that the imposed dens fracture decreased construct stiffness as compared with the intact case. Overall, the techniques that used screws for cervical anchors provided greater stiffness than the wiring techniques. Also, the system that used occipital screws with C2 pedicle screw fixation demonstrated the greatest construct stiffness for all rotations. Puttlitz used the finite element model of the occiput-C1-C2 complex to investigate the biomechanics of a novel hardware system (Fig. 6.3). Finite element models representing combinations of cervical anchor type (C1–C2 transarticular screws vs. C2 pedicle screws) and unilateral versus bilateral instrumentation were evaluated. All models were subjected to compression with pure moments in flexion, extension, or lateral bending. Bilateral instrumentation provided greater motion reductions than the unilateral hardware. When used bilaterally, C2 pedicle screws approximate the kinematic reductions and hardware stresses (except in lateral bending) that are seen with C1–C2 transarticular screws. The finite element model predicted that the maximum stress was always located in the region where the plate transformed into the rod. Thus, the author felt that C2 pedicle screws as described by Goel and Laheri should be considered as the primary method of treatment or as an alternative to C2–C1 transarticular screw usage when bilateral instrumentation is applied.

The occipitoatlantoaxial complex is a unique structure that offers extensive motion while allowing vertebrae...
to be interlocked, forming an amazingly stable three-dimensional structure. The stability of the normal segments is provided mainly by the ligamentous complex due to the lack of intervertebral disks and the transverse plane orientation of the atlanto-occipital and atlantoaxial joints.

Of these two joints, the atlantoaxial complex is more easily destabilized in certain pathological conditions. Causes of atlantoaxial instability include trauma, tumor, congenital malformation, and inflammatory diseases. Significant atlantoaxial instability is a potentially serious progressive condition that, if untreated, may result in local pain, myelopathy, or ultimately death. Surgical intervention is often indicated to realign and stabilize the segment and to decompress the neural structures if necessary.

Different strategies to fix the atlantoaxial complex can be found in the literature. Commonly available fixation techniques to stabilize the atlantoaxial complex are posterior wiring procedures (e.g., Brooks and Gallie fusions), interlaminar clamps (Halifax), and transarticular screw (Magerl technique), either alone or in combination, as well as Goel’s technique of C1-lateral mass and C2-pars screw fixation.

Posterior wiring procedures and interlaminar clamps are obviously easier to accomplish. However, these do not provide sufficient immobilization across the atlantoaxial complex. In particular, posterior wiring procedures place the patient at risk of spinal cord injury because of the sublaminar passage of wires into the spinal canal. Interlaminar clamps offer the advantage of avoiding the sublaminar wire hazard and have more rigid biomechanical stiffness than posterior wiring procedures.

Transarticular screw fixation (TSF), in contrast, affords a stiffer atlantoaxial arthrodesis than posterior wiring procedures and interlaminar clamps. TSF does have some drawbacks, including injury of vertebral artery, malposition, and screw breakage. Furthermore, body habitus (obesity or thoracic hyperkyphosis) may prohibit achieving the low angle needed for screw placement across C1 and C2. Recently, a technique of Goel incorporating screw and plate/rod fixation (SRF) that minimizes the risk of injury to the vertebral artery and allows intraoperative reduction has been reported. The configuration of this technique, which achieves rigid fixation of the atlantoaxial complex, consists of lateral mass screws at C1 and pedicle screws at C2 linked via longitudinal plates/rods with constrained coupling devices.

The purpose of study by Kuroki and associates was to compare the biomechanical stability impaired to the atlantoaxial complex by either the TSF or SRF technique and to assess how well these methods withstand fatigue in a cadaver model. In the unilateral fixations, the SRF group was stiffer than the TSF group in flexion loading, but there were no evident differences in other directions. In the bilateral fixations, SRF was more stable than TSF, especially in flexion and extension. These results were similar to those reported by Richter et al., yet different from those of Lynch et al. The instrumentation procedure (screw length, type of constrained coupling device, etc.), the destabilization technique, and the condition of the specimens might have an influence on the results. In the current study, when stabilizing the atlantoaxial segments, all screws were placed bicortically in both techniques in accordance with the procedure described by Goel and Laheri. Previous work has demonstrated that bicortical cervical vertical screws are superior to unicortical screws in terms of pullout strength and decreased wobble. Most surgeons, however, prefer unicortical screwing at C1 and C2 levels to reduce the risk of penetration during surgery. This could affect the outcome. We initially connected the screw to the plate/rod using the oval-shaped constrained coupling device recommended for use in C1 and C2 vertebrae. The stability was not judged adequate, however, so we altered the procedure to use the stiffer circle-shaped constrained coupling device. With regards to the destabilization procedure, there are three typical methods: sectioning of intact ligaments, odontoid fracture, and odontoïdectomy. In the present study, the atlantoaxial complex was destabilized by ligament transection to simulate ligamentous instability, whereas Lynch et al. used odontoïdectomy. Furthermore, the bone quality of specimens affects the screw–bone interface stability. These factors were possibly reflected in other results. However, results were not statistically different between TSF and SRF, so they could be interpreted equivalent in terms of effective stabilization when compared with the intact specimen.

In unilateral TSF and SRF, the fixed left lateral atlantoaxial joint acted as a pivot in left axial rotation and as a fulcrum in left lateral bending, thus leading to an increase in motion. This motion could be observed with the naked eye.

Stability in flexion and extension of the bilateral TSF group was inferior to that of the SRF group. Henriques et al. and Naderi et al. also reported a similar tendency. Henriques et al. thought it most likely due to the transarticular screws being placed near the center of motion between C1 and C2. This was judged to be another reason that the trajectory of the screws is consistent with the motion direction of flexion and extension. So, if TSF is combined with some posterior wiring procedures, the stability in flexion and extension will increase.

**Conclusion**

The stability (or instability) of the human spine is integral to the diagnosis and treatment of patients with spinal disorders. The stability of the spine as portrayed by its motion characteristics can be determined through the use of clinical and radiographic criteria or other methods of determining the orientation of one spinal vertebra with respect to another. The instability can be the result of injury, disease, or other factors, including surgery. Therefore, it is necessary to become familiar with recent findings and suggestions that deal with the instability that can result from such procedures. The prevalence of spinal fusion and stabilization
procedures to restore spinal stability is continuously increasing. This chapter has presented many of the contemporary biomechanical issues germane to stabilization and fusion of the spine. Because of the variety of devices available, various testing protocols have been developed in an attempt to describe the mechanical aspects of these devices. These investigations reveal comparative advantages (and disadvantages) of the newer designs to existing hardware. Subsequent in vivo testing, specifically animal models, provides data on the performance of the device in a dynamic physiological environment. All of the testing, in vitro and in vivo, helps to build confidence that the instrumentation is safe for clinical trial. Future biomechanical work is required to produce newer devices and optimize existing ones, with an eye toward reducing the rates of nonfusion and pseudarthrosis. In addition, novel devices and treatments that seek to restore normal spinal function and loading patterns without fusion continue to necessitate advances in biomechanical methods. These are the primary challenges that need to be incorporated in future biomechanical investigations. Finally, one has to gain understanding of the effects of devices at the cellular level and undertake outcome assessment studies to see if the use of instrumentation is warranted for the enhancement of the fusion process.

References


Placement of instrumentation for stabilization of the craniocervical junction requires that fixation points and constructs be tailored to individual patient anatomy and that pathological movements be controlled. The latter are usually composed of some combination of anteroposterior translation, axial, sagittal, and coronal plane translation, as well as vertical translation (cranial settling).

### Fixation Points

#### Occiput

Formerly, fixation to the occiput was generally accomplished by means of stainless steel wire, generally 18 gauge, looped through adjacent burr holes or through the diploic space. Although this provided some degree of stability, several problems were noted, including pull-through, dural laceration with cerebrospinal fluid leakage, and difficult placement after suboccipital craniectomy. As a result, screw-based mechanisms of occipital fixation were developed. 1, 2

Bicortical placement of occipital screws is preferred. The pullout strength of bicortical screws has been noted to be 50% greater than that of unicortical screws; the pullout strength of unicortical screws is similar to that of 18-gauge wire. 3 Screw location is equally important, as screws placed in the midline demonstrate greater insertional torque and greater pullout strength than screws placed more laterally. 4 Despite the fact that longer screws may often be placed in the suboccipital region, 3.5-mm titanium screws longer than 7 mm typically break before pulling out, 5 suggesting that the use of larger-diameter screws should be considered when possible. Also, though shorter, laterally placed occipital screws may offer less resistance to pullout, and constructs incorporating multiple laterally placed screws may perform equivalently to those with midline screws. There does not appear to be an advantage to the use of constrained (locking) screws in the suboccipital region. 6

Promising alternative techniques have been described. Mingsheng et al. described a technique for the placement of screws into the diploic space of the occipital bone. 7 Although no biomechanical data were presented, the use of screws with a mean length of nearly 26 mm was found to be possible. Pait et al. described a technique for “inside-outside” screw placement in the suboccipital region, in which a threaded stud is placed through a burr hole with the threads facing outward. 8 The pullout strength in synthetic bone was found to be superior to wire cables and bone screws. 9

#### Occiput–C1

In 2001, Grob described a technique for direct posterior occiput–C1 transarticular fixation, describing its use in a patient with occiput–C1 dislocation. 10 In the case described, the construct was reinforced with a posterior occiput–C2 plate. Gonzalez et al. described a technique for direct fixation of the occiput–atlas using transarticular screws. 11 Though allowing a significantly greater range of motion (ROM) at the occiput–C1 in flexion and extension in pure moment testing than an occipital plate–C2 construct, it was concluded that the technique was a useful alternative, especially in cases where suboccipital bone is deficient, and there is not significant instability at C1–C2. 11 Incorporation of posterior occiput–C1 transarticular screws into larger constructs, though theoretically possible, has not been described. Goel and Laheri discussed the use of a C2 pars screw in isolation or in combination with a C1 lateral mass screw for the fixation of the cervical end of the occipitocervical plate/rod. 1

Dvorak et al. described a technique for direct anterior occiput–C1 fixation, noting that this construct was as effective as two posterior fixation techniques in lateral bending and axial rotation but less effective when tested in flexion and extension. 12 Given that instability in sagittal plane rotation is frequently encountered in pathological conditions affecting the craniovertebral junction, it is interesting to note the reported clinical successes.

#### Atlas

Traditionally, fixation to the atlas was generally accomplished by means of sublaminar stainless steel wire or, more recently, braided stainless steel or titanium cable. This technique remains useful in cases where the C1 arch is not deficient, fractured, or removed for decompression. It should be kept in mind, however, that the complication rate of sublaminar wire placement in the cervical spine has been estimated to be ~7%. 13
Goel and Laheri first described the use of screws in the lateral mass of C1 in 1988, reporting excellent outcomes in 30 patients with atlantoaxial subluxation. Some authors have recently introduced polyaxial screws using the basic surgical technique described by Goel. These techniques obviate some of the deficiencies associated with sublaminar wire placement, particularly in the absence of a viable C1 lamina.

Landmarks for the placement of C1 lateral mass screws have been studied in detail. Gupta and Goel performed a cadaveric anatomical study of the C1 lateral mass and C2 nerve root, noting that the mean distance from the midline to the midpoint of the C1 lateral mass was 17.6 mm, and the mean width of the posterior arch at the point of the vertebral artery overpass was 4.7 mm. With an entry point above the C2 nerve root (or the space available after the sectioning of the C2 root), at the junction of the C1 posterior arch and the midpoint of the posteroinferior part of the C1 lateral mass, screws can be placed with a medial angulation of 9.8 to 21.6° and a superior angulation of 17.8 to 28.8°, depending on the individual patient’s anatomy. Using this technique, a screw thread length of 19.1 to 25.9 mm can be accommodated within the lateral mass. Resnick et al. examined computed tomography (CT) scans in 50 consecutive patients without known pathology of the craniocervical junction and found that placement of screws into the lateral mass of C1 was possible in every instance. Although C2 nerve root irritation as a result of C1 lateral mass placement inferior to the C1 arch is a potential complication, the incidence has been reported to be 12%, and the symptoms are temporary. C1 lateral mass screws, when placed using a standard entry point and trajectory, exhibited a pullout force of 1818.16 N, comparable to that of C2 pedicle screws. Incorporation of C1 lateral mass screws into a longer occipitocervical instrumentation construct, though possible in many instances, may be cumbersome secondary to crowding of screw heads at the occipitocervical rod bend (Fig. 7.1).

A variant technique for placement of C1 lateral screws has been described by Tan et al., using an entry point 18 to 20 mm lateral to the midline and 2 mm superior to the inferior border of the arch. The authors determined that a screw with a mean length of 29.65 mm could be placed in 92% of 50 anatomical specimens. The primary limiting factor was the thickness of the posterior arch in the region of the vertebral artery groove.

**Axis**

The use of sublaminar wires or cables remains a viable option for fixation at C2, particularly in the setting of diminutive or malformed posterior elements, so long as the C2 lamina remains structurally intact. However, the development of several screw-based fixation options has greatly decreased the general applicability of this technique.

Screws placed into the pars interarticularis of C2 have gained wide acceptance as caudal anchors for occipitocervical instrumentation constructs. Following a modification of the technique used in the subaxial cervical spine, screws are placed using an entry point at the junction of the inferior border of the lamina and the midpoint of the inferior facet process of C2, with a trajectory 15° medial and parallel to the superior surface of the lateral mass (usually −35° superior) in the sagittal plane. Bicortical screw purchase is not used. In case of a deep vertebral artery groove, screw length may be shortened to avoid injury. A minor, though potentially useful variant of this technique employs a less medially directed trajectory, similar to that of a C1–C2 transarticular screw (see the later description), through the lateral mass of C2. This technique allows the later conversion to a C1–C2 transarticular screw, if necessary.

![Fig. 7.1 Potential fixation points at C1 and C2. C1 lateral mass screw (A), C1–C2 transarticular screw (B), C2 intralaminar screw (C), and offset connector between the C2 intralaminar screw and the longitudinal element (D). Note that the head of the C1 lateral mass screw falls very near the bend in the longitudinal element.](image-url)
Goel and Laheri described placement of screws into the pedicle of C2 (Fig. 7.2). A great deal of confusion exists in the literature regarding the nomenclature C2 “pedicle.” For the purposes of this discussion, a C2 pedicle screw is defined as one that passes through the C2 pedicle along its axis and terminates within the body of C2 (Fig. 7.3). Usually with the assistance of intraoperative neuronavigation techniques, the pedicle of C2 may be cannulated using a 39° medial trajectory parallel to the inferior end plate in the sagittal plane. Radiographic analyses have shown that 91% of C2 pedicles could accept a screw at least 4 mm in diameter. C2 pedicle screws have been shown to have significantly higher resistance to pullout than C2 pars interarticularis screws and to provide caudal fixation equivalent to C1–C2 transarticular screws in finite element modeling. C2 pars interarticularis and pedicle screws are readily incorporated into longer occipitocervical constructs.

Goel and Kulkarni placed the screw into the spinolaminar junction, incorporating the thickness of the spinous process. Wright described the technique for placement of intralaminar C2 screws. Using an entry point at the base of the C2 lamina, crossing screws may be placed into the cancellous bone between the cortices (Fig. 7.4). Gorek et al. demonstrated equivalent ROM reduction in C1–C2 constructs anchored by intralaminar C2 screws and C2 pars interarticularis screws and to provide caudal fixation equivalent to C1–C2 transarticular screws in finite element modeling. C2 pars interarticularis and pedicle screws are readily incorporated into longer occipitocervical constructs.

Fig. 7.2a, b Lateral x-ray with the head in (a) flexion and (b) extension showing lateral mass plate and screw fixation. Segmental bone fusion can be observed.
pedicle screws. When employed as a salvage technique, C2 lamina screws exhibit a pullout strength at least equivalent, if not superior, to screws placed into the pars interarticularis of C2. Though incorporation into longer constructs generally requires three-plane rod bending or special offset connectors, C2 lamina screws may be particularly useful in situations where the C2 lateral mass or pedicle is attenuated. The placement of the screw at the spinolaminar junction as described by Goel and Kulkarni appears to provide more stability to the construct than the placement of the screw in the lamina.

**C1–C2 Transarticular Screws**

C1–C2 transarticular screws were initially developed for stabilization of the C1–C2 segment and are excellent, even when placed unilaterally, at controlling axial plane rotation at that location. Bilateral or even unilateral C1–C2 transarticular screw fixation, in combination with intraspinous bone graft wiring, has been shown to be adequate in the majority of patients for the treatment of isolated atlantoaxial instability. Screws are placed using an entry point at the level of the inferior border of the C2 lamina and the midpoint of the ipsilateral pars interarticularis of C2. Using intraoperative fluoroscopy or neuronavigation, a pilot hole is drilled with a slightly medial trajectory through the lateral mass of C2, across the C1–C2 articulation, and into the lateral mass of C1, using the anterior arch of C1 as a fluoroscopic target.

Not all patients possess anatomy at C1–C2 suitable for the placement of transarticular screws. Potential impediments to placement include irreducible deformity and the presence of a deep vertebral artery groove. Radiographic analyses by Lehman et al. showed that C1–C2 transarticular screws could be placed in 113 of 120 (94%) sides examined in patients without rheumatoid arthritis. Abou Madawi et al. examined the vertebral artery groove in 50 anatomical specimens, noting that in 11 (22%) the groove was deep enough to preclude the placement of a C1–C2 transarticular screw on at least one side. This correlates well with the study of Paramore et al., in which it was postulated, based on CT data, that 18% to 23% of patients may not be suitable for C1–C2 transarticular screw placement on at least one side.

**Constructs**

Multiple authors have commented that, given the current sophistication of occipitocervical instrumentation systems, the design of an occipitocervical instrumentation construct should be dictated more by clinical than biomechanical concerns. Nevertheless, certain principles regarding their design can be deduced from clinical and laboratory investigations.

Whenever possible, cranial anchors should be screw-based. Constructs incorporating midline suboccipital screws have been shown to offer significantly higher stiffness in extension and axial rotation than constructs incorporating stainless steel rectangles anchored with stainless steel cables. Likewise, whenever possible, screw-based fixation points should be used at C2. To control flexion and extension, a construct incorporating screw-based points of fixation at the occiput and C2 is
necessary. In the study of Hurlbert et al., only constructs with these two characteristics exhibited greater stiffness in sagittal plane rotation than uninjured specimens. With regard to the type of C2 screw used, it appears that C1–C2 transarticular screws and C2 pedicle screws have some advantages over other types. Occipitocervical constructs anchored by C1–C2 transarticular screws performed as well as or better than those anchored by sublaminar cables, C2 pars, or C2 lamina screws in ROM testing. C2 pedicle screws, not tested in the study by Wolfia et al., appear to be an excellent alternative. In a biomechanical study by Oda et al., constructs incorporating C1–C2 transarticular screws or C2 pedicle screws offered significant biomechanical advantages compared with sublaminar wiring or lamina hooks, with C2 pedicle screws offering superior stiffness in all loading modes, including anteroposterior translation. Puttlitz et al. in fact reported that a construct incorporating suboccipital screws, C1 lateral mass screws, and C2 pedicle screws showed similar reduction in motion when compared with a construct incorporating suboccipital screws and C1–C2 transarticular screws.

When the placement of C1–C2 transarticular screws or C2 pedicle screws is not possible or practical, intralaminar screws or pars interarticularis screws at C2 are an excellent option. In the kinematic study of Wolfia et al., constructs anchored by C2 pars screws performed equivalently to the C1–C2 transarticular screw-based constructs in all modes except lateral bending. In fact, any of the seven constructs tested, including those anchored by sublaminar cables at C2, reduced ROM in all planes when compared with intact specimens.

Concurrent fixation to C1 as described by Goel and Laheri, though offering some potential advantages, particularly with regard to the control of sagittal plane rotation, has not been shown to be biomechanically advantageous in pure moment testing. Although technically possible in many instances, the addition of C1 lateral mass screws does not appear to significantly reduce ROM in the short term, except perhaps when constructs are anchored by C2 lamina screws.

The choice of longitudinal element does appear to have an effect on construct stiffness and ROM reduction. In addition to allowing greater flexibility with regard to screw placement and a wide variety of fixation options (screws, hooks, and cable anchors), screw-rod constructs have been shown to yield lower ROM, higher stiffness, and higher failure load than cable- or lateral plate–based constructs. It also appears that rod diameter is an important parameter, with Anderson et al. reporting that ROM reduction in modern rod-based occipitocervical instrumentation constructs is influenced by rod diameter, with larger-diameter rods resulting in the greatest degree of reduction.

Finally, when occipitocervical constructs are extended caudally to the subaxial or thoracic spine, Cheng et al. showed that ROM in lateral bending and axial rotation is less with constructs employing fixation at each vertebra compared with those skipping levels.

Alignment

Alignment after occipitocervical stabilization merits careful consideration. Patients undergoing such a procedure will have lost significant mobility of the head and neck. At the extreme, patients fixated in too flexed or too extended a position may be unable to safely maintain the head in a neutral position, with difficulty ambulating or swallowing. Phillips et al. examined the occipitocervical angle (between McRae’s line and the superior end plate of C3) in the neutral, flexed, and extended position in 30 patients without known pathology. Mean values were 44.0, 24.2, and 57.2°, respectively. Using a slightly different technique (between McGregor’s line and the superior end plate of C2), Matsunaga et al. examined the occipitocervical angle in 120 normal volunteers, noting a slightly higher (retroverted) angle in women than in men, as well as a gradual decline with age. In addition, in a series of patients with rheumatoid arthritis who underwent occipitocervical fusion, 80% of those who developed kyphosis or swan neck deformity within 5 years of occipitocervical fusion had fixed and abnormally elevated occipitocervical angles, and 12 of 13 with fixed and abnormally decreased occipitocervical angles developed subaxial subluxation. It was recommended that the occipital angle should be set, using the latter technique, in the range of 0 to 30° intraoperatively.

Conclusion

This chapter outlined the core principles relating to occipitocervical instrumentation, with an emphasis on how specific knowledge of the regional anatomy, biomechanics, and kinematics may be used to enhance outcome. There is no optimal instrumentation construct for all patients, as this is precluded by variations in anatomy and pathology. The surgeon’s challenge, therefore, is to intelligently choose from the available fixation options to build the most secure construct for any given patient.

References


Radiological investigations of various anomalies of the craniovertebral junction (CVJ) were previously based on the assessment and measurement of several different lines and angles, drawn on plain roentgenograms (craniometry). With the availability of cross-sectional imaging techniques like computed tomography (CT) and magnetic resonance imaging (MRI), assessment of pathologies of the CVJ and their effects on the cervicomedullary neural structures has become more refined.

### Magnetic Resonance Imaging of the Craniovertebral Junction

MRI, with its multiplanar capabilities and high soft tissue contrast resolution, has become the mainstay in the radiological evaluation of the CVJ. It is especially useful because it reveals the cord, soft tissue, ligamentous, and vascular anatomy in detail. Sagittal T1-weighted MR images are of greatest value in the assessment of the CVJ (Fig. 8.1).

MRI must be done in all patients presenting with neurological symptoms. Signal abnormalities due to compressive effects on the cervicomedullary junction, impingement on the lower cranial nerves, and compromised adjacent vertebral arteries can be assessed using MRI. Dynamic MRI also can detect cases of cord compression that are not seen in the neutral position and thus is diagnostic in all cases of mobile atlantoaxial instability. Hypoperfusion in the cerebellum due to vertebrobasilar insufficiency in CVJ anomalies has been documented, and MR angiography can help assess the site and nature of the occlusion.

### Computed Tomography of the Craniovertebral Junction

CT can be used to assess structural bony abnormalities, differentiate congenital clefts from fractures, and as an adjunct to MRI in assessing arthropathies. Axial high-resolution, thin-section CT shows the bony anatomy in detail. Sagittal and coronal images (Fig. 8.2), as well as three-dimensional reconstructions, are useful in assessing the relations of bony structures with respect to each.

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**Fig. 8.1a, b** Sagittal T1-weighted magnetic resonance imaging (MRI) (a) and line diagram of normal anatomical landmarks (b). 1 Nasion, 2 tuberculum, 3 basion, 4 opisthion, 5 posterior border of the hard palate, 6 odontoid tip, and 7 anterior arch of the atlas.
other. CT myelography with the use of intrathecal watersoluble contrast medium to delineate the dural sheath is not often indicated except in conditions where there is a contraindication for the use of MRI.

### Craniometry: The Essential Lines and Angles

Various lines and angles can be used to assess the relationship of bony components of the CVJ to one another. Some of the more useful lines employed in the measurement of the CVJ are outlined here.

#### Chamberlain Line

This line joins the posterior margin of the hard palate to the opisthion (the posterior lip of the foramen magnum) on a lateral radiograph of the skull. The anterior arch of the atlas vertebra lies below this line. In normal individuals, the tip of the dens should be no more than 5 mm above this line (Fig. 8.3a,b).

#### McGregor Line

The McGregor line, which joins the posterior margin of the hard palate to the undersurface of the occiput, can be...
Fig. 8.3c, d  Sagittal T1-weighted MRI (c) and line diagram of the Wackenheim clivus baseline (d). 1 Normally, this line is tangential to the posterior margin of the dens or intersects the posterior one third of the dens. 2 Clivus canal angle. In normal individuals, this angle is > 150°

obtained from a lateral radiograph. In normal individuals, the anterior arch of the atlas vertebra lies below the line, and the tip of the dens should be no more than 7 mm above the line. The mean position of the odontoid peg when assessed on sagittal MR images has been found to lie 1.2 mm below the Chamberlain line and 0.9 mm below the McGregor line.4

■ Welcher Basal Angle

The Welcher basal angle is formed at the junction of the nasion–tuberculum and the tuberculum–basion tangents. The basion is the anterior lip of the foramen magnum. The angle is normally less than 140°. In platybasia, the angle measures greater than 140° (Fig. 8.3a,b).

■ Wackenheim Clivus Canal Line

The Wackenheim clivus canal line is created by extrapolating the line of the posterior surface of the clivus (Fig. 8.3c,d). Normally, this line is tangential to the posterior margin of the dens or intersects the posterior one third of the dens.

■ Clivus Canal Angle

The clivus canal angle is formed at the junction of the line created by extrapolating the line of the posterior surface of the clivus (Wackenheim clivus canal line) and the posterior vertebral body line (Fig. 8.3c,d). In normal individuals, this angle is greater than 150°.

■ Atlanto-occipital Joint Axis Angle (Schmidt Angle)

This angle is measured in the anteroposterior view or on a coronal CT/MRI. It is formed at the junction of the lines traversing the atlanto-occipital joints. An angle measuring less than 124° is seen in occipital condyle hypoplasia (Fig. 8.4).

■ Digastric Line (Fischgold Line)

This line is measured on frontal projections and joins the origins of the digastric muscles at the medial base of the mastoids. The tip of the dens lies below the digastric line (Fig. 8.4).

■ Bimastoid Line

The bimastoid line lies caudad to the digastric line and joins the tips of the mastoid processes. Normally, the tip of the dens lies up to 10 mm above this line.

■ Klaus Height Index

The Klaus height index is determined by drawing a Twining line from the internal occipital protuberance to the tuberculum sellae as a baseline. From this a perpendicular line is drawn to the tip of the dens. This index measures 40 mm. If the index is 34 mm or less, a diagnosis of basi-
lar invagination is made. Goel et al. measured the Klaus height index on MRI and found it to be much more accurate than the conventional measurements based on plain x-rays. The tentorium could be clearly identified on MRI and the distance of the tip of the odontoid from the line of the tentorium indicated the height of the posterior cranial fossa.

**Modified Klaus Height Index**

This measures the height of the posterior fossa. The distance between the Twining line and the plane of the foramen magnum is measured. Normally, this value is at least 30 mm.

**McRae Line**

The McRae line is represented drawn from the basion to the opisthion. Normally, it measures 40 mm.

**Spinolamellar Line**

The spinolamellar line is drawn from the interoccipital ridge above and downward along the fused spinous process portions of C2–C3 below. This curvilinear line should intersect the fused posterior arch of the atlas. If the atlas lies anterior to this line, posterior compression of the spinal cord is usually present.

**Foramen Magnum–Basion Angle**

This angle lies between a line connecting the posterior arm of the plane of the foramen magnum and the point at which it crosses the basion with an anterior arm to the nasion. Normally, it is 163 ± 3.4°. It is abnormal if the value is more than 180°.

**Francesconi Invagination Index**

The Francesconi invagination index is measured by subtracting the occipitoforaminal distance from the occipitobasal distance. The occipitoforaminal distance is measured by drawing a perpendicular from the Twining line to the plane of the foramen magnum. The occipitobasal distance is measured by drawing a perpendicular from the Twining line to the McGregor line. Normally, the Francesconi invagination index is 3 to 10 mm. In basilar invagination, this distance is more than 10 mm.

**Goel’s Omega Angle**

A line is drawn along the hard palate. Another line drawn parallel to this passes through the center of the base of the axis. A third line is drawn from the center of the base of the axis along the tip of the odontoid process. The angle between the second and third lines is the modified omega angle (Fig. 8.5).

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Fig. 8.4a, b Coronal CT bone algorithm (a) and line diagram of the atlanto-occipital joint axis angle (Schmidt angle) (b). 1 An angle measuring < 124° is seen in occipital condyle hypoplasia. 2 The tip of the dens lies below the digastric line.

Fig. 8.5 A line is drawn along the hard palate. Another drawn parallel to this traverses along the midpoint of the base of the C2 vertebra. The angle of the odontoid process on this line is the modified omega angle.
Distance between the Odontoid Tip and the Pontomedullary Junction

The distance of the tip of the odontoid from the pontomedullary junction, as measured on MRI, was observed by Goel et al. to be a useful index in defining the reduction of the posterior cranial fossa bone size.

Kulkarni and Goel’s Vertical Atlantoaxial Instability Index

The vertical atlantoaxial instability index, as described by Goel and Kulkarni, measures the vertical relationship of the atlas and axis (Fig. 8.6). A horizontal line is drawn through the lower end plate of the axis. A second line is drawn parallel to this and tangential to the lower border of the anterior arch of the atlas. A third line is drawn parallel to these lines and tangential to the superior margin of the dens. The shortest distance between the first two lines (x) is divided by the shortest distance between the first and third lines (y). Depending on the severity, the vertical instability is graded from 1 (mild) to 3 (severe).

Anomalies of the Craniovertebral Junction

Congenital anomalies (Table 8.1) and acquired conditions (Table 8.2) affecting the CVJ, including genetic and developmental ones, have been tabulated.

Congenital Anomalies Affecting the Craniovertebral Junction

Congenital anomalies of the CVJ may occur separately, or they may be associated with conditions such as Down syndrome and achondroplasia.

Anomalies of the Occiput

Most anomalies of the occiput are associated with a decrease in the height of the skull base.

Platybasia

The kyphotic notch in the base of the skull is of interest to anatomists, anthropologists, and radiologists. Platybasia refers to a decrease in the basal kyphosis or flattening of the base of the skull. Objectively, platybasia is diagnosed when the Welcher basal angle exceeds 140°. Platybasia usually occurs in conjunction with basilar invagination (Fig. 8.7). The two are not synonymous, however, and it may occur in isolation, in which case there are no neurological signs or symptoms.

Basilar Invagination and Basilar Impression

These are occipital dysplasias with upward displacement of the margins of the foramen magnum into the posterior fossa. The medial rims curve upward, whereas the

Table 8.1 Congenital anomalies affecting the craniovertebral junction

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<th>Anomalies of the Occiput</th>
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more lateral or paracondylar occipital bones curve downward. There is a reduced posterior fossa volume with an irregularly shaped foramen magnum, a short vertical clivus, and a high odontoid position. Basilar invagination (Fig. 8.7) is a primary growth disturbance with dissociated development of the skull and premature fusion of the sutures leading to occipital hypoplasia. It can also be defined as prolapse of the cervical spine into the base of the skull, as suggested by von Torklus and Gehle. Syringohydromyelia and Chiari I malformations, as well as atlantoaxial dislocations (Fig. 8.8), may be associated with basilar invagination. Basilar impression is a secondary or acquired condition resulting from softening of the occipital bone. It is seen in conditions such as rickets, osteomalacia, and Paget disease and results in an actual “impression” of the skull base. Basilar impression may result in severe compression of the brainstem.

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<th>Table 8.2 Acquired conditions affecting the craniovertebral junction</th>
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<td><strong>Inflammatory, Infectious, and Degenerative</strong></td>
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<tr>
<td>Psoriasis</td>
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<td>CPPD disease</td>
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<tr>
<td>Tuberculosis</td>
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<td>Osteoarthritis</td>
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<td>Pseudotumor</td>
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Abbreviation: CPPD, calcium pyrophosphate deposition.

Occipital Condyle Hypoplasia

In occipital condyle hypoplasia, the occipital condyles are flattened (Fig. 8.9). Thus, the height of the skull base is reduced, resulting in disruption of the Chamberlain line (basilar invagination) and an abnormally increased atlanto-occipital joint axis angle.

The condition is commonly bilateral, but occasionally it may be unilateral. Differences in the height of the occipital condyles are compensated for by a lateral deviation of the cervical spine, the head remaining therefore in the midline. Clinically, condylar hypoplasia limits or may even abolish movement at the atlanto-occipital joint and may occasionally lead to compression of the vertebral artery as the result of excessive posterior gliding of the occiput in relation to the atlas.

Fig. 8.7a, b Coronal CT showing basilar invagination (a). The tip of the dens lies above the bimastoid line. Sagittal CT shows the Wackenheim clivus baseline intersecting the anterior one third of the dens (abnormal) (b). Associated platybasia is evident. The Welcher basal angle is >140°.
Fig. 8.9 Coronal CT showing occipital condyle hypoplasia. Flat hypoplastic occipital condyles are fused with lateral masses of C1 bilaterally.

Fig. 8.8a–c Sagittal T1-weighted MRI in flexion (a) and extension (b) showing an atlantoaxial dislocation. Note the widened predental space (seen better on the flexion image). Associated basilar invagination with occipitalization of C1 can also be seen. Three-dimensional CT. Note the wide atlantodental space in the atlantoaxial dislocation (c).
Basioccipital Hypoplasia

Basioccipital hypoplasia may be mild or severe, depending on how many of the four occipital sclerotomes have not formed. The hypoplasia decreases the length of the clivus; hence, the term short clivus. The Chamberlain line is violated, and associated basilar invagination is common.

Chiari Malformations

Chiari malformations comprise brain changes characterized by downward displacement or elongation of the brainstem and cerebellar tonsils through the foramen magnum. Hydrocephalus is variably present and mild. There are two main presentations.

A type I malformation usually presents in adulthood with headache and neck pain. Imaging shows tonsillar herniation, mild hydrocephalus, and variable syringomyelia (Fig. 8.10).

A type II malformation presents in a more dramatic manner, with stridor, feeding problems, and apnea in infancy and early childhood and nystagmus and cranial nerve palsies in the older child.

A type III Chiari malformation is rare. It was first reported by Chiari as a single case of cervical spina bifida combined with multiple spinal and brainstem anomalies. Plain radiographs are not helpful in the diagnosis, but they are capable of showing associated skeletal malformations. MRI currently is the key to making a definitive diagnosis. Low-lying, triangular cerebellar tonsils and elongation and kinking of the fourth ventricle with a sharp clivoaxial angle are classic findings. Syringomyelia is easily demonstrated by MRI as a spinal cord cavitation. The cavity may be focal, usually cervical or cervicodorsal, or a holocord.

Fig. 8.10a–c  (a) Sagittal T1-weighted MRI revealing an Arnold-Chiari malformation. Herniation of cerebellar tonsils through the foramen magnum can be seen. An associated syrinx is noted within the cervical cord. (b) Conventional myelogram and (c) axial CT myelogram. The dye is obstructed and displaced due to herniated cerebellar tonsils.
Anomalies of the Atlas

Assimilation of the Atlas

Assimilation of the atlas, also known as occipitalization of the atlas, occipitocervical synostosis, or occipitocervical fusion, refers to congenital fusion of the atlas to the occiput (Fig. 8.11). The fusion ranges from total occipital incorporation of the atlas (Fig. 8.11a) to a partial bony or fibrous atlanto-occipital band. The most common type encountered is fusion of the anterior arch of the atlas to the anterior margin of the foramen magnum (Fig. 8.11b). In this scenario, the atlanto-occipital joint is nonfunctioning, and flexion occurs at the atlantoaxial joint. Often, the posterior arch of the atlas is absent. In complete assimilation, midsagittal MR images reveal the characteristic comma shape of the anterior margin of the foramen magnum, or basion (Fig. 8.11d).

Occipital Vertebrae

The occipital bone is formed by the union of four or five somites, which normally fuse together to encircle the foramen magnum. The last occipital somite, or proatlas, may fail to incorporate itself fully into the
occiput, resulting in an occipital or proatlas vertebra. Manifestations of occipital vertebrae include the third condyle, paramastoid process, epitransverse process, and various occipital ossicles.\textsuperscript{20,21} The \textit{third condyle} is the most common form of occipital vertebra. An anterior midline bony process located between the two occipital condyles and contiguous with the anterior foramen magnum, the third condyle extends a variable distance caudally. It is generally in the midline, but it may be paramedian. The third condyle may form an articulation with the apex of the odontoid process or anterior arch of the atlas. It is a regular feature in lower animals. There is an increased incidence of associated \textit{os odontoideum}.$^{22}$ The third condyle may be visualized on plain radiographs, although thin-section CT is the best technique for demonstration of this anomaly. MRI is indicated in the rare instances where there are symptoms of brainstem complications.

\section*{Paracondylar, Paramastoid, and Epitransverse Processes}

At C1, a transverse process may enlarge superiorly at its distal end and either fuse with the occiput or have an accessory joint with the occiput. This enlargement is termed an \textit{epitransverse process}. If a process arises from the occiput near a mastoid condyle and fuses or has an accessory joint with the transverse process, the anomaly is called a \textit{paramastoid} or \textit{paracondylar process}. Either condition may limit the range of motion of the upper cervical spine and may be a contraindication for cervical manipulation in the region.

\section*{Posterior Arch Anomalies}

Total or partial aplasia of the posterior arch of the atlas is rare, and when it occurs in isolation, it is usually asymptomatic. Clefts of the arch of the atlas are more commonly encountered. Midline posterior arch rachischisis is more common than lateral clefts. On an open-mouth odontoid view, posterior arch rachischisis may be superimposed on lateral clefts. On an open-mouth odontoid view, posterior arch rachischisis may be superimposed on the axis, simulating a fracture. Transverse thin-section CT can resolve the problem. Because the posterior arch of the atlas lacks a true spinous process, the term \textit{spina bifida of the atlas} is incorrect, and the term \textit{posterior arch rachischisis} is encouraged.$^{16}$

\section*{Ponticles of the Atlas}

The posterior ponticle represents ossification of the atlanto-occipital membrane adjacent to the vertebral artery. It is an archlike structure superior to the posterior arch of C1. It is usually bilateral but may be found unilaterally. If it is incompletely ossified, it is referred to as a \textit{partial posterior ponticle}. The “hole” formed by this variant, the odontoid process, houses the vertebral artery. It is thought that the posterior ponticle produces vertebrobasilar insufficiency symptoms; therefore, proper orthopedic testing is required before cervical manipulation.

\section*{Anomalies of the Axis}

Most anomalies of the axis are confined to the odontoid process.

\subsection*{Os Odontoideum}

The term \textit{os odontoideum} refers to an independent osseous structure lying cephalad to the body of the axis in the location of the odontoid process$^{24}$ (Fig. 8.12a,b). This finding may cause significant upper cervical instability and is an absolute contraindication to cervical spine manipulation. The degree of instability must be assessed by comparison of flexion and extension studies.$^{25}$

In the setting of acute injury to the cervical spine, it is important to differentiate between an acute fracture of the base of the dens (Fig. 8.12c) and an \textit{os odontoideum}. This distinction must be made because treatment differs for each condition. Acute dens fractures are potentially unstable and must be immediately immobilized to prevent neurological injury. Patients with \textit{os odontoideum} may also have neurological instability; however, they do not need immediate treatment if neurological signs and symptoms are absent. Hypertrophy of the anterior arch of the atlas, which occurs
in os odontoideum, is a reliable method to distinguish this anomaly from fracture of the dens in an acute setting. The hypertrophy is seen as enlargement of the arch, increased cortical thickness, and a convex posterior border. The hypertrophy is presumably a reaction to chronic stress at an unstable atlantoaxial junction. Also, in the presence of os odontoideum, the body of the axis has a well-corticated convex upper margin, whereas in an acute fracture of the dens, the upper margin of the body of the axis is sharp and uncorticated.

Many authors feel that a significant fraction of os odontoideum may actually be old, ununited dens fractures.

### Acquired Conditions Affecting the Craniovertebral Junction

Acquired conditions affecting the CVJ include inflammatory infectious and degenerative arthropathies. Various tumors and vascular disorders have also been considered.

### Inflammatory Infectious and Degenerative Disorders

Rheumatoid arthritis is the most common arthropathy, the others being psoriasis, CPPD (calcium pyrophosphate deposition) disease, and osteoarthritis. Common infectious diseases include tuberculosis and fungal infections.

#### Rheumatoid Arthritis

Rheumatoid arthritis affecting the occipitoatlantoaxial region manifests in the form of subluxations, odontoid erosions, and apophyseal joint erosions. Subluxations result from ligament laxity because of hyperemia and local synovitis. Anterior atlantoaxial subluxation is the most common and is indicated by an atlantoaxial interval exceeding 2.5 mm. Vertical subluxation (cranial settling) may be associated with atlantoaxial impaction and causes bulbomedullary compression, which is well visualized on MRI. Lateral (rotatory) and posterior subluxations may also occur. Odontoid erosions as a consequence of synovial inflammation in adjacent articulations are best seen on CT. The dens may be reduced to a small osseous spicule (Fig. 8.13a). MRI demonstrates a pannus, which appears as a periodontoid soft tissue mass (Fig. 8.13b). It shows enhancement on contrast images. Functional MRI has been advocated for cord compromise not seen on routine studies, where images in flexion demonstrate the compression.
Psoriasis

Cervical cord changes in psoriasis are similar to those seen in rheumatoid arthritis. Atlantoaxial subluxation and periodontal pannus may cause compression on the cervical spine and are indistinguishable from rheumatoid arthritis on MRI.

CPPD

CPPD (articular chondrocalcinosis or pseudogout) causing periodontoid mass lesions is a distinct clinical disease entity and is probably underdiagnosed. Nodular deposits appearing isointense to neural tissue on T1-weighted images and iso- to hyperintense on T2-weighted images are seen. They exhibit peripheral enhancement on postcontrast scans due to surrounding distended and inflamed synovial tissue. CT scans may show small areas of calcification. Calcification may also be seen within the transverse ligaments on axial CT scans. Diagnosis can be established based on this distinctive radiological appearance. Atlantoaxial subluxations similar to those seen in rheumatoid arthritis may result. The absence of joint erosions differentiates CPPD from rheumatoid arthritis.

Fig. 8.13a–c T1-weighted sagittal MRIs in flexion (a) and extension (b), and T1-weighted coronal MRI showing rheumatoid arthritis (c). The odontoid is reduced to a small osseous spicule due to tip erosions. An atlantoaxial dislocation is evident on flexion and extension images. A periodontal pannus is seen. This would exhibit darkening on T2-weighted images. An active pannus shows enhancement upon injection of intravenous gadolinium.
Tuberculosis

Tuberculosis of the CVJ is a rare entity, accounting for only 0.3 to 1.0% of tuberculous spondylitis. Organisms usually spread retrograde from a retropharyngeal abscess to the craniovertebral joints. Changes seen on MRI may be due to granulation tissue, abscess, or chronic osteomyelitis. There is a destruction of bone and ligaments leading to occipitocervical or atlantoaxial instability. Signal changes within the cord and intracanalicular extension of a tubercular abscess may also be seen on MRI.

Osteoarthritis

Degenerative changes at the cervico-occipital junction are well visualized on thin-section CT. Odontoid osteophytes, as well as calcification of the transverse ligament, have been seen. Synovial cysts may be seen that compress the cord.

Pseudotumor

Periodontoid extradural mass lesions have been detected in elderly patients presenting with progressive neurological deterioration. These have been confused with tumors, especially meningiomas. Histologically, however, they comprise acellular degenerative fibrocartilaginous material. It is thought that they result from repeated degenerative posterior longitudinal or transverse ligament damage and attempted repair. Edema, fibrocartilaginous metaplasia, and fibrovascular ingrowth during repair have been implicated in the formation of these progressively growing masses. They appear as smooth homogeneous hypo- to isoattenuating lesions on T1-weighted and hypoattenuating on T2-weighted images with none or very subtle contrast enhancement.

Tumors

Tumors preferentially localized to the CVJ include chordomas, meningiomas, and neurofibromas. Metastases are more common than primary tumors. Other intramedullary tumors encountered are ependymomas, astrocytomas, and hemangioblastomas. Malignant neoplasms include chondrosarcomas and nasopharyngeal carcinomas.

Metastases

Metastases, especially from lung, breast, and prostate carcinomas, involve the CVJ. Primary lesions from the colon, uterus, bladder, and kidney need to be considered when CVJ metastases are encountered. The loss of normal marrow signal on T1-weighted images is the hallmark. Soft tissue (Fig. 8.15) and epidural extension is seen on CT and MRI. Contrast imaging clearly delineates the extent of involvement.

Chordoma

Chordoma is the most common benign tumor seen at the CVJ. It appears as a well-defined enhancing soft tissue density lesion with lobulated margins arising from the clivus. Calcification and adjacent bone destruction are best seen on CT. A chordoma appears hypointense on T1-weighted and hyperintense on T2-weighted images. MRI clearly depicts extension of the lesion into surrounding structures. The loss of normal clival marrow density indicates invasion.
Meningiomas

Meningiomas in the CVJ arise from the leptomeninges at the clivus, the foramen magnum, or the anterior spinal canal. Symptoms are due to cord or lower cranial nerve compression. Meningiomas appear well defined and iso- to hyperdense on plain CT scans; some of them show calci- fication. MRI shows an extra-axial mass, hypointense on T1-weighted and hyperintense on T2-weighted images. Homogeneous enhancement with a contiguous enhancing “dural tail” is seen (Fig. 8.16).

Nerve Sheath Tumors

Schwannomas and neurofibromas at the CVJ enlarge the associated foramina and may cause mass effects on the cervicomedullary junction or lower cranial nerves. These are isointense on T1-weighted images, hyper- intense on T2-weighted images, and exhibit intense enhancement.

Chondrosarcoma

Skull base chondrosarcomas may occur as primary les- sions or may complicate chondromas or enchondromas.

Nasopharyngeal Carcinoma

Nasopharyngeal carcinoma commonly spreads by direct ex- tension and causes skull base and CVJ invasion. Replacement of normal fatty marrow on T1-weighted images is charac- teristic. Nasopharyngeal carcinomas are isointense on T1- weighted images and hyperintense on T2-weighted images and enhance on administration of contrast.

Other Conditions

Vascular Lesions

Vascular aneurysms at the CVJ may be confused with tumors. Vessels, however, show a signal void on MRI due to the effect of flowing blood. Dynamic contrast-enhanced MRI is useful in delineating vascular malformations.
Atherosclerotic changes within vertebral arteries at the CVJ are best seen on MRI.

- **Amyloid**

Cervical affection in amyloidosis is a feature of dialysis-associated β₂-microglobulin amyloid. Amyloid deposits in the disks, paravertebral ligaments, and synovial tissues, with a predilection for the craniocervical junction. A destructive spondyloarthropathy is seen. Amyloid deposits and the resultant lytic lesions exhibit a predominantly low signal on all sequences, which helps differentiate them from inflammatory, infectious, and neoplastic lesions. Periodontal soft tissue causes atlantoaxial erosions and destruction.

- **Down Syndrome**

Atlantoaxial subluxation due to transverse atlantal ligament laxity is a feature seen in Down syndrome. A predental interval of 4 to 5 mm in children up to 9 years and 2 to 3 mm in older children is considered the upper limit of normal. Other CVJ anomalies associated with Down syndrome are atlanto-occipital instability, os odontoideum, odontoid hypoplasia, basioccipital hypoplasia with invagination, rotatory atlantoaxial subluxation, and posterior atlas arch hypoplasias. MRI may reveal increased signal on T2-weighted images within the cord, a sequela of chronic instability.

- **Achondroplasia**

Achondroplasia is an autosomal dominant congenital skeletal dysplasia characterized by a decreased rate of enchondral bone formation. Neurological and respiratory symptoms due to stenosis at the foramen magnum are seen.

- **Klippel-Feil Syndrome**

Block vertebrae are characteristic of Klippel-Feil syndrome. Along with these, CVJ anomalies, including basilar invagination, odontoid hypoplasia, platybasia, Chiari I malformation, and atlanto-occipital assimilation, may be seen.
■ Paget Disease

Paget disease, or osteitis deformans, is characterized by abnormal bone remodeling secondary to bone replacement by soft weak connective tissue. This results in basilar impression with compression of the posterior fossa and cervicomedullary junction, as well as a platybasia-like appearance due to remodeling of the occiput.

■ Osteogenesis Imperfecta

Osteogenesis imperfecta is an inherited connective tissue disorder with a defect in collagen type 1 metabolism. Neurological manifestations are because of brainstem compression and hydrocephalus resulting from basilar impression and platybasia.

References

34. Crockard HA, Sett P, Geddes JF, Stevens JM, Kendall BE, Pringle JA. Damaged ligaments at the craniovertebral junction presenting as an extradural tumour: a differential


Atlantoaxial Dislocation

Chapter 9
Congenital and Developmental Anomalies of the Craniovertebral Junction

Chapter 10
Atlantoaxial Fixation Using Lateral Mass Plate and Screws

Chapter 11
Atlantoaxial Transarticular Fixation Techniques

Chapter 12
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Chapter 15
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Chapter 18
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Chapter 22
Techniques of Occipitocervical Fixation
The congenital and developmental bony anomalies and abnormalities that affect the craniovertebral complex may result in neural compression, vascular compromise, and abnormal cerebrospinal fluid dynamics. The basis for understanding the osseous abnormalities in this region is derived from knowledge of the embryology, developmental anatomy, and biomechanics of the craniovertebral junction (CVJ). This chapter discusses the embryology and normal development, as well as the abnormal developmental pathology, of the CVJ and its implications.

### Classification of Craniovertebral Junction Abnormalities

A wide spectrum of congenital, developmental, and acquired abnormalities exist at the CVJ and may occur singularly or in multiples in the same individual. The pathology of these abnormalities is extensive. An attempt at a working classification has been provided in Table 9.1, but it must be appreciated that there are overlapping causes within this classification.

#### Embryology and Development at the Craniovertebral Junction

The bony cranial base is developed by a process of enchondral ossification, in which a cartilaginous framework is first developed and subsequently resorbed with further deposition of bone caused by distorting forces, such as brain development and the development of the eye. The cranial base, as well as the clivus, elongates by sutural growth at the sphenoccipital and spheno-petrosal synchondrosis. There is further sutural growth along the lateral portion of the cranial base. The facial

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**Table 9.1 Classification of craniovertebral junction (CVJ) abnormalities**

<table>
<thead>
<tr>
<th>I. Congenital anomalies and malformations of the CVJ</th>
<th>II. Developmental and acquired abnormalities of the CVJ</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>I.A Malformations of the occipital bone</strong></td>
<td><strong>II.A Abnormalities at the foramen magnum</strong></td>
</tr>
<tr>
<td>1. Manifestations of the occipital vertebra</td>
<td>1. Secondary basilar invagination (e.g., Paget disease, osteomalacia, rheumatoid cranial settling, renal-resistant rickets)</td>
</tr>
<tr>
<td>a. Clivus segmentations</td>
<td>2. Foraminal stenosis (e.g., achondroplasia)</td>
</tr>
<tr>
<td>b. Remnants around the foramen magnum</td>
<td><strong>II.B Atlantoaxial instability</strong></td>
</tr>
<tr>
<td>c. Atlas variants</td>
<td>1. Errors of metabolism (e.g., Morquio syndrome)</td>
</tr>
<tr>
<td>d. Dens segmentation anomalies</td>
<td>2. Down syndrome</td>
</tr>
<tr>
<td>2. Basilar invagination</td>
<td>3. Infections (e.g., Grisel syndrome)</td>
</tr>
<tr>
<td>3. Condylar hypoplasia</td>
<td>4. Inflammatory (e.g., rheumatoid arthritis)</td>
</tr>
<tr>
<td>4. Assimilation of the atlas</td>
<td>5. Traumatic occipitoatlantal and atlantoaxial dislocation; os odontoidideum</td>
</tr>
<tr>
<td><strong>I.B Malformations of the atlas</strong></td>
<td>6. Tumors (e.g., neurofibromatosis, syringomyelia)</td>
</tr>
<tr>
<td>1. Assimilation of the atlas</td>
<td>7. Miscellaneous (e.g., fetal warfarin syndrome, Conradi syndrome)</td>
</tr>
<tr>
<td>2. Atlantoaxial fusion</td>
<td></td>
</tr>
<tr>
<td>3. Aplasia of the atlas arches</td>
<td></td>
</tr>
</tbody>
</table>
bones, on the other hand, and the majority of the cranium develop by intramembranous ossification. This development bypasses the intermediate cartilaginous phase characteristic of the development of the bony cranial base.5,6

There are 42 somites that are formed by the fourth week of gestation. There are 4 occipital somites, 8 cervical, 12 thoracic, 5 lumbar, 5 sacral, and 8 to 10 coccygeal pairs.3,7,8 Each somite differentiates into an outer dermatome and inner myotome and a medial sclervertebral body. These ventromedial bilateral cells migrate toward the midline and surround the notochord. Each sclerotome will develop the fissure of Ebner, which is a central cleft that divides a loose collection of cells cranially from a dense cellular collection caudally. In this development, the cells from the fissure of Ebner migrate toward and encase the notochord to become the precursors of the intervertebral disks.9

The superior half of one sclerotome will unite with the lower half of its neighbor, thus forming the earliest manifestation of the vertebral body. The first four sclerotomes, however, will not follow this course, but instead essentially fuse to form the occipital bone and the posterior portions of the foramen magnum. Simultaneously, vascularization of the occipital bone begins, along with differentiation of the ganglia and vascular tissue. The hypoglossal artery and the first cervical arteries demarcate the caudal occipital segment. The nervous system begins to differentiate at this time.

During the fifth and sixth weeks of gestation, further differentiation of the various parts of the brain and spinal cord takes place. The roof of the fourth ventricle, however, thins out in the midline to form the foramen of Magendie and laterally the foramen of Luschka.4,8 This occurs as an opening in approximately the seventh week when a connection between the fourth ventricle and the subarachnoid space is established.

The occipital sclerotomes correspond to the segmental nerves that group together to form the hypoglossal nerve with a path through the individual foramina within the bone. The first two occipital sclerotomes ultimately form the basiocciput. The third sclerotome is responsible for the exoccipital bone, which forms the jugular tubercles. The key sclerotome in the understanding of CVJ abnormalities is the fourth occipital sclerotome, or the proatlas (Figs. 9.1 and 9.2).

The hypocentrum of the fourth occipital sclerotome forms the anterior tubercle of the clivus. The centrum itself forms the apical cap of the dens and the apical ligament. The neural arch component of the proatlas divides into a rostral ventral segment and a caudal segment. The cruciate ligament and the alar ligaments are condensations of the lateral portion of the proatlas. The ventral portion of the proatlas forms the anterior margin of the foramen magnum, as well as the occipital condyle and the midline third occipital condyle. The caudal division of the neural arch of the proatlas forms the lateral atlantal masses and the superior portion of the posterior arch of the atlas.

The first spinal sclerotome forms the atlas vertebra. It is modified from the remaining spinal vertebra in which the centrum is separated to fuse with the axis body and thus forms the odontoid process (Fig. 9.2). The neural arch of this spinal sclerotome proceeds to form the posteroinferior portion of the atlas arch.4 At times, the hypochordal bow, instead of disappearing, may survive and join with the anterior arch of the atlas to form a variant with an abnormal articulation, which then exists between the inferior clivus, the anterior arch of the atlas, and the apical segment of the odontoid process.2

Recent evidence with computed tomography (CT) evaluation of the atlas has shown that several ossification centers are present in the development of the atlas.2 The lateral masses must be present at birth. A complete ring of the atlas should form by 3 years of age. Abnormal development is observed with skeletal dysplasia, such as
achondroplasia, spondyloepiphyseal dysplasia, and Gold- enhar syndrome, and in genetic abnormalities, such as Down syndrome.

During embryogenesis, the hypocentrum of the second spinal sclerotome disappears. The axis body is formed by the centrum, and the division of the neural arch forms from the facets and the posterior arch of the axis vertebra. Thus, the dens appears from the first sclerotome, whereas the terminal portion of the odontoid process arises from the proatlas. At birth the odontoid process is separated from the body of the axis vertebra by a cartilaginous band that represents a vestigial disk and is later referred to as the neural central synchondrosis. This is crucial in the understanding of the formation of os odontoideum. The neural central synchondrosis lies below the surface of the superior articular facets of the axis and does not represent the anatomical base of the dens. This synchondrosis is present in most children younger than 3 to 4 years of age and disappears by 8 years of age. The odontoid process is observed at birth but does not fuse to the base of the axis. The tip of the odontoid is not ossified at birth and is not observed on lateral radiographs. It is represented by a separate ossification center usually observed at 3 years of age and fuses with the remainder of the dens by 12 years of age.

The posterior fossa expansion occurs because of enchondral resorption, sutural growth, and bone accretion. The growth of the basal aspect of the clivus elongates the foramen magnum, resulting in tonsillar ectopia. Abnormalities that occur with atlas assimilation and basilar invagination, and, more importantly, their natural history. Thus, a wide variety of abnormalities exist. These may occur by themselves or involve both the osseous and neural structures. It appears that insult to these structures may occur between the fourth and seventh week of intrauterine life, resulting in a combination of abnormalities consisting of failure of segmentation, failures of fusion of different components of each bone, hypoplasia, and ankylosis.

There is a high incidence of both anterior and posterior spina bifida of the atlas (C1), as well as os odontoideum in connective tissue diseases, such as mucopolysaccharidosis, Down syndrome, and Morquio syndrome. This results subsequently in atlantoaxial subluxation. It is possible that because of the excessive abnormal head movements in the embryo between the 50th and 53rd day, the process of chondrification becomes impaired, resulting in both anterior and posterior spina bifida at C1. This has been alluded to in the discussion of the development of the atlas. Spinal trauma in children younger than 8 years of age is mainly centered at the CVJ because of the high fulcrum of neck motion. This results in ligamentous injuries more than fractures. However, odontoid fractures in this age group are usually observed as avulsion injuries with separation of the neural central synchondrosis.

### Table 9.1

| Implications of Craniovertebral Abnormalities |

Table 9.1 provides a practical classification of the most frequently encountered congenital craniovertebral anomalies, which are divided into those that are present at birth (congenital) and those that have an abnormal embryology leading to symptomatic abnormalities during early childhood and into adulthood (developmental abnormalities). The immediate relevance of this classification is the understanding of the basis of abnormalities that occur with atlas assimilation and basilar invagination, and, more importantly, their natural history. Thus, a wide variety of abnormalities exist. These may occur by themselves or involve both the osseous and neural structures. It appears that insult to these structures may occur between the fourth and seventh week of intrauterine life, resulting in a combination of abnormalities consisting of failure of segmentation, failures of fusion of different components of each bone, hypoplasia, and ankylosis.
The growth of the posterior fossa, especially the clivus, continues past late adolescence and provides a rationale for the need to continue observing children who have undergone occipitocervical stabilization or a craniovertebral decompression. The downward growth of the brain and the elongation of the posterior fossa and the clivus may re-create a ventral bony abnormality later in life, despite a satisfactory previous ventral decompression at the CVJ performed during the first two decades of life. We have observed this to occur, although infrequently, in our patients with ventral or dorsal posterior fossa decompression.

In considering the epidemiology affecting the CVJ, it is important to keep in mind that infants with Goldenhar syndrome, skeletal dysplasias, and Conradi syndrome will have abnormalities at the CVJ. It should be suspected also in infants who present with torticollis. Diseases such as Down syndrome have a 14 to 20% incidence of atlantoaxial dislocation. Once the stage is set by congenital craniovertebral abnormalities, developmental and acquired phenomena may supervene, producing atlantoaxial instability and subsequently basilar invagination. This is more common in developing countries, where heavy loads are carried on the head from childhood. An erroneous diagnosis of “congenital dislocations” thus appears in the literature. Likewise, upper respiratory infections can cause stiff neck, torticollis, and ligamentous instability and may come to attention later in developing countries than in places where medical attention is readily available. For this reason, it seems that abnormalities of the CVJ are more frequently encountered in populous and less advantaged areas of the globe.

Marin-Padilla and Marin-Padilla in 1981 demonstrated that the basichondrocranium of fetuses with hindbrain malformations, such as Chiari syndrome, is shorter than normal and elevated in relation to the axis of the vertebral column. The shortness of the basichondrocranium of these fetuses is attributed to the underdevelopment of the occipital bone, especially noticeable in the basal component. The defect results in a small posterior fossa, inadequate to contain the developing nervous structures at that region. Unfortunately, this information, though provided in the literature, received very little attention. The elongation of the odontoid process, referred to as a “dolicho-odontoid process,” is explained by the depression at the basiocciput, resulting in a secondary form of basilar impression observed in clinical Chiari I malformations. These changes have been reproduced experimentally in pregnant hamsters by a single dose of vitamin A given early in the morning of the eighth day of gestation, thus producing the typical Chiari malformations as well as the axial skeletal dysraphism.

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**Clinical Presentation**

Clinical presentation of craniovertebral abnormalities is diverse due to the compromise of the brainstem, cervical spinal cord, cranial nerves, cervical roots, and vascular supply. The symptoms of craniovertebral dysfunction may be insidious and may present as false localizing signs. Infrequently, a rapid neurological progression is followed by sudden death. More often than not, there is an antecedent history of minor trauma that sets off a pattern of symptoms and signs that may progress at a galloping pace. The most common neurological symptoms and signs are listed in Table 9.2.

Abnormalities of the CVJ are often associated with an abnormal general physical appearance. The head may be tilted to one side, as seen in patients with rotary luxation of the atlas and the axis or the classic triad of the Klippel-Feil syndrome. The latter consists of an abnormally low hairline posteriorly, limitation of neck motion, and a short neck. Facial asymmetry and webbing of the neck are seen in conjunction with this syndrome. At times scoliosis is present. It is not uncommon to see children with a small fixed stature. There is an increased incidence of craniovertebral abnormalities in diseased states such as spondyloepiphyseal dysplasia, achondroplasia, and related diseases of dwarfism.

The most common symptom is neck pain originating in the suboccipital region, with radiation to the cranial vertex occurring in 85% of patients. There are false localizing signs with abnormalities in this region that are usually motor; these include monoparesis, hemiparesis, paraparesis, and quadriparesis. The central cord syndrome is often seen in children with basilar invagination in whom the myelopathy mimics a lower cervical spinal cord disturbance.

Sensory abnormalities are manifested with neurological deficits related to posterior column dysfunction. Brainstem and cranial nerve deficits cause abnormalities such as dysphagia and sleep apnea. Not uncommonly, internuclear ophthalmoplegia is present, leading to a misdiagnosis of mesencephalic and upper pontine disturbances. Downbeat nystagmus is present in strictly

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**Table 9.2  Signs and symptoms of craniovertebral anomalies (insidious or rapid onset of symptoms and signs)**

| Head tilt |
| Short neck, low hairline, limitation of neck motion |
| Web neck |
| Scoliosis |
| Features of skeletal dysplasias |
| Neck pain and posterior occipital headache |
| Basilar migraine |
| Hand or foot isolated weakness |
| Quadriplegia/paraplegia/monoplegia |
| Sensory abnormalities |
| Nystagmus: usually downbeat and lateral gaze |
| Sleep apnea |
| Repeat aspiration pneumonia, dysphagia |
| Tinnitus and hearing loss |
| Vertigo |

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The list of clinical presentations includes various neurological symptoms and signs associated with craniovertebral abnormalities, emphasizing the importance of early detection and appropriate treatment to prevent further complications.
compressive lesions of the craniovertebral border with or without an associated hindbrain malformation.

Basilar migraine affects ~25% of children with basilar invagination and compression of the medulla. This is usually due to the compression of the vertebrobasilar arterial system. These symptoms will reverse with decompression of the area and stabilization. The excessive mobility of the unstable CVJ can cause repeated trauma to the anterior spinal artery and the perforating vessels of the upper cervical cord and the medulla oblongata, as well as the basilar and vertebral arteries. Unfortunately, this may lead to spasm or occlusion and attendant neurological deficit. Proatlas segmentation abnormalities commonly have this presentation. The most common neurological deficit encountered in affected children is myelopathy. The most common cranial nerve dysfunction seen is hearing loss, occurring in 25% of cases. There is an increased incidence of this finding in the Klippel-Feil syndrome. Unilateral or bilateral paralysis or dysfunction of the soft palate or the pharynx may lead to repeated bouts of aspiration pneumonia, as well as poor feeding and inability to gain weight. Vascular symptoms, such as intermittent attacks of altered consciousness, transient loss of visual fields, confusion, and vertigo, appear in 15 to 25% of patients with congenital or developmental abnormalities at the CVJ. This may be provoked by extension of the head or rotation of the head, as with manipulation of the head and neck.1,21–23

### Neurodiagnostic Imaging

The diagnostic procedure of choice is magnetic resonance imaging (MRI), but plain cervical spine radiographs with lateral flexion and extension views are essential to gain a better understanding of the biomechanics.11 MRI is used to identify the neural abnormalities as well as osseous compression and to recognize any instability, if present. In this examination, flexion and extension dynamic views are obtained in the parasagittal dimension in both T1- and T2-weighted modes. The axial views supplement this. The effects of cervical traction are also documented with MRI to visualize the neural-osseous relationships. MR angiography (MRA) and CT angiography can provide a better understanding of the vasculature, as well as the vascular deformities that may occur with motion of the CVJ.

Three-dimensional CT is extremely useful in defining the abnormalities, as well as in allowing coronal and sagittal sectioning. This author has found CT to be invaluable in recognizing the presence or absence of epiphyseal growth plates in toddlers and young children and in assessing the extent of fusion and segmentation defects, as well as “missing” osseous components. Spondyloysis and spondyloolisthesis are best appreciated with three-dimensional studies.

Each of these imaging modalities provides complementary information to define the craniovertebral deformity. Thus, the factors that influence the specific treatment of CVJ abnormalities are as follows: (1) the reducibility of the bony lesion (i.e., the ability to restore anatomical alignment and relieve compression on the neural structures); (2) the mechanics of compression and the direction of encroachment; (3) the cause of the pathological process, as well as the presence of hindbrain herniation, syrinx, and vascular abnormalities; and (4) the presence of abnormal ossification centers and epiphyseal growth plates.

The primary aim of treatment is to relieve compression at the cervicomedullary junction. Stabilization is paramount in reducible lesions to maintain the neural decompression. Irreducible lesions require decompression at the site where the compression has occurred; these can be divided into the ventral and dorsal compression states. In the former, the operative procedure is ventral decompression through a transpalatal route, the Le Fort drop-down maxillotomy, the lateral extrapharyngeal route, and in severe cases via a mandibular rerouting. In dorsal compression states, a posterolateral decompression is required. If instability is present after decompression, posterior fixation is mandated for stability. It is thus necessary to select the operation or combination of operations for each particular individual based on a clear understanding of the pathophysiology and the functional anatomy.

### Specific Clinical States

#### Proatlas Segmentation Failure or Manifestations of Occipital Vertebrae

Malformations and anomalies of the most caudal of the occipital sclerotomes are caused by proatlas segmentation failures. These abnormalities surround the foramen magnum and usually involve the posterior arch of the atlas.4,24–27 A hindbrain herniation was associated in 33% of individuals in one series.2 At times, the proatlas component of the dens may fail to separate from that portion that forms the basiocciput of the clivus. In this situation, the anterior arch of the atlas comes to rest above the axis.2 At times, the proatlas abnormality is united with the clivus, grossly distorting the cervicomedullary junction ventrally. Variations of this may be observed in the midline, ventrally, laterally, and, at times, dorsally. Thus, one may see paramesial invagination.

Of the 5200 patients in the CVJ database of this author at the University of Iowa, 75 patients were identified with proatlas segmentation abnormalities.2 This is separate from atlas assimilation. Eighty-five percent of such abnormalities present between the first and second decade of life. In our series, the earliest presentation was 3 years of age, and the oldest was 23 years.1 Spastic quadripleasia was the presenting symptom in 80% of patients and lower cranial nerve palsies in 33% of patients. Vascular presentation of vertebrobasilar system dysfunction was observed in 40%. Minor trauma precipitated neurological deficit in 60% of individuals.
Three-dimensional CT provides the best definition of this abnormality and in combination with MRI provides the best understanding. Earlier in our series, pluridirectional tomography was used in the frontal and lateral projections. A hindbrain herniation was present when the posterior fossa volume was reduced, especially by distortion of the vertical height of the posterior fossa (Fig. 9.3). Thus, the treatment of the condition rests with the precise definition of the anatomical and pathological abnormality, relief of neurovascular compression, and prevention of recurrence by stabilization. The surgical approach depends on the manner of encroachment on the neurovascular structures.

Assimilation of the Atlas and the Klippel-Feil Syndrome

Atlas assimilation is defined as failure of segmentation between the fourth occipital sclerotome and the first spinal sclerotome. This anomaly occurs in 0.25% of the population and may be unilateral, segmental, focal, or bilateral. In most instances, it occurs in conjunction with abnormalities such as the Klippel-Feil syndrome. Basilar invagination is the secondary phenomenon. The finding of atlas assimilation was present in more than 500 individuals who were evaluated for craniovertebral abnormalities. A hindbrain herniation occurred in 38% and was caused by reduced posterior fossa volume. Segmentation failures of the second and third cervical vertebrae, in association with atlas assimilation, lead to an excessive load on the atlantoaxial motion segment, which subsequently becomes unstable. This was also found in a series by Baba et al. They reviewed 57 patients with the Klippel-Feil syndrome and found that those patients with C2–C3 segmentation failure had occipitocervical instability and were also found to have odontoid “dysplasia.” Of these 57 patients with the Klippel-Feil syndrome, 21 had progressive neurological symptoms, and 19 underwent an operation. In addition, they felt that degenerative changes at the unfused segments in the cervical spine and a narrow bony canal were high-risk factors in the development of neurological compromise.

Initially, there is reducible atlantoaxial instability present. The sequence of events is that there is pannus formation around the odontoid process with the reducible dislocation. However, as the child grows, there is grooving behind the occipital condyles caused by the upward migration of the axis. This is then followed by a reducible basilar invagination up to approximately the age of 14 to 15 years. This was seen in our initial series published in 1995. As these children age, the lesion becomes an irreducible basilar invagination. During the phase of reducibility and partial reducibility, a prolific granulation...
tissue mass crowns the odontoid process in an attempt to reduce the excursion. This, in turn, compounds the compression of the cervicomedullary junction.

In the irreducible basilar invagination there is an associated horizontally oriented clivus and the abnormal grooving that occurs behind the occipital condyles also pushes up the cranial base resulting in platybasia and a short horizontal clivus. This leads to complete irreducibility of the lesion. Thus, a child who comes for evaluation between 4 and 16 years of age has a better chance of having a reducible atlantoaxial dislocation or a reducible basilar invagination than a fully grown adult. The upward migration of the cranial base and the reduction of the vertical height of the posterior fossa leads to an acquired hindbrain herniation syndrome. Hence, an operative procedure that relies on posterior fossa decompression without addressing the potential for instability below the age of 20 years can lead to unfortunate results (Fig. 9.4).

Torticollis has been a presenting symptom in children with unilateral atlas assimilation. This is critical with head manipulation that occurs during general anesthesia for patients who require placement of drainage tubes in the tympanic membrane and for adenoidectomy. In this situation, the trunk seems to stay in one position, while the head is rotated 90° to either side. As a result, the patient may present with secondary rotary dislocation of the atlas on the axis. It thus behooves the treating physician to be aware of these problems, especially in children with Klippel-Feil syndrome.

The Klippel-Feil syndrome has a classic triad of short neck, webbed neck, and a low hairline with limitation of neck motion. In this syndrome, deafness, high arch palate, facial palsy, and cardiovascular abnormalities are common. Abnormal rib fusions and scoliosis are observed, and 30% of individuals have genitourinary abnormalities. Thus, the recommended treatment for reducible atlantoaxial dislocation or reducible basilar invagination is stabilization. Should a hindbrain herniation be present, posterior fossa decompression should be done with fusion. If irreducible basilar invagination is present, ventral decompression is warranted. Unfortunately, if a posterior procedure for decompression is made in the face of ventral decompression, 30% of these individuals have an unfavorable outcome. This is because the ventral abnormality acts as a “peg” impinging on the cervicomedullary or pontomedullary junction when the patient is positioned for the prone procedure. Additionally, fusion in the flexed position compromises the ability to perform a satisfactory ventral decompression.

It is important to reiterate that the ability to reduce the invagination is age related. The presence of syringohydromyelia with hindbrain herniation and basilar invagination should not sway the neurosurgeon to perform a posterior operative procedure alone. In most instances, the syringohydromyelia disappears once the ventral abnormality has been corrected.

Fig. 9.4a, b
a Midsagittal T2-weighted MRI of the craniocervical region. This 14-year-old boy underwent dorsal posterior fossa decompression for hindbrain herniation (Chiari I) syndrome. He worsened over 6 months with formation of a new cervical cord syrinx and brainstem myelopathy. This reversed after ventral transoral decompression.

b Two-dimensional computed tomography (CT) reconstruction of CVJ. There is a posterior fossa occipital decompression. The ventral abnormality had not been addressed.
**Atlas Abnormalities**

Failure of development and failure of segmentation of the atlas result in abnormal articulation among the clivus, atlas, and odontoid process. Variations may consist of a partial absence of the posterior arch of the atlas, bifid anterior atlas, or posterior atlas, or a combination. A bifid anterior atlas and a posterior atlas result in the two halves of the atlas acting like a complex Jefferson fracture with lateral displacement.\(^{2,3,29-34}\) This condition should be considered grossly pathological if it is present beyond the age of 3 to 4 years and should be addressed.

We have reviewed 160 normal CT scans of the CVJ junction in subjects between the age of birth and 4 years.\(^2\) Our conclusion was that the atlas should be a complete ring by 3 years of age. In cases where the atlas persists as being separate with abnormal dynamics, an operative intervention or fusion should be performed. Neurological deficit was observed in 16 infants with a bifid anterior and posterior arch of the atlas or with the absent anterior and posterior arches except for preservation of the lateral atlantal masses. Placement in a custom-built cervical collar and bracing through 3 years of age have allowed for reformation of the anterior atlas arch and stabilization of the CVJ in 80% of children. It is thought that continued motion in an abnormal situation will prevent the formation of the absent segments and thus lead to further neurological deficits.

A persistent bifid anterior and posterior arch of the atlas beyond the age of 3 or 4 years is observed in skeletal dysplasias, Conradi syndrome, Down syndrome, Goldenhar syndrome, and atlas assimilation. In our series, the presentation was torticollis and plagiocephaly. Neurological dysfunction presented as paresis, apnea, and failure to thrive. The ideal form of imaging is three-dimensional CT and MRI.\(^4\) As previously mentioned, the treatment in our series consisted of bracing until 4 years of age, with repeat three-dimensional CT on a yearly basis. If instability was still present beyond age 4 years, an occipitocervical bony arthrodesis was accomplished.

**Basilar Invagination**

Basilar invagination is a primary developmental defect implying prolapse of the vertebral column into the skull at the base. It is usually associated with occipitalization, assimilation, and blocked vertebra.\(^1\) Hindbrain herniation syndrome occurs in 33% of individuals with basilar invagination.\(^27\)

The term *basilar invagination* is not synonymous with *platybasia*. Similarly, the term *basilar impression* has been used interchangeably. Basilar impression is an acquired form of invagination secondary to softening of the skull, as in diseased states such as osteogenesis imperfecta, rickets, Paget disease, hyperparathyroidism, Hurler syndrome, and acro-osteolysis (Hajdu-Cheney syndrome) (Fig. 9.5). Platybasia refers to an abnormal obtuse basal angle formed by the clivus and the anterior skull base. There are no symptoms or signs attributable to platybasia alone.

There are two types of basilar invagination.\(^12,35,36\) In the ventral variety, there is shortening of the basiocciput so that the clivus is short and horizontally oriented, thus displacing the plane of the foramen magnum in an upward direction compared with the spinal column. In
the paramesial type, condylar hypoplasia may be present so that the clivus becomes dorsally displaced into the posterior fossa but may be of normal length. The occipital hypoplasia may be unilateral and leads to torticollis. The distinction between these two types is not clinically rigid because an admixture may often occur.

Basilar invagination is commonly associated with an odontoid process invaginating into the posterior fossa. The true odontoid process itself is small. However, the body of the axis is elongated. Thus, it appears that there is an “elongated dolicho-odontoid process” on the lateral midsagittal MRI or CT. Of significance is an abnormal clivus–odontoid articulation. This resultant abnormal angle indents into the pontomedullary or cervicomedullary junction in a ventral manner.

The hindbrain herniation associated with basilar invagination occurs in 30 to 33% of individuals. In this situation, the ventral compression of the cervicomedullary junction by the bony abnormality should be relieved by ventral decompression, which must be performed before any posterior surgical procedure. If a posterior procedure is done before relieving the ventral compression, 30% of individuals have an unfavorable outcome.

The ability to reduce the invagination is age related, as stated earlier. The presence of syringohydromyelia with hindbrain herniation and basilar invagination should not sway the neurosurgeon to perform a posterior operative procedure first.

### Aplasia–Hypoplasia of the Dens

In this anomaly, a rudimentary dens may be present or completely absent. Theoretically, the cruciate ligament is incompetent, leading to atlantoaxial instability. This is quite common in patients with atlas assimilation with segmental failure of C2 and C3. In these individuals, the axis body is abnormal, but the dens itself may be hypoplastic. Significant vascular compromise from stretching and distortion of the vertebral arteries has been seen in such lesions. Chronic atlantoaxial dislocation in this situation may result in the formation of granulation tissue at the site of luxation with compression of the cervicomedullary junction.

### Os Odontoideum

Os odontoideum is an independent bone in the dens and is above the axis body. It is not an isolated dens. It exists apart from the hypoplastic dens. Radiographically, the os has smooth borders, and a small dens is always present. It is located in the position of the odontoid process, where it may fuse with the clivus. The gap between the axis and the free ossicle usually extends above the level of the superior facets of the axis, thus making this an acquired rather than a congenital abnormality. The entire complex leads to an incompetence of the cruciate ligament and subsequently to atlantoaxial instability.

In our series of 445 patients symptomatic with os odontoideum, trauma was identified in 175 patients younger than age 4 with a previously recognized normal odontoid process. At times, the odontoid may be associated with an unrecognized fracture in children younger than 5 years with a previously normal odontoid structure, as observed in our series. Os odontoideum may be associated with nontraumatic situations with ligamentous laxity, such as Morquio and Down syndromes. Symptomatic patients were found to have instability in all planes. The biomechanics of os odontoideum are complex. It is different with each individual, in that, in some situations, the flexed position may be best to relieve compression of the cervicomedullary junction,
whereas in others, it is the extended position. Thus, the biomechanics must be carefully studied.

Irreducible dislocation was found to be caused by pannus and the cruciate ligament, which slipped between the os odontoideum to rest in front of the superior portion of the dens and the axis ([Fig. 9.6]). Thus, both the dens and the axis body create the compressive mechanism on the ventral cervicomedullary junction and not the ossicle alone.

In severe chronic dislocations, the os may become fixed with severe basilar invagination. At its worst, os odontoideum has significant implications regarding compression of the cervicomedullary junction. We feel strongly that all patients with recognized instability at the CVJ and associated os odontoideum should undergo stabilization. Furthermore, the fusion will need to be spanned between the occiput, C1, and C2 in 50% of individuals. This can be recognized best at operation. In the remaining 50%, we presently use lateral mass screw-rod fixation or transarticular screw fixation between C2 and C1 ([Fig. 9.7]). Seventy-two individuals in our series presented with worsening following atlantoaxial arthrodesis and required extension of the fusion to the occiput.2

The References


Atlantoaxial Fixation Using Lateral Mass Plate and Screws

Atul Goel and Vinod Laheri

The treatment of patients with atlantoaxial dislocation is a surgical challenge, and achieving a successful outcome for these patients is gratifying. The complications of surgery, however, are potentially lethal.

The techniques of fixation and stabilization of the craniovertebral region evolved during the 20th century as the anatomy and the biomechanical properties were evaluated and understood. Various methods of fixation have been described and used successfully in the treatment of atlantoaxial instability. In 1988, we described an alternative plate and screw technique of fixation of the lateral masses of the atlas and axis vertebrae (Fig. 10.1). We also described simultaneously a technique of occipitocervical fixation, in which the occipital fixation was done with screws, and the cervical end of the fixation was done with either a C2 lateral mass screw alone or in addition to C1 lateral mass screws, as shown in Fig. 10.2. Our technique is gaining wide acceptance and is being used by most large units in which patients with craniovertebral anomalies are treated, although Magerl’s technique, which combines interspinous wiring with transarticular screw fixation and midline fixation techniques, are still used. Several clinical and biomechanical studies have been performed, and the lateral mass plate and screw fixation technique as discussed by us has been uniformly identified to be safer and stronger when compared with other similar fixation techniques. Recently, some authors have modified our technique and have recommended polyaxial screws instead of monoaxial screws, as well as rods instead of a plate.

Our method uses fixation with a plate and direct implantation of screws in the lateral masses of both the atlas and the axis. Fixation of the subaxial spine has been achieved with the use of similar interarticular plates and screws. The lateral masses of the atlas and axis are considerably larger and stronger than any other lateral masses of vertebrae in the rest of the spine and can be used effectively. Firm, multidirectional stabilization is possible with the use of four screws in addition to plates. With our experience now exceeding 650 cases, we are convinced that our technique of atlantoaxial fixation is biomechanically strong, technically easier, safer for the neural structures, and results in remarkable clinical and radiological improvement.

Indications

All cases having atlantoaxial instability can be suitable for operation by this technique. Although pathology and deformities of bones in the region may sometimes make
the operation a little difficult, considering the remarkable stability that it provides, an attempt can always be made to perform this technique. The procedure can be performed safely even in the presence of torticollis and/or assimilation of the atlas. This technique can be used in cases with “fixed” and rotatory dislocation and in group A basilar invagination.

**Contraindications**

There are no specific contraindications to the performance of lateral mass plate and screw fixation, as long as the lateral masses of the atlas and axis are normal. We have observed that such a fixation is possible even in cases where there is lateral mass destruction or erosion or in cases where there is significant osteoporosis. Abnormal course of the vertebral artery within the facet of the axis and in the vicinity of the posterior arch of the atlas may rarely preclude the use of our technique.

**Operative Technique**

**Traction and Positioning**

Cervical traction is given prior to induction of anesthesia, and the weights are progressively increased to approximately one fifth of the total body weight, or 3 to 7 kg. The patient is placed prone with the head end of the table elevated to ~35° (Fig. 10.3). The turning of the patient from the supine position to the operative prone position is done carefully by firmly stabilizing the neck by both hands and under continuous traction. This is because any abnormal movement of the neck under anesthesia, when the neck muscles are relaxed, can critically compromise the cord. Cervical traction stabilizes the head in an optimally reduced extension position and prevents any rotation. The traction also ensures that the weight of the head is directed toward the direction of the traction, and pressure over the face or eyeballs by the headrest is avoided. Although the head is placed on the headrest, it is essentially “floating,” as the traction pulls the head away from the headrest. Elevation of the head end of the table, which acts as a countertraction, helps in reducing venous engorgement in the operative field. The body parts are adequately protected by the use of soft and firm cushioning pads placed on the operating table. Particular attention is given to avoid undue pressure over the penis, testicles, and breasts.

**Incision and Dissection**

The suboccipital region and the upper cervical spine are exposed through an ~8 cm longitudinal midline skin incision centered on the spinous process of the axis. The spinous process of the axis is identified, and the attachment of paraspinal muscles to it is sharply sectioned. The C2 laminae are widely exposed subperiosteally, and the dissection is followed laterally over the pedicles. Use of an operating microscope facilitates the dissection and adds remarkable safety and precision to the entire surgical procedure. Actual vertebral artery exposure is unnecessary either lateral to the pars of the axis or superior to the arch of the atlas.

**Sectioning of the C2 Ganglion**

The C2 ganglion is placed transversely over the atlantoaxial joints. The large ganglion is widely exposed, then sectioned and resected. The ganglion is closely related to the vertebral artery on its lateral aspect, and all dissection in the region must be done under direct vision. On some occasions, the ganglion can even be mobilized superiorly or inferiorly, and sectioning can be avoided. However, sectioning of the ganglion provides a wide panoramic exposure of the lateral masses of the atlas and axis, along
with the atlantoaxial joint region. The consequence of numbness related to the ganglion sectioning is marginal and easily tolerated by the patient (Fig. 10.4).

### Venous Bleeding

Large venous plexuses and sinuses in the region and in the extradural space can result in troublesome bleeding and on some occasions can be a cause of significant difficulty during the surgical procedure. Excessive coagulation in the region can easily be avoided. A relatively quick and sharp dissection in the region is helpful in minimizing exposure in the region and in reducing overall blood loss. Packing of the region with Surgicel and Gelfoam can assist in the control of venous bleeding. The packing is generally required in the extradural space of the spinal cord and in the region lateral to the ganglion. As packing can control the bleeding, it is sometimes helpful to pack the region and change the side of the dissection and return after a period of time. Such a strategy can assist in reducing blood loss.

### Joint Opening, Distraction, and Graft Insertion

The joint capsule is cut sharply, and the articular surfaces of the joints are exposed. The articular surfaces are distracted with the help of varying sizes of osteotomes. The osteotome is introduced into the joint with its flat end and is then turned 90° to effect distraction. The articular end plate cartilage is denuded widely with a microdrill, and pieces of bone harvested from the iliac crest are stuffed into the joint space. The lateral aspect of the lamina and part of the pars of the axis are drilled to make the posterior surface of the lateral mass of the axis relatively flat so that the metal plate can be placed snugly and parallel to facet bones (Fig. 10.5). Drilling also helps in placing the plate with ease, in reducing the size of the plate, and in placing the screw
10 Atlantoaxial Fixation Using Lateral Mass Plate and Screws

Fig. 10.5a–c
a Photo showing the atlas vertebra. Note the thick facets.
b Photo showing the axis vertebra.
c Photo showing the alignment of the atlas and the axis.

more superiorly and almost directly into the lateral mass of the axis.

Hardware Insertion

Metal screws are implanted into the drilled guide holes in the lateral mass of the atlas and axis through a two-holed adequately selected and shaped double-compression stainless steel or titanium plate, ~2 cm in length.

Screw Insertion into the Atlas

First, a screw is placed into the facet of the atlas. It is directed at an angle of ~15° medial to the sagittal plane and 15° superior to the axial plane. The preferred site of screw insertion is at the center of the posterior surface of the lateral mass, 1 to 2 mm above the articular surface. Whenever necessary, careful drilling of the posterior or inferior surface of the lateral aspect of the posterior arch of the atlas in relation to its lateral mass can provide additional space for placement of the plate and screw. The screw may even be implanted from the articular surface of the lateral mass of the atlas. Such a site is useful more frequently in children. We have also placed the screw directly into the posterior arch of the atlas that traversed into the facet. Elevation of the vertebral artery off the arch of the atlas may or may not be required for screw implantation.

Screw Insertion into the Axis

Screw implantation in the axis needs precise direction of insertion, because of the intimacy of the vertebral artery. As discussed in an earlier article on this subject, the pars interarticularis can be divided into nine quadrants (Fig. 10.6).6 The superior and medial compartment can be used for the interarticular technique of screw implantation. The direction of screw implantation must be sharply medial and superior and should be toward the tubercle of the anterior arch of the atlas located in the midline. The medial surface of the pars of the axis is identified before implantation of the screw. The screw is directed at an angle ~25° medial to the sagittal plane and 15° superior to the axial plane. The angle of screw implantation varies, depending on the local anatomy and the size of the bones. The quality of cancellous bone in the lateral masses of the atlas and axis in the proposed trajectory of screw implantation is usually good, providing an excellent purchase of the screw.

Implant Specifications

The devices used to cut and mold the small plates are commercially available. Precut and premolded plates are also available. The plates and screws can be of stainless steel or titanium. The screws used in adult patients are 2.9 mm in diameter and 2.7 mm in diameter in pediatric patients. The length of the required screw is calculated on the basis of the size of the lateral masses observed on

Fig. 10.6 Photo showing the axis vertebra. Nine quadrants have been drawn. The medial and superior quadrants are safest for screw insertion.
preoperative radiological studies. The approximate lengths of the atlas screws in adults are 22–26 mm and in children, 18–22 mm. The screws in the atlas and axis are almost similar in length. The lateral masses of the atlas and axis are firm and cortical in nature and, although preferable, it is not mandatory that the screws engage both the posterior and anterior cortices. If the screw traverses beyond the anterior cortex, it will lie harmlessly in the anteriorly displaced soft tissue. Although injury to the carotid artery and pharyngeal wall is possible in such cases, the chances of such injury are extremely low, and the tissues usually get displaced by the advancing screws.

### Onlay Graft Placement and Closure

All the muscle attachments to the C2–spinous process are sharply cut (Fig. 10.7). Surface drilling of the posterior elements of the atlas and axis is performed to provide a bed for onlay graft placement. Large pieces of corticocancellous bone graft from the iliac bone are then placed in the adequately prepared receptor area of the posterior arch of the atlas and the lamina of the axis. After the wound is closed, cervical traction is removed (Figs. 10.8, 10.9, and 10.10).

Text continues on page 90
Fig. 10.9a–f

a Lateral radiograph with the head in the extension position showing complete reduction of the dislocation.

b Lateral film of the craniocervical junction with the head in the flexion position showing atlantoaxial dislocation.

c Three-dimensional computed tomography (CT) scan showing atlantoaxial dislocation.

d Postoperative CT scan showing the screws in the lateral mass of the atlas and axis.

e Postoperative radiograph showing reduction and fixation using C1 and C2 lateral mass plate and screw fixation.

f Coronal image of the postoperative CT scan showing the screws in the lateral masses of the atlas and axis.
Fig. 10.10 a–e

a Lateral radiograph with the head in the extension position showing complete reduction of the dislocation.

b Lateral film of the craniovertebral junction with the head in the flexion position showing atlantoaxial dislocation.

c Postoperative radiograph showing reduction and fixation using C1 and C2 lateral mass plate and screw fixation.

d Postoperative CT scan showing the reduction of dislocation.

e Coronal image of the postoperative CT scan showing the screws in the lateral masses of the atlas and axis.
Patients are mobilized within a few days of surgery. They wear hard cervical collars for 3 months, and all physical activities related to the neck are restricted for this period.

**Neuronavigation**

Neuronavigation can be used to great advantage in selecting the best site and direction of screw insertion. The appropriate size of the screw, including its thickness and length, can also be suggested by the navigation. With the current technology, C2 pars screw insertion can be suitably guided by navigation equipment (Fig. 10.11).

**Complications and Avoidance**

The most dreaded complication of the procedure is injury to the vertebral artery. Because the direction of the screws is medial and not anterior, the possibility of injury to the vertebral artery is less with our technique than with Magerl’s technique. Consulting appropriate anatomical information of the region in general and in the case in question is the only way to avoid this complication. The vertebral artery can be injured during the process of lateral dissection of the C2 ganglion. The other potential point of injury is during the insertion of the screw in the axis. In the latter situation, to control the bleeding, one has to pack the bleeding bone hole with bone wax. One can then prepare for an alternative site of screw insertion or use another method of atlantoaxial fixation. Screw implantation can be sometimes rapidly completed through the same bleeding hole. Respect and care of all neural and vascular tissues and employment of precise technique are critical to success. In cases where there are smaller tears in the artery, these can be directly sutured. However, continuous venous bleeding in the region can sometimes make this procedure difficult. In cases where vertebral artery laceration has been encountered, the possibility of formation of delayed pseudoaneurysm should be kept in mind. Digital subtraction angiography, identification of the aneurysm, and coil occlusion of the aneurysm or of the artery may be necessary. Sacrifice of the vertebral artery in the craniocervical region was nonconsequential in our 11 cases in which the artery was lacerated during surgery and subsequently was sacrificed.

The resection of the large second cervical ganglion widely exposes the atlantoaxial joint and the posterior surfaces of the atlas and axis. This is an important surgical step in our technique. There is no significant or annoying clinical symptom related to the sectioning of the ganglion. It may be that patients are so satisfied with regard to their limb function that they ignore the anesthesia in the nape of the neck and the occipital scalp area.

**Comparison with Other Techniques**

In our technique, longer purchase of screws in both the atlas and the axis is possible, unlike in Magerl’s technique. The entire length of the screw is introduced into the lateral mass of the atlas and axis, and the implantation procedure can be performed with direct visualization of the lateral mass of the atlas and axis. In Magerl’s interarticular screw fixation technique, only one screw is used, the screw implantation in the atlas is not performed under direct visualization, and only part of the screw engages the lateral masses of the atlas and axis. In our technique, because the screw implantation is not as acutely angled superiorly as
in Magerl’s interarticular technique, the skin incision does not need to be extensive. In addition, there is no need for flexion of the patient’s neck during the procedure to obtain the proper trajectory, and an inadvertent injury to the hypoglossal nerve is thus avoided. The screw in the axis is directed medially away from the course of the vertebral artery. In Magerl’s technique, however, the screw has to be directed more superiorly than medially to engage the lateral mass of the atlas. Most authors who use Magerl’s technique indicate the need for midline fixation of the posterior arch of the atlas and the lamina or spinous process of the axis to provide optimal mechanical stability. Because no midline fixation is necessary, although it is possible, with our technique, occasional problems of exposure and dissection of the posterior arch of the atlas are avoided. The avoidance of the need to introduce wire underneath the arch of the atlas imparts safety to the neural structures. Some of the stabilization methods described earlier in which bone grafts and interlaminar wires or methylmethacrylate are used eventually produce stability but require a prolonged period of immobilization and have a significant failure rate. The biomechanical advantage afforded by our technique is suggested by our successful results. The segmental fixation of the atlantoaxial dislocation preserved the movement of the rest of the cervical spine and occipitoatlantal joint. In addition, it does not prevent the growth and elongation of the neck in children, in contrast to multilevel fixation techniques.

### Advantages of Lateral Mass Fixation

The advantages of the lateral atlantoaxial articular fixation include segmental stabilization at the site and fulcrum of the dislocation. There is a possibility for midline decompression wherever indicated. In rare situations, lateral fixation is the only available procedure when the posterior arch of the atlas is congenitally deficient, cartilaginous, or broken after trauma or sublaminar wiring.

Opening the atlantoaxial joint, denuding the articular cartilage, and stuffing bone chips within the joint provide remarkable stabilization by themselves. These steps in the procedure provide a large ground for bone fusion. Articular arthrodesis was observed to be crucial for the stability of the construct and for successful bone fusion. Additional bone graft is placed over the posterior arch of the atlas and the lamina of the axis after adequately preparing the host bone by decortication, thus providing a wide area for bone fusion. Moreover, the opening of the joint provides an avenue to manipulate the bones and reduce fixed and rotatory dislocations and basilar invagination.

Thus, the principle advantages of our technique include the following:

- The technique provides direct treatment to the fulcrum of the movements located at the atlantoaxial joint.
- The removal of the articular cartilage and the stuffing of bone graft within the joint obstruct all movements, stabilize the region, and provide additional space for bone fusion.
- The fusion is segmental. The problem is atlantoaxial dislocation, and the treatment is atlantoaxial fixation.
- The fixation is firm and rigid, as it involves screw implantation in the strong bones of the facets of the atlas and axis. Such a fixation provides an appropriate environment for bone fusion.
- The method can be used in children, even when other methods are not possible.
- All midline procedures can be additionally performed.
- There is a possibility of manipulating and distracting the facets that can affect reduction of mobile and fixed atlantoaxial dislocation and of basilar invagination.
- The atlantoaxial fixation can be done even in cases where there is assimilation or occipitalization of the atlas.
- As the screw implantation is carried out separately in the facet of the atlas and the pars of the axis, the procedure is safer as regards the vertebral artery.
- The entire procedure is away from the neural structures. The avoidance of introduction of any wire underneath the arch of the atlas and axis adds remarkable safety. Because tightening of wires is not involved, the dangers of incomplete tightening and overtightening are avoided.

### Treatment in Specific Situations

#### Failed Midline Fixation Procedures

Midline fixation techniques using sublaminar wires and metal loops and rings are still widely employed. Different types of fixation material, including bone graft, acrylic material, and other bone substitutes, are used.

The treatment of a failed midline fixation can be a formidable surgical problem. The incomplete bone fusion leads to difficulty in its removal and appropriate exposure. The exposure of a previously treated arch of the atlas and lamina of C2 may be difficult. In such a situation, lateral mass fixation using our technique is a reasonable surgical option. Because the lateral masses are not exposed during the earlier surgery, this region is relatively virgin and can be exposed with relative ease. The feasibility of joint manipulation and distraction even under such conditions makes lateral mass fixation an option.

### Fixation in Children

We have treated over 115 children younger than 12 years using the atlantoaxial fixation technique. The youngest was 3 months of age. There are some special issues of concern in treating children with atlantoaxial dislocation. The internal fixa-

Text continues on page 94
Fig. 10.12a–f

a Preoperative CT scan showing atlantoaxial dislocation.
b CT scan with sagittal cut passing through the lateral masses showing atlantoaxial dislocation.
c Postoperative radiograph following first surgery showing occipitocervical fixation with wire and artificial bone material. The fixation is in the dislocated position.
d CT scan confirms fixation in the dislocated position.
e Postoperative radiograph showing fixation in the reduced position.
f CT scan with a sagittal cut passing through the lateral masses showing plate and screw fixation in the reduced position.
Radiograph (infant 3 months old) with the neck in flexion showing atlantoaxial dislocation.

Radiograph with the neck in extension showing complete reduction of the dislocation.

T1-weighted magnetic resonance (MR) image showing a syrinx in the craniovertebral region.

T2-weighted MR image showing the syrinx.

Postoperative radiograph showing fixation using plates and screws.
tion procedure, for example, has to be biomechanically strong, as children tend not to tolerate external support, such as a collar, traction, or halo brace. Limitation of the range of neck movements for a period of at least 6 to 12 weeks that is necessary for full bone fusion is difficult to achieve by any external arthrodesis in children. The fixation has to be segmental in nature, as any fixation procedure that includes the occipital bone and the subaxial spine not only is biomechanically inferior, but limits the range of neck movements and in the long term hinders the growth and full development of the neck.

Congenital atlantoaxial dislocation is usually associated with a range of complex anomalies. These include the absent, cartilaginous, or bifid nature of the arch of the atlas and the lamina of the axis. Traumatic fracture of the arch of the atlas or the lamina of the axis is relatively rare, but it can lead to significant difficulty for all midline surgical fixation methods. Wire tightening cannot be done with appropriate pressure due to the nature of the bone structures. Over-tightening and fixation in hyperextension of the atlas and overreduction have been reported. Considering the delicate nature and the small size of the bones and neural structures of children, several authors have reported complications related to sublaminar wire introduction under the lamina of the axis, particularly under the arch of the atlas.

All of these concerns in children and the drawbacks with midline fixation procedures are circumvented by the lateral mass plate and screw fixation method. The lateral masses of both the atlas and the axis are formed earlier during embryological development and are significantly large, strong, and well developed, even in children. Although congenital anomalies of the facets have been reported, they are relatively rare.

Wide exposure of the joint is possible after sectioning of the large C2 ganglion. Sectioning of the ganglion can be avoided more frequently in children, as the tissues are elastic and stretchable. Drilling and denuding of the articular cartilage of the facet joint and placement of large pieces of bone graft provide mechanical stability to the region.

On some occasions, venous bleeding can be profuse and challenge the skills of the surgeon. The sudden nature of blood loss can critically affect the hemodynamics in children. Blood loss should be anticipated, however, and the anesthetist should be appropriately warned.

Exposure of the posterior surface of the inferior facet of the atlas for insertion of the screw into the facet of the atlas can sometimes be difficult in small children due to the bone size. In such a situation, the screw is inserted through the articular surface of the facet of the atlas. The direction of the screw insertion into the pars interarticularis of the axis needs to be precise and should be directed medially and superiorly toward the base of the odontoid process.

The success rate (100%) of fusion with our technique speaks of the biomechanical advantage of this procedure. In three patients in our series, lateral mass fixation was possible on only one side, but still a firm bony fusion was achieved. The technique provided firm fixation of the region, and no rigid external arthrodesis was necessary. The children in our series were given a four-poster hard cervical collar, and all activities involving neck and head were restricted. The segmental nature of the fixation proved another advantage. Drilling the articular cartilage and inserting bone graft in the joint at the site of the fulcrum of all the movements in the region added stability. Because the occipitoatlantal and entire subaxial region joints are preserved and are functional using this technique, neck movements are almost full. The technique also allows unrestricted growth of the child’s neck size. Another plus is avoidance of sublaminar wiring, which added to the safety to the procedure. Sectioning of the C2 ganglion, however, was necessary to provide appropriate exposure of the region and allow completion of the procedure under direct vision.

In our series, it was observed that, although all patients had some numbness in the suboccipital region, the area affected progressively decreased. Numbness in the nape of the neck is better tolerated in children. Also, improvement in numbness as a result of functional takeover by adjoining scalp nerves is better and quicker in children. At follow-up, none of the patients complained of any disability related to numbness.

### Double Insurance Atlantoaxial Fixation

In 2007, we presented an alternative to atlantoaxial fixation and joint distraction for the treatment of both atlantoaxial dislocation and basilar invagination, which incorporates the advantages of both the interarticular and transarticular techniques and provides a firm fixation of the region ([Fig. 10.14]). Onlay and interfacetal bone grafts subsequently produced bony fusion.

### Surgical Indications

The technique of atlantoaxial fixation employed in the series was used randomly, and cases were not consecutive. Case selection was primarily based on the local anatomical situation gauged after the exposure of the region and manipulation of the joint in cases of atlantoaxial dislocation with or without the presence of basilar invagination. The size of the pedicle and the location of the facets after their exposure and manipulation determined case selection.

### Operative Technique

Plate and screw fixation of the region is performed by combining the interarticular technique and Magerl’s technique ([Fig. 10.14]). A two-holed stainless steel plate is used measuring ~20 to 25 mm in length. The lateral aspect of the lamina of the axis and the pedicle is drilled using a microdrill to make the area flat so that the plate can be placed flush with the bones. Drilling also reduces the size of the plate and helps in placing the atlas screw more superiorly and almost directly into the lateral mass of the axis.
The screws are 2.6 to 2.8 mm in diameter and measure 20 to 28 mm in length. Axis screws are longer. Screws are first passed through the holes in the plate into the lateral mass of the atlas, and the plate is adjusted over the lateral mass. A screw is then passed in a transarticular fashion, as described by Magerl. The transarticular screw implantation is done keeping the local anatomy in perspective and the facet of the atlas under direct vision. The site and angle of screw implantation in both the atlas and the axis are modified to suit the local anatomical situation. The medial surface of the pars is identified before implantation of the screw. Additional bone graft is placed between the posterior elements of the C1–sub occipital bone complex and C2 after decorticating the host bone area with a burr.

The combined transarticular/interarticular technique provides strong fixation and stabilization of the region and an optimum environment for bony fusion. It allows implantation of both the atlas and axis screws under direct vision and alteration of their trajectory depending on the local anatomical situation. The technique also allows placement of bone pieces and spacers within the joint. Firm, multidirectional stabilization is possible with the use of four screws in addition to plates. The technique is particularly suited for cases with basilar invagination, in which the facet of the atlas is placed more rostrally and has an anterior slant (Figs. 10.15 and 10.16). The orientation of facets makes transarticular fixation easier in such a situation. In the described procedure, the internal fixation appeared to be firm, permitting no movements, and minimal or no external immobilization was found to be necessary.19

Fig. 10.14a, b
a Drawing showing the construct. A metal plate is placed flush to the lateral masses of the atlas and axis after adequately preparing the host area. A screw is passed directly into the facet of C1 through a hole in the plate. A transarticular screw is placed in C2, as described by Magerl. A bone graft with or without a spacer is placed in the articular cavity.
b Posterior view of the drawing.

Fig. 10.15a–c
a CT scan showing an atlantoaxial dislocation in a 45-year-old man.
b Postoperative scan showing reduction of the dislocation.
c Postoperative CT scan showing the fixation. A lateral mass screw is placed directly into the C1 facet, and a transarticular screw is placed in C2.
**Conclusion**

We think that our technique has great potential in the treatment of atlantoaxial dislocation, and if it is learned adequately and performed successfully, the results are extremely gratifying. An understanding of the anatomy of the region in general and of the case in question is the crucial key to success.

**References**


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**Fig. 10.16a–c**

a CT showing fixed atlantoaxial dislocation with basilar invagination and odontoid in a 24-year-old woman.

b Postoperative CT scan showing the C1 screw and the spacer placed in the atlantoaxial joint.

c Medial section of the CT scan showing the transarticular screw at C2 and the direct screw in C1.
The posterior upper cervical spine C1–C2 transarticular screw fixation technique was introduced by Magerl in the 1980s and has been widely published.\textsuperscript{1–4} It was developed because of partially unsatisfactory results with wiring techniques.\textsuperscript{5,6} Magerl’s transarticular posterior technique was conceptually a further development of the lateral transarticular screw fixation, which had been developed in 1971 by Barbour, who used a bilateral neck approach instead.\textsuperscript{7–9} Furthermore, this lateral approach to the C1–C2 joints was anatomically more demanding and more prone to complications than a simple posterior midline approach due to the anatomical dissection involved.

In the 1990s, we developed an anterior approach for transarticular C1–C2 screw fixation, which again represented progress in terms of surgical access, as it avoids posterior paravertebral muscle dissection, a possible source of postoperative neck pain.\textsuperscript{7,10,11} For this anterior transarticular screw fixation, the simple, well-established unilateral anterior approach is chosen in the virtual space between the trachea, esophagus, and long muscles on the medial side and the sternocleidomastoid muscle with the underlying neurovascular bundle on the lateral side. When we first published our data, we realized that others had come to the same idea and published their experiences even earlier in the neurological literature.\textsuperscript{12}

This approach is extremely atraumatic and is recommended in elderly patients (supine position, very little blood loss) and in combination with an anterior odontoid screw fixation or anterior plating of the middle cervical spine.

### Posterior Techniques

#### Surgical Principles

The original transarticular screw fixation as described by Magerl\textsuperscript{1} allows rigid stabilization of the C1–C2 segment in all directions. The screw enters from the posterior aspect of the massa lateralis of C2, passes through the subchondral bone of the superior articular surface of C2, and ideally crosses the joint gap of C1–C2 to enter the subchondral bone of the inferior C1 joint surface. Consequently, the fixation point of the transarticular screws is located exactly in the center of the facet joints of C1–C2, exposing this screw only minimally to flexion, extension, and rotational forces.

This screw is not a compression screw but a positioning screw, thus avoiding compression of the roots within the C1–C2 foramen by narrowing the foramen. The screw is under minimal adverse load, and breakage is extremely rare even when formal fusion is lacking.

#### Biomechanics

There have been several studies investigating the biomechanical behavior of C1–C2 screw fixation in comparison with other fixation systems.\textsuperscript{2,13–22} The C1–C2 transarticular screw with interspinous graft is a standard technique for atlantoaxial fixation\textsuperscript{15} and can be considered equivalent to Goel and Laheri’s technique of screw-plate/rod systems that are anchored in the lateral mass of C1 and C2.\textsuperscript{16} The transarticular fixation of C1–C2 prevents lateral bending and axial rotation better than current posterior cable-secured graft constructs, which are somewhat better in terms of flexion and extension. This is true only when the transarticular screw fixation is not combined with an interspinous graft.\textsuperscript{18} Similar in vitro biomechanical tests for five different C1–C2 constructs confirm the above findings and demonstrate clearly that the Gallie or Brooks-Jenkins cable fixation alone may not be adequate for atlantoaxial arthrodesis. The combination of the Brooks-Jenkins fixation with one transarticular screw is superior to the construct of a Gallie fusion with one transarticular screw fixation.\textsuperscript{19} Richter et al. tested six fixation systems for the atlantoaxial segment and came to the conclusion that simple transarticular screws from C2 to C1 are best for stabilization, lateral bending, and axial rotation.\textsuperscript{20} When a Gallie fusion or claw was used, additional stability was obtained. The authors concluded that the combination of transarticular screw fixation with a Gallie fusion and Goel and Laheri’s technique of lateral mass fixation are the best fixation techniques of all those tested.\textsuperscript{20} They also mentioned that lateral mass screws (Goel and Laheri’s technique) or claw fixation together with isthmic screws is a valid alternative in case transarticular screws are not feasible.\textsuperscript{20}

#### Surgical Technique

##### Surgical Approach

The patient is positioned in the prone position, and the position of C1–C2 is checked using lateral image intensifier control. The alignment of C1–C2 is facilitated by traction...
either with Gardner-Wells tongs/halo or a Mayfield clamp, the latter of which controls translation more easily. The neck is flexed as much as possible to facilitate insertion of the screws, and an image intensifier is used to exclude redislocation in case of a fracture dislocation (Fig. 11.1).

A midline incision is performed from the occiput to the tip of the spinous process of C3. The arch of C1, spinous process, laminae, and inferior articular processes of C2 are exposed subperiosteally. In case of a persistent anterior dislocation of C1 over C2, a reduction is done by pushing on the spinous process of C2 and/or by pulling gently on the posterior arch of C1, either with a towel clamp or with a sublaminar wire or suture sling. Persistent posterior dislocation would require opposing forces.

It is recommended that the surgeon gently expose the inner side of the isthmus and pedicle of C2 and stay medially as close as possible to this landmark. A small dissector is used to expose the cranial surface of the lamina and isthmus of C2 by careful subperiosteal dissection up to the posterior capsule of the atlantoaxial joint (Fig. 11.2a). Medial to the isthmus, the atlantoaxial membrane is visible. The laterally situated vertebral artery is not exposed. It is further recommended to get a computed tomography (CT) scan of the C1–C2 segment before a surgical intervention to rule out an anomalous course of the vertebral artery.

### Standard Technique

Using lateral image intensifier control, a 2.5-mm-long drill bit is inserted in a strictly sagittal direction. The oscillating attachment prevents soft tissues from being wrapped around the drill bit. The entry point of the drill bit is at the lower edge of the caudal articular process of C2 (Fig. 11.2b). The drill bit goes through the isthmus near its posterior and medial surface. It then enters the lateral mass of the atlas close to its posteroinferior edge. Anteriorly, the drill perforates the cortex of the lateral mass of C1. The screw length is measured, and the direction of the screw canal is checked using an image intensifier (Fig. 11.2c).

The 3.5-mm cortex screws are inserted after checking with a 3.5-mm tap across the C1–C2 joint; the anterior cortex of C1 must not be tapped (Fig. 11.2d). Proper caudocephalad drilling may be difficult because the neck muscles and the upper torso prevent the correct placement of the drill bit. Gently pulling the spinous process of C2 cranially with a towel clamp facilitates drilling. It is often necessary to drill through a distal percutaneous stab wound to place the drill in the correct angle of ~60°. Prepping and draping of the upper thoracic spine are necessary to be able to enter the drill bit through a stab incision.23–25

Drilling in a horizontal direction must be avoided because

- At the level of C2, the vertebral artery runs upward anteriorly to the C1–C2 joint and could easily be damaged.
- The screw could exit C2 anteriorly and not enter the atlas.
Postoperative Care

Patients are immobilized in a firm collar for a period of 6 to 8 weeks but are allowed to remove the collar for daily care. After 6 to 8 weeks, the collar can be discarded when resting. If additional posterior wiring has been used, a soft collar can be worn instead of a firm collar.

Cannulated Screw Technique

As in direct anterior fixation of the odontoid, we used the cannulated screw technique for both posterior and anterior transarticular screw fixation. This technique also has been used and published by others. With Kirschner (K) wires as drill guides, we performed the surgery in a minimally invasive manner. Through bilateral stab wounds, a K wire can be placed under anteroposterior and lateral simultaneous image intensifiers from the described entry points toward the lateral projection of the C1 arch. The interspinous graft can then be placed between the C1 and C2 through a small midline incision. This technique has been further elaborated by the Ulm group. More recently, an image-guided technique was presented; however, this technique is far from being used on a routine basis.

Using lateral image intensifier control, a 1.2-mm K wire is inserted with a surgical drill guide in a strictly sagittal direction into each hole. The entry point of the K wire is at the lower edge of the caudal articular process of C2. The length of the screw is established with a special ruler by measuring the protruding part of the guidewire. Before inserting the cannulated screw over the K wire, the entrance for the screw is prepared by a special 3.5-mm cannulated countersink to facilitate the starting purchase of the screw. The appropriate 3.5-mm self-tapping, cannulated cancellous bone screw (same as for odontoid fixation, usually ~40–45 mm long) is inserted over the guidewire. The progress of the screw must be observed under lateral image intensifier control to ensure that the K wire does not migrate proximally beyond the C1 arch and the screw does not push away (anteriorly and upward) the lateral mass of C1, consequently dilating the C1–C2 joint.

In severe degenerative diseases, the sclerotic subcortical bone of the C2 joint may prevent insertion of the self-drilling screw. In this case, a 2.7-mm cannulated drill bit is used to cross the joint, and a cannulated fully threaded 3.5-mm screw can be inserted after regular tapping of the predrilled screw hole.

Postoperative Care

Patients are immobilized in a firm collar for a period of 6 to 8 weeks but are allowed to remove the collar for daily care. After 6 to 8 weeks, the collar can be discarded when resting. If additional posterior wiring has been used, a soft collar can be worn instead of a firm collar.

Fusion

Following bilateral screw fixation, according to the above described techniques, a posterior C1–C2 fusion is performed, preferentially Brooks-Jenkins or Gallie fusion. According to Magerl’s technique, it is preferable to supplement the graft with a posterior wire or cable, as this increases the stability of the fixation and hence the fusion rate. Instead of a wire, a nonresorbable suture can be used.
When there is a defect or fracture of the posterior arch of C1, a fusion of the atlantoaxial joint must be performed. For visualization of the atlantoaxial joint, K wires are drilled into the posterior aspect of the lateral mass of the atlas. For this purpose, the greater occipital nerve is retracted cranially. The soft tissues containing the greater occipital nerve and its accompanying venous plexus are retracted simultaneously. The atlantoaxial joints are exposed by opening the posterior capsule, thus making the C1–C2 joint visible (Fig. 11.4). The articular cartilage of the posterior half of the facet joint is removed with either a small chisel or a sharp curet, after which the joints are packed with cancellous bone, and the screws are inserted.

**Advantages and Disadvantages**

The advantages of transarticular C1–C2 screw fixation are biomechanical superiority to the wiring techniques, the possibility of maintaining reduction, and the lack of need of integrity of the posterior arch of C1 for the fixation technique. Furthermore, this technique can be easily combined with a Gallie fusion, thus obviating the need for relevant external immobilization.

The disadvantages include the fact that this is a demanding technique and that there is a potential risk of bleeding due to injury to the venous plexus located lateral to the entry point of the screws or due to injury of the vertebral artery. This is particularly true in anomalies of the vertebral artery course in relation to the massa lateralis of C1 and C2. An aiming device may facilitate insertion of the screws. It may be difficult to apply this technique in patients with an upper thoracic kyphosis because the necessary inclination of the screws may be in conflict with the kyphosis, and the insertion of the screw therefore may be limited.

Depending on the obliquity of the screw insertion, a complete reduction of the C1–C2 joint may be necessary. When there is erosion of the transverse process of C2, the vertebral artery may change direction; this aberrant trajectory of the vertebral artery may make this technique impossible. In such a case, bilateral transarticular screw fixation may not be possible. An option may be unilateral transarticular screw fixation in a fracture or a defect of the posterior arch of C1, C1–C2 fusion is achieved by removing the posterior half of the C1–C2 joint and its cartilage. The greater occipital nerve (not shown) must be retracted cranially, to expose the C1–C2 joint. With a small curet, the cartilage can be removed, and bone graft can be added to the C1–C2 joint.

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**Case Illustration**

Transarticular screw fixation of C1–C2 with Gallie-type titanium cable fixation in a 55-year-old rheumatoid patient is shown in Fig. 11.5.

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**Fig. 11.5a–c**

a A 55-year-old man with rheumatoid arthritis predominantly of the C1–C2 segment and pannus formation of the odontoid.
b, c Postoperative radiographs with transarticular screw fixation and Gallie-type wire fixation without any decompression. The immobilization of the C1–C2 segment may lead to resolution of the pannus.
transarticular screw fixation combined with posterior C1–C2 wiring.

**Indications**

The following disorders may qualify for posterior transarticular screw fixation:

- Acute and chronic atlantoaxial instability, mostly due to rupture of the transverse ligament of C2 following trauma or inflammatory destruction (Fig. 11.6)³⁻⁴¹⁻⁴³
- Degenerative arthritis of the C1–C2 joint (Fig. 11.7)³

- Rheumatoid arthritis (isolated) of the atlantoaxial junction (Figs. 11.5 and 11.8a, b)³⁻⁴¹⁻⁴³ with more extensive involvement of the cervical spine (atlanto-occipital migration) (Fig. 11.8c, d)
- Malformations or posttraumatic deformities³⁻⁴¹⁻⁴²⁻⁴⁴⁻⁴⁵
- Jefferson fractures³⁻⁴⁶
- In rare cases, this technique may be applied in metastatic tumor surgery and infections (Fig. 11.9)³⁻⁴¹
- Oblique odontoid fracture type II (fracture line from posterosuperior to anteroinferior, where a direct screw fixation of the odontoid is not possible and an anterior transarticular screw fixation is not an option)³⁻⁴⁻³⁻⁴¹⁻⁴³

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**Fig. 11.6a–c** Ligamentum transversum rupture with C1–C2 subluxation in the sagittal plane.
- a, b Proof of instability by the flexion–extension view.
- c The treatment of choice is transarticular screw fixation of C1–C2.

**Fig. 11.7a–d** A 44-year-old woman with a 20-year postoperative follow-up after transarticular screw fixation and posterior Gallie-type fusion of C1–C2 with a nonresorbable suture (no wire) of a posttraumatic degenerative atlantoaxial arthritis. No adjacent segment disease is noted. The patient remained asymptomatic.
Fig. 11.8a–d

a Early stage of atlantoaxial rheumatoid arthritis with translational frontal instability and severe neck pain. Minimal pannus formation is seen.

b Unisegmental transarticular fixation.

c Rheumatoid arthritis involving the occipitoatlantal and atlantoaxial joints with vertical migration and sagittal instability of the C1–C2 joint (arrows).

d Postoperative image of the occipitocervical fixation with a Y plate, including transarticular C1–C2 screws, with the right screw too far lateral but still within the massa articularis of C1 and no damage to the vertebral artery. The wire holds the arch of C1 toward the plate.
Results

Generally speaking, the results of most series with posterior transarticular screw fixation of the atlantoaxial joint published in the literature are good. Complications reported include vertebral artery injury, unilateral insufficient screw purchase mostly due to anomalies, screw pullout or breakage, and neurological or vascular damage. Strokes and transient ischemic attacks have been mentioned in the context of screw fixation. Even when there is screw malposition (~2%), it is usually not clinically relevant. This technique can be applied safely in children. A survey of the American Association of Neurological Surgeons/Congress of Neurological Surgeons on disorders of the spine peripheral nerves reported the results of 847 active members. Of 101 respondents, 2492 C1–C2 transarticular screws were placed in 1318 patients. Thirty-one patients (2.4%) had known vertebral artery injuries, and an additional 23 patients (1.7%) were suspected of having injuries. However, only 2 (3.7%) of the 54 patients with known or suspected vertebral artery injuries exhibited subsequent neurological deficits, and only 1 (1.9%) died of bilateral vertebral artery injury. Other iatrogenic complications included dural tears, screw fractures, screw breakout, fusion failure, infection, and suboccipital numbness. Therefore, the risk of a vertebral artery injury (both suspected and known cases) was 4.1%, or 2.2% per screw inserted. The risk of neurological deficit consequently was 0.2% per patient, or 0.1% per screw, and the mortality rate was 0.1%.

The management of a vertebral artery injury includes either placing the patient under observation or carrying out an immediate postoperative angiography with possible balloon occlusion or vessel repair under the microscope. Goel and Gupta draw attention to the fact that it is the course of the vertebral artery within the facet of the axis that predisposes it to risk of injury during screw implantation. Therefore, it is crucial to study the course of the vertebral artery at the C1–C2 complex to avoid potentially fatal injury of the vertebral artery.

Anterior Technique

Surgical Principle

The anterior C1–C2 screw fixation provides anterior stability and is as stable as a posterior transarticular fixation in all clinically significant planes of motion. The access for anterior fixation is less traumatic than the posterior approach with its dissection of the paravertebral muscles.

Biomechanics

In early 2000, we investigated the biomechanical behavior of the anterior transarticular screw fixation of C1–C2 and compared it with the posterior technique. The only significant difference in cadaver studies was found for flexion–extension, where the cerclage wire added some additional strength to the posterior transarticular fixation. Otherwise, we did not find any significant difference between anterior and posterior fixation techniques.

Surgical Techniques

The initial part of the approach and technique is identical to anterior odontoid screw fixation. The patient is in a supine position, and two image intensifiers are necessary to identify the midline of the upper cervical spine as well as the lateral projections of the joints. Without the help of these intensifiers, the technique cannot be performed safely.

The head is placed in the extended position to facilitate access to C2 and screw insertion. The use of a Mayfield headrest makes reduction of a possible fracture and dislocation easier than Gardner-Wells tongs combined with a horseshoe headrest. In case of persistent anterior displacement of C1 in relation to C2, and therefore an incongruency of the C1–C2 joints, further reduction of the C1–C2 complex can be achieved by pushing directly on the anteriorly displaced C1–C2 segment through the mouth with the index finger.
Surgical Approach

An anterior medial approach is used. The surgical incision should not be started until the C1–C2 segment is satisfactorily reduced. The placement of the incision is determined by placing a long K wire along the side of the neck in the intended direction of the screws and viewing lateral projection on the image intensifier. The transverse incision can then be made in the neck where the K wire is likely to exit the skin (in most cases at the level of C4–C5).

Surgical Procedures

The vertebral column is exposed anteriorly by blunt dissection until the inferior edge of the body of the second cervical vertebra is identified. Two radiolucent Hohmann retractors are then inserted on each side of the odontoid into the C1–C2 joints to expose the body and the facet joints of the axis (Fig. 11.12). Today we apply the cervical SynFrame (Synthes, Oberdorf, Switzerland) to expose the operating field, which is delineated by the position of the Hohmann retractors and blade in a stable ring configuration (Fig. 11.13). The anterior portion of both atlantoaxial joints is then denuded of cartilage using a small angled curet under direct view and fluoroscopic guidance (Fig. 11.14a).

The landmarks for the insertion of the screws for the anterior C1–C2 fixation are identified. The starting point for the C1–C2 stabilization screw lies under the superior overhanging lip of the lateral mass of C2, 5 mm lateral to the base of the odontoid process or 8 to 10 mm from the axis midline. Under anteroposterior and lateral fluoroscopic control, the guidewire is advanced in a maximum 25° lateral and, anteroposteriorly, 20° sagittal direction and engaged in the lateral masses of C1 (Fig. 11.14b, c). The lengths of the K wires are measured, the screw holes are overdrilled, and 3.5- or 4-mm cannulated self-tapping titanium screws are inserted (Fig. 11.14d, e). Due to the saucerlike shape of the C1 lateral mass, the image intensifier picture overestimates the length of the transarticular screw. Therefore, it is useful to have a preoperative measurement on the CT scan of the appropriate length of the screws to be selected. The anterior transarticular C1–C2 screws are usually 1.5 to 2.5 cm long.
Fig. 11.13a–d  Application of the cervical SynFrame to expose the operating field, which is delineated by the position of the Hohmann retractors and blade in a stable ring configuration, making a surgical assistant complementary.

Fig. 11.14a–e  

a  The anterior portion of both atlantoaxial joints is denuded of cartilage using a small angled curet under direct visualization and fluoroscopic guidance.

b, c  The landmarks for anterior C1–C2 fixation are identified. The starting point for the C1–C2 stabilization screws lies on the undersurface of the overhanging lip of the lateral mass of C2, 5 mm lateral to the base of the odontoid process or 8 to 10 mm from the midline. Under anteroposterior and lateral fluoroscopic control, the guidewire is advanced in a 25° lateral direction and engaged in the lateral mass of C1.

d, e  The length of the K wires are then measured, the screw holes are overdrilled, and 3.5- or 4.0-mm cannulated self-tapping titanium screws are inserted. Due to the saucerlike shape of the C1 lateral mass, the image intensifier picture overestimates the length of the transarticular screw. Therefore, it is useful to have a preoperative measurement on the CT scan of the appropriate length of the screws to be selected. The anterior transarticular C1–C2 screws are 1.5 to 2.0 cm long.
Postoperative Care

Postoperative immobilization with a soft collar is usually sufficient when there are no other pathologies, which may require a more stable external immobilization.

Advantages and Disadvantages

The advantages of this technique are that it does not require prone positioning, and it is a less traumatic approach than a posterior approach, in which the posterior paravertebral muscles need to be dissected. The technique may, be demanding, however, and proper equipment (two image intensifiers) is mandatory.

The disadvantages are the setup for anterior C1–C2 screw fixation, which requires two C arms. Also, the technique requires that the patient be brought in a sufficiently lordotic position, so that there is no interference during drilling and positioning the screws in a patient with a high sternum. A preoperative K wire can be held in place using a lateral radiograph projection to investigate whether the inclination of the K wire trajectory interferes with the level of the sternum.10,12,48,49

Indications

- A failed anterior odontoid screw fixation
- Instability of C1–C2 (e.g., in a rupture of the ligamentum transversum)49
- Unstable Jefferson fractures60
- Arthritis of the C1–C2 joint (degenerative arthritis)
- Isolated rheumatoid arthritis of C1–C2
- Occasionally for fixation in metastatic tumor60 or infections in combination with anterior plate fixation of the middle and lower CS
- Some forms of malformations (os odontoideum)

Furthermore the approach, which is used for this technique, is basically identical for direct screw fixation of odontoid fractures and can be combined with such a procedure in some selected cases (Fig. 11.15).

Results

There are no big series available to definitively confirm the harmless nature of this surgical technique. Since our first publication,10 we have operated on 18 patients according to this technique. We did not encounter any major complication, nor did we see any screw loosening or breakage. The fact that no formal fusion of the C1–C2 joint is done is not reflected in signs of nonunion, although real bony consolidation of the joint could not be demonstrated, at least not in the observation period of 2 years, at which point we stop following patients when they do well clinically. The case published by Apostolides et al. is very similar to our first case (Fig. 11.13), which healed uneventfully.12 Since Barbour described his bilateral anterolateral transarticular screw fixation for C1–C2

Case Illustration

Figure 11.15 shows an odontoid fracture type II with a combined injury of the C1–C2 joint on the left side, where a direct screw fixation of the odontoid fracture would not be sufficient to immobilize the C1–C2 fracture. Therefore, a combined direct screw fixation of the odontoid and C1–C2 anterior screw fixation was done.

Fig. 11.15a–f
a–d Combination of an odontoid fracture type II and an intra-articular fracture of the left massa lateralis of C2 (arrow).

E, f Anterior transarticular screw fixation and anterior odontoid screw fixation through the same access.
in 1971, only a few articles have reported data using his mostly modified technique.\(^9\) Simmons and du Toit used this technique and presented results with a modification.\(^8\) A further modification of the Barbour technique was applied successfully by Lesoin et al. in seven cases.\(^6\) All these techniques, however, are not the same as that described by Apostolides et al. and later by us.\(^10,12\)

Koller et al.,\(^49\) Cacciola et al.,\(^47\) and Goel and Gupta\(^31\) published meticulous anatomical analysis of the relationship of the massa lateralis of C1 and C2, more specifically, the projection of the vertebral artery groove on the massa lateralis of C2.\(^31,47,49\) With the technique that we developed, using a K wire with self-tapping cannulated screws, the chance to come in contact with the vertebral artery groove is almost nil, as the entry point is \(-5\) to \(8\) mm from the midline of the odontoid at the groove where the facet joint lip of C2 overlaps the massa lateralis anteriorly. The course in the C2 massa lateralis is short compared with the position of the screw, which is positioned in the lateral mass of C1. As demonstrated by Koller et al., it is crucial not to tolerate a rotation of C1 relative to C2, because the screw may not enter the C1 massa lateralis.\(^49\) It seems to us that the angle of the screw insertion in the frontal plane should never exceed \(25^\circ\) and \(-15\) to \(20^\circ\) in the sagittal plane to avoid the vertebral artery groove. Similar indications are given by the anatomical studies done by Lu et al.\(^51\)

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Transarticular Fixation Combined with Other Techniques

A posterior transarticular fixation may be combined with either an occiput–C2 fixation or a long posterior fixation using rods or plates (Fig. 11.16). This is usually the case in tumor reconstruction or in rheumatoid arthritis with multisegmental fixation and frequently including the occiput in the fixation. (Figs. 11.8, 11.9, and 11.16).

The use of transarticular C1–C2 screws allows the surgeon to create a block between the C1 and C2 vertebrae and to fix this block against the occiput or the lower cervical spine. When using plates, it is advisable to fix the C1–C2 joint first through the plate, then fix the plate to the occiput. However, the plate and the rod need to be modeled to meet the angulation between the occiput and the cervical spine.

An anterior transarticular screw fixation may be combined with an anterior direct odontoid screw fixation in case of a combined injury with odontoid fracture and dislocation between C1 and C2, including an injury to the C1–C2 joint (Figs. 11.13 and 11.15). In 1994, Goel and Karapurkar described a transoral metal plate fixation that extended from the clivus to the C2–C3 vertebral body.\(^52\) It may also be an option to include C1–C2 anterior screw fixation with an anterior plate of the middle and lower cervical spine in long fixations as in an anterior approach. The latter configuration has not been systematically investigated.

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**Fig. 11.16a–f**

a–d: Atlanto-occipital and atlantoaxial posttraumatic arthritis. Occipitoaxial fixation with a Y plate, including C1–C2 transarticular screws.

e, f: Occipitocervical and subaxial rheumatoid arthritis stabilized by plates, including transarticular C1–C2 screws, to form a stable atlantoaxial block and a sublaminar wire of the posterior arch of C1 to fix the C1–C2 block against the plate, which is fixed to the occiput by cortical screws.
Conclusion

Over the last 20 years the atlantoaxial transarticular fixation techniques of the upper cervical spine have become an established surgical tool to treat not only complex but also routine pathologies at the craniocervical junction.

Increasingly we use the anterior transarticular screw fixation for isolated atlantoaxial arthritis or instabilities because of the minimal trauma caused by this type of surgery. If the atlantoaxial fixation is part of a more extensive craniocervical stabilization or if the unisegmental fixation is accompanied by posterior decompression, the application of the posterior transarticular screw fixation is recommended. Both techniques retain consistently good result with low morbidity and complication rates when the surgical technique is used in a standardized way and follows the established technical guidelines.

References

Cervical instrumentation surgery started in the early 20th century for management of unstable cervical spine disorders. However, progression of cervical instrumentation surgery has been slow in comparison with instrumentation surgery in the thoracic and lumbar spine. Simple instrumentation such as spinous process wiring and lateral mass wiring had been the representative fixation anchor for stabilization of the cervical spine for ages. Occipitocervical fixation had been similarly dependent on simple cervical fixation until the last decade of the 20th century. However, new fixation anchors for cervical instrumentation surgery appeared in the late 1980s and early 1990s, including the lateral mass screw and pedicle screw.¹⁻³ These rigid cervical fixation anchors allowed major changes in the field of cervical instrumentation surgery, and instrumentation surgery for cranio cervical disorders almost simultaneously assumed a new aspect. This chapter reviews the history of cranio cervical instrumentation surgery and several methods of fixation, and introduces the authors' technique of occipitocervical instrumentation.

## Historical Review of Occipitocervical Stabilization Surgery

A report by Pilcher in 1910 seems to be the first description of occipitocervical fusion. He reported an unintentional occipitocervical fusion after open reduction of atlantoaxial dislocation by exposing the occiput and the upper cervical spine.⁴ Foerster in 1927 conducted the first intended occipitocervical fusion with the use of a fibular strut graft without internal fixation for management of atlantoaxial instability caused by fracture of the odontoid process.⁵ In 1935, Kahn and Yglesias described the use of the autologous iliac crest for bone grafting in occipitocervical fusion.⁶ Cone and Turner reported a patient who had undergone a fusion from the occiput to the sixth cervical vertebra.⁷ They used wires placed through occipital burr holes to the spinous process of the cervical vertebra. Subsequently, several occipitocervical fusion techniques supplemented by wires had been developed. Robinson and Southwick described an onlay graft bone method in 1960. Wires passed from two burr holes in the occiput through the foramen magnum accomplished fixation of the grafted bone. The grafted bone was also stabilized by wires to the atlas, axis, and third cervical vertebra.⁸ Since then, several modifications of wiring techniques have been employed.⁹⁻¹⁰ Besides these reports, Perry and Nickel, as well as Newman and Sweetnam, reported occipitocervical fusion using chipped bone grafts without internal fixation. They postoperatively kept patients on prolonged bed rest with skull traction, followed by rigid external support, such as a halo cast.¹¹⁻¹² Despite the use of wiring techniques, postoperative stability of the fixed occipitocervical complex was insufficient, and bony union depended heavily on rigid postoperative external support and/or prolonged bed rest.

In the next stage of development of occipitocervical instrumentation surgery, in the 1980s and 1990s, application of a metal rod for use as a longitudinal connector and use of sublaminar wires and laminar hooks for cervical fixation were started. Luque introduced sublaminar wiring for correction of scoliosis in the thoracic and lumbar spine.¹³ This relatively rigid anchor was also used for cervical and occipitocervical fixation; however, initially, bone graft, such as the ilium, was used as the longitudinal connector, which did not supply sufficient stability to the fixed occipitocervical spine. Therefore, several types of metal rods were employed instead. In 1987, Flint et al. reported using Hartshill rectangular loop rods for occipitocervical fixation in 10 patients.¹⁴ A rectangular rod was fixed to the cervical vertebra via stainless steel sublaminar wires, and a firm collar was then used for postoperative external support. The authors noted no cases of pseudarthrosis. Since then, several series of occipitocervical fixation using metal rods and sublaminar wiring have been reported.¹⁵⁻¹⁷ According to these descriptions, the rods were contoured to correct kyphosis or maintain lordosis. One of the major disadvantages of using sublaminar wiring for cervical or occipitocervical fixation is the need for extension of construct to fixed segments when laminae had been removed already or if a laminectomy was required for decompression. Another disadvantage of sublaminar wiring is that single-level anchoring is not sufficient to obtain satisfactory stability or to correct a deformity (Fig. 12.1).

More recently, occipitocervical fixation using hooks or clamps as cervical anchors has been attempted.¹⁸⁻²⁰ However, these procedures have not become popular probably because of fear of setting the metal into the spinal canal, causing spinal cord complications (Fig. 12.2). Callahan et al. performed occipitocervical fixation using rods fixed to the lateral mass using wires passed through the articular facets.²¹ This procedure was useful for patients with missing laminae. In 1988, Goel et al. described the use of occipital screws for occipitocervical fixation.²² With the advent of screws, wire fixation of the lateral masses is not considered an option for occipitocervical fixation.
Fig. 12.1a–c  Occipitocervical fixation using cervical sublaminar wiring. Single-level cervical anchor using sublaminar wires is not sufficient to obtain stability or to correct a deformity.

a  The patient with os odontoideum underwent occipitocervical fixation using C2 sublaminar wiring and a rectangular rod.
b  Prior to surgery, anterior subluxation of C1 was partially corrected by skull traction.
c  However, the correction was not maintained, and subluxation developed again shortly after surgery.

Current Procedures of Occipitocervical Stabilization

Recent Development of Occipitocervical Instrumentation Surgery

Many reports have demonstrated that simple onlay graft techniques with wiring do not provide sufficient stability for occipitocervical fixation. In addition, occipitocervical stabilization using wiring usually requires longer fixation, especially for patients who need additional posterior decompression and for those whose laminae were removed in earlier surgeries. Therefore, more rigid internal fixations that do not require laminae for stabilization have been popularized. In the 1990s, several procedures were developed using atlantoaxial transarticular screws, lateral mass screws, and pedicle screws in the cervical spine. Regarding occipital anchors, burr holes for setting screws or hooks are generally placed paramedially on the occiput. According to the morphologic studies of the occiput by Zipnic et al. and by Ebraheim et al., the midline
of the occiput is thicker than the lateral portion, with the thickest portion of the midline (~1.55 mm average thickness) at the center of the external protuberance.\(^{23,24}\) Therefore, considering the stability of the screw and the minimal risks to neurovascular structures inside the skull, placement of occipitocervical instrumentation into the paramedian of the occiput has been recommended (Fig. 12.3).

The C1–C2 transarticular screw proposed by Maggerl and the fixation procedure involving the C2 pedicle screw and C1 lateral mass screws developed by Goel and Laheri have been considered the most rigid internal fixations for atlantoaxial arthrodesis.\(^{22,25–27}\) Grob and colleagues reported using C1–C2 transarticular screws as the anchor for occipitocervical fixation.\(^{28,29}\) A Y-shaped plate was used to connect the occiput and the bilaterally inserted transarticular screws. In 1993, Smith et al. reported their series of occipitocervical fixation using atlanto-occipital reconstruction plates and screws. Screws inserted into the lateral masses were used as the cervical anchor.\(^{30}\) In 1994, Goel and Laheri first described an occipitocervical fixation technique in which the cervical end of the plate was fixed with the help of a C2 pedicle screw alone or in conjunction with C1 lateral mass screws, and the occipital end of the plate was fixed with the help of screws.\(^{22,28}\) Use of C2 pedicle screws to fix the cervical end of the occipitocervical fixation has become a popular method. In 1995, Goel and Achawal described occipitocervical fixation simultaneously with foramen magnum decompression.\(^{31}\) They made a small bone flap incorporating the foramen magnum and turned the flap inside out. The technique was labeled a foramen magnotomy.\(^{32}\)

Abumi et al. reported a series of cases with occipitocervical fixation using a combination of occipitocervical rods and cervical pedicle screws.\(^{33}\) In their series, screws were inserted into the C2 pedicles for occipitoatlantoaxial fixation; more caudal pedicles were employed for fixation from the occiput to the middle or lower cervical spine in several cases. Occipitocervical fixation using C1–C2 transarticular screws, pedicle screws, or lateral mass screws is available for use in patients who require one-stage posterior cervical decompression and occipitocervical stabilization, because it requires no laminae for stabilization. In all these cases of cervical anchors used for occipitocervical fixation, both the stabilizing capability of an unstable segment and the pullout strength of the lateral mass screw are inferior to those of C1–C2 transarticular screws and pedicle screws.\(^{34–37}\) Oda et al. demonstrated by biomechanical study that C2 pedicle screws and C1–C2 transarticular screws provide significantly higher stabilizing effects when compared with sublaminar wiring and laminar hooks.\(^{38}\)
Despite the high capability for achieving occipitocervical fusion with stabilizing atlantoaxial transarticular screws or C2 pedicle screws, it is difficult to apply a distraction force for correction of alignment. In addition, further correction of atlantoaxial subluxation after insertion of atlantoaxial transarticular screws cannot be obtained. Therefore, correction of vertical subluxation of the odontoid process may be insufficient, and reduction of “irreducible” atlantoaxial dislocation may be difficult by an occipitocervical stabilization procedure using atlantoaxial transarticular fixation. Grob et al. conducted a statistical analysis of surgical reduction of occipitocervical malalignment. According to the results in their published study on occipitocervical fusion using the C1–C2 transarticular screw fixation, no significant differences between the preoperative values of upward migration of the odontoid process and those at the time of follow-up were noted, and 3 of 28 patients remained neurologically unchanged. Goel et al. also attempted reduction of vertical odontoid migration in cases with basilar invagination by using C2 pedicle screws and occipital fixation using screws. In a series of cases treated by Abumi et al., reduction of upward migration evaluated by the McRae line was 4.2 mm on an average, and only 1 of the 19 patients who had myelopathy before surgery remained in the same neurological grade and the remaining 18 patients obtained neurological recovery. The reduction of upward migration of the odontoid process may enhance the decompressive effect by realignment at the atlantoaxial dislocation and may produce better neurological improvement than other procedures, including occipitocervical fusion using the C1–C2 transarticular screw fixation. More recently, Goel and colleagues recommended opening of the atlantoaxial joint and direct distraction of the facets to achieve reduction of the basilar invagination and or irreducible atlantoaxial dislocation.

### Occipitocervical Disorders Requiring Reconstructive Surgery

Disorders requiring surgical intervention in the cranio-cervical junction include rheumatoid arthritis, trauma, metastatic or primary bone tumor, infectious disorders, and instability or deformity by congenital abnormalities. The primary indication for occipitocervical fixation is mechanical instability at the occipitocervical junction. With the development of instability, patients suffer intractable pain and/or neurological deficit. Conservative treatment is usually ineffective for these conditions. Neurological deficits at the cranio-cervical junction in many patients are caused by compression of neural tissues by tumors, by deformity, by instability, or by a combination of factors. For patients with tumors, if stability of the cranio-cervical junction has been compromised, or surgical extirpation of the tumor destroys important stabilizing factors at the junction, cranio-cervical reconstruction must be performed.

Many patients who require craniocervical fixation have a combination of deformities in the sagittal plane. These consist of anterior translation of the atlas on the axis, vertical subluxation of the odontoid process, and flexion deformity caused by anterior subluxation or dislocation of the occipitotantal complex on the axis (Fig. 12.4). Therefore, the ventral aspect of the junction of the medulla oblongata and the spinal cord is compressed by the tip of the cranially migrated odontoid process. In some patients with a severely anteriorly translated atlas, the tip of the odontoid process anteriorly and the posterior arch of the atlas posteriorly impinge on the neural elements. If anterior atlantoaxial dislocation is the cause of neurological deficits, ventral decompression is required. However, direct anterior decompressive procedures by a transoral or a mandible-splitting approach involve complicated perioperative management and a risk of infection. It is difficult to obtain solid fusion with these procedures even with postoperative support by

![Fig. 12.4 Mechanism of nerve compression at the cranio-cervical junction. Many patients who require craniocervical fixation have combined deformities in the sagittal plane. These consist of anterior translation of the atlas on the axis (white arrow), vertical subluxation of the odontoid process (black arrow), and flexion deformity caused by anterior subluxation or dislocation of the occipitotantal complex on the axis (curved arrow).](image-url)
rigid external fixation, such as a halo vest or cast. Therefore, direct anterior decompressive procedures are usually followed by additional posterior instrumentation with bone grafting.

**Occipitocervical Stabilization Using Cervical Pedicle Screw Fixation**

**Cervical Pedicle Screw Placement for Occipitocervical Fixation**

The combined use of cervical pedicle screws and occipitocervical rods for reconstruction of occipitocervical lesions provides sufficient correction of malalignment of the craniocervical junction by application of the combined force of extension and distraction. As a result of the reduction, indirect decompression of the ventral portion of the medulla oblongata is obtained by decrease of the mechanical stress at the anterior aspect of the neural tissue. 33

**Pedicle Screw Insertion in the Cervical Spine**

In occipitocervical fixation using cervical pedicle screws, many patients can be managed by occipitoatlantoaxial fixation, avoiding extension of the fusion level more caudad than C2. However, vertebrae below C2 should be used for pedicle screw insertion in patients who require longer fixation for reduction or stabilization, or for the patients who do not have a suitable C2 pedicle for screw insertion. 44

**C2 Pedicle Screw**

The cranial margin of the lamina of C2 is the landmark for the point of screw penetration in C2. To confirm the screw insertion point in C2, a slightly curved small spatula can be inserted into the spinal canal along the cranial margin of the C2 lamina to the medial surface of the pedicle of C2 (Fig. 12.5a). The angle for the C2 pedicle screw insertion should be 15 to 25° medial to the midline in the transverse plane (Fig. 12.5b). Prior to screw insertion, a pedicle probe should be inserted to create a route for the insertion of the tap and screw. Lateral C-arm projection should be used to confirm the screw insertion point and direction and to control the depth of the pedicle probe, tap, and screws.

**C3–C7 Pedicle Screws**

The points of screw penetration for C3–C7 pedicles are lateral to the center of the articular mass and close to the inferior margin of the inferior articular process of the cranially adjacent vertebra. The lateral margin of the articular mass of the cervical spine has a notch at the level of the pedicle. 45 The pedicles are located below the lateral vertebral notch at C2, at or slightly above the notch at C3–C6 (Fig. 12.6). The anatomical axis of the pedicle to the midline in the transverse plane is −45° from C3 to C6 and less in C7. However, the anteroposterior length of the cervical pedicle is shorter than the lumbar or thoracic spine. Therefore, surgeons have more freedom for the insertion angle and can insert a screw at a smaller angle (25–45°) in the cervical spine (Fig. 12.7).

The cortex at the point of screw insertion is penetrated with a high-speed burr. The surgeon can see the pedicle entrance directly in many cases by enlarging the insertion hole using a high-speed burr and a
After creating the insertion hole, a small pedicle probe, tap, and screws are inserted into the pedicle with the help of lateral C-arm projection to confirm the direction and insertion depth (Fig. 12.8b,c,f). Prior to screw insertion, the surgeon can confirm the proper creation of the screw path using a pedicle sounder (Fig. 12.8e). The cortex of the cervical pedicles is always thinnest laterally toward the vertebral artery. Therefore, the surgeon should keep this in mind while probing and tapping the pedicle and placing the screws. Especially while being pulled out, the tap can easily shift to a direction parallel to the sagittal plane by paravertebral muscle force, and the lateral wall of the pedicle with the thinner cortex can be violated. The direction of the tap must be kept the same as the insertion angle by application of force lateral against the directional force of the paravertebral muscle (Fig. 12.9). A drill bit must never be used to penetrate the cortex of the lateral mass or to make a hole for screw advancement.

The intended angle of screw insertion in the sagittal plane is parallel to the cranial end plate for pedicles of C5–C7 and at a slightly cephalad direction in C2–C4, according to the screw’s angulation in the sagittal plane. The direction of the C2 pedicle screw is almost perpendicular to the ventral surface of the C2 vertebral body (Fig. 12.10). The neurocentral junction in the cervical spine, which is near the base of the pedicle in the vertebral body, is sometimes harder to pass with the pedicle probe than in the thoracic and lumbar spine. In such cases, the junction can be perforated with a Kirschner wire to make a path for the pedicle probe into the vertebral body.

Occipitocervical Rod Application and Correction of Deformity

After insertion of the pedicle screws, decortication of the posterior cortex of the lateral masses and residual laminae is performed with a burr. Washers 2.5 or 3.5 mm in height are placed on the C2 pedicle screws to reduce anterior translation of the atlas on the axis and to allow space for monocortical onlay bone grafting between the rod and the lateral mass. The occipitocervical rods are contoured at the plate–rod junction to reduce hyperflexion at the occipitocervical junction. Self-tapping screws fix the plate portion of the rod onto the occiput. The outer table of the occiput must be penetrated by a diamond burr with a 2-mm-diameter head. The inner table of the occiput can be penetrated by screws; however, a screw that is too long can cause injury to the underlying neural or vascular structures. The caudal end of the rod is cut to a proper length, leaving adequate length to allow distraction force to correct upward migration of the odontoid process. Hyperflexion alignment of the occipitoatlantoaxial complex is corrected by application of extension force created by tightening of the nut to the pedicle screws (Fig. 12.11a). If reduction of vertical subluxation was not sufficient by application of
Fig. 12.8a–f Creation of a screw insertion hole.

a–c Lateral C-arm projection is helpful to confirm the screw insertion point and the direction of the screw. The probe and tap must be advanced between two broken lines. IAP inferior articular process.

d The cortex at the point of insertion is penetrated with a high-speed burr. The surgeon can see the pedicle entrance directly in many cases by enlarging the insertion hole using a high-speed burr and small curet (white arrow). IAP inferior articular process, SP spinous process, open arrow facet joint.

e, f The surgeon can confirm proper creation of the screw path using a pedicle sounder.
extension force, upward migration of the odontoid process can be reduced by application of distraction force between the plate portion of each rod and the head of each screw inserted into the pedicle of C2 or C3 using a spreader (Fig. 12.11b). These reduction maneuvers by application of the combined forces of extension and distraction are conducted alternately first on one side and then on the other to avoid excessive force being applied to one side of the screws. After reduction by application of proper force, rods and screws are fixed by tightening the set screw in the connectors. A large monocortical iliac bone is trimmed and packed between the two rods as a bridging graft between the occiput and the axis. Chipped cancellous and cortical bones are grafted on the remaining exposed lamina and lateral masses over the fused levels.

Text continues on page 120
Case 1 (Fig. 12.12) is a patient with a lesion at the craniocervical junction caused by rheumatoid arthritis. The patient was managed by occiput-C2 fixation.

Fig. 12.12a–d Case 1.

a The patient with rheumatoid arthritis suffered from vertical subluxation of the odontoid process and anterior subluxation of C1 on C2, causing neurological complications.

b Sagittal reconstruction computed tomography (CT) demonstrates invagination of the odontoid process into the foramen magnum.

c Combined force of extension and distraction reduced both the vertical and atlantoaxial subluxation.

d Pre- and postoperative magnetic resonance imaging (MR) demonstrates improvement of anterior spinal cord compression by reduction of vertical subluxation of the odontoid process. A preoperative cervicomedullary angle of 115° improved to 143° postoperatively. Preoperative hyperlordosis at the lower cervical spine was corrected spontaneously by correcting the flexion deformity at the craniocervical junction. The angle between the lines on the ventral side of the cervical spinal cord and the medulla oblongata on MRI serves as the cervicomedullary angle.
Case 2 (Fig. 12.13) is a patient with a lesion at the craniocervical junction caused by rheumatoid arthritis. The patient was managed by occiput-C4 fixation.

**Fig. 12.13a–d** Case 2.

a. The patient with rheumatoid arthritis had marked vertical subluxation of the odontoid process and atlantoaxial subluxation. Alignment of the lower cervical spine was remarkably hyperlordotic. She had preoperatively severe spinal cord dysfunction.

b. CT images show invagination of the odontoid process into the foramen magnum and destruction of the C2 vertebral body by rheumatoid lesion.

c. Preoperative MRI demonstrates compression of the nerve at the junction of the spinal cord and the medulla oblongata by cranially migrated odontoid process.

d. Screws were inserted into C3 and C4 pedicles, avoiding screw insertion into the destructed C2 vertebral body. Correction of the flexion deformity and vertical subluxation of the odontoid process were satisfactory, and preoperative hyperlordosis in the lower cervical spine was corrected spontaneously. Neurological disturbance improved significantly.
Case 3 (Fig. 12.14) is a patient with atlantoaxial subluxation caused by C5 odontoideum. The patient was managed by occiput-C2 fixation.

**Fig. 12.14a–c Case 3.**

a A 14-year-old girl with os odontoideum suffered from severe neck pain and quadriparesis. Flexion-extension films demonstrate instability at the occipitoatlantoaxial segments with posterior subluxation of the atlas on the axis.

b Preoperative T2-weighted MRI shows posterior subluxation of C1 on C2 and high signal intensity in the spinal cord at the level of C1–C2.

c The patient underwent occipitocervical fixation using occipitocervical rods and C2 pedicle screws. A computer navigation system was used to insert small-diameter screws into the C2 pedicles.

**Combined Surgery of Anterior Release and Posterior Fixation**

Most patients who require occipitocervical fixation can be managed by posterior surgery alone. However, those with a fixed or extremely rigid flexion deformity caused by fixed atlantoaxial dislocation after injuries or congenital deformities require a longer fixation than occipito–C2 segments to correct flexion deformity even when using pedicle screw fixation.

In 2006, Wang et al. reported on their procedure involving anterior atlantoaxial release and reduction by skull traction followed by posterior fixation using cervical pedicle screws for irreducible atlantoaxial dislocation. Among their 33 patients with irreducible atlantoaxial dislocation, 26 required occipitocervical fixation, and 7 were managed by atlantoaxial fixation. In the first stage of surgery, Wang and colleagues performed a transoral anterior release of the bilateral atlantoaxial joints without resection of the odontoid process under continuous intraoperative skull traction. After confirmation of reduction, the patient was turned to the prone position, and a posterior occipito–C2 fixation using C2 pedicle screws and occipitocervical plates or atlantoaxial fixation using C1–C2 transarticular screws or a combination of C1 lateral mass screw and C2 pedicle screws was done. The most caudally fixed vertebra was C2 in all patients. Their results demonstrate that the combined procedure
Case 4 (Fig. 12.15) illustrates this procedure.

**Fig. 12.15a–d  Case 4.**

**a** The patient had a congenital malformation that consisted of occipitalization of the atlas, assimilation of C2 and C3, vertical subluxation of the odontoid process, and atlantoaxial subluxation. The subluxation was irreducible.

**b** Preoperative MRI demonstrates nerve compression by invagination of C2, anterior subluxation of C1 on C2, and subaxial spinal canal stenosis.

**c** The surgeon performed anterior atlantoaxial release and reduction by skull traction followed by posterior fixation using a cervical pedicle screw for irreducible atlantoaxial dislocation. A laminectomy from C3 to C6 was conducted to address lower cervical canal stenosis.

**d** Postoperative MRI demonstrates improvement of the cervicomедullary angle and resolution of nerve compression by the tip of the odontoid process. (Courtesy of Professor Wang Chao, Third Hospital of Beijing University Medical Center, Beijing, China.)
enables reduction and fixation of irreducible atlantoaxial dislocation by shorter segmental instrumentation than a single posterior surgery. They emphasized that most of the so-called irreducible/fixed atlantoaxial dislocation could become reducible by anterior release and that the posterior short-segment fixation could achieve ideal reduction (Fig. 12.15). Great attention must be given while turning the patient to the prone position, however, as the atlantoaxial joint is unstable after the anterior release.

### Complications of Occipitocervical Fixation Using Posterior Screw Instrumentation

Posterior screw instrumentation procedures are associated with risks to the neurovascular structures. Screw insertion points, direction, and depth must be carefully controlled. Abumi et al. and Kast et al. reported neurovascular complications during cervical pedicle screw use.\(^{51-53}\) Cho et al. demonstrated one patient who had brainstem infarction caused by lateral mass screw insertion.\(^{54}\) According to these reports, the rate of neurovascular complication is not very high; however, the possibility of serious complications caused by screw insertion cannot be completely eliminated. Cerebrospinal fluid leakage resulting from too long of a tap penetrating the inner table of the occiput is usually stopped by the inserted screw.

Besides the complications directly attributable to screw insertion, radiculopathy caused by iatrogenic foraminal stenosis was reported by Abumi et al. and by Heller et al.\(^{52,55}\) They mentioned that the radiculopathy was due to correction of the kyphotic deformity or to reduction of the anterior translation; the radiculopathy was corrected in most patients with a foraminotomy. The neural foramina in patients with degenerative disorders or rheumatoid arthritis are sometimes stenotic preoperatively. Therefore, surgeons must not apply excessive shortening force at the segment with neural foraminal stenosis due to degenerative changes during correction of kyphosis. Use of a washer under the plate/rod with cranial vertebral screws is helpful for excessive reduction of anterior translation. Otherwise, a prophylactic foraminotomy is recommended for patients with marked stenosis of the neural foramen. Reconstructive computed tomography (CT) in an oblique plane provides useful information regarding the condition of the neural foramen. Once radiculopathy has occurred postoperatively, the position of the inserted screw must be checked using CT. If the screw is not a causative factor, foraminotomy for nerve root decompression must be conducted as soon as possible.

The diameter of the pedicles in some patients is too small to allow screw insertion.\(^{48,49}\) Preoperative oblique projections on plain radiographs are valuable for evaluating the pedicle size. In oblique projection plain films, contralateral pedicles are seen as an oval, projected onto the vertebral bodies, showing the outer and inner diameter of the pedicles (Fig. 12.16). CT evaluations (adjusted to the bone windows) are essential to assess the pedicle morphometry and determine the pedicle size, which allows the surgeon to choose the appropriate pedicle screw diameter, length, and direction in the coronal plane.

Preoperative evaluation of the morphology of the vertebral artery is important in preventing serious complications involving the artery.\(^{51,52}\) The incidence of ischemic brain complications caused by unilateral obstruction of the vertebral artery is low.\(^{56}\) However, if the dominant vertebral artery is injured, serious neurological complications can occur. CT and magnetic resonance imaging (MRI) provide information regarding the right-left dominance and anatomical variations of the vertebral artery. Magnetic resonance angiography must be used for patients with evidence of abnormalities of the vertebral artery or in whom these abnormalities are suspected (Fig. 12.17).

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**Fig. 12.16** Oblique film showing the pedicle cavity. Preoperative plain films in bilateral oblique views are valuable for evaluating the pedicle size. In oblique projection plain films, contralateral pedicles are seen as an oval projected onto the vertebral body.
Occipitocervical Stabilization Using Other Cervical Fixation Anchors

Occipitocervical fixation using C1–C2 transarticular or cervical pedicle screws allows good stabilization; however, the risk of neurovascular injury, especially to the nerve roots or the vertebral artery, cannot be completely eliminated. For patients with abnormal morphological conditions of the vertebral artery such as a high-riding artery, C1–C2 transarticular screw and pedicle screw fixation carries a higher risk than in patients with a normal vertebral artery. The pedicle of the axis is of sufficient diameter to place a screw in the majority of patients. However, if the size of the C2 pedicle is considered too small to insert a screw safely, more caudal pedicles must be selected for cervical anchorage, or an alternative occipitocervical fixation procedure must be selected.

Recently, alternative screw techniques using C2 lamina and cervical transarticular facet screws below C2–C3 have been reported (Fig. 12.18). Occipitoatlantoaxial fixation can be performed using C2 lamina screws or screws placed in the base of the spinous process of C2 or at the spinolaminar junction, as described by Goel and Kulkarni. However, connecting the C2 lamina or spinolaminar screws to the screws inserted into the pedicles or lateral masses in the middle or lower cervical spine may be difficult for patients who require a longer occipitocervical fixation. Transarticular facet screws can also be used for occipitocervical fixation. The disadvantage of using transarticular facet screws in occipitocervical fixation is the unavoidable need for extension of the fusion level, thus sacrificing the caudal facet joints of the vertebrae in which the screw is inserted. In addition, the mechanical stability of transarticular facet and lateral mass screws is not as strong as C1–C2 transarticular or pedicle screws. Using transarticular facet screws or lateral mass screws, a larger number of motion segments need to be fused for stabilization than when using C1–C2 transarticular or pedicle screws for reconstruction of the craniocervical junction. Besides these screw fixation procedures, Grob and Gonzalez et al. reported occipitocervical fixation using occipitoatlantal transarticular screws for atlanto-occipital dislocation. Such procedures may be useful for patients who cannot undergo occipitocervical fixation using lateral plate–rod fixation procedures (Fig. 12.19).

Computer-assisted Screw Placement

Computer-assisted navigation systems have been developed for use in cervical spine surgery. Such systems can be employed specifically for cervical pedicle screw insertion. Ludwig et al. conducted a comparative study of current computer-assisted technology and fluoroscopy-based technique in the laboratory. They showed that the use of a computer-assisted image guidance system did not enhance the safety or accuracy of placing pedicle
Fig. 12.18a–c Occipitoatlantoaxial fixation using a C2 lamina screw.

a  The patient with rheumatoid arthritis requiring occipito–C2 fixation had an extremely small left pedicle of C2.

b, c  A screw was inserted only into the right pedicle of C2, and a lamina screw was placed for C2. (Courtesy of Itaru Oda, MD, Hokkaido Orthopaedic Memorial Hospital, Sapporo, Japan.)
However, their computer-assisted system (Stealth Station, Sofamor-Danek, Memphis, Tennessee) only helped in navigating a screw guide tube at the insertion point on the bone surface and did not guide the actual tip of the pedicle probe, tap, or screw within the pedicle.

Abumi et al. have developed a custom-made computer-assisted guidance program for cervical pedicle screw insertion using the Stealth Station. The system, which facilitates each step of probing, tapping, and screw insertion within the vertebra, is more accurate than the fluoroscopy-assisted technique for patients with extremely small pedicles or for those with indistinct screw insertion points due to degenerative or destructive changes or previous posterior surgery. Kotani et al. reported that real-time, three-dimensional instrument/screw tip information provided by a computer navigation system was helpful in improving the safety and accuracy rates of pedicle screw placement in the cervical spine (Fig. 12.20). Further modifications and developments in technology will increase the systems’ safety records and refine surgical techniques.
Conclusion

Reconstructive surgery in the craniocervical junction has significantly improved over the past 10 years with the development of specialized instrumentation for this region. However, neurovascular complications can still occur because of misunderstanding of the pathological condition of the spine, inefficient surgical technique, and inattentive surgical handling. Surgery in the craniocervical junction using transarticular, pedicle, and lateral mass screws as cervical anchors is technically demanding. Only doctors who have sufficient experience in cervical spine surgery and with instrumentation in the thoracic and lumbar spine should perform these procedures. Further developments in technology using computer assistance will increase the safety of instrumentation surgery at the craniocervical junction.

References


Fig. 12.20 Computer-assisted cervical pedicle screw placement. A custom-made computer-assisted guidance program for cervical pedicle screw insertion using the Sofamor-Danek Stealth Station was used for a patient with abnormal pedicle conditions, such as marked degenerative changes, scoliotic deformity, and extremely small pedicle. The system facilitated each step of probing, tapping, and screw insertion.
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13 Odontoid Screw Fixation
Francesco Cacciola, Nicola Desogus, and Nicola Di Lorenzo

According to the classification of Anderson and D’Alonzo, direct osteosynthesis of type II and shallow type III odontoid fractures by means of an anteriorly inserted screw along the major axis of the dens and across the fracture line is an elegant and efficient technique to provide stabilization and restore normal function. Despite its description as early as 1980 by Nakanishi and in 1982 by Böhler, reporting his 8 years’ experience, however, it appears that this technique has not been fully adopted by centers dealing with this pathology. The vicinity of the fractured dens to vital structures and the impossibility of directly visualizing the structures that are worked upon still make some surgeons opt for conservative approaches or other, more movement-restricting fixation techniques. It is beyond the scope of this chapter to discuss the value of conservative treatment or to compare different surgical techniques in the management of odontoid fractures; however, an attempt is made here to clarify the indications for this procedure as well as to describe the important steps necessary for a good technical outcome. This should help readers who do not yet practice this technique to decide whether they want to include it in their own armamentarium.

Indications for Odontoid Screw Fixation

When considering whether or not to perform an odontoid screw fixation, the most important reason in its favor is the procedure’s success rate of ~80% to 90% in terms of functional restoration and stabilization. This obviously applies to appropriately selected patients and a meticulous surgical technique. In general, though, this rather straightforward operation delivers favorable results with relatively few risks.

In selecting patients for this procedure, three points need to be considered: the type of fracture, the timing of the surgery, and the age of the patient.

Fracture Type

Fracture type and the amount of displacement are important factors in selecting a patient for surgery.

According to the classification of Anderson and D’Alonzo, type II and “shallow,” or rostral, type III fractures are appropriate for odontoid screw fixation. Apfelbaum et al. suggested a subclassification of type II fractures according to the orientation of the fracture line. According to this subclassification, horizontal and oblique anterosuperiorly oriented fracture lines on the sagittal plane predict higher fusion rates than oblique anteroinferiorly oriented fracture lines. This subdivision, however, has not been addressed in series by other authors. Given that the overall fusion rate in the oblique anteroinferior group is still 75%, this information is probably not indispensable, but it can be of help in making difficult decisions.

The axis needs to be carefully examined for additional fractures or fracture lines, as this could have negative repercussions on screw purchase. The same goes for the fractured dens fragment. Type IIA fractures (comminution of the dens) might not warrant sufficient screw purchase, which is particularly important at the cortical tip of the dens that needs to be engaged by the screw.

As far as the amount of displacement is concerned, the literature shows that values ≥4 to 6 mm seem to represent a threshold beyond which the risk of nonfusion becomes significantly and probably unacceptably high with conservative treatment. Finally, a magnetic resonance imaging scan of the area to check for integrity of the transverse ligament is indicated even though not indispensable, in our opinion, due to the relatively low association of ligamentous rupture with an odontoid fracture. However, because of the common availability of this imaging technique and its routine use in spinal trauma in many centers, particular attention to the state of the transverse ligament should be paid. In case of a clear rupture, this would represent a contraindication to odontoid screw fixation.

Timing of Surgery

The general rule of the earlier the fusion, the better surely also applies to this type of procedure. Evidence from the literature seems to suggest that there are no differences in fusion rates during the first 6 months, whereas after 18 months, fusion rates clearly drop. This information is based on a series published by Apfelbaum et al. and has led to a division of patients as “early” or “late.” The fact that there is essentially no information on patients who had a fracture between 6 and 18 months is because such patients were not included in that study (or, to our knowledge, in any other study).

Knowing, however, that it apparently does not make a difference whether a patient undergoes fusion immediately
or after 6 months is surely helpful, as it gives the surgeon the option of trying conservative treatment in those patients deemed appropriate while still having time to do fixation should that treatment fail.

### Age

Although there is some controversy in the literature, generally it is agreed that the patient’s age influences the outcome of odontoid screw fixation. The younger the patient, the more likely it is that fusion may be obtained by external immobilization. Sherk et al. reported a series of 35 children (younger than 7 years), with only 1 failed fusion after halo immobilization. The older the patient, the more the fusion rate seems to correlate inversely. Lennarson et al. showed in one of their studies that the nonunion rate in patients older than 50 years is 21 times higher than in those who are younger.

Even if one tends to not believe in the inverse relationship between age and fusion rate in conservative management, the important message surely seems to be that older patients do at least as well as younger patients with surgery; this can be important when tailoring a specific treatment plan. In older patients, compliance with external immobilization such as a halo jacket and the resulting overall movement restriction can have significant negative repercussions on outcome and the patient’s general health. We therefore tend to favor surgery in older patients, taking only unfitness for general anesthesia and severe osteopenia as contraindications.

### Surgical Technique

The following is a description of the operative technique used in odontoid screw fixation outlining the most significant general steps that need to be followed. Various manufacturers offer different systems to carry out this operation, and some specific steps might thus differ according to the specific system. It is not our intent to specifically describe the use of any of these systems but to outline the important points that are necessary for a smooth and successful operation.

### Patient Positioning and Setup

Under general anesthesia, the patient is intubated with an endotracheal endoscope. This can be accomplished via either the nose or the mouth, making sure that a nonar-mored tube is used. The patient is then positioned neutrally supine on the operating table. The patient’s head can be secured in a three-pin fixation device, rest on a horseshoe, or positioned directly on the operating table with a towel roll under the neck. What is most important is that the head remains immobile throughout the procedure.

Once the patient is thus positioned, two C-arm image intensifiers are brought in and centered on the C1–C2 complex, with one obtaining a lateral view and the other an anteroposterior view (Fig. 13.1).

In case of dislocated fracture, the patient’s head should be gently manipulated under lateral fluoroscopy in an attempt to reduce the fracture as much as possible, then secured in the desired position. The two C-arms should remain in position during the entire procedure, as frequent imaging is crucial during the various steps. To obtain a good anteroposterior view, it is often helpful to obtain open mouth views by inserting a radiolucent mouth opener or bite blocks in the patient’s mouth.

Once these steps are accomplished, the C-arms are both in a position to obtain good views, and the patient’s head is well secured and in the correct position to obtain as much fracture reduction as possible, the patient and the equipment are draped in the usual fashion.

![Fig. 13.1 Intraoperative photograph showing patient positioning for odontoid screw fixation. Note the biplanar fluoroscopy left in place throughout the entire procedure.](image-url)
Approach to the Lower Edge of the Axis and the Screw Insertion Site

The approach to the anterior cervical spine is identical to a standard approach to the subaxial spine for an anterior cervical discectomy. A horizontal skin incision is made roughly at the level of C5, extending from the midline to slightly beyond the medial border of the sternocleidomastoid, usually on the right side of the patient or according to surgeon preference (Fig. 13.2).

After division of the platysma, the fascia of the sternocleidomastoid is sharply incised along its medial border. Blunt dissection along the natural planes and medial to the carotid sheath leads down to the anterior plane of the cervical spine. At this stage, both longus colli muscles are partially lifted from the vertebral plane, and a self-retaining blade retractor is firmly positioned underneath them. Up to this point, the approach is identical to an anterior discectomy.

Once the lateral retractors are inserted, the vertebral plane is followed rostrally until the inferior border of C2 is palpated. At this stage, craniocaudal self-retaining retractor blades are inserted.

Under fluoroscopic control, a Kirschner wire is inserted through the incision, advanced to the anteroinferior border of C2, and hit with a mallet (Fig. 13.3). On the anteroposterior plane, the K wire should sit in the center of the odontoid process; on the lateral view, it should already be angulated in a way that its projection will go through the major axis of the dens and penetrate its posterior half. The central position on the anteroposterior view is chosen because we prefer the insertion of a single screw, as the use of two screws, from our experience, offers no advantage.

Once the K wire is in place, it can be used to slide a cannulated drill guide over it and anchored on the cervical spine. With some systems, the K wire is actually fully advanced through the whole length of C2 with a drill, thus creating the trajectory for the screw. Other systems replace the K wire once the drill guide is in place and use a 2-mm drill bit to drill the screw trajectory. Continuous fluoroscopic control in anteroposterior and lateral projections is mandatory (Figs. 13.4 and 13.5).
What is important at this stage, whether a K wire or a drill bit is used to create the screw hole and canal, is to measure the depth of penetration into C2 and the dens, as this will determine the screw length that needs to be inserted. Different instrument systems have different devices and means of accomplishing that.

Once the screw hole is created and the depth of penetration measured, insertion of the screw can follow.

### Screw Insertion

There are essentially two types of screws: lag screws and fully threaded screws. In our opinion, lag screws are indicated in the majority of cases because they allow for the possibility of reducing and compressing a fracture fragment. Once the screw head is engaged against the inferior border of C2, the threaded part continues to deliver the fragment downward upon turning, with no change in position. The only situation where lag screws may be contraindicated is an anteroinferior oblique fracture line of the dens, as the compression can lead to malalignment of the fracture (Fig. 13.6). When using lag screws, the exact measurement of the needed screw length prior to insertion becomes particularly important.

At the stage of screw insertion, there is only one more important point to be observed and that is to make sure that the screw engages and traverses the cortex of the fractured dens fragment. A protrusion of the screw a couple of millimeters beyond the dens cortex is safe and mandatory for good purchase and to avoid later screw pullout.

Again, this needs to be closely monitored on fluoroscopy.

### Postoperative Care

The issue of whether or not to suggest a postoperative cervical collar is not resolved. In a fracture fixed with a lag screw of the correct length that has appropriately traversed the dens cortex and offers good purchase in a good-quality bone, a collar is most likely superfluous, whereas poor bone quality and less good purchase in a noncompliant patient would warrant immobilization in a rigid cervical collar for 6 to 8 weeks. Again, the final decision is most appropriately based on the inclusion of specific patient features and needs.

### Conclusion

Odontoid screw fixation can be a very efficient and rewarding procedure for both the patient and the surgeon. Correct patient selection is an important step in this procedure, and appropriate corroboration of the guideline recommendations with the particular features and needs
of the individual patient can lead to a 90% success rate with minimal need for patient immobilization (Fig. 13.7).

The surgical technique is straightforward, and as long as all steps are correctly performed and good fluoroscopic visualization is guaranteed throughout the entire procedure, the risk of any surgery-related complication is very low. In our experience, as well as in the literature, apart from the possibility of retraction-related problems associated with the standard anterior approach to the cervical spine, no morbidity or mortality directly related to odontoid screw fixation has been reported.2–26

References


Fig. 13.7 Decision-making flow chart for the management of type II and shallow type III odontoid fractures.
Joint “Jamming” as a Treatment for Atlantoaxial Dislocation and Basilar Invagination

Atul Goel

This chapter discusses an alternative technique for atlantoaxial fixation that involves blocking, or “jamming,” movements of the atlantoaxial joint.¹ The technique features forcible impaction of spiked titanium metal spacers and bone graft within the distracted atlantoaxial facet joints. Wires, metal loops, rods, and screws are not used for fixation. Onlay and interfacetal bone grafts subsequently produce bone fusion. The technique simulates to an extent interbody fixation of the subaxial spine in cases of spinal instability and spondylolisthesis. Joint jamming as a stand-alone method or in combination with other fixation methods could provide firm stabilization in selected cases of atlantoaxial dislocation and basilar invagination.

Role in Atlantoaxial Dislocation

Our successful experience suggests that joint jamming can be used in highly select cases of atlantoaxial dislocation. We are still not entirely convinced that it can be a stand-alone method of atlantoaxial fixation in all cases. It can, however, be a useful method of fixation when used in combination with other posterior fixation techniques. The utility and indications of use of this technique are still under evaluation.

Indications

Our preferred technique for mobile atlantoaxial dislocation is Goel and Laheri’s technique of lateral mass plate and screw fixation, as discussed.²⁻⁶ In our experience, we have observed that in select cases, placement of the bone graft and titanium spacers by themselves made the region stable, and the need for additional plate and screw fixation could be avoided. Patients with atlantoaxial dislocation following trauma and those having only subtle or moderate recent-onset abnormal mobility of the joint and “normal taut” ligamentous assembly were more suitable for the described technique. The excessive mobility of the atlantoaxial joint and the laxity of ligaments in cases with congenital and long-standing dislocation probably make them unsuitable for the described technique in isolation, but it can still be employed in combination with most other methods of fixation. Direct operative observation of the nature of the facets and the joint and the stability of the impacted spacer within the joint space also determine if any additional fixation device is necessary.

Surgical Technique

The basic surgical steps in exposure of the region have been described in our articles on the subject.²⁻⁸ Cervical traction is given prior to induction of anesthesia, and the weights are progressively increased to −4 to 5 kg, or one-eighth of the patient’s total body weight. The patient is placed prone with the head end of the table elevated to −35°. The atlantoaxial facet joints are widely exposed on both sides after either sectioning or elevating superiorly the large C2 ganglion. The joint capsule is excised, and the end-plate articular cartilage is widely removed using a microdrill. The facets are distracted with the help of varying sizes of osteotomes, which are introduced in the joint with their sharp edge and then turned 90° to effect distraction. The joint space is assessed, and a specially designed spiked titanium spacer suitable for the region is impacted, or “jammed,” into the joint space using suitable instruments. The average size of the spacers used measures 12 mm in length, 10 mm in breadth, and 6 mm in height. Customized titanium spacers have multiple small holes and are tapered on one side for easier insertion during placement in the joint space (Figs. 14.1, 14.2, 14.3, and 14.4). The base of the spacer has a transverse slot that assists in placement and stabilization of the instrument used for its impaction into the
Fig. 14.1a–e  Images of a 56-year-old female patient.

a  Preoperative computed tomography (CT) scan with the neck in flexion shows atlantoaxial dislocation. Assimilation of the atlas can be observed.

b  Preoperative CT scan with the neck in extension showing reduction of the atlantoaxial dislocation.

c  Sagittal image of the postoperative CT scan showing the spacer within the atlantoaxial facet joint.

d  Coronal image showing the spacers within the facet joints on both sides. Distraction of the facets can be observed.

e  Lateral radiograph with the neck in flexion with spacers in both facet joints 28 months after surgery. Solid bone fusion can be seen.
joint cavity. Introduction of the bone graft pieces within the joint cavity provides stability to the implant and additional opportunity for bone fusion. Multiholed titanium spacers allow bone incorporation and fusion across the distracted joint space. Corticocancellous bone graft pieces harvested from the iliac crest are stuffed into the space available around the spacers. Additional bone graft is placed over the posterior elements of C1 and C2 after decorticating the host bone area with a burr. Postoperatively, the traction is discontinued, and the patient is placed in a four-post hard cervical collar for 3 months; all physical activities involving the neck are restricted during this period. However, sitting and standing are encouraged, as during these maneuvers, the weight of the head helps in further impaction of the spacers and stability of the joint.

Although the biomechanical properties need to be evaluated further, the technique of facet distraction as described here simulates several successfully employed and evaluated interbody fixation techniques used for the subaxial spine. From our experience, it appears that distraction of the facets increases stability, as multiple ligamentous structures and axial loading of the spine contribute to stability. The extent of the stability achieved is significantly enhanced by the use of specially designed spiked spacers. The release of distraction after introduction of the spacers and bone graft assists in further impaction of the spikes of the spacers into the facets.
Fig. 14.3 Picture showing spacers of various sizes. The spacers are made of titanium and have spikes. There are multiple holes within the body of the spacers for assistance with bone healing. The spacers are tapered at the leading edge to assist insertion within the joint. The base of the spacer has a transverse slot for stability of a suitable osteotome during hammer insertion.

Fig. 14.4 Images of a 17-year-old male patient.

a Preoperative CT scan showing basilar invagination. Assimilation of the atlas can be observed.
b Preoperative CT scan showing the abnormal alignment of the atlantoaxial joint.
c Coronal image of the CT scan showing the craniovertebral junction and the joints.
d Axial section of a magnetic resonance imaging (MRI) scan showing marked rotation of the odontoid process. CVJ, craniovertebral junction.
Fig. 14.4e–i

e  Postoperative CT scan showing the reduction of the basilar invagination and improvement in the craniovertebral alignments.

f  Postoperative sagittal cut through the atlantoaxial joint showing marked realignment of the facets and the joint.

g  Postoperative sagittal cut showing the spacer in the atlantoaxial joint and realignment.

h  Postoperative coronal section showing spacers in both of the atlantoaxial joints.

i  Postoperative radiograph (lateral view) showing spacers in both of the atlantoaxial joints. Evidence of bone fusion is noted.
and stabilization. The distraction of the facets and impaction of the spacer and bone graft can be technically challenging, but they are certainly less complicated than interarticular and transarticular screw fixation methods. Because no screws are implanted, the problem of vertebral artery injury is avoided.

Between January 2003 and January 2007, six patients underwent the discussed method of fixation. All six patients had posttraumatic mobile and reducible atlantoaxial dislocation. The mean follow-up period was 16 months (range 5–47 months). Successful atlantoaxial stabilization and ultimate bone fusion were achieved in all patients and were documented with dynamic radiography. There were no neurological, vascular, or infective complications. During the follow-up period, all patients showed neurological recovery, and there was no indication of implant migration or failure, suggesting the effectiveness of the operation.

Because no wire, screws, plates, or rods were used for fixation, as is the norm in other fixation procedures, the extent of stability provided by the implant will have to be assessed by a larger series done over a longer period of time. The issues of the most appropriate size and type of spacer and the extent of optimum distraction necessary also will have to be evaluated further.

### Role in Basilar Invagination

Despite the fact that several techniques are now available for the treatment of basilar invagination, the search for the most appropriate option continues.

### Indications

Joint jamming and joint distraction using specially designed metal spacers can be an alternative form of treatment for group A basilar invagination and for patients having rotatory dislocation.

### Surgical Technique

The basic technique is the same as the one described above. Exposure of the joint in cases of basilar invagination, however, is more difficult, as the joint is positioned significantly rostrally. Basilar invagination has been considered by several authors to be a “fixed” anomaly. Dynamic imaging in this condition seldom shows evidence of abnormal movements of the atlantoaxial joint or increased compromise of the spinal canal dimension on flexion of the neck. Fixation of the region is recommended by several authors after transoral decompression, as bone resection and ligamentous laxity after surgery are said to destabilize the region. It appears to us that the instability of the atlantoaxial joint is the primary cause of basilar invagination. A history of trauma preceding the clinical events, a predominant complaint of pain in the neck, and improvement in neurological symptoms following the institution of cervical traction suggest “vertical” instability of the craniovertebral region. The process of basilar invagination has been likened to lumbosacral spondylolisthesis. Several authors have successfully treated lumbosacral spondylolisthesis using interbody cages/spacers, resulting in distraction, reduction, and fixation of the listhesis. The combined size of the facets of the atlas and axis is more than the size of the cervical vertebral bodies. The facets of the atlas and axis are significantly stronger and heavier than the cervical vertebral bodies. Meniscus and the articular cartilage by drilling and subsequent distraction of the joint. The stability of the atlantoaxial joint is not excessive in cases of basilar invagination, as in congenital mobile atlantoaxial dislocation; therefore, the spacer is more stable within the joint, and migration becomes less probable.

The procedure provides sufficient stability to the region, and any other fixation method can be avoided. However, if the surgeon finds the stability inadequate, all other described techniques of fixation, both midline and lateral, can be employed simultaneously. Although we still consider simultaneous fixation of the region with plates and screws in the lateral masses as the more optimum form of treatment, in our experience with select cases, the stability provided by the spacers was so firm that all other forms of fixation were avoided. In some cases of basilar invagination, exposure of the facet of the atlas for screw insertion may be technically difficult, particularly in cases with assimilation of the atlas. Because screw insertion in the facets of the atlas and axis is not involved, the technique becomes simpler, and vertebral artery injury is thus avoided.

The issues regarding the appropriate size and type of spacer, as well as the extent of optimum distraction necessary, will have to be further evaluated.

### Conclusion

The technique of joint distraction and jamming probably has some potential in the treatment of select cases of atlantoaxial dislocation and basilar invagination. Future improvements and innovations in the design of spacers and instrumentation will make this technique biomechanically more stable and surgeon friendly.
Fig. 14.5a–e  Images of an 11-year-old male patient.
a  Preoperative MRI showing basilar invagination (transoral surgery was already attempted) and cord changes.
b  Coronal MRI showing marked rotation of the craniovertebral junction.
c  Sagittal section of CT scan showing basilar invagination and fixed atlantoaxial dislocation.
d  Sagittal image showing a spacer in the atlantoaxial joint.
e  Coronal section showing spacers in both of the atlantoaxial joints and distraction.
Fig. 14.6a–g Images of a 15-year-old female patient.

a Preoperative MRI showing basilar invagination and fixed atlantoaxial dislocation.
b Preoperative CT scan showing relatively minor basilar invagination and fixed atlantoaxial dislocation.
c Postoperative CT scan showing incomplete but significant reduction of basilar invagination and atlantoaxial dislocation.
d Sagittal image through the atlantoaxial joint showing the spacer.
e Sagittal image through the joint showing the distraction and opening up of the joint.
References


Fig. 14.6f–g
f Coronal image of CT scan showing the spacers in both of the atlantoaxial joints.
g Postoperative radiograph showing the spacers in the joints.
Typical pathologies for occipitocervical fusion (OCF) include congenital and developmental malformations, rheumatoid arthritis, primary bone tumors, metastatic disease, and trauma. Although rates of OCF have decreased with the advent of effective medical therapy for inflammatory arthritides, OCF will remain an essential tool for spine surgeons in select cases. Posterior instrumentation specifically designed for OCF is available from several vendors, and assorted fixation strategies have been advocated and tested biomechanically. Some authors have reported novel anterior constructs, although these have not gained widespread popularity. This chapter touches briefly on the history of OCF and relates preferred methods.

## Background

Initial experience with occipitocervical fusions reflected the need to address decompression and stabilization in patients with rheumatoid arthritis with compressive myelopathy secondary to basilar invagination, ventral pannus, or instability at the atlantoaxial joint. Some early reports also relate experience treating congenital and developmental anomalies. Given the progressive nature of rheumatoid arthritis and the deleterious effects of corticosteroids on bone, many surgeons advocated fusion at the first sign of clinical symptoms (myelopathy and neck pain) and/or demonstration of instability. The procedure was often applied to patients with pure C1–C2 instability because of the high rate of pseudoarthrosis with C1–C2 fusions. Surgeons argued that the added immobility from incorporation of the occipitoatlantal joint in a C1–C2 construct was clinically inconsequential. Lacking advanced instrumentation, early fusion strategies ranged from non-structural onlay grafts of morcellized iliac crest to wired cortical strips of the ilium or tibia, followed by lengthy immobilization and traction.

Early results with patients with rheumatoid arthritis—which arose as the index population for this procedure—were disappointing, with high rates of pseudoarthrosis/reoperation and high morbidity and mortality. As the development of fixation techniques continued, various surgeons reported their experience with wiring techniques and supplementation with acrylic. In the 1980s, rigid plate and loop systems using wires or cables for fixation were introduced. The incorporation of rigid internal devices led to earlier mobilization and decreased the need for external immobilization. Ultimately, success with screw fixation was reported. For the cervical end of the fusion, Grob et al. reported the use of C1–C2 transarticular screws, and Goel and Laheri reported the use of a C2 pars screw in isolation or in combination with C1 lateral mass screws. Both groups fixed the occipital end of the implant with screws. Today successful outcomes with OCF are related to improvements in techniques and instrumentation, as well as a changing patient population. One recent surgical series reported an 87% rate of improvement in myelopathy and a 97% fusion rate in a group of 69 patients undergoing OCF for a variety of indications. Goel and Kulkarni reported atlantoaxial lateral mass plate and screw fixation even in cases where there was assimilation of the atlas.

The current trend toward a reduction in the implementation of occipitocervical fusion has been heavily influenced by the advent of effective medical therapy for rheumatoid arthritis, the employment of highly effective C1–C2 fusion strategies, and the recognition of significant morbidity from immobilization of the craniovertebral junction. The first two developments have significantly reduced the number of patients for whom OCF is indicated, and the third has pushed surgeons to opt for OCF only as a last resort. Still, craniovertebral instability is unlikely to disappear completely, and, as such, OCF is likely to remain an essential tool for a subset of patients with relatively severe and complex pathology.

## Indications

The indications for OCF are globally defined as pathological conditions resulting in instability of the articulation of the atlas and the skull base and the treatment of an iatrogenic instability resulting from a decompressive procedure. Typical disease processes that produce such situations are inflammatory arthritis (in particular, rheumatoid arthritis), trauma (occipital condyle–C1 [0–C1] dislocation and complex high cervical fractures), malignancy (primary bone tumors or metastases), congenital malformations (dysgenesis of the C1 lateral masses and occipital condyles), and developmental disorders (Grisel and Down syndromes). The anatomical abnormality can range from a reducible instability that requires reduction
and fixation, either in the vertical plane, such as basilar invagination with rheumatoid arthritis, or in the sagittal plane, such traumatic O–C1 dislocation, to a compressive pathology whose surgical decompression results in instability requiring fusion (rheumatoid pannus and malignancy). Often the decision involves the relative merits of extension of a high cervical construct to the occiput. For example, in cases of complex C1–C2 fractures, where C1–C2 instability is combined with destruction of the C1 lateral masses, OCF may be considered as a primary procedure or as a backup strategy.

## Instrumentation

Current OCF strategies usually entail fixation of a rigid contoured rod to multiple points in the occipital bone and C2 at a minimum, with the addition of other fixation points as needed depending on pathology, technique, and surgeon preference. Fixation may be achieved using a variety of techniques.

### Rod/Wiring Technique

An older but still effective technique is a pure rod and cable construct. This technique is attributed to Ransford et al. and was discussed by Menezes. It can be accomplished with minimal equipment and at low cost. Exposure of the craniovertebral junction and the cervical spine down to C3 is followed by a small suboccipital craniectomy of ~2 × 2 cm, saving bone for subsequent arthrodesis. Burr holes are drilled around the edge of the craniectomy to create fixation points for cables in the occipital bone. Menezes prefers four burr holes of 6 to 8 mm placed in the occipital bone, two rostral and two lateral to the craniectomy. The rostral burr holes are 1 cm off the midline and 1 cm above the craniectomy, and the lateral burr holes are 1 cm above the foramen magnum and 1 cm lateral to the craniectomy. The dura is freed between the craniectomy and the burr holes and under the lamina of the vertebra to be fused. Sublaminar cables are passed under C1, C2, and C3 (if necessary). A loop titanium rod, which is preconformed. Sublaminar cables are passed under C1, C2, and the burr holes and under the lamina of the vertebra to accommodate the rod vector dictated by the trajectory of the cervical fixation points.

### Screw Rod/Plate Constructs

Although cabling remains an important tool, most surgeons prefer screw fixation points because of increased rigidity and durability. Several current instrumentation systems have been specifically designed for OCF, and these offer the convenience of contoured rods and malleable plates. All of these constructs offer rigid occipital bone screw fixation with integration into cervical screw rod/plate constructs. The use of screws in the cervical spine, however, remains “off-label” according to the U.S. Food and Drug Administration. Designs vary mostly in terms of whether a single-piece contoured loop is used versus two curved rods whose biomechanics are integrated with cross-linkers. The significant technical advance in dedicated OCF systems lies in the ability to create rigid, integrated constructs with multiple screw fixation points in the cervical spine and the occipital bone.

### Occipital Bone Screws

Perhaps the greatest advancement in occipitocervical instrumentation has been the development of occipital bone screws. The thickness of the occipital bone is greatest in the midline; hence, the term occipital keel. The strength of the cortical bone in this area is reflected in a unicortical screw pullout threshold that is equivalent to bicortical screws elsewhere in the cervical spine. Although bicortical occipital screws are twice as strong, they are associated with such complications as sinus and cerebellar injuries, and cerebrospinal fluid leak. Given the strength and safety of unicortical screws, most surgeons feel that bicortical occipital purchase may be desirable but is not routinely necessary.

Placement of occipital screws is advised just off the midline to take advantage of the thickest part of the occipital bone. Anatomical studies have supported placement of occipital bone screws at least 1 cm off midline and 1 cm inferior to the superior nuchal line to avoid sinus injury. Screws placed too laterally or too inferiorly may encounter thin bone to the extent that the bone thickness may exceed the shortest screw length in many systems. Generally speaking, it is advisable to place cervical fixation prior to the occipital screws, as these can be placed to accommodate the rod vector dictated by the trajectory of the cervical fixation points.

### C1 and C2 Fixation

Although extension of OCF constructs below C2 may be desirable for individual pathologies, it is not routinely necessary. Immobilization of the occiput relative to C2 is required at a minimum, and a variety of strategies have been described. One popular strategy has been a combination of occipital screws and C1–C2 transarticular screws. This construct has the advantage of direct fixation of all three segments—occiput, C1, and C2—and the pullout resistance conferred by fixation through four cortical surfaces at the caudal anchor. C1–C2 transarticular screws are believed to confer more risk to the vertebral arteries, however, and
their necessity, particularly in adult patients, has been called into question.

A variety of other strategies are possible and may include combinations of other fixation types, such as C1 lateral mass screws, C2 pars screws, and C2 laminar/spinolaminar screws. Many biomechanical studies have examined the relative impact of incorporation of these fixation strategies into occipitocervical constructs with occipital bone screws. These studies have failed to find significant differences when comparing constructs with occipital screws and C1–C2 transarticular screws versus C2 pars screws, or C1–C2 Magerl transarticular screws versus Goel C1 lateral mass/C2 pars screws. One study of OCF constructs that compared cervical cabling with various screw fixation strategies, including C1–C2 transarticular screws, C2 pars screws, and C2 laminar screws—each with and without C1 lateral mass screws—found very similar immobilization profiles among all of the screw constructs and minimal impact with the addition of C1 lateral mass screws. Perhaps the most noteworthy conclusion of these studies is the high performance of C2 pars-occipital screw constructs without C1 fixation, a construct that is considered the safest, most straightforward of all options. Ultimately, there is no construct that can be applied universally, as constructs need to be tailored to each patient. Choices may be limited by patient anatomy. Highly unstable patients may prompt the surgeon to use additional fixation points, either caudally or at C1. Individual pathologies may preclude certain fixation options. Where fixation points are desired and screws are not possible, hooks and cables may be needed.

### Instrumentation Systems

#### Overview

Several instrumentation companies offer hardware specifically designed for OCF. The majority of these involve dedicated semimalleable plates for occipital fixation that can connect to cervical and thoracic fixation points via rods. These systems are all very similar in principle and offer the ability to generate relatively seamless constructs from the occiput to the cervical and thoracic spine. Evolution of these systems can be traced to an effort to address two specific technical hurdles. The first involves contouring the occipital plate and rod to fit individual patient anatomy. The second relates to designing a rod/plate system with the flexibility to join fixation points in the occiput that are midline or parasagittal and fixation points in the cervical spine that are significantly more lateral. Some of the major spinal instrumentation companies’ products are described below. This list is not exhaustive. Companies that offer similar systems are Biomet Microfixation Inc. (Jacksonville, Florida), Aesculap Implant Systems Inc. (Center Valley, Pennsylvania), and Zimmer Spine Inc. (Minneapolis, Minnesota).

### Vertex Max

The Vertex Max system (Medtronic Sofamor Danek, Inc., Memphis, Tennessee) consists of a precontoured plate with a 3-mm rod (Fig. 15.1a) and an independent occipital plate with midline and lateral screw insertion sites (Fig. 15.1b). The occipital plate has lateral slotted connectors to facilitate attachment to the rod from the cervical portion of the construct. A custom set with “inside out” occipital screws is also available. This system offers multiaxial screws in 3.5-, 4.0-, and 4.5-mm diameters that permit up to 45° of angulation. The cervical rod is 3.2 mm in diameter. Also, a partially threaded multiaxial screw was designed for C1 lateral mass fixation that theoretically may reduce C2 nerve root irritation.

### Mountaineer

The Mountaineer system (DePuy Spine Inc., Raynham, Massachusetts) includes an inverted Y-shaped occipital plate, favored-angle polyaxial screws, and medial/lateral-angled screws, as well as an adjustable hinged occipitocervical rod (Fig. 15.2a). The occipital plate allows for three midline screws and comes with an optional washer extension for additional occipital fixation points.
The occipital plate comes in three sizes (31, 37, and 45 mm), is malleable, and has two rod connectors that can rotate and slide 4 mm to facilitate rod attachment. The favored-angle polyaxial screws are available in 3.5- and 4.0-mm diameters, and the polyaxial head provides a 60° cone of freedom with an additional 15° at certain angles. The medial/lateral-angled screws are available in 3.5-, 4.0-, and 4.35-mm diameters. A partially threaded C1 screw is also available. In addition to the hinged occipitocervical rod, precontoured 3.5-mm rods are available.

**Oasys**

The Oasys system (Stryker Spine, Allendale, New Jersey) consists of two separate occipital plates, each of which has four anchor holes and one slot for placement of occipital screws. Each plate has a proximal foot that connects and locks onto the cervical rod (Fig. 15.3). The plate-to-foot angle is fixed at 100° or 130°, eliminating the need for manual rod contouring. The plate is also malleable. Polyaxial screws are available in 3.5- and 4.0-mm diameters and allow for conical screw angulation of up to 110°. The
cervical rod diameter is 3.5 mm, and a partially threaded C1 screw is available as well.

**Protex-CT**

The Protex-CT system (Globus Medical, Inc., Audubon, Pennsylvania) offers small occipital plates with holes for one or two occipital bone screws. These can be combined as needed for occipital bone fixation (Fig. 15.4). Screws are available in 3.5-, 4.0-, and 4.5-mm diameters that offer 80° total angulation. Prebent rods are available, as are tapered transition rods for the cervicothoracic junction that range from 3.7-mm diameter to 4.0- to 6.5-mm diameter in 0.5-mm increments.

**Axon**

The Axon system (Synthes, Inc., West Chester, Pennsylvania) features a rod/plate with four holes that transitions to a 3.5-mm rod for cervical fixation. The plate is malleable. This system provides polyaxial 3.5- and 4.0-mm self-tapping screws with 60° cone angulation. A partially threaded C1 rod is available, as are 3.5/5.0-mm and 3.5/6.0-mm dual diameter rods for cervicothoracic constructs.

**OctaFix**

The OctaFix system (Abbott Spine, Inc., Austin, Texas) consists of a contoured horseshoe-shaped plate that is contiguous with two rods. The rods can be cut depending on the length of the cervical portion of the construct. The plate has three midline holes and three lateral holes on each side for placement of occipital screws, which are available in 3.5- and 4.0-mm diameters; 4-mm screws for C1–C2 fixation are 4 mm in diameter and attach to the rods via grooved connectors that allow for unrestricted sagittal and coronal angulation.

**Conclusion**

Although the prevalence of patients with instability of the craniovertebral junction requiring occipitocervical fusion has declined, OCF will remain an essential tool for spine surgeons. Currently available instrumentation systems have greatly facilitated this endeavor through the development of hardware designed specifically for this anatomical region. Important biomechanical studies exist to help guide the surgeon in selecting the appropriate fixation strategy for each patient. These advancements are reflected by improvements in outcomes in recent clinical series.

**References**


The transoral approach has been used to address disorders of the craniocervical junction. Over the past half-century, several surgeons have described their experience with the approach and reported varying degrees of success. In this chapter, the transoral approach for decompression and resection of the odontoid is discussed. The indications for transoral resection of the odontoid process are in selected cases having basilar invagination, nonreducible bony compression of the spinal cord, or soft tissue pannus causing severe ventral compression and resulting in spinal cord contusion with rapidly progressive myelopathy. Ventral soft tissue pannus causing compression without cord contusion or rapidly progressive myelopathy may be treated instead with a posterior decompression/fusion, which typically leads to a reduction of the size of the pannus over time. The transoral approach for intradural lesions is controversial.

Modifications to the traditional transoral approach have been described by various authors, including Menezes, Crockard, and Sonntag, in an effort to improve exposure and reduce complications. In this chapter, we outline the advantages and disadvantages of the transoral technique based on past experience.

### Operating Room Configuration and Patient Positioning

Operating room setup and positioning of the patient are critical to the success of the transoral approach. Prior to intubation, the patient is positioned on the operating table awake with his or her head in extension on a horseshoe headrest in the position required for surgery. In rare instances, a Mayfield head holder or halo ring may be used if instability is of concern. Awake hyperextension allows the surgeon to optimally position the neck while also ensuring the safety of positioning from a neurological standpoint. Maintaining a neutral, nonrotated head position is critical to maintaining orientation intraoperatively. Subsequently, an awake, fiberoptic oral intubation is performed with an armored endotracheal tube.

In general, tracheostomy is rarely necessary in patients who will undergo transoral surgery. Prophylactic tracheostomy is performed only in patients who have Down syndrome or those with small oral apertures (≤3 cm diameter, depending on the rostrocaudal extent of the pathology to be decompressed). These patients have both redundant posterior pharyngeal wall tissue and a decreased ability to coordinate their pharyngeal musculature following odontoidectomy.

Patients are given high-dose penicillin or clindamycin prior to starting surgery and in the perioperative period. Dexamethasone may also be given prior to surgery to decrease both neural and airway edema.

In patients with severe myelopathy, neural monitoring consisting of somatosensory evoked potentials (SSEPs) and/or motor evoked potentials (MEPs) is employed. When neuromonitoring is used, a preoperative baseline study is obtained prior to incision.

Cranial traction may be used if necessary to optimize anatomical reduction of the craniocervical junction (Fig. 16.1); in most cases, we do not need to employ traction.

To maximize ergonomics for a right-handed surgeon, the patient is positioned with his or her left side toward the anesthesiologist. The endotracheal tube is placed in the midline of the mouth. The C-arm fluoroscope is positioned to obtain cross-table lateral cervical images. The surgeon stands behind the patient, and the scrub nurse is positioned to the right side of the patient. The operating microscope base is positioned to the left-hand side of the surgeon (Fig. 16.2).

### Operative Considerations

Patients with nonreducible craniocervical junction bony compression or ventral soft tissue compression with spinal cord contusion are candidates for transoral odontoidectomy. The body habitus of the individual patient must be taken into account before embarking upon the procedure. The patient should be able to open his or her mouth widely enough to accommodate the transoral retraction systems. In our experience, the minimal jaw excursion to perform an odontoidectomy is 3 cm or, as a good rule of thumb, two fingerbreadths. A working corridor of this size will provide the surgeon with access from the inferior clivus to the C2–C3 interspace. Some patients with more limited jaw excursion may require transmaxillary or transmandibular approaches. Patients must have good dentition, and oral hygiene must be optimized to minimize the risk of infectious complications. Patients who satisfy these criteria are good candidates for transoral odontoidectomy.

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Surgical Technique for Transoral Odontoidectomy

A Spetzler-Sonntag or similar retractor is placed with an appropriate tongue blade to retract the tongue and endotracheal tube inferiorly. Care is taken to avoid trapping the tongue against the teeth. The retractor is opened as widely as possible to allow maximum exposure of the posterior pharyngeal area (Fig. 16.3). The anesthesiologist administers agents that create neuromuscular blockade to allow for wider mandibular opening.

Once the Spetzler-Sonntag retractor is in place, a red rubber catheter is placed through one of the nares and is sutured to the uvula (Fig. 16.4). The catheter is then pulled up through the nose, thus retracting the uvula and soft palate superiorly; this retraction improves exposure of the upper portion of the posterior pharyngeal wall overlying the tip of the odontoid and prevents secretions from flowing into the incision. This maneuver is a critical step, allowing the surgeon to avoid making soft palate incisions in most cases. It is the soft palate incision that results in postoperative dysphagia and dysphonia in some patients.

The superior soft palate retractor on the Spetzler-Sonntag system is applied after the uvula is retracted superiorly to provide even further superior exposure...
Fig. 16.3 Artist’s illustration of placement of the Spetzler-Sonntag retractor. The tongue and endotracheal tube are retracted inferiorly. Care is taken to avoid trapping the tongue against the teeth. Note that the soft palate and uvula are still obstructing the surgeon’s view of the dens (arrow).

Fig. 16.4 Artist’s illustration from the operating surgeon’s viewpoint of the placement of a red rubber catheter through a naris with attachment of the distal end to the uvula with a suture. Retraction of the red rubber catheter allows for elevation of the uvula and soft palate.
At this point, a clear view from the inferior clivus to C2 is obtained.

The posterior pharynx is then infiltrated with 1% lidocaine with epinephrine. Subsequently, lateral fluoroscopy is used to identify the area of the posterior pharyngeal mucosal wall overlying the odontoid process. The anterior tubercle of C1 may also be palpated. The incision is typically 1.5 to 2 cm in length and is carried through the posterolateral pharyngeal constrictor muscle in the midline raphe (Fig. 16.6) over C2 and the anterior tubercle of C1. A full-thickness incision must be made through the mucosa, and the integrity of the superolateral mucosa should be maintained to facilitate closure. Our initial incision is shorter than what is ultimately needed for the odontoidectomy because the retractors that are subsequently placed tend to extend the rostrocaudal length of the incision when they are opened. Once the incision has been created, we do not touch the remainder of the oral cavity outside the incision. We avoid tracking secretions and oral bacteria into the incision with this “no touch” technique.

The Crockard self-retaining retractor is placed in the midline pharyngeal incision to retract the mucosal and pharyngeal constrictor muscles and spread laterally to expose the anterior arch of C1. We then use electrocautery to skeletonize, in a subperiosteal fashion, the anterior surface of the arch of C1 (Fig. 16.7). The fluoroscope is again used to confirm anatomical landmarks. The use of image guidance is optional, and in most cases we do not use computerized neuronavigation systems.

Once the arch of C1 has been exposed (Fig. 16.8), we identify the midline and drill and remove the anterior arch of C1 to expose the anterior portion of the odontoid process. The arch removal should be wide enough to expose the shoulders of the odontoid process; this typically requires removal of two thirds of the anterior arch of C1. Inadequate resection of the anterior portion of the C1 ring is a common mistake. The shoulders of the odontoid process are a critical landmark for the surgeon and must be visualized. Preoperative computed tomography (CT) scans should be obtained to ensure that there are no variations in the normal anatomical course of the vertebral artery in the foramen transversarium of C1 and C2.

After exposure of the odontoid process, an angled curet is used to detach the apical and alar ligaments at the top of the odontoid process. Fluoroscopy is used to identify how far posteriorly the angled curet may be placed without violating the spinal canal. These ligaments act as a tether and will prevent removal of the tip of the dens if they are not severed. Removal of these ligaments at the apex of the dens establishes the superior boundary for bone removal and allows for subsequent resection of the top of the odontoid process. In patients with significant basilar invagination, accessing and removing these ligaments can be difficult; traction is often necessary in these cases to gently mobilize the top of the odontoid process inferiorly.

Once the ligaments have been detached, a “top-down” removal of the odontoid process is performed by drilling the dens using an eggshell drilling technique with the Midas...
Fig. 16.6 Illustration from the surgeon’s viewpoint after placement of the Spetzler-Sonntag retractor blades. The soft palate has been elevated by the superior blade. The red rubber catheter has been tractioned and is secured to the Spetzler-Sonntag frame with a clamp. A linear incision is then created in the midline posterior pharyngeal wall overlying the area of the odontoid.

Fig. 16.7 Artist’s illustration from the operating surgeon’s viewpoint. The Crockard retractor is placed in the pharyngeal incision and spread laterally to expose the anterior arch of C1. The Bovie cautery is used to skeletonize the anterior arch of C1.
resorption of the soft tissue pannus following posterior fixation\textsuperscript{13,19–21}; therefore, aggressive resection of the pannus may not be necessary if posterior fixation is to be placed. Because posterior fixation is indicated for most patients, complete resection of the soft tissue pannus, if present, is not required. All loose fragments of soft tissue pannus are resected, but the deeper layers on the surface of the dura are left behind. Aggressive removal of soft tissue pannus risks injury of the underlying dura and potential leakage of cerebrospinal fluid (CSF). CSF leakage in the setting of the transoral odontoidectomy may result in meningitis, which can be fatal.

Once we feel that we have achieved an adequate removal of bone and pannus, we then inject iohexol dye into the resection cavity and obtain a lateral fluoroscopic radiograph to confirm the extent of our decompression. The spread of the dye helps to reveal any remaining remnants of the dens. In cases where neuronavigation is used, the image guidance system is helpful to identify any remnants of the dens. If we are satisfied with the bone removal, we then proceed with closure.

The posterior pharyngeal mucosa and muscle are closed by reapproximating them with interrupted 3.0 chromic suture in a single- or double-layer fashion. The posterior pharynx is irrigated with a small amount of antibiotic solution. Bone wax is not used, as the wound is considered contaminated, and the wax theoretically could provide a nidus for infection. Only the incision is irrigated, not the entire oral cavity. Excessive oral irrigation may allow fluid to enter the airway and lungs.

The nasal red rubber catheter is removed. The uvula falls into its original place. To allow for nutrition after a surgery, a Dobhoff feeding tube is then passed through one of the nares and down into the esophagus under direct vision while the Spetzler-Sonntag retractor is still in place. The Dobhoff tube is stitched to the lateral wall of the nostril to prevent accidental dislodgment. The retractor systems are then removed, and the tongue and lips are inspected. Cortisone cream is typically applied to the tongue and lips to reduce postoperative swelling. The tongue is massaged to restore circulation. Typically, the tongue retractor is not released during the procedure because the operation can be performed in $\approx 90$ minutes. Instances of tongue edema following surgery typically resolve within 2 or 3 days.

The instability created by the removal of the odontoid process\textsuperscript{9,10,22} has been established in several previous studies. Therefore, posterior stabilization is performed as a second procedure on the same day, immediately following the transoral odontoidectomy. This is performed without changing the endotracheal tube. The patient is fixedated in skull clamps and positioned prone onto chest rolls, on a separate operating table for the posterior portion of the procedure. For cases with significant basilar invagination or occipitocervical instability, an occipitocervical fusion is indicated.\textsuperscript{23} For cases without significant basilar invagination and only C1–C2 instability, Magerl’s transarticular C1–C2 screw fixation or Goel’s C1 lateral mass screws with

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**Fig. 16.8** Artist’s illustration from the surgeon’s viewpoint of the exposed anterior arch of C1. Drilling of the anterior arch can now commence.
C2 pars/pedicle screw constructs\textsuperscript{15-23} are preferable. Posterior fusions should incorporate iliac crest autograft or rib autograft.

After completion of both anterior and posterior procedures, the patient is monitored in the intensive care unit (ICU). Patients typically remain intubated in the ICU for 2 or 3 days. They are extubated only after a cuff leak (breathing around a deflated endotracheal cuff) is confirmed in the ICU on postoperative day 2 or 3. The anesthesiologist typically removes the endotracheal tube over a tube changer, which is left in place for a minimum of 1 hour. The tube changer allows for easy reintubation should the need arise.

\textbf{Surgical Technique for Transoral Odontoidectomy with Mandibular Split}

In rare cases, the transoral approach may be combined with a mandibular splitting procedure to obtain additional exposure of the C1–C3 region. This approach is useful for cases of radical tumor excision in that region.

The procedure is best performed by a multidisciplinary team, with the mandibular split performed by ear, nose, and throat specialists. The mandibular split requires the creation of an incision on the lip and chin. A midline incision of the lower lip is made and then curved around the chin in a C shape. A self-retaining retractor is then placed, and a wire saw is used to divide the mandible in the midline (between the lower incisors). Once split, the mandible can be spread apart to offer a broader exposure of the oral cavity.

The tongue is split in the midline raphe using a low setting on the electrocautery. The two halves of the tongue are then spread apart; this allows for maximum access to the posterior pharyngeal wall. The posterior pharyngeal wall is also opened with a curvilinear incision rather than the standard midline mucosal incision previously described. The purpose of this change is to avoid pressure on the incision if a cage is to be implanted for structural support in the midline of the C1–C3 area.

Once the resection of the C1–C3 area is complete, the mucosa is closed using the technique previously described. The tongue is reapproximated using resorbable suture, and the mandible is plated together with mini-plates and screws. The lip and chin incision are also closed with resorbable suture.

\textbf{Complication Management}

\textbf{Cerebrospinal Fluid Leaks}

CSF leaks encountered during the course of a transoral surgery can have potentially devastating consequences. Meningitis due to oral bacteria invading the CSF and resulting in death has been reported with this complication.\textsuperscript{8} To avoid CSF leaks, the aggressive removal of the soft tissue pannus often seen in patients undergoing transoral odontoidectomy should be avoided. Any loose pannus tissue is typically removed, but it is not mandatory to remove all pannus up to the dura. Studies have shown spontaneous resolution of the pannus following posterior fusion and stabilization in these patients.

If CSF leakage occurs during surgery, the leak is sewn closed. Fibrin glue or a dural sealant and a small piece of synthetic dural substitute are placed over the area of the leak to reinforce the repair. Serious consideration should be given to placement of a lumbar drain in patients with CSF leaks. However, excessive CSF drainage with a lumbar drain may create negative pressure at the pharyngeal incision, which may pull oral secretions and bacteria into the spinal fluid pathway.

\textbf{Vertebral Artery Injury}

The surgeon must always be cognizant of the location of the vertebral artery in relation to the planned bony resection of the arch of C1. A preoperative CT scan of the occiput to C3 is obtained to assess for any anatomical variations in the position of the foramen transversarium of C1 and C2. Occasionally, a more medially positioned foramen will be identified on these studies, and the surgeon must be aware of this anatomy to avoid drilling into the foramen and risking injury to the vertebral artery. This danger is especially high in patients who have a rotary subluxation at C1 and C2. In addition, the surgeon should be aware that the vertebral artery typically takes an anteromedial course at the level of the C2–C3 disk, and caution must be exercised to avoid lateral dissection at the level of the C2–C3 disk.

If the vertebral artery is injured during the removal of the anterior arch of C1 or the dens, the bleeding should be packed off and tamponaded with Gelfoam. A direct repair of the vessel should not be attempted. An intraoperative (if available) or immediate postoperative angiogram is then performed to evaluate the vessel injury and for consideration of endovascular occlusion of the vessel.

\textbf{Spinal Cord Injury}

Patients with severe myelopathy are at risk of worsening their motor function following any spinal decompressive procedure. To minimize this risk, patients are positioned while awake on the operating table to ensure they do not experience any new neurological symptoms when placed in the position for surgery. In addition, SSEP and MEP monitoring is employed during surgery on patients with severe myelopathy.

The importance of maintaining a patient’s preoperative mean arterial pressure at all times during induction and surgery should be impressed upon the anesthesia
staff preoperatively. Severely myelopathic patients typically undergo placement of an arterial line prior to surgery to allow for intensive monitoring of their mean arterial pressure. Avoiding hypotension in such patients is of paramount importance, as even temporary hypotension can cause cord ischemia, resulting in a worsening of the neurological state.

## Conclusion

The transoral technique is a viable and useful option in the treatment of irreducible ventral craniocervical junction bony compression. The technique described provides excellent exposure to achieve the surgical objective while minimizing potential complications. As with many approaches, the surgeon experiences a learning curve with this technique, but with careful planning and meticulous technique, the procedure may be performed with excellent outcomes.

## References

Advancements in skull base surgical techniques and improved understanding of microsurgical anatomy permit aggressive surgical excision of the lesions at the craniovertebral junction. The anterolateral, lateral, and posterolateral approaches provide a wide, sterile operative field, as opposed to the potentially contaminated oral space, and are preferred by many neurosurgeons. In addition, recent developments in spinal instrumentation provide neurosurgeons with many opportunities and avenues of posterior craniocervical fixation, such as Magerl’s C1–C2 transarticular fixation and Goel’s C1 lateral mass and C2 pars screws with plate or rod.

To perform these procedures safely, management of the vertebral artery is the most fundamental surgical step. In this chapter, we explain the microsurgical anatomy of the vertebral artery and the suboccipitocervical muscles and present the surgical techniques to expose and transpose the vertebral artery.

### Surgical Anatomy of the Vertebral Artery

The vertebral artery is divided into four segments. V₁ runs from the origin at the subclavian artery to the transverse process of C6. V₂ lies between the transverse foramen of C6 to C2. V₃ is from the transverse foramen of C2 to the dural entrance at the foramen magnum. V₄ is the intradural part.

Understanding the microanatomy of V₃ is especially essential for surgical approaches to lesions at the craniovertebral junction (CVJ). After passing through the transverse foramen of C2, V₃ runs upward and passes through the C1 transverse foramen (vertical portion), then turns medially to traverse above the lateral aspect of the posterior arch of C1. It enters the vertebral canal by passing below the lower, arched border of the posterior atlanto-occipital membrane (horizontal portion) and finally goes obliquely upward and pierces the dura mater at the foramen magnum (oblique portion). All along V₃, the vertebral artery is surrounded by the vertebral venous plexus and a periosteal sheath covering the plexus. At the dural penetration of the vertebral artery, the periosteal sheath is in continuity with the outer layer of the dura and also tightly adherent to the adventitia of the artery, making a sort of distal fibrous ring. The adventitia of the vertebral artery is fixed at only two segments; one is at this part, and the other is at the transverse process of C6. This anatomical characteristic prevents injury to the vertebral artery from various neck movements.

Muscular branches originate from the horizontal portion of V₃ to supply the deep paravertebral muscles. Some of them may need to be divided to expose the entire course of V₃ or to mobilize and transpose the segment. The posterior meningeal artery also originates at V₃ just before dural penetration. In rare cases, the posteroinferior cerebellar artery has an extradural origin. This may be evaluated preoperatively to avoid an ischemic complication (Fig. 17.2).

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**Fig. 17.1** Illustration demonstrating the anatomy of the vertebral artery. The V₁ segment of the vertebral artery is from the origin at the subclavian artery to the transverse process of C6. V₂ runs from the transverse process of C6 to the transverse process of C2. V₃ is from the transverse process of C2 to the dural entrance at the foramen magnum. V₄ is the intradural part.
Surgical Anatomy of the Suboccipital and Posterior Paravertebral Muscles

Although individual muscles need not be dissected separately at surgery, precise anatomical knowledge of all suboccipital and posterior paravertebral muscles, including their origin and insertion, is indispensable for operative exposure and management of the vertebral artery.\(^1\) \(^{10}\) The suboccipital triangle is the most important surgical landmark for exposure of \(V_3\). This triangle comprises three deep muscles: the superior oblique, rectus capitis posterior major, and inferior oblique. The superior oblique muscle originates at the inferior nuchal line and inserts on the transverse process of C1, the rectus capitis posterior major muscle originates from the inferior nuchal line and inserts on the spinous process of C2, and the inferior oblique muscle connects the transverse process of C1 to the spinous process of C2. The floor of the triangle is formed by the posterior atlanto-occipital membrane and the posterior arch of C1. In the triangle, the horizontal portion of \(V_3\) and the C1 nerve root lie in a groove on the upper surface of the lateral part of the posterior arch of C1 (Fig. 17.3a).

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**Fig. 17.2a, b** Illustrations showing the detailed surgical anatomy of \(V_3\) and surrounding structures. The distal side of the horizontal (HP) and oblique portion (OP) of \(V_3\) is covered by the posterior atlanto-occipital membrane (PAOM). PCEV posterior condylar emissary vein, VP vertical portion of \(V_3\), VVP vertebral venous plexus, YL yellow ligament.

**Fig. 17.3a, b**

a. Illustration delineating the relationship between the vertebral artery and the suboccipital triangle. IO inferior oblique muscle, IJV internal jugular vein, PAOM posterior atlanto-occipital membrane, RCPM rectus capitis posterior major muscle, SO superior oblique muscle, YL yellow ligament.

b. Illustration demonstrating the deep muscles that attach to the transverse process of C2. IJV internal jugular vein, LS levator scapulae muscle, RCL rectus capitis lateralis muscle, SC splenius cervicis muscle.
Understanding of the anatomy of the deep muscles inserting on the C1 transverse process is also essential when the vertebral artery is exposed from the anterolateral or lateral direction. The rectus capitis lateralis muscle running from the jugular process to the C1 transverse process serves as a natural barrier to the internal jugular vein located just in front of the C1 transverse process. The splenius cervicis and levator scapulae muscles are landmarks for the vertical portion of V3 and the C2 ventral root (Fig. 17.3b).

Exposure and Transposition of the Vertebral Artery

For CVJ surgery, the vertebral artery can be exposed using two different techniques of muscle dissection. In the anterolateral or lateral approach, following a lateral neck incision, deep muscles inserting on the C1 transverse process are dissected and reflected in a single layer in a lateral-to-medial direction. In the posterolateral approach, a medial-to-lateral reflection of the deep suboccipital and cervical muscles is performed. This technique is relatively straightforward and is described here.

The patient is placed in the lateral “park bench” position, with the patient’s head fixed using a three-pin head holder. The head position should be decided according to the location and extension of the region, but lateral neck extension is critical to provide a wide surgical space between the foramen magnum and the C1 posterior arch. The skin incision starts in the midline at the level of the C4 spinous process and is then directed upward to the external occipital protuberance. It turns laterally just above the superior nuchal line to reach the base of the mastoid process, then turns downward along the posterior ridge of the mastoid process to end just below the tip of the mastoid process (Fig. 17.4). The suboccipital and paraspinal muscles are dissected by splitting the nuchal ligament in the midline at the level of the C4 spinous process and is then directed upward to the external occipital protuberance. It turns laterally just above the superior nuchal line to reach the base of the mastoid process, then turns downward along the posterior ridge of the mastoid process to end just below the tip of the mastoid process (Fig. 17.4). The suboccipital and paraspinal muscles are dissected by splitting the nuchal ligament in the midline and are then gently reflected subperiosteally from the occipital bone, the C1 posterior arch, and the C2 lamina in a medial-to-lateral direction. This subperiosteal reflection gradually exposes the atlanto-occipital membrane, a tough, thick membrane connecting the inferior margin of the foramen magnum and the superior margin of the C1 posterior arch. The yellow ligament is also revealed between the C1 posterior arch and the C2 lamina (Fig. 17.5). Sharp longitudinal incisions on these membranes are initiated in the midline, then extended laterally along the inferior margin of the foramen magnum, the superior and inferior margin of the C1 posterior arch, and the superior margin of the C2 lamina. This incision is the first step in safely exposing V3 (Fig. 17.6a). The open door–like flap of the incised membrane, which is not adherent to the underlying epidural structures, can be easily reflected laterally. Thus, the epidural fat, venous plexus, and medial part of the horizontal portion of V3 are exposed (Fig. 17.6b). The vertebral artery is entirely covered with the periosteal sheath and venous plexus in this area. The muscular branches of the vertebral artery arising from the horizontal segment may be coagulated and sacrificed for a more lateral exposure, thus exposing the transverse process of C1 and the horizontal portion of V3. In some cases, the condylar emissary vein around V3 prevents lateral exposure of the vertebral artery. This vein should be coagulated and sacrificed just behind the condylar fossa to avoid unexpected venous bleeding.
After dealing with the muscular branches and condylar emissary vein, the C1 posterior arch and the horizontal portion of V₃ are widely exposed up to the lateral end of the transverse process of C1 (Fig. 17.7). This is the second key step. In this step, the rectus capitis posterior major and minor, superior oblique, and inferior oblique muscles, which form the suboccipital triangle, have been already reflected in a single layer. The rectus capitis lateralis, a short muscle from the C1 transverse process to the jugular process, is left undissected at the lateral end of the operative field. The muscle is a natural barrier and protects the jugular vein, which is located just anterior to it.

At the interlaminar space between C1 and C2, a flap of ligamentum flavum is reflected laterally, and the atlanto-occipital membrane, epidural fat, and venous plexus are uncovered at the lateral side of the dural tube. The enlarged venous plexus can be easily managed by low-power coagulation using blunt-tip bipolar forceps. In this space, the C2 root running behind the vertical segment of V₃ is identified. Following the C2 root laterally allows safe exposure of the vertical segment of V₃ between C2 and the C1 transverse process. The C2 root can be sacrificed to perform complete transposition of V₃.

The mastoid process, condylar fossa, occipital condyles, foramen magnum, C1 posterior arch, C2 lamina, and V₃, are completely exposed using this technique.

### Transposition of V₃

The V₃ segment may be entirely transposed to expose the regions at the ventral aspect of C1. The C1 transverse foramen is first unroofed with a small rongeur or a diamond bur (Fig. 17.8). The vertebral artery, which is covered by the venous plexus and periosteal sheath, is then carefully dissected from the C1 transverse foramen and arterial sulcus in the C1 lamina and C1 articular process. After this dissection, the vertical portion and the caudal part of the horizontal portion of the vertebral artery is completely freed. At the rostral part, the
periosteal sheath covering the vertebral artery is continuous with the periosteum over the C1 articular process, occipital condyle, and condylar fossa (Fig. 17.9a). The periosteum is incised at the condylar fossa and meticulously reflected downward to the horizontal segment of V3, which frees the rostral part of the vertebral artery (Fig. 17.9b). During the procedure, the vertebral artery can be controlled extraperiosteally as long as possible to avoid bleeding from the venous plexus, which lies inside the periosteal sheath. The enlarged venous plexus in the periosteal sheath can also be easily and safely controlled by low-power coagulation using bipolar forceps, which does not damage the underlying vertebral artery. After completion of this step, the entire course of V3 is liberated from C1 to C2 and can be transposed inferomedially to approach the ventral side of the articular process of C1, the anterior arch, and the C2 odontoid process (Figs. 17.10 and 17.11).

Several regions ventral to the medulla oblongata and the foramen magnum require a more aggressive trans-
position of the vertebral artery. The intra- and extradural vertebral artery can be transposed by cutting the dural fibrous ring in a round shape at the junction of $V_3$ and $V_4$.

### Conclusion

Complete mastery of vertebral artery management enhances surgical safety at the CVJ. In this chapter, we described how to expose the vertebral artery in a medial-to-lateral direction, but after precise understanding of the vertebral artery and the surrounding muscle anatomy, it appears that there is no technical difference between that and lateral-to-medial dissection.

In recently prevalent atlantoaxial fixation techniques, such as transarticular screw fixation and lateral mass screw fixation, $V_3$ exposure is key to increasing surgical safety.
References

Endoscopic sinus surgery was developed by Alfred Hirschmann and Etienne Escat in the early 1900s. However, only after a better understanding of the physiology and pathology of the nose and sinuses provided by Walter Messerklinger and the development of appropriate instruments and the rod-lens systems by Harold Hopkins in the 1960s did endoscopes establish their position in sinonasal surgery. David Kennedy coined the term functional endoscopic sinus surgery (FESS) in 1985 and, along with Heinz Stammberger and Wolfgang Draf, popularized the use of modern endoscopy for the paranasal sinuses in the 1980s. The use of endoscopes in neurosurgery had a different evolution. In an attempt to improve visualization, Gerald Guiot in 1963 was the first neurosurgeon to use the endoscope in transsphenoidal surgery. The endoscopes at this time, however, were still inadequate, and when Jules Hardy finally established the transsphenoidal route in 1967, he incorporated the operating microscope for demonstrating the superiority of accessing the pituitary fossa transsphenoidally as opposed to transcranially.

In the late 1970s, Apuzzo and colleagues, as well as Bushe and Halves, reintroduced the endoscope as an adjunct to the microscope for resection of pituitary lesions with extrasellar extension. Endoscopically “assisted” transsphenoidal microsurgery has since been reported by various authors, stressing the advantages of visualization around corners, particularly for tumors that extend beyond the sella. The endoscope, however, often limits the working space, and there is limitation of maneuverability of the endoscope within the speculum when the microscope is the primary form of visualization. This limitation led to the development of a fully endoscopic, non-speculum-assisted endonasal approach to the ventral skull base resulting from collaboration between neurological and otolaryngology–head and neck surgeons.

The endoscope was first used as the only visualizing tool in skull base surgery for pituitary lesions in the early 1990s. In 1992, Jankowski and colleagues reported their experience with three cases using a pure endoscopic transsphenoidal approach to the sella. Others subsequently confirmed the feasibility of pure endoscopic approaches to the sella. Paolo Cappabianca and Enrico de Divitiis were extremely important in fostering and disseminating the technique.

As a natural progression, various centers around the world began to perform pure endoscopic approaches to the sella. With the panoramic view offered by the endoscope allowing visualization beyond the sella, combined with otolaryngologic and neurological surgeons working together, endoscopic skull base surgery was born.

In a similar evolution to the transsphenoidal approach, the anterior approach to the rostral cervical spine for treatment of the craniovertebral junction was first explored in 1935 by German. Since then, the transoral approach using microscopic visualization has been used extensively for a wide range of pathological entities, including basilar invagination, rheumatoid arthritis with craniovertebral settling and/or pannus, odontoid fractures, tumor (both extradural and intradural), and odontoid hypoplasia. Only recently was the endoscope introduced to transoral surgery as a secondary tool to the microscope, and the feasibility of endoscopic endonasal approaches to the cervicomedullary junction initially demonstrated in cadavers by Alfieri et al.

In late 1998 at the University of Pittsburgh, we began to pursue the expanded fully endoscopic endonasal approach to the ventral skull base in a systematic fashion. Working as a team of otolaryngologic and neurological surgeons over the past 10 years, we have performed over 1000 endoscopic endonasal skull base procedures. As part of this, the first endoscopic endonasal approach for the cervicomedullary junction was reported in 2004.

Indications (Patient Selection)

Clivectomy

A transclival approach is used for endonasal resection of extra- and intradural lesions, such as chordomas, chondrosarcomas, meningiomas, and even some craniofaryngiomas. The approach is defined by partial (upper, middle, or lower third) or complete removal of the clivus (panclivectomy). The upper third consists of the dorsum sellae in the
midline and the posterior clinoïd processes paramedially. The posterior clinoïd processes can be removed either intradurally or extradurally via a combined trans- and subsellar corridor by first performing a pituitary transposition. This consists of elevating the pituitary gland (either intra- or extradurally) superiorly to access the underlying bony structures. Removal of these structures can provide access to the basilar artery and interpeduncular cistern.

The middle clivus can be directly accessed at the posterior aspect of the sphenoid sinus. Its resection is limited laterally by both ascending paraclival internal carotid arteries (ICAs). This lateral access can be extended by exposing and retracting the vertical portion of the ICAs to some extent. If the bone drilling continues inferiorly, the lower third of the clivus is limited laterally by the fossae of Rosenmüller and the torus tubarius. However, by sacrificing the eustachian tubes, the bony resection is limited laterally by the hypoglossal canals. A panclivectomy can extend all the way from the dorsum sellae and posterior clinoïds to the basion at the foramen magnum.

Transodontoid Approach

Lesions involving or adjacent to the clivus may extend caudally to the cervicomedullary junction. The anterior cervicomedullary junction provides a unique surgical challenge. Access is difficult from a posterior or lateral approach due to intervening neural structures and mechanical stability concerns. Anterior, transoral approaches have their own set of difficulties, including poor rostral access, potential for palatal insufficiency, delayed feeding, and oral bacterial contamination. The endoscopic endonasal route avoids these problems.

Extension of the transclival approach can continue caudally to the anterior atlas, dens, and superior body of the second cervical vertebra. Caudal access via the nose is restricted by the angle created with the endoscope and instruments pivoting on the hard palate. To determine caudal access preoperatively, the Kassam line can be drawn on a midsagittal computed tomography (CT) or magnetic resonance imaging (MRI) scan from the tip of the bony nasal bridge to the posterior hard palate and extended to the cervicomedullary junction (Fig. 18.1).

This approach can be used for resection of the odontoid process in degenerative/inflammatory diseases, pannus formation, and basilar invagination or to allow for exposure of the ventral medulla and upper cervical spinal cord for tumor resection. Foramen magnum meningiomas and rheumatoid pannus are examples of lesions that can be treated using this approach.

The approach is defined by the removal of the odontoid process of the axis (second vertebra). This approach is an extension of the transclival approach and may require partial removal of the clivus, especially in the setting of basilar invagination. The lower third of the clivus is exposed, as well as the anterior arch of C1, after dissection of the nasopharyngeal mucosa and the longus capitis muscles. The anterior arch of C1 is removed with a high-speed drill and rongeurs to expose the articulation with the odontoid process. The odontoid process can then be resected in a fashion similar to that previously used with transoral approaches. Lateral access is limited by the eustachian tubes, although these can be retracted or even resected, at which point lateral access is limited by the hypoglossal canals in the condyle and the parapharyngeal ICAs.

![Fig. 18.1: Midsagittal computed tomography (CT) scan showing the Kassam line drawn from the tip of the bony nasal bridge to the posterior hard palate and extending to the cervicomedullary junction.](image-url)
Description of the Technique

Planning

Frameless stereotactic image guidance is used in all expanded (endoscopic) endonasal approaches to the skull base. Image guidance is of value in corroborating the visual impression of the surgical anatomy, especially critical neurovascular structures, and helps to define a targeted resection and ensure adequate exposure. A high-resolution CT angiogram is used for most skull base surgeries, as it allows for the simultaneous visualization of osseous, vascular, and soft tissue anatomy. Increasingly, we use image fusion of CT and MRI scans to take advantage of the best features of each: CT for the bony anatomy of the cranial base and MRI for intracranial tumor visualization.

Operating Room Setup

The surgeons are positioned on the right side of the patient opposite the anesthesia team. The surgical technician or nurse is positioned toward the foot of the bed. This arrangement gives the surgeons unrestricted access to the nasal region. Electrical cords and suction tubing are directed away from the surgical field toward the head and foot of the bed to minimize interference with surgical instruments.

Head pin fixation is used to ensure the lack of intraoperative movement of the head, especially during drilling and neurovascular dissection. The head is fixed following endotracheal intubation, with the neck in slight and neurovascular dissection. The head is minimally tilted to the left, and the face turned to the right side of the surgical bed as possible to limit the surgeon’s reach. The bed can be angled in the room so that the foot of the bed goes away from the surgeons, allowing for even more space.

Neurophysiological monitoring of cortical function (somatosensory evoked potentials) with or without brainstem function (brainstem evoked responses) is routinely performed in all cases where dura is exposed or dissection near the carotid or vertebral arteries is performed. In addition, neurophysiological monitoring can identify changes in cerebral blood flow that may occur with blood loss or changes in blood pressure and alert the anesthesiologist to make adjustments. Cranial nerve electromyography is often performed, particularly to monitor cranial nerve (CN) VI during upper third clival resections and CN XII during condyle dissection. If a lateral extension of the approach is planned in the infrapatrous area, CN VII and VIII are monitored, as well as CN IX, X, and XI, when dissection is planned near the jugular foramen.

A nasal decongestant, such as oxymetazoline 0.5%, is applied topically to the nasal mucosa using cottonoids. The skin of the external nose and nasal vestibule, as well as the abdomen (fat graft donor site), is preppe with a povidone antiseptic solution. The patient is given a third- or fourth-generation cephalosporin for peroperative antibiotic prophylaxis.

Initial Approach

The endoscope is introduced at the “12 o’clock” position of the nostril (usually the right) and is used to retract the nasal vestibule superiorly. This elongates the nostril and increases the available space for other instruments. A suction tip is generally introduced at the “6 o’clock” position on the same side. Dissecting instruments are introduced through the left nasal cavity. A suction irrigation sheath or irrigation applied by an assistant or cosurgeon cleans the lens of the scope and preserves visualization without removing the scope for frequent cleaning. If for any reason a bimanual (preferably binarial) approach cannot be performed, then the surgery should be modified to allow bimanual dissection or aborted. Furthermore, we discourage the use of an endoscopic holder for all expanded endonasal approaches.

Widening of the nasal corridor is achieved initially by out-fracturing of the inferior and middle turbinates, followed by removal of the right middle turbinate to provide room for the endoscope. Injection of vasoconstrictors is optional and is performed according to the surgeon’s preference.

For the past 2 years we have been using a nasoseptal vascularized mucosal flap for reconstruction of the skull base defect. Because it has to be harvested during the exposure, surgical planning is crucial. For cases in which dura mater will be opened for tumor exposure, the decision is simple, and the flap is elevated immediately without hesitation. Recently, we have experienced less nasal morbidity during the postoperative period for patients who received a vascularized mucosa covering the exposed sphenoid sinus bone. Posterior crusting is converted to anterior crusting, which can be easier to access and treat.

We use unipolar electrocautery with an insulated needle tip to incise the septal soft tissues. Two parallel incisions are performed following the sagittal plane of the septum. One follows the maxillary crest, and a parallel incision follows a line 1 cm below the most superior aspect of the septum to preserve the olfactory epithelium and function. These parallel incisions are joined anteriorly by a vertical incision usually placed just anterior to the anterior head of the inferior turbinate. Posteriorly, the superior incision is extended laterally inferior to the natural sphenoid ostium. The inferior incision extends laterally on the superior margin of the choana. These cuts are critical and must be done properly to preserve the vascular supply via the posterior nasal artery(ies). Elevation of the mucoperichondrium using a Cottle dissector proceeds from anterior to posterior after ascertaining that all incisions have been carried through the peristeum and periosteum. Elevation of the flap from the anterior face of the sphenoid sinus is completed preserving the vascular pedicle between the sphenoid ostium sinus and choana.
The flap is usually stored in the nasopharynx; however, for exposure of the cervicomедullary junction, we prefer to tuck it in the maxillary sinus after an antrostomy is performed, which facilitates dissection by moving the flap out of the way.

The nasoseptal flap is pedicled on the posteronasal segment of the artery. The mucosal pedicle goes from the roof of the choana to the sphenoid ostium. Inferiorly, it can be extended laterally in the floor of the nasal cavity if needed; or more often, it is harvested up to the transition between the septum and the floor.

### Rostral Clivus

Clival approaches can be broken down based on division of the clivus into thirds. The rostral third includes the dorsum sellae and posterior clinoids. The middle third consists primarily of the clival recess of the sphenoid sinus and Dorello’s canal. The caudal third is the remainder located inferior to the level of Dorello’s canal at the petroclival junction down to the foramen magnum.

Access to the rostral clivus begins with access to the sella. The dorsum sellae and posterior clinoids help create the boundaries of the rostral clivus. These structures can be accessed via either an intra- or extradural approach.

The intradural approach essentially involves the en bloc elevation of the contents of the sella, providing access to the interpeduncular cisterns and basilar artery through a pituitary transposition. This approach is useful for lesions with retrosellar or retrolabyrinthine extension.

This approach begins with the complete exposure of the sella, planum, and tuberculum sellae and the junction of the sella with the clivus, followed by complete removal of the bone overlying the pituitary gland and inferior and superior intercavernous sinuses. These and the following steps are done essentially to allow safe mobilization of the pituitary gland and prevent its compression against a bony margin. The superior intercavernous sinus (SIS) is cauterized using bipolar cautery and transected. This allows opening of the diaphragma and subsequent stalk mobilization. The entire pituitary gland is exposed and the diaphragma is cut along the midline at its point of attachment to the sella to expose the stalk. The diaphragma is then cut in a paramedian direction to release the stalk circumferentially. Care must be taken not to damage the superior hypophyseal arteries. Next, the lateral connective tissue between the gland and the cavernous sinus is carefully dissected, freeing the gland while carefully preserving vital blood supply via superior hypophyseal branches. The gland can now be safely elevated, exposing the dura overlying the dorsum sellae and posterior clinoid. This dura harbors the posterior intercavernous sinus and is coagulated (bipolar cautery) and dissected, with further hemostasis achieved using fibrillar collagen/cottonoid or morcellized Gelfoam. The posterior clinoid is then drilled using a 1- or 2-mm diamond bit until eggshell thin, dissected free from its dural attachments, and then removed carefully to avoid injury to the carotid and CN III located laterally.

For petroclival lesions without retrosellar or more rostral extension, the dorsum sellae can be removed via a completely extradural approach. This is obviously preferred for extradural lesions and petroclival intradural lesions that do not reach the interpeduncular cistern superiorly. In this approach, the planum is left intact, but the sella and SIS are exposed. The clival recess and upper clivus are drilled with a 3- or 4-mm coarse hybrid diamond bit between the paraclival carotid protuberances. Next, the gland and its overlying dura are elevated together (it is at this point that the removal of bone overlying the SIS is critical, as it allows the free elevation of the gland). Constant awareness of the carotid is critical throughout any and all expanded endonasal approaches. The dorsum sellae and occasionally the posterior clinoids can be reached and dissected, exposing the posterior clival dura that harbors the basilar plexus. Once more, careful hemostasis is necessarily observed using morcellized Gelfoam, fibrillar collagen, and bone wax. Overpacking should be avoided, given the proximity of critical neurovascular structures.

### Caudal Clivus/Panclival Approach

Access to the most caudal portion of the clivus is often necessary, as part of a complete clival resection. Exposure therefore begins as usual with wide bilateral sphenoidotomies. However, it is important to mention that there are situations in which we perform a direct inferior approach to the cervicomедullary junction without opening the sphenoid sinus.

It is important to have awareness and even exposure of anatomical landmarks (e.g., ICA) and create access for identification of the pterygoid plates and vidian canal. Modifications specific to caudal clival access begin with removal of the most inferior attachment of the nasal septum to the sphenoid rostrum. In addition, the removal of the posterior 1 to 2 cm of the septum should be continued caudally, allowing binarial work in this lower field. The total field is now defined by the sphenoid sinus rostrally, the hard and soft palate caudally, and the pterygoid plates and eustachian tubes laterally.

The depth of the field is composed of nasopharyngeal mucosa overlying basopharyngeal fascia. The basopharyngeal fascia is stripped from the underlying clivus. This is typically a somewhat difficult and tedious process. It can be facilitated by monopolar electrocautery during the initial, central dissection, but caution should be used laterally, as one approaches the pharyngeal segment of the carotid artery, posterolateral to the eustachian tubes. A Cottle elevator, “true cut” rongeurs, and specially designed scissors and bipolar cautery may aid this portion.

The clivus is then drilled using a 3- or 4-mm coarse bit. Bleeding from the cancellous bone can be controlled with bone wax applied on a cottonoid once the inner cortex is reached. The inner cortex is removed with a combination...
of drilling and Kerrison rongeurs. The lateral extent of bone and/or tumor removal can be achieved through a combination of the aforementioned approaches. When removing the midline clival bone rostral to the level of the vidian nerve, it is imperative to drill only in the midline, between the carotid canals, until these are well defined.

Once the overlying clivus is removed in the midline, the underlying dura and its basilar venous plexus are exposed. The basilar plexus can bleed profusely when the dura is opened, particularly if it has not been thrombosed by the tumor. Given this potential, the face of the dura should be thoroughly coagulated and segmentally opened in the midline, using microfibrillar collagen (Avitene) or morcellized Gelfoam to control bleeding. The lateral dural opening, under the horizontal segment of the petrous carotid, is extended using specially designed endoscopic scissors or Kurze scissors until the eustachian tubes are encountered just as they disappear obliquely into the skull base. Lateral opening of the dura superior to this segment (at the level of the carotid genu between the petrous and vertical/paracaval ICAs) should be done, as always, under direct visualization, bearing in mind the position of the abducens nerve as it travels in this space to enter Dorello’s canal just medial, superior, and dorsal to the anterior carotid genu and paracaval ICA.

The sequence for intradural dissection in this region begins with identification of the vertebral artery, which is then followed to the vertebralbasilar junction (VBJ). The abducens nerve can be identified, as expected, at the VBJ bilaterally. The basilar artery can then be followed in a rostral direction to provide exposure to the remainder of the posterior circulation, pons, and CN V, VI, VII, VIII, IX, and X. If exposure of the oculomotor nerve is needed, a posterior clinoidectomy, as described above, will need to be completed.

This approach is effective for clival tumors, especially chordomas, chondrosarcomas, sinonasal tumors, plasmacytomas, and osseous and intradural meningiomas. Once again, tumor characteristics, such as consistency and vascularity, do not contradict endoscopic resection. These techniques have been used for all of the above lesions, as well as hemangioblastomas, angiofibromas, and arteriovenous malformations.

### Cervicomedullary Junction

The exposure is the same as the inferior clival exposure described above, with fascial stripping extending to the ring of C1 and the rostral body of C2 if accessible. Care should be taken to avoid violation of the ligamentous bands between the caudal clivus/foramen magnum and the ring of C1. This will prevent inadvertent dural or neurovascular violation, as there is often an interdigitating dural fold in this area. The lateral exposure is limited by the hypoglossal canals. Exposure of the medial portion of the occipital condyle provides a boundary. Once again, care should be taken with the lateral dissection, bearing in mind the course of the vertebral artery. Bleeding from muscle or venous plexus can be controlled with bipolar cautery or Avitene “sandwiches.”

The ring of C1 can be drilled with a 3- or 4-mm coarse diamond bit until eggshell thin. This last portion is removed safely with a 1- or 2-mm Kerrison rongeur. Similarly, the dens can be resected, if needed, by drilling a central trough, followed by in-fracturing of the thinned cortex. The base of the dens should not be transected first, as this will leave a free-floating tip, increasing the difficulty and risk of removal. After careful sectioning of the tectorial membrane and overlying ligaments, these can be laterally retracted and resected or retracted with bipolar cautery to expose the dura. Dural opening can be extended with endoscopic scissors.

The addition of this exposure provides the caudal access needed for foramen magnum tumors, such as meningiomas, and is also ideal for the removal of arthritic pannus.

### Case Illustration

**Craniovertebral Junction: Extradural**

A 75-year-old man presented with 8 months of multiple falls and gait instability. An MRI and CT of the cervical spine revealed a pannus at the level of the odontoid causing displacement and impingement of the cervicomедul- lary junction (Fig. 18.2). After medical evaluation, the patient was taken to the operating room for a combined anterior endoscopic endonasal approach for odontoidectomy and pannus resection, followed by a posterior occipital cervical fusion. The endoscopic endonasal procedure consisted of removal of the anterior arch of C1, the dens, and the extradural pannus. At this point, it was apparent that the abundant pannus had penetrated the dura. This was carefully dissected, the dural opening was enlarged slightly, and the intradural portion was examined and resected. Reconstruction was performed using a nasoseptal flap augmented by a fat graft. The patient underwent posterior fusion the same day.

The postoperative period was uneventful. Postoperative MRI and CT were obtained during the initial 24 hours after surgery (Fig. 18.3). Lumbar drainage was employed and was removed on the fifth day after surgery without any sign of cerebrospinal fluid (CSF) leakage. The patient was discharged neurologically unchanged on the seventh postoperative day.
A 69-year-old woman presented with nonspecific symptoms of loss of taste and smell. General physical examination was normal. Neurological examination was unremarkable with the exception of hyperreflexia. MRI findings revealed a tumor compatible with a foramen magnum meningioma (Fig. 18.4). Based on the fact that she had early signs of brainstem compression on imaging and clinical examination, resection was recommended. Given the completely ventral location of the lesion, located between both vertebral arteries, as confirmed by CT angiogram (Fig. 18.5), an endoscopic endonasal transclival/transodontoid approach was offered and performed. Surgery was executed without complications. The clivus was drilled from the level of the rostrum to expose the superior portion of the tumor down to the foramen magnum and tip of the dens with the help of image guidance (Fig. 18.6). A middle condylectomy was performed on the patient’s left side (Fig. 18.7), where the tumor was more eccentric, to allow for better exposure and control of the left vertebral artery. Once the bony resection was satisfactory, the dura mater was opened in the midline and then expanded laterally (Fig. 18.8). The tumor was immediately under the dura, and the initial step was direct identification of the VBj (Fig. 18.9), followed by the anterior spinal arteries...
Fig. 18.8 Intraoperative view with a 0° endoscope during a transclival approach for resection of a meningioma. At this point, the dura (D) was opened in the midline, and the tumor (T) is exposed. The arachnoid (A) can be visualized superiorly in the field.

Fig. 18.9 Intraoperative view with a 0° endoscope during a transclival approach. Once the tumor (T) was partially debulked and the superior portion was resected, then the vertebrobasilar junction was exposed. Note the close relationship of both vertebral arteries (V) forming the basilar artery (B).

Fig. 18.5a, b Preoperative CT angiogram showing a contrast-enhancing tumor located ventral to the cervicomedullary junction compatible with a foramen magnum meningioma. Note the location of the tumor between both vertebral arteries. (a) Sagittal and (b) coronal reconstructions.

Fig. 18.4a–c Preoperative MRI with contrast showing a contrast-enhancing tumor located ventral to the cervicomedullary junction compatible with a foramen magnum meningioma. (a) Coronal and (b, c) axial T1 images at two different levels.

Fig. 18.6 Intraoperative view with a 0° endoscope during a transclival approach. Note the dura (D) already exposed dorsally and the foramen magnum (FM) inferiorly.

Fig. 18.7 Intraoperative view with a 0° endoscope during a transclival approach. At this point, the anterior arch of C1, along with the medial aspect of the occipital condyle, is being removed with a drill.
(Fig. 18.10). Once both vertebral arteries were identified, the dissection proceeded in either direction (rostrally and caudally) following the vertebral arteries (Fig. 18.11) with exposure of the brainstem and spinal cord (Fig. 18.12). The resection was subtotal, with residual inferiorly located behind the odontoid process (Fig. 18.13). The decision was made not to resect C2 to avoid destabilization and the need for craniocervical fusion.

The postoperative period was complicated by a pulmonary embolism, which was successfully managed clinically with anticoagulation. The patient was seen in follow-up at 3 months without any complaints and was neurologically intact.
Craniovertebral Junction: Panclival and Corridor Surgery

A 3-year-old boy presented to a local hospital after being struck in the head with a baseball bat. CT scan revealed a large posterior fossa mass. Upon referral to the University of Pittsburgh Medical Center, further questioning disclosed a long history of ataxia and clumsiness, especially when compared with the coordination of his triplet brothers. His parents also reported 2 months of progressive dysphagia with profound drooling causing spoilage of his clothing.

On neurological examination, the child was awake and alert. Pertinent positive findings on examination included decreased gag reflex bilaterally, decreased phonation, mildly brisk reflexes, and ataxia with the inability to perform tandem gait. The remainder of his examination was within normal limits.

A brain MRI revealed a large mass, likely originating from the skull base with submucosal extension into the nasopharynx and oropharynx and exhibiting significant brainstem compression and mass effect on the fourth ventricle with resultant mild supratentorial ventriculomegaly (Fig. 18.14). The basilar artery was displaced posteriorly and encased by the tumor. A CT angiogram of the head and neck was obtained to better define the vascular and bony anatomy surrounding the tumor, and these images were used for frameless stereotaxy. Because the etiology of this lesion was uncertain and the mass was visibly accessible within the posterior pharynx, a minimally invasive approach was pursued to obtain an initial tissue diagnosis. All stages of tumor resection were performed with frameless stereotactic guidance using both CT and MRI technology (Stryker Navigation, Kalamazoo, Michigan). The surgical approach to this tumor included a sublabial incision for access to the nasal cavity due to the small aperture of the anterior nares (Fig. 18.15). Separate endoscopic stages were performed for tumor debulking over several months. Superolateral exposure included the petroclival synchondroses and pterygoid canals. The atlanto-occipital ligaments were resected and the anterior foramen magnum was completely removed to expose the occipital condyles. Intraoperative image guidance (Stryker Navigation) displayed the route, as our pointer is seen resting on the posterior right edge of the foramen magnum (Fig. 18.16). The remainder of the retropharyngeal tumor was removed laterally until the contents of the parapharyngeal space and the jugular veins were exposed bilaterally. The tumor was noted to have multiple septations. The dura was opened, and the tumor resection for stage 2 was more extensive, achieving decompression of the brainstem. Pathology revealed a chordoma.

The patient did suffer a brief, transient hemiparesis that resolved spontaneously. The residual tumor was intimately associated with the vertebrobasilar vasculature. The patient had a mild transient right hemiparesis, which completely resolved within 1 week. His dysphagia and decrease in phonation improved with resolution of drooling and marked improvement in imbalance/ataxia.

Due to the proximity to the cervicomedullary junction, the residual volume of tumor was determined to be too large to safely irradiate. Given the findings noted above and the position of the vertebral arteries, this portion of the tumor was felt to be best accessed through bilateral posterolateral, transcervical, and transcondylar approaches.

The patient underwent bilateral transcervical, transcondylar approaches through the carotid–vertebral window for further tumor resection. Using a retroauricular curvilinear partial Fisch incision, a neck dissection and mobilization of the digastric and sternocleidomastoid muscles along with the carotid sheath anteriorly provided access into the posterior triangle of the neck. After the dissection of the paraspinal musculature, the transverse processes of C1, C2, and C3 were identified and removed. The vertebral artery was transposed...
into a posteromedial position, which allowed the lateral mass of C1 to be removed. Tumor resection followed superiorly to the dural space, and the occipital condyle was drilled to provide further exposure and access to the hypoglossal foramen. Figure 18.17 demonstrates the essence of usage of corridors, as the midline tumor is shown schematically being approached from the endoscopic endonasal route and the posterior/lateral components being accessed via the carotid–vertebral window. After the destabilizing approaches, the patient was turned prone under neurophysiological monitoring, and an instrumented occiput-C2 fusion was performed.

The multiplicity of resections allowed for tumor settling into accessible locations so that it could be resected using the appropriate corridor in an easier and safer fashion. It is critical that the surgical team approaching such tumors be well versed in open as well as endonasal approaches so that they are not limited merely by their anatomical knowledge and comfort. Adjunctive therapy with proton beam radiotherapy is planned for the treatment of the small amount of residual disease (Fig. 18.18).
Fig. 18.17 This drawing depicts the essence of corridor surgery for the skull base surgeon. Note the retroclival tumor (brown) that is accessible through endonasal endoscopy, along with the tumor extending inferoposteriorly (green) into the upper cervical spine that is accessible through the carotid–vertebral window (arrow). (Reprinted with permission from Pirris SM, Pollack IF, Snyderman CH, et al. Corridor surgery: the current paradigm for skull base surgery. Childs Nerv Syst 2007;23:377–384.)

Fig. 18.18a, b A 3-year-old patient’s postoperative (a) sagittal T1-weighted with contrast image and (b) axial T2-weighted image revealing ~90% removal of the tumor, which is stable and, most importantly, with persistent adequate decompression of the brainstem and fourth ventricle. The residual tumor (arrow) was originally adherent to the basilar artery and its perforators.

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**Conclusion**

The clivus and paracloval areas can be accessed via an endoscopic, endonasal route, as an expansion of techniques for accessing the sella. Complete knowledge of the associated sinonasal, bony, vascular, and neural anatomy is essential for the safe performance of these procedures. In addition, binarial access with bimanual instrument manipulation is key for many portions of these approaches. For these reasons, as well as perioperative management, a team approach, including an otolaryngologist and a neurosurgeon, is critical to the development and performance of these procedures.

The techniques described here allow access to the clivus, petroclival junction, and cervicomedullary junction with minimal or no neural manipulation. A wide range of skull base lesions involving these areas can be accessed for biopsy, debulking, or complete resection as warranted. These techniques have an inherent learning curve that must be acknowledged. The team performing expanded approaches should first achieve comfort and skill with standard sellar lesions prior to attempting more complex work. This is necessary for acquiring skill as well as the teamwork required when working with angled endoscopes in tight areas.

Finally, these approaches merely offer another corridor for resection of skull base, clival, and cervicomedullary junction lesions. They are another weapon in the arsenal of the surgical team and should be applied carefully and not broadly to all skull base tumors. Cases should be chosen carefully, as these techniques are naturally well suited, though not restricted, to tumors that displace neurovascular structures laterally, which is the majority of clival lesions. If applied with care, these approaches hold great value in the management of clival and cervicocranial junction lesions, with the potential for decreased morbidity.

**References**


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19 Image-guided Surgery in the Craniovertebral Junction and Upper Cervical Spine

Petr Suchomel and Ondrej Choutka

Scott D. Boden pointed out: "The challenge and obligation of any new technology must be to solve an unsolved problem, enable the physician to perform an otherwise undoable task, or significantly facilitate the performance of a common task. We must all ensure that we do not encourage the triumph of technology over reason." This is particularly true in the field of spinal surgery with its rapid development of navigational techniques over the past decade. This technological evolution not only brought about improved safety and accuracy of various techniques but also allowed less experienced surgeons to perform technically more demanding procedures. Despite this, the majority of experts correctly emphasize that image-guided spinal surgery is a supplement to, not a replacement for, the individual surgeon's experience and judgment.

All procedures involving the upper cervical spine (UCS) and craniovertebral junction (CVJ) are technically demanding. The presence of vitally important structures and the unique anatomy of this region call for experience, excellence, and the most sophisticated technical solutions available. Numerous surgical techniques for treating pathologies of the CVJ and UCS have been described in the literature. Therefore, the introduction of new types of imaging and guidance techniques does not substantially contribute to further development of such approaches. However, there are certain types of operations where the extent of pathology is not directly visible, the resection volume of pathological tissue cannot be predicted, or the implant trajectory is out of sight of direct vision. In such situations, simple fluoroscopy is very helpful; however, its mono- or biplanar nature cannot provide the same three-dimensional (3D) control. Hence, new techniques that use image and real-time computer guidance have the potential to significantly improve the safety and accuracy of such procedures.

When it comes to spatial orientation, it is important to have a comprehensive knowledge of not only the target structures, but also all the potential surrounding dangers. Historically, neurosurgeons have always tried to reach deeply located lesions via the safest and least destructive route. Frame stereotaxy and frameless navigational systems proved to be indispensable in this task. Navigation in cranial surgery is simplified by the initially stable relationship of the brain to the skull. It allows for accurate surface registration prior to the intervention. Numerous attempts to use similar techniques in the spine were relatively successful in the past; however, the mobile nature of spinal elements prevented the use of stereotaxy in this region. The development of modern imaging techniques (computed tomography [CT], magnetic resonance imaging [MRI], and isocentric fluoroscopy), together with improved computer software solutions, opened the door to a new way of thinking about spinal surgery in three dimensions.

The most suitable type of computer guidance system used in image-guided surgery is dependent on the task at hand. The requirements are different in, for example, oncologic procedures, where localization, tumor extent, and demarcation of the tumor's borders have to be outlined, and the extent of resection may need to be continuously monitored, than for trauma cases, where mobile fragments are registered based on preoperative scans, but any deformity correction cannot be followed in real time without data re-registration.

It is important to realize that inaccuracies in instrumentation of the UCS or CVJ may result in three types of morbidity—neurological, vascular, and mechanical.

Relevant Anatomy

A detailed knowledge of the anatomy prior to surgery is necessary to avoid unacceptable surgical aggression or implant misplacement. All imaging modalities of the UCS are used to assess the individual anatomy and its variations prior to surgery. Plain radiographs (including lateral and transoral views) can demonstrate bony anomalies and/or deformities. CT, especially thin-cut studies with 2D or 3D reconstructions, are essential for bony anatomy analysis. Furthermore, the use of specific CT guidance protocols allows for virtual preoperative planning that is capable of assessing the feasibility and safety of screw implantation (e.g., implant length and width relative to the accepting bone). MRI is more frequently used to depict soft tissue anatomy and pathology (e.g., disks, ligaments, and neural structures). Dynamic or cinegraphic MRI can demonstrate the relationship of the spinal envelope to its contents in flexion, extension, or other movements. For example, a damaged transverse ligament can be directly visualized.
Osseous anatomy of the occipital bone is important whenever occipitocervical fusion is being considered. Most of the currently available constructs use occipital plates with either lateral (squamous part of bone) or medial (occipital ridge) screw holes. It is important to pay attention to the thickness of the bone, as well as the position of the confluence of the sinuses. Medially placed occipital screws provide stronger purchase due to thicker bone. Screws can be optimally placed in the skull with the aid of image guidance (CT-based cranial navigation, intraoperative CT, or isofluoroscopy); however, its routine use is probably not necessary for this task.
Clivus

Virtual navigation of the clival region is frequently used for skull base tumor localization and for intraoperative monitoring of the extent of tumor resection. Whenever clival screws are considered, the position, angle, and thickness of the clivus needs to be assessed prior to surgery. This structure can be virtually navigated from cranial registration. Navigation can also help to localize the clival edge as an anatomical landmark in complex transoral surgeries.

Atlas

The first cervical vertebra has no body, and the only strong structures for potential implant anchorage are the lateral masses. They can be included in anterior\(^2\)\(^,\)\(^23\) or posterior atlantoaxial transarticular fixation,\(^4\) or they can act as isolated anchors in different constructs.\(^2\)\(^,\)\(^24\)\(^,\)\(^25\) Although the vertebral artery can potentially be violated during anterior exposures, implant-related injury is usually limited to posterior techniques.\(^26\)\(^–\)\(^28\) This risk may be increased due to vertebral artery course anomalies.\(^29\)\(^–\)\(^32\) Venous bleeding from vessels surrounding both C2 nerve roots can be avoided by subperiosteal dissection\(^2\) and better positioning of the patient on the surgical table.\(^25\) Recent publications warn of other “hidden” risks during lateral mass screw placement,\(^26\)\(^,\)\(^33\) especially if bicortical screw purchase is necessary to strengthen the construct.\(^26\) The carotid artery and hypoglossal nerve are also at risk. The hypoglossal nerve lies 2 to 3 mm lateral to the middle of the anterior aspect of the C1 lateral mass.\(^23\) The internal carotid artery varies in location but can lie within 1 mm of the potential screw exit point.\(^26\) Therefore, it is recommended to place such screws in the posteromedial sublaminar part of the lateral mass; also, the trajectory should be slightly medial. In general, the so-called screw entry safe zones (both anterior and posterior) have been established;\(^2\)\(^,\)\(^23\)\(^–\)\(^25\)\(^,\)\(^34\)\(^–\)\(^37\) and preoperative thin-cut CT and/or CT/MRI angiography may be helpful in selected cases with suspected vascular anomaly.

Axis

The unique anatomy of the second cervical vertebra represents the most challenging structure in the UCS. In particular, the variability of the vertebral artery course and its modification under pathological conditions are the subject of many articles.\(^29\)\(^,\)\(^38\)\(^–\)\(^40\) A high-riding vertebral artery groove is described in up to 23% of patients.\(^29\)\(^,\)\(^31\)\(^,\)\(^37\)\(^–\)\(^41\) The axis, in comparison to the subaxial vertebrae, has the largest pedicles but the least developed lateral masses; hence, the pedicle appears more suitable for screw placement.\(^42\)\(^–\)\(^44\) This assumption is not supported by the work of Resnick et al.\(^36\) and Yoshida et al.\(^45\) Measuring the distances of the vertebral artery groove from the possible screw trajectory, they found that the C2-pedicle screw placement has nearly the same potential risk of vertebral artery injury as transarticular screw placement, and they recommended a preoperative 3D computerized evaluation to choose the best surgical technique. Numerous anatomical exclusion criteria for safe transarticular screw placement have been described. Abou Madawi et al.\(^31\)\(^,\)\(^39\) stated that a 3.5-mm screw cannot be introduced if the internal height of the C2 lateral mass, measured on CT reconstructed images, between the highest point of the vertebral artery groove and the superior facet surface is < 2 mm (Fig. 19.3). This statement is questionable, as the screw trajectory does not necessarily intersect the measured area, and it can be adjusted for adequate placement.\(^46\)

Measuring the dimensions of the isthmus of the pars interarticularis in the real screw trajectory modeled on 3D software is more reliable.\(^40\)\(^,\)\(^47\)\(^,\)\(^48\) Mandel et al.\(^40\) analyzed C2 vertebrae of 205 human cadavers and found that 10% of specimens had an isthmus < 5 mm wide and 5 mm high. This is less than was expected from related clinical articles. Bloch et al.\(^47\) studied 17 cadaveric spines and determined that the risk of injury to the vertebral artery can be decreased to 5.9% with the use of a navigational system if the entry point and screw trajectory are modified individually based on preoperative 3D planning. Under these circumstances, the isthmus height can be 4.0 mm for safe placement of a 3.5-mm screw. We also have to consider that size differences depend on the individual’s gender, race, and even the side on which the procedure is being done.\(^29\)\(^,\)\(^37\)\(^,\)\(^38\)\(^,\)\(^46\)\(^,\)\(^47\)\(^,\)\(^49\)\(^,\)\(^50\)

Fig. 19.3  Internal height of a C2 lateral mass on coronal plane reconstruction. Arrows indicate distance between the highest point of the vertebral artery groove and the surface of the superior facet.
In conclusion, the height of the internal isthmus should be $>5$ mm on CT sagittal reconstructions and 5 mm lateral from the internal spinal canal border (Fig. 19.4). This indirectly implies that the isthmus width is $>5$ mm. In challenging cases, the possible trajectory should be tested in a 3D spatial program changing the entry point and screw path.\textsuperscript{48} If a C2 pedicle screw is considered, the size and angle of the individual pedicle should be measured on a CT reconstruction directly and the trajectory planned accordingly.\textsuperscript{36,49} In questionable or borderline situations, CT angiography should be done to localize the course of the vertebral artery exactly. The space occupancy ratio of the vertebral artery/bony groove was measured by Cacciola et al.\textsuperscript{29} as 79\% (34–100\%).\textsuperscript{30} Anterior safe zones of the axis were defined by Kandziora et al.\textsuperscript{35} and Koller et al.\textsuperscript{23}

![Fig. 19.4 Internal height of the C2 pars interarticularis isthmus (arrows) measured on sagittal reconstruction 5 mm laterally from the internal border of the spinal canal. (a) Insufficient and (b) sufficient space for the introduction of a 3.5-mm screw.](image)

**Technique Description**

**Virtual Image-guided Surgery**

After a positive experience with cranial image guidance, Foley and Smith introduced virtual image-guided surgery to the spine in 1994.\textsuperscript{13,51} The main problem was the lack of correlation between reliable skin surface registration markers (fiducials) and bony structures of the spine because of skin and spine movement. This was solved with the registration of bony anatomical landmarks of the dorsal spine in association with dynamic reference array (DRA) directly attached to the target vertebra.\textsuperscript{13,14,52,53} Most surgeons at the time were using virtual image-guided surgery for lumbar pedicle screw placement.\textsuperscript{13,14,54,55} Later, it became evident that in the lumbar or thoracic spine, implant navigation could be more accurate than traditional methods,\textsuperscript{30,55,57} and the technique also began to be used in more delicate structures of the UCS.\textsuperscript{15,16,58–60}

In principle, according to the specific protocol, the anatomical data have to be obtained by thin-sliced (1.0–1.5 mm) CT or MRI and transmitted to the computer workstation. DRA must be firmly connected to the target area so as not to hinder the operation. The electro-optical camera connected to the computer workstation then registers the position of the navigated vertebra and the instruments being tracked. The space orientation of tracked entities is marked by either passive arrays (reflective spheres) or active arrays (light-emitting diodes). The virtual picture is shown on the computer output display. The surgery itself follows the virtual preoperative plan and can be controlled in 3D on the screen.

All available systems work with images obtained either before the surgery or during the operation after exposure of the target structures. This means that the anatomical data set is obtained prior to intervention or implant introduction. Intersegmental movement during positioning of the patient or the surgery itself can change the virtual data set. Therefore, only one vertebra can be virtually guided, and any change in the DRA connection has to be updated with the help of new manual or automated registration.

**Planning in Cervical Surgery**

The acquired set of anatomical data is so comprehensive that the workstation software allows the surgeon to make a plan for any intervention. In the UCS and CVJ, 3D analysis usually focuses on the feasibility of C2 screw acceptance, the course of the vertebral artery, the localization of the spinal cord and brainstem, the position and thickness of
the clivus, and the planning for tumor approach and resection. Such planning allows the surgeon to determine the feasibility of complex procedures. Virtual planning is often recommended even when further navigation is not possible or required.\cite{38,39,48}

**Preoperative CT-based Virtual Image-guided Surgery**

This is the most accurate method of virtual spinal bone navigation, especially effective in anatomically difficult regions. If the registration process can be done precisely enough (1.5-mm accuracy), image guidance can be used for all known procedures of screw introduction in the UCS and CVJ. It is most frequently used in transarticular C1–C2 fusions\cite{15,16,58,61,62} and the published results describing precision of screw placement are better with navigation than using conventional techniques.\cite{30,63–65}

**Major Drawbacks**

A specific protocol calling for preoperative CT is necessary, which increases the cost if CT adequate for diagnosis already exists but is not compatible with the image-guided system. The time for registration is long, there is a learning curve, and only one vertebra can be navigated; therefore, a separate registration process for each vertebral level is unavoidable.

**Intraoperative CT-based Virtual Image-guided Surgery**

The CT scanner is located in the operating room, and the registration process is automated. The scans are obtained in the final surgical position. Inaccuracy caused by intersegmental movement can be minimized.\cite{51} Postinstrumentation images allow for an intraoperative check of the position.

**Major Drawbacks**

Drawbacks include the cost of the mobile scanner, ergonomic problems (draping, patient manipulation, etc.), the necessity of a special transparent table, and the fact that movable parts of the spine (fracture fragments) cannot be visualized in changed positions without new registration.

**Fluoroscopy-based (Two-dimensional) Virtual Image-guided Surgery**

This is a method combining fluoroscopy, familiar to all spine surgeons, with image guidance techniques.\cite{14,51,66} The accuracy of instrumentation and virtual guidance is increased in only one plane at a time. Its usefulness was not described in UCS and CVJ surgery. Nevertheless, the radiation exposure is much less than in classical fluoroscopy.

**Major Drawbacks**

The guidance depends on the quality of images obtained by fluoroscope. This can be a problem in shadow areas of the upper thoracic spine or in obese patients. Precise images of the complex UCS anatomy cannot be obtained regularly. Bone structure is also not clearly visible in patients with osteopenia and deformity.

**Isocentric Fluoroscopy-based Virtual Image-guided Surgery**

The isocentric C-arm automatically rotates around the patient, creating multiple fluoroscopic images in the surgical position while maintaining the relevant spinal anatomy at its center.\cite{67,68} Product software then reconstructs the images to provide coronal, axial, and sagittal views of the anatomy. The final image quality is not as high as CT, but it is higher than simple fluoroscopy. If the isofluoroscopy unit is connected to an image guidance workstation, the images for registration can be obtained after final positioning of the patient. Spinal exposure prior to registration is not necessary, and the surgeon-driven registration process is completely obviated, thus opening the field for minimally invasive or even percutaneous surgery.\cite{30,67} The accuracy is frequently good enough for UCS and CVJ surgery.\cite{41,44,68,69}

Axial plane tomography images that are received are reconstructed three-dimensionally. If connected to the navigation system, the real-time position of spinal elements and implants can be transmitted to the computer (real-time automatic registration), and any subsequent navigation procedure is actualized and more accurate. Furthermore, the final position of the implants can be checked directly at the end of implantation.\cite{68} It is also important to note that the radiation dose is reduced to 57% to 77%\cite{68} in comparison with standard CT protocols.

**Major Drawbacks**

The quality of images is reduced compared with CT, especially in obese patients, and the scan volume is limited (12 cm³)—approximately four cervical and three lumbar vertebrae.

**Intraoperative MRI-based Virtual Image-guided Surgery**

Similarly, intraoperative changes can be updated with MRI.\cite{70} This is often used in brain surgery in glioma resections, where not only the volumetric analysis and border line definition but also the functional or angiographic reimaging can be very helpful. This feature can be used effectively in resection of intramedullary tumors but with difficulties in the navigation of bony spine structures.
Major Drawbacks

Drawbacks include the cost of the MRI scanner, size of the hardware, necessity of a nonmagnetic environment (room, instruments, and anesthesiology equipment), and ergonomic problems that are more expressed in MRI-based systems than in CT. MRI is good for soft tissue imaging, bad for bone.\(^ {71} \)

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### Real-Time Image-guided Surgery

Real-time image-guided surgery would ideally allow for continuous monitoring of the progress of surgical intervention. Although the current technology does not allow for real-time feedback, it does offer an immediate check of each step during surgical intervention. It also offers improved safety. The surgeon can adapt strategy according to changes caused by surgery or movement of target structures. For example, a change in bone structure after resection of a tumor, the extent of decompression, deformity reduction, and fracture fragment reposition are all automatically updated on successive scans. The angle and length of used drills or screws can also be modified. During consecutive steps of surgery, vascular or neural injury risk is minimized, depending on the amount of surgery (e.g., resection/reduction) done between repeated scans. Real-time image guidance has clearly opened the way for safe, minimally invasive or even percutaneous spine surgery. The improved safety and accuracy are counterbalanced by the time necessary for repeated scans, the need for a good radiographer or radiologist, the higher radiation exposure, the need for special table hardware connections for CT or MRI, and the surgeon’s discomfort caused by ergonomic problems.

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### Isofluoroscopy: Three-dimensional Fluoroscopy

With the technical improvements seen with isofluoroscopy, there has been an increasing number of articles describing the direct intraoperative control of implant introduction and extent of bone resections.\(^ {58,69} \) The isocentric C-arm is not as large as mobile CT scanners and offers some flexibility with regards to patient positioning and hardware use (standard operating tables with a Mayfield head holder can be used). Scan acquisition times are significantly shorter than those associated with CT or MRI.\(^ {57,72} \) The step-by-step nature of the procedure allows for the angle and the depth of drilling, as well as optimal screw positioning, to be monitored in real time. Good visualization of bony structures on fluoroscopy makes isofluoroscopy amenable to very delicate procedures, such as transpedicular or transarticular C2 screw placement and odontoid compressive osteosynthesis.\(^ {58,69} \) Real-time navigation can be combined with a virtual system if necessary. For example, the placement of an odontoid compressive screw can be guided by a combination of lateral fluoroscopy and isocentric C-arm virtual guidance instead of biplanar fluoroscopy.\(^ {58} \) Other important advantages of real-time imaging are the ability to account for motion of bone during reduction in cases of instability or deformity corrections and the ability to visualize the final position of fracture fragments after reduction.

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Major Drawbacks

The quality of scans is entirely dependent on the quality of acquired fluoroscopy images, which can be inadequate for obese patients, as well as those with osteopenia and deformities. Increased radiation exposure is another major disadvantage.

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### CT Real-Time Navigation

The use of direct spinal CT guidance in the past was common in percutaneous interventions\(^ {73,74} \) but was limited to procedures performed in stationary scanners in radiology departments and suites adapted for surgery and general anesthesia.\(^ {18,75} \) The development of mobile CT scanners connected to adapted radiolucent surgical tables allows surgeons to move to real operating rooms and perform larger open surgeries.\(^ {75,76} \) The range of potential real-time CT application is similar to that of isofluoroscopy; however, CT images are definitely of a higher quality than any fluoroscope currently available. Very fine bony anatomical structures can be visualized and reconstructed in 3D by computer software. Better accuracy and data actualization of this technique have made it possible for percutaneous procedures to be done in the UCS.

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Major Drawbacks

Early scanners have a small gantry and thus limited working space, acquisition times are longer than for the isocentric C-arm, a special operating table may be required, and there are ergonomic problems.

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### MRI Real-Time Navigation

The use of open MRI surgical systems has been referred to extensively in the literature as intraoperative MRI.\(^ {1,71} \) When the surgical area of interest involves soft tissues and is more important than the bony anatomy, MRI technology is the modality of choice. This is particularly true in cranial surgery; however, it can also be important in localization of soft tissue spinal pathologies. Extra- or intradural tumors, intramedullary tumors, and CVJ anomalies compressing the neural tissue can be imaged very well. During surgery, the extent of the tumor and the progress of its resection can be followed. In CVJ surgery, the adequacy of neural tissue decompression is clearly visualized.

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Major Drawbacks

The surgeon’s working space is limited, operative time is longer than in any other computer-guided method, and a
“nonmagnetic” room has to be equipped with compatible instruments and anesthetic equipment. Additionally, metallic implants create image artifact, the quality of images is lower in comparison with conventional MRI, and bone discrimination is limited. The startup costs currently limit the widespread application of this technology in spine surgery.

**Indications and Authors’ Experience**

Computer guidance is indicated in surgery involving the UCS and CVJ whenever the safety and accuracy of a procedure can be significantly improved. This is of particular importance when it comes to navigating invisible areas.

**Compression of Neural Structures at the Craniovertebral Junction**

Neural structures can be compromised by developmental or acquired deformity, trauma, inflammation, or tumor (Fig. 19.5). Localization and the extent of decompression can be monitored with virtual or real-time MRI or CT. In cases of pure bony compression, isocentric C-arm is sufficient.

**Navigation of the Clivus and Foramen Magnum**

Cranial image-guided surgery can be helpful in localizing the clival edge during transoral procedures. This is particularly useful when screws are to be anchored into the clivus. Image guidance allows a direct measure of the bone thickness at the point of screw entry (Figs. 19.6 and 19.7). Navigation may be used during skull base procedures involving occipital condyles and is sometimes indicated in atlanto-occipital fusions when the position of the hypoglossal canal is of importance.

**Atlas Computer Navigation**

The easily approachable and anatomically simple first cervical vertebra is usually not navigated even if it is a
part of complex instrumentations. Nevertheless, modern technology opened up a new application of percutaneously treated fractures of the first cervical vertebra. The case illustration demonstrates an example of minimally invasive real-time CT guidance.

## Computer Guidance of the Axis

Image-guided surgery can be used for any type of axis screw placement. The anterior approach can be complicated by a difficult registration process and unstable DRA attachment to the navigated vertebra. The posterior virtual image-guided surgery described below can be problematic in cases of deformity correction and fracture dislocations. Disadvantages of image-guided surgery involving the axis include the fact that the mobile part of the spine or fracture fragment cannot be registered preoperatively, perioperative data update is time-consuming, and the system depicts only one actual position of the target in space. This is particularly apparent in posterior reduction of hangman’s or atypical miscellaneous fractures of the axis. Real-time CT-based guidance was developed to overcome these issues.\(^1\) Using CT-based systems, Judet transpedicular screw compressive osteosynthesis\(^3,4,3,4,4,7,8\) can be performed effectively in patients with reducible Effendi type II fractures without disk bulge or, more appropriately, in Effendi type I injuries, where the fracture gap is \(\geq 3\) mm on the CT scan. The procedure itself is performed under general anesthesia, and the UCS is entered through a standard midline approach. The entry points are determined according to the computer-planned optimal trajectory. The gantry of the scanner is tilted to be in the same plane as the proposed implant path. Gradual

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**Case Illustration**

A middle-aged man presented with neck pain and no neurological deficits after a fall. He was diagnosed with a two-point fracture of the atlas with a slight unilateral lateral mass “overhang.” The transverse atlantal ligament was intact on CT and MR images. Given the displacement and intra-articular nature of the fracture, percutaneous, direct real-time CT-guided osteosynthesis with fracture reduction was performed under general anesthesia using a CT scanner (GE Sytec 3000; GE Healthcare, Milwaukee, Wisconsin) in the radiology department. An appropriate screw trajectory was planned (Fig. 19.8), and fracture reduction and fixation (direct osteosynthesis) was achieved using a Kirschner wire and a cannulated lag screw technique (Fig. 19.9). Anatomical alignment of the lower articular surface of C1 (Fig. 19.10) was achieved in a relatively short procedure (90 min) with no associated intraoperative morbidity. Follow-up (18 mo) demonstrated satisfactory alignment and healing of the intra-articular fracture (Fig. 19.11). The patient was pain-free and had no limitation of neck rotation with minimally restricted flexion and extension. The advantage of intraoperative, real-time CT guidance was well demonstrated in this case, allowing for a safe, minimally invasive fracture fixation in the UCS. Without the current technology, such intervention would not be possible.
Fig. 19.8 A 1.1-mm Kirschner wire gradually passes through the C1 lateral mass according to the preoperative plan. (a) Plan of the appropriate screw trajectory. (b, c) Gradual wire introduction with final anterior atlas cortex penetration. (d) K wire attached to the battery-operated drill being introduced percutaneously.
Fig. 19.9a–d  A cannulated screw being inserted over the K wire.

a  Cannulated drill with external 2-mm diameter guided along the wire percutaneously.
b–d  Gradual advancement of the screw along the K wire.

Fig. 19.10a, b  Reduction of the fracture gap by the bicortical lag screw.
a  Before final tightening.
b  After final compression of the fracture site.
screw introduction (2-mm steps) is monitored with consecutive CT scans. Final tightening of the lag screw results in fracture compression, and the most appropriate screw length can be chosen based on navigation (Fig. 19.12). The ongoing development of mobile scanners, isocentric C-arms, and computer software will allow for similar procedures to be done percutaneously in the near future.

Guidance of C1–C2 Fusions

Currently, there are two commonly used methods of C1–C2 fixation offering immediate stability and high fusion rates and obviating the necessity for external bracing: the atlantoaxial screw fixation technique developed by Magerl in 19874 and Goel’s2 method of solid connection between the C1 lateral mass screw and the C2 transpedicular screw (Fig. 19.13). Both techniques involve passage of a screw through the axis, which is a potential source of morbidity. Anatomical studies36,45 do not seem to favor either of these techniques with respect to possible vertebral artery injury; however, it would appear logical that the transpedicular screw is safer to place due to direct visibility of the internal isthmus ridge and better angle of introduction. The few, rather sparse, studies documenting the results of Goel’s method of C1–C2 fixation do not describe a single case of vertebral artery injury.2,24,25 However, in the retrospective survey of the American Association of Neurological Surgeons/Congress of Neurological Surgeons, Wright and Lauryssen28 identified a 4.1% incidence of recognized vertebral artery injuries during transarticular screw fixation. The actual incidence could be even higher because of low survey response (26.6%) and the possibility of unknown vertebral artery tears. Due to collateral flow from the other vertebral artery, the incidence of neurological deficit was only 0.2%. In the same series, the screw misplacement rate was as high as 15%. In the second-largest series, Gluf et al.27 found the incidence of neurological deficit to be 2.6%, but one patient died. Conversely, the Swiss retrospective study79 describing

![Fig. 19.11a–c Images at 18 months demonstrate a satisfactory alignment and healing of the intra-articular fracture.](image)

- Axial CT scan of the healed fracture.
- Sagittal reconstruction.
- Plain lateral radiograph.

![Fig. 19.12a–c Axial CT scans and a plain radiograph showing a successful real-time CT-guided compressive osteosynthesis of hangman’s fracture.](image)

- One of the initial scans with partially introduced K wires.
- Final compression of the fracture with lag screws.
- Postoperative lateral radiograph.
161 surgeries did not reveal any vertebral artery injuries, but the tap was not regularly used; therefore, some injuries could be unrecognized.

Vertebral artery injury can occur if the trajectory of drilling or screw placement is too low. This is often caused by a desire to reach the anterior tubercle of the atlas as a fluoroscopy target. The anatomy of the C2 vertebra, rather than the position of the atlas, needs to guide screw trajectory in patients with rheumatoid arthritis with settling of the atlas (Fig. 19.14), after odontoid process resections (C1 anterior arch included), or in nonreduced dislocations. Therefore, it would appear very reasonable in those situations to use a surgical navigation system to guide the screw through the isthmus of C2.

When a CT-based virtual image-guided surgery system is used, the image set is obtained by a CT scanner according to a specific protocol and transferred to the workstation. Although the data need to be comprehensive, the focus should be on the C2 vertebra to create a precise 3D model. The virtual preoperative planning can then start, and the feasibility of the pars to accept a trans-isthmic screw can be tested (Fig. 19.15). Safe placement of a 3.5-mm screw should be possible as long as a 4- to 5-mm corridor exists along the projected path. The clearly visible and anatomically defined fiducial points are then determined (Fig. 19.16) for registration purposes. The patient is positioned on the surgical table, and all necessary hardware (C-arm, workstation, screens, etc.) are ergonomically positioned. A standard midline upper cervical approach is used to expose C1 and C2, including C2–C3 joints. The bone surface of previously determined fiducials has to be clear of soft tissue. A DRA frame is firmly attached to

**Fig. 19.13a, b** Lateral radiographs of a commonly used methods of atlantoaxial fixation.
* a Magerl’s transarticular fusion with posterior grafting.

**Fig. 19.14a–c** An incorrect trajectory of a second screw being inserted in a patient with rheumatoid arthritis and atlas settling. Injury to the vertebral artery caused by a tapper was successfully treated with a short plug screw.
* a Atlas settling (sagittal MRI) with the lower line demonstrating the wrong and the upper line the correct direction of drilling.
* b The tapper in a position of vertebral artery injury.
* c A short screw introduced as a plug to stop the vertebral artery bleeding.
the C2 spinous process in such a way as to not hinder the operation and be visible to the optoelectronic camera. The registration of the C2 spatial position is then performed, and instruments are tracked. The virtual picture can be continuously compared with the actual visible anatomy to avoid any registration mistakes caused by vertebral movement (e.g., after drilling of the first screw hole).

Atlas dislocation (if not reduced by positioning of the patient) can often be reduced by open C1–C2 wire fixation or C2 traction. The authors prefer to do this step first not only to reduce the dislocation but also to limit any movement of the atlas during drilling and tapping. Such movement could disrupt the continuity of the screw path and screw passage through an articulation. Screw entry points are marked with a high-speed drill in planned positions, and a tracked drill is navigated through the largest isthmus available first. The diameter of the pars interarticularis is not the only rate-limiting aspect for a correct cannulation of the pars. Frequently, a sharp entry angle necessitates the creation of a separate stab wound and the use of a trocar, cannulated system to correctly deliver the isthmus screw. This seems to be the case in obese patients and those with a thoracic kyphotic deformity.

The hole is tapped with a tracked tapper, and an appropriately long screw (chosen according to the preoperative surgical plan) is placed. Usually, the accuracy of a navigated procedure is further confirmed by lateral fluoroscopy. Anatomical knowledge and a direct visualization of the internal isthmus ridge serve as a second line of defense against inadvertent events. Correct implant position should

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**Fig. 19.15a–c** The feasibility of an insertion of transarticular C1–C2 screw is assessed using three-dimensional (3D) virtual CT and navigation software.

- **a** A 3D model in which the angle and entry point of a drill are simulated.
- **b** Sagittal reconstruction of a possible transisthmic screw trajectory (C1–C2 subluxation is not reduced in this patient with rheumatoid arthritis).
- **c** Oblique axial projection of the possible screw trajectory.

**Fig. 19.16** Screen shot of a computer workstation (StealthStation Treon Plus, Medtronic Surgical Navigation Technologies, Louisville, Colorado) documenting the localization of fiducial points during the registration process.
be checked on the first postoperative day with a thin-cut cervical CT, together with upright plain radiographs (including lateral and transoral views), to confirm the stability of the construct under axial loading (Fig. 19.17).

Despite the real concern of injury to the vertebral artery during instrumentation of the UCS, as raised by multiple anatomical studies, navigational systems were not used in the largest published series of Goel’s method of C1–C2 fixation to date. Virtual or real-time image guidance is indeed helpful during atlantoaxial fixation, especially when dealing with thin pedicles identified during preoperative evaluation. It is the authors’ opinion that 3D measurements and optimal pedicle screw trajectory planning should be done whenever this surgery is being considered.

**Conclusion**

Although it is clear that no navigational technology is a substitute for a surgeon’s knowledge and experience, new developments in imaging and guidance techniques can substantially contribute to the accuracy and safety of current surgical performance. This is of significant importance when it comes to anatomically delicate areas, such as the upper cervical spine and craniocervical junction. In the near future, image-guided application processes will become faster with better real-time data acquisition, allowing a broader range of minimally invasive or even percutaneous and robotic surgeries of the UCS and CVJ.

**References**


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**Fig. 19.17a–c** Postoperative CT reconstructions showing optimal transarticular screw placement.

a The entire screw path on oblique transverse reconstruction showing an adequate screw position and length.

b A safe position of the screw in the right isthmus.

c The vertebral artery groove is not violated in the left pars interarticularis (note the reduced position of the atlas in the same patient as in Fig. 19.15).


With the recent advances in neurosurgery and neuroimaging, a more aggressive approach to lesions that were once regarded as inoperative is now possible. The craniovertebral junction (CVJ) is one of the most complex and challenging areas in skull base and spinal surgery. It is essential for neurosurgeons to avoid damage to critical neural and vascular structures while removing lesions in the CVJ. Intraoperative neurophysiological monitoring can contribute to safe surgical performance.

Intraoperative neurophysiological monitoring comprises evoked potential monitoring and functional mapping. Evoked potential monitoring involves the continuous observation of the functional integrity of a specific neural pathway. It enables direct feedback in terms of the integrity of neural function and informs the neurosurgeon as to what is going on during surgery. Functional mapping covers neurophysiological localization of a specific neural structure or pathway in the surgical field. It guides the neurosurgeon through the functional anatomy. Thus, the neurosurgeon can avoid direct damage to neural structures. To perform functional mapping, the neurosurgeon has to interrupt surgery from time to time to perform stimulation. This is the shortcoming of functional mapping. In general, evoked potential monitoring and functional mapping are combined. This is not always the case in CVJ surgery, as most of the surgical pathology is in the extradural space, and functional mapping cannot be applied unless cranial or peripheral nerves are involved. Evoked potential monitoring thus serves a more important role than functional mapping during this part of CVJ surgery.

In this chapter, the author describes clinical aspects of intraoperative neurophysiological monitoring in surgery for the CVJ that was performed mainly at the National Center for Child Health and Development (NCCHD) in Tokyo, Japan. Special emphasis is placed on clinical application of motor evoked potential (MEP) monitoring and its usefulness for surgery in and around the CVJ.

A 15-year-old boy diagnosed with Hajdu-Cheney syndrome had several previous surgeries at other institutions, including foramen magnum decompression and syringosubarachnoidal shunt. However, progressive gait disturbance, tetraparesis, and intractable headache forced him to live a bedridden life. Swallowing disturbances developed slowly, and he was referred to our institution. Preoperative computed tomography (CT) and magnetic resonance imaging (MRI) showed a marked basilar impression, with the clivus running horizontally and the tip of the odontoid process penetrating the brainstem. Because of the decompression of the suboccipital bone that was previously done, the cerebellar hemispheres hung over the upper cervical spine. Due to this condition, anterior decompression through the transoral transpalatal approach was chosen as a surgical treatment.

The question here was to identify the best intraoperative neurophysiological monitoring method that could be applied during the anterior decompression procedure performed in this critically ill young patient. Conventional evoked potential monitoring, such as the somatosensory evoked potential (SEP) or the auditory brainstem response (ABR), may be helpful in such cases to monitor the functional integrity of the brainstem and spinal cord. However, the neurosurgeon’s main concern during such surgery is that the motor function can be completely lost if further compression to the brainstem occurs. The answer in this case was MEP monitoring. Figure 20.2 shows pre- and postoperative recordings of the MEP, and Figure 20.3 demonstrates continuous MEP monitoring during the transoral transpalatal anterior decompression. Although there was some reduction of the MEP amplitude by the end of the surgery, the MEP remained robust throughout the procedure, and the boy awoke with no further neurological deterioration. Postoperative CT and MRI revealed satisfactory anterior decompression of the brainstem and spinal cord after surgery. Preoperatively, syringomyelia was present, and it decreased in size after surgery was performed. The boy received occipitocervical/upper cervical instrumentation 1 month later. He was discharged and was able to walk with the aid of a walker.

Case Illustration

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Fig. 20.1 A 15-year-old boy with Hajdu-Cheney syndrome with severe basilar impression.

a Preoperative computed tomography (CT) and magnetic resonance imaging (MRI) demonstrate that the odontoid process seems to penetrate the brainstem.

b Postoperative neuroimaging shows satisfactory brainstem decompression after surgery. The cervicothoracic syrinx also improved after surgery.

Fig. 20.2 Pre- and postoperative motor evoked potentials (MEPs) show approximately the same features of the muscle MEP recorded from the abductor pollicis brevis (APB) muscles. The results indicate that the motor function of the patient should be preserved during the decompression surgery.
Intraoperative Neurophysiological Monitoring: Motor Evoked Potentials

MEPs are elicited by transcranial electrical stimulation and recorded from either the spinal epidural space or the limb muscles. Electrodes for transcranial electrical stimulation are placed at C3 and C4 (10–20 international electroencephalographic [EEG] electrode system). The anode is a stimulating electrode, contrary to peripheral nerve stimulation. When unilateral stimulation is preferred, a C3/C4+ versus Fz− or C3/C4+ versus Cz− stimulating montage is used to elicit MEPs (Fig. 20.4). Parameters for transcranial stimulation are given in Table 20.1.

The MEP recorded from the spinal epidural space (epidural MEP) was obtained through the catheter electrode inserted percutaneously on the lower cervical or upper thoracic level (Fig. 20.5). The spinal epidural MEP consists of D (direct) and I (indirect) waves. The D wave is elicited by activation of the cortical motoneurons directly, and the I wave from its transynaptical activation via the cortical interneurons. Single transcranial stimulation can evoke several I waves following the D wave. Therefore, because a D wave reflects specific information regarding the functional integrity of the corticospinal tract (CST) and is less influenced by anesthesia and other nonsurgical factors, it is used for intraoperative MEP monitoring.

The advantages of epidural MEP are that the D wave amplitude is usually stable, and its change correlates with the damage to the CST. Additionally, the D wave can be elicited by a single transcranial stimulus, and muscle...
relaxants can be used during surgery without influencing D wave monitoring. The disadvantages are that the placement of the recording electrode into the spinal epidural space is an invasive procedure, and evaluation of the individual CST is difficult.

MEP recorded from the limb muscles (muscle MEP) is obtained from electrodes placed at the target muscle. The preferable muscle for muscle MEP monitoring is the abductor pollicis brevis (APB) for the upper extremity, and the tibialis anterior (TA) or the abductor hallucis brevis (AHB) for the lower extremity. Transcranially, a short train of stimuli is applied to elicit muscle MEP because during anesthesia, multiple descending drives are necessary to bring the resting potential of the α-motor neurons up to the firing level, which will consequently transmit signals to the peripheral nerves and muscles. Figure 20.6 shows an example of muscle MEP recorded from an 18-year-old male patient with Chiari I malformation. Muscle MEP was elicited by transcranially applying a short train of stimuli (C1/C2, train of five stimuli) and continuously recording from the bilateral APB, TA, and right diaphragm.

The advantage of muscle MEP monitoring is that it enables surgeons to evaluate the individual motor function of each extremity without using an invasive procedure, such as placing an electrode in the spinal epidural space. The disadvantages are that the amplitude of muscle MEP is often unstable, and it fluctuates during surgery. In addition, muscle relaxants are not used with anesthesia, and total intravenous anesthesia is required for intraoperative monitoring of muscle MEP. It is necessary to confirm that all responses are present before starting the MEP monitoring by checking the train of four stimulation techniques being applied to the peripheral nerve. This is important to do so as to exclude any influence of residual

Table 20.1 Parameters for transcranial stimulation for motor evoked potentials

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Position of electrodes</td>
<td>C3/C4 or C1/C2</td>
</tr>
<tr>
<td>Waveform</td>
<td>Square wave</td>
</tr>
<tr>
<td>Train</td>
<td>N = 5</td>
</tr>
<tr>
<td>Interstimulus interval</td>
<td>2–4 msec</td>
</tr>
<tr>
<td>Duration of stimulation</td>
<td>0.5 msec</td>
</tr>
<tr>
<td>Frequency</td>
<td>4.3 Hz</td>
</tr>
<tr>
<td>Intensity</td>
<td>Suprathreshold intensity (&lt; 200 mA)</td>
</tr>
</tbody>
</table>

Fig. 20.4 Placement of electrodes for transcranial electrical stimulation for MEP monitoring based on the 10–20 international electroencephalographic (EEG) system.

Transcranial electrical stimulation
Position of electrodes for stimulation

Muscle MEP
Transcranial electrical stimulation

Motor evoked potentials
Train of stimuli
0.5 msec
2–4 msec

Epidural MEP
Single stimulation
0.2–0.5 msec

Recording from epidural space
D wave
I wave

Recording from muscles
Train of stimuli
Duration: 0.5 msec
ISI: 4 msec

Fig. 20.5 Difference between the muscle MEP and epidural MEP. The muscle MEP is evoked by a transcranial train of stimuli and recorded from limb muscles. The epidural MEP is evoked through a single transcranial stimulus and recorded from a percutaneously inserted electrode placed in the epidural space. In general, the recorded response from the epidural electrode consists of a D wave followed by one or more I waves.
muscle relaxant used for intubation purposes or that may be accidentally administered later.

### Clinical Application of MEPs for CVJ Surgery

MEP monitoring is a practical intraoperative neurophysiological procedure for surgeries of the CVJ. Information obtained from MEP monitoring can be useful feedback on the well-being of the nerve structure at critical stages of the surgery.

The first step of CVJ surgery is the positioning of the patient’s head. Flexion or rotation of the neck is usually required for better access to the lesion. Manipulations of the neck under anesthesia can produce further compression of the spinal cord in some patients. MEP monitoring will help ensure that positioning of the patient on the operating table does not further compromise nerve structures. Figure 20.7 shows a complex atlantoaxial dislocation associated with os odontoideum. The hypoplastic C1 lamina and superior left facet are invaginated into the posterior fossa. MEP is recorded before and after positioning of the patient on the operating table to confirm that the lesion does not produce further compression of the spinal cord at the CVJ (Fig. 20.8).

![18 year old boy: Chiari I malformation with syringomyelia](image1)

**Surgery:** Suboccipital decompression with dural plasty

**Train of stimuli**

- **Intensity:** 160 mA
- **Train:** x5
- **ISI:** 4 msec
- **Rate:** 4.3 Hz
- **Average:** x4

![100 uV](image2)

**Fig. 20.6** An 18-year-old male patient with Chiari I malformation. The MEP was recorded from the upper extremity (APBs), lower extremities (tibialis anterior muscles, TAs), and right diaphragm by transcranial train of stimuli. For this patient, electrodes for the transcranial stimulation were placed on the C1 (anode) and C2 (cathode).

![Fig. 20.7 Preoperative MRI and CT scans of a 4-year-old girl with atlantoaxial dislocation due to os odontoideum](image3)

The cervicomedullary junction was severely compressed. Details of the bony anomaly were revealed by the three-dimensional (3D) CT scan. Part of the hypoplastic C1 lamina was invaginating into the foramen magnum, contributing to the cervicomedullary compression. 1, os odontoideum; 2, base of the odontoid process; 3, part of the hypoplastic right C1 lamina; 4, dislocated left C1 superior facet.
Fig. 20.8 Intraoperative position of the patient (upper left). A bone flap harvested from the skull used for the occipital-C2 posterior fixation (lower left). MEP monitoring shows no significant change in amplitude before and after positioning, and the MEP remains the same amplitude after surgery (right).

Fig. 20.9 On the left: 3D and sagittal reconstruction of CT scans. Atlantoaxial subluxation was corrected, and no stenosis at the foramen magnum was observed. On the right: records of continuous MEP monitoring during the surgery demonstrate stability of response.
MEPs were continuously monitored during the occipital-C2 fixation (Fig. 20.9). When passing a thread at the foramen magnum or C2 lamina, the surgeon must be sure that the procedure is safe. Continuous MEP monitoring during surgery can help avoid neural damage.

Finally, MEP monitoring helps confirm that surgery was successful in terms of preserving the motor function even before the patient wakes up from anesthesia. In general, if the spinal MEP amplitude remains > 50% from the baseline value, the patient’s motor function will be the same as that seen preoperatively. MEP amplitude < 50% indicates that the patient has suffered serious motor deterioration.9,10 Regarding muscle MEP monitoring, the presence of muscle MEP indicates that motor function was preserved. The loss of muscle MEP amplitude does not necessarily suggest that motor function is lost if at the same time spinal MEP is present. This indicates that there is simply transient postoperative motor deterioration that will recover later. For a more precise prediction of motor outcome, it has been strongly recommended that both spinal and muscle MEPs be monitored.11

Case Illustration

A 1-year-old baby girl diagnosed with achondroplasia was referred to the NCCHD because of developmental delay in motor function. Her parents noticed marked snoring during sleep with the head extended. MRI and CT revealed prominent stenosis at the foramen magnum and spinal cord compression. Before positioning the patient on the operating table, the MEP was recorded, and she was turned prone with the neck kept in a neutral position. The MEP showed no apparent deterioration after positioning (Fig. 20.10). The MEP was continuously monitored during surgery. When the foramen magnum decompression was started, the MEP amplitude suddenly augmented, then significantly deteriorated (Fig. 20.11). It was speculated that the partial decompression caused focal spinal cord compression at the edge where the decompressed dura bulged. Foramen magnum decompression was quickly accomplished. Consequently, the muscle MEP amplitude increased with lower amplitude, when compared with the baseline at the end of foramen magnum decompression. The amplitude of muscle MEP gradually recovered thereafter until the end of surgery. The patient showed no neurological deterioration. Her locomotion improved after surgery, and she was able to walk after 1 month.

**Fig. 20.10**  The chart on the left shows electrodes for transcranial MEP monitoring placed at C3 and C4. The position of the patient and the head are shown in the center and right charts. The MEP demonstrated no change before and after positioning and was maintained throughout the surgery.
After this experience with transient loss of MEP, the author modified the surgical technique of foramen magnum decompression. Instead of decompressing from one side, the whole range of bone over the foramen was thinned out first, then peeled out in a short period of time. Since then, no MEP deterioration during foramen magnum decompression has been encountered.

Neurophysiological Mapping at the CVJ

Functional mapping of the neural structure/pathway during surgery is relatively straightforward compared with evoked potential monitoring using MEP. The area of concern in the surgical field is stimulated by a mono- or bipolar stimulating probe, and the muscle response is recorded as an electromyogram response. The author prefers to use a monopolar stimulating probe because a bipolar probe needs to have contact with the nerve tissue on both ends, and in a narrow surgical field, this could be difficult. The types of anesthetics used are the same as those for monitoring muscle MEP, and total intravenous anesthesia without the use of muscle relaxants is recommended. The intensity of electrical stimulation should be \( < 2 \) mA when the spinal cord or brainstem is directly stimulated. After obtaining muscle response, the intensity of stimulation is then adjusted to the suprathreshold level. When the nerve root is directly stimulated, a stimulation intensity of 1 to 2 mA is sufficient to elicit the muscle response. The intensity of stimulation in a nonaffected motor root, which elicits a muscle response, can be as low as 0.2 mA. Parameters for nerve root stimulation are shown in Table 20.2.

Application of mapping techniques in CVJ surgery is indicated mainly for the intradural part of the surgery. Tumors involving the cranial nerves are the best indication for using mapping techniques. The procedure is

<table>
<thead>
<tr>
<th>Table 20.2</th>
<th>Parameters for nerve root stimulation</th>
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</thead>
<tbody>
<tr>
<td>Cathode</td>
<td>Handheld monopolar probe</td>
</tr>
<tr>
<td>Anode</td>
<td>Cervical muscles in the operative field or Fz</td>
</tr>
<tr>
<td>Waveform</td>
<td>Square wave, single pulse</td>
</tr>
<tr>
<td>Duration of stimulation</td>
<td>0.2 msec</td>
</tr>
<tr>
<td>Frequency</td>
<td>0.5–4.0 Hz</td>
</tr>
<tr>
<td>Intensity</td>
<td>0.2 mA or more for the motor root</td>
</tr>
<tr>
<td></td>
<td>0.4–2.0 mA for brainstem mapping</td>
</tr>
</tbody>
</table>

Fig. 20.11  Left: severe stenosis and spinal cord compression at the foramen magnum (upper), which was satisfactorily decompressed after surgery (lower). Right: continuous MEP monitoring during surgery. The MEP augmented and suddenly disappeared in the middle of foramen magnum decompression, then slowly recovered.
Intraoperative Neurophysiological Monitoring in Surgery for the Craniovertebral Junction

Functional mapping of the cervical roots and the spinal accessory rootlets is an indispensable tool for intradural rhizotomy for spasmodic torticollis. Figure 20.12 shows an example of functional mapping in a patient with spasmodic torticollis. Following the mapping, the spinal accessory rootlets, which elicit muscle response in the sternocleidomastoid muscle, were cut to treat torticollis.

Intraoperative Neurophysiological Monitoring for Tumors Involving the Brainstem

If a tumor is located in the intramedullary part of the brainstem or the upper cervical cord, brainstem mapping (BSM) would be indicated before reaching the tumor. Details of BSM have been reported elsewhere, and a brief summary will be described later in this chapter.13–16

Regarding MEP, monitoring the functional integrity of the cranial motor nerves as well as the CST is required. The muscle MEP recorded from cranial motor nerve innervated muscles is referred to as corticobulbar tract (CBT) MEP.17 Parameters for transcranial stimulation for CBT MEP are approximately the same as that for muscle MEP. Muscles for CBT MEP monitoring are the same as those used for BSM (the orbicularis oculi and oris for the facial nerve, the intrinsic tongue muscle for the hypoglossal nerve, and, in some cases, the retropharyngeal muscle or the cricothyroid muscle for the glossopharyngeal/vagus nerves). Care should be taken to distinguish between direct stimulation of the cranial motor nerves and stimulation via CBT. To exclude possible direct stimulation, it is recommended that a single transcranial electrical stimulus be delivered first. If the muscle response is recorded following a single stimulus, then the response is regarded as being generated by direct stimulation of the motor cranial nerves. Only the muscle response evoked following a train of transcranial stimulation can be accepted as CBT MEPs.

Conclusion

Intraoperative neurophysiological monitoring can help prevent/diminish neurological injuries in CVJ surgery. In general, a combination of both evoked potential monitoring and functional mapping techniques is important to preserve the functional integrity of the central nervous system and motor cranial nerves. However, application of these techniques would depend on the lesion to be operated. Evoked potential monitoring, especially MEP monitoring, would be enough for extradural lesion surgery. If the lesion is located in the intradural space, a combination of MEP monitoring and functional mapping, such as nerve root mapping or BSM, is preferred. Identification of
A 2-year-old boy was referred to the NCCHD due to vomiting and altered consciousness. Neuroradiological evaluation revealed a tumor extending around the brainstem through the lateral recess of the fourth ventricle. Following endoscopic third ventriculostomy and temporary installation of ventricular drainage, direct surgery was scheduled. Because the tumor was located in the fourth ventricle and extended toward the cerebellopontine angle, potential damage to the cranial motor nuclei and their peripheral parts, especially the right facial nerve, was highly probable. It was decided to monitor the CBT MEPs together with muscle MEPs from the upper extremity. The ABR was also monitored, but only intermittently, when required to evaluate gross brainstem functional integrity.

Figure 20.13 demonstrates the initial evaluation of CBT MEP. As shown on the left side of the figure, there was no muscle response by applying a single stimulus at the intensity of 90 mA. Small responses were elicited when stimulated with the train of five stimuli with an intensity of only 40 mA. Steady muscle responses from the face, tongue, and hands were recorded when the stimulation intensity was increased to 70 mA (Fig. 20.14). CBT MEP was continuously monitored during the tumor resection, and it remained stable until the end of surgery (Fig. 20.15). After removing part of the tumor, for the tumor extending in the right lateral recess, a cerebellar peduncle BSM was performed. The brainstem and the floor of the fourth ventricle were stimulated repeatedly with recording of muscle responses to locate the nuclei of the cranial motor nerves (Fig. 20.16). By the end of tumor resection, CBT MEP remained robust (Fig. 20.15). The patient awoke without any disturbance in facial or lower cranial nerve function. Postoperative MRI revealed gross total resection of the tumor extending to the brainstem (Fig. 20.15).

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![Fig. 20.13](image) A single transcranial electrical stimulus evoked no muscle response even at 90 mA. Small compound muscle action potentials were recorded with a transcranial train of stimuli at 40 mA. The response was regarded as corticobulbar tract (CBT) MEPs.
Intraoperative Neurophysiological Monitoring in Surgery for the Craniovertebral Junction

Fig. 20.14  CBT MEP recorded before and after surgery showed no change.

Fig. 20.15  Continuous monitoring of CBT MEP. Images show the tumor, which extended to the left lateral recess of the cerebellopontine angle. The patient showed no neurological deficit after the tumor was totally resected.
Fig. 20.16 After exposure of the floor of the fourth ventricle, brainstem mapping (BSM) was performed. The intraoperative photograph is from BSM using a monopolar electrode. The graphs show muscle recordings during BSM. The response obtained in different cranially innervated muscles depends on the position of the monopolar electrode on the floor of the fourth ventricle.

References

Occipitocervical fusion can be done successfully, especially with the recent advent of internal fixation devices available for treatment of instability. Occasionally, the spine surgeon will be faced with having to revise one of these constructs. As with all surgeries requiring a revision surgery, the first steps are recognizing the complication and identifying the underlying reason for failure of the previous surgery.

**Background**

The occipitocervical junction is a complicated interaction between the cranium and the upper cervical spine. It comprises the atlanto-occipital and atlantoaxial joints and is the most mobile section of the cervical spine, stabilized only by capsuloligamentous attachments. As a result, it poses significant challenges for fusion options, and attempts to stabilize this region require a thorough understanding of the anatomy. This becomes even more imperative when assessing possible reasons for failure and performing revision surgery.

The occiput has several bony landmarks that are advantageous to identify. The external occipital protuberance (EOP) is a dense ridge of bone that marks the thickest portion of the cranium. It measures 11.5 to 15.0 mm thick in men, 10.0 to 12.0 mm in women. The superior nuchal line is a similar thickened ridge that runs laterally from the EOP. The inferior nuchal line runs parallel but inferior to the superior nuchal line. The transverse sinus lies at the superior nuchal line, and the confluence of the transverse sinus is at or above the EOP; caution must be taken to avoid placing screws that may disturb the intracranial contents there. The occiput also takes an acute curve anteriorly from the superior nuchal line to the foramen magnum. The occiput interacts with the cervical spine through two articular condyles on either side of the foramen magnum. The occiput occupies a significant portion of the suboccipital bone and the cervical spine. Furthermore, these constructs must be able to accommodate adjustments in positioning the patient intraoperatively into an appropriately neutral occipitocervical position prior to final tightening and preparation for fusion. Consideration should also be given to postoperative care and the need for immobilization, depending on the rigidity of the construct.

**Historical Techniques for Occipitocervical Fixation**

Historically, internal fixation was inadequate to fix the spine rigidly, and external immobilization was often necessary to provide stability. With current instrumentation, rigid external immobilization is usually not necessary. Although many of the techniques described below are...
no longer used in the United States, we include a brief description of them because, when performing revision surgery in this region, readers may encounter them. Occasionally, one of these techniques may be necessary during revision surgery, if all of the available fixation points have been used or destroyed with previous operations.

### Onlay Bone Graft without Internal Fixation

Onlay bone grafting was the first technique described for occipitocervical fusion. Stability until a bony fusion was achieved was with external fixation—cranial tongs with bed rest, a Minerva jacket, and/or a postoperative halo vest. Foerster first described this technique in 1927. A fibular graft was inserted between the occiput and C7 to stabilize a progressive atlantoaxial dislocation after an odontoid fracture. Graft choice and methods to fashion the graft have varied. In general, autograft has been favored, but allograft has also been used. Techniques have varied from wedges to struts spanning the occiput to C2 or from the occiput to various levels in the subaxial spine. Different types of grafts have been used, including cancellous and corticocancellous strips or wedges (unicortical, bicortical, or tricortical), as well as cortical struts, such as the fibula, humerus, and femur. Elia et al. reported their results with onlay grafting without internal fixation. They used the autologous iliac crest and reported an 89% fusion rate. Although a successful fusion can be obtained, most surgeons are skeptical that such a high fusion rate can be obtained without internal fixation. An important criticism of onlay grafting alone is the need for external traction/immobilization, which can be difficult for many patients to tolerate. Another limitation of this technique is in malalignment reduction or maintenance of reduction. Postoperative traction is also concerning in patients with ligamentous instability because distracting forces may be contraindicated. Some surgeons continue to employ this technique in children because of the tremendous osteogenic potential in this patient population, as well as the difficulty with internal fixation in pediatric patients with inherently small osseous structures. Most surgeons would still recommend at least wiring in these patients to assist with stability until a fusion is achieved.

### Onlay Bone Graft with Wiring

The first modern reference to internal fixation in the cervical spine is from 1891 in a report by Hadra, who performed cervical spinous process wiring. Wiring techniques of C1 and C2 developed and usually involved sublaminar placement at C1 and either sublaminar or spinous process wiring of C2. The first published study in which wire was used in fixation of bone graft to the occipital cervical junction was by Cone and Turner in 1937. Different occipital wiring techniques have been tried. Passage of wire through the foramen magnum and out through burr holes in the occiput can be complicated by the very adherent dura to the bony margin of the foramen magnum. To avoid this, the foramen magnum is often widened posterocentrally to ease the passage of wires out of the adjacent burr holes. Wertheim and Bohlman popularized a technique that avoided burr holes in the occiput by using a trough at or near the EOP and tunneling wire beneath the outer table. This technique circumvented the problems associated with passing the wire through burr holes and potential cerebrospinal fluid (CSF) leaks. A second wire was placed sublaminar at C1, and a third was passed through a drill hole in the spinous process of C2. Autologous corticocancellous strips of the ilium were fixed with this triple-wire technique. In Wertheim and Bohlman’s study, 13 patients underwent an occipitocervical fusion using this technique, and all 13 were noted to have a solid fusion at 2 to 7 years’ follow-up. Despite such good reports, wiring does not provide rigid fixation; therefore, patients still require postoperative treatment with a halo for supplementary external fixation, and many surgeons believe that an improved fusion rate is obtained with more rigid internal fixation (Fig. 21.1).

### Rod and Wire Technique

In 1986, Ransford et al. introduced the first rigid internal fixation of the occipitocervical junction, which used a looped Luque steel rod to bridge the occiput to the cervical spine. The rod was carefully contoured to contact the occiput and cervical lamina for increased rigidity and was fixed with occipital and sublaminar wires. Bone graft assisted in fusion. Other surgeons have made modifications to this technique, including the use of a threaded Steinmann pin and specially designed threaded rods, which arguably improve wire fixation by not allowing the wires to slip on the rod as easily. The rod and wire technique provides more rigid fixation and allows surgeons to reduce and hold reductions at occiput–C1 and C1–C2. Despite the relatively rigid fixation of this construct, however, many surgeons still recommend supplementing the internal fixation with a postoperative halo. Another limitation to the rod and wire technique is seen in patients in whom a posterior decompression has been performed, as well as those who have posterior element fractures, because the typical sublaminar or spinous process wiring would be compromised (Fig. 21.2). Surgeons have reported on the use of facet wiring in these cases, but this fixation has not been commonly used. There is also some concern over the use of wire fixation in patients with osteoporosis and rheumatoid arthritis, as there is a significant risk of screw cut-out with posterior element wiring alone. Moskovich et al. reported on 150 patients with rheumatoid arthritis who presented with vertical instability and multilevel involvement. The patients were treated with a contoured occipitocervical loop affixed with sublaminar wires and no bone grafting. The authors found that there was no clinical difference between those who were fused and those with bone...
grafting. They went on to assume that the clinical improvement might be attributable to stabilization of the joint rather than to osseous fusion. Apostolides et al. reported a 97% success rate for achieving a stable occipitocervical construct in 39 patients. They concluded that rigid segmental fixation of the craniovertebral junction using a wide-diameter, contoured, threaded Steinmann pin and supplemental autograft creates excellent fusion with minimal complications.

Fig. 21.1a–d
a, b Lateral and anteroposterior radiographs of a patient who had undergone an occipitocervical arthrodesis with wires in the remote past. Although this often resulted in successful fusions, it required postoperative halo immobilization. Upon presentation to us, there was an obvious pseudarthrosis. In addition, there was subaxial spondylosis and radiculopathy.

c, d Postoperative anteroposterior and lateral radiographs using modern instrumentation. The revision was performed with a skull plate that was then attached to a screw-rod system to fixate the cervical spine. This provides a rigid construct that obviates the need for a halo and increases the likelihood of a solid fusion.

■ Rod and Hook Technique

More recently, Faure described an occipital fixation technique in which hooks are placed back to back in the same burr hole, with one hook positioned up and the other down. This created an occipital clamp construct. The technique may be most useful in patients with significant osteoporosis, in revision occipital instrumentation cases, or when there is significant thinning of the occipital bone.
Another technique that may be recommended in this difficult patient population was described by Pait et al., who used an inside-out inverted bolt in the occiput, with the head of the bolt against the inner table of the occiput. This provided a broad surface area for fixation in the occiput and could be useful in patients with significant osteoporosis. One problem with this system, however, is that revisions can sometimes be challenging. Because the burr holes for the bolts are so large, much of the surface area for revision fixation and bone grafting is compromised (Fig. 21.3).

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c Magnetic resonance imaging (MRI) demonstrates severe cord compression due to basilar invagination and compression between C3 and the posterior fusion mass, showing kyphosis.

d, e Clinical radiographs demonstrating a lateral tilt as well as a kyphotic deformity.

f Intraoperative photograph. The skull is to the left and the spine to the right. There was a solid fusion mass connecting the skull to the spine. We decompressed the foramen magnum by burring through the cranial portion of the fusion mass and the posterior rim of the foramen magnum. In addition, we completely burred down the lateral aspects of the fusion mass until the dura was visualized. Then, we burred partially through the caudal aspect of the fusion mass to create a hinge, similar to a laminaplasty hinge. The fusion mass was then lifted up to decompress the foramen magnum and proximal cord.

We then placed three plate-rod constructs with screws into the lateral masses and pedicles, as well as into the fusion mass and the midline. This provided multiple points of fixation. In addition, using a different system than was used in the initial operation allowed us to achieve fixation into a different part of the skull.
Plate with Screw Fixation Techniques

The development of rigid fixation of the occipitocervical junction began with plate and screw fixation of the cervical spine, in particular, C1 and C2. Magerl and Seeman popularized C1–C2 transarticular screw placement, which could be combined with fixation of the lower cervical vertebrae using plates that extend distally and lateral mass screws in the subaxial cervical vertebrae. Other C1–C2 screw fixation techniques were needed, as transarticular screws proved to be technically challenging, even contraindicated, in patients with a small C2 pars interarticularis and in those with an aberrant vertebral artery. The decision as to whether a C2 pars can accept a transarticular screw is best evaluated with sagittal reconstructions of a computed tomography (CT) scan. Goel and colleagues were the first to introduce a technique that has become very popular for fixation of C1–C2 and avoids the potential challenges and complications associated with transarticular screws. They described plating/rod placement with fusion of C1–C2 using pedicle screws at C2 and lateral mass screw placement at C1.

Once rigid fixation of C1–C2 and the subaxial cervical spine was achieved, a truly rigid construct could then be developed to cross the occipitocervical junction. In 1991,
Grob et al. rigidly fixed the occipitocervical junction with plating from C1–C2 to the occiput using transarticular screws at C1–C2 and occipital screws. Goel and Laheri subsequently modified the fixation of the cervical end of the construct by advocating the use of insertion of a C2 pars screw either in isolation or in combination with a C1 lateral mass screw. These rigid fixation methods allowed for the discontinuation of routine postoperative halo placement, as well as an arguably increased rate of fusion.

Plating of the occipitocervical junction has been criticized primarily for three reasons. First, surgeons have found it difficult to contour the rigid plates to match the acute sagittal angle of the occipitocervical junction while maintaining alignment between the plate on the occiput and properly placed screws in the lateral aspect of the cervical spine (Fig. 21.4). Second, hole spacing in plates dictates where screws can be placed, which has led to suboptimal placement of screws that leads to suboptimal fixation. Third, plates need to be placed more lateral in the occiput so that they line up for screw placement in the upper cervical and subaxial spine. This lateral placement dictates that shorter screws be used for fixation of the plates to the occiput, as the thickness of the occiput has been shown to decrease as one deviates both laterally and inferiorly from the external occipital protuberance. The lateral occipital thickness in the region of the cerebellar fossa can be as little as 3 mm, which would require special short screws with short thread pitch. This placement could potentially lead to weaker fixation and higher risk of CSF leaks and epidural/subdural hematomas. The use of Y-type plates allows more centralization of the occipital screws, but this plating technique still requires difficult contouring.

**Combined Rod-Plate Fixation**

The development of rod fixation with multiaxial screws for the cervical spine was a major advancement in cervical fixation. Screw placement in the cervical spine was no longer dictated by the hole spacing in plates. Instead, screws could be placed independently and in a more anatomically ideal position, which improved fixation. The technique of rod-screw fixation of the cervical spine was extended to the occiput with rods that transitioned to occipital plates (Fig. 21.5). Plating continues to be the fixation of choice for the occiput, as it allows rigid fixation while maintaining a relatively low profile on the occiput, where coverage can be an issue. Although the cervical rod that transitions to an occipital plate provides significant improvements when compared with simple plating of the occipitocervical region, this technique can still require much contouring. The rod is easier to bend than a plate, but it still needs a very acute sagittal bend to match the profile of the occipitocervical junction. The plate needs to be in intimate contact with the occiput for improved fixation, and the rod needs to be contoured down to the cervical screws so that the screws are loosened/backed out when the rod is captured to the screw heads. This transition rod-plate also needs to be contoured in the coronal plane with an attempt to medialize fixation on the occiput, where screws can be longer due to the increased thickness of the occiput in that location. Despite the significant advancement over simple plating, some surgeons still find this combined rod-plate design somewhat difficult to use because of the contouring that is needed.

**Independent Occipital Fixation and Cervical Rod Instrumentation**

The most recent advancement has been the development of instrumentation that allows the surgeon to plate or fixate the occiput independently and subsequently connect this to the cervical rod-screw construct (Fig. 21.5b). Plates are often precontoured to fit the occiput, which makes placement easier. Some systems allow screws to be placed in the occiput near the EOP, where the skull is thicker, providing greater screw-plate fixation. These centrally placed occipital plates then have lateral offset connectors for cervical rod attachment, so minimal coronal bend is needed to join the relatively lateral cervical fixation to the occipital plate. This technique allows ideal placement of screws in both the occiput and the cervical spine. Given the acute angle at the occipitocervical junction, prebent rods are available to help with contouring. More recently, an adjustable rod hinge has been used in the occipitocervical junction so the sagittal angle of the rod can be easily adjusted. Once the proper sagittal angle is achieved, the hinge is locked into position and very minimal rod contouring is required.
Other systems feature a plate with a distal connection, which attaches to the rod. Yet another uses individual eyelets that fixate into various points in the skull, then connect individually to the rod. All of these systems have vastly improved occipitocervical fixation.

### Pseudarthrosis

Pseudarthrosis, which means “false joint,” is more reliably described as the presence of motion and failure of union at the intended site of fusion. Some key factors that should be considered are the surgical fixation technique, type of instrumentation, type of bone graft (allograft vs. autograft and cancellous vs. cortical), number of vertebral levels involved, external immobilization utilized, and patient factors, including general health, history of inflammatory disease (e.g., rheumatoid arthritis), use of tobacco, and overall patient compliance.

The reported rates of nonunion have varied from 0% to 50% using wires and plates. Others have even questioned whether a fusion might be necessary in certain cases, such as in patients with rheumatoid arthritis. As mentioned above, Moscovich et al. reported on the successful long-term use of a Ransford loupe without an arthrodesis in rheumatoid patients.

Fortunately, pseudarthrosis at the occipitocervical region is relatively rare. With rigid fixation, one can avoid bed rest and halo vest immobilization and still achieve excellent results. There is very little reported in the literature with regards to revisions secondary to pseudarthroses. However, several authors have advocated using autogenous bone graft, cortical-only grafting techniques, and allograft material. Additionally, bone morphogenetic proteins (BMPs) may help achieve fusions in recalcitrant nonunions.

The best way to diagnose a pseudarthrosis is with reformatted high-resolution fine-cut CT images. Low-resolution images can give the false impression of contiguous bone across the fusion mass in the presence of a pseudarthrosis. Plain radiograph and other imaging modalities usually can detect only very obvious pseudarthroses.

Once a symptomatic pseudarthrosis is identified with correlation of radiographic findings and complaints of pain and/or instability with or without neurological changes, the revision procedure must address the area of pseudarthrosis, as well as investigate the cause. Patients with rheumatoid arthritis have higher rates of pseudarthrosis.
due to their history of steroid use. Similarly, patients who have undergone radiation therapy to the occipitocervical region will have higher rates of pseudarthrosis. In this “higher risk” group, it is imperative to be meticulous when preparing the fusion bed and performing the decortication. Although less frequently needed with the newer instrumentation that is available, these patients may even warrant external immobilization postoperatively.

Depending on which method of fixation was utilized to gain access to the pseudarthrosis, a significant amount of effort will be dedicated to dissection and instrumentation removal (Fig. 21.7). If a wiring technique was used initially, a plating and screw technique is a viable option. Sometimes screws can be placed into the fusion mass if the lateral mass or pedicles are not usable (Fig. 21.3g).

Fig. 21.6a–c
a, b This patient had undergone multiple operations, and the treating surgeon had removed the existing occipitocervical instrumentation under the impression that there was a solid fusion. However, as Fig. 21.6b illustrates, the computed tomography (CT) scan clearly demonstrates a pseudarthrosis of the occipitocervical region. Although spiral CT is the best method to assess for a nonunion, it must be high resolution, as it can be difficult to visualize around instrumentation. Poor resolution can give the false impression of contiguous bone.

c Revised occipitocervical arthrodesis.
Failure of Fixation

Fixation failure can be the result of complications associated with insertion of instrumentation or direct mechanical failure of fixation. Insertion is technique and surgeon dependent, whereas direct mechanical failure of fixation is structure related, for instance, osteopenic or osteoporotic bone, gross instability, and pseudarthrosis. The earliest techniques that used stainless steel wires to secure bone grafts had wire breakage in up to 13% of cases. More recently, braided titanium cables have replaced wires due to their greater tensile strength, but they can also potentially loosen or break.

Screws have been used to secure plates or rods to the occiput, as well as to lateral masses of the cervical spine, but they can be a source of fixation failure. Screws can be improperly placed, pull out, or fracture. They have also been placed in the pedicles of C2 and across the C1–C2 joint. Such screws tend to be longer, and the failure rate is generally less than that with lateral mass and occipital screws, but they too can be associated with pullout and fracture. In the presence of screw fracture, a pseudarthrosis is also likely present.

Biomechanical studies have demonstrated that the greatest pullout strength is at the occipital protuberance. Studies have also found that bicortical screw placement had 50% greater pullout strength than wire or unicortical screw placement, but screws placed unicortically at the occipital protuberance offered acceptable pullout strength without the potential for complications seen with bicortical screw or wire placement.

Rod and plate fracture is rare. The force that is required to cause a fracture in a rod or plate will generally cause failure at the wire or screw level first. Extreme
bends in the rod or plate can increase the risk of breakage. Rod and plate contouring should be achieved with small incremental bends rather than a large acute bend, and recontouring the same rod or plate should be avoided. This is especially true with modern titanium rods, which are highly notch-sensitive. If a titanium rod is bent in one direction and subsequently bent back, it is highly prone to breaking at that spot. Therefore, it is usually prudent to discard that rod and use a new one instead of rebending it (Fig. 21.8).

Construct design is a critical factor in fixation failure. The surgeon must have a clear understanding of the capabilities and limitations of the hardware construct selected.

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**Fig. 21.8a–g**

- **a** Preoperative lateral radiograph of a patient with rheumatoid arthritis. This patient had syncopal episodes when forward-flexing her neck, and it was found that she had occipitocervical and subaxial subluxations with myelopathy.
- **b** A preoperative sagittal MRI demonstrating a high-intensity signal behind the cranial portion of the dens, consistent with subluxations seen on flexion-extension views. The patient had compression at the foramen magnum as well as in the subaxial spine.
- **c** We performed a circumferential operation and occipitocervical fusion.
- **d** Unfortunately, within 1 year following surgery, one of the rods broke. This was due to a pseudarthrosis and the fact that we had contoured the rod in one direction and back in the opposite direction, which caused notching of the titanium rod. Titanium rods should be bent in only one direction. If a rod has been overbent, it should be discarded.
Occipitocervical Malalignment

Malalignment of the occipitocervical junction can be debilitating to the patient and may require revision surgery. The best solution is to avoid this problem at the time of the initial surgery by taking meticulous measures to ensure proper positioning on the operating table. The patient may have been inadvertently placed into a flexed, extended, or rotated position. Abnormal flexion poses a challenge for respiration, as well as proper swallowing. Abnormal extension and rotation can cause associated problems with balance and vision. Matsunaga et al showed that an abnormal occipitoaxial angle, with normal being between 0 and 30° measured from the occiput to C2, may contribute to the development of postoperative kyphosis and subaxial subluxation in patients with rheumatoid arthritis. They also advised including abnormal “buckling” cervical vertebrae in the construct to avoid later subaxial subluxation. Extreme flexion, extension, or rotation may compromise the canal space and result in neurologic deficits. Once a satisfactory position is achieved, a lateral radiograph should be obtained prior to final fixation and arthrodesis.

Surgical Planning

Preoperative surgical planning is done with a CT scan and reformatted CT images. The external occipital protuberance thickness, as well as C2 pedicle orientation, is measured. The presence of a ponticulus is also noted. If there is any question of a previous vertebral artery injury, a CT angiogram is done to evaluate the integrity of the vertebral arteries.

We prefer to position the patient on a Jackson frame in a prone position, with the head suspended in midair using Gardner-Wells tongs. Twenty pounds of traction keeps the head in a proper position. This setup allows intraoperative manipulation of the head–neck angle without having to...
make the minute adjustments that Mayfield tongs require. Precise positioning of the head and neck can be achieved for the duration of surgery. If the patient is in a halo preoperatively, the halo attachment can be used in lieu of the tongs. Once the patient is properly positioned, a lateral radiograph is obtained, and the occipitocervical angle is compared with the preoperative angle showing the patient looking straight ahead. A lateral image is repeated with the final fixation in place to ensure that the alignment has not changed. If an occipitocervicotoracic fusion is performed, it is even more imperative that the angle of the head and neck be perfect, as the patient cannot make any compensatory adjustments for minor iatrogenic malalignments. We prefer to place the head in a few degrees of flexion so that the patient can still see the front of his or her body (Fig. 21.3k). Hair at the base of the head is shaved 5 to 10 cm above the palpable occipital protuberance.

Surgical Technique

Approach

A posterior midline incision is performed, likely through a previous incision line. The occiput, posterior ring of C1, and posterior elements of C2, including the spinous processes, vertebral arches, and lateral masses of the lower cervical spine levels to be included in the fusion, are exposed subperiosteally. At the level of C1, care is taken to avoid exposing more than 1.5 cm laterally from the midline to prevent injury to the vertebral artery. If transarticular screw fixation at C1–C2 is necessary, the isthmus of C2 is also exposed bilaterally.

Extension of the Fusion

In the revision setting, the extension of fusion should be kept as short as possible while including all pathological segments. The occipitocervical junction can be immobilized with an occiput-to-C2 fusion. If there is additional subaxial instability, the fusion may be extended to include these levels. Many current systems have polyaxial screws available for purchase into the lateral mass and/or pedicles in the subaxial spine.

Bone Grafting

Autograft is the gold standard for occipitocervical arthrodesis. When using autograft, we prefer either the iliac crest or rib. If the iliac crest is used, we harvest a large corticocancellous structural graft from the outer table. The piece must be large enough to bridge from the occiput down to C2. We harvest additional cancellous bone, which we place underneath this structural piece. For longer constructs into the subaxial spine, we prefer to harvest a rib. The patient’s back is prepped so that a rib below the scapula can be harvested. Either one long rib or two short ribs can be harvested to bridge from the occiput to the cervical spine. The natural curvature of the rib makes it ideal to fit in the concavity between the occiput and the cervical spine. We prefer not to use allograft alone for occipitocervical arthrodeses.

We typically use local autograft and BMPs in procedures. It should be noted that this is considered off-label use of BMP, and there are no peer-reviewed data to support its use in such cases. In cases of metastatic disease, no bone grafting is used, and the construct can be augmented with polymethyl methacrylate bone cement.

Postoperative Management

As mentioned above, most patients do not require rigid external immobilization postoperatively. However, if the surgeon is not confident about the fixation, consideration should be given to using a halo vest postoperatively. In patients with adequate fixation, we usually immobilize with a soft cervical collar postoperatively for 6 weeks. A recent study by Chin et al. demonstrated that the more rigid collars stabilize the chin so well that during eating, the jaw cannot drop down. Instead, the head moves backward when the mouth opens, so that motion is concentrated in the upper cervical spine. In contrast, a soft collar allows the chin to drop, and there is less motion across the occipitocervical joint. In cases of severe osteoporosis, a four-poster brace with a chin plate or Minerva orthosis may be used when the patient is not eating, and a soft collar can be used during meals.

Complications

Injury to the vertebral artery can be catastrophic and can occur while drilling or tapping a hole for screw placement. The risk is greatest during the placement of transarticular screws at C1–C2. If an injury does occur, it can be addressed by either direct repair or tamponading the bleeding, with the intended screw aligned in the original trajectory. Direct repair is technically more demanding and results in greater blood loss. Postoperatively, a vertebral angiogram should be obtained to help manage a possible dissection or false aneurysm and prevent a potential cerebrovascular embolism. It is also important to refrain from attempting to place a second screw on the contralateral side.

Injuries to the dura and neural elements are most common during placement of fixation, although they can occur with exposure and decompression procedures. Typically, they are due to cable, wire, and, screw insertion. Injuries while passing cables and wires can be avoided by using large burr holes or laminotomy at the side of insertion. The epidural space should be palpated to confirm the presence of a plane between the osseous structure and the dura. When passing a cable or wire, even tension should be applied to both ends to avoid redundant cable
pressing on the dura. If a durotomy is caused by drilling in the occiput, wax followed by the intended screw should be placed. At other sites, if a direct dura repair is not feasible, DuraGen and DuraSeal may be used.

**Conclusion**

Revision surgery of the occipitocervical junction and the upper cervical spine can be technically challenging. With the development of spinal instrumentation, surgeons are faced with more options in the primary and revision settings. It is crucial to recognize the potential challenges of occipitocervical arthrodesis and to manage complications and the need for revision surgery.

**References**

With the advancement of segmental atlantoaxial fixation techniques, the indications for occipitocervical fixation have been reduced. However, occipitocervical fixation is still a commonplace operation. In this chapter, we discuss our strategy and modifications of the conventional modes of occipitocervical fixation.

Indications for Occipitocervical Fixation

The occipitoaxial joint is among the strongest joints of the body. The strength of the ligaments and the orientation of the joint surface provide remarkable stability and mobility to the region. We have identified that, in most cases of craniovertebral junction region–related instability, it is the atlantoaxial joint that is unstable whereas occipitoaxial instability is infrequent or rare. In cases with trauma and in congenital instability of the region, atlantoaxial instability is the principle issue in question. In cases of craniovertebral instability related to rheumatoid arthritis, the issue of occipitocervical instability is debated, whereas the presence of atlantoaxial dislocation or instability is a suboptimal operation and can be avoided. The technique of manipulation and distraction of the atlantoaxial joint can be the key to a variety of craniovertebral instability–related problems like reducible and irreducible atlantoaxial dislocation, basilar invagination, rheumatoid arthritis, and similar such problems. Craniovertebral realignment can be possible after such a joint manipulation and distraction. Although technically relatively difficult, it is possible to perform direct screw implantation in the facet of atlas, even in the presence of assimilation of the atlas. Accordingly, it is crucial for the surgeon to clearly identify the presence or otherwise of occipitoaxial instability prior to resorting to occipitocervical fixation. The improvements in understanding the anatomy of the craniovertebral junction and advancements in the techniques of atlantoaxial fixation have limited the indications of occipitocervical fixation.

Direct Atlantoaxial Fixation in the Presence of an Occipitalized or Assimilated Atlas

An occipitalized or assimilated atlas is relatively common. An occipitalized atlas is usually associated with fusion anomalies of adjoining bones in the region and with basilar invagination. We had earlier analyzed 190 cases of basilar invagination treated between 1987 and 1997 and isolated 153 (77%) having occipitalization of the atlas. Occipitalization is frequently associated with maldevelopment of the occipital bone, reduced length of the clivus and platybasia, occipital condylar and adjoining bone hypoplasia, and complete or incomplete fusion of the occipitoaxial joint. Fusion of the C2–C3 spinal elements and a range of Klippel-Feil spinal abnormalities are frequently associated. Occipitalization of the atlas is usually associated with basilar invagination and compression of the cervicomedullary cord by the odontoid process. Transoral surgery followed by posterior fixation is the generally adopted protocol for such an anomaly. Several methods have been discussed. These include use of the occipital squama for wire or screw fixation of the occipital end of the implant (plate, rods, or metal loops). Jain et al. described the technique of drilling the occipital bone close to the foramen magnum and formation of an artificial arch of the atlas, which is subsequently used for atlantoaxial fixation. Although occipitocervical fixations have been performed for more than 50 years using various types of instrumentation, and each type has its own advocates, it is fair to say that an ideal, universally applicable, and acceptable technique has not evolved. Crockard et al. after reviewing the literature, remarked that occipitocervical fixation is always difficult.

Although all authors do not uniformly agree, the craniovertebral region in the presence of an occipitalized atlas has been observed by us to be potentially unstable. Complete or incomplete occipitalization of the atlas is frequently seen in cases of basilar invagination. Mobile and reducible atlantoaxial dislocation in the presence of an occipitalized atlas is not common and has been reported by us earlier. In a more recent publication, we discussed that fusions in the craniovertebral junction are more frequently above (assimilation of the atlas) and below (C2–C3 fusion) the site of neural compression by the odontoid process. The bone fusions may not be due...
to an embryonic dysgenesis but are a result of chronically reduced neck size. Although direct screw implantation into the lateral masses of the atlas in the presence of occipitalization of the atlas is a relatively difficult surgical procedure, we found that the screw purchase and stability achieved were much stronger than screw implantation or wire fixation in the occipital squama, which is a relatively thin shell of bone. Direct application of screws to the atlas and axis, thus using the firm purchase in their thick and large corticocancellous lateral mass, provided a biomechanically firm fixation of the region. The drilling of the articular cartilage of the atlantoaxial joint and placement of the bone graft provided an opportunity for joint distraction, reduced the dislocation and basilar invagination, and helped in the ultimate bone fusion of the joint. The relatively strong and stubby spinous process of the axis facilitated screw implantation. Screws that were implanted in the base of the spinous process or spinolaminar junction and those that extended into the substance of the lamina were the strongest in their purchase. Preoperative computed tomography (CT) scan and intraoperative navigation assisted in the identification of the thickness of the spinous process and in determining the best site and direction of screw implantation. The selection of the site for screw implantation varied and was primarily based on the shape and thickness of the components of the spinous process. The spinous process was first denuded of all its ligamentous attachments and periosteum. The strongest part of the spinous process was at its base at the spinolaminar junction. The safety of the trajectory of the screw implantation was verified with intraoperative navigation and direct physical observation. The screw diameter ranged from 2.5 to 2.7 mm, and the screw length ranged from 8 to 12 mm. In cases where the screw will subsequently traverse into the lamina of the axis, the longer screw length ranging from 20 to 26 mm can be used. In some cases, the spinous process can be so thick that two or more screws can be implanted into its substance.

Screw Implantation in the Spinous Process and Spinolaminar Region of the Axis

The feasibility of direct screw implantation in the spinous process or in the spinolaminar region for occipitocervical fixation was first described by Goel and Kulkarni in 2004. The relatively strong and stubby spinous process of the axis facilitated screw implantation. Screws that were implanted in the base of the spinous process or spinolaminar junction and those that extended into the substance of the lamina were the strongest in their purchase. Preoperative computed tomography (CT) scan and intraoperative navigation assisted in the identification of the thickness of the spinous process and in determining the best site and direction of screw implantation. The selection of the site for screw implantation varied and was primarily based on the shape and thickness of the components of the spinous process. The spinous process was first denuded of all its ligamentous attachments and periosteum. The strongest part of the spinous process was at its base at the spinolaminar junction. The safety of the trajectory of the screw implantation was verified with intraoperative navigation and direct physical observation. The screw diameter ranged from 2.5 to 2.7 mm, and the screw length ranged from 8 to 12 mm. In cases where the screw will subsequently traverse into the lamina of the axis, the longer screw length ranging from 20 to 26 mm can be used. In some cases, the spinous process can be so thick that two or more screws can be implanted into its substance.

Text continues on page 225

Fig. 22.1a–c
a Lateral radiograph with the head in flexion showing an occipitalized atlas and atlantoaxial dislocation.
b Lateral radiograph with the head in extension showing reduction of the dislocation.
c Postoperative radiograph with the head in flexion showing atlantoaxial fixation using the plate and screw method.
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Fig. 22.2a–e
a Lateral radiograph with the head in extension showing an occipitalized atlas.
b Lateral radiograph with the head in flexion showing atlantoaxial dislocation.
c T2-weighted magnetic resonance imaging (MRI) showing severe cord compression.
d Postoperative radiograph showing atlantoaxial fixation using plate and screws.
e Postoperative axial computed tomography (CT) scan showing the direction of screw insertion into C2.
Screw Implantation in the Spinous Process and Spinolaminar Region of the Axis

Fig. 22.3a–c
a Lateral radiograph with the head in flexion showing assimilation of the atlas and atlantoaxial dislocation.
b T1-weighted MRI showing a Chiari 1 malformation and syringomyelia.
c Postoperative radiograph showing plate and screw fixation.

Fig. 22.4 Lateral radiograph showing craniovertebral fixation. The fixation on one side is with a lateral mass plate and screw. On the contralateral side of the fixation, a screw has been inserted into the spinous process of the axis to stabilize the cervical end of the plate, and wire has been used to stabilize the occipital end of the plate.
Fig. 22.5a–c
a  Lateral radiograph showing basilar invagination, atlantoaxial dislocation, and assimilation of the atlas.
b  CT scan showing the craniovertebral anomaly.
c  Lateral radiograph showing craniovertebral fixation following transoral surgery. The occipitocervical fixation has been done on one side by stabilization of the cervical end of the plate with a C2 pars screw and the occipital end with screws. The fixation is done on the contralateral side by fixation of the cervical end of the plate by C2-spinolaminar screw insertion and the occipital end by wires.

Fig. 22.6a–e
a  CT scan showing assimilation of the atlas and basilar invagination.
b  MRI showing severe cord compression.
Foramen Magnotomy Following Foramen Magnum Decompression

Foramen magnotomy was described by Goel and Achawal in 1995. The occipital bone flap was reversed, denuded of pericranium, and roughened with the help of a drill. The upper end of the reversed bone flap was sutured to the bone defect. The convex outer surface of the occipital bone with the edge of the foramen magnum was now concaved outward. Bone graft taken from the iliac crest was placed into the articular cavity of the facet joints, and longer pieces were placed in the midline over the foramen magnotomy flap. (Fig. 22.7)

Foramen magnotomy, as described, uses the curvature of the occipital bone to provide adequate and safe decompression of the craniovertebral region, while preserving the bone in the midline. The superior edge of the magnotomy bone flap is a tight fit and can be sutured in place, while the rest of the flap swings away from the neural structures with little danger of recurrent compression by the bone flap. The large bone surface available is used for the placement of onlay bone graft, which could be critically important for the bony fusion and the ultimate stability of the region. Such an expansive foramen magnotomy procedure can be performed in the case of Chiari malformation without atlantoaxial dislocation, as the procedure provides adequate decompression by increasing the diameter and volume of the foramen magnum. However, this may not be important or even necessary in such cases. Occasionally, instability of the cervical spine develops following posterior fossa decompression in cases of Chiari malformation. A reversed foramen magnotomy operation can be considered if instability of the region is feared.

Posterior fossa craniectomy with or without upper cervical laminectomy (foramen magnum decompression) is the most frequently performed surgical procedure for Chiari malformation. In several reviews, Goel et al. argued that opening the dura is not necessary and can be counterproductive. They suggested that dura is an expansile structure and is not a compressive factor by itself.

Fixation of the Occipital End of a Plate or Rod

The occipital end of a multiholed plate or rod can be done with the help of wires or screws. Fixation of the occipital end with screws was first described by Goel and Laheri in 1987 (Fig. 22.8). The site of screw implantation can be assessed on the basis of preoperative CT scan images. The thinnest part of the occipital bone is the occipital squama over the cerebellar hemispheric convexity. The bone is
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thicker near the midline, particularly in the region of the occipital crest. The bone is also thicker in the region closer to the inferior and superior nuchal lines, in the region of the transverse sinus, and laterally closer to the mastoid process. Screw insertion can be done in an oblique fashion so that a relatively longer purchase of the screw is available. Insertion of a thicker (3.2–3.4 mm) screw with a blunt tip is usually done. Insertion of 2 to 4 mm of the screw inside the inner surface of the occipital bone is safe.

Fixation of the occipital end of the implant with wires is done by drilling two small holes by the side of the occipital end of the implant. The wire is then maneuvered through the holes (Figs. 22.9 and 22.10). Serrations within the implant or holes in a plate can be used for anchoring the implant.

Anterior Transoral Occipitocervical Fixation

Transoral metal fixation from the clivus to the cervical vertebral body affecting occipitocervical fixation was first described by Goel and Karapurkar in 199516,17 (Figs. 22.11, 12, and 13). Several implants have been described recently for atlantoaxial fixation. However, with the current available methods of posterior occipitocervical and atlantoaxial fixation, transoral insertion of metal implants is less popular.

Figure 22.11 shows occipitocervical fixation of the metal plate done from the mouth following transoral odontoidecotomy and decompression. The superior end of

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Fig. 22.7a,b
a. Illustration showing the types of occipitoaxial plate and screw fixation. On the right side, occipitoaxial is shown and occipitoaxial plate and screw fixation is shown on the left. The dashed line shows the site of the bone flap for a foramen magnotomy.

b. Illustration to show a foramen magnotomy. The bone flap is reversed. The superior edge of the reversed flap is stitched to the occipital bone while the inferior edge swings away from the neural structures.

Fig. 22.8 Line drawing showing occipitocervical fixation. The occipital end is fixed with screws. The screws in the cervical end are placed in the pars of the axis and in the facet of the atlas. On the right side, the screw is placed in the pars of the axis.
Fig. 22.9a, b
a T1-weighted MRI showing basilar invagination, atlantoaxial dislocation, and Chiari I malformation.
b Postoperative radiograph showing occipitocervical fixation. The fixation of the cervical end is done by placing a screw in the C2 pars. The occipital end of the plate is fixed with wires.

Fig. 22.10a, b
a Lateral radiograph showing basilar invagination and atlantoaxial dislocation. Assimilation of the atlas can be seen.
b Following transoral decompression, posterior fixation was done. Metal ring was appropriately molded. The occipital end and the axial ends of the ring were fixed with wire.

Fig. 22.11a–c
a Lateral radiograph showing marked basilar invagination.
b MRI showing severe basilar invagination.
Fig. 22.11c
Lateral radiograph showing transoral occipitocervical fixation following odontoidectomy. The upper end of the plate is fixed to the clivus with two screws, and the cervical end is fixed to the C3 vertebra.

Fig. 22.12a, b
a T2-weighted MRI showing severe basilar invagination.
b Transoral decompression followed by iliac crest bone fixation of the region can be seen.

Fig. 22.13b–d
a CT scan showing basilar invagination.
b T2-weighted MRI showing basilar invagination and cord compression. Evidence of posterior foramen magnum decompression can be seen.
the plate was fixed to the clivus with screws, and the inferior end of the plate was fixed to the body of the second or third cervical vertebra. The plate was appropriately molded to suit the anterior craniocervical curvatures. The potential of infection of any implant placed from the mouth is high; whenever possible, such an endeavor should be avoided if an alternative solution is available. On the positive side, high vascularity of the nasopharyngeal wall has an excellent healing potential and can accept grafts and implants. The clivus can form a strong ground for fixation of the plate by screws. In this procedure, the length of the screws was measured on evaluation of preoperative CT scans.

**Conclusion**

For any kind of instability, segmental fixation is crucial. For atlantoaxial instability, atlantoaxial fixation should be done. In cases having primarily atlantoaxial instability, inclusion of the occipital bone and subaxial cervical spine in the fixation procedure can compromise the strength of the fixation procedure. Long fixations that include occipital bone and multiple cervical spine segments need to be evaluated critically and carefully. Stabilization of the most mobile part of the body and provision of opportunity for bone arthrodesis can be most effective when limited spinal segments are incorporated in the fixation process. Currently, our primary indications to perform occipitocervical fixation are narrowed to cases having extensive regional bone destruction by tumors and, infrequently, in cases with extensive tuberculosis of the region.

**References**


**Fig. 22.13c–d**

- Following transoral decompression, occipitocervical fixation was done using screw implantation in the clivus and in the C3 vertebral body.
- Open-mouth views showing the plate and screw fixation.
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Chiari malformations are named after the Austrian pathologist Hans Chiari, who published two seminal reports on hindbrain herniation in 1891 and 1896. Chiari provided detailed pathological descriptions from postmortem cases, with hindbrain descent through the foramen magnum associated with hydrocephalus. The current classification of Chiari malformations involves four distinct subtypes (Table 23.1).

The focus of this chapter will be on the most common Chiari malformation, type I. The definition of this anomaly has evolved since Chiari’s original description of tonsillar herniation occurring with hydrocephalus. Most cases now are diagnosed in adults without hydrocephalus. Unfortunately, the term Chiari I malformation currently applies to virtually any type of tonsillar herniation except for cases related to myelodysplasia (Chiari II) or cervical encephalocele (Chiari III). This practice does not emphasize the etiologic factors that produce the tonsillar herniation and can lead to inappropriate treatment. The current definition of the Chiari I malformation depends on a single criterion of tonsillar descent \( > 5 \) mm, based on a magnetic resonance imaging (MRI) diagnosis.

Although tonsillar herniation is the common anatomical marker, it is important to recognize that Chiari I malformations are heterogeneous conditions with different pathophysiological mechanisms, which can often cause overlapping symptoms. Tonsillar herniation may arise from congenital causes with small posterior cranial fossae (classic Chiari I malformations and craniosynostotic syndromes) and acquired causes with normal posterior cranial fossae (hydrocephalus, Paget disease, posterior fossa tumors, tethered cord, and spinal hypotension). Because of the varied manifestations for Chiari malformations, there have been differences in treatments and surgical outcomes. To appropriately treat these patients, each patient’s tonsillar descent must be evaluated in the setting of morphometric analysis of the posterior fossa and stability of the craniovertebral junction. Ultimately, successful management depends on appropriate patient selection, tailoring the surgical intervention to treat the underlying anatomical disorder, and complication avoidance.

Chiari malformations are the leading cause of syringomyelia (Fig. 23.1). The association between Chiari malformations and syringomyelia was first recognized by Russell and Donald in 1935 and Lichtenstein in 1943. The early surgical strategies for syringomyelia were directed at the syrinx cavity, and there was little attention given to any associated hindbrain anomalies. It was Gardner and Goodall in the 1950s who were credited with demonstrating that correction of the hindbrain hernia by suboccipital decompression could lead to improved surgical outcomes. Gardner’s investigations appropriately directed attention to the craniovertebral junction.

According to Gardner’s hydrodynamic theory, a syrinx develops as a result of obstruction of the outlets of the fourth ventricle, which causes a caudal cerebrospinal fluid (CSF) pulse wave, producing a “water hammer” effect that dilates the lumen of the central canal. As it became known, Gardner’s operation included a foramen magnum decompression with plugging of the obex to close the hypothetical communication between the syrinx and the fourth ventricle. His technique revealed significant clinical improvement in some patients, and his operation laid the basis for contemporary surgical management of syringomyelia occurring with Chiari malformations.

Williams and others noted that decompression alone was sufficient for good clinical results. His critical evaluation of the Gardner technique revealed that plugging the obex was an additional source of morbidity and occasional mortality. Williams developed an alternative explanation that obstructions of CSF, occurring at the level of the foramen magnum, produce a dissociation of pressure between the cranial and spinal CSF compartments. Williams theorized that the pressure gradient

<table>
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<td>Type I</td>
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between cranial and spinal compartments “sucks” fluid from the fourth ventricle into the central canal, as a consequence of a relatively lower CSF pressure caudal to the block. Valsalva maneuvers, such as sneezing, coughing, and straining, accentuate this pressure gradient phenomenon by producing changes in venous volume and pressure.

With the advent of MRI technology, the classical theories of Gardner and Williams have been challenged. Although MRI firmly established the diagnosis and the link between Chiari malformations as the leading cause of syringomyelia, there remains some question regarding the pathogenesis of these disorders. Unlike Gardner’s initial contention, it has become clear from analyses of pathological autopsy specimens that only a minority of syrinxes communicate directly with the fourth ventricle, and that most syrinxes are separated by the fourth ventricle by a long segment of syrinx-free spinal cord. Further advances in MR technology improved the temporal imaging resolution, particularly during the cardiac cycle. Oldfield et al. demonstrated that the tonsils behave like a piston moving during systole and diastole. During systole the tonsils are impacted into the foramen magnum, leading to a CSF pressure wave in the spinal subarachnoid space which drives CSF into the cord through Virchow-Robin spaces or the dorsal roots. Although some questions remain, Oldfield’s theory has gained wide acceptance.

Toward a better understanding of the pathogenesis, there were observations that patients with Chiari I malformations have a relatively small posterior cranial fossa. With a small cranial vault, the normal-sized brain matter is constricted, causing tonsillar descent through the foramen magnum. This tonsillar ectopia leads to obstruction.
of the CSF flow at the foramen magnum and a noncommunicating syringomyelia in 20% to 65% of cases. Further morphometric studies involving quantitative comparisons of the size of the posterior cranial fossa confirmed this theory.12,13 Goel et al. in 1998 described additional radiological morphometric parameters and discussed the clinical symptoms and therapeutic implication of the reduction in posterior cranial volume and its relationship with pathogenesis of initially Chiari I malformation and subsequently syringomyelia.13

By the early 1990s (100 years after Chiari’s initial description), there emerged multiple treatments for syringomyelia associated with Chiari malformations. There were proponents for a variety of different approaches, which included (1) foramen magnum decompression only, (2) foramen magnum decompression with plugging of the obex, (3) syrinx shunting only, and (4) simultaneous decompression and syrinx shunting. With the proliferation of different surgeries, there has been great difficulty in assessing outcomes. Operative strategies for treating Chiari I malformation and syringomyelia have yet to be fully standardized. The traditional approach is to perform a posterior fossa decompression comprising a suboccipital craniectomy, upper cervical laminectomy, and duraplasty. Unfortunately, specific details, such as the size of the craniectomy, the extent of the laminectomy, and the preferred intumescence of duraplasty (e.g., cadaveric dura, pericranium, bovine pericardium, or synthetic material), are rarely discussed in the literature. Also controversial are additional steps, such as lysis of adhesions, plugging of the obex, terminal ventriculostomy, drainage of the fourth ventricle, leaving the dura open, and resection or shrinkage of the cerebellar tonsils. No one procedure has been uniformly successful, and it is estimated that a significant improvement in preoperative symptoms and a reduction in syrinx size occurs in only 40% to 60% of patients.

In surveys sent to international pediatric neurosurgeons16 and American pediatric neurosurgeons,17 suboccipital decompression was considered the standard surgical procedure. The majority of respondents favored routine dural opening at surgery and closure with a pericranial or synthetic patch graft. This technique has been adopted by many surgeons, because it improves flow of CSF at the level of the foramen magnum.

Contemporary neurosurgical debate rests on whether or not to perform a duraplasty. The controversy arises from a higher associated morbidity with duraplasty and intradural manipulations. As a result, some surgeons advocate bony decompression only, as there appears to be a subset of patients who do respond to this intervention.13,17,18,20 In effect, the surgeon’s decision is often determined by balancing the risk of a complication against the risk of undertreatment, necessitating a return to the operating room.

Duraplasty and intradural manipulations have been associated with higher morbidity in certain series. When the arachnoid is opened, there is an increased risk of bleeding and adhesion formations, which can lead to arachnoiditis, pseudomeningoceles, hydrocephalus, and persistent syringomyelia. In turn, these conditions may cause persistence in symptoms or new posterior fossa syndrome complaints. The advantage of opening the dura is that it provides the necessary exposure to allow for internal decompression. The effect of chronic severe foramen magnum impaction by the cerebellar tonsils is the formation of arachnoidal adhesions, which may be the primary pathological focus. The adhesions may be quite pervasive, involving the brainstem, posteroinferior cerebellar artery, and spinal cord. Microlysis of the adhesions is an important part of the internal decompression. Additionally, we advocate tonsillar reduction if the obex area is closed and if there is no evidence of pulsatile flow of the CSF from the fourth ventricle. At the end of the operation, the tonsils are ideally positioned slightly above the level of the putative foramen magnum. Those surgeons who elect not to open the dura13,17 may not address the potential significant impact of arachnoidal scarring.

Most surgeons have moved away from various intradural techniques, including posterior fossa stenting, catheterization, and plugging of the obex, which have not been associated with improved outcomes.21,22 The one popular exception is tonsillar shrinkage or complete tonsillar amputation. Some have argued that neural tissue should be preserved whenever possible.23 Reduction of the cerebellar tonsils appears to be well tolerated. At the time of surgery, cystic changes are often apparent as a consequence of chronic compression and ischemia.24 There is evidence that the cerebellar tonsils have no neurological function, and bilateral tonsillectomy is not associated with neurological deficits.

Although the majority of surgeons who perform duraplasties close the dura, there have been some reports with good outcomes supporting leaving the dura opened25 or stitched laterally to the muscles.26 According to the authors, it is important to preserve the arachnoid plane. Postoperative complications were related to arachnoidal violations. Limonadi et al. reported that a dura-splitting decompression compared with duraplasty can result in reduced operative time, hospital stay, and cost with equivalent early outcome.27 The counterargument is that one return to the operating room for incomplete decompression would significantly tip the cost-effective analysis in favor of duraplasty.

Duraplasty material is another variable. Duraplasties include autologous and nonautologous graft materials. Autologous grafts may be harvested from the fascia lata, ligamentum nuchae, and pericranium. Nonautologous materials include, in decreasing order of use, bovine pericardium, cadaveric dura, and synthetics. These products may be favorable because they decrease operative time. We prefer autologous pericranium because of the decreased incidence of inflammatory reactions and CSF leaks.28,29

With all of these variables, perhaps the central question is what is necessary and sufficient in an operation to
achieve surgical goals. To help answer this question, we have adopted the use of color Doppler ultrasonography (CDU) to guide us intraoperatively. Many of the technical decisions are patient specific. The particular variables in our practice include the size of the craniectomy, the levels of the laminectomy, the degree of tonsillar reduction, and the size of the duraplasty. CDU is an important tool because it gives real-time feedback and intraoperative confirmation of the restoration of CSF flow before surgery is completed. Some surgeons have also reported that an intraoperative ultrasound may help in deciding whether or not to remove bone only or perform a craniectomy with duraplasty.

### Indications

In this era of MRI and other commonly available imaging technologies, Chiari malformations and syringomyelia are being diagnosed with increasing frequency. In a study of 2000 MRI scans, Chiari malformations comprised 0.9% of unexpected, asymptomatic brain abnormalities on imaging. Although the correct diagnosis can usually be established by an MRI scan of the cervical spine, the following workup is strongly recommended: (1) an MRI scan of the brain to rule out hydrocephalus and other causes of acquired tonsillar herniation (Fig. 23.2), (2) an MRI scan of the thoracolumbar spine to rule out spinal cord tethering (Fig. 23.3), (3) cine-MRI to assess CSF velocity/flow at the cervicomedullary junction (Fig. 23.4), and (4) a three-dimensional CT scan to reveal any bony variations (e.g., an incomplete bifid C1 lamina or occipital–C1 assimilation) (Fig. 23.5). The anatomical factors that need to be assessed are the level of cerebellar tonsillar descent, degree of cervicomedullary compression and foramen magnum impaction, presence of skeletal anomalies (basilar impression, platybasia, odontoid alignment, pannus formation, joint hypermobility, and craniocervical instability), and disturbance of CSF circulation (hydrocephalus or syringomyelia).

Patients develop symptoms related to Chiari malformations from two primary mechanisms: direct neural compression and disturbance of CSF flow. The most common symptom is the suboccipital headache that may radiate to the vertex, behind the eyes, or to the shoulders and neck. Cranial nerve signs may include impaired gag reflexes, facial sensory loss, and vocal cord paralysis. Ocular, otoneurological, and cerebellar disturbances are also varied and common. Pediatric patients may demonstrate different clinical manifestations. The youngest patients may present with poor Karnofsky scores related to failure to thrive, because of poor oral intake. Other pediatric patients may have vague behavioral problems before they are diagnosed. Some adolescent patients may present after having a workup for scoliosis, which is associated with syringomyelia. Finally, an incidental Chiari malformation may be found after a traumatic event. These patients are first given conservative treatment, which often involves pain management and clinical monitoring of the syrinx if present.

After conservative remedies have been exhausted, the three main indicators for surgical interventions are poor Karnofsky score (≤ 70), new-onset or progression of syringomyelia (particularly syringes occupying > 75% transverse diameter), and severe neurological deficit.

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**Fig. 23.2 MRI of the brain revealing hydrocephalus with secondary tonsillar herniation and syringomyelia.**
Fig. 23.3a, b MRI with (a) T1-weighted sagittal view of the lumbar spine in a 35-year-old man with spinal dysraphism and conus extending to S1 and (b) T2-weighted axial view of the thoracic spine revealing nonexpansile syringomyelia.

Fig. 23.4 Cine-MRI of the brain revealing significant obstruction of cerebrospinal fluid (CSF) flow at the level of the foramen magnum.
Surgical Goals

The goals of Chiari surgery are fourfold:

1. Adequate decompression of the cervicomedullary junction, which includes neural structures and relief of CSF block
2. Creation of normal-sized cisterna magna and retrocerebellar spaces
3. Establishment of optimal CSF flow between cranial and spinal compartments
4. Stabilization of the craniocervical junction if excessive joint hypermobility exists

Approximately one third of patients with Chiari I malformations have anterior encroachment of the foramen magnum by a retroflexed odontoid process or basilar invagination. Thus, before formulating a surgical plan for such patients, a decision needs to be made whether posterior decompression, anterior decompression, or both is the optimal treatment. The presence of clinically significant anterior compression of the cervicomedullary junction by a retroflexed or invaginated odontoid is a contraindication for primary decompression of the posterior fossa in some cases (Fig. 23.6). Anterior compressions are divided into two groups by cervical traction. Irreducible compressions are managed by transoral odontoidectomy, then a craniovertebral fusion at the time of posterior decompression. More recently, Goel and Sharma described a technique of distraction of the facets of the atlas and axis for reducing the “irreducible” compressions. If technical subtleties of the technique are adequately learned, transoral surgery can be avoided entirely. Reducible compressions are managed by a posterior decompression and fusion in one or two stages.

Recently, we recognized an association between Chiari I malformations and hereditary disorders of connective tissue. Those patients with both disorders are predisposed to developing symptoms attributable to craniocervical instability, which we have termed functional cranial settling. If the functional cranial settling is reducible by a trial of invasive cervical traction, these patients will respond well to posterior decompression and fusion.

The indications for reoperation on patients with failed Chiari surgery are the same as those for patients undergoing primary operations. In patients undergoing reoperation for failed Chiari surgery, there is radiographic evidence of one or more of the following findings: underdecompression of the posterior fossa, overdecompression of the posterior fossa with...
Cerebellar ptosis, pseudomeningocele formation, surgical meningocele, cranial settling, basilar invagination, and hydrocephalus.

**Description of Surgical Technique**

At the Chiari Institute (Great Neck, New York), the surgical management of Chiari malformation involves (1) a craniectomy of moderate size that increases the volume of the underdeveloped posterior fossa and (2) an expansile duraplasty employing autologous pericranium. A cervical laminectomy is performed to expose the distal tips of the cerebellar tonsils as demonstrated by intraoperative ultrasound. If the cerebellar tonsils are impacted or deeply herniated through the foramen magnum, it is common practice to lyse adhesions and shrink the tonsillar tips with bipolar coagulation. These and other surgical decisions are guided intraoperatively by the use of B-scan real-time ultrasound, somatosensory evoked potentials (SEPs), and color Doppler imaging that has been modified for measurement of CSF flow and velocity. What follows is a description of a tailored operative technique for Chiari I malformation using intraoperative CDU.

**External Decompression**

The patient is placed prone on chest rolls. The head is fixed in a Mayfield head holder and gently flexed under SEP monitoring. Transcutaneous cardiac pacemaker electrode pads are placed to protect against intraoperative cardiac arrhythmias that may occur during intradural manipulations. The posterior occipital and cervical spine are exposed through a midline incision that extends approximately two fingerbreadths above the inion to the third cervical spine. The supraocciput exposure is...
necessary for harvesting of a pericranial graft for dura-plasty (Fig. 23.7).

A suboccipital craniectomy is performed using a combination of high-speed drilling, rongeurs, and Kerrison punches. The size of the bony decompression is guided by CDU. After a small suboccipital opening is made, the atlanto-occipital membrane is excised to facilitate imaging. Thereafter, the craniectomy is enlarged in a step-wise manner to expose the dura overlying the area of cerebellar impaction, as demarcated by compressed or obliterated subarachnoid spaces. The superior limit of the craniectomy is never above the prepyramidal fissure. Laterally, the craniectomy is widened to create a nearly circular bony opening, which is generally as wide as the spinal canal. An excessive amount of bony removal may lead to cerebellar ptosis, requiring additional operative intervention.

After completion of the craniectomy, a decision is made regarding whether or not to perform a laminectomy. The determining factors are the extent of tonsillar herniation and the required length of the dural incision. CDU is used to establish the true position of the tonsillar tips, which is typically 3 to 6 mm lower than predicted by MRI. In patients with intermediate herniations (12–15 mm), a C1 laminectomy is routinely performed. Herniations > 15 mm generally require a standard C1 laminectomy and a partial or complete C2 laminectomy. Bony decompression is not limited at the expense of exposure for dural decompression. Occasionally, the relationship of the cerebellar tonsils to bony landmarks is altered by anomalies such as assimilation of the atlas (Fig. 23.5b, c).

CDU Imaging before Opening of the Dura

Before the dura is opened, CDU is used for anatomical orientation and to establish baseline measurements of the CSF flow (Fig. 23.8a, d). Structures that are routinely insonated include the cerebellar tonsils, the uvula, the medulla, the upper cervical spinal cord, both vertebral arteries, both posteroinferior cerebellar arteries, the marginal sinuses, the fourth ventricle and its choroid plexus, the parenchymal arteries and veins, and bridging vessels suspended by the arachnoid. The identification of aberrant vascular anatomy, asymmetrical herniations, and neural displacements helps reduce the risk of surgical error. In patients undergoing reoperation for failed Chiari surgery, information concerning the location and extent of meningocerebral scarring is invaluable in planning dissection strategies.

CSF circulation at the cervicomedullary junction is assessed immediately before opening the dura (Fig. 23.8b, e). The following measurements are made and stored: (1) the size and volume of the cisterna magna, (2) the size and volume of the dorsal cervical theca between C1 and the

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**Fig. 23.7a–d** Summary of the surgical approach for posterior fossa decompression of a Chiari malformation. (Drawings with permission from MediVisuals, Inc.)

- **a** Location of the posterior occipitocervical incision.
- **b** Completion of a craniotomy, upper cervical laminectomy, and dural opening with initiation of arachnoid dissection.
tonsillar tips, (3) CSF velocity/flow in the cisterna magna, (4) CSF velocity/flow in the dorsal cervical theca, and (5) CSF velocity/flow in the premedullary cisterns. Cisternal magna volumes $< 0.5 \text{ cm}^3$ and CSF flow velocities in the range of 0 to 0.8 cm/second anterior and posterior to the cervicomedullary junction are typical baseline findings.

## Dural Opening

The dura is opened with a Y-shaped incision across the marginal sinuses unless CDU imaging suggests a safer line of entry. The arachnoid is left intact, and imaging is repeated in appropriate cases to analyze whether duraplasty alone might be sufficient treatment. Opening the dura invariably results in some re-expansion of the cisterna magna, but a significant improvement in CSF flow is rarely observed. A simple duraplasty without additional steps is performed in patients who met the following criteria: cisterna magna volume of at least 4 cm$^3$, CSF velocity/flow of at least 2 cm/second, and CSF tracings demonstrating bidirectional movement with vascular and respiratory variations.

## Internal Decompression

The arachnoid is opened, and the CSF is allowed to drain spontaneously. We use magnification to visualize the area. The arachnoid is resected widely, and adhesions to the cerebellar tonsils, posteroinferior cerebellar arteries, and spinal cord are coagulated and divided (Fig. 23.7b). The tonsils are mobilized, and the posteroinferior cerebellar arteries are protected with moist cotton patts. On the basis of the direct inspection, a decision is made whether or not to shrink the cerebellar tonsils. The tonsils are not shrunken if the obex area is open and a pulsatile flow of CSF could be observed to exit from the fourth ventricle into the dorsal cervical theca. In most cases, the tonsils are reduced with low-voltage, bipolar coagulation until the tonsillar tips are positioned at or slightly above the putative level of the foramen magnum (Fig. 23.7c).

## Expansile Duraplasty

The dura is closed with autogenous pericranium, which had been previously harvested rostral to the craniectomy site, along the occiput. A graft to $\sim 5 \text{ cm}$ in length and 2.5 cm in width is usually sufficient to produce a competent retrocerebellar space. The graft is anchored to the poles of the incision and sewn in place with continuous locking sutures of 5-0 Gore-Tex (Fig. 23.7d). Before the last suture is tied down, the retrocerebellar space is inflated with 30 to 40 mL of sterile saline to expand the graft and to eliminate intradural air bubbles, which can degrade CDU images. Valsalva maneuvers are performed to ensure a watertight closure. CSF leaks are corrected by oversewing the suture line.
Fig. 23.8a–f  Color Doppler ultrasound (CDU) images of the cervicomedullary junction in a patient with Chiari I malformation undergoing posterior fossa decompression. (a, b) Sagittal and (c–f) axial views of 10 and 8 mm asymmetric (left to right) tonsillar herniation. CDU images (b) after external decompression and (e) before opening the dura reveal minimal CSF flow caudal to the cerebellar tonsils. (c, f) After internal decompression with lysis of the arachnoid, shrinkage of the tonsils, and duraplasty, CDU images reveal improved CSF flow, with peak CSF velocity of 3 to 4 cm/second. There are optimal CSF flow characteristics with bidirectional movement, arterial pulsations, and respiratory and venous variations.
CDU Imaging after Closing the Dura

After dural closure, CDU imaging is repeated to assess the goals of surgery. Optimal CSF flow is found to have the following characteristics: a peak velocity of 3 to 5 cm/second; bidirectional movements; and a waveform exhibiting arterial, venous, and respiratory variations. Figure 23.8 shows typical findings before and after lysis of the arachnoid, shrinkage of the tonsils, and duraplasty. Postoperative neuroimaging demonstrates a normal-sized cisterna magna and unrestricted CSF flow anteriorly and posteriorly through the foramen magnum in most patients. Overly generous duraplasties and iatrogenic meningoceles are associated with suboptimal CSF flow velocities (<1 cm/s). The problem could usually be corrected by tightening the graft with reinforcing suture or by adding a restrictive graft. Excessive CSF flow velocities (>8 cm/s) are most often encountered during reoperations in which the dura is densely scarred and thickened. Such observations are consistent with principles of fluid mechanics governing rates of flow through spaces of varying compliance and cross-sectional area.

Wound Closure

The paraspinal muscles are brought together in layers. Rarely, in patients at risk for a CSF leak, a lumbar drain is inserted after surgery.

Our Experience

Posterior fossa decompression with duraplasty reliably treats the syringomyelia associated with Chiari I malformation (Fig. 23.9). The syrinx decreases within 3 to 6 months, as observed by others. Symptomatic improvement is variable. Headache, strength, and spasticity improve more consistently compared with sensory changes and cranial nerve dysfunction. Table 23.2 summarizes our experience with the aforementioned technique at the Chiari Institute. Revision surgeries were referred from outside hospitals. When there was extensive leptomeningeal scarring, from either infection or hemorrhage, persistent symptomatic syringomyelia may occur, requiring direct shunting. Of the 263 cases of syringomyelia, we had to perform syrinx shunting in only 3 cases. The unusually low incidence of CSF leak in our experience (0.3%) was felt to be a direct function of the surgical technique used to repair thin and ectatic dura and for sealing duraplasty stitch holes.

Chiari Revision Surgery

In balancing risks versus benefits for any procedure, it is often useful to review those complications that may lead to additional surgery. There are several pathophysiological mechanisms for development of problems after surgery for Chiari malformations. First, there may be complications directly related to the index procedure, which include transcutaneous CSF leaks, pseudomeningoceles, defective duraplasties, meningoceles, cerebellar prolapse, infection, extensive adhesions, cranial settling, and cranio cervical instability. Second, there may have been a failure to achieve the original goals (underdecompression with persistent tonsillar herniation, persistent impaction of the CSF spaces and neuroanatomical structures at the foramen magnum, persistent and enlarging syringomyelia cavities, etc.).
Third, there may have been previously unrecognized pathology, not treated at the time of decompression (e.g., hydrocephalus, pseudotumor cerebri, basilar invagination, or cranial settling). These complications provide an impetus for further understanding of this complex disorder.

### Functional Cranial Settling

Recently, we described a new syndrome of occipitoatlantoaxial hypermobility, cranial settling, and Chiari I malformation in patients with hereditary disorders of connective tissue (HDCT). Not only does this functional cranial settling occur with patients with HDCT, but it is also present in some patients who have undergone previous surgery or suffered a traumatic event, such as a cervical whiplash injury. Patients in whom there is a clinical suspicion of functional cranial settling undergo a trial of invasive cervical traction. Invasive cervical traction allows testing of the reducibility of the cranial settling as well as the patient’s symptoms. Cranial tongs are applied in the operating room after propofol sedation and injection of a local anesthetic. After the patient has awakened fully, cervical traction is performed with the patient sitting in an 80° Fowler position. Using graduated weights, the head is extracted upon the neck under fluoroscopic guidance, and symptoms and neurological findings obtained immediately before testing are assessed. Once an optimum level of relief is achieved, fluoroscopic images are obtained to compare morphometric measurements of the osseous structures at the craniovertebral junction in the supine, upright (sitting), and extraction positions. The invasive cervical traction (ICT) test is defined as positive if optimal extraction produces an 80% or greater relief of baseline symptoms. Patients with a positive ICT undergo an occipital cervical

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**Table 23.2** Surgical outcome after tailored posterior fossa decompression using intraoperative color Doppler ultrasonography at the Chiari Institute, 2002–2007

<table>
<thead>
<tr>
<th>Variable</th>
<th>Chiari I Malformation</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of patients</td>
<td>714</td>
</tr>
<tr>
<td>Male/Female</td>
<td>179/525</td>
</tr>
<tr>
<td>Mean age (years)</td>
<td>32.5</td>
</tr>
<tr>
<td>Syringomyelia*</td>
<td>263 (37%)</td>
</tr>
<tr>
<td>Cervicothoracic syrinx</td>
<td>167</td>
</tr>
<tr>
<td>Holochord syrinx</td>
<td>96</td>
</tr>
<tr>
<td>Primary decompression</td>
<td>372 (52%)</td>
</tr>
<tr>
<td>Revision of failed</td>
<td>342 (48%)</td>
</tr>
<tr>
<td>Chiari surgery</td>
<td></td>
</tr>
<tr>
<td>Postoperative decrease</td>
<td>260 of 263</td>
</tr>
<tr>
<td>in size of syrinx</td>
<td></td>
</tr>
<tr>
<td>Failures requiring</td>
<td>3</td>
</tr>
<tr>
<td>syringopleural shunting</td>
<td></td>
</tr>
</tbody>
</table>

*Excludes distal thoracic terminal syrinxes below T5.

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**Fig. 23.10a, b** (a) Intraoperative and (b) lateral plain radiographs of a patient who had undergone staged posterior fossa decompression with duraplasty and occipital cervical fusion.
fusion (Fig. 23.10), using the same position that obtained the idealized position during the traction test.

### Persistent Syringomyelia

Shunting can be performed for treatment of persistent syringomyelia when the standard operations, such as posterior fossa decompression for Chiari malformations, fail to achieve symptomatic relief. Syringostomy has a simple and immediate appeal for direct treatment of spinal cavitation; however, syrinx shunts often fail, have poor long-term results, and have high initial complication rates. Furthermore, insertion of the shunt invariably results in some degree of spinal cord injury. There are significant technical variations within syrinx shunt management, including (1) placement of the syringostomy (midline vs. dorsal root entry zone), (2) location of the distal catheter (subarachnoid vs. pleural vs. peritoneal space), and (3) selection of the catheter (soft vs. rigid vs. siliconized). These drainage procedures are not an effective first-line solution for syringomyelia caused by hindbrain herniation. Instead, syrinx drainage should be reserved as a salvage procedure if the initial foramen magnum decompression fails.

### Conclusion

The diagnosis of syringomyelia has undergone a dramatic transformation with the advent of MRI. It is now understood that syrinx formation in Chiari malformations depends on obstruction of the subarachnoid pathways at the level of the foramen magnum. Treatment strategies have evolved with improved radiographic and clinical outcomes. In our experience, the most effective procedure with minimal complications has been a tailored osseous decompression of the cranio cervical junction, duraplasty employing autologous pericranium, and additional intradural steps (microlysis or arachnoidal adhesions, tonsillar shrinkage) as determined by intraoperative color Doppler ultrasonography.

### References


38. Wetjen NM, Heiss JD, Oldfield EH. Time course of syringomyelia resolution following decompression of Chiari malformation type I. J Neurosurg Pediatr 2008;1:118–123

History

Chiari Malformations

Hans Chiari was a professor of pathology at The University of Prague, Czechoslovakia, and later at the University of Strasbourg. In 1891, his initial work on what would become known as Chiari malformation was published, and 5 years later, he published a classification system that is still in use today.1 In the 1896 publication, he mentioned Cleland and Arnold, who made incidental observations compared with Chiari’s thorough study of the malformation. Arnold’s name was added to the type II malformation to coin the term Arnold-Chiari malformation by Schwalbe and Gredig, in 1907, writing from Arnold’s laboratory in Heidelberg. Unfortunately, this has led to the loose application of the term Arnold-Chiari malformation to both type I and II conditions, and subsequently much confusion. The term Chiari II malformation is clear and the preferred terminology.

Syringomyelia

The original Greek word from which syringomyelia is derived means tubular cavitation of the spinal cord. In 1546, Estienne described an abnormal, fluid-filled space within the spinal cord. It was later described in continuity with the fourth ventricle and labeled syringomyelia by Olivier d’Angers in 1824.2 Abbe and Coley were the first to decompress a syringomyelic cavity, in 1892. But it was only in the 1950s that Gardner postulated the hydrodynamic theory and craniocervical decompression as a surgical method of treating syringomyelia when associated with craniocervical abnormalities.3

Definitions and Classification

Chiari Malformations

Chiari’s classification of these craniocervical malformations is based on the degree of hindbrain herniation and location (Table 24.1). In Chiari I malformation, there is cerebellar tonsillar herniation through the foramen magnum. There may be associated syringomyelia. Other abnormalities that could be present include an elongated aqueduct, a stretched fourth ventricle, and a kinked lower medulla. The upper cervical nerve roots usually project inferiorly, and the posterior fossa is shallow.

Chiari II malformation has many associated congenital abnormalities of the craniospinal axis. It is typically associated with meningomyelocele and hydrocephalus. Apart from the cerebellar tonsils, the cerebellar vermis, fourth ventricle, and medulla herniate through the foramen magnum. In more severe deformities, the pons is also displaced below the foramen magnum. The upper cervical roots project superiorly, the pons is elongated, and the fourth ventricle is slitlike. Usually, the tentorium cerebelli is low lying and hypoplastic, with the superior cerebellum

Table 24.1 Classification of Chiari malformations

<table>
<thead>
<tr>
<th>Herniation</th>
<th>Associated Anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>Foramen magnum tonsils only</td>
</tr>
<tr>
<td>Type II (Arnold-Chiari)</td>
<td>Foramen magnum tonsils, vermis, fourth ventricle, medulla, and pons</td>
</tr>
<tr>
<td>Type III</td>
<td>Meningocele High cervical, suboccipital</td>
</tr>
<tr>
<td>Type IV</td>
<td>None Same as type I</td>
</tr>
<tr>
<td>Acquired Chiari</td>
<td></td>
</tr>
</tbody>
</table>

displaced upward through a wide incisura. Other dural abnormalities are hypoplasia and fenestrations in or partial absence of the falx cerebelli, with tight apposition between the cerebral hemispheres. Abnormalities of the skull include lacunae of the skull, erosion of the postero-medial aspect of the petrous pyramid (petrous scalloping), and enlargement of the foramen magnum.

To help differentiate Chiari I from Chiari II malformations, it is generally agreed that cerebellar ectasia in the presence of a neural tube defect, regardless of associated abnormalities or age of onset, constitutes Chiari II malformation.

In Chiari III malformation, there is herniation of the cerebellum and brainstem into a high cervical or sub-occipital meningocele. In Chiari IV malformation, there is cerebellar hypoplasia without cerebellar herniation. These last two categories are rare with limited neurosurgical relevance and will not be discussed further.

The iatrogenic, or acquired Chiari, malformation was not a recognized entity at the time of Chiari’s original classification. It has gained clinical significance and come into the literature as a result of neurosurgical interventions. Cerebellar herniation occurs through the foramen magnum (usually normal in size) due to a change in cerebrospinal fluid (CSF) dynamics from posterior fossa/upper cervical spinal cord surgery or a lumboperitoneal shunt.

### Syringomyelia

A cyst involving the central canal and therefore lined with ependyma is known as hydromyelia, whereas a cyst outside the central canal is known as syringomyelia. The differentiation is therefore histological and not relevant in terms of clinical differentiation or management. Both are considered as syringomyelia for this chapter.

Milhorat classified syringomyelia based on autopsy and findings on magnetic resonance imaging (MRI) as communicating, noncommunicating, atrophic, and neoplastic types (Table 24.2). The atrophic and neoplastic types are not relevant to this chapter.

In communicating syringomyelia, there is dilatation of the central canal, which is also in continuum with the fourth ventricle. Usually there is hydrocephalus due to obstruction to the CSF pathway distal to the fourth ventricle outlets. The central canal therefore acts like a “fifth ventricle.” Communicating syrinxes are less prone compared with noncommunicating cavities to dissect or rupture paracentrally, and therefore usually asymptomatic or associated with only minor neurological findings. Etiological conditions include postmeningitic and posthemorrhagic hydrocephalus, complex hindbrain anomalies, such as Chiari II malformation and encephalocele, and Dandy-Walker cysts.

In noncommunicating syringomyelia, there is either dilatation of the central canal, which does not communicate

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**Table 24.2  Classification of syringomyelia**

<table>
<thead>
<tr>
<th>Classification</th>
<th>Location of Syrinx</th>
<th>Clinical Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>I Communicating</td>
<td>Dilation of the central canal in communication with the fourth ventricle</td>
<td>Communicating hydrocephalus (posthemorrhagic, postmeningitic) Complex hindbrain malformations (Chiari II, encephalocele) Dandy-Walker cyst Chiari malformations Basilar invagination Spinal arachnoiditis (posttraumatic, postmeningitic) Extramedullary compressions (spondylosis, tumors, cysts) Tethered cord Acquired tonsillar herniation (hydrocephalus, intracranial mass lesions, craniosynostosis)</td>
</tr>
<tr>
<td>II Noncommunicating dilation of the central canal</td>
<td>Dilation of the central canal not communicating with the fourth ventricle; often dissects into the spinal cord parenchyma</td>
<td>Spinal cord trauma Ischemia/infarction Intramedullary hemorrhage Degenerative changes occurring in conjunction with cord atrophy or secondary to cord atrophy</td>
</tr>
<tr>
<td>II Noncommunicating parenchymal cavitation</td>
<td>Primary spinal cord parenchymal cavitations</td>
<td>Spinal cord trauma Ischemia/infarction Intramedullary hemorrhage Degenerative changes occurring in conjunction with cord atrophy or secondary to cord atrophy</td>
</tr>
<tr>
<td>III Atrophic (syringomyelia ex vacuo)</td>
<td>Ex vacuo dilatation of the central canal, microcysts, intramedullary cleft</td>
<td>Spinal cord trauma Ischemia/infarction Intramedullary hemorrhage Degenerative changes occurring in conjunction with cord atrophy or secondary to cord atrophy</td>
</tr>
<tr>
<td>IV Neoplastic</td>
<td>Within the tumor</td>
<td>Cystic degeneration of intramedullary tumors (e.g., astrocytomas and ependymomas)</td>
</tr>
</tbody>
</table>

with the fourth ventricle, or a separate extracanalicular syrinx that arises in the spinal cord parenchyma and does not communicate with the central canal or fourth ventricle (primary parenchymal cavitation as a result of spinal cord injury due to trauma, ischemia/infection, or spontaneous intramedullary hemorrhage).

Noncommunicating central canal dilatation is associated with obstruction to the CSF pathways at or below the level of the foramen magnum, the most common etiology by far being the Chiari I malformation. Other etiological factors are basilar invagination, spinal arachnoiditis, extramedullary compressions, tethered cord, and acquired tonsillar herniation. Unlike a communicating syrinx, these syringes often dissect into the spinal cord parenchyma, usually to the dorsolateral quadrant of the spinal cord, and can extend to the pial surface to communicate with the subarachnoid space. Also, unlike a communicating syrinx, they are often symptomatic with neurological findings correlating to the anatomy of cavitation demonstrated on MRI.

#### Etiology and Pathogenesis

**Chiari I Malformations**

Chiari described hindbrain anomalies in infants and preterm births. Therefore, subsequent descriptions of tonsillar herniation in adults assumed the condition to be present since birth. A difficult labor can cause excessive molding of the skull, thereby pressing the brain into the foramen magnum, with anoxia, hemorrhage, and hydrocephalus setting the stage for chronic tonsillar herniation. There have been reports of familial aggregation of Chiari malformation and/or syringomyelia, implying an underlying genetic basis, with an autosomal dominant and predominant autosomal recessive inheritance pattern. Morphometric studies of the posterior cranial fossa have shown that the posterior fossa is small due to an underdeveloped occipital bone. Because the posterior fossa contents are of normal size, they are “squeezed” by the underdeveloped occipital enchondrium, with a secondary downward shift of the brain and upward shift of the cerebellar tentorium. Basilar invagination due to a more severely underdeveloped occipital enchondrium results in worsening of the overcrowded posterior cranial fossa. There also may be a role of craniospinal CSF pressure gradient in the pathogenesis of Chiari I malformations. Due to failed neurulation, a dorsal myeloschisis is created, leading to excessive ventricular CSF drainage through the defect. The ventricular system is therefore unable to distend normally and does not exert its normal inductive effect of pressure and volume on the surrounding mesenchyme and endochondral bone formation. This results in a small posterior fossa, and the developing cerebellum and brainstem herniate through the incisura and foramen magnum. As opposed to Gardner’s unifying theory, in which hydrocephalus is felt to initiate tonsillar and brainstem herniation, McLone suggested that hydrocephalus arises as a result of hindbrain herniation. CSF outflow can be impaired by “collapse-induced” developmental abnormalities (blockage) due to the cerebral aqueduct, obstruction of the outlets of the fourth ventricle, or obstruction at the level of the incisura. It is also possible that the abnormally low CSF pressure, as a result of overdrainage, may have an adverse effect on developing arachnoid granulations, leading to hydrocephalus, especially after closure of the neural tube defect.

**Chiari II Malformations**

The dysraphic condition occurring in the setting of Chiari II malformations has been proposed to arise from many causes, including the arrest of normal closure of the neural tube, overgrowth during neurulation, reopening of a closed neural tube, secondary to altered CSF dynamics, and maternal vitamin B6 (thiamine) levels. McLone proposed a unifying theory to explain the development of Chiari II malformations.

**Syringomyelia**

Gardner’s and other earlier theories on the pathogenesis of syringomyelia assumed that there was a communication in all cases between the fourth ventricle and the syrinx. These early theories therefore focused on the transmission of pressure from the intracranial space into the central canal, via the fourth ventricle.

**Cephalocranial Disproportion Theory**

Shunting procedures, common to all such cases, can be associated with the arrest of skull growth. The cranial contents grow faster than the surrounding cranium, and skull growth is again stimulated as the brain continues to grow and fill the space previously occupied by the ventricular system, but not before a transient rise in intracranial pressure produces herniation of the cerebellar tonsils through the foramen magnum.

**Craniospinal Pressure Gradient Theory**

According to this theory, the artificially created pressure gradient due to the relatively lower intraspinal pressure caused by the spinal shunting or drain is the driving force for the tonsillar descent. For acquired Chiari malformations, the conversion of a lumbar shunt to a ventricular shunt may eliminate the downward craniospinal pressure gradient, allowing the reversal of the tonsillar descent or at least arresting further migration.

**Acquired Chiari Malformations**

Two theories have been proposed to explain the development of acquired Chiari malformations.
Gardner’s Hydrodynamic Theory

Prior to Gardner’s theory put forth in the late 1950s, the popular belief was that syringomyelia was the result of cystic degeneration in nests of glial proliferation, scattered throughout the spinal cord. Gardner postulated that in the presence of obstruction to the foramina of Luschka and Magendie, there would be exaggerated ventricular pulsations from the choroid plexus. At 6 to 8 weeks of the embryonic period, there could be failure to develop permeability to CSF flow in the roof of the fourth ventricle. The subsequent hydrocephalus was felt to cause posterior displacement of the primitive transverse sinus and herniation of the hindbrain and cerebellum through the foramen magnum. Because of obstruction at the foramina of Luschka and Magendie, the “water-hammer” effect of the choroid plexus–driven CSF flow through the obex caused dilation of the central canal and subsequent syringomyelia. Therefore, Gardner proposed relieving outflow obstruction through lysis of arachnoidal adhesions and treatment of the syrinx by plugging the obex. However, Gardner’s theory cannot explain syrinx formation in most cases where there is no hydrocephalus, and the fourth ventricle outlet is patent.

Craniospinal Pressure Dissociation Theory

This theory was proposed by Cameron and later modified by Williams. Tonsillar herniation caused obstruction to the caudal flow of CSF, from the cranium at the foramen magnum. This resulted in a craniospinal pressure gradient, causing CSF to flow from the fourth ventricle into the central canal, with subsequent syrinx formation. Differences from Gardner’s theory are outlined in Table 24.3.

“Slosh” and “Suck” Effects

Unlike Gardner’s theory, syrinx expansion was felt to be the result of increases in intrathoracic pressure and not intracranial pressure. Sneezing/coughing caused expansion of the epidural spinal veins and moved CSF from the spinal canal into the cranial subarachnoid spaces through the foramen magnum. In this process, the syrinx was compressed, and its fluid was pushed up and down the central canal, expanding the syrinx cavity (“slosh” effect). At the end of the Valsalva maneuver, the tonsillar herniation prevented the return of CSF from the cranial cavity into the spinal subarachnoid space. Therefore, the pressure gradient (higher intracranial to lower intraspinal) continued to be maintained, forcing further CSF from the fourth ventricle, through the obex, and into the central canal, leading to further syrinx expansion (“suck” effect). This hypothesis is supported by gradients in excess of 100 mmHg, documented after coughing episodes.

“Milking” Action at the Level of the Foramen Magnum

Du Boulay showed that in response to cerebral hemisphere expansion due to arterial inflow during systole, there were periodic movements of CSF in the basal cisterns. He suggested that in the presence of tonsillar herniation, these movements resulted in a ball-valve plugging of the foramen magnum by the cerebellar tonsils. Therefore, during systole, the tonsillar impaction resulted in CSF from the basal cisterns being diverted into the fourth ventricle and the central canal. During diastole, retraction of the tonsils and brainstem resulted in a “milking” action at the foramen magnum, causing further movement of the CSF along the central canal. Such a repetitive activity, over time, could lead to the formation and expansion of a syringomyelia. During coughing, a large upward displacement of CSF throughout the spinal axis and up to 50% reduction in the anteroposterior diameter of the thecal sac occurred; therefore, an increase in spinal venous pressure was felt to be the main factor in syrinx expansion following a Valsalva maneuver.

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Table 24.3 Differences between Gardner’s hydrodynamic and Williams’ craniospinal pressure dissociation theories

<table>
<thead>
<tr>
<th>Obstruction to cerebrospinal fluid flow</th>
<th>Formation of syringomyelia</th>
<th>Due to scarring of the fourth ventricle outlet foramina</th>
<th>At the foramen magnum, due to tonsillar herniation</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Gardner’s Hypothesis</strong></td>
<td><strong>Williams’ Hypothesis</strong></td>
<td>Hydrodynamic theory: arterial pulse pressure from the choroid plexus acts from within the neuraxis, through the central canal centrifugally, to form and expand the syrinx</td>
<td>Craniospinal pressure dissociation theory: relatively prolonged intrathoracic venous distention acting initially on the syrinx from outside (“slosh”) and then from within (“suck”)</td>
</tr>
</tbody>
</table>

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Ball and Dayan’s Theory

Ball and Dayan found that in the walls of syrinx cavities were prominent small arteries and veins, as well as dilated perivascular spaces around the cavities. They
felt the dilated perivascular spaces were likely to represent enlarged Virchow-Robin spaces. When India ink was injected into the lumen of the syrinx, it was found to spread out around blood vessels and reach the subarachnoid space. The authors therefore postulated that, with obstruction at the foramen magnum, syringomyelia could develop as a result of CSF dissecting into the spinal cord through dilated Virchow-Robin spaces.

Rennels et al. demonstrated that horseradish peroxidase in the CSF penetrates rapidly and deeply into the Virchow-Robin spaces at various sites of the central nervous system, and the penetration is dependent on systolic pulsations in the CSF. Stoodley and coworkers subsequently confirmed these observations.

Oldfield’s Theory

Oldfield and coworkers further expanded this theory. According to their mechanism, during cardiac systole, there is a pistonlike downward movement of the cerebellar tonsils at the foramen magnum onto the partially entrapped spinal CSF. The resultant spinal subarachnoid heightened systolic CSF pressure and increased pulse pressure cause bulk movement of the fluid from the subarachnoid CSF to the spinal cord, through the perivascular spaces, causing formation of a syrinx. This action also compresses the cord and causes pulsatile flow in the syrinx, leading to expansion of the syrinx.

Levine’s Theory

Levine postulated that the fluid in the syrinx is not from CSF in the fourth ventricle or spinal subarachnoid space. Activities such as assuming an erect posture, coughing, and straining, as well as pulsatile fluctuations of CSF pressure during the cardiac cycle, result in a transient but abrupt increase in CSF pressure above the level of the foramen magnum, in the presence of subarachnoid obstruction (e.g., due to a Chiari malformation). This results in changes in transmural venous and capillary pressure favoring dilation of vessels below and collapse of vessels above the block. This in turn produces mechanical stress on the spinal cord, particularly caudal to the block. Repetition of this stress over a prolonged period of time can produce tissue breakdown and partial disruption of the blood–spinal cord barrier, leading to accumulation of a plasma ultrafiltrate of protein-poor fluid.

Pre-Syrinx State

There have been a few reports of symptomatic cord swelling with Chiari I malformation, and also of syringomyelia associated with tonsillar impaction at the foramen magnum preceded by an interval of cord swelling. It is therefore possible that the cord swelling occurs due to accumulation of excess CSF in the extracellular space, and syringomyelia is produced by the coalescence of the excess extracellular fluid. In some cases, accumulation of the CSF could begin at an isolated segment of a patent central canal.

Chiari I Malformation without Syringomyelia

Why are some Chiari I malformations not associated with syringomyelia? It appears that in these cases, the CSF systolic flow is shortened and therefore insufficient to maintain a hypertensive condition long enough to result in syrinx formation.

Pathology

Chiari Malformations

The foramen magnum usually causes an indentation on the tonsils, dividing the intracranial tonsils from the extracranial herniated tonsils. The tonsils are also usually flattened (Figs. 24.1 and 24.2). The midbrain can be enlarged, low positioned, and have an exaggerated cleft between it and the pons anteriorly. The pons can also be low lying. Abnormalities of the posteroinferior cerebellar artery (PICA) can be present. The common anomaly is for the cranial loop of the PICA to lie below the foramen magnum. There is often a thick transverse cervicomedullary dural band, arachnoid adhesions with occlusion of the foramen of Magendie, and upward nerve root deviation.

Histopathological examination of paraffin-embedded specimens of herniated tonsils has shown disorganization of all three cellular layers of the cerebellum. In Chiari II malformations, there are often displaced islands of Purkinje, granular, and basket cells deep within the cerebellar white matter. There are also neuronal dropout in all three cortical layers and a reactive astrocytic proliferation.

Fig. 24.1 Postmortem case of Chiari I malformation. Note the compressed, gliotic tonsils and the effect of the shallow posterior fossa on the appearance of the cerebellum. (From Engler GL, Cole J, Merton WL, eds. Spinal Cord Diseases: Diagnosis and Treatment. New York: Marcel Dekker; 1998. Reprinted by permission of Routledge/Taylor & Francis Group, LLC.)
The pia–arachnoid is markedly thickened in the region of the herniation and contains prominent thin-walled vessels and arteriovenous shunts. It has been postulated that these vascular anomalies can cause ischemia or anoxia of the brainstem and the resulting symptomatology.

Postmortem studies of the brainstem below the level of the foramen magnum in children with meningomyelocele and Chiari II malformations show vascular engorgement, hemorrhage, focal areas of neuronal and axonal loss, hemosiderin deposition, and gliosis. Thickening and adhesions between the pia and arachnoid are seen, causing partial obliteration of the subarachnoid space. Cleftlike spaces are seen in the inferior vermis communicating with the fourth ventricle. Other abnormalities are thickened interthalamic adhesions, hypoplasia of the falx cerebri, partial obliteration of the interhemispheric fissure, cortical heterotopia projecting into the walls of the lateral ventricle, cerebral microgyria, and aqueductal stenosis and forking.

**Syringomyelia**

The cervical cord is the most common site to be affected, usually the upper cervical region. It can extend into the thoracic region from a cervical origin. The cavities are incompletely lined by ependyma, the remainder lined by subependymal astrocytic glial cells. Glial tissue septa can divide the cavity into variable compartments. Communication between an eccentrically located syrinx cavity and the central canal can often be demonstrated (**Fig. 24.3**).

**Epidemiology and Clinical Presentation**

**Chiari I Malformations**

With the introduction of noninvasive screening using MRI, the prevalence of asymptomatic Chiari I
In Milhorat et al.’s series of 364 patients, 15 ± 37% had lifelong complaints of headache and clumsiness, and 24% felt that trauma preceded their symptoms.

Most cases are sporadic, with occasional reports of familial aggregation of Chiari malformation and/or syringomyelia, implying an underlying genetic basis in these familial clusters. The predominant pattern of inheritance in these familial cases is autosomal recessive, although an autosomal dominant pattern is also seen.

Milhorat et al. reviewed the clinical features in 364 patients,15 which are summarized in Table 24.4. The common presenting complaint is suboccipital neck pain. This is usually a severe, crushing type of pain that can be pounding in character when severe, but otherwise nonthrobbing. It characteristically worsens with physical exertion and Valsalva maneuvers, such as sneezing, coughing, and straining, along with changes in head position, possibly due to an increase in intracranial pressure. Women of menstrual age exhibit worsening of headache in the week prior to menstruation. The headache is probably the result of dural irritation or stretching of the upper cervical nerve roots.

Symptoms and/or signs of spinal cord dysfunction were found in 94% of patients with associated syringomyelia. In patients with Chiari malformation without syringomyelia, it was less severe and found in 66% (Table 24.5).

Sudden cardiorespiratory arrest, sometimes followed by death, has been associated with Chiari I malformations. Children with Chiari I malformations may be more prone to spinal cord injury and often have scoliosis when syringomyelia is present.

One-quarter to one-half of patients with symptomatic Chiari I malformations have associated skeletal, cerebral, or craniofacial anomalies (Table 24.6).

### Chiari II Malformations

Neural tube defects occur in ~1:1000 live births in North America. The incidence in the United Kingdom and Hungary is higher. About 10% to 20% of these children with meningo(myelome)elocyst un develops symptoms due to cranial nerve, cerebellar, or brainstem dysfunction. Because ~90% of these children have hindbrain herniation of varying degrees at autopsy, there may be a large asymptomatic group in this population as well. There is a higher incidence when the neural tube defect is at or above L3. In patients born with neural tube defects, hindbrain anomalies are the leading cause of death. Symptoms of Chiari II malformation present in infancy, usually before 3 months of age. As a result of the established connection between neural tube defects and folic acid, folic acid diet supplementation could lead to up to 50% reduction in the incidence of Chiari II malformations.16

The clinical presentation for Chiari II is similar to that of Chiari I malformations. However, the associated hydrocephalus plays an important part in the clinical presentation. Symptoms usually develop within the first 3 months of life and include stridor, apnea, and feeding difficulties. Dysphagia is manifested as prolonged feeding time, pooling of secretions in the oropharynx, and coughing or choking episodes during feeding. Because the brainstem abnormalities are more severe in Chiari II compared with Chiari I malformations, apneic episodes, vocal cord paralysis, and a weak cry occur more frequently.

When symptoms arise later in childhood or adolescence, clinical features include hemiparesis, quadriparesis, opisthotonos, and ocular findings such as oscillopsia and nystagmus. The clinical features and lower motor neuron findings of the associated dysraphic state can obscure the long tract signs due to brainstem or spinal cord involvement.

### Acquired Chiari Malformations

Acquired Chiari malformation usually results from some form of neurosurgical intervention. The common procedures related to its development are insertion of a lumboperitoneal shunt for treatment of communicating hydrocephalus or benign intracranial hypertension and external lumbar drain for temporary CSF diversion. Retrospective data suggest that acquired types may be asymptomatic in up to 70% of cases. Symptomatic cases are rare, occurring in ~0.25% to 10%.

### Table 24.4 Clinical symptoms and signs in Chiari I malformations

<table>
<thead>
<tr>
<th>Clinical Picture</th>
<th>Frequency (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Suboccipital pain</td>
<td>81</td>
</tr>
<tr>
<td>Ocular symptoms</td>
<td>78</td>
</tr>
<tr>
<td>Otoneurological symptoms</td>
<td>74</td>
</tr>
<tr>
<td>Nystagmus</td>
<td>26</td>
</tr>
<tr>
<td>Lower cranial nerve, brainstem, and cerebellar disturbances</td>
<td>52</td>
</tr>
<tr>
<td>Spinal cord dysfunction with associated syrinx (see Table 24.5)</td>
<td>94</td>
</tr>
<tr>
<td>Spinal cord dysfunction without associated syrinx (see Table 24.5)</td>
<td>66</td>
</tr>
</tbody>
</table>

The clinical presentation of acquired Chiari malformation is similar to that of Chiari I. However, it can be dramatic, with quadripareisis and cardiopulmonary arrest due to acute foramen magnum syndrome, following acute foramen magnum herniation.

Syringomyelia occurs in ~30% to 40% of Chiari I malformations, although an incidence of up to 85% has been reported. In Chiari II malformations, the frequency may be less. Syringomyelia does not appear to be related to the degree of tonsillar herniation; in fact, it may occur more often with an intermediate degree of herniation. When present, syringomyelia is responsible

### Table 24.5  Spinal cord dysfunction in Chiari malformation with and without syringomyelia

<table>
<thead>
<tr>
<th>Clinical Symptoms and Signs (n = 364)</th>
<th>Chiari Malformation with Syringomyelia (%) (n = 238)</th>
<th>Chiari Malformation without Syringomyelia (%) (n = 126)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Affected patients</td>
<td>94</td>
<td>66</td>
</tr>
<tr>
<td>Sensory Symptoms</td>
<td>74</td>
<td>32</td>
</tr>
<tr>
<td>Paresthesia/hyperesthesia</td>
<td>55</td>
<td>23</td>
</tr>
<tr>
<td>Nonradicular segmental pain</td>
<td>52</td>
<td>19</td>
</tr>
<tr>
<td>Analgesia/anesthesia</td>
<td>38</td>
<td>12</td>
</tr>
<tr>
<td>Burning dysesthesia</td>
<td>30</td>
<td>9</td>
</tr>
<tr>
<td>Poor position sense</td>
<td>33</td>
<td>2</td>
</tr>
<tr>
<td>Impaired temperature sense</td>
<td>66</td>
<td>40</td>
</tr>
<tr>
<td>Motor Symptoms</td>
<td>39</td>
<td>13</td>
</tr>
<tr>
<td>Weakness</td>
<td>12</td>
<td>2.4</td>
</tr>
<tr>
<td>Spasticity</td>
<td>24</td>
<td>12</td>
</tr>
<tr>
<td>Atrophy</td>
<td>22</td>
<td>7</td>
</tr>
<tr>
<td>Other Symptoms</td>
<td>29</td>
<td>12.5</td>
</tr>
<tr>
<td>Trophic phenomena</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Urinary incontinence</td>
<td>43</td>
<td>14</td>
</tr>
<tr>
<td>Impotence (adult male patients)</td>
<td>30</td>
<td>0</td>
</tr>
<tr>
<td>Fecal incontinence</td>
<td>25</td>
<td>4</td>
</tr>
<tr>
<td>Sensory Findings</td>
<td>61</td>
<td>20</td>
</tr>
<tr>
<td>Analgesia/anesthesia</td>
<td>47</td>
<td>38</td>
</tr>
<tr>
<td>Dissociated sensory loss</td>
<td>26</td>
<td>6</td>
</tr>
<tr>
<td>Impaired position sense</td>
<td>11</td>
<td>1</td>
</tr>
<tr>
<td>Motor Findings</td>
<td>45</td>
<td>14</td>
</tr>
<tr>
<td>Weakness</td>
<td>24</td>
<td>6</td>
</tr>
<tr>
<td>Impaired fine motor function</td>
<td>25</td>
<td>1</td>
</tr>
<tr>
<td>Increased muscle tone</td>
<td>13</td>
<td>3</td>
</tr>
<tr>
<td>Atrophy</td>
<td>1</td>
<td>25</td>
</tr>
<tr>
<td>Abnormal Tendon Reflexes</td>
<td>Hyperreflexia</td>
<td>18</td>
</tr>
<tr>
<td>Clonus or Babinski sign</td>
<td>7</td>
<td>57</td>
</tr>
<tr>
<td>Hyporeflexia</td>
<td>1</td>
<td>59</td>
</tr>
<tr>
<td>Other Findings</td>
<td>Trophic changes</td>
<td>4</td>
</tr>
</tbody>
</table>


### Table 24.6  Associated anomalies seen in cases of symptomatic Chiari I malformation

<table>
<thead>
<tr>
<th>Skeletal</th>
<th>Atlanto-occipital assimilation</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Platybasia</td>
</tr>
<tr>
<td></td>
<td>Basilar invagination</td>
</tr>
<tr>
<td></td>
<td>Abnormal odontoid segmentation</td>
</tr>
<tr>
<td></td>
<td>Fused cervical vertebrae</td>
</tr>
<tr>
<td></td>
<td>Cervical ribs</td>
</tr>
<tr>
<td></td>
<td>Fused thoracic ribs</td>
</tr>
<tr>
<td></td>
<td>Kyphoscoliosis (syrinx associated)</td>
</tr>
<tr>
<td>Cerebral</td>
<td>Lipoma of the corpus callosum</td>
</tr>
<tr>
<td></td>
<td>Septo-optic dysplasia</td>
</tr>
<tr>
<td></td>
<td>Aqueductal stenosis</td>
</tr>
<tr>
<td>Craniofacial</td>
<td>Choanal atresia</td>
</tr>
</tbody>
</table>

Syringomyelia occurs in ~30% to 40% of Chiari I malformations, although an incidence of up to 85% has been reported. In Chiari II malformations, the frequency may be less. Syringomyelia does not appear to be related to the degree of tonsillar herniation; in fact, it may occur more often with an intermediate degree of herniation. When present, syringomyelia is responsible
for the clinical presentation, rather than the tonsillar herniation.

The clinical presentation of Chiari-associated syringomyelia is presented in Table 24.5. The common presenting features of numbness, weakness, and pain in the upper extremities are similar to those seen in patients with syringomyelia due to any cause. Pain or paresthesias can be aggravated by Valsalva-like maneuvers raising the intrathoracic pressure, such as sneezing and coughing. In children, scoliosis is often present due to the syringomyelia from Chiari malformation.

### Diagnosis

#### Chiari Malformations

MRI is noninvasive and has become the gold standard for the radiological diagnosis of Chiari malformation and syringomyelia (Fig. 24.4). In addition, phase-contrast MRI gives information for quantitative and qualitative CSF flow analysis. This can prove useful in determining alterations in normal CSF flow dynamics in patients with Chiari I malformations; it can also help document normalization of flow parameters postoperatively.

#### Localizing the Level of the Foramen Magnum Using MRI

Fat within the bone marrow gives off high signal on T1 images and can be recognized within the clivus and basiocciput. At the lowest end of the clivus (basion) and the posterior margin of the foramen magnum (opisthion), the cortical bone gives off a low signal, which contrasts with the high signal from the bone marrow. This line from the basion to the opisthion is used to define the foramen magnum.

#### MRI Criteria for the Diagnosis of Chiari I Malformation

These have been developed studying symptomatic patients with Chiari I malformations compared with normal controls. They are as follows: (1) unilateral or bilateral tonsillar herniation ≤ 5 mm below the foramen magnum; (2) bilateral tonsillar herniation 3 mm below the foramen magnum, if accompanied by other features consistent with Chiari I malformation, such as a syringomyelia or cervicomedullary kinking; and (3) the absence of Chiari II malformation, prior cranial or cervical spinal surgery, previous shunting procedure, or intracranial mass.

#### Normal Position of the Cerebellar Tonsils

The cerebellar tonsils are usually located 1 to 3 mm above the level of the foramen magnum. In 14% of normal patients, the tonsils extend below the foramen magnum by up to 3 mm. Tonsillar herniation < 2 mm may not be of clinical significance.

Chiari II malformations are usually described in the background of a known meningomyelocele. MRI remains the diagnostic procedure of choice in this group. The degree of tonsillar herniation is usually more severe, and the other brain abnormalities have been discussed previously.

#### Syringomyelia

MRI has revolutionized the diagnosis of syringomyelia and intramedullary lesions. Prior to the MRI era, myelography and delayed-contrast computed tomography (CT) imaging defined syringes poorly and missed them in 1 to 5 of every 10 patients. On MRI, the signal characteristics of the syrinx cavity are similar to CSF: low signal on T1- and high signal on T2-weighted images. Septations can often be visualized on sagittal images. The radiological extent of the syrinx, however, does not necessarily correlate with the duration or severity of symptoms or clinical findings.

#### Swallowing Studies

Swallowing studies such as cine-esophagogram and pharyngoesophageal manometry are useful to determine if...
supplemental nutrition is necessary and the type of alternate feeding technique required. They are also useful for assessing progress postoperatively.

**Treatment**

**Chiari I and II Malformations**

There are no good randomized trials to form an evidence-based management system for Chiari malformations. Obviously, the management is guided by the individual patient’s clinical presentation (and not the radiological findings alone), the experience of the treating surgeon, and the historical experience of others. More and more patients are being detected on MRI with asymptomatic Chiari malformations; such patients are probably best dealt with by observation and follow-up. Progressive or disabling symptomatology is the common indication for surgery. Our surgical approach is as follows. Surgical decompression of the Chiari malformations consists of decompression of the foramen magnum (removal of 1.5 cm of bone on either side of the midline, and 2.5 cm superiorly from the foramen magnum) and usually a C1 and/or C2 laminectomy (Fig. 24.5). The laminectomy is carried down inferiorly as low as it is necessary to reach beyond the lower pole of the herniated tonsils. A Y-shaped durotomy is made, keeping the arachnoid intact (Fig. 24.6), and a dural substitute or fascia lata graft is used and sutured to the dural edges to achieve a watertight dural closure (Fig. 24.7). This is reinforced with fibrin glue and Gelfoam. This procedure has been shown to yield good results and is aimed at dealing with the cause of the problem—obstruction at the craniocervical junction.\(^{13,22}\)

We do not attempt to release arachnoidal adhesions, remove the tonsils, shunt the syrinx, or plug the obex at primary surgery. Patients who remain symptomatic or deteriorate with a persistent or enlarging syrinx are considered for a shunt procedure. Larger bony decompressions have been described, but care should be taken, as this can increase the risk of cerebellar subsidence and recurrence of symptoms.\(^{22,23}\)

There seems to be a subset of patients whose symptoms will resolve and syrinx cavity will decrease with the removal of bone only. These patients seem to undergo a volumetric increase in the posterior fossa, but it is difficult to identify these patients. Perioperative use of ultrasound has been described to decide on the extent of this surgery. After doing a bony decompression, if the ultrasound findings show inadequate decompression, scouring of the outer layer of the dura can be done to promote dural expansion and, if inadequate, be followed by durotomy and duraplasty.\(^{13,22,24}\)

In the few patients who present with hydrocephalus, consideration may be given to performing a ventriculoperitoneal shunt.

**Acquired Chiari Malformations**

Removing the lumboperitoneal shunt usually results in resolution of the symptoms and improvement in tonsillar descent at the foramen magnum. If CSF diversion continues to be necessary, it can be done with a ventriculoperitoneal shunt or some other diversion technique. If the ventricles are small, stereotactic placement of the ventricular catheter can be done. In cases of symptomatic acquired Chiari malformation following posterior fossa decompression, duraplasty may be necessary.

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**Fig. 24.5** Intraoperative photograph showing the typical extent of bony decompression done prior to dural opening for a Chiari malformation.
Syringomyelia

In the presence of a Chiari malformation, the first line of treatment of a symptomatic syrinx should be decompression of the craniocervical junction. If this procedure fails, then shunting of the syrinx can be considered. In such a case, the syrinx should be entered and the shunt tube placed at the thoracic extension of the syrinx (T3 or lower) or the lowest cervical level where the syrinx is accessible. This is to prevent the potential for upper limb dysfunction and decrease the length of tube required for a cystopleural or cystoperitoneal shunt. A two- or three-level laminectomy is performed, the dura is opened, and the syrinx is entered through a small myelotomy at the dorsal root entry zone. A midline myelotomy can be performed, but this increases the risk of bilateral posterior column deficit. The two short ends of the T tube are placed in the syrinx, while the long end is tunneled and placed in the pleural or peritoneal space. The pleural space is closer to the site of surgery, requiring a shorter shunt tube, and has a relatively negative pressure compared with the peritoneum; it is thus our preferred site. Although syringosubarachnoid shunting is still actively advocated by surgeons,25–27 others have had poor results, probably due to scarring in the subarachnoid space leading to tube blockage.

Fig. 24.6 Intraoperative photograph showing the intact arachnoid after the dural opening. The lower end of the tonsil is visible, confirming adequate decompression.

Fig. 24.7 Intraoperative photograph showing a dural patch sutured to the edges of the dural defect.
A lumboperitoneal shunt with or without myelotomy has been shown to be useful in the treatment of patients with syringomyelia even in the presence of Chiari malformations, some of whom had undergone previous craniocervical decompression. However, we feel that it is best used not at all or as a last resort of treatment following failure of craniocervical decompression and in all syrinx shunting procedures in severely symptomatic patients. This is because of the potential for aggravating a preexisting Chiari malformation with a small risk of disastrous consequences. In addition, only a few patients have been reported with short-term follow-up using this technique.

**Morbidity and Mortality**

**Craniocervical Decompression**

In the early days of neurosurgery, there were significant mortality and morbidity associated with decompression for Chiari malformations. More recent experiences have shown that it is a relatively low-risk procedure.

Respiratory depression is the most serious postoperative complication and can result in death. In survivors, it usually resolves within the first week of surgery. The incidence of CSF leak and aseptic meningitis has been reduced with the practice of duraplasty. The proponents of partial splitting of the dura, of course, claim that these complications can be completely avoided. Other complications are postoperative wound hematoma, weakness and sensory loss that may be temporary or permanent, and cranial nerve palsies.

**Syringomyelia Shunt Procedures**

Complications for procedures involving shunting of the syrinx include meningitis, shunt infections and obstruction, overdrainage leading to headache, and worsening neurological deficit. Although complication rates as high as 26% have been reported, more recent series have shown a low rate of complications. Reoperation rates are high (17–19%), although most of these occurred in early series. Avoiding placement of the distal end of the shunt in the dorsal subarachnoid space and meticulous surgical technique to avoid spillage of blood into the subarachnoid space may reduce the incidence of reoperation.

**Prognosis**

**Chiari I Malformations**

The natural history of patients with Chiari I malformations is largely unknown. Few case reports have shown spontaneous resolution of syringomyelia along with Chiari malformations and other craniocervical junction lesions, such as basilar invagination.

Patients with symptomatic Chiari I malformations treated nonsurgically have a comparable proportion to those who deteriorate or remain stable when observed for several years. Most patients, including those surgically treated, show long periods of stability interrupted by stepwise neurological deterioration. Apart from a few case reports in nonsurgically treated patients, spontaneous remission of symptoms has not been recorded. In contrast, patients undergoing surgery show improvement routinely. Although surgery is definitely beneficial, there are no well-designed studies to show that surgery alters the natural course of Chiari malformations. Given the beneficial effects of surgery, it is probably unethical not to surgically treat symptomatic patients with or without syringomyelia in whom there is progressive symptomatology.

Headache and neck pain, which are the most common symptoms of Chiari I malformations, are relieved in < 90% of patients who have undergone craniocervical decompression. Cerebellar dysfunction also improves well following surgical decompression. Paresthesias and sensory deficits improve or resolve in ~40 to 100% of patients. Scoliosis remains stable or improves in 70% to 100% of patients following craniocervical decompression with or without a syrinx drainage procedure.

**Chiari II Malformations**

The long-term outcome for patients with Chiari II malformations treated with surgery depends mainly on the severity of associated anomalies. Retethering following myelomeningocele repair can lead to neurological or urological deterioration. Again, the natural history is not completely understood here, and patients remain free from neurological deterioration for prolonged periods following surgery.

The mortality rate in infants with symptomatic Chiari II malformations is high (≥ 50%) in both the surgical and nonsurgical groups, with the survivors doing well. In children and adolescents with symptomatic Chiari II malformations, the prognosis may be as good as that of adults with type I malformations.

**Brainstem Involvement**

Resolution of dysphagia following craniocervical decompression is found in 30 to 70% in large series, although it has been reported to occur in up to 100% of cases. As expected, the postoperative outcome depends on the preoperative function. Advanced degree of brainstem involvement can result in ventilatory dependence and death; therefore, early surgical intervention is crucial in these patients. In patients who present late in the course of the disease or progress relentlessly despite surgery, recurrent pneumonia is the most common cause of death.
Overall

Favorable outcomes for craniocervical decompression with or without syrinx shunting range from 50 to 85%. In most cases, the syrinx associated with the Chiari I malformation reduces in size, and more than 50% of syrinxes can disappear by 6 months following surgery. Longer-term follow-up has shown worsening in 25% of patients in older series and is better (worsening in 6%) in more recent series. Although delayed deterioration has been reported in up to 20% of patients who improved following surgery, others have not shown such a high rate.

Neurological Deterioration in the Postoperative Setting

This can arise from a variety of causes, such as postoperative hematoma, meningitis (aseptic or bacterial), pseudomeningocele, occipital–C1–C2 instability, recurrence of syringomyelia, “slumping” or “sagging” of the cerebellum due to excessive decompression of the posterior fossa, and inadequate decompression of the posterior fossa with persistent tonsillar herniation.

Conclusion

Chiari malformations cover a spectrum of disorders with varying degrees of cerebellar herniation. The Chiari I malformation usually presents in adulthood, compared with Chiari II, which presents in childhood. Chiari II can be differentiated from Chiari I malformation, as it is associated with meningocele. Lumboperitoneal shunting can lead to symptomatic acquired Chiari malformation. Dysphagia is a sign of brainstem dysfunction, and patients with dysphagia should be treated early and aggressively with surgical intervention. MRI is the investigation of choice for Chiari malformation and syringomyelia. The cervical spine should be routinely imaged in all cases of Chiari malformations, because of the high incidence of associated syringomyelia. Although a variety of surgical procedures exist for the treatment of Chiari malformations with or without syringomyelia, craniocervical decompression followed by duraplasty is probably the procedure of choice. Long-term follow-up has shown good results in the majority of patients.

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Syringomyelia is a frequently encountered clinical and radiological entity. It is associated with a range of pathological conditions, including congenital craniovertebral anomalies, tumors, infections, trauma, and spinal deformities. Syringomyelia is rarely or never an acute phenomenon and is generally encountered in long-standing and persistent space-occupying or space-reducing lesions and in situations with subtle but persistent instability. Obstruction to the normal flow of spinal or cranial cerebrospinal fluid (CSF) by posttraumatic or infective arachnoiditis is also a less frequently encountered cause of syringomyelia. Acute injuries, acute infections, rapidly growing malignant tumors, mobile dislocations, and acute posttraumatic deformities are generally not associated with syringomyelia. The clinical presenting features are long-standing and relentlessly progressive.

Our analysis of the issue suggests that syringomyelia or hydrocephalus of the cord is never a primary phenomenon. It is always secondary to an obvious or an unidentified pathology. The understanding of the pathophysiology of syringomyelia is crucially important to design a suitable treatment strategy and prognosticate the ultimate outcome. We concluded from our earlier study that syringomyelia is a natural protective phenomenon that balances the disturbed pressure situation in an attempt to reduce neural compromise to the minimum. Although syrinx formation by itself can have clinical sequel, the entire process works for safeguarding the general interest of patients. On the basis of our experience, we are convinced that syrinx drainage as a primary procedure should never be considered as a therapeutic option. All attempts must be made to identify and treat the primary pathological cause. Syrinx drainage or shunting without dealing with the primary pathology can result in disastrous clinical consequences.

### Relevant Embryology

Syringomyelia is commonly associated with the spinal abnormality integral to the complex of basilar invagination and Chiari malformation. From our previous study, we concluded that Chiari malformation and related pathological events could be primarily attributable to the maldevelopment of the occipital bone and overcrowding of the normally developed cerebellum within a smaller posterior cranial fossa.

Neural development is initiated and completed early in embryonic life. Almost all named arteries and cranial nerves and much of the substance of the cerebral and cerebellar hemisphere are formed before any spicule of bone develops. Bone formation is almost an afterthought. Musculoskeletal development of the craniovertebral junction occurs in the latter third of fetal life when the cervical flexure of the fetus is reversed. This development precedes the delivery of the child, and the basic formation of the craniocervical junction is completed when the infant starts holding his or her head at 3 months. The cerebellar mass continues to grow in bulk and texture at least until 1 year of age.

We observed that the craniovertebral maldevelopment results in reduced length of the clivus (i.e., the sphenoid part of the clivus is formed relatively normally, whereas the occipital part is formed incompletely) and platybasia. Occipital condylar hypoplasia, nonformation or inadequate formation of the occipitoaxial joint, occipitalization of the atlas, fusion of the atlantoaxial joint and C2–C3 spinal elements, and a range of Klippel-Feil spinal anomalies are also frequently associated. The entire complex of the odontoid process, atlas, and clivus is located rostrally so that the volume of the posterior cranial fossa is effectively reduced. Basal mesodermal maldevelopment results in rostral positioning of the plane of the foramen magnum and eventually in group II basilar invagination.

### Pathophysiology

Basilar invagination and associated basal maldevelopment, with resultant reduction in the volume of the posterior cranial fossa, are the primary events in syringomyelia pathology. There is usually no demonstrable structural abnormality of the brainstem, cerebellar hemisphere, or fourth ventricle, suggesting that the neural development in these patients remained unaffected in the embryonic dysgenesis. The presence of the normal bulk of the cerebellum in the reduced-volume posterior cranial fossa results in the herniation of part of the cerebellum into the foramen magnum, an entity labeled Chiari I malformation. Essentially, it appears that syringomyelia is a tertiary event to primary basilar invagination and secondary Chiari malformation. Long-standing pulsatile pressure of the herniated tonsil into the foramen magnum can result in compression of
the cervicomedullary cord. Apart from physical attempts to increase the volume of the foramen magnum by bone erosion and membranous thinning and expansion, the body probably provides a cushioning counterbalance of fluid from the spinal end in the form of syringomyelia so that the pressure of the tonsils on the cervicomedullary cord is minimized. The signs and symptoms related to the syrinx are more predominant than those related to brainstem compression by tonsillar herniation. The primary aim of the entire process of syrinx formation is to reduce the compression of the brainstem at the level of the foramen magnum by the herniating cerebellum.

We analyzed cases from our departmental archives (Seth G.S. Medical College and K.E.M. Hospital, Parel, Mumba, India) and observed that there was a group of such cases that were treated by syringosubarachnoid shunt surgery as the primary modality of therapy. Although symptoms related to spinal cord dysfunction improved after surgery, symptoms related to brainstem and upper cervical cord compression worsened. In a way, a syrinx protects the vital neural structures at the level of the craniovertebral junction at the cost of the spinal cord.

The mechanism of formation of a syrinx within the cord could be similar to the formation of hydrocephalus within the brain. In our earlier studies, we suggested that tumor obstructive hydrocephalus could be a natural protective phenomenon of the body. The hydrocephalus and an increase in supratumoral intracranial pressure are attempts to reduce the pressure of the tumor over vital brain structures, such as the brainstem and hypothalamus. Similarly, it appears that syringomyelia could be a phenomenon acting as a counterbalance to the relentless pressure exerted by a Chiari malformation.

Rationale of Treatment

Our observations suggest that the treatment strategy in such cases should be directed toward increasing the posterior cranial fossa volume by posterior fossa volume expansion or foramen magnum decompression. The routine involving opening of the posterior cranial fossa dura, resection of the cerebellar tonsils, arachnoidal sectioning, and syringostomy is unnecessary. It appears that if the primary problem of basilar invagination is dealt with early in the course of the disease, the secondary and tertiary events will regress spontaneously.

Primary syringomyelia, in which the cause is not identifiable, is a relatively complex clinical situation. The syringomyelia in such cases simulates normal pressure hydrocephalus. The intrasyrinx pressure is relatively low in such cases, and the results of any kind of treatment are far from gratifying (Fig. 25.1).

When there is syringomyelia and Chiari malformation but no definite basilar invagination (Figs. 25.2, 25.3, and 25.4), efforts can be made to identify the cause of the Chiari malformation. In such cases, it might be possible that the cerebellar mass is larger than normal. Even in such cases, volume expansion of the posterior cranial fossa is required. The theory that the dura or dural bands can act as a compressive factor appears unacceptable and does not correlate well with the pathogenic events. The dura is an expansile structure and can never be a compressive factor.

Primary syringomyelia secondary to primary cord pathology, such as a tumor or kyphosis (Figs. 25.5, 25.6a, and 25.7), also appears to be a secondary and protective phenomenon. Treatment in such cases should be directed toward the pathology and not toward the syringomyelia. Our observation is that if the syringomyelia is treated without dealing with the primary pathology, more often than not the outcome is poor, and the patient can be harmed.

Conclusion

Syringomyelia turns out to be a good teacher, provided we learn to trust nature, best enshrined in the phrase *Vis medica trix Naturae*. We have not even scratched the surface...
Fig. 25.2 T1-weighted MRI showing basilar invagination, Chiari I malformation, and syringomyelia.

Fig. 25.3a,b
a T1-weighted MRI showing basilar invagination, Chiari I malformation, and syringomyelia. The herniated part of the tonsil shows a cystic cavitation.

b T2-weighted MRI showing basilar invagination, Chiari I malformation, and syringomyelia.
Fig. 25.4a–f
a  T1-weighted MRI of an infant showing dumbbell-shaped syringomyelia.
b  T2-weighted MRI showing the syrinx.
c  Lateral radiograph with the head in a flexed position showing atlantoaxial dislocation.
d  Radiograph with the head in extension showing complete reduction of the dislocation.
e  Postoperative radiograph with the head in flexion showing fixation of the dislocation in a reduced position.
f  Radiograph with the head in extension showing the fixation.
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Fig. 25.5  MRI showing a large intramedullary tumor and an associated syrinx.

Fig. 25.6a–c
a  T2-weighted MRI showing kyphosis, cord compression, and syringomyelia.
b  T1-weighted image.
c  MRI of the upper spine showing an extensive syrinx.
of the hidden mysteries in this seeming abnormality. Any urge to deflate a syrinx should be tempered by the need to exonerate the syrinx in the court of intellectual appeal. Syrinxes are not only innocent, but useful as well.\textsuperscript{14,15}

References

The surgical management of congenital craniovertebral anomalies is complex due to the relative difficulty in accessing the region, the critical relationships of neurovascular structures, and the intricate biomechanical issues involved. Basilar invagination forms a prominent component of the craniovertebral anomalies. Chiari malformation and syringomyelia are the most common associates of basilar invagination and are the soft tissue components of dysgenesis. Basilar invagination has been a subject of clinical interest for a long time. Various classical presentations have referred to this issue. Plain radiological and tomographic parameters have been used to diagnose basilar invagination for many years. There has been a renewed interest in the normal anatomy and the pathological lesions of the craniovertebral junction with the development of imaging by high-resolution computed tomography (CT) and magnetic resonance imaging (MRI). Improved imaging has provided an opportunity to clearly observe the bony abnormality and the distorted neural and vascular relationships. Dynamic MRI and CT have helped in the evaluation of the pathology of basilar invagination, in the assessment of biomechanics of the joints, and in the formulation of a surgical strategy. Despite the clarity of imaging, controversy regarding the management of basilar invagination continues. Even the natural history has not been clearly elucidated in the literature. The surgical indications for a given approach, together with the timing of the surgical stages, are still under discussion.

### Classification into Groups I and II

In 1997, we presented a classification system for basilar invagination that divided it into two discrete categories. This classification helped to improve understanding of the pathology and pathogenesis of the anomaly, selection of the surgical treatment, and prediction of the outcome.\(^1\) The analysis was based on a study of 190 cases of basilar invagination surgically treated in our department within a 10-year period (1988–1997). Based on a single criterion of the absence or presence of Chiari malformation, the anomaly was classified into group I or II, respectively.

Essentially, group I included cases where there was invagination of the odontoid process into the foramen magnum, causing the odontoid process to indent into the brainstem (Figs. 26.1 and 26.2). The tip of the odontoid process has invaginated into the brainstem.
odontoid process distanced itself from the anterior arch of the atlas or the inferior aspect of the clivus. The distancing of the odontoid process from the anterior arch suggested the presence of instability in the region and atlantoaxial dislocation. The angle of the clivus and the posterior cranial fossa volume were essentially unaffected in these cases. In group II, the assembly of the odontoid process, the anterior arch of the atlas, and the clivus migrated superiorly in unison, resulting in reduction of the posterior cranial fossa volume, which was the primary pathology in these cases. The Chiari malformation or herniation of the cerebellar tonsil was considered to be a result of reduction in the posterior cranial fossa volume (Figs. 26.3, 26.4, 26.5, and 26.6).

In 1998, we first defined the clinical implication of association of small volume of the posterior cranial fossa with Chiari malformation.¹ In our current review of cases with Chiari malformation, we identified a subgroup of patients in whom there was clear radiological evidence of instability of the region.
that was manifested by distancing of the odontoid process away from the anterior arch of the atlas, as well as radiological features matching those of group I cases. Considering this current evaluation, we have proposed a new classification for basilar invagination that is discussed in Chapter 27.

The following is a discussion of how these two groups were identified and how they affected the management of basilar invagination. Our opinion is that identification of these two groups is important, as they determine the pathology and direct the treatment planning.

### Diagnosis

#### Radiologic Criteria

#### Chamberlain Line

Basilar invagination was diagnosed when the tip of the odontoid process was at least 2 mm above the Chamberlain line. Measurements of the Chamberlain line on lateral sagittal reconstruction pictures of CT and sagittal MRI were seen to be reliable and accurate. The analysis of
basilar invagination in the two groups on the basis of the Chamberlain line suggested that the basilar invagination is much more severe in group II than in group I (Fig. 26.7).

### Distance between the Odontoid Tip and the Pontomedullary Junction

The distance of the tip of the odontoid from the pontomedullary junction, as observed on MRI, was seen to be a useful index to define the reduction in the size of the posterior cranial fossa bone. The distance was markedly reduced in group II cases, whereas it was relatively large in group I cases.

### Atlantodental or Clivodental Interval

In group I cases, it was seen that the odontoid process migrated superiorly and posteriorly into the foramen magnum and distanced itself from the anterior arch of the atlas and the inferior end of the clivus. As judged from the atlantodental or clivodental interval, there was an element of “fixed” atlantaxial dislocation in these cases. Actual mobility of the atlantoaxial joint on flexion and extension of the neck can be demonstrated only rarely. In group II, the alignment of the odontoid process with the anterior arch of the atlas and with the inferior aspect of the clivus remains normal.

### Wackenheim Clival Line

The tip of the odontoid process was significantly superior to the Wackenheim clival line in group I cases. In group II cases, the relationship of the tip of the odontoid process and the lower end of the clivus and the atlantodental and clivodental interval remained relatively normal. In a majority of cases, the tip of the odontoid process remained below the Wackenheim clival line and the McRae line of the foramen magnum. The basilar invagination thus resulted from the rostral positioning of the plane of the foramen magnum in relation to the brainstem (Fig. 26.7).

### Platybasia

The association of platybasia and basilar invagination is well known. In his initial study, Chamberlain referred to both of these entities as being synonymous. Subsequently, various authors have questioned the clinical significance of platybasia and have considered it to be of anthropological significance only. The superior position of the odontoid process was associated with a more horizontal angulation and shortening of the clivus. Klaus also identified two groups of basilar invagination on the basis of the clival line of Wackenheim. He noted that in basilar invagination associated with platybasia, the tip of the odontoid process almost never reaches the Wackenheim clival line, whereas in a steeply shelving or normal clivus, the line from the dens often reaches or even overshadows it. Platybasia was seen in both groups but was relatively less in number and severity in group I. From the study of group II cases, it appears that platybasia was as important as invagination of the odontoid process in causing the anterior concavity of the brainstem and in reducing the volume of the posterior fossa. Marin-Padilla concluded that the Chiari-like deformities reflect the effects of clival and occipital molding, which act mainly anteriorly. Platybasia did not directly result in any neurological symptoms, but it participated with basilar invagination in critically reducing the volume of the posterior cranial fossa.

### Posterior Cranial Fossa Volume

The Klaus height index, as measured on MRI, was seen to be much more accurate than the conventional measurements based on plain radiographs. The tentorium could be clearly identified on MRI, and the distance of the tip of the odontoid from the line of the tentorium indicated the height of the posterior cranial fossa. On the basis of the Klaus index, the posterior fossa height was found to be markedly reduced in group II cases, whereas it was only moderately affected in group I cases.

### Omega Angle

Although not frequently used, the omega angle or the angulation of the odontoid process from the vertical axis as described by Klaus was found to be a useful guide.
invagination was noted
Occipitalization of the atlas associated with basilar
the angle from the vertical axis that was a
described a modi
Fig. 26.8 Diagram showing the modi
À
ed omega angle. A line is drawn along the hard palate (line A, upper line). Another line is drawn parallel to this line that traverses through the mid-point of the base of the C2 body (line B, lower line). A line is drawn from the midpoint of the base of C2 along the tip of the odontoid process. The angle between this line and line B is the modified omega angle.
described a modified omega angle as the measurement of the angle from the vertical axis that was affected by flexion and extension of the neck. A line was drawn traversing through the center of the base of the axis parallel to the line of the hard palate. The line of the hard palate was unaffected by the relative movement of the head and the cervical spine in these “fixed” craniocervical anomalies (Fig. 26.8). Facial hypoplasia or hard palate abnormality was not seen in any case in this series and did not affect the measurements. The omega angle depicted the direction of displacement of the odontoid process. It was severely reduced in group I cases, whereas it was much larger in group II cases. The reduction in the omega angle in group I cases indicated that the odontoid process had tilted toward the horizontal and was posteriorly angulated in group I cases, whereas it was nearly vertical and superiorly migrated in group II cases.

Brainstem Girth

The effective brainstem girth measured on MRI was a useful additional parameter. Whereas the brainstem girth was markedly reduced in group I cases, it was only marginally affected or unaffected in group II cases, indicating that there was no direct brainstem compression as a result of the odontoid process in the latter group.

The anterior concavity of the brainstem was smooth in curvature in group II cases, whereas it was acute in group I cases, the angle being formed by the tip of the odontoid process. In group I, the brainstem distortion was directly a result of indentation of the odontoid process.

Occipitalization of the Atlas

Occipitalization of the atlas associated with basilar invagination was noted first by Rakitansky (cited by Grawitz) and has since been referred to frequently. Many authors have regarded assimilation as a characteristic feature of basilar invagination. The assimilation of the atlas can be partial or incomplete.

Association with Chiari Malformation

The association of Chiari malformation, syringomyelia, and basilar invagination has been the subject of various studies. Pillay et al. classified symptomatic Chiari malformation on the basis of the presence of syringomyelia. We observed that the more significant the basilar invagination and platybasia were in group II cases, the more the reduction was in the posterior cranial fossa volume, and consequently, the more marked was the tonsillar herniation. However, Chiari I malformation existed in all cases. The presence and the extent of the syrinx did not correlate with the severity of basilar invagination or Chiari malformation. In general, it appeared that syringomyelia will result when the craniovertebral bone anomaly is subtle and the symptoms are long-standing.

On the basis of the radiological study, it appeared that in group I, there was an invagination of the cervical spine into the base of the skull. The definition of basilar invagination, that is, prolapse of the spine into the base of the skull, as suggested by von Torklus and Gehle, was noted to be appropriate only for this group. In group II cases, the clivus was invaginated along with the cervical spine into the posterior cranial fossa. This probably resulted primarily in the reduction of volume of the posterior fossa and secondarily in the herniation of its neural contents into the foramen magnum. The basilar invagination, in all cases, resulted in an anterior indentation of the brainstem. An associated lateral or paramesial invagination and an isolated lateral invagination were not encountered. Lateral invagination is always associated with a marked torticollis of the head. In none of the cases was a “posterior” basilar invagination seen. Schmidt and Fisher recognized both an anterior and a medial type of basilar impression. In posterior basilar impression, according to Virchow, the posterior edge of the foramen magnum is tilted upward. Our current analysis suggests that the basilar invagination is always anterior. Frequently, the invagination has a tilt on one side, giving an impression that the basilar invagination is lateral. The bone deformation and torticollis are secondary phenomena and will essentially depend on the nature of the tilt of invagination.

Demographic and Clinical Criteria

Age Distribution

A significant majority of group I patients were adolescents. Group II patients were older at the time of presentation, with the average age being 27 years.
Sex Distribution

Male predominance has also been observed in some of the earlier described series. Klaus observed equal sex distribution. Levy et al. and the Mayo Clinic series found a female predominance in cases with Chiari malfunction.

Duration of Symptoms

The presentation was relatively acute in group I cases, whereas it was long-standing and slowly progressive in group II cases. In group II cases, the onset of symptoms and their evolution were insidious. Thus, a careful inquiry was needed to pinpoint the timing of the appearance of the initial symptoms. In the majority of cases, the time of onset was already forgotten. This made it difficult to establish with certainty the age at which the complaints began. The average duration of symptoms in the series of Chiari malformation in adults, as discussed by Levy et al., was 3.1 years.

Precipitating Factors

Trauma of varying severity was a noteworthy precipitating factor in group I cases. Trauma seldom plays any role in precipitating the symptoms in group II cases. The fact that trauma influenced the acute development of symptoms pointed toward an element of instability of the craniovertebral region in group I patients. Infections probably precipitate the events in a few patients in group I cases. Although long-term carrying of loads on the head as a precipitating factor has been recorded in some of the previous studies from India, it was not seen to be of any significance in our series.

The analysis of the symptoms that caused the patients to seek medical advice disclosed that in group I cases, the pyramidal symptoms formed a dominant component. Kinesthetic sensations were also affected in a large majority of cases. In group II cases, the spinothalamic sensory dysfunction and ataxia were as significant as the motor and deep sensory system affection. Similar observations were recorded in the large series of cases of basilar impression and Chiari malformation studied by de Barros et al. Neck pain and spasm of the muscles at the nape of the neck were frequent findings in group I patients. The pain usually was constant, but in most instances, it became worse on movement of the neck. This feature suggested strain on the muscles secondary to subtle instability of the region. Dysfunction of the 9th to 12th cranial nerves was present in ~10% of cases in both groups.

Associated Clinical Features

Mere inspection of the patients with basilar invagination was of diagnostic value in the majority of cases in both groups (Figs. 26.9, 26.10, and 26.11).

Fig. 26.9a, b
a Photo of a 2-year-old boy having a short neck and basilar invagination.

b Profile of the same boy.
The symptoms and signs in group II basilar invagination appeared to be directly related to the “crowding” of neural structures at the foramen magnum. Although the dimensions of the foramen magnum were large and sometimes larger than even in a normal state, the volume of its contents and probably the “pulsatile” compression of the structures at the foramen magnum resulted in neurological symptoms. The markedly reduced girth of the brainstem in group I cases clearly showed that direct compression of the brainstem by the odontoid process caused the neurological symptoms. Central cord symptoms and related signs were noted in cases associated with syringomyelia.

**Treatment**

**Effect of Cervical Traction**

Following traction, the Chamberlain line, the omega angle, and the craniocervical angulation reduced in group I cases, indicating the reason for clinical improvement. The clinical improvement following traction was an important guide to the prognosis. It suggested the presence of relative instability of the craniovertebral region in group I cases and stability in group II cases. We have earlier suggested that transoral odontoidectomy in such group I cases would result in clinical recovery. However, as per our current understanding, we now advocate distraction and fixation of the atlantoaxial facets in an attempt
to realign and stabilize the craniovertebral junction in group I basilar invagination. Such a variation in the treatment will obviate the need for transoral or posterior bony decompression. This issue is the basis of our modified classification for the entity of basilar invagination and has been discussed elsewhere (see Chapter 27).

**Surgery**

Considerable debate exists about which surgical options are the best for the management of basilar invagination. The treatment of basilar invagination in the presence of Chiari II malformation has not been clearly outlined and is confused by the fact that some authors are currently recommending anterior surgery for even this type. It appears from the analysis of the results in our series that patients in group II benefited by foramen magnum bony decompression. The procedure resulted in amelioration of symptoms and at least an arrest in the progression of the disability. None of the patients in this group had a delayed worsening in the neurological condition following foramen magnum decompression. Driesen reported that during operations for craniovertebral anomalies, he often had to remove noticeably thickened pieces of bone from the posterior edge of the foramen magnum. In our series, the suboccipital bone and posterior rim of the foramen magnum and the dura overlying the herniated cerebellar tissue were thin in a significant number of cases. This probably was related to the chronic pressure changes secondary to the reduced posterior cranial fossa volume. The bulbous lipping of the posterior rim of the foramen magnum represents the rudiments of the posterior arch of the atlas assimilated into the occipital bone. Various authors have recommended that to achieve maximal decompression, it is necessary to open the dura mater and to cut all constrictive dural and arachnoidal bands. Some authors have recommended leaving the dura open, whereas others have recommended the placement of a graft. More recent articles do not recommend resection of the herniating tonsils or even sectioning of adhesions around it. Our opinion is that dural incision and opening are not necessary in group II cases following bony decompression of the foramen magnum. We are convinced that the dura is an expansile structure and can never be a compressive factor. Opening of the dura not only is unnecessary but also subjects the patient to an increased risk of cerebrospinal fluid fistula. It turns an otherwise simple operation into a relatively complex and dangerous surgical maneuver. The fact that dural opening was not necessary while performing posterior fossa or foramen magnum decompression was first described by Goel et al. in 1998.

The treatment of a syrinx in the presence of a Chiari malformation is also controversial. Logue and Edwards reported 75 patients treated with craniovertebral decompressions for Chiari malformations and syringomyelia. Patients were divided into two groups: one treated with decompression only, leaving the arachnoid intact, and one treated with Gardner’s procedure of opening the fourth ventricle and plugging the upper cervical canal. The authors concluded that muscle plugging did not seem to change the results. Levy et al. agreed along the same lines. Logue and Edwards noticed that there was no significant need for performing a syringosubarachnoid shunt following craniovertebral decompression. Subsequent studies have questioned the need for syrinx drainage surgery following foramen magnum decompression. Driesen reported that muscle plugging did not seem to be removed while performing a syringosubarachnoid shunt following craniovertebral decompression. Driesen reported that muscle plugging did not seem to be a necessary procedure in cases with syringomyelia. Transoral odontoidectomy with resection of the superior half or third of the C2 body was a gratifying surgical procedure in group I patients. Following this surgery, there was clinical recovery in all patients in this group. However, the long-term clinical outcome following the twin operation of craniovertebral decompression followed by posterior stabilization was seen to be inferior to the clinical outcome following our current operation that involves craniovertebral realignment without any bone, dural, or neural decompression. In group I cases, the odontoid process was low-lying although posteriorly angulated. Surgery was helped by cervical traction, as it reduced the posterior angulation and helped pull the odontoid process inferiorly.

The analysis of the pathology, the surgical experience, and the results suggest that anterior transoral decompressive surgery is relevant in group I patients (Fig. 26.11), although we currently recommend craniovertebral realignment in these cases. Foramen magnum decompression was appropriate in group II patients. Some authors are of the opinion that lesions anterior to the neural axis are best treated with an anterior surgical approach. Proponents of anterior surgery for anterior compression have advocated anterior transoral surgery even for group II cases. In a significant proportion of group II patients in this series, the smooth anterior indentation of the brainstem was caused not only by the high odontoid process, but also by the dorsally and horizontally positioned clivus. If the aim of surgery is to obliterate the anterior indentation in these patients, a large part of the clivus also has to be removed. In our group II cases, where transoral surgery was performed, the surgical aim was resection of the odontoid process and part of the body of the axis. The worsening neurological deficits following transoral surgery in group II patients were related to the markedly difficult surgical procedure. The surgery in these cases was more difficult when compared with group I cases, due to the high position of the odontoid and the inability to pull the odontoid process caudally by cervical traction. In the majority of patients, a part of the hard palate had to be removed to achieve exposure. Some authors have recognized the difficulties involved in the transoral surgical procedure in such cases and have recommended an additional maxillotomy to approach the high odontoid process. Additional removal of the clival bone, which,
in our opinion, appears to be necessary to achieve complete obliteration of the anterior indentation of the brainstem, can be an extremely formidable operation, and the validity of such an endeavor can be argued. Although transoral surgery has improved remarkably over the past 2 decades, the potential complications related to it can be life threatening. The cause of the symptoms appears to be crowding of the neural structures at the level of the foramen magnum, and the surgery should be directed toward decompression of the bone components of the foramen magnum and not toward obliteration of the anterior curvature of the brainstem. Logue and Edwards concluded from their study that Chiari malformation was of paramount importance in producing symptoms in patients with basilar invagination, inasmuch as patients with the two diseases respond to decompression of the cerebellar tonsils even when medullary angulation goes uncorrected. Dickman et al. had to perform posterior decompression surgery in each of their four cases after an anterior decompression due to persistence of compressive symptoms in cases of basilar invagination associated with Chiari malformation. Our impression is that posterior foramen magnum decompression is the optimum operation in group II basilar invagination, and it results in enduring clinical improvement and stabilization of symptoms.

## Conclusion

The division of basilar invagination on the basis of the presence or absence of a Chiari malformation is embryologically, pathologically, and clinically relevant. The symptoms in group I cases are related to brainstem invagination by the odontoid process. In group II cases, although the brainstem is posteriorly displaced, it is not indented by the odontoid process. The principal cause of neurological symptoms in group II is the crowding of neurological structures in the foramen magnum. Transoral surgery is most appropriate in group I cases. Posterior atlantoaxial or occipitocervical fixation is mandatory after transoral decompression. Posterior foramen magnum decompression is adequate in group II cases. Although foramen magnum decompression can be done by an anterior route as well, the operation is more tedious, and the decompression of the neurological structures may not be as complete as by a posterior route. The anomaly is of a “fixed” variety in a majority of cases due to lateral fusion or malformation of the joints. These patients can safely be observed after either anterior or posterior decompression surgery.

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**Fig. 26.12a, b**

a  Lateral radiograph showing group I basilar invagination. There is partial occipitalization of the atlas and “fixed” atlantoaxial dislocation.

b  Occipitocervical fixation using a metal loop after transoral odontoidectomy.
Fig. 26.13a–e
a  T1-weighted MRI showing basilar invagination.
b  T2-weighted MRI.
c  MRI showing a marked torticollis.
d  T2-weighted MRI showing a torticollis.
e  Posterior fixation and craniovertebral realignment using the plate and screw technique.
Table 26.1  Salient distinguishing features between groups I and II

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<tr>
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<th>Group I</th>
<th>Group II</th>
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<tr>
<td>Odontoid–atlas relation</td>
<td>Distance increased, odontoid angled posteriorly and indentering into brainstem</td>
<td>Odontoid–atlas alignment not changed, odontoid superiorly directed and not directly indentering into brainstem</td>
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<tr>
<td>Posterior fossa volume</td>
<td>Marginally reduced</td>
<td>Significantly reduced and main pathology</td>
</tr>
<tr>
<td>Syringomyelia</td>
<td>Rare</td>
<td>Frequent</td>
</tr>
<tr>
<td>Chiari I malformation</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Basilar invagination</td>
<td>Moderate</td>
<td>Severe</td>
</tr>
<tr>
<td>Surgical approach</td>
<td>Transoral decompression followed by posterior fixation or craniocervical junction realignment surgery</td>
<td>Posterior decompression of the foramen magnum</td>
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In Chapter 26, we discussed the subject of basilar invagination and presented a classification for these anomalies based on the presence or absence of Chiari I malformation. On the basis of the possible pathogenetic factors, we suggested a specific treatment protocol for each of the two groups. With our improved understanding of the subject, we reclassified basilar invagination into two groups (groups A and B) based on parameters that determined an alternative treatment strategy. In group A, there is a “fixed” atlantoaxial dislocation, and the tip of the odontoid process invaginates into the foramen magnum and above the Chamberlain line, the McRae line of the foramen magnum, and the Wackenheim clival line. The definition of basilar invagination, or prolapse of the cervical spine into the base of the skull, as suggested by von Torklus and Gehle, is suitable for this group of patients. In group B, alignment of the odontoid process and clivus remains normal despite the presence of basilar invagination and other associated anomalies. In this group, the tip of the odontoid process is above the Chamberlain line but below the McRae and Wackenheim lines. Radiological findings suggest that the odontoid process in group A patients results in direct compression of the brainstem. Analysis on the basis of the Chamberlain line and on the distance from the odontoid tip to the pontomedullary junction shows that basilar invagination is only mild or moderate in these cases. Modified omega angle measurements suggest that the odontoid process tilts horizontally rather than rostrally. Despite the clinical evidence of instability of the craniovertebral joint, abnormal atlantoaxial mobility can be identified on dynamic radiology only in a minority of cases. Essentially, in group A basilar invagination, there is an element of instability of the region that is manifested by the tip of the odontoid process distancing itself from the anterior arch of the atlas or the lower end of the clivus. Some group A patients have a Chiari malformation, and this feature differentiates the present classification from the earlier classification. In this group, the atlantoaxial joints are “active,” and their orientation is oblique, as shown in Fig. 27.2a, instead of the normal horizontal orientation. We have found similarities in C1–C2 facets with spondylolisthesis seen in the subaxial spine (Fig. 27.2b). It appears to us that the atlantoaxial joint in such cases is in an abnormal position as a result of a mechanical problem rather than...
A congenital abnormality of the bones, and progressive worsening of the dislocation is probably secondary to increasing “slippage” of the facets of the atlas over the facets of the axis. The slip of the atlas over the axis appears to be accentuated by trauma. With our experience in handling atlantoaxial joints, we have realized that the joint in these cases is not “fixed” or “fused” but is mobile, and in some cases hypermobile, and is probably the prime cause for basilar invagination. The history of trauma preceding the clinical events, the predominant complaint of pain in the neck, and the improvement in neurological symptoms following institution of cervical traction suggest “vertical” instability of the craniovertebral region. In group B, the atlantoaxial joints are either fused or normally aligned (Fig. 27.3).

**Fig. 27.2a, b**
- **a** CT scan showing spondylolisthesis of the C1 facet over the C2 facet. This is probably the cause of basilar invagination.
- **b** Illustration showing L5 over S1 spondylolisthesis.

**Fig. 27.3a, b**
- **a** T2-weighted MRI showing invagination of the odontoid along with the clivus. The alignment of the odontoid tip with the clivus is not changed.
- **b** T1-weighted MRI showing group B basilar invagination.
We recently studied the treatment of group A basilar invagination and discussed the feasibility of manipulation and distraction of the facets of the atlas and axis, as well as reduction of the basilar invagination and fixation of the atlantoaxial joint.7–10 Our current experience with the technique in over 200 cases makes us convinced that distraction and direct lateral mass fixation of the atlantoaxial joint is the ideal form of treatment in group A basilar invagination, and transoral surgery can be avoided entirely. The technique results in realignment of the facets into a horizontal position and in realignment of the entire craniovertebral junction (Fig. 27.4).

In our series, the majority of patients with group A basilar invagination (58%) had a history of minor to major head injury prior to the onset of the symptoms. The pyramidal symptoms formed a dominant component. Kinesthetic sensations were affected in 55% of cases. Spinthalamic dysfunction was less frequent (36%). Neck pain as a major presenting symptom was seen in 77% of cases. Torticollis was present in 41% of cases. The analysis of radiological and clinical features suggests that the symptoms and signs were a result of brainstem compression by the odontoid process.

Surgical Experience and Technique

All patients in our series underwent joint manipulation surgery (Figs. 27.5, 27.6, 27.7, 27.8, 27.9, and 27.10). No patient underwent a transoral decompression as a first stage operation. The basic surgical steps of the joint manipulation surgery are the same as discussed in our papers on lateral mass plate and screw fixation of the atlantoaxial joint. The exposure of the atlantoaxial joint in cases with basilar invagination is significantly more difficult and technically challenging when compared with a normally aligned atlantoaxial joint encountered during the treatment of posttraumatic instability. The joint is significantly rostral in location and the microscope needs to be appropriately angled. Due to bony abnormalities of the region and frequently encountered rotation, the orientation can easily be lost. Cervical traction is given prior to induction of anesthesia and the weights are progressively increased to approximately one fifth of the total body weight. The patient is placed prone, with the head end of the table elevated to ~35°. Use of neuronavigation assistance facilitated the dissection and added safety to screw implantation.

Text continues on page 284
Fig. 27.5a–f

a  Photo of a wheelchair-bound 15-year-old boy. The patient’s neck is remarkably reduced in size.
b  Axial CT scan showing the occipital bone riding up to the level of the petrous bone.
c  Preoperative T1-weighted MRI showing basilar invagination, fixed atlantoaxial dislocation, and indentation of the brainstem.
d  Preoperative CT scan showing marked basilar invagination and fixed atlantoaxial dislocation.
**Fig. 27.5e–f**
e Postoperative CT scan showing reduction of the basilar invagination and atlantoaxial dislocation. Note the change in the bony alignments of the craniovertebral junction. The alteration in the relationship of the anterior arch of the atlas and the clivus to the C2 body and odontoid process can be appreciated. Metal artifacts can be observed.

**Fig. 27.6a, b**
a Preoperative CT scan showing the C2 body and odontoid and its relationship with the anterior arch.

b Postoperative scan after distraction surgery. The alignment of the craniovertebral junction is now nearly normal.

d Picture taken 4 months after surgery. The child can now stand with support.
Fig. 27.7a–d

a CT scan of a 16-year-old boy showing basilar invagination and fixed atlantoaxial dislocation.

b CT scan showing the oblique alignment of the atlantoaxial joint.

c Postoperative CT scan showing the changes in the alignment of the craniovertebral region. Note the changes in the relationship of the odontoid process with the clivus.

d Postoperative CT scan through the atlantoaxial joint. Note the partial realignment of the facets and the spacer.
**Fig. 27.8a, b**
a Preoperative CT scan showing marked basilar invagination.
b Postoperative CT scan showing significant realignment.

**Fig. 27.9a–d**
a Preoperative CT scan showing basilar invagination, fixed atlantoaxial dislocation, and assimilation of the atlas.
b Postoperative CT scan showing realignment of the craniovertebral junction.
c CT scan showing oblique angulation of the facets of the atlas and axis.
d Postoperative CT scan showing realignment of the facets, fixation with plates and screws, and distraction with the help of spacers.
The atlantoaxial facet joints are widely exposed on both sides after sectioning of the large C2 ganglion. The exposure of the facet of the atlas was significantly difficult as in several patients there was an assimilation of the atlas resulting in a rostrally located C1 facet. The joint capsule is excised and the articular cartilage is widely removed using microdrill. The joint on both sides is distracted using an osteotome. The flat edge of the osteotome is introduced into the joint and it is then turned vertical to effect distraction. The status of the dislocation and of basilar invagination is evaluated by intraoperative radiographic control. Corticocancellous bone graft harvested from the iliac crest is stuffed into the joint in small pieces. Specially designed titanium spacer or hydroxyapatite blocks are used in selected cases as strut graft and stuffed into the joints to provide additional distraction and stability. Plate and screw fixation of the region is subsequently performed by the interarticular technique. A two-holed stainless steel plate is used measuring 15 to 20 mm in length. The screws are 2.4 to 2.8 mm in diameter and measure 20 to 26 mm in length. Screws are passed bilaterally through the holes in the plate into the lateral mass of the atlas and axis (Figs. 27.5, 27.6, 27.7, 27.8, and 27.9). In cases where lateral mass plating cannot be completed because of anatomical or surgical/technical limitations, a C1–C2 transarticular screw fixation method described by Grob and Magerl\textsuperscript{15} or an occipitoaxial fixation\textsuperscript{11,14} can be done. The point of entry and the direction of the screw for the transarticular fixation are altered to suit the complex local anatomy in these cases. Additional bone graft is placed between the posterior elements of C1 and C2 after decorticating the host bone area with a burr. Postoperatively, the traction is discontinued, and the patient is placed in a four-post hard cervical collar for 3 months, with all physical activities involving the neck restrained during this period.

### Special Surgical Issues

#### Venous Bleeding

Venous bleeding in the lateral gutter can be a formidable challenge. It is generally more profuse in cases with basilar invagination. Judicious packing with Surgicel or Gelfoam and relatively quick surgical dissection in the region can limit blood loss during dissection.
Vertebral Artery Management

The presence of occipitalization of the atlas can lead to an anomalous course of the vertebral artery over the posterior surface of the facet of the atlas, leading to considerable difficulty in dissection. Preoperative angiographic studies are important to understand the course of the vertebral artery in relationship to the bone in the region. During surgery, it is important to identify the C2 pedicle and avoid any dissection lateral to it. The dissection and subsequent resection of the C2 ganglion should be under direct vision, as the vertebral artery can sometimes traverse parallel to the course of the ganglion or under the posterior arch of the atlas. It is not necessary to actually dissect and isolate the vertebral artery during surgery. Identifying the probable course of the artery and avoiding the region are generally sufficient. However, in some situations, dissection, isolation, and manipulation of the vertebral artery may be necessary to expose the region for instrumentation. In cases where there is injury to the vertebral artery during dissection, control of bleeding can become difficult. Temporary clip application and direct suturing of the artery can be attempted. However, in cases where such suturing is not possible, coagulation and sacrifice of the artery appear to be a safe option. Venous bleeding in the vicinity makes any kind of direct vertebral artery handling difficult. If the suspicion of vertebral artery injury is there during the screw insertion procedure, the bleeding should be stopped with bone wax, and an alternative site for insertion can be selected. However, application and screw insertion can be completed through the same hole. Appropriate drilling of the bones hindering the exposure can provide a suitable view to the region.

Results of Surgery

Clinical neurological improvement varies, but in most cases, it is remarkable and satisfying. All of our patients had sustained neurological improvement to varying degrees, suggesting the effectiveness of the operation. Following surgery, the alignment of the odontoid process and the clivus and the entire craniovertebral junction improved. The tip of the odontoid process receded in relationship to the Wackenheim clival line, Chamberlain line, and McRae line, suggesting reduction in basilar invagination. The posterior tilt of the odontoid process, as evaluated by the modified omega angle, was reduced after the surgery. We could obtain varying degrees of reduction of basilar invagination and atlantoaxial dislocation. The extent of distraction of the joint and the subsequent reduction in basilar invagination were more significant in younger than in older patients. In addition to the odontoid process changing its angulation, the angulation of the clivus became more normal, suggesting that the entire craniovertebral junction was realigned by the procedure.

The extent of neurological improvement observed by us after using the technique described here appeared to be far more satisfactory and sustained when compared with the improvement obtained after transoral surgery and subsequent occipitocervical fixation. Despite the complex and deformed nature of anomalies encountered in the series, there were no postoperative vascular, neurological, or infective complications. In three patients, the reduction of the basilar invagination was not entirely satisfactory, and delayed transoral surgery was performed to decompress the region (Fig. 27.11). No patient needed reexploration for failure of fixation of the implant. Immediate postoperative and follow-up radiographs confirmed fixation and fusion, along with reduction of basilar invagination. Torticollis improved significantly following surgery in all patients. No patient complained of numbness in the suboccipital region, but on a leading question acknowledged having a patch of suboccipital numbness.

![Fig. 27.11a–d](image1)

- Preoperative MRI showing marked basilar invagination.

![Fig. 27.11c–d](image2)

- Preoperative CT scan showing marked basilar invagination.
Craniovertebral Realignment for Group A Basilar Invagination Associated with Syringomyelia

In one study, we classified syringomyelia and suggested a specific treatment protocol on the basis of possible pathogenetic factors. We suggested that syringomyelia is a tertiary response to a primary craniovertebral anomaly in the form of basilar invagination that leads to secondary Chiari I malformation as a result of reduction in the volume of the posterior cranial fossa. Accordingly, posterior fossa bony decompression was considered optimum in the treatment of this subgroup of patients.

We identified cases of syringomyelia in which there are associated bony abnormalities of the craniovertebral region that include “fixed” atlantoaxial dislocation or those having group A basilar invagination. This select group of patients were treated by reduction of the atlantoaxial dislocation and basilar invagination and by direct lateral mass plate and screw atlantoaxial fixation. No bony or dural decompression or neural manipulation of any kind was done in these cases.

The majority of patients with Chiari malformation-related syringomyelia without any bony anomaly of the craniovertebral region have hyporeflexia of the upper extremities and spastic lower extremities. The presence of spastic quadriplegia in all of our patients in this study suggests that the symptoms were related to the compression of the brainstem from the invaginated dens rather than due to syringomyelia. It was observed that the patients were relatively young, neck pain formed a part of the symptom complex, and the motor symptoms and ataxia were far more prominent in cases having the complex of malformation that included group A basilar invagination, Chiari I malformation, and syringomyelia than in cases with a similar complex but without bony anomalies of the craniovertebral region. It appears that when the angulation of the facets is not as acute or is only marginally affected, the progress of basilar invagination is slow and over several years, providing an opportunity for a syrinx to develop.

It was observed that in cases of syringomyelia where there was “fixed” atlantoaxial dislocation with or without the association of group A basilar invagination and Chiari malformation, an attempt could be made to realign the bones in the craniovertebral junction. As observed by us earlier, it appears that the atlantoaxial joint in such cases is in an abnormal position as a result of congenital abnormality of the bones and that progressive worsening of the dislocation is probably secondary to increasing slippage of the atlas over the axis. The fact that there was remarkable clinical improvement following the reduction of the atlantoaxial dislocation and basilar invagination indicates that the complex of atlantoaxial dislocation, basilar invagination, and syringomyelia is probably secondary to craniovertebral instability. Group A basilar invagination was less severe in cases with associated syringomyelia than in cases where there was no syringomyelia. The conduct of surgery and joint manipulation was relatively easier in these cases.

Following surgery, the alignment of the odontoid process and clivus, as well as the entire craniovertebral junction, improved. We could obtain different degrees of reduction of basilar invagination and atlantoaxial dislocation. The atlantoaxial alignments became more normal, and the tip of the odontoid process receded in relationship to the Wackenheim clival line and Chamberlain line, sug-
gesting reduction in basilar invagination and atlantoaxial dislocation. The posterior tilt of the odontoid process, as evaluated by the modified omega angle, was reduced after surgery. All patients exhibited sustained neurological improvement to varying degrees, suggesting the effectiveness of the operation. Stainless steel plates, nonlocking screws, and custom-made spacers were used due to the higher costs of branded material. Because of the type of metal used in the procedure, the effect on syringomyelia could not be confirmed (Figs. 27.12 and 27.13).

**Fig. 27.12a–g**

a Preoperative radiograph showing basilar invagination and fixed atlantoaxial dislocation.

b Preoperative MRI in a 36-year-old man showing severe basilar invagination, fixed atlantoaxial dislocation, and Chiari I malformation. Posterior decompression was done at an earlier stage.

c CT scan showing basilar invagination and fixed atlantoaxial dislocation. C2–C3 fusion can be seen.

d A lateral sagittal cut through the joint showing the dislocation.

e Postoperative CT scan showing reduction of basilar invagination and atlantoaxial dislocation.
Fig. 27.13a–f

a  T1-weighted MRI showing basilar invagination, Chiari I malformation, and syringomyelia.

b  T2-weighted MRI.

g  Postoperative radiograph showing plate and screw fixation. A metal spacer can be seen.

f  CT scan showing plate and screw fixation, with the distracted joint harboring a metal spacer. Note the reduction of the dislocation.
Treatment of Torticollis

Torticollis is a common symptom in all forms of basilar invagination. No specific treatment modality has been described to treat this symptom. Transoral decompression or a posterior fixation procedure can provide decompression of the region and fixation, but torticollis remains untreated. We observed that reduction of the atlantoaxial dislocation and of basilar invagination by manual distraction of the atlantoaxial joint presents an ideal opportunity for reduction of torticollis (Fig. 27.14). Although differential distraction of the joints was not attempted in our patients, this procedure may provide additional reduction of torticollis.

Progressive Basilar Invagination after Transoral Odontoidectomy

Worsening basilar invagination as a cause of failure of transoral odontoidectomy has been reported.17–19 Such worsening has been seen even when a posterior fixation was performed (Fig. 27.15).18 Previous investigators related the progressive instability to the transoral surgical procedure, damage to or manipulation of the bones, and disruption of transverse and both alar ligaments.17–19 Dickman et al. reported increased anteroposterior translation of the atlas as well as increased total angular range of motion after odontoidectomy.17 They identified that in cases in which the anterior arch of the atlas was
Fig. 27.14a–e
a T2-weighted MRI showing basilar invagination and cord compression.
b Coronal MRI scan showing marked torticollis.
c Postoperative radiograph showing fixation using plate and screws and distraction of the facets using a spacer.
d Photo of the boy prior to surgery showing marked torticollis.
e Photo of the boy following surgery showing significant reduction of torticollis.
sectioned, horizontal separation of the lateral masses of C1 occurred. On this basis, they suggested preservation of the anterior arch of the atlas during odontoidectomy. Naderi and Pamir also believed that the integrity of the C1 contributes to the stabilization of the craniovertebral junction after odontoidectomy. Spetzler et al. suggested that a partial resection of the anterior arch might minimize the progressive adverse effect of odontoidectomy. Sawin and Menezes suggested prolonged external orthotic immobilization with a Minerva brace and observed symptomatic improvement and arrest in the progression of the deformity.

In our experience, we have found that the atlantoaxial joint in these cases is not “fixed” or “fused” but is mobile, and in some cases is hypermobile, and is probably the prime cause of basilar invagination. The history of trauma preceding the clinical events, the predominant complaint of pain in the neck, and the improvement in neurological symptoms following the institution of cervical traction suggest vertical instability of the craniovertebral region. Transarticular or interarticular methods of posterior fixation that directly stabilize the joint appear to stall the progression of the disease.

Our analysis reaffirms the view that in select cases of basilar invagination associated with fixed atlantoaxial dislocation, the process is progressive in nature and is probably a result of an unusual inclination of the facets of the atlas and axis. Posterior fixation appears to be mandatory after transoral decompression in such cases. Distraction of the facets and direct articular joint atlantoaxial fixation in a distracted position may be an ideal method of treatment of the atlantoaxial dislocation and of basilar invagination.
Conclusion

Our results in the treatment of group A basilar invagination are promising and encourage us to undertake further study on the subject. The procedure described here is technically demanding and anatomically precise, but if it is learned adequately and performed successfully, the neurological outcome is extremely gratifying.

Although our experience with the described technique in patients with syringomyelia is limited, the results suggest that realignment and direct fixation of the atlantoaxial joint may have a place in the treatment of select cases of syringomyelia.

References

We identified a select group of patients having group I basilar invagination in which there was complete reduction of basilar invagination on extension of the head without the need of any cervical traction. We labeled this group of patients as having “vertical mobile and reducible” atlantoaxial dislocation. Considering that the imaging characteristics with the head in a flexed position are of group I basilar invagination, the term mobile and reducible basilar invagination can also be used to classify the clinical condition. It is critical to differentiate this group of patients from other group I basilar invagination cases that have “fixed” atlantoaxial dislocation, as the treatment of the two clinical entities is discrete.

### Clinical Features

Based on our understanding of the presence of mobile and reducible vertical dislocation, as well as the use of dynamic computed tomography (CT), we identified 8 out of 64 patients with group I basilar invagination seen between January 2006 and March 2008 as having vertical mobile and reducible dislocation. Pain at the nape of the neck, spasm of the neck muscles, and a short neck were the main symptoms. Patients ranged in age from 8 to 54 years (mean 24 years). There were five male and three female patients. A short neck and torticollis were clinical findings in all patients and were present since early childhood. In six patients, there was a history of relatively moderate degree of trauma at the time of onset of major neurological symptoms. Torticollis exaggerated after the injury in all cases. The history of trauma ranged from 15 days to 4 years (mean 6 months) prior to diagnosis and treatment. All patients had different degrees of neck pain, neck muscle spasm, and spastic quadriparesis. Sensory symptoms were relatively mild and predominantly included bilateral upper and lower extremity paresthesias and kinesthetic sensation deficits.

### Radiological Features

The Wackenheim clival line and McRae line of the foramen magnum were used to evaluate the basilar invagination or the superior migration of the tip of the odontoid process in relationship to the clivus or to the anterior arch of the atlas. To grade the degree of vertical dislocation, we used our previously described vertical atlantoaxial instability index. Most patients were adolescent or middle-aged. The dislocation was generally more severe in younger patients. Because the compression of the cervicomedullary cord is not as acute as is seen in horizontal atlantoaxial dislocation, the symptoms are relatively mild and long-standing.

### Vertical Instability

Vertical instability of the atlantoaxial joint has been identified and more often linked with cranial settling associated with rheumatoid arthritis. It has also been linked by some authors to basilar invagination secondary to congenital anomaly in the region. Reduction of basilar invagination on institution of cervical traction has been observed by us and by others. Such a reduction suggests the presence of vertical instability. We had earlier identified a relatively inclined profile of the facets of the atlas and axis to be the primary cause of basilar invagination. Progressive “slip” of the facet of C2 over C1 could be the cause of spondylolisthesis, a phenomenon that eventually results in basilar invagination. In all cases with vertical mobile and reducible dislocation, the atlantoaxial joint was markedly inclined, an anomaly that appeared to be primarily responsible for incompetence of the lateral masses and abnormal vertical mobility. Although several authors consider congenital basilar invagination as a “fixed” anomaly, others have identified it as the result of vertical craniocervical instability. Essentially, it appears that cases with group I basilar invagination can be subclassified as having either fixed atlantoaxial dislocation (where the basilar invagination does not disappear on neck movements) or vertical mobile and reducible atlantoaxial dislocation (where the basilar invagination disappears on extension of the neck).

### Treatment

The more accepted method of treatment of group I basilar invagination with fixed atlantoaxial dislocation is transoral decompression of the odontoid process and subsequent
craniovertebral stabilization. For this group of patients, we advocate atlantoaxial joint distraction (with or without the assistance of metal spacers), reduction of the dislocation, and subsequent fixation. However, in group I basilar invagination with vertical mobile and reducible atlantoaxial dislocation, fixation of the atlas and axis in the reduced position suffices, and distraction of the facets is not necessary (Figs. 28.1, 28.2, 28.3, 28.4, and 28.5). A treatment algorithm for cases with basilar invagination is given in Fig. 28.6.

We used our plate and screw fixation technique to stabilize vertical atlantoaxial dislocation. On direct intraoperative observation, it was clear that the joint was active and mobile and that the articular surfaces were functional. The articular surfaces were drilled and denuded of the articular cartilage, and iliac crest bone graft was introduced into the joint cavity to effect bone fusion. Immediate postoperative and long-term results following successful stabilization were extremely gratifying.

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Fig. 28.1a–f  Investigations in a 54-year-old woman.
a  Computed tomography (CT) sagittal reconstruction with the head in flexion shows vertical dislocation (or basilar invagination).
b  CT scan with the head in extension documents reduction of the dislocation.
c  Radiograph of the craniovertebral junction with the head in flexion shows evidence of basilar invagination and atlantoaxial dislocation.
d  Radiograph with the head in extension reveals reduction of the dislocation.
Fig. 28.1e–f

**e** Postoperative CT scan shows fixation of the atlantoaxial dislocation in a reduced position.

**f** Postoperative radiograph shows fixation with plate and screws.

Fig. 28.2a–f Images of an 8-year-old girl.

**a** T2-weighted magnetic resonance imaging (MRI) showing extensive basilar invagination and cervicomedullary cord compression.

**b** CT reconstruction with the head in a flexed position revealing severe basilar invagination and vertical atlantoaxial dislocation. The Wackenheim clival line (A–B) and the McRae line of the foramen magnum (C–D) show the relationship with the odontoid process.
Fig. 28.2c–f

c CT scan with the head in extension showing reduced dislocation. Note the altered relationship of the odontoid process with the McRae and Wackenheim lines.
d CT scan with the sagittal cut passing through the atlantoaxial joint. An abnormal oblique orientation of the joint can be appreciated.
e Postoperative CT scan showing fixation of the dislocation using the lateral mass plate and screw fixation method. Reduction of the vertical dislocation can be appreciated.
f Postoperative sagittal CT scan showing plate and screw fixation and realignment of the joint.
Fig. 28.3a–d Images of a 12-year-old boy.

a CT scan (sagittal image) with the head in flexion revealing basilar invagination.

b CT scan with the head in extension showing reduction of basilar invagination.

c CT scan with the cut traversing through the joint. An abnormal configuration of the joint can be appreciated.

d Postoperative scan showing fixation in a partially reduced position.
Fig. 28.4a–d  Investigations in a 21-year-old woman.
a  CT scan with the head in flexion showing vertical atlantoaxial dislocation.
b  CT scan with the head in extension documenting complete reduction of the dislocation.
c  Postoperative CT scan showing fixation in the reduced position.
d  Postoperative CT scan showing plate and screw fixation of the facets.
Fig. 28.5a–g Investigations in a 22-year-old woman.

a CT scan with the head in flexion revealing vertical atlantoaxial dislocation.

b CT scan with the head in extension showing complete reduction of the dislocation.

c CT scan with a sagittal cut through the facets showing an inclined alignment of the facets.

d T1-weighted MRI showing the vertical atlantoaxial dislocation (basilar invagination).

e T2-weighted MRI documenting the dislocation.

f Postoperative CT scan showing reduction of the dislocation.

g Sagittal image through the facets showing plate and screw fixation.
**Conclusion**

Considering the number of cases seen by us in a relatively short period of time, it appears that vertical mobile and reducible atlantoaxial dislocation is a discrete clinical entity. The movements of the odontoid process in a vertical plane result in indentation of the cervicomedullary cord and related symptoms. From direct observation, it appears that all cases with basilar invagination should undergo dynamic flexion-extension computerized imaging to determine the presence or absence of vertical instability and reducibility. Stabilization and fixation in reduced and anatomical positions result in a cure.

**References**

Musculoskeletal Changes in Basilar Invagination

Atul Goel and Abhidha Shah

Several abnormalities of the neck and spine are associated with basilar invagination. Short neck, low hairline, web-shaped neck muscles, torticollis, reduction in the range of neck movements, and other physical variations have been described as hallmarks of basilar invagination. Several bone fusions and deformities and platybasia have also been recorded. Neck pain, muscle spasms, and restriction of neck movements are frequently recognized and suggest potential instability of the region.

In 1912, Maurice Klippel and André Feil discussed a triad of observations of extensive cervical vertebral fusions in a 46-year-old patient with a short neck, low hairline, and restricted neck movements. This combination of symptoms, labeled the Klippel-Feil triad, is commonly used synonymously with basilar invagination. Several authors have commented on the importance of external appearance in identifying anomalies of the craniovertebral junction. The general understanding is that musculoskeletal features, such as a short neck and bony variations, are the result of embryonic dysgenesis and are the primary abnormalities that lead to odontoid compression on the craniocervical cord.

We have speculated that basilar invagination is secondary to abnormally inclined alignment of the facets of the atlas and axis. The progressive slippage of the atlas over the axis secondary to this malalignment, a process that is similar to spondylolisthesis seen in the lumbosacral spine, results in invagination of the odontoid process into the craniocervical cord. The abnormal inclination of the facets of the atlas and axis appears to be the result of congenital malformation of the bones. It is also possible that this inclination is due to acquired causes, such as neck muscle weakness during early infancy, trauma to the neck during inappropriate delivery practices, and inability to spontaneously correct these deformities due to weakness of the muscles at the nape of the neck due to protein/calorie malnutrition. The progressive nature of the anomaly of basilar invagination has been alluded to by other authors.

We recently studied 170 patients with group I basilar invagination, treated with atlantoaxial joint distraction in our department. We analyzed the physical and radiological changes that occur in these patients following surgery.

Our analysis revealed that odontoid compression of the cord is the primary event and that all musculoskeletal alterations are secondary protective mechanisms of the body aimed at reducing the effect of neural compression. All of these secondary physical abnormalities are reversible following surgery that involves decompression of the cord and stabilization of the region. Essentially, it appears that these abnormalities are not a result of embryonic dysgenesis and are only secondary adaptive changes.

Clinical Features

Patients with basilar invagination have several physical changes, such as reduced neck size, torticollis, and exaggerated lordosis of the cervical spine, as well as reduced craniospinal angulation. These patients usually present with neck pain, weakness of extremities, paresthesias, hoarseness of voice, nasal regurgitation, and bowel/bladder symptoms.

In the majority of cases, there is a history of moderate to severe trauma prior to the onset of symptoms. In most cases, the patient or his or her family had noticed the patient’s short neck since early childhood. Although some degree of torticollis is also present since early childhood, this symptom is exaggerated after trauma or after the onset of clinical neurological symptoms.

Our patients’ ages, clinical features, and duration of symptoms are summarized in Table 29.1.

Physical and Radiological Parameters

Computed tomography (CT), magnetic resonance imaging (MRI), and dynamic radiography are the main imaging technologies used with these patients.

In most patients, imaging shows evidence of reduced disk space height, significant posterior cervical osteophyte formation, assimilation of the atlas, single- or multiple-level cervical fusions, and an increase in the spinal subarachnoid space both above and below the level of maximum neural compression at the tip of the odontoid process.

The landmarks used for measurements of various indices were based on a review of original work by Van Gilder et al. and those used in our earlier studies. They are shown in Figs. 29.1, 29.2, and 29.3.

Craniovertebral Height

The craniovertebral height is measured by a modification of the Klaus posterior fossa height index. A line is drawn from the tuberculum sellae to the torcula. From the midpoint of this, a line is drawn that connects to the midpoint of the inferior surface of the body of C5.
Care is taken to select the images that are in a neutral neck position. C5 is selected to assess the lower limit of the neck, as in a significant percentage of our cases, the investigations did not show the C6 and C7 vertebrae.

### Cervical Height

The cervical height is measured by a line drawn from the tip of the odontoid process to the midpoint of the base of C5 (Fig. 29.1).

### Cervical Lordosis

Cervical lordosis is measured by a modification of the omega angle (Fig. 29.2). The line of the hard palate is taken as a fixed line parameter, and a parallel line is drawn to it that passes from the center of the base of C3. The angle of the odontoid process is measured on this line. The base of C3 is selected instead of C2, as discussed by us elsewhere, because in several cases there are C2–C3 fusions.

---

**Table 29.1 Clinical characteristics of patients**

<table>
<thead>
<tr>
<th>Total no. of patients</th>
<th>170</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age (y)</strong></td>
<td></td>
</tr>
<tr>
<td>0–10</td>
<td>8</td>
</tr>
<tr>
<td>11–20</td>
<td>48</td>
</tr>
<tr>
<td>21–30</td>
<td>58</td>
</tr>
<tr>
<td>31–40</td>
<td>40</td>
</tr>
<tr>
<td>41–50</td>
<td>12</td>
</tr>
<tr>
<td>&gt; 50</td>
<td>4</td>
</tr>
<tr>
<td><strong>Duration of symptoms</strong></td>
<td></td>
</tr>
<tr>
<td>0–6 mo</td>
<td>62</td>
</tr>
<tr>
<td>6–12 mo</td>
<td>28</td>
</tr>
<tr>
<td>13–24 mo</td>
<td>26</td>
</tr>
<tr>
<td>25–36 mo</td>
<td>18</td>
</tr>
<tr>
<td>37–48 mo</td>
<td>21</td>
</tr>
<tr>
<td>49–60 mo</td>
<td>0</td>
</tr>
<tr>
<td>61 mo–15 y</td>
<td>15</td>
</tr>
<tr>
<td><strong>Presenting symptoms</strong></td>
<td></td>
</tr>
<tr>
<td>Neck pain</td>
<td>110</td>
</tr>
<tr>
<td>Paresthesias</td>
<td>62</td>
</tr>
<tr>
<td>Weakness</td>
<td>170</td>
</tr>
<tr>
<td>Hoarseness of voice or nasal regurgitation</td>
<td>14</td>
</tr>
<tr>
<td>Bladder or bowel disturbance</td>
<td>41</td>
</tr>
<tr>
<td><strong>Sensations</strong></td>
<td></td>
</tr>
<tr>
<td>Normal sensations</td>
<td>55</td>
</tr>
<tr>
<td>Only posterior column</td>
<td>35</td>
</tr>
<tr>
<td>Only spinothalamic tract</td>
<td>18</td>
</tr>
<tr>
<td>Posterior column and spinothalamic tract</td>
<td>62</td>
</tr>
</tbody>
</table>
Craniocervical Angle

On a lateral radiograph, a line is drawn along the clivus corresponding to the Wackenheim clival line (Fig. 29.3). Another line is drawn along the posterior surface of C2–C3. The angle between these two lines is identified as the craniovertebral angle.

Measurements are made on all three forms of investigation (plain radiographs, CT, and MRI) and are then averaged. Because stainless steel metal implants are used in the majority of our cases for fixation, postoperative MRI was not possible.

Neck size is assessed by two parameters. A line is drawn from the inion to the tip of the spinous process of C7. Another line is drawn from the angle of the mandible to the medial end of the clavicle. Both measurements are done in a neutral neck position.

Clinical photographs help in assessing the alterations in the degree of torticollis.

Table 29.2 lists the various abnormalities that were observed on imaging, and Tables 29.3, 29.4, and 29.5 show the various postoperative changes in the parameters assessed in our patients. Figures 29.4, 29.5, and 29.6 illustrate the various musculoskeletal changes associated with basilar invagination. On MRI, in addition to the features observed in Table 29.2, large and dilated subarachnoid spaces were observed anterior to the cord both above and below the point of maximum neural compression at the tip of the odontoid process in at least 66% of cases (Figs. 29.4a and 29.5b). Although there were spondylotic bone changes, neural compression by the osteophytes was not prominent because of the presence of “buffering” by enlarged subarachnoid spaces.

Reduced neck size has been considered diagnostic of basilar invagination. In our series, partial or complete fusion of the vertebral bodies appeared to be directly related to long-standing reduction of the disk space height. This feature was apparent by the range of reduction of the disk space height in these cases. Spondylotic changes with formation of osteophytes indenting into the cervical subarachnoid space at multiple levels are observed in these patients. These “spondylotic” changes are disproportionately more common when related to patients’ ages.

### Table 29.2 Radiological characteristics of patients

<table>
<thead>
<tr>
<th>Abnormality</th>
<th>No. of Patients (N = 170)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Partial or complete assimilation of atlas</td>
<td>123</td>
</tr>
<tr>
<td>Fusion</td>
<td></td>
</tr>
<tr>
<td>C2–C3</td>
<td>44</td>
</tr>
<tr>
<td>C3–C4</td>
<td>3</td>
</tr>
<tr>
<td>C4–C5</td>
<td>3</td>
</tr>
<tr>
<td>C3–C4–C5</td>
<td>5</td>
</tr>
<tr>
<td>Osteophytes</td>
<td></td>
</tr>
<tr>
<td>C2–C3</td>
<td>12</td>
</tr>
<tr>
<td>C3–C4</td>
<td>18</td>
</tr>
<tr>
<td>C4–C5</td>
<td>6</td>
</tr>
<tr>
<td>Chiari malformation</td>
<td>28</td>
</tr>
<tr>
<td>Syringomyelia</td>
<td>25</td>
</tr>
</tbody>
</table>

### Table 29.3 Extent of change in craniovertebral and cervical height

<table>
<thead>
<tr>
<th>Increase in Height (cm)</th>
<th>Craniovertebral Height (No. of Patients)</th>
<th>Cervical Height (No. of Patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>24</td>
<td>49</td>
</tr>
<tr>
<td>0–1</td>
<td>40</td>
<td>86</td>
</tr>
<tr>
<td>1–2</td>
<td>55</td>
<td>25</td>
</tr>
<tr>
<td>2–3</td>
<td>36</td>
<td>7</td>
</tr>
<tr>
<td>3–4</td>
<td>12</td>
<td>3</td>
</tr>
<tr>
<td>4–5</td>
<td>3</td>
<td>–</td>
</tr>
</tbody>
</table>

### Table 29.4 Changes in the craniospinal angle and modified omega angle in cervical lordosis

<table>
<thead>
<tr>
<th>Alterations in Angle (Degrees)</th>
<th>Craniospinal Angle (No. of Patients)</th>
<th>Modified Omega Angle (No. of Patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>21</td>
<td>15</td>
</tr>
<tr>
<td>0–10</td>
<td>31</td>
<td>48</td>
</tr>
<tr>
<td>11–20</td>
<td>72</td>
<td>63</td>
</tr>
<tr>
<td>21–30</td>
<td>40</td>
<td>38</td>
</tr>
<tr>
<td>30–40</td>
<td>7</td>
<td>6</td>
</tr>
</tbody>
</table>

Fig. 29.3 Line A is drawn along the clivus, and line B is drawn along the posterior surface of C2–C3. The angle between these lines is the **craniospinal angle**.
Table 29.5  Extent of increase in cervical height

<table>
<thead>
<tr>
<th>Change in Height (cm)</th>
<th>Number of Patients (N = 41)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0–1</td>
<td>15</td>
</tr>
<tr>
<td>1–2</td>
<td>10</td>
</tr>
<tr>
<td>2–3</td>
<td>12</td>
</tr>
<tr>
<td>3–4</td>
<td>3</td>
</tr>
<tr>
<td>4–5</td>
<td>1</td>
</tr>
</tbody>
</table>

Misdiagnosis of cervical spondylotic disease and erroneous treatment for cervical osteophytes are possible in such cases. The spondylotic changes are more marked and prominent at the level of C2–C3, or in cases where there is C2–C3 fusion, at the level of C3–C4. Fusions are also more prominently observed both above (partial or complete assimilation of the atlas) and below (C2–C3 fusion) the point of maximum neural compression at the tip of the odontoid process. The anterior subarachnoid space is remarkably dilated both above and below the point of maximal neural compression by the odontoid process.

Surgery

All patients in our series underwent atlantoaxial facet distraction fortified by plate and screw fixation by the surgical steps elaborated earlier. In more than half of all patients, an acrylic, stainless steel metal plate, or titanium metal “spiked” spacer was used in addition to bone graft for distraction of the facets. In about a third of patients, only bone graft with no metal spacer/distractor was used. In 12 patients, distraction/fixation of the facets was done with metal spacers, and no additional atlantoaxial fixation with plates and screws was performed.

Postoperative Musculoskeletal Changes

Radiological assessment shows that the craniovertebral height increases in > 90% of patients, and in 30% of patients the increase is > 2 cm. The most remarkable change is the reversal of craniospinal angulation and alignments. The angle of the clivus in relationship to the spine becomes more normal, as can be appreciated in Figs. 29.4, 29.5, and 29.6. The reversal of posterior lordosis and increase in the height of the disk space are responsible to a great extent in directly increasing the height of the neck. The disk space height increases more significantly in the upper cervical vertebrae. In cases where there is no assimilation of the arch of the atlas, the distance between the arch of the atlas and the suboccipital bone increases most remarkably. This increase in the disk space height following surgery is suggestive of the fact that spondylotic changes and even bone fusions are potentially reversible.

Fig. 29.4a–h
a  T2-weighted magnetic resonance imaging (MRI) of an 11-year-old girl showing basilar invagination.

b  T1-weighted MRI.

Fig. 29.4c–h
Fig. 29.4c–h

- **c** Computed tomography (CT) scan showing basilar invagination, partial assimilation of the atlas, and C2–C3 fusion.
- **d** Lateral radiograph with the neck in flexion.
- **e** Lateral radiograph with the neck in extension showing the hyperlordosis of the cervical spine.
- **f** Postoperative CT scan. Note the craniovertebral and cervical spinal realignment and increase in the craniovertebral and neck height.
- **g** Postoperative radiograph with the neck in extension. Note the recovery in posterior cervical lordosis and neck size.
- **h** Postoperative radiograph with the neck in flexion.
The evaluation of the cervical lordosis is done by modification of the Klaus omega angle\(^a\) and modified omega angle.\(^a\) Exaggerated posterior cervical lordosis or hyperextension of the neck was observed in \(\geq 90\%\) of patients.\(^a\) The craniospinal angle and cervical lordosis recover remarkably following surgery (Table 29.4). Considering the alterations in the angle of the clivus, it appears that if conventional parameters are used, platybasia also recovers significantly.

The neck size is measured as the distance between the external occipital protuberance and C7 spinous process in a neutral position and the distance between the angle of the mandible and the medial end of the clavicle. Table 29.2 shows the postoperative increases in neck length seen in our patients. The neck size increases in the majority of patients following surgery. The increase in the neck height and other parameters are disproportionately more when compared with that achieved locally by direct distraction of the facets of the atlas and axis.

Although there are some methods discussed in the literature to measure the extent of torticollis, it was assessed by clinical photographs in our series. Torticollis recovers in all patients to some extent. In our series in \(\approx 70\%\) of patients, this recovery was, to say the least, remarkable.\(^a\) The recovery in torticollis occurred even
though no special operative measure was adopted to correct it. The reversal of torticollis suggests that the altered neck angulation is probably a mechanism that attempts to minimize the neural compression. The mechanism appears to simulate the well-described phenomenon of development of scoliosis following acute spinal disk prolapse. The neck movements also become more supple and free despite the craniovertebral fixation.

Several authors have opined that congenital and developmental musculoskeletal abnormalities and anomalies that affect the craniovertebral junction complex can result in a range of neural compression. From the analysis of postoperative musculoskeletal changes that occur, it appears that neural compression at the craniovertebral junction is primary, and the majority of osseous abnormalities are probably secondary in nature and unrelated to embryonic dysgenesis. A variety of changes that probably occur in the bones and soft tissues appear to be directly related to the natural adjustments that operate to minimize the compression of the cord by the indentation of the odontoid process and by the instability of the region. The restriction of neck movements, reduction of the neck size, and alterations in the craniospinal angulation probably lead to large subarachnoid spaces in relationship to the cervical
cord and to the brainstem. All these natural responses probably allow the cord a relatively stretch-free traverse over the indenting odontoid process. Reduction of the disk spaces, osteophyte formation, and incomplete and complete cervical fusions and alterations in the craniospinal and cervical angulations appear to be directly related to the reduction in the neck size. The reduction in the disk space height and fusions are more prominently seen in the upper cervical vertebrae. Also, cervical fusions and assimilation of the atlas may be related to long-standing and progressive reduction in the disk space height.

**Conclusion**

Considering that several of the physical alterations in the neck occur in early childhood, it appears that the process of neural compression is recognized and the process of natural adjustments is initiated early in life. The stage of neural decompensation probably occurs later in life and is frequently initiated by some trauma. The nonsymptomatic presence of short neck, torticollis, and other such features should alert the physician and the patient about the possibility of the presence of basilar invagination and indentation of the neural tube by the odontoid process.

Reduction in the neck size and alterations in its physical form and functional ability in cases with basilar invagination appear to be acquired anomalies and are probably a protective response of the body to minimize the stretch of the cord over the indenting odontoid process. The presence of cervical spondylotic changes, vertebral body fusions, and alterations in craniospinal angulations are probably related to the reduced neck size and torticollis. All these changes are potentially reversible following a successful decompression and fixation of the region.

**Case Illustration**

A 20-year-old man presented with a short neck and torticollis since birth. For about 2 years, he had neck pain and progressive quadriparesis. Investigations done 1 year ago had shown basilar invagination, "fixed" atlantoaxial rotatory dislocation, assimilation of atlas, and fusion of C2 and C3 vertebrae. There was clear evidence of cord compression due to large osteophytes opposite the C3–C4 disk space (Fig. 29.7a, b). Although the cord was humped over by the odontoid process, no cord changes were evident at this level. Repeat investigations at the time of admission showed similar radiological features. Considering the presence of clinical and radiological features of instability of the craniovertebral region, a stabilization procedure was planned that...

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**Fig. 29.7a–d**

a. T2-weighted MRI showing basilar invagination, atlantoaxial dislocation, assimilation of atlas, C2–C3 fusion, and hyperintensity in the cord at the level of C3–C4.

b. CT scan showing basilar invagination, atlantoaxial dislocation, assimilation of the atlas, C2–C3 fusion, and relatively large posterior osteophytes at the level of C3–C4.
would have had incorporated occipital bone and the C3–C4 vertebra. However, during operation, a marked instability was identified at the craniovertebral-atlantoaxial joint, while the C3–C4 vertebrae appeared remarkably stable. The patient was treated with distraction, reduction, and fixation of the craniovertebral instability that included basilar invagination and “fixed” atlantoaxial dislocation. Specialized spacers and bone graft pieces harvested from the iliac crest were impacted in the distracted atlantoaxial joint and direct plate and screw fixation incorporating C1 facet and C2 pars was performed. The C2–C3 and C4 spinous processes revealed no evidence of instability and no fixation of the region was attempted. He was placed on a hard cervical collar for 3 months. Postoperatively, the patient had remarkable clinical improvement. He was able to walk unaided swiftly, within a few days of surgery. Because stainless steel plates and screws were used for fixation, the status of the C3–C4 region cord compression could not be analyzed (Fig. 29.7c, d).

The case illustrates the presence of C2–C3 vertebral body fusions, large osteophytes opposite the C3–C4 space, high cervical disk protrusion, and atlas assimilation in a patient with basilar invagination and “fixed” atlantoaxial dislocation. The craniovertebral anomaly in this patient was the primary pathology and the C3–C4 disk region compression a secondary phenomenon. Treatment of the primary pathology probably resulted in the relief from a factor that caused a secondary effect on the cord.

References

“Fixed” (Irreducible) Atlantoaxial Dislocation
Atul Goel and Dattatraya Muzumdar

Among other causes, atlantoaxial dislocation can result from trauma, inflammation, and congenital anomalies. Although shorter-duration dislocations often reduce on extension of the neck, long-standing dislocations may become fixed with time. The incidence of fixed atlantoaxial dislocation is relatively low, and the literature on the subject is available only as isolated reports. The treatment protocol in such cases has not yet been fully agreed upon.

This chapter will review the nomenclature, pathogenesis, and treatment options available for fixed atlantoaxial dislocation, with a special emphasis on the utility of joint manipulation and distraction in such patients. According to our current hypothesis, based on observations in surgically treated cases, we now believe that all patients with “fixed” dislocations have pathologically abnormal movements that cause micromotion and cord compression. All patients labeled as having fixed dislocations and neurological deficits can be candidates for manipulation of the atlantoaxial joint, craniovertebral realignment, and reduction and fixation of the dislocation.

Nomenclature
An atlantoaxial dislocation can be described as either mobile or fixed. In mobile dislocation, there is complete radiographic reduction of the dislocation on full extension of the neck or after institution of cervical traction. The dislocation is described as fixed when no radiographic reduction occurs even on full neck extension or after institution of cervical traction. In a select group of patients, a dislocation considered fixed on awake traction may reduce when traction is applied under general anesthesia. Therefore, some authors attempt reduction by traction under general anesthesia before labeling an entity as fixed.

Etiopathogenesis
Atlantoaxial dislocation may be congenital in nature or secondary to trauma to the region. It may also occur secondary to rheumatoid or degenerative osteoarthritis of the atlantoaxial joints. Irreducibility of atlantoaxial dislocation is probably the result of a gradual and progressive process rather than an acute episode. Various causes of irreducibility have been suggested. Some authors speculate that prolonged atlantoaxial dislocation results in contraction of the anterior muscles, ligaments, and capsules of the atlantoaxial joint and subsequent scar formation. Scar tissue around the dens and ossification may also be seen in this type of contraction. The development of scar tissue in these cases prevents reduction by nonoperative methods, such as skull traction. Entrapment of the transverse ligament after fracture at the base of the odontoid process, fusion anomalies of the region, and other such causes have also been implicated. Subin et al. believed that, in chronic fixed atlantoaxial dislocation, granulation tissue, callus around the dens, scarring between the dens and the axis, and locking of the atlantoaxial lateral joints are the major reasons for the irreducibility. Congenital os odontoideum and fracture at the base of the odontoid process are frequent accompaniments of fixed atlantoaxial dislocation, and in many of these cases, an exact differentiation between the congenital or traumatic etiology may not be possible.

Surgical Options
A variety of methods to treat reducible atlantoaxial dislocation have been described in the literature. Procedures involving screw implantation into the lateral masses have been popular and include either the transarticular method described by Grob and Magerl in 1987 or the interarticular method of fixation described by us in 1988. Both the transarticular and interarticular methods of fixation are technically challenging operations, but they provide remarkable stability to the region.

The treatment protocol for fixed atlantoaxial dislocation is, however, not adequately streamlined in the literature. Various authors have suggested a transoral bony decompression followed by a posterior fixation as the safest method of treatment of this complex anomaly. Wang et al. theorized that the dislocation in these patients might be reduced by anterior transoral atlantoaxial joint release without the need for odontoid resection. Treatment by posterior decompressive procedures has also been reported but may be associated with a high complication rate. Other authors have reported success with stand-alone transoral decompression of the region, without the need for subsequent posterior fixation.

In a substantial number of patients, the irreducibility is due to partial fusion of the atlas-axis joint space; in these patients, attempts at reduction with techniques that do
not involve opening of the joint usually fail. Our method of joint distraction followed by interarticular lateral mass plate and screw fixation involves wide exposure of the atlantoaxial facet joint after sectioning of the C2 ganglion. The articular cartilage of the joint is widely removed, the joint space is distracted, and bone graft is introduced in the joint space. This direct atlantoaxial facet distraction facilitated by traction and radiographic control results in reduction of the fixed dislocation in a significant number of cases. Thus, we observed that even in cases where a fusion was evident on imaging, an attempt can be made to manipulate and reduce the joint. Cervical traction followed by occipitocervical fixation may not be effective in a large percentage of these cases. On the other hand, direct opening of the joint and manual distraction are a far more effective procedure. We observed during several operations that in these cases the region is not entirely fixed and is instead abnormally mobile. The excessive and unnatural mobility of the joint is probably the cause of compressive symptoms. It was also observed that fixation, even if it was in an incompletely reduced position, resulted in symptomatic recovery.

**Intraoperative Joint Distraction for Reduction of Fixed Atlantoaxial Dislocation**

**Surgical Technique**

The basic steps of the surgery are the same as discussed in our articles on similar subjects. The patient is placed prone, with the head end of the table elevated to −35°. The head-high position provides countertraction and helps reduce venous engorgement in the operative field. Cervical traction is instituted prior to anesthesia and is continued. The atlantoaxial facet joints are opened up after excising the capsule and exposed on both sides widely after sectioning of the large C2 ganglion. Large veins in the region of the C2 ganglion and lateral masses frequently cause troublesome bleeding. The venous bleeding can be controlled by employing judicious use of diathermy and packing of the extradural space and the space lateral to the facets with Surgicel and/or Gelfoam. The articular cartilage is widely removed using a microdrill. The joints on both sides are distracted using an osteotome. The distraction is maintained by placement of bone graft pieces harvested from the iliac crest. Whenever necessary, the distraction of the joint can be maintained with the help of specially designed multiholed titanium blocks used as spacers or strut grafts that are placed or impacted into the joints (Fig. 30.1). The size of the spacers used depends on the space available within the distracted joint space. In our initial cases, hydroxyapatite blocks were used, which were later replaced by titanium spacers in subsequent cases. Spacers measure 12 mm in length, 10 mm in breadth, and 4 mm in height. These customized titanium spacers have multiple small holes and are tapered at one end for easier placement during insertion in the joint space. Holes in the spacers allow bone incorporation across the prepared joint space. Morcellized bone graft harvested from the iliac crest is inserted into the distracted joint space on all sides and into the holes of the spacer. Plate and screw fixation of the region is subsequently performed by the interarticular technique. A two-holed stainless steel plate is used measuring 15 to 20 mm in length. The screws measure 2.4 to 2.8 mm in diameter and 20 to 28 mm in length. Screws are passed bilaterally through the holes in the plate into the lateral mass of the atlas and pars of the axis (Figs. 30.2, 30.3, 30.4, 30.5, and 30.6). Axis screws are tightened first, then the atlas screws are tightened simultaneously on both sides. This maneuver, which is akin to reduction of lumbar spondylolisthesis using pedicle screws and plates/rods, helps to reduce the forward slip of the atlas over the axis. The reduction is assessed on the table by intraoperative fluoroscopy. Our experience with neuronavigation suggests that this technology can help to make the procedure significantly safe as regards the vertebral artery and in selecting the best trajectory for screw implantation. Postoperatively, the traction is discontinued, and the patient is placed in a four-poster hard cervical collar for 3 months; furthermore all of the patient’s physical activities involving the neck are restrained during this period.

**Rationale**

We observed that introduction of bone graft with or without the assistance of a spacer within the atlantoaxial joint, subsequent fixation with a plate, and direct implantation of screws in the atlas and axis can sustain the reduction and provide a basis for a solid bone fusion. When necessary, the screw-tightening procedure in the atlas and axis can be modified to effect a reduction with the technique described for spondylolisthesis.
Lateral radiograph of the craniovertebral region in flexion in a 45-year-old woman showing severe atlantoaxial dislocation. A pseudarthrosis at the base of the odontoid process can be seen.

Lateral radiograph in extension shows no reduction of the dislocation.

Computed tomography (CT) scan showing os odontoideum and atlantoaxial dislocation.

CT scan showing degenerative changes in the atlantoaxial joints on both sides.

Postoperative radiograph showing fixation by plate and screws in the reduced position.

Fig. 30.2a–e
Fig. 30.3a–g

a  Lateral radiograph with the head in flexion of a 45-year-old man showing atlantoaxial dislocation.

b  Lateral radiograph with the head in extension showing fixed atlantoaxial dislocation.

c  CT scan of the patient shows atlantoaxial dislocation.

d  CT scan with the sagittal section passing through the joint showing an inclined alignment.

e  Postoperative CT scan showing complete reduction of the dislocation.
Intraoperative Joint Distraction for Reduction of Fixed Atlantoaxial Dislocation

Fig. 30.4a–g Imaging of a 22-year-old man.

a Lateral radiograph of the craniovertebral region with flexion of the neck showing atlantoaxial dislocation. (There was no history of any significant trauma.)

b Lateral radiograph in extension shows no reduction of the dislocation.

c T2-weighted magnetic resonance imaging (MRI) showing severe cord compression.

g Postoperative radiograph showing plate and screw fixation.
Fig. 30.4d–g

d CT scan showing the atlantoaxial dislocation. Os odontoideum can be seen.
e Postoperative CT scan showing reduction of the dislocation.
f Postoperative CT scan with a cut through the atlantoaxial joint. The plate and screws and the spacer can be seen.
g Postoperative radiograph taken 2 years after surgery with flexion of the neck showing reduction of the dislocation and lateral mass plate and screw fixation. Spacers on two sides can be seen.

Fig. 30.5a–h

a Preoperative radiograph of a 15-year-old boy with the head in a flexed position showing marked atlantoaxial dislocation.
b Preoperative radiograph with the head in extension showing fixed atlantoaxial dislocation.
Fig. 30.5c–h

c Preoperative CT scan showing marked dislocation.
d Sagittal cut of CT scan showing facet dislocation.
e T1-weighted MRI showing the dislocation and marked stretch of the ligaments.
f Postoperative CT scan showing significant but incomplete reduction.
g Postoperative CT with the sagittal cut through the lateral masses. Realignment of the facets and fixation with plate and screws are observed.
h Postoperative radiograph showing the plate and screw fixation.
Fig. 30.6a–e  Imaging of a 50-year-old man.

a  Radiograph with the head in flexion showing fracture of the odontoid process and fixed atlantoaxial dislocation.

b  Radiograph with the head in extension showing persistent dislocation.

c  Preoperative MRI showing the dislocation and cord signal intensity changes.

d  Postoperative CT scan showing reduction of the dislocation following distraction of the atlantoaxial joint.

e  Postoperative CT scan showing C1–C2 fixation of the facets.
Monoaxial screws and custom-made plates were used in the present series.

The technique of atlantoaxial joint manipulation and reduction has been discussed before. Subin et al. believed that an anterior transoral approach could release the compression of the cord and also directly loosen the atlantoaxial joint. This goal is accomplished by removal of the callus, scars, and granulation tissue so that it is possible to reduce the dislocation during and after surgery. Subin et al. believed that direct release of the locked lateral joints of the atlas and axis and the excision of the articular cartilages between the superior and inferior surfaces of the lateral joints could bring about unlocking as well as eventual fusion at this level without the need for any secondary posterior fusion. The authors achieved postoperative atlantoaxial reduction by continuous biaxial skull traction and maintained the reduction in a cast for 3 to 4 months.

Various authors have performed atlantoaxial facet joint manipulation in rotatory atlantoaxial dislocation and effected a reduction of the dislocation. Schmidek et al. used a transoral route, Crockard and Rogers employed an extreme lateral approach, and Goto et al. released the atlantoaxial lateral facet joint by transoral surgery and followed it up by a posterior fixation using interlaminar clamps.

We could obtain a satisfactory reduction without the need for additional anterior or posterior decompressive surgery in the majority of our cases. The space provided to the spinal cord by the release of the facet joints and reduction of the dislocation and stabilization afforded to the atlantoaxial region helped in neurological recovery. It was observed that anatomical reduction could be better achieved in cases that had fixed atlantoaxial subluxation in the presence of os odontoideum or odontoid fracture. In cases not associated with the above features, it was generally observed that the reduction was partial probably due to the presence of the relatively unyielding nature of the organized fibrous tissue in the predental region. The patients were not placed on any long-term traction or external rigid immobilization protocol. The alignment of the bony architecture was in an acceptable position with enough space available for the spinal cord. An anterior transoral decompression procedure is possible in a subsequent stage if the alignment after the distraction surgery or the neurological recovery of the patient is less than satisfactory. We have observed that the procedure of distraction and attempts toward reduction of the dislocation as discussed are safe, and an anterior transoral procedure could be done whenever it is felt to be necessary.

**Conclusion**

In our experience, there may be a place for reduction of fixed atlantoaxial dislocation and subsequent fixation without the removal of any bony spinal element. The procedure is technically demanding and anatomically precise, but if it is learned adequately and performed successfully, the neurological outcome is extremely gratifying.

**References**

31  Rotatory Atlantoaxial Dislocation

Abhidha Shah, Atul Goel, Shradha Maheshwari, and Antonio Figueiredo

Atlantoaxial dislocations are divided into anterior, posterior, vertical, and rotational types.\textsuperscript{1–3}  Destructive, infective, traumatic, or degenerative involvement of the craniovertebral region can rarely result in lateral dislocation of the facet of the atlas over the axis. In this chapter, we discuss the rotational types of atlantoaxial dislocations.

Physical neck rotation (or torticollis) is of three types: positional, spasmodic, and facetal rotatory.

\section*{Positional Torticollis}

This is related to positional alteration in the craniovertebral junction over a prolonged period of time as well as indentation of the odontoid process into the cervicomедullary cord. Such an indentation is seen in patients with group I basilar invagination. The physical neck alterations simulate the changes in spinal posturing secondary to disk herniation. The torticollis seen in patients with group I basilar invagination is present in most cases. The long-standing torticollis seen in such cases appears to be part of a protective alteration that occurs as an attempt to prevent the spinal cord from relentlessly and progressively reducing the spinal canal. Our recent observations suggest that such a torticollis (and even short neck) is reversible following decompression and stabilization of the craniovertebral junction.\textsuperscript{4}

\section*{Treatment}

Direct manipulation of the atlantoaxial joint and distraction of facets with bone graft, with or without titanium metal spacers, have been shown to result in reduction of basilar invagination and fixed atlantoaxial dislocation (Figs. 31.1 and 31.2). We have observed remarkable reduction of torticollis by this method of treatment (Fig. 31.2). Differential distraction of the facets can also result in direct reduction of torticollis (Fig. 31.1). Torticollis reduces even after transoral decompression of the odontoid process.

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\textbf{Fig. 31.1a–f} Images of a 20-year-old woman. She had torticollis to the right side since birth. For 2 years, she had progressive weakness of all four limbs. When admitted, she needed support to walk and to carry out all routine activities of her life. She had spastic grade 4 quadriparesis.

\textbf{a} Computed tomography (CT) scan (sagittal cut) shows evidence of basilar invagination.

\textbf{b} Coronal T1-weighted magnetic resonance imaging (MRI) shows lateral indentation of the spinal cord.

---
Fig. 31.1c–f

C Coronal CT scan showing marked tilting of the atlas.
D Postoperative CT scan showing reduction of basilar invagination.
E Sagittal CT scan showing a large spacer within the atlantoaxial joint.
F Coronal CT scan showing the spacer on the side of the torticollis. Reversal of torticollis can be observed by reduction of the tilt of the atlas and its facets.

Fig. 31.2a–i Images of a 26-year-old woman. She had neck pain, tingling, numbness, and weakness in all four limbs for 3 years. The disability was progressive. The patient had marked quadriplegia and could not perform any useful activity by herself. She had spastic grade 3 quadriplegia.

A T1-weighted MRI showing marked basilar invagination, Chiari I malformation, and syringomyelia.
B T2-weighted MRI.

Fig. 31.2c–i
Rotatory Atlantoaxial Dislocation

Fig. 31.2c–f

c  CT scan reconstruction showing basilar invagination.
d  Axial CT scan showing marked torticollis and the odontoid process deviated on one side of the midline.
e  CT scan showing torticollis.
f  Another view of the CT scan (coronal view) showing torticollis.

Fig. 31.2g–i
**Spasmodic Torticollis**

The torticollis in this group is related to sternocleidomastoid muscle spasm ([Fig. 31.3](#)). The pathology in these cases and the treatment remain controversial. Botulinum toxin injections in the spasmodic muscles have been found to be a satisfactory treatment modality in resistant cases.

**Facetal Rotatory Torticollis**

Torticollis in this group is related to rotatory atlantoaxial dislocation secondary to rotatory dislocation of the facet of the atlas over the facet of the axis ([Figs. 31.4 and Fig. 31.5](#)). This group forms a discrete entity and is the subject of discussion in this chapter. Locking of the atlantoaxial facets results in rotatory atlantoaxial dislocation. Such a dislocation has been identified more commonly in young children. Although several pathogenetic factors have been considered, the exact cause of the phenomenon is unclear. The dislocation is an acute event that usually follows an episode of relatively minor trauma. Some patients had throat and paranasal sinus infections or tonsillitis at the time of the event. However, an exact correlation between possible infection, trauma, and atlantoaxial dislocation is only speculative.

**Historical Background and Nomenclature**

Rotatory dislocation of the atlantoaxial joint was first described by Sir Charles Bell in 1830 and Corner in 1907. Wortzman and Dewar introduced the term *atlantoaxial rotatory fixation-subluxation* in 1968. Text continues on page 329.
Fig. 31.3  Spasmodic torticollis in a 5-year-old child.

Fig. 31.4a–m  Images of a 10-year-old boy. He had torticollis following a tap on the back of the head by a friend at school. An attempt at reduction by traction failed. Observation for a period of 2 months did not result in recovery of torticollis.

a  Three-dimensional anterior CT scan showing rotatory dislocation.

b  Posterior CT scan showing the rotation.
Fig. 31.4c–h

c  Axial CT scan showing rotatory dislocation.
d  Sagittal CT scan showing anterior displacement of the facet of C1 over the facet of C2.
e  Sagittal CT scan showing the tilt of the odontoid process.
f  Three-dimensional CT scan showing the odontoid process.

g  Postoperative CT scan showing reduction of the tilt of the odontoid process.
h  Sagittal CT scan showing fixation of the facets of the atlas and axis in a reduced position.
Fig. 31.4i–m
i  Sagittal CT scan showing fixation of the facets of the atlas and axis in a reduced position.
j  Axial CT scan showing screws passing through the facets of the atlas. Reduction of torticollis can be observed.
k  Coronal image showing lateral mass plate and screw fixation using plate and screws.
l  Picture showing torticollis.
m  Postoperative picture showing the neck of the patient in a good position.
Fig. 31.5a–i  Images of a 5-year-old girl. She developed sudden onset torticollis while playing at school.

a  Picture showing marked torticollis.
b  Axial CT scan showing partial rotatory dislocation. The facet of the atlas on one side acts as a pivot, and the contralateral facet of the atlas is in an anteriorly dislocated position. The process results in a rotatory dislocation.
c  Coronal CT scan showing the atlantoaxial facets on one side in alignment; on the contralateral side, the facet of the atlas is not aligned with the facet of the axis.
d  Sagittal image showing the dislocation of the facet of the atlas over the facet of the axis.
e  Three-dimensional image of the CT scan showing rotatory atlantoaxial dislocation.
Postoperative axial image of the CT scan showing the implant and the realignment of the facets.

Coronal image showing the facets of the atlas and axis in normal alignment.

Image showing plate and screw fixation of the atlantoaxial region and the facetal realignment.

Picture showing the neck in a normal position.
Rotatory atlantoaxial dislocation can be subdivided into reducible or irreducible types. Reducible rotatory atlantoaxial dislocation is classified as one in which the dislocation reduces on dynamic images or after institution of cervical traction. It is usually incomplete, when some part of the facet of the atlas is in contact with the facet of the axis. Complete rotatory dislocation can also reduce on traction, but the possibilities of its reduction are relatively less. Mobile and reducible rotatory dislocation can be treated by conservative observation for ~3 months using halo traction or a cervical collar. If during that period the rotatory dislocation reduces and remains reduced on dynamic imaging, there is no need for surgery. Otherwise, surgical fixation of the rotatory dislocation in a reduced position should be performed. In cases with irreducible rotatory dislocation, an attempt can be made to reduce the dislocation by local distraction and reduction by manual realignment.

The main presenting symptom of rotatory atlantoaxial dislocation is torticollis of the neck; neurological symptoms may be marginal. The torticollis may be painless. Surgical treatment for rotatory dislocation is to fix the dislocation in the reduced or maximally reduced position. Some surgeons feel that segmental fixation of the dislocation, even if it is in a dislocated position, allows the subaxial spine to move better, and torticollis can be expected to recover.

### Biomechanics

The occipitoatlantoaxial joints are the most complex joints of the axial skeleton. This joint complex forms part of the primary pillar of the spine that provides stability and mobility to the head. The primary movement of the atlanto-occipital articulation is flexion-extension; of the atlantoaxial joints, rotation (Table 31.1). The geometry of the lateral atlantoaxial articular surfaces is such that they are convex with horizontal orientation, thus able to permit rotation. The maximum rotation between the atlas and the axis is ~45°. When rotation exceeds 45°, the inferior facet of the atlas interlocks over the superior facet of the axis. If the transverse ligament is deficient, this facet interlock may occur even with rotation < 45°. It has also been seen that if the transverse ligament remains intact, the anterior arch of the atlas will not subluxate over the axis unless there is > 65° of rotation.

The alar ligaments limit axial rotation and side bending of the CO–C1–C2 complex. The left alar ligament limits rotation of C1 and the head to the right, and the right alar ligament limits rotation to the left. The vertebral arteries located in the foramen transversarium are not affected by the extremes of normal rotation, but they can be severely compromised by excessive rotation, especially if combined with anterior displacement. The fact that the vertebral artery has a dynamic relationship with the bones during rotational movements was first discussed by us. The laxity of the vertebral artery in the region assists in facilitating a range of neck movements.

### Etiopathogenesis

#### Predisposing Factors

Several predisposing factors have been identified for rotatory atlantoaxial dislocation, including upper respiratory tract infections, trauma, surgery of the head and neck, genetic diseases, and ocular infections. In addition, up to 24% of cases of rotatory atlantoaxial dislocation can occur without an obvious predisposing cause. Rheumatoid arthritis, Down syndrome, Morquio disease, and an assortment of congenital cervical anomalies have also been associated with it. There is a definite preponderance of rotatory atlantoaxial dislocation in the pediatric population. The higher incidence of rotatory atlantoaxial dislocation in children may be attributed to the shallower and more horizontally oriented joint surface, the relative elasticity of the ligaments, the not yet fully developed neck muscles, and a relatively large head.

#### Cause of Fixation

The cause of fixation remains obscure, as no anatomical or autopsy evidence is available. Wittek suggested that there is an effusion in the synovial joints producing stretching of the ligaments. Coutts proposed the theory that synovial fringes, when inflamed or adherent, may block reduction, whereas Fiorani-Gallotta and Luzzatti believed that the deformity is due to rupture of one or both alar ligaments and the transverse ligament. Watson Jones postulated hyperemic decalcification with loosening of the ligaments. Grisel suggested that muscle contracture might follow an upper respiratory tract infection and be a factor. Hess and associates concluded that there is a combination of factors, including muscle spasm, that prevent reduction in the early stages. Wortzman and Dewar postulated damage to

### Table 31.1 Range of motion at the occipitoatlantoaxial joints

<table>
<thead>
<tr>
<th>Occipitoatlantal joint</th>
<th>Degrees</th>
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<tr>
<td>Combined flexion and extension</td>
<td>25</td>
</tr>
<tr>
<td>One side lateral bending</td>
<td>5</td>
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<tr>
<td>One side axial rotation</td>
<td>5</td>
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<table>
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<tr>
<th>Atlantoaxial joint</th>
<th>Degrees</th>
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<tbody>
<tr>
<td>Combined flexion and extension</td>
<td>20</td>
</tr>
<tr>
<td>One side lateral bending</td>
<td>5</td>
</tr>
<tr>
<td>One side axial rotation</td>
<td>40–45</td>
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the atlantoaxial articular processes of unknown nature. Fielding and Hawkins suggested that reduction is probably obstructed in the early stages by swollen capsular and synovial tissues and by associated muscle spasm. If the abnormal position persists because of a failure to achieve reduction, ligament and capsular contractures develop and cause fixation.

## Radiographical Features

Three-dimensional computed tomography (CT) scanning is the gold standard investigation to diagnose and confirm rotational atlantoaxial dislocation. The exact alignment of the facets and their displacement can be evaluated. Dynamic CT scan, with the head and neck in flexed and extended positions, clearly delineates the pathology and assists in determining the appropriate surgical treatment modality.

Magnetic resonance imaging (MRI) assists in confirming the findings of CT scans and demonstrates features that would suggest compression and displacement of the neural and vascular structures.

Plain radiograph investigations have become less relevant with the increasing use of CT scans and MRI.

The degree of neck rotation or tilt in patients having rotatory dislocation is measured as an angle between a line that courses from the nasion, tip of the nose, and symphysis menti to the horizontal. The angle is expressed as the degree of tilt from the vertical.

## Clinical Features

Because of the rarity of the pathology, the infrequency of neurological involvement, and the difficulty in obtaining adequate radiographs, the diagnosis may be difficult to make and is usually delayed. The acuity of the neck deformity in an otherwise healthy and neurologically normal child is the hallmark of the problem. Pain is not a prominent presenting symptom. With more severe degrees of subluxation or dislocation, torticollis may be noted, and the patient may present with the typical “cock robin” posture, with the head tilted toward one side and turned (rotated) toward the other and in slight flexion. For example, in a right-sided unilateral anterior rotatory subluxation (right-sided atlas lateral mass subluxated anteriorly), there is lateral bending of the head and neck to the right and rotation of the head to the left. Attempting to turn the head opposite the direction it faces is difficult; however, rotating it further in the direction in which it faces is possible. On palpation, the C2 spinous process may be prominent and deviated to the side to which the chin is pointed as a result of the lateral tilt of the head or from counterrotation of C2; this is done in an attempt to realign the head.

In the acute setting, it is important to distinguish between true rotatory atlantoaxial dislocation and muscular torticollis. In rotatory atlantoaxial dislocation, the sternomastoid spasm is on the side of the chin, whereas in muscular torticollis, the spasm is on the contralateral side. In rotatory dislocation, the sternomastoid spasm on the side of the chin is secondary or reflexive, as if the muscle is attempting to correct the deformity.

Neurological involvement, fortunately, is uncommon; however, occipital neuralgia may occur because the greater occipital nerve runs in close proximity to the C1–C2 facet capsule.

## Classification

### Fielding and Hawkins Classification

Fielding and Hawkins classified rotatory atlantoaxial dislocation into four types.

#### Type I

*Rotatory atlantoaxial dislocation with no anterior displacement of the atlas (atlantodens interval ≤ 3 mm)*

The transverse ligament is intact, and the dens acts like a pivot. The rotation of the atlas on the axis is > 45°. This is the most common type of rotatory atlantoaxial dislocation.

#### Type II

*Rotatory atlantoaxial dislocation with anterior displacement of the atlas by 3 to 5 mm*

This type is associated with deficiency of the transverse ligament and unilateral anterior displacement of one lateral mass of the atlas, while the opposite joint is intact and acts as the pivot. In these patients, there is abnormal anterior displacement of the atlas on the axis, and the amount of fixed rotation is in excess of normal maximum rotation. The rotation usually exceeds 40°.

#### Type III

*Rotatory atlantoaxial dislocation with anterior displacement of the atlas by > 5 mm*

This type is associated with disruption of both the transverse and alar ligaments. Both lateral masses of the atlas are displaced anteriorly, one more than the other, thus producing the rotated position. The rotation is > 40°, and the atlas is displaced forward from the odontoid by > 5 mm.

#### Type IV

*Rotatory atlantoaxial dislocation with posterior displacement of the atlas*

This type is rare and occurs when the odontoid process is deficient or fractured. There is a posterior shift of
one or both lateral masses of the atlas, one of them shifting more than the other, thus causing rotation.

### Goel's Classification of Facetal Locking

Facetal locking can be divided into two types: rotatory and translatory.

### Rotatory Locking

Rotatory locking is when the head is in torticollis or a rotated position. It is considered partial when the facet of the atlas of only one side is dislocated anteriorly over the facet of the axis. The facet on the contralateral side is not abnormally positioned and acts as a pivot for the dislocation (Fig. 31.5). Rotatory locking is considered complete when the facets of the atlas on both sides are dislocated over the facets of the axis in a rotatory fashion. The process results in the facet of the atlas being positioned anterior to the facet of the axis on one side and posterior to it on the other side (Fig. 31.5).

### Translatory Dislocation

Translatory dislocation is a condition in which the facets of the atlas on both sides are dislocated anterior to the facets of the axis. Such a dislocation results in a fixed flexion neck deformity (Fig. 31.6). A posterior translation is also technically possible.

### Treatment

Traditional treatment guidelines based on the classification of Fielding and Hawkins⁹ are as follows:

- **Type I** lesions, which are the most benign, may be treated with cervical spine immobilization after reduction.

- For **type II** lesions in children, if reduction is achieved within 14 days, subsequent immobilization with a halo is recommended. For type II lesions associated with occipitoatlantal dislocation and/or when the diagnosis is delayed for more than 14 days, a posterior atlantoaxial or occipitoatlantoaxial arthrodesis is recommended.

- **Type III** and **IV** lesions are treated with posterior arthrodesis after reduction.

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**Fig. 31.6a–i** Images of a 12-year-old boy. He developed torticollis following a minor fall.

- a Lateral radiograph showing translatory dislocation. The C1 is dislocated anteriorly.

- b Three-dimensional CT scan showing the translatory dislocation of C1 over C2.
Fig. 31.6c–f

c  Sagittal image showing the dislocation of the facet of C1 over the facet of C2.
d  Axial image showing the facets of the atlas on both sides dislocated anterior to the facets of the axis.
e  Coronal image showing the atlantoaxial malalignment.
f  Photo of the boy showing him holding his head with his thumb under his chin to look ahead.

Fig. 31.6g–i
Fielding and Hawkins\textsuperscript{9} described children in whom the diagnosis of rotatory atlantoaxial subluxation was not made for months and in whom traction was unsuccessful in achieving a satisfactory reduction or relief of the symptoms. They performed an arthrodesis of the atlas and axis in these patients and achieved resolution of the torticollis. They made no attempt intraoperatively to realign the atlas on the axis. Manipulative reduction of rotatory atlantoaxial subluxation may be indicated in select patients with recent onset of torticollis, but there is less risk when reduction is achieved by bed rest or by traction without manipulation.

Phillips and Hensinger\textsuperscript{13} recommended a treatment plan that is based on the duration of the torticollis. Patients who have had a rotatory subluxation of less than 1 week's duration can be treated with immobilization in a soft collar and bed rest for 1 week. Close follow-up is essential. If reduction fails to occur spontaneously, then the patient should be hospitalized and given traction. In patients with rotatory subluxation longer than 1 week but less than 1 month in duration, hospitalization and cervical traction are indicated. After reduction, immobilization for 4 to 6 weeks is recommended. In patients with subluxation of more than 1 month's duration, cervical traction can be attempted for up to 3 weeks. If reduction does not occur, an arthrodesis should be performed.

There are only isolated reports mentioning surgical treatment strategy in cases with locked atlantoaxial facets. Schmidek et al.\textsuperscript{25} used a transoral route, and Crockard et al.\textsuperscript{26} employed an extreme lateral approach to remove obstructing ligamentous or bone structures within the joint. Goto et al.\textsuperscript{27} released the atlantoaxial facets.
lateral facet joint by a transoral surgery and followed it up by a posterior fixation using interlaminar clamps. Continuous monitoring of the neurological status of the patient is essential during any manipulative reduction. After reduction, some form of cervical immobilization is used to prevent recurrent subluxation. Immobilization presumably allows the stretched or edematous ligaments to heal and to return to their normal length. Full, pain-free motion of the neck should be restored before external immobilization is discontinued.

Some authors have discussed the usefulness of fixation in the abnormal position. They observe that segmental fixation of the dislocation, even if it is in a dislocated position, allows the subaxial spine to move better, and torticollis can be expected to recover. Our technique involves sectioning of the C2 ganglion, wide exposure, and opening up of the atlantoaxial joint and unilateral or bilateral manipulation of the atlantoaxial facets. It was observed that external traction may not be successful in relocating the facets, as the effect of traction was dissipated more to the lower neck than to the site of the problem in the region of the joint.

Goel’s Technique of Craniovertebral Realignment for Cases with Locked Facet

Under anesthesia, the patient is placed under cervical traction, the weights ranging from 3 to 6 kg. The head end of the table is raised ~35° to provide countertraction by the body. The position assists in reducing the venous congestion in the operative field, particularly in the vein-loaded region of the lateral masses. The exposure of the atlantoaxial joint in cases with facet malalignment is significantly more difficult and technically challenging when compared with a normally aligned atlantoaxial joint encountered during treatment of posttraumatic instability. However, it has been observed that intraoperative traction results in an improvement in the alignment of the facets in all cases, when compared with their preoperative status. The atlantoaxial facet joints are widely exposed on both sides after sectioning of the large C2 ganglion. Such a wide exposure provides an opportunity to observe the status of the facets and for direct manipulations. The joint capsule is excised, and the articular cartilage is widely removed using a microdrill. The joints on both sides are distracted using an osteotome. The flat edge of the osteotome is introduced into the joint; it is then turned to the vertical orientation to effect distraction and further manipulation. The size of the osteotomes varies depending on the local situation. The sequence of manipulation of the facets varies, but in general, the more affected facet is treated first. Whenever necessary, distraction is done simultaneously on both sides.

Although visualization of the region with high-definition radiography or intraoperative CT scanning is possible, such investigations were not performed in this series. Direct observation of the position and alignment of the articular surfaces of the facets of the atlas and axis was the key to facetal realignment. Another indicator of successful realignment was the location of the tip of the spinous process of the axis in relationship to the midline of the arch of the atlas. Corticocancellous bone graft harvested from the iliac crest was stuffed into the joint in small pieces. Subsequent fixation of the joint with the help of interarticular screws and a metal plate provided a biomechanically firm fixation and sustained distraction and realignment. In cases where the facet of the atlas was dislocated anteriorly and manual realignment was incomplete, the screw (with the plate) was first inserted into the lateral mass of the axis, then tightened. The atlas screw was then tightened over the fixed plate. This variation resulted in posterior movement of the facet of the atlas. A similar technique has been described in the treatment of spondylolisthesis. Larger pieces of corticocancellous bone graft were placed in the midline over the arch of the atlas and the lamina of the axis after appropriately preparing host bone.

Postoperatively, the traction is discontinued, and the patient is placed in a four-post hard cervical collar for 3 months. All physical activities involving the neck are restrained during this period. After 6 months, all activities, including sports, are permitted in an unrestrictive manner.

We employed the technique of opening of the joint, distraction and manipulation of the facets, and reduction of rotatory atlantoaxial dislocation. The technique is relatively complex and tedious, particularly in neurologically intact patients. However, reduction of the rotatory dislocation is possible with this technique (Figs. 31.4 and 31.5). Holding and manipulating the spinous process of the axis, the posterior arch of the atlas, and the facets of the atlas and axis can result in reduction of the rotatory dislocation.

Conclusion

Rotatory or translatory atlantoaxial dislocation and related neck torticollis can be a physically crippling disorder. The difficulties associated with exposure and manipulation of the atlantoaxial joint for reduction of the rotatory dislocation, particularly in neurologically intact patients, make surgery for this problem difficult and controversial. The proposed surgical procedure necessitates fixation of the joint. A technique that will reduce dislocation and permit movements of the atlantoaxial joint needs to be identified.

References

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# Trauma, Degenerative Disorders, and Infections

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The craniovertebral junction (CVJ) is formed by three main bony structures: the basiocciput and the first and second cervical vertebrae. The relations among these bony elements are constant because of the complex array of associated ligaments. The ligaments are the main structures responsible for stability in this region. Maintaining the appropriate alignment is important in protecting the upper spinal cord and medulla and both vertebral arteries.

This chapter covers the traumatic conditions that most often affect the CVJ and related surgical treatments. A thorough understanding of the local anatomy and biomechanics of this region is needed to treat the pathology in this location effectively.

### Anatomy and Biomechanics

The occipital condyles articulate with the lateral masses of C1 in cup-shaped joints that facilitate motion primarily for flexion and extension. This joint is the largest contributor to flexion and extension from any single motion segment in the cervical spine, but its role during axial rotation and lateral bending is minimal. C1 rests on top of the C2 facets, which resemble relatively flat shoulders with a lateral and inferior slope. The configuration of C1 on top of C2 predicts and facilitates its axial rotation and minimizes lateral bending and flexion-extension at this joint. The second cervical vertebra is also known as the axis. Its vertical portion, the odontoid process, which is an essential part of this complex, forms the pivot point about which the majority of the axial rotation of the head occurs.

This complicated anatomical configuration is kept in functional alignment by the complex associated ligamentous array (Fig. 32.1). The cruciate ligament consists of two portions, one vertical and one horizontal. Its fibers are interwoven in a cross. The vertical segment extends from the posterior aspect of the body of C2 and attaches rostrally on the anterior aspect of the foramen magnum. The horizontal band corresponds to the transverse ligament, which has two lateral insertions on each side of the medial aspect of the lateral mass of C1 and another insertion on its own synovial joint behind the odontoid process. The transverse ligament serves as a “seat belt” to prevent the odontoid process from posterior translation.

The paired alar ligaments run from the lateral and superior aspect of the odontoid process and fan out to the inferior aspect of each occipital condyle. Their main function is to stretch simultaneously when the head rotates to either side, thereby preventing excessive axial rotation at the occiput–C1–C2 complex.1 The apical ligament extends from the tip of the odontoid process and fans out to the anterior rim of the foramen magnum. Its main function is to keep the atlas under compression between the axis and the occipital bone. Some doubt its importance because its presence is inconsistent.2
The function of a second group of ligaments is less clear. The tectorial membrane, the most rostral extension of the posterior longitudinal ligament (PLL), inserts on the anterior rim of the foramen magnum. Its main function is to serve as a restraint during flexion of the head. The main function of the cranial extension of the anterior longitudinal ligament (ALL), known as the anterior atlanto-occipital membrane, is to restrain hyperextension of the head.

In summary, the stability of the CVJ primarily is imposed by a complex array of ligaments that apply high tension on the occiput–C2 complex to maintain its integrity while still allowing a wide range of motion.

Traumatic injuries of the CVJ need to be studied and evaluated for treatment as a group. From a methodological perspective, it is useful to organize CVJ injuries as primary bony injuries, primary ligamentous injuries, and combined injuries involving both ligaments and bones.

### Bony Injuries

#### Condylar Fractures

Anderson and Montesano classified condylar fractures into three types: type I comminuted fractures, type II skull base fractures that extend into the occipital condyle, and type III avulsion fractures (Fig. 32.2). In general, types I and II are considered stable fractures, which may heal with treatment by cervical immobilization (collar) alone. Type III fractures compromise the ligaments. They are usually unstable and require rigid immobilization.  

#### Atlas Fractures

A wide variety of fractures can occur at C1 (Fig. 32.3). The key factor that determines treatment is the integrity of the transverse ligament. In general, most C1 fractures heal satisfactorily with immobilization. Cervical collars provide minimal immobilization at the CVJ and should be used in cases of nondisplaced fractures (Fig. 32.4). Mildly displaced fractures require sterno-occipital mandibular immobilization (SOMI) or a halo brace, as do severely comminuted fractures, which can render the transverse ligament physiologically incompetent. In such cases, a halo brace or surgical fixation is indicated. Patients treated in a halo brace should be followed closely. If fusion fails to occur, they should undergo surgical stabilization. Figure 32.4 compares the range of motion of various cervical orthoses.

#### Axis Fractures

C2 fractures can be classified as odontoid fractures, hangman’s fractures, and miscellaneous fractures (Fig. 32.5).
Fig. 32.3a–g  There are a variety of atlas fractures.

a The four-part ring or burst fracture is classically referred to as a Jefferson fracture.

b The comminuted lateral mass fracture is extremely common. It creates C1–C2 instability by rendering the transverse ligament incompetent because it detaches the tubercle for insertion of the transverse ligament. More stable patterns of injury include (c) the unilateral ring fracture, (d) linear lateral mass fracture, (e) posterior ring fracture, (f) anterior arch fracture, and (g) contralateral ring fracture. The major determinant of stability of these injuries is whether the transverse atlantal ligament is anatomically and physiologically intact. (Used with permission from Barrow Neurological Institute.)
Type I odontoid fractures occur when the tip of the odontoid process is avulsed. This type of fracture is rare and usually heals well when treated with a semirigid orthosis, such as a Minerva brace. Type II odontoid fractures occur at the base of the dens and involve minimal displacement (<6 mm). Their fusion rate ranges from 85% to 90% when treated with a halo brace. Fractures associated with >6 mm of displacement have an 80% chance of nonunion if treated only with an external orthosis. Consequently, early surgical fixation is recommended. Type III odontoid fractures compromise the vertebral body of C2 and usually heal with treatment in a halo brace. These patients must be closely followed. If there are no signs of fusion after 3 months of immobilization, surgery should be considered.

Displaced acute type II odontoid fractures can be treated with odontoid screw fixation. This technique requires the transverse ligament to be intact. If the ligament is disrupted, posterior C1–C2 fusion is indicated. This procedure requires visualization of the odontoid process in lateral and anteroposterior projections, which is facilitated by using two C-arms (Fig. 32.6). When available, frameless stereotactic image guidance can be useful.

Odontoid screw fixation includes a standard anterior cervical exposure of the C2–C3 interspace from the anterior cervical incision at C4–C5 (Fig. 32.7). A pilot hole is drilled at the base of C2 (Fig. 32.8), which is cannulated across the fracture with a Kirschner (K) wire. Next, a cannulated drill is used to insert a partially threaded screw to obtain a lag effect. Sasso et al. found that the stability of fixation with either one or two screws is surprisingly equivalent.

We prefer a single cannulated lag odontoid screw for the fixation of type II odontoid fractures. This technique has two primary advantages: it provides immediate...
reduction, and fusion of the odontoid process does not affect the axial rotation of C1–C2. During exposure, a tubular retraction system can be used to retract soft tissue."}

### Combined C1–C2 Fractures

Combined fractures should be analyzed individually. In general, however, combined injuries can be managed with a halo brace except when the transverse ligament is disrupted or when a type II odontoid fracture with > 6 mm of displacement is present. For such cases, early surgical fixation should be considered.

### Ligamentous Injuries

Occipitoatlantal dislocation (OAD) and atlantoaxial dislocation (AAD) are well-documented, devastating injuries that are usually fatal. In both cases, enormous distractive energy is applied and pulls the head vertically. In the case of OAD, this energy separates the skull base from C1. In the case of AAD, C1 is separated from C2. In the worst-case scenario, a combined OAD and AAD injury, C1 is isolated from the occiput and from C2 (Fig. 32.9). How the CVJ is distracted at two different levels by the same type of mechanism is unknown, as is the site at which the energy of the injurious force is absorbed.

The complex ligamentous array at the CVJ keeps C1 sandwiched between the occiput and C2. During pure
Fig. 32.6  Organization of the operating room showing the relative positions of the patient, personnel, and dual-image intensifier. (Used with permission from Barrow Neurological Institute.)

Fig. 32.7a, b

a  The patient is positioned supine with the head and neck extended to facilitate placing the screw into the tip of the dens. A transverse incision is made over the C4–C5 interspace to provide a horizontal trajectory, parallel to the anterior surface of the spine.

b  The appropriate screw angle with respect to the patient’s chest, as shown, cannot be achieved in a patient with a short neck or barrel chest. (Used with permission from Barrow Neurological Institute.)
axial distraction of the CVJ, different types of injury will manifest, depending on where the vertical portion of the cruciate ligament is sectioned (Fig. 32.9). In both OAD and AAD, the transverse ligament is likely intact because the injurious force is distractive along the spinal axis. These patients seldom show evidence of anteroposterior subluxation. If all the vertically compressive ligaments (apical, alar, and tectorial membrane) are sectioned, and the vertical portion of the cruciate ligament is ruptured above the transverse ligament, the result is OAD (Fig. 32.9). If the articular capsules between C1 and C2 and the vertical segment of the cruciate ligament inferior to the transverse ligament fail, distraction would occur at C1–C2. The result would be AAD instead of OAD.

We hypothesize that if lesions occur both above and below the transverse ligament, a combined OAD/AAD injury or OAD alone will occur (Fig. 32.9).

■ Occipitomentatal Dislocation

OAD is a devastating injury that very few individuals survive.11,12 Multiple landmarks are used to establish this diagnosis accurately. A Power’s ratio > 1 is one of the most sensitive, objective measurements used to identify this type of injury.11,13 Unfortunately, no radiographic method is completely accurate for detecting OAD.14

The diagnosis of OAD in children is more challenging than in adults. On sagittal and coronal CT reconstructions, Pang et al. measured the distance between the occipital condyle and the lateral mass of C1 in normal children. A separation > 2 mm was highly suggestive of OAD.15,16 If the distance is > 4 mm, OAD is confirmed.15,16

■ Treatment

OAD implies a severe ligamentous disruption that will not heal by itself;17 immediate reduction and internal fixation are required. The ideal fixation system would provide enough support while compromising only the involved segments (occipitomentatal joint). Fixation should be immediate, and no hardware would be inserted into the spinal canal, which can itself be abnormally compressed.18

Different systems and techniques have been described for fixation of the CVJ. These systems can be
classified into two broad groups: techniques that use wires and those that involve plates and screws. Wiring and screw techniques imply anchorage to the suboccipital surface connected to a rod and plate secured to the spine, typically with sublaminar wiring. A contoured braided loop is a simple, inexpensive, and useful resource for fixating the CVJ to the desired level (Fig. 32.10). However, the laminae of the involved levels must be intact so that the wires can be tightened around them. Several techniques incorporate plates and screws. Screws can be inserted into the occipital bone, or epidural “washers” can be connected to a frame anchored to the spine with transarticular (C1–C2) screws. Occipital screws risk penetration of the inner table of the calvarium or the dura.

Biomechanically, screw fixation is superior to fixation with wire or cable. After numerous repetitive cycles, wire fixation tends to lose strength from fatigue. Unlike cables or wires, which fixate the bone through a cerclage effect, screws rigidly engage bone. However, their efficacy depends on the quality of the bone in which they are inserted because the bone–screw interface is the weakest point of screw fixation.
Fig. 32.10b–e

- **b** Bender used to create a uniform U shape. It is also used to bend the rod to the desired curvature to contour to the shape of the CVJ.
- **c** Close-up of the rod bender shows the three different aperture orifices that can be rotated to bend the contoured rod into the U shape.
- **d** Contoured Steinmann pin affixed to the CVJ with laminar wires and to the skull through burr holes.
- **e** Example of the bone graft placed under compression between the occiput and C2 and wired to secure its position.

(From Apostolides PJ, Sonntag VKH, Dickman CA. Occipitocervical wiring techniques. In Dickman CA, Spetzler RF, Sonntag VKH, eds. Surgery of the Craniovertebral Junction. New York: Thieme; 1998:800. d and e Used with permission from Barrow Neurological Institute.)
In the case of pure OAD with no other superimposed injuries, the best available treatment from a biomechanical perspective is fixation and fusion of the compromised segment. That is, the occipitoatlantal joint could be fixed with transarticular screws crossing the occipitoatlantal joint (C1–occiput) (Fig. 32.11) or with a C1 lateral mass screw and interconnected with a rod fixed to a plate affixed to the skull with screws (Fig. 32.12). Because the thickest part of the suboccipital bone is involved, we prefer systems that use keel screws. These techniques preserve the normal range of motion during flexion-extension and axial rotation at C1–C2.

Occipitoatlantal transarticular screws were first reported by Grob in 2001. These screws provide direct rigid screw fixation of the occipitoatlantal joints bilaterally. This technique is based on a principle similar to that underlying the Magerl technique for C1–C2 transarticular screw fixation.

At our institution, the anatomy and biomechanics of occipitoatlantal fixation with transarticular screws were investigated in terms of the feasibility and safety of inserting screws in bony structures in relation to the surrounding vascular structures (vertebral arteries), hypoglossal nerves, and spinal cord.

The occipitoatlantal joints must be realigned intraoperatively. The gap should be reduced. This reduction can be performed preoperatively by placing the halo under compression. The entry point of the screws is the midpoint of the C1 lateral mass posteriorly under the sulcus arteriosus of C1 (the same entry point for C1 lateral mass screws). The trajectory of the screw is oriented rostrally in the sagittal plane through the atlanto-occipital joint into the occipital condyle. The target on a lateral radiograph projection is an imaginary point 1 cm rostral to the anterior arch of C1. The trajectory of the screw is positioned to avoid the hypoglossal canal, spinal canal, and vertebral arteries. The fixation is performed with a cannulated screw technique using a K wire to anchor the dislocated bone followed by drilling and placement of the screw to fixate the bones directly. The technique is performed under fluoroscopic guidance. Intraoperative navigation and intraoperative computed tomography (CT) can also be used. A piece of autologous iliac crest bone graft is wired to the surface of the occiput into the posterior arch of C1 as in a C1–C2 fusion.

The disadvantages of transarticular screw fixation include the potential to injure the vertebral artery in the sulcus arteriosus.

An alternative to fusing the occiput–C1 complex is a technique that uses C1 lateral mass screws interconnected with rods to a plate affixed to the skull through keel screws. This construct provides good stability but to a lesser extent than transarticular screws, especially during extension and lateral bending.

Appropriate preoperative measurement of the occipital keel is key to preventing cerebellar hemorrhage, epidural hematomas, and leakage of cerebrospinal fluid.

Fig. 32.11a–c The C1–occipital transarticular screw fixation.
- **a** Lateral projection shows the trajectory and purchase into the occipital condyle.
- **b** Coronal view shows the medially tilted trajectory with an entry point just under the sulcus arteriosus. The bone graft is left under compression between C1 and the suboccipital surface.
- **c** Sagittal CT reconstruction of a patient with an isolated OAD shows the screw trajectory and its position just below the hypoglossal foramen. (Used with permission from Barrow Neurological Institute.)
**Atlantoaxial Dislocation**

The mechanisms underlying AAD and OAD have many similarities. In both cases, the transverse ligament is undamaged (Fig. 32.9). Based on the position and function of the cruciate ligament, damage to its vertical portion is unavoidable in a vertical distraction injury. The vertical portion is disrupted below the transverse portion. Intact fibers keep C1 apposed to the skull base, and an isolated C1–C2 vertical distraction injury results.

Vertical instability at C1–C2 has been attributed to severe ligamentous injury without evidence of major fracture (Fig. 32.13). The amount of energy required to distract the C1–C2 complex is tremendous: Complete anatomical section of the spinal cord has even been reported. Acute severe and devastating neurological damage reflects injury to the associated cranial nerves, long tracts, or vascular structures (primarily to the vertebral arteries with subsequent thrombosis). On magnetic resonance imaging (MRI), short tau inversion recovery (STIR) (Fig. 32.14) sequences may show increased signal intensity at the atlantoaxial joint. This finding supports the hypothesis that C1–C2 vertical distraction and AOD are part of the same spectrum of injuries that share a common distractive mechanism.

The diagnosis of AAD represents a challenge. Radiological evaluation of the craniovertebral junction is primarily directed toward the identification of fractures and not ligamentous injuries. Yet OAD and AAD occur after the disruption of ligaments. Orthogonal reconstructions, including coronal and sagittal views of CT scans, are required to identify vertical distraction. We have found that the most reproducible characterization of the C1–C2 articulation in the vertical plane is the lateral mass interval (LMI), which measures the distance from the lateral mass of C1 to the lateral mass of C2. An LMI > 2.6 mm in the coronal plane is highly indicative of AAD. The separation can be confirmed on MRI. 24

MRI serves to verify vertical distraction injuries suspected from CT. STIR sequences are very sensitive and demonstrate blood or fluid distributed within the C1–C2 joint space, especially when sagittal and coronal reconstructions are performed. Imaging the coronal plane shows signal abnormalities distributed throughout the C1–C2 joint space to best advantage.

**Treatment of C1–C2 Instability**

Because vertical AAD and OAD share features, they should be treated as part of the same spectrum of distracting injuries. Measures that reproduce the mechanism of trauma, such as cephalic traction and collars with extension, should be avoided to prevent neurological worsening or the appearance of new deficits.

Isolated AAD (e.g., without additional fractures or OAD) should be treated with C1–C2 fusion. Either C1–C2 transarticular screws or C1 lateral mass and C2 pedicle/pars screws with interspinous bone graft and wiring should be used.
Theoretically, C1–C2 pedicle screws connected with an adjustable rod can be used to correct the distraction. Partially threaded, C1–C2 transarticular screws are ideal for providing a lag effect to reapproximate C2 to C1 and can be used with posterior fusion and wiring as was done in our patient.

**C1–C2 Transarticular Screws**

Described by Magerl, C1–C2 transarticular screw fixation provides immediate fixation compromising only the affected levels. The patient’s head is fixated in a Mayfield head holder (Integra Neurosciences, Plainsboro, New Jersey).
with the neck flexed. Alignment is verified with real-time fluoroscopy before the head is positioned in slight flexion.

Subperiosteal dissection is performed exposing the C2–C3 facet, the pars interarticularis of C2, and the posterior arch of C1. A pilot hole is drilled 2 to 3 mm lateral to the medial aspect of the C2–C3 facet joint (Fig. 32.15a). The appropriate trajectory is identified on a lateral radiograph by using the anterior tubercle of C1 as the target (Fig. 32.15b). A K wire is inserted under fluoroscopic guidance with a slight medial trajectory (0–10°). It is important to dissect the medial aspect of the C2 pars so that it is available as a landmark. Once the K wire is introduced and has pierced the C1–C2 joint, a cannulated drill is used, followed by insertion of a cannulated screw. The surgical assistant must hold the proximal wire so that it does not advance inadvertently. Once the screws are in position, a braided cable wire is introduced and passed under C1, then under the spinous process of C2. The wire holds the bone graft in position while fusion develops.

Adequate preparation of the bone graft is key to obtaining fusion. The graft is carved carefully and positioned under compression between C1 and C2. A single wire is passed under the lamina of C1, with the loop embracing the spinous process of C2 and secured over the bone graft (Fig. 32.16).

Potential complications can be minimized with careful preoperative planning and measurement of the appropriate distances for screw trajectories. Although biomechanically superior to the C1 lateral mass, placement of transarticular
screws in the C2 pars can be contraindicated by anatomy. Paramore et al.\textsuperscript{26} found that up to 23\% of at least one side of C2 vertebral bodies were inappropriate for transarticular screw placement because of high-riding transverse foramina. If the patient’s anatomy precludes inserting a screw into the C2 pars, the construct can be extended inferiorly into C3, skipping C2\textsuperscript{27} on one side or bilaterally.

### C1 Lateral Mass and C2 Pars Interarticularis Fixation (Goel’s Technique)

This technique is indicated for C1–C2 instability in patients with an anomalous vertebral artery that precludes placement of a transarticular screw (~20\% of the population).\textsuperscript{157} This screw fixation technique provides rigid internal fixation of the atlantoaxial joint and is biomechanically equivalent to transarticular screws.\textsuperscript{28} Several clinical series have reported excellent fusion outcomes with this technique, which is combined with the placement of autologous bone graft to encourage arthrodesis.\textsuperscript{29}

The screws are positioned in the bone as follows. The dorsal aspect of the C1 posterior arch is dissected. The dissection is extended inferior to where the sulcus arteriosus meets the lateral mass of C1 (Fig. 32.17). A large...
venous plexus usually covers the lateral mass of C1. Bleeding is easily controlled with Nu-Knit (Johnson & Johnson, Arlington, Texas) or Flo-Seal (Baxter, Deerfield, Illinois). A pilot hole is drilled into the lateral mass along a slight superior and medial trajectory (< 10°). Screws are inserted into the lateral mass in a unicortical fashion centered within the lateral mass. The medial aspect of the lateral mass of C1 is used as a reference landmark. If available, frameless stereotactic guidance is useful. Screws are inserted into the C2 pars interarticularis. After the medial aspect of C2 pars has been identified as a reference, a pilot hole is drilled 1 to 2 mm lateral to the C2–C3 joint. The hole is tapped, and the screw is then inserted. Fluoroscopic imaging is used to direct the screw toward an imaginary line posterior to C2. The screw fixation procedure can be performed unilaterally or bilaterally. If the anatomy of the contralateral joint is suitable, a transarticular screw can be placed on that side. Autologous graft is wired between the posterior arches of C1 and C2 to provide long-term stability to the construct.

### Combined Occiput–C1 and C1–C2 Injuries

Patients who survive this devastating injury can undergo occipitocervical fusion with any of the methods described above. OAD and atlantoaxial vertical distraction share the same physiopathology of severe injury at the CVJ, and few survive the injuries. We hypothesize that these two injuries occur because the vertical injurious force manifests as a traumatic lesion at different levels of the same ligamentous complex.

If C1 is unattached to the occipital condyles and C2, occipitocervical fixation that includes the subaxial cervical spine as part of the construct would be required. The potential for accelerating degenerative disease is present at the uncompromised levels; however, such a construct increases the fixation in the construct, thereby improving the chances of fusion. We have described the simultaneous use of occiput–C1 and C1–C2 transarticular screw fixation in a patient with a combined injury. 50

### Transverse Ligament Injuries

As discussed above, the transverse ligament serves as a seat belt for the odontoid process, preventing the odontoid from posterior axial translation and potential compression of the upper spinal cord. Incompetence of the transverse ligament is suspected when the atlantodental interval (ADI) is > 5 mm in children or > 3 mm in adults. A separation between the lateral mass of C1 and the odontoid process > 7 mm in the coronal plane is highly suggestive of failure of the transverse ligament, a relationship known as Spence’s rule. 31 The ligament can fail at different locations (Fig. 32.18). The sites of failure serve as the basis for classifying injuries and have therapeutic implications. In type I injuries, the ligament ruptures from within itself. Because ligaments heal poorly, 13 such injuries usually require surgical fixation. Type II injuries occur when the ligament is intact, but the bony tubercle on the lateral mass of C1 is fractured. These injuries can occur as part of a C1 comminuted lateral mass fracture or as a teardrop avulsion of the ligamentous insertion onto the C1 lateral mass. Bone-to-bone interface is present in this type of injury. Therefore, the likelihood of healing with external rigid fixation (i.e., in a halo brace) is ~80%. Surgery remains an option for patients who fail halo fixation after 3 months of immobilization. The treatment options for C1–C2 fixation are the same as described above.

On axial gradient echo MRIs, the transverse ligament appears as a continuous low-intensity signal band that spans from one tubercle to the other (Fig. 32.19). 32

### Selection of Fixation for CVJ Fusion

For each of the ligamentous and bony injuries described above, it is possible to fixate the occipitoatlantal joint, atlantoaxial joint, both joints, or both joints plus adjacent lower cervical motion segments to achieve bony fusion using a variety of surgical hardware constructs. In general, better immobilization of the joint(s) provides a better environment for fusion, although the optimal degree to which motion must be eliminated is unknown.

Fusion of the occipitoatlantal joint without fusion of the atlantoaxial or more caudal joints is feasible by at least two techniques: transarticular screws and occipital keel–C1 lateral mass screws–rods. However, especially as part of a wired construct, isolated short segment fixation limits motion less than when the segment is wired within a longer construct of the same components (Fig. 32.20). Screw constructs tend to provide better immobilization in most directions of loading than wired constructs, but often the screws provide a pivot point about which motion restriction is less effective, for example, poor resistance to flexion-extension by occipitoatlantal transarticular screws. Addition of a wired structural graft overcomes this deficiency and effectively prevents flexion and extension.

Fusion of the atlantoaxial joint without fusion of the occipitoatlantal joint or lower cervical spine can be achieved by several screw constructs, all of which provide better resistance to motion than a wired interspinous graft (Fig. 32.21). Addition of an interspinous graft to a construct, especially isolated constructs at C1–C2, in all cases provides some improvement in resistance to flexion and extension, although the improvement in resistance to other motions is often unremarkable. Because of the near equivalence of biomechanics among techniques, selection of the appropriate screw construct
Fig. 32.18  Classification of injuries to the transverse atlantal ligament. Type I injuries disrupt the ligament substance in its medial portion (type IA) or at its periosteal insertion (type IB). Type II injuries disconnect the tubercle for insertion of the transverse ligament from the C1 lateral mass involving a comminuted C1 mass (type IIA) or avulse the tubercle from an intact lateral mass (type IIB). (Used with permission from Barrow Neurological Institute.)

Fig. 32.19a, b

a  On a gradient echo MRI, a normal transverse ligament appears as a low-intensity band.
b  MRI shows a torn transverse ligament near its insertion on the lateral mass of C1.
(From Dickman CA, Greene KA, Sonntag VKH. Traumatic injuries of the craniovertebral junction. In: Dickman CA, Spetzler RF, Sonntag VKH, eds. Surgery of the Craniovertebral Junction. New York: Thieme; 1998:182. Used with permission from Thieme.)
may largely be a decision based on the surgeon's comfort level (perceived risk) with the procedure, anatomical suitability of the procedure for each patient, and compatibility of the hardware with other implanted components.

**Conclusion**

Even the most rigid fixation construct will fail if bony fusion does not occur. Requirements for arthrodesis include rigid immobilization, a vascularized bed to promote graft incorporation, and autogenous bone graft. The iliac crest, rib, and calvarium have been used to promote fusion at the CVJ.

Imaging of the CVJ has improved dramatically with advances in CT and MRI and the availability of multiplanar reconstructions and three-dimensional models. It is likely that the incidence of pathology found at this location is increasing at least partially because of the improvements in imaging. Traumatic injuries to the CVJ should be considered part of a full-spectrum understanding of the close relations among the occiput, C1, and C2 and their interconnecting ligaments. Treatment should be individualized to reflect factors such as age, comorbidities, the local anatomy, and the surgeon’s experience.
Fig. 32.21 Mean angular range of motion of the atlantoaxial joint in various directions of loading in human cadaveric spine specimens under an applied load of 1.5 Nm using nonconstraining pure moments without compressive follower loads. Results are compiled from various studies performed at the authors' institution evaluating surgical fixation. Error bars show standard deviations. (Used with permission from Barrow Neurological Institute.)

References


Rheumatoid arthritis (RA) is a chronic, relapsing, inflammatory autoimmune disorder characterized by symmetrical erosive synovitis of multiple peripheral joints with a varying degree of systemic involvement. RA is relatively common, affecting 0.8% to 2.0% of the world’s population and 2 million people in the United States alone. The cervical spine is the second most frequently affected region (after hands and feet). Women are affected more often than men (3:1), although men have a greater risk of advanced cervical spine involvement.

Garrod first described RA of the cervical spine as a clinical entity. The prevalence of cervical spine involvement in RA ranges from 25% to 80%, depending on the diagnostic criteria applied. Three types of deformities occur commonly in rheumatoid cervical spine. Atlantoaxial subluxation (AAS) or instability is the most common type, accounting for nearly 65% of the deformities (Fig. 33.1). Rostral migration of the dens (basilar invagination/cranial settling) is the second most common deformity, seen in 20% of patients with RA (Fig. 33.2). Subaxial subluxation is the third most common pathology, occurring in 15% of patients (Fig. 33.3).

Forty to 85% of patients with RA have neck pain. Radiographic evidence of instability is observed in a similar percentage of patients, although only 7% to 13% of them develop neurological deficits. Kauppi and Hakala found that in a Finnish population-based series of 98 patients with RA (mean disease duration 11 years), 33% of patients presented with AAS and 27% with atlantoaxial impaction (AAI). Pellicci et al. in 1981 reported that, although 80% of their patients with RA and cervical spine involvement demonstrated radiographic progression, 36% had neurological progression. Fifty percent of patients with radiographic instability were asymptomatic in the series by Collins et al. Approximately 10% of patients with RA die of unrecognized spinal cord or brainstem compression.

The mutilating disease subset of patients with RA, as classified by Olerud et al., usually have global involvement of the cervical spine with associated AAS, subaxial sub-
luxation, and vertical subluxation. The natural course of the disease and the survival rates for this subset are also very discouraging, as shown in Omura’s series, where all patients (n = 6) with the mutilating disease type of RA who were managed conservatively died within 3 years.11

Predictors of Cervical Spine Involvement

No single factor is entirely predictive of progressive cervical spine disease in RA. Nevertheless, disease duration, rapid joint erosiveness, elevated C-reactive protein levels, seropositivity, HLA-DRB1 (human leukocyte antigen–DR beta 1) susceptibility, and severe peripheral joint involvement, including arthritis mutilans, are, in general, suggestive of a more aggressive disease process.12

It is important to understand the various classification systems used in the literature to grade functional capacity, neurological deficit, and pain in patients with craniovertebral junction/upper cervical spine pathology. With these schemes, the outcome from the variety of treatment modalities can be compared in a standardized fashion. The common classification schemes are listed in Tables 33.1, 33.2, 33.3, and 33.4.

Table 33.1 American Rheumatism Association classification of functional capacity

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<th>Class</th>
<th>Description</th>
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<tr>
<td>1</td>
<td>Complete: ability to perform all usual activities without handicap</td>
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<tr>
<td>2</td>
<td>Adequate: ability to perform some activities despite handicap, discomfort, or limited motion at one or more joints</td>
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<tr>
<td>3</td>
<td>Limited: little or no activities of usual occupation or self-care</td>
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<td>4</td>
<td>Incapacitated, largely or wholly: bedridden or confined to a wheelchair; little or no self-care</td>
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Table 33.2 Ranawat classification of neurologic deficit

<table>
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<tr>
<th>Class</th>
<th>Description</th>
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<tbody>
<tr>
<td>I</td>
<td>Pain, no neurologic deficit</td>
</tr>
<tr>
<td>II</td>
<td>Subjective weakness with hyperreflexia and dysesthesia</td>
</tr>
<tr>
<td>IIIa</td>
<td>Objective weakness, long tract signs, but ambulatory</td>
</tr>
<tr>
<td>IIIb</td>
<td>Nonambulatory and quadriparetic</td>
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Pathomechanics of the Rheumatoid Cervical Spine

The cervical spine, in particular, is affected in RA because of the large number of articulations and their significant mobility. Inflammation of the synovial membrane leads to pannus formation (overgrowth of hyaline cartilage and periarticular inflammation) (Fig. 33.4), which in turn results in bone erosion and synovial cysts. This then leads to joint laxity and subluxation. In the cervical spine, it affects the atlantoaxial joint, periodontoidal ligaments, facet (zygo-apophyseal) joints, uncovertebral joints, retrodental bursa, interspinous ligaments, and ligaments around the atlas. The craniovertebral junction (CVJ) often becomes unstable because of the regional dependence on ligamentous structures for stability. The resulting

Table 33.3 Ranawat scale of pain in rheumatoid arthritis

<table>
<thead>
<tr>
<th>Level</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>0</td>
</tr>
<tr>
<td>Mild</td>
<td>1 (requiring aspirin)</td>
</tr>
<tr>
<td>Moderate</td>
<td>2 (requiring cervical collar)</td>
</tr>
<tr>
<td>Severe</td>
<td>3 (pain not relieved by either aspirin or collar)</td>
</tr>
</tbody>
</table>

hypermobility at occiput–C1 and C1–C2 is due to damage to the cruciform-alar ligament complex, the transverse ligament, and the joint capsules. Degenerative arthritic changes are superimposed on the CVJ, which remains hypermobile. Asymmetric wear results in the superimposition of degenerative arthritis on the region, especially the atlantoaxial articulation. This results in severe AAS, rotatory subluxation, dorsal and lateral AAS, craniocervical scoliosis, and basilar invagination. The atlas often slips over the axis in a ventral-caudal direction and causes a greater upward movement of the dorsal arch of the atlas, resulting in an increased anteverted C1–C2 kyphosis angle. The asymmetrical wear and subluxation ultimately result in the formation of a hypertrophic cicatrix, described as pannus. The pannus itself may be compressive, especially when it is magnified by coronal or sagittal plane malalignment at the CVJ. If these degenerative changes are symmetrical, the patient experiences symmetrical basilar invagination. When only one lateral mass is affected by the disease process, fixed rotational tilt of the head to the affected side may occur.

The most common deformity is AAS, representing 60% to 65% of rheumatoid cervical subluxations. The majority are ventral, 20% are lateral, and 10% are posterior. Posterior AAS is typically associated with erosion or fracture of the dens, and although this is thought to be more benign than anterior AAS, it actually carries a higher risk of spinal cord compression due to less restricted backward translation of the free-floating atlas.

In contrast, in the subaxial spine, the uncovertebral joints are the predominant focus of the inflammatory process. Disk collapse and autofusion/ankylosis are typical. Subluxation and pannus formation occur later, as the ligaments become involved. Subaxial subluxation usually occurs late in the course of the disease and tends to affect multiple vertebral levels, sometimes causing a classic “stepladder” or “staircase” deformity.

**Cranial Settling or Vertical Translocation**

Cranial settling is also termed basilar invagination, atlantoaxial impaction, or odontoid vertical migration. Horizontal AAS almost always precedes vertical translocation, usually by 6 years or so. If chronic inflammation affects both atlantoaxial facet joints, their cartilage and bony surfaces may be destroyed. The weight of the skull then forces the atlas downward over the axis, causing cranial settling (Figs. 33.2 and 33.5). Vertical translocation, or

<table>
<thead>
<tr>
<th>Grade</th>
<th>Radiculopathy</th>
<th>Myelopathy</th>
<th>Gait</th>
<th>Hand Function</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Present</td>
<td>Absent</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>I</td>
<td>Present</td>
<td>Present</td>
<td>Normal</td>
<td>Slight</td>
</tr>
<tr>
<td>II</td>
<td>Present</td>
<td>Present</td>
<td>Mildly abnormal</td>
<td>Functional</td>
</tr>
<tr>
<td>III</td>
<td>Present</td>
<td>Present</td>
<td>Severely abnormal</td>
<td>Unable to button</td>
</tr>
<tr>
<td>IV</td>
<td>Present</td>
<td>Present</td>
<td>With assistance only</td>
<td>Severely limited</td>
</tr>
<tr>
<td>V</td>
<td>Present</td>
<td>Present</td>
<td>Nonambulatory</td>
<td>Useless</td>
</tr>
</tbody>
</table>

ascent of the odontoid peg, is primarily due to C1 lateral mass collapse and may also be partly due to destructive changes in C2 and occipital condyles. Casey et al. reported from Crockard’s series that the physical constraint of the ring of the atlas as it slides down the axis leads to decreased mobility, thus in turn leading to decreased pannus formation and a decreasing atlantodens interval (ADI). Diminishing ADI thus should not be considered a sign of amelioration of the patient’s condition. In fact, it suggests increasing vertical translocation, heralding an increased bony compression of the brainstem and spinal cord. This may be associated with a shortened dens because of the erosive process affecting C2.

**Histology of Affected Joints**

Synovitis with fibrinoid changes of the cervical spine is similar to that of peripheral joints. The transverse ligament becomes insufficient because of inflammatory erosion of the dorsal surface of the dens granulation tissue originating in the synovial joints between the transverse ligament and the dorsal surface of the dens. This loss of tensile strength and stretching of the transverse ligaments initiates AAS. A similar inflammatory response of the synovium may affect the uncovertebral joints, retrodental bursa, facet joints, ligaments around the atlas, and even the intervertebral disk if invaded by synovium. O’Brien et al., in their histological analysis of tissue obtained from transoral resection of the dens, identified two types of synovium: type I, chronic active rheumatoid synovium, and type II, end-stage rheumatoid synovium. The latter was associated with decreased spinal cord cross-sectional area, myelopathy, and greater evidence of craniocervical junction osseous destruction. This suggests that ligamentous destruction is followed by replacement of the rheumatoid synovium with fibrous tissue, whereas the osseous structures reveal severe destruction secondary to mechanical stability, rather than to an acute inflammatory process. Therefore, it appears that mechanical degeneration rather than an inflammatory rheumatoid process is responsible for the deteriorating neurological picture in chronic RA at the CVJ.

**Imaging in Rheumatoid Arthritis of the Spine**

**Cervical Spine Radiographs**

Radiographs are usually the first line of imaging used to assess the degree of cervical spine involvement from RA. Neutral views, in combination with flexion/extension and rotation, are helpful in identifying and measuring atlantoaxial subluxation (Fig. 33.6), basilar invagination, and subaxial subluxation, as well as for a rough assessment of the extent of osteoporosis. Various measurements, such as ventral ADI, dorsal ADI (Fig. 33.7), and the space available for the spinal cord, can be determined from radiographs, although none are reliable to predict neurological deterioration or to assess the extent of spinal cord compression.

Indications for anteroposterior and lateral radiographs of the cervical spine in patients with RA include:

- Prolonged cervical symptoms > 6 months
- Neurological signs or symptoms
- Scheduled procedures requiring endotracheal intubation
- Rapidly progressive carpal or tarsal bone destruction
- Rapid overall functional deterioration

**Evaluation of Basilar Invagination**

**McGregor Line**

The McGregor line is drawn on a lateral plain radiograph from the hard palate to the base of the occiput. Vertical settling of the occiput is defined as migration of the odontoid > 4.5 mm above the McGregor line (Fig. 33.8).
The odontoid process is divided into three equal parts in the sagittal plane. If the ventral ring of the atlas is level with the middle third or the caudal third of the odontoid process, a diagnosis of basilar invagination is made (Fig. 33.9).

Redlund-Johnell Criterion

The distance between the McGregor line and the midpoint of the caudal margin of C2 is referred to as the Redlund-Johnell criterion. If it measures < 29 mm in women or < 34 mm in men, it indicates basilar invagination (Fig. 33.10).

McRae Line

The McRae line is drawn across the foramen magnum from the basion to the opisthion. A protrusion of the tip of the dens above this line is suggestive of basilar invagination (Fig. 33.11).

Chamberlain Line

The Chamberlain line is drawn from the dorsal edge of the hard palate to the opisthion. If the tip of the dens lies > 3 mm above this line, basilar invagination is suspected (Fig. 33.11).

Wackenheim Line

The Wackenheim line is drawn along the rostral surface of the clivus. A protrusion of the tip of the dens dorsal to this line indicates basilar invagination (Fig. 33.11).

Ranawat Criterion

The Ranawat criterion is measured on lateral plain films. A line is drawn from the pedicles of C2 superiorly along the vertical axis of the odontoid until it intersects a line...
connecting the anterior and posterior arches of C1. If this measures $\leq 15$ mm in men or $\leq 13$ mm in women, basilar invagination is suspected (Fig. 33.12).

It is often difficult to identify the tip of the dens on plain radiographs. Therefore, the aforementioned radiographic criteria that do not rely on the identification of the tip of the dens for diagnosing basilar invagination are more sensitive.

The sensitivity and specificity of these criteria are quite low (70%) if used individually. Riew et al., however, reported that if the Clark station, the Redlund-Johnell criterion, and the Ranawat criterion are positive for basilar invagination, the sensitivity was 94%, with a negative predictive value of 91%, regarding the diagnosis of basilar invagination.

**Computed Tomography**

Thin-cut computed tomography (CT) of the CVJ, with reformatted coronal and sagittal views, has become the standard imaging modality for both the initial evaluation of RA of the spine and postoperative follow-up. It provides a three-dimensional appreciation of the extent of subluxation or instability, the extent of basilar invagination, the nature of ventral compression (osseous vs. soft tissue) (Fig. 33.13), the diameter of the spinal canal, and the space available for the spinal cord. It is an extremely important imaging modality for delineating the relation of the vertebral artery to the pars interarticularis, the definition of bony defects, and bone quality.

**Magnetic Resonance Imaging**

Magnetic resonance imaging (MRI) (Figs. 33.4 and 33.5) gives direct information about the pannus, the nature of ventral compression (whether osseous or soft tissue), the extent of
Fig. 33.13 Sagittal computed tomography (CT) of the cervical spine shows a large retrodental pannus causing spinal cord compression.

cord compression, subarachnoid space obliteration, spinal cord signal changes, and the extent of basilar invagination. Dynamic MRI of the cervical spine in flexion and extension modes has been suggested by some authors, as MRI in the neutral position alone may be misleading with regard to the amount of spinal cord compression. T1- and T2-weighted MR images, along with contrast-enhanced T1-weighted spin echo sequences, are generally required. Hypervascular pannus appeared as high signal intensity on T2-weighted graded images with and without contrast, whereas fibrous pannus showed reduced signal intensity on T2-weighted, gradient-recalled echo images and on pre- and postcontrast T1-weighted spin echo images. Such information regarding the differentiation of fibrous versus hypervascular pannus may be useful, as dorsal fusion alone may not cause the ventral mass of fibrous hypovascular pannus to disappear. Conversely, active pannus and synovitis associated with hypervascular tissue may disappear with dorsal stabilization alone. Goel theorized that retroodontoid pannus is a result of laxity and kinking of ligaments secondary to reduction of joint space and lateral mass collapse. He showed that distraction of the facets resulted in immediate postoperative disappearance of the pannus.

Natural History of Rheumatoid Arthritis of the Craniovertebral Junction

Knowledge of the natural history of the cervical spine and the CVJ in RA is limited by the difficulty in performing long-term clinical follow-up studies. With the advent of the new disease-modifying antirheumatic drugs (DMARDs), spine surgeons encounter far fewer patients with severe rheumatoid involvement. Neva et al. reported objective evidence that rheumatoid AAS can be retarded or even prevented with early and active therapy with DMARDs. Weissman et al. found that spinal cord compression was more common when AAS = 9 mm was present (24% vs. 2%), when there was associated basilar invagination (20.0% vs. 7.3%), and when there was an associated lateral subluxation. Rana reported a retrospective review of 41 patients with AAS followed for 10 years: 61% had no change, 27% had progressive subluxation, and 12% had improvement at final follow-up. Boden et al. retrospectively reviewed the records of 73 patients observed for an average of 7 years. Of the 31 patients (42%) who did not have paralysis (all managed nonsurgically), none developed significant neurological deficit during the observation period. Of the 42 patients (58%) in whom paralysis developed, 35 (48%) were treated operatively. Of the seven patients treated nonoperatively for their paralysis, none improved, and six of seven declined neurologically. All seven died within 4 years, five from spinal cord compression. In contrast, 25 of the 35 patients (71%) who underwent surgery had experienced marked neurological recovery. Crockard and Grob reported that 50% of patients with rheumatoid myelopathy will be dead in a year. Once myelopathy becomes manifest, conservatively treated patients have a much higher mortality rate. The cumulative survival of patients with myelopathy is 28 to 38% at 10 years after surgery. Approximately 10% of patients with RA died of unrecognized spinal cord or brainstem compression. The natural course of cervical lesions in patients with RA with mutilating disease is distressing, with myelopathy and quadripareisis developing within 3 years and death within 7 years in a series of 21 patients reported by Sunahara et al. Therefore, it is evident that although many cases of rheumatoid cervical spine are asymptomatic, careful follow-up examination is necessary to identify patients with early neurological signs.

Clinical Features

Clinical features in RA of the CVJ are highly variable. Pain is the most common clinical feature and is seen in 40% to 80% of patients with RA. It is felt in cervical, temporal, occipital, and retro-orbital regions. Compression of the greater occipital nerve may result in suboccipital pain, and subtle signs of trigeminal nerve involvement may be the first indication of CVJ involvement. Patients often complain of a “clunking” sensation in the neck or feeling as if their head is falling forward on flexion of the neck. Pain is exaggerated with flexion or rotation movements. Neurological symptoms such as paresthesias, anesthesia, loss of proprioception, and lightning shock sensation (Lhermitte sign) on bending of the neck may be seen, but sphincter disturbances are rare and usually occur late in the course of disease. Neurological manifestations are difficult to interpret due to severe peripheral arthritis, loss of joint motion, and muscle wasting or joint pain. Hence, the diagnosis of myelopathy may be delayed significantly (up to 6 months from the first neurological signs). Myelopathy, once it develops, is usually progressive, and even sudden death from medullary or cervical cord compression may occur. In summary, common symptoms of RA affecting the CVJ are nonspecific, and serial examinations are often the best and only objective way to detect subtle changes in the development of myelopathy.
Indications for Surgery
Surgical intervention is clearly indicated in patients with intractable pain and/or symptoms initiating neurological deficits.\textsuperscript{35–37}

Intractable Pain
Pain is the most common surgical referral. In a few cases, medical management fails, and pain may become persistent and intractable. Such pain may be purely axial or include a radiculopathy (usually C2). A cervical collar and traction may be employed. Arthrodesis is indicated if all medical and conservative measures fail. The type of surgery (i.e., fusion alone or with associated decompression) depends on pathology and imaging findings.

Neurological Deficits
Surgery is universally recommended if patients have neurological deficits and are medically fit for intervention. The etiology of deficits may be due to cranial settling, AAS, or retrodental pannus, either independently or in combination. Cervical collars and traction usually are not sufficient.\textsuperscript{38} There is ample evidence in the literature suggesting that prognosis is poor once myelopathy develops.\textsuperscript{16,29} Studies have also demonstrated that patients with more severe neurological deficits have greater morbidity and mortality rates. Hence, early surgical intervention is recommended when deficits are subtle and before more profound neurological decline ensues.\textsuperscript{39,40} Cervical myelopathy and instability, especially if occurring after a decompressive laminectomy or dens resection, are indications for urgent surgical intervention.

It is a challenge to manage patients with clear imaging evidence of cervical instability, but without neurological deficits and pain. These patients form a subset who may be candidates for “prophylactic” surgery to prevent neurological deficits in the future (see discussion below). Surgical intervention, whether prophylactic or not, may be associated with significant morbidity and mortality in patients with RA. A risk–benefit assessment should be performed prior to offering surgery. It is therefore important to identify the predictors of paralysis to determine which patients may benefit most from an early prophylactic surgical procedure.

Predictors of Impending Paralysis in Patients with Cervical Instability

Anterior Atlantodental Interval
Normal anterior atlantodental interval (AADI), or the distance between the dens and the anterior ring of C1, is usually < 3 mm. Although this interval increases with increasing AAS, it can be unreliable for two reasons: patients with even up to 8 to 9 mm of AADI may not be neurologically impaired; and as RA progresses, especially with the development of cranial settling, AADI may, in fact, decrease.

Posterior Atlantodental Interval
Boden et al. showed that the posterior atlantodental interval (PADI), or the distance between the dens and the posterior ring of C1, is a better predictor of impending paralysis. In their study, PADI < 14 mm is predictive of neurological injury.\textsuperscript{29}

Cervicomedullary Angle
Bundschuh et al. found a strong correlation between cervicomedullary angles < 135° and myelopathy and paralysis in patients with RA.\textsuperscript{41}

Space Available for the Spinal Cord
This is measured on MRI. If it is < 13 mm, the spinal cord is compressed, and the patient is at risk for myelopathy.\textsuperscript{42}

Spinal Cord Diameter
If the diameter of the spinal cord is < 6 mm in flexion, surgery is indicated. Basilar invagination in combination with spinal cord compression or severe AAS places the patient at risk for impending paralysis.

Predictors of Surgical Recovery
Age, gender, duration of paralysis prior to surgery, and preoperative AADI do not strongly correlate with postoperative recovery.

Preoperative Deficit
In 116 patients with basilar invagination and myelopathy who underwent surgery, Casey et al. showed that 45% improved by one or more Ranawat grades.\textsuperscript{43} They also demonstrated that patients who improved to Ranawat class I or II had a stable course overall in the long-term follow-up. Conversely, patients who were Ranawat class IIIA or IIIB had a less satisfactory course, with many dying within the follow-up period. In their study, they found that preoperative severity of
myelopathy, spinal cord cross-sectional area, and degree of vertical translocation influenced the final neurological grade.

PADI

Boden et al. recorded complete neurological recovery in all patients who had paralysis and a PADI of at least 14 mm. They also observed improvement by at least one Ranawat grade if the PADI was 10 to 14 mm and no recovery if < 10 mm.\(^\text{29}\)

Pseudarthrosis

Patients with pseudarthrosis are less likely to experience complete pain relief.

Significant Basilar Invagination

Significant basilar invagination is a negative predictor of neurological recovery.

Screening and Conservative Management

Surgical intervention is advocated for patients with intractable pain and/or neurological deficits. Surgery is also indicated in patients who are at risk of impending neurological deficits (see discussion above). The remainder should be conservatively managed, with screening checks at regular intervals to identify patients at risk. The goals of management of this group are to avoid the development of an irreversible neurological deficit, to avoid onset of sudden death due to unrecognized spinal cord compression, and to avoid unnecessary surgery.\(^\text{44}\)

Atlantoaxial Subluxation

It is safe to continue careful observation of patients with known AAS but without neurological deficits, as long as PADI is = 14 mm. If PADI is < 14 mm, MRI should be performed to evaluate spinal cord compression and to determine the thickness of the pannus. Surgery in the form of atlantoaxial arthrodesis should be considered if the space available for the spinal cord (SAC) is < 13 mm or if the cervicomедullary angle is < 135°.

Atlantoaxial Subluxation with Basilar Invagination

This group requires a closer and more aggressive evaluation, as neurological morbidity increases with progressive invagination. MRI is performed if radiographs demonstrate evidence of basilar invagination; it is used to assess the extent of cord/medullary compression and the thickness of the pannus. Patients with isolated and fixed (nonprogressive) basilar invagination without spinal cord compression can be observed after explaining the potential risks.

Subaxial Subluxation

Patients with subaxial subluxation, and PADI of 14 mm on film, but no neurological deficits can be followed up with interval radiographs and observation. MRI is performed if PADI is < 14 mm. Surgical arthrodesis is recommended if segmental mobility is > 3.5 mm or if the SAC is < 13 mm.

Surgical Management of Rheumatoid Arthritis

The primary goals of surgical management are restoration of neurological function and relief of pain. The secondary goals are prevention of further neurological decline and instability, restoration of alignment, correction of deformity, and attainment of a solid arthrodesis. Management should be determined on a case-by-case basis and thus individualized. A proposed management algorithm for the management of rheumatoid involvement of the CVJ is presented in Fig. 33.14. Malnutrition, chronic anemia, osteopenia, decreased endurance, and the side effects of steroids and other disease-modifying drugs place most of the patients at higher baseline risk for surgery. Hence, proper selection of surgical candidates is of utmost importance. Therefore, preoperative evaluation by anesthesiologists and internists is recommended. Fiberoptic awake intubation, as well as intraoperative neurophysiological monitoring, must be considered in all cases.

Preoperative Skull Traction

Traction is recommended for all patients with cranial settling or AAS that does not reduce on voluntary flexion-extension radiographs (Fig. 33.15). It is used for at least 24 to 48 hours to align the osseous anatomy and to relieve neural compression. Prolonged periods of traction do not necessarily improve the extent of reduction. Most reductions occur within 1 to 2 days.\(^\text{45}\) Moreover, prolonged traction and bed rest in already debilitated, malnourished patients with RA can lead to further postoperative morbidity, usually pulmonary in nature. Traction is applied via a halo ring or Gardner-Wells tongs, starting at 5 lb (2.3 kg), with a gradual increase to 12 to 15 lb (5.4–6.8 kg). Periodic radiographic evaluation to verify and quantify the degree of reduction achieved and to detect overdistraction/atlanto-occipital dislocation is mandatory. Traction is contraindicated for complex rotary subluxations and dorsal occipitooatlantoaxial dislocations.
Atlantoaxial Fusion/Fixation

Indications

Select cases of AAS (described below) and retrodental pannus causing spinal cord compression with or without ventral decompression are the common indications for atlantoaxial fusion and fixation. AAS is surgically managed by atlantoaxial arthrodesis. The indications for this surgery in AAS are the presence of neurological deficits, impending neurological compromise, and intractable occipital headache with demonstrated atlantoaxial instability. Severe AAS seldom results in life-threatening neurological complications. Hence, selection of cases for surgery is dependent on factors such as degree of pain and development of neurological deficits. AAS > 9 to 10 mm in the presence of intractable occipitocervical pain or the development of neurological deficits merits surgical arthrodesis. This is most commonly achieved by a dorsal technique, such as C1–C2 transarticular screws, Brooks-Jenkins fusion, or Gallie fusion, or a combination of hooks and clamps. The dorsal approach is favored, as it allows easy access to multiple levels and provides a variety of bony points for fixation. However, it is worth mentioning that dorsal fusion alone, in the presence of irreducible ventral pathology, can be associated with adverse outcomes.

Dorsal Wiring

Dorsal wiring techniques alone have typically been unsuccessful in patients with RA, unlike in other populations, because of significant nonunion rates. Such techniques are also not feasible in patients who have had a C1 laminectomy.

C1–C2 Transarticular Screw Fixation

This is the best method for obtaining arthrodesis in reducible AAS cases, provided the lateral atlantal masses are intact and the quality of bone is satisfactory. Preoperative reconstructed CT images are very helpful in assessing the width of the pars interarticularis at C2, as well as in excluding patients with significant erosion/atrophy and osteoporosis of the lateral mass of C1 for adequate purchase of transarticular screws. Similarly, any encroachment of the vertebral artery groove into the pars interarticularis can be identified preoperatively to avoid potential vertebral artery injuries. The safety of C1–C2 transarticular screw fixation can be augmented by aligning three points (depicted as dots in Fig. 33.16) in the sagittal plane: the insertion site, the dorsal-rostral aspect of the C2 facet, and the upper one third of the ventral C1 arch (Fig. 33.16a). However, if these points are not linear (in line), the first two points instead of the first and third points should be used (Fig. 33.16b). This

Fig. 33.15a, b  Sagittal CT of the cervical spine shows (a) basilar invagination prior to traction and (b) reduction of invagination with traction.
avoids infringement of the vertebral artery foramen by the screw trajectory that would have occurred if the first two points had been used (Fig. 33.16c).

Transarticular fixation (Fig. 33.17) provides immediate fixation of the atlantoaxial articular complex and also eliminates the majority of rotational motion at this joint. To further increase the ability of transarticular screws to withstand flexion and extension forces, a dorsal tension band construct, such as a Gallie or Brooks-Jenkins technique, is recommended. Madawi et al. reported an 87% fusion rate and an 8% rate of vertebral artery injury in 61 patients (37 patients with RA). Haid et al. reported a 96% fusion rate without any vascular or neurological injuries in 75 consecutive patients (30% of whom had RA) with transarticular screws supplemented by interspinous graft. Gluf’s group reported a 98% fusion rate in 191 patients (63 patients with RA), with 5 patients sustaining vertebral artery injuries. If AAS is irreducible, pars or isthmic screws in C2 with sublaminar wires around C1 can be used to provide adequate segmental fixation.

### Halifax Interlaminar Clamps

Halifax interlaminar clamps have not been shown to offer any advantage over wiring techniques, with respect to fusion rates. However, clamps may be a safer alternative in patients with severe spinal canal stenosis.

### Wire and Graft Arthrodesis

The Gallie fusion technique involves using a modified H-shaped bone graft that is secured by midline wiring, whereas the Brooks-Jenkins method involves wedge compression grafts using sublaminar wires (Fig. 33.18).

### Goel’s Atlantoaxial Distraction Technique

Goel et al. reported a new technique of direct manual distraction of facets of the atlas and axis and stabilization by placement of bone graft and metal spacers within the joint, followed by Goel’s direct interarticular C1-lateral mass and C2 pars plate and screw fixation for management of both basilar invagination and atlantoaxial instability.

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**Fig. 33.17** Screen shot of image-guided navigation for transarticular screw placement.
dislocation secondary to RA. Fixation to the occiput or bony decompression was not necessary, and none of the nine patients with RA and basilar invagination treated by this technique had delayed neurological decline during follow-up.53–56

**Bony Fusion**

Pseudarthrosis or fusion in a suboptimal position is relatively common, occurring in 33% to 50% of all patients who undergo dorsal fusion. Hence, some authors even questioned the role of routine bone grafting after instrumentation.57 Long-term results have shown persistent relief of pain even in cases of nonfusion, indicating that fibrous stabilization is adequate for pain relief.58 As expected, patients on long-term steroid therapy were more likely to develop pseudarthrosis.

**Bone Grafting for Fusion**

Allograft provides a suboptimal fusion substrate for dorsal fusion in patients with RA. Bi- or tricorticate iliac crest autograft is ideal, although some surgeons advocate autogenous rib graft, citing fewer donor site complications with similar fusion rates.59 Calvarial grafts are used by some surgeons when occipitocervical fusion is performed, especially in children.60

**Extension of Atlantoaxial Arthrodesis**

It is extremely important to determine the extent of fusion prior to entering the operating room. If there is associated cranial settling, especially if there is impingement upon the spinal cord, fusion extension to the occiput should be strongly considered. However, the decision to extend the fusion to below C2 should be carefully considered. There exists a significant difference in adjacent segment degeneration depending on whether or not the construct is limited in caudal extent to C2 or whether the construct extends further subaxially. Because of the ligamentous laxity precipitated by the disease process and the iatrogenic concentrated stresses applied to levels adjacent to the fusion, patients with RA tend to show precocious degeneration/subluxation of the adjacent segment joints. Olerud et al., however, reported that adjacent level degeneration is more observed in the subaxial spine after cranio cervical fusion that includes the upper subaxial spine.10 This is especially the case in patients who have significant pre-existing subaxial spine disease.58,61 However, this is not the case with isolated cranio cervical fusion that is limited to C1–C2. The likelihood of developing subaxial instability may decrease with early stabilization of an AAS.62 Hence, it is recommended that for isolated AAS, isolated atlantoaxial arthrodesis be performed, if possible. On the other hand, if instability or deformity is global, it may be better to “fuse long.” Occiput to T1–T2 instrumentation provides stabilization of the entire cervical spine to take advantage of superb anchor points provided by thoracic pedicle screws. It is important to reiterate that like other cervical spine operations, long subaxial fusions should not end at C7, but should extend to T1–T2.

**Occipitocervical Fusion**

**Indications**

Occipitocervical fusion in RA is indicated for patients with basilar invagination or cranial settling with myelopathy or impending neurological compromise, combined atlantoaxial and subaxial instability, inability to obtain fusion to the dorsal ring of the atlas (due to laminectomy or insufficient bone stock), or nonunion from a prior atlantoaxial arthrodesis, as well as following transoral decompression.63 Fusion significantly impacts upon the range of motion. It eliminates 30° of flexion/extension and the first 35 to 40° of lateral rotation.64 Suboccipital
decompression with or without C1 laminectomy is performed depending on the nature of pathology and the site of compression of the neuraxis.

## Fixation Techniques

Wires used to secure strut grafts were the first devices employed in occipitocervical fusion procedures. They were associated with high failure rates, providing minimal rigidity and no significant immediate stability.

Rod and wire constructs provide limited resistance to axial loading. Implant and construct failure is common.

Threaded occipitocervical titanium loops use sublaminar cables for fixation (Fig. 33.19).

Plate and screw constructs provide a significant improvement in overall multiplanar stability. Fixed hole-to-hole distances make insertion difficult at times.

Rod and screw constructs employ polyaxial screws, rods, and plates for fixation to the skull. Currently available systems include

- The OctaFix occipitocervical fixation system (Spinal Concepts, Inc., Austin, Texas) uses a contoured occipital plate, which supports the skull against vertical translation and offers incorporation of polyaxial screws to dual titanium rods.
- The modular Oasys occipitocervicothoracic system (Stryker Spine, Allendale, New Jersey) incorporates titanium polyaxial screws, hooks, and rods with occiput plates.
- The CerviFix system (Sofamor-Danek, Memphis, Tennessee) uses two separate titanium occipital plate-rod constructs with hooks, along with C2 pars and subaxial lateral mass screws.
- The Summit system (DePuy AcroMed, Raynham, Massachusetts) uses T- or Y-shaped occipital plates that can be secured to titanium rods (Fig. 33.20).
- The Mountaineer system (DePuy AcroMed) is an upgrade of the Summit system.

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**Fig. 33.19** Intraoperative photograph of a contoured occipitocervical loop secured with sublaminar wires, as well as C1–C2 transarticular and C3 lateral mass screws.

**Fig. 33.20a, b** (a) Intraoperative photograph and (b) postoperative anteroposterior view of occiput–C2 fixation with the Summit system. Note the C2 translaminar screws used for fixation.
Transoral Decompression

Indications

Transoral decompression is indicated for large ventral masses causing spinal cord compression (Fig. 33.23). Such pathology includes periodontoid pannus, upward migration of the dens, and irreducible atlantoaxial kyphosis. Arrest of the vertical migration of the dens and regression of the size of the periodontoid pannus have been well documented. Hence, transoral decompression may not be necessary unless progressive myelopathy is present. Casey et al., in their review of 67 transoral decompressions in patients with RA, felt that the major component of ventral compression in patients with vertical translocation is osseous in nature. Hence, they proposed ventral decompression with posterior fusion in these patients. However, they did not observe a statistically significant difference in outcome in patients who underwent transoral decompression and dorsal fusion, compared with patients who had a dorsal fusion alone.

Complications

Complications involving occipital screw placement include cerebellar parenchymal or subdural hematomas. The inside-outside technique in which cranial bolts are placed from inside the occiput to the outside was reported by Sandhu et al. with good results (Fig. 33.21). Specific complications of rod systems include rod pistoning into the posterior fossa (Fig. 33.22).

Stabilization after Dens Resection

Transoral decompression should be accompanied by a stabilization procedure, either at the same setting or as a staged procedure. Fang et al. did not perform secondary...
fixation after transoral decompression, but they did use prolonged external fixation in a cast or halo, with a high nonunion rate.

Dorsal atlantoaxial wire fixation and fusion may be used to supplement transoral dens resection. A tendency, however, for ventral instability persists, as the circular wires by themselves may not be efficient in preventing translatory displacement (due to a “parallelogram effect”).

Several surgeons follow transoral decompression with occipitointalantoaxial fusion. Good results have been reported with this strategy. The major disadvantages with this technique are a substantial restriction of flexion and extension and the chronic overload of subaxial segments. Another alternative is that of atlantoaxial transarticular screw fixation. This strategy limits the fusion to C1–C2 alone, while providing augmented biomechanical stability.

It should be kept in mind that all of the above-mentioned dorsal fusion procedures require that the patient be turned from the supine to the prone position during the period of diminished stability, with a theoretical risk of injury to the spinal cord.

## Ventral Plate Fixation

Ventral craniovertebral and upper cervical plating is complicated by two factors: the relatively weak screw fixation points and the risk of infection secondary to transoral contamination (Fig. 33.24). Additionally, transoral upper cervical plate fixation is less stable than combined ventral and dorsal reconstruction procedures. This limits their use as a “free-standing” stabilization procedure. Hence, Kerschbaumer’s group performed anterior Harm’s plate fixation (after transoral decompression) to provide immediate stabilization for irreducible atlantoaxial...
kyphosis in rheumatoid patients. They observed screw loosening in two of three patients in their earlier series and subsequently combined ventral plating with a dorsal Brooks-Jenkins fusion. They reported improvement of pain and an increased range of motion in 12 patients followed for 26 to 77 months postoperatively.73 Goel and Karapurkar reported a case of ventral plating from the clivus to C3 after transoral dens resection.74 Several other ventral atlantoaxial plate systems have been described, including the subarcicular atlantoaxial locking plate reported by Kandziora et al., who found that it provided better fixation.75

Ventral Transarticular Screw Fixation

Ventral transarticular screw fixation strategies are limited predominantly by the risk associated with screw violation of adjacent vascular and neural structures, as well as the risk of infection. Their use, therefore, is significantly restricted (Fig. 33.24).

Contemporary Controversies in Management of Rheumatoid Arthritis of the Craniocervical Junction

Is prophylactic surgery justified? Because most patients with cervical spine involvement in RA are asymptomatic, conventional practice has been to reserve cervical or occipitocervical fusion for those with worsening neurological deficits or intractable axial pain. It is debatable, however, whether prophylactic surgery should be performed in patients without neurological abnormalities, but with associated imaging evidence of severe instability, to prevent catastrophic neurological complications. Tanaka et al. followed 50 patients with AAI, but without neurological compromise, for 24 years. Twenty-six of these patients had prophylactic surgery, and 24 were managed conservatively.76 They reported a 1.7-fold decrease in survival for conservative treatment (probability of survival being 96% after 24 years in the surgical group vs. 52% in the conservative group), better pain relief and functional recovery, and decreased rates of vertical settling and AAS. They also found that mortality in conservatively managed patients is higher if the disease is mutilating and if susceptible factors in both of the HLA-DRB1 alleles are present.

Is surgery justified for Ranawat IIIB patients? Many authors found that patients who are Ranawat classes IIIA and IIIB have a less satisfactory postoperative course, with many dying soon after surgery. This strongly suggests that surgical intervention should be performed before permanent damage to the spinal cord has occurred.6 Nonambulatory myelopathic patients (IIIB) fare particularly poorly after surgery, with up to 25% mortality within 6 months. Casey et al. even made a point that surgery for these IIIB patients is “too much, too late,” as they observed that this subset of patients fared worse in terms of complication rate, hospital stay, functional outcome, and ultimate survival.77 A meta-analysis of all series that reported outcomes in IIIB patients showed that 60% of patients were able to walk again (i.e., improving to at least IIIA), which is certainly a huge benefit in terms of the quality of life conferred on them by surgery.78 Neck pain improved universally in all series. Hence, it can be argued that aggressive surgical intervention in select IIIB patients is justified, given the dismal progression of the disease with conservative management.

Is transoral decompression necessary in rheumatoid disease of the CVJ? There is irrefutable evidence in the literature that retrodental pannus regresses and vertical dislocation is arrested following dorsal fusion alone.66,67 Hence, it is justified to limit surgery to dorsal instrumentation and fusion alone, and to follow these patients clinically and radiologically with follow-up MRI. Transoral decompression can be performed later if there is further neurological deterioration or progression of spinal cord compression from ventral pathalogy.

There are no prospective studies comparing the clinical outcome in patients treated by ventral decompression versus dorsal atlantoaxial fusion. Very few transoral decompressions are currently performed for rheumatoid pathology.

Conclusion

Rheumatoid arthritis is a common autoimmune inflammatory disorder that commonly affects the CVJ and upper cervical spine. Patients with rapid joint erosion, elevated C-reactive protein levels, seropositivity, and HLA-DRB1 susceptibility are particularly prone to early or severe involvement of the cervical spine. Atlantoaxial subluxation, basilar invagination, and subaxial subluxation are the three most common forms of craniocervical involvement in RA. Although many of these patients may be asymptomatic, attention should be paid to the predictors of impending paralysis. Patients with RA are disabled with other peripheral joint deformities; thus, it is often difficult to identify early changes suggestive of cervical myelopathy. Surgical outcome is often rewarding, particularly if performed before myelopathy appears. Patients who are candidates for surgical intervention should be carefully selected, with attention paid to anatomical and bony detail of the spinal segments involved.

References

4. Garrod AE. A Treatise on Rheumatism and Rheumatoid Arthritis. London: C Griffin; 1890
The atlantoaxial joints are the most mobile in the body. The lateral masses of the atlas and axis form a pillar of stability and mobility for the neck and head. Like all other joints, they are subject to arthritis. With the general aging of the population, the issue of arthritis is becoming more relevant. Osteoarthritis of the atlantoaxial joint is a well-defined phenomenon that eventually results in atlantoaxial instability. The process of joint degeneration and instability is progressive and extends over several months to years. This instability is probably the result of degeneration of the articular cartilage, reduction of the joint space, and secondary incompetence of the ligaments controlling the movements.1–3

Degenerative arthritis of the atlantoaxial joint is rarely discussed, and most of the literature on the subject is in the form of isolated reports.1,4–9 Difficulty in obtaining good quality images and in direct visual assessment of the joint has probably made the diagnosis and evaluation of arthritis relatively less common. We retrospectively identified 108 patients who were diagnosed as having atlantoaxial instability secondary to degenerative osteoarthritis of the atlantoaxial joints on the basis of presenting clinical features, radiological imaging, and direct observation of the joint status during surgery that involved opening, manipulation, and fixation of the joints. These patients were treated in our department of neurosurgery between 1990 and 2008.10 The following discussion is based on this experience.

Clinical Features

Pain in the neck on movement often forms the earliest and most prominent symptom. The more classical pattern is of an elderly patient presenting with symptoms of pain in the neck on movement, along with restriction of neck movement and gradually progressive quadriparesis over a few days to several months. The incidence and severity of quadriparesis vary in different reports; this variation reflects the duration of the disease before the diagnosis is clinched. Sensory symptoms are relatively mild and typically include bilateral upper and lower extremity paresthesias and kinesthetic sensation deficits. A history of mild to moderately severe trauma, from a few days to several years prior to diagnosis, may be present in a substantial number of patients. In most of these cases, the symptoms progress from the time of trauma. Palpable crepitis with motion has also been identified as a presenting symptom.4 Carrying of heavy weights over the head has been shown to cause increased incidence of cervical spondylotic changes and basilar invagination.11

Radiological Features

Among the more constant radiological features is reduction of the height of the lateral mass complex due to reduction of the joint space (Figs. 34.1 and 34.2). Erosion of the bones of the lateral masses and body of the atlas and axis is usually observed in more chronic situations.12 Degenerative erosion of the facets of the atlas and axis, the odontoid process, and the body of the axis, as well as periodontoid ligamentous degenerative changes, is also frequent (Figs. 34.3, 34.4, 34.5, 34.6, 34.7, 34.8, 34.9, 34.10, and 34.11). The changes seen in our series of 108 patients are summarized in Table 34.1.

The term basilar invagination in cases of craniovertebral arthritis has been used synonymously with cranial settling and vertical odontoid migration.11,13–15 In our experience, basilar invagination was only mild to moderate, and in none of the cases did the tip of the odontoid process migrate >9 mm above the Chamberlain line. This is unlike the basilar invagination observed in rheumatoid arthritis cases in which the superior migration of the odontoid process is significantly greater because of the more severe lateral mass bone collapse. In our series, the alignment of the facets was horizontal and not oblique, as was observed in cases with congenital basilar invagination in our earlier reports.16,17

Atlantoaxial joint arthritis is expected to eventually result in atlantoaxial instability in all cases. However, in those presenting early, this may not be evident.4 In our experience, all patients presented with mobile atlantoaxial dislocation, and more than half showed hypermobility. The dislocation may only be partially reducible due to the presence of nonyielding tissues around the odontoid process. The subtlety of instability may make the diagnosis difficult in some cases.
Fig. 34.1a–e Images of a 65-year-old male patient.

a Preoperative computed tomography (CT) scan with the head of the patient in flexion shows the atlantoaxial dislocation and basilar invagination. Erosion of the dens and peri-odontoid degenerative tissue can be seen. Osteophyte-like ossification in the region of the apical ligament and atlantodental joint can be observed. Degenerative changes in the cervical spine are seen.

b Preoperative CT scan with the head in an extended position showing reduction of the dislocation.

c Three-dimensional CT scan showing ossification near the apical ligament at the site of its attachment to the clivus.

d Postoperative CT scan shows realignment of the craniovertebral junction and fixation.

e Postoperative radiograph showing fixation with plate and screws and bone graft.
Fig. 34.2a–h Images of a 58-year-old female patient.
a Preoperative radiograph with the head in flexion showing marked atlantoaxial dislocation.
b Preoperative radiograph with the head in extension showing partial reduction of the dislocation.
c Preoperative CT scan showing the dislocation. Erosive changes in the dens and degenerative changes in the cervical spine can be seen.
d T1-weighted magnetic resonance imaging (MRI) showing the dislocation. Preodontoid degenerative tissue can be seen.
e Postoperative CT scan showing fixation in an incompletely reduced position.
Fig. 34.2f–h

f Postoperative CT with the sagittal cut through the lateral masses. A metal spacer is seen in the distracted joint, in addition to fixation with plate and screws.

g Coronal cut showing spacers in the atlantoaxial joints and screws in the facets of the atlas and axis.

h Postoperative radiograph showing the plate and screw fixation. Metal spacers can be seen.

Fig. 34.3a, b Images of 57-year-old male patient.

a Preoperative CT scan showing extensive degenerative changes in the periodontoid region and in the facets.

b Coronal cut showing degenerative changes in the lateral masses. Reduction of joint space height and osteophyte formation in the facets can be observed.
Fig. 34.4 Coronal CT scan showing degenerative changes in the atlantoaxial joints of a 68-year-old man. Degeneration is more marked on one side, where erosive changes are observed in the facets of the atlas and axis.

Fig. 34.5a, b Images of a 70-year-old male patient.
- CT scan showing degenerative periodontoid changes. Calcification of the anterior occipitoatlantal membrane can be seen.
- Coronal image showing degenerative changes in the atlantoaxial joints. Osteophytes can be seen in the facets of the atlas and axis.

Fig. 34.6a, b Images of a 67-year-old male patient.
- Preoperative CT scan showing extensive periodontoid degenerative changes. Atlantoaxial dislocation and basilar invagination can be seen.
- Postoperative scan showing reduction and fixation of the region.
Fig. 34.7a–d Images of a 68-year-old female patient.

a  T2-weighted MRI showing degenerative periodontoid changes. Elevation of the posterior longitudinal ligament/tectorial membrane is clearly visible.

b  T1-weighted MRI showing the periodontoidal soft tissue and degenerative changes.

c  CT scan in flexion showing subtle basilar invagination and atlantoaxial dislocation.

b  Postoperative CT scan showing reduction of the dislocation and basilar invagination.
Fig. 34.8  Image of a 49-year-old female patient. Paraodontoid degenerative change is seen.

Fig. 34.9a–e  Images of 61-year-old male patient.

a  T1-weighted MRI showing a retro-odontoid ligamentous degenerative tumorlike mass. The posterior longitudinal ligament/tectorial membrane can be seen elevated posteriorly by the mass.

b  T2-weighted MRI showing the retro-odontoid hypointense mass. Signal intensity changes in the cord can be seen.

c  Lateral radiograph in flexion shows subtle atlantoaxial dislocation.
Fig. 34.9d–e

d  Lateral radiograph in extension showing incomplete reduction of the dislocation.

Fig. 34.10a–g Images of a 65-year-old male patient.
a  Radiograph with the neck in a flexed position shows moderate atlantoaxial dislocation.
b  Radiograph with the neck in extension showing incomplete reduction of the dislocation.
c  CT scan showing basilar invagination and atlantoaxial dislocation. Osteophyte-like changes are seen in the region of the apical ligament.

e  Postoperative radiograph showing atlantoaxial fixation. The reduction of the dislocation is incomplete.
Degenerative pannus, also referred to as articular, ganglion, synovial, or juxtafacet cyst, may arise from the degenerating synovial lining of any joint in the body. Pannus related to atlantoaxial joint arthritis probably represents degenerative ligaments and secondary osteophyte-like tissue formation in the periodontoid region. Degenerating tissues are typically isointense on T1-weighted magnetic resonance imaging (MRI) and iso- to hypointense on T2-weighted MRI (Fig. 34.9) and do not enhance on contrast administration. In some cases, the retro-odontoid mass may have a tumorlike appearance, resulting in posterior “buckling” of the posterior longitudinal ligament or the tectorial membrane and indentation of the cord substance (Figs. 34.7 and 34.9). Such a retro-odontoid “pseudotumor” is a well-defined entity. We recently reported a case of rheumatoid arthritis having both atlantoaxial dislocation and basilar invagination. In this case, we identified laxity and buckling of the posterior spinal ligaments as a cause of retro-odontoid pannus formation. This feature was confirmed by the dramatic reduction of the pannus immediately following surgery that involved distraction of the facets of the atlantoaxial joint. Retro-odontoid ligamentous hypertrophy appears to be related to laxity of ligaments due to reduced atlantoaxial height and secondary to progressive degenerative changes in the region. The pathogenesis of the degenerative changes simulates to an extent the formation of posterior osteophytes in cases with spinal degeneration. It appears that the presence of retro-odontoid ligamentous degenerative hypertrophy in an elderly person can be diagnostic evidence.
Fig. 34.11a–i Images of a 78-year-old patient.

a Radiograph with the neck in a flexed position showing atlantoaxial dislocation.

b Radiograph with the neck in extension showing incomplete reduction of the dislocation.

c CT scan showing basilar invagination and atlantoaxial dislocation. Osteophyte-like changes are seen in the region of the apical ligament.

d CT scan with the sagittal section traversing the atlantoaxial joint. It shows arthritis and dislocation of the atlantoaxial joint and chronic arthritic changes in the rest of the spine.

e Coronal section of CT showing degenerative changes in the atlantoaxial joint.
Fig. 34.11f–i
f  T1-weighted MRI showing periodontoid degenerative soft tissue and atlantoaxial dislocation leading to cord compression.
g  T2-weighted MRI showing periodontoid degenerative soft tissue and atlantoaxial dislocation leading to cord compression. Degenerative changes in the rest of the spine are evident.
h  Postoperative CT scan showing reduction of the dislocation and of the basilar invagination.
i  Radiograph with the head in flexion showing the fixation.
that suggests atlantoaxial instability, even when such instability is not clearly visualized on radiological imaging. In general, it is observed that retro-odontoid ligamentous degenerative changes are identified and are thicker in cases where the atlantoaxial instability is less marked and the entire degenerative process is more chronic in nature. As opposed to cases of rheumatoid arthritis, where the process is more chronic or long-standing, bone destruction of the facets of the atlas and axis and osteomalacia are not as pronounced in cases of degeneration-related arthritis. Degeneration of the atlantodental joint may be observed in a minority. Cervical spondylotic degenerative disease is likely to be present to varying degrees in the rest of the spine in most cases.

### Surgery

For basilar invagination and retro-odontoid ligamentous hypertrophy, transoral decompression and subsequent posterior fixation has been the accepted treatment protocol. However, several authors have reported the futility of direct surgical resection of the retro-odontoid mass with regression of the size of the retro-odontoid ligamentous hypertrophy on conservative treatment or after posterior fixation. In 2004, we described an atlantoaxial lateral mass plate and screw fixation without surgical removal of the retro-odontoid pseudotumor. Although some surgeons still prefer to resect the retro-odontoid degenerative mass, the current consensus appears to be that in such cases stabilization of the atlantoaxial joint is sufficient and results in regression of the retro-odontoid mass. Some surgeons advise incorporation of the occipital bone and cervical vertebrae up to C3 and C4 in the occipitocervical fixation and feel that such a fixation is necessary to avoid cranial settling. We observed from our experience that such an extended fixation may not be necessary. Our patients were treated by the lateral mass plate and screw method described by us earlier for both fixed and reducible atlantoaxial dislocation. An attempt may be made to distract the facets and restore the height of the lateral mass by impaction of bone chips harvested from the iliac crest alone or by additional impaction of spiked metal spacers. Distraction of the facets not only assists in the reduction and fixation of the atlantoaxial dislocation and basilar invagination but also stretches the buckled posterior spinal ligaments that appear to play a significant role in the pathogenesis of retro-odontoid ligamentous degenerative hypertrophic masses.

Upon opening the joint, the presence of a relatively rough, pale yellow articular surface devoid of end-plate, reduction of the joint space, and evidence of instability of the region are the more prominent observations. On reviewing the postoperative images, it was apparent that in all cases in our series there was reduction of the atlantoaxial dislocation and basilar invagination (Figs. 34.1 and 34.2). The postoperative reduction of the dislocation was significant but incomplete in about half of the cases out of those in which the reduction of the dislocation was incomplete during preoperative evaluation. This was probably because of the presence of a nonyielding periodontoid ligamentous degenerative mass. Despite the incomplete reduction, the treatment resulted in an immediate postoperative and sustained neurological recovery. This improvement appeared to be related to elimination of abnormal mobility, realignment of bones, and reduction of the indentation by the retro-odontoid mass. Postoperative computed tomography (CT) scans revealed evidence of reduced soft tissue shadow in the periodontoid space. The neurological recovery seen after the treatment of osteoarthritis-related atlantoaxial dislocation and basilar invagination appeared to be significantly more gratifying than that seen after treatment of rheumatoid arthritis–related atlantoaxial dislocation and basilar invagination.

#### Anterior Transoral Release Followed by Posterior Atlantoaxial Fixation

Because the reduction of the dislocation was incomplete in a large proportion of cases, our current strategy of surgical treatment in this condition is to approach the

### Table 34.1 Radiological findings

<table>
<thead>
<tr>
<th>Serial No.</th>
<th>Finding</th>
<th>Incidence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Reduction of the height of the atlantoaxial lateral mass complex</td>
<td>100</td>
</tr>
<tr>
<td>2</td>
<td>Erosion of the bones of the atlas and axis</td>
<td>76</td>
</tr>
<tr>
<td>3</td>
<td>Periodontoid degenerative tissue</td>
<td>90</td>
</tr>
<tr>
<td>4</td>
<td>Periodontoid osteophyte-like bone formation</td>
<td>28</td>
</tr>
<tr>
<td>5</td>
<td>Retro-odontoid tumor-like ligamentous hypertrophy indenting into the cord</td>
<td>13</td>
</tr>
<tr>
<td>6</td>
<td>Mobile atlantoaxial dislocation</td>
<td>100</td>
</tr>
<tr>
<td>7</td>
<td>Subtle mobility</td>
<td>46</td>
</tr>
<tr>
<td>8</td>
<td>Hypermobility</td>
<td>54</td>
</tr>
<tr>
<td>9</td>
<td>Completely reducible</td>
<td>59</td>
</tr>
<tr>
<td>10</td>
<td>Incompletely reducible</td>
<td>41</td>
</tr>
<tr>
<td>11</td>
<td>Basilar invagination</td>
<td>68</td>
</tr>
</tbody>
</table>
A 61-year-old man presented with complaints of progressive quadriparesis and neck pain persisting for ~1 year. He had difficulty in performing manipulations with the hands and needed support to walk. He reported a fall from a height of ~6 m resulting in hyperextension of the neck 2 years before. Except for severe neck pain that lasted for ~1 week, there were no other significant posttraumatic sequelae. He had no history of any systemic disease. There was no evidence of rheumatoid arthritis or any other joint-related generalized disease. Neurological examination found spastic grade 3 or 4 quadriparesis but no gross sensory abnormality. MRI showed a retro-odontoid process midline lesion indenting into the craniomedullary cord, appearing as isointense on T1-weighted and hypointense on T2-weighted images (Fig. 34.9a, b). Dynamic radiography revealed subtle mobile and incompletely reducible atlantoaxial dislocation (Fig. 34.9c, d). The patient was treated with lateral mass atlantoaxial plate and screw fixation. Satisfactory but incomplete reduction was achieved (Fig. 34.9e). The patient showed remarkable neurological and symptomatic recovery. The improvement was sustained, and at the follow-up examination 14 months after surgery, the patient was almost asymptomatic.

Craniovertebral region transorally in the first stage of the operation. Surgery is done under cervical traction. The operation involves decompression and radical resection of compressive degenerative tissue in the periodontoid region, without resection of any part of the bone. Reduction of the dislocation is confirmed on radiological imaging. The patient is then turned prone, and a posterior fixation as discussed earlier is completed. This strategy appears to be a viable option in the treatment of arthritis-related “fixed” atlantoaxial dislocation.

Conclusion

Atlantoaxial joint osteoarthritis should be considered in the differential diagnosis in elderly patients being investigated for the symptom of neck pain with or without progressive quadriparesis. Identification of a periodontoid ligamentous degenerative mass suggests the presence of instability of the atlantoaxial joint.

Our experience confirms that in these cases, osteoarthritis of the atlantoaxial joint is the primary event that results in the secondary development of atlantoaxial instability and basilar invagination and periodontoid ligamentous degeneration. Direct opening of the joint and introduction and impaction of bone chips within it provide remarkable joint stability and a wide ground for bone fusion, along with an opportunity for realignment of atlantoaxial dislocation and basilar invagination. From our successful experience, we are convinced that atlantoaxial joint fixation is the key to treatment of osteoarthritis of the atlantoaxial joints.

References

Atlantoaxial dislocation and basilar invagination are both commonly associated with rheumatoid arthritis involving the craniovertebral region. Numerous methods to achieve decompression and stabilization of the region have been described in the literature. We present the feasibility of an alternative technique of craniovertebral bone alignment by distraction of the facets of the atlas and axis with or without the addition of direct lateral mass plate and screw atlantoaxial fixation for management of both basilar invagination and atlantoaxial dislocation secondary to rheumatoid arthritis. We have discussed this technique in our reports on the subject.1–5

Seropositive rheumatoid arthritis has been identified in ~0.9% of the white adult population of the United States and 1.1% of the adult population in Europe.6,7 Of these, as many as 10% of patients may need an operation for atlantoaxial subluxation and basilar invagination. Such figures are not available from India, as the incidence of rheumatoid arthritis affecting the craniovertebral junction appears to be significantly less, and in our institute, congenital anomalies form the bulk of craniovertebral pathology.

The term basilar invagination has been used synonymously with cranial settling and vertical odontoid migration.8–10 Basilar invagination is commonly associated with atlantoaxial dislocation, and the complex results in a significant degree of neck pain and myelopathy. Occipitocervical fixation has been observed to provide stabilization to the craniovertebral region, and the clinical outcome has been uniformly reported to be satisfactory.3,8,10–12 Different instrumentation and methods have been adopted to secure the occipitocervical fixation. Recently, several authors have reported success after transarticular atlantoaxial fixation.11,12 Some reports have indicated that atlantoaxial fixation is the treatment of choice even in the presence of retro-odontoid pannus. Some authors have reported arrest of the vertical migration of the basilar invagination and regression of the size of the retro-odontoid pannus after posterior fixation.10,12 For basilar invagination, transoral decompression and subsequent posterior fixation has been the most accepted treatment protocol. Menezes et al.10 observed that traction in cases with basilar invagination and atlantoaxial subluxation results in a significant improvement in the craniovertebral alignment. They suggested the use of halo traction for maintaining the distracted and reduced state of both basilar invagination and atlantoaxial subluxation during positioning for surgery to avoid cord injury.

Clinical Series

Between November 2001 and January 2008, 15 patients with rheumatoid arthritis involving the craniovertebral junction were treated in our neurosurgery department.4,5 Nine had basilar invagination and “fixed” atlantoaxial dislocation, and six had a retro-odontoid pannus and mobile and incompletely reducible atlantoaxial dislocation. Patients ranged in age from 24 to 78 years. Seven patients were men, and eight were women. Neck pain and spastic quadriplegia were the most prominent symptoms.

Procedures

All patients underwent distraction of the facets of the atlas and axis and attempts toward reduction of both basilar invagination and atlantoaxial dislocation (Figs. 35.1, 35.2, and 35.3) by the techniques described by us earlier.1–5 In 11 patients, distraction of the facets was followed by fixation using lateral mass plate and screws. In four patients, only distraction of the facets using custom-made spiked spacers was performed, and no lateral mass fixation using a plate and screws was done (Figs. 35.1 and 35.2). No patient underwent anterior transoral decompression or a posterior foramen magnum bony decompression.

Selection Criteria for the Type of Surgery

In general, we feel that distraction and additional fixation using lateral mass plate and screws should be performed wherever feasible. This is particularly true in younger patients and in cases where the lateral mass bones could provide sufficiently strong purchase for the screws. However, when the mobility is subtle, and the lateral masses are either diseased or osteoporotic, only distraction may be sufficient.

Text continues on page 395
Fig. 35.1a–e
a  Preoperative computed tomography (CT) scan showing marked basilar invagination.
b  Coronal view showing the atlantoaxial joint and lateral masses.
c  Postoperative CT scan showing partial reduction of the basilar invagination.
d  Postoperative coronal view showing the spacers in the facet joint on both sides.
e  Postoperative sagittal image showing the spacer in the joint space.
Fig. 35.2a–h  Images of a 24-year-old male patient.
a  Plain radiograph of a 24-year-old man, with the head in flexion showing marked atlantoaxial dislocation.
b  Radiograph with the head in extension showing persistent atlantoaxial dislocation.
c  T2-weighted magnetic resonance imaging (MRI) showing basilar invagination and atlantoaxial dislocation, as well as marked compression of the cervicomedullary cord.
d  CT scan showing severe atlantoaxial dislocation and basilar invagination.
e  Scan showing overriding of the facet of the atlas on the axis (spondyloptosis).
Fig. 35.2f–h
f Postoperative CT scan showing partial but significant reduction of both atlantoaxial dislocation and basilar invagination.
g Postoperative scan showing the facets of the atlas and axis in alignment and fixation using plates, screws, and a spacer.
h Postoperative radiograph showing atlantoaxial fixation with plate, screws, and spacers.

Fig. 35.3a–h
a CT scan showing atlantoaxial dislocation and mild basilar invagination.
Fig. 35.3b–f
b T2-weighted MRI showing retro-odontoid pannus, lax posterior spinal ligament, and evidence of cord compression.
c T1-weighted MRI showing retro-odontoid pannus and cord compression.
d Postoperative CT scan showing reduction of the atlantoaxial dislocation and basilar invagination.
e Sagittal CT scan showing the spacer impacted within the atlantoaxial joint.
f Coronal CT scan showing the spacers within both of the atlantoaxial joints.

Fig. 35.3 g–h
Surgical Technique

Intraoperative Joint Distraction and Lateral Mass Atlantoaxial Fixation

The basic steps of surgery have already been discussed previously and are summarized here. Cervical traction is given prior to induction of anesthesia, and the weights are progressively increased to approximately one fifth of the total body weight. The atlantoaxial facet joints are widely exposed on both sides after sectioning of the large C2 ganglion. The joint capsule is excised, and the articular cartilage is widely removed using a microdrill. The joints on both sides are distracted using an osteotome. The status of the atlantoaxial dislocation and of basilar invagination is evaluated by intraoperative radiographic control. Large pieces of corticocancellous bone graft harvested from the iliac crest and titanium metal plate spacers are stuffed into the joints (Figs. 35.1, 35.2, and 35.3). The size of the spacers used depends on the space available within the distracted joint space allowing maximum reduction of atlantoaxial dislocation and basilar invagination. Morcellized bone graft is stuffed into the distracted joint space on all sides and into the sheaths of the spacer. Plate and screw fixation of the region is subsequently performed by the interarticular technique (Fig. 35.2). Additional bone graft is placed between the posterior elements of C1 and C2 after decorticating the host bone area with a burr. Postoperatively, the traction is discontinued, and the patient is placed in a four-post hard cervical collar for 3 months; all physical activities involving the neck are restrained during that period.

Stand-alone Intraoperative Joint Distraction (Joint Jamming)

We treated four patients with rheumatoid arthritis involving the craniovertebral junction by only distraction of the facets. The main reason for selection of such an operative procedure was considerable destruction of the facets of either or both the atlas and axis. Surgery involved attempts to reduce the basilar invagination and restore the height of the “collapsed” lateral mass by manual distraction of the facets of the atlas and axis and forced impaction of titanium spacers in the joint, in addition to bone graft harvested from the iliac crest. The procedure also provided stabilization of the region. No other fixation procedure involving wires, screws, plates, and rods was performed simultaneously.

Results

Follow-up ranged from 1 to 98 months (average 32 months). All patients improved to varying degrees following surgery. There were no intraoperative or postoperative vascular, neurological, or infective complications. During follow-up, none of the patients suffered delayed neurological
worsening warranting transoral or posterior decompression or any other kind of surgical procedure. No patient needed re-exploration for fixation failure. Immediate postoperative and follow-up radiographs confirmed fixation and fusion and reduction of the basilar invagination.

In all cases, craniovertebral realignment and stabilization without any bone decompression were successful. Wide removal of the atlantoaxial joint capsule and articular cartilage by drilling and subsequent distraction of the joint by manual manipulation provided a unique opportunity to obtain reduction of the basilar invagination and of atlantoaxial dislocation. The use of spiked spacers provided distraction and stability to the region. In select cases, the stability to the region was such that no additional fixation procedure was felt to be necessary. Maintenance of the joint in a distracted and reduced position with the help of bone graft and metal spacers and subsequent fixation of the joint with the help of interarticular screws and a metal plate provided a biomechanically firm fixation and sustained distraction. Multiholed titanium spacers were chosen to allow bone incorporation and fusion across the distracted joint space. The fixation was seen to be strong enough to sustain the vertical, transverse, and rotatory strains of the most mobile region of the spine. Following surgery, the alignment of the odontoid process, anterior arch of the atlas and clivus, and entire craniovertebral junction became more normal. The tip of the odontoid process receded in relationship to the Wackenheim clival line and Chamberlain line, suggesting reduction in the basilar invagination. The posterior tilt of the odontoid process, as evaluated by a modified omega angle, was reduced after the surgery. The segmental nature of fixation in our cases was seen to provide additional scope for local stabilization, assist bone fusion, and permit near complete neck movement in the postoperative period. Biomechanical advantage afforded by our technique is suggested by our successful fusion results. Many surgeons advise incorporation of the occipital bone and cervical vertebrae up to C3 and C4 in the occipitocervical fixation and feel that such a fixation is necessary to avoid cranial settling. Several authors advocating atlantoaxial fixation have found no reason to incorporate the occipital bone and lower cervical vertebrae in the fixation procedure.

Our patients were relatively young when compared with the similar subgroup reported in the literature. We observed that the bone quality of the atlas and axis facets for purchase of the screws was satisfactory, allowing the possibility of distraction and stabilization with screw implantation. Our impression is that rheumatoid arthritis is a disease of the articular capsule and bones forming the atlantoaxial joint. The articular capsule and the subchondral bone may be affected, leading to varying degrees of subluxation and basilar invagination. Such joint affection can lead to lateral mass collapse, as has been identified by some authors in similar cases. In such a situation, treatment of the joint appears to be the most rational form of therapy.

The patients showed remarkable sustained neurological and radiological improvement. In the postoperative phase, patients used a hard cervical collar and were advised to limit activities related to neck movement for a period of 3 months. However, activities such as assisted walking in the house were permitted. Some authors advise a more rigid postoperative immobilization protocol, and others recommend the use of halo traction after occipitocervical instrumentation.

### Case Illustration

A 66-year-old woman who had seropositive rheumatoid arthritis and was on long-term steroid therapy presented with gradually progressive quadriparesis for the past 18 months. Examination revealed spastic grade 4 quadriparesis. She could walk with support and performed routine activities with considerable difficulty. She did not have any sensory deficit. Computed tomography (CT) and magnetic resonance imaging (MRI) showed basilar impression with destruction of the facets of the atlas and axis (Fig. 35.1a, b). The facets were not suitable for screw implantation. There was no retro-odontoid pannus. The patient underwent surgery in the prone position under cervical traction by the technique described by us earlier. The atlantoaxial joints were exposed after sectioning of the C2 ganglion. The joint capsule was excised, and the articular cartilage was removed using a microdrill. The joints on both sides were distracted using an intervertebral spreader. The joint space was assessed, and the largest-size titanium spacer suitable for the region that allowed maximum reduction of the basilar invagination was impacted into the joint space using suitable instruments. Large pieces of corticocancellous bone graft harvested from the iliac crest were stuffed into the space available and all around it. The stability achieved after the impaction of the spacers was firm, and the need for an additional midline surgical procedure was not necessary. Postoperatively, the traction was discontinued, and the patient was placed in a four-post hard cervical collar for 3 months. Postoperatively, symptoms improved. Postoperative CT and MRI showed significant but incomplete reduction of the basilar invagination (Fig. 35.1c-e). At 24-month follow-up, the patient showed significant clinical recovery and was able to walk on her own.
Case Illustration

A 24-year-old man with seropositive rheumatoid arthritis presented with neck pain and progressive weakness of all four limbs lasting 3 years. On admission, the patient had grade 3–4 spastic quadripareisis. He was unable to walk and was bedridden (Ranawat grade IIIB). Dynamic radiograph of the cervical spine showed fixed atlantoaxial dislocation. CT and MRI showed basilar impression, fixed atlantoaxial dislocation, and marked compression of the cervicomedullary cord (Fig. 35.2a–e). The patient underwent surgery in the prone position, under cervical traction, which was instituted prior to the induction of anesthesia. The atlantoaxial facet joints were widely exposed on both sides after sectioning of the large C2 ganglion. The joint capsule was excised, and the articular cartilage was widely removed using a microdrill. The joints on both sides were distracted using appropriate-sized osteotomes. The status of the atlantoaxial dislocation and basilar invagination was evaluated by intraoperative radiographic control. Large pieces of corticocancellous bone graft harvested from the iliac crest and titanium metal plate spacers were stuffed into the joints. Morcellized bone graft was stuffed into the distracted joint space on all sides and into the sheaths of the spacer. Plate and screw fixation of the region was subsequently performed by the interarticular technique. A two-holed stainless steel plate was used measuring 15 to 20 mm in length. The screws were 2.4 to 2.6 mm in diameter and measured 16 to 22 mm in length. Screws were passed bilaterally through the holes in the plate into the lateral mass of the atlas and axis. Additional bone graft was placed between the posterior elements of C1 and C2 after decorticating the host bone area with a burr. Postoperatively, traction was discontinued, and the patient was placed in a four-post hard cervical collar for 3 months. Symptoms improved postoperatively. Post-surgery CT showed partial but significant reduction of the atlantoaxial dislocation and basilar impression (Fig. 35.2f–h). At follow-up, the patient showed significant improvement in his neurological status and was able to walk without support (Ranawat grade II).

Retro-odontoid Pannus

It was not possible to assess the extent of regression of pannus in our earlier cases, as stainless steel metal plates were used for fixation. However, we recently encountered a case in which we had used titanium and MRI-compatible spacers for distraction of the atlantoaxial joint, and the postoperative status of the craniovertebral region could be assessed by MRL.13 In this case, the retro-odontoid pannus completely resolved immediately following lateral mass distraction surgery. A brief report of this case is presented here.

Reduction of the retro-odontoid pannus in the immediate postoperative phase following distraction of the facets suggests that retro-odontoid pannus could be a result of laxity of the ligaments in the region. Distraction results in restoration of the tautness of these ligaments and reduction or obliteration of the indentation caused by the retro-odontoid pannus. Placement of spacers within the joint results in restoration of the height of the lateral mass and provides vertical stability to the atlantoaxial joint.

Conclusion

Lateral mass manipulation, fixation, and distraction with or without lateral mass plate and screw fixation may be an ideal form of surgical treatment for rheumatoid arthritis affecting the craniovertebral junction. It simultaneously treats atlantoaxial dislocation, basilar invagination, and the retro-odontoid pannus.

Case Illustration

A 35-year-old man was seropositive for rheumatoid arthritis and had multiple joints involved with disease. He had undergone several surgical therapies for several joints. He was on intermittent steroid drug treatment for ~5 years. For ~2 years, he was considerably disabled and had developed contractures of all four limbs. When admitted, he had spastic grade 3–4 quadripareisis and could walk with great difficulty using crutches. According to Ranawat's classification, the disability was considered grade IIIA. MRI and CT showed retro-odontoid pannus, fixed atlantoaxial dislocation, and basilar invagination with significant cord compression (Fig. 35.3a–c). The patient underwent surgery in the prone position and under cervical traction with the technical steps discussed by us earlier. The atlantoaxial joint was exposed after retracting the C2 ganglion superiorly. The facets were distracted using appropriate-sized osteotomes. The articular surfaces were drilled to resect the articular cartilage widely. A spiked titanium spacer 4 mm thick was impacted forcibly into the distracted joint. Multiple bone chips harvested from the iliac crest were inserted into the joint space along with the spacer. The patient recovered immediately after surgery, and the spasticity was reduced to a considerable extent. As per the protocol, postoperative CT and MRI were done on the day after surgery, after ~20 hours. The investigations showed reduction of the basilar invagination and atlantoaxial dislocation. MRI showed complete regression of the retro-odontoid pannus (Fig. 35.3g). At 18-month follow-up, the patient had satisfactory clinical recovery and could walk unaided (Ranawat grade II).
References

Spinal tuberculosis (TB) occurs in < 1% of patients with tuberculosis and in ~6% patients with extrapulmonary tuberculosis. Among patients with tubercular spondylitis, craniovertebral junction (CVJ) TB constitutes only 0.3% to 1.0% of cases. The worldwide resurgence of tubercular infection, especially with a high incidence of human immunodeficiency virus (HIV) infection, and the process of “reverse migration” have made this disease common not only in the endemic regions but also in areas where it was until now relatively unknown.

CVJ is the most mobile segment of the spine. CVJ TB may produce severe osteoligamentous destruction leading to atlantoaxial dislocation (AAD), cranial settling, cervical canal impingement, and compression of vital cervicomedullary structures. Destabilization of the joint complex by destroyed bones, ligaments, and articular surfaces results in abnormal translational and rotational movements. Even when the tuberculous infection responds to antituberculous therapy, the presence of abnormal mobility may cause recurrent cervicomedullary compression.

In this chapter, we present a brief review of the clinical and radiological features of CVJ TB and describe the management protocol.

Classification

Clinical Manifestations

Ramamurthi, quoting Boyd’s textbook of pathology, wrote: “There is no organ that TB cannot affect, there is no symptom that it cannot produce, and there is no disease that it cannot simulate.”

According to the reported literature, subaxial TB may have neurological involvement in up to 50% of patients, whereas CVJ TB may have neurological involvement in 63% to 70%. The most common manifestations are severe neck pain (with or without suboccipital pain), restricted neck movements, and torticollis. The severity of neck pain, feeling of instability at the upper cervical spine, and restricted neck movements often force these patients to support their chin constantly with their hands. Myelopathy, manifesting as various grades of weakness and spasticity, often associated with spinthalamic tract and posterior column dysfunction, is frequently present. The other important neurological manifestations are paresis of the ninth and tenth cranial nerves manifesting as dysphagia, nasal regurgitation, and hoarseness of voice; hesitancy; precipitancy of micturition; and sensation of incomplete evacuation. The respiratory status of these patients is often compromised and may be evaluated at bedside by a single breath count < 10 and a breath-holding time < 10 seconds. Often these patients have a history of systemic signs of TB in the form of weight loss, night sweats, and fever. A history of endemicity is very important, as these patients often live in an area endemic for tubercular infection. They often have a history of contact with a person with TB or may have a past history of partially or even completely treated TB. TB at other sites may also be present. Commenting on the correlation between spinal cord compression and neurological deficits, al-Mulhim et al. stated that < 50% narrowing of the cervical spine produces only mild to moderate deficits, and > 75% narrowing causes severe deficits. Because neurological deficits in CVJ TB are a result of cervicomedullary compression and instability, predicting and prognosticating the progression of neurological deficits on the basis of the extent of cervicomedullary compression may lead to a false sense of security. Even patients with minimal demonstrable compression may present with severe deficits. During the stages of healing, the persistence of various grades of subluxation may not correspond with either the degree of neural recovery or the quality of mechanical stabilization achieved. Fang et al. reported sudden death associated with CVJ TB probably due to unrecognized instability at the CVJ.

Edwards et al. listed the myriad presentations of CVJ TB:

1. Direct compression of the neuraxis by tubercular abscess or granulation tissue: these are usually extradural; however, intradural and intramedullary lesions have been reported.
2. Osteoligamentous destruction causing either AAD or upward translocation of dens causing secondary compression of the medulla
3. Combination of abscess formation with AAD
4. Abscess formation presenting with lower cranial nerve palsy or spinal nerve root compression leading to occipital pain with or without paresthesias
5. Dysphagia or airway compromise due to mechanical obstruction from the tubercular mass, which can extend to the posterior triangle or mediastinum
6. Neck pain, stiffness, occipital headache, torticollis, fever, night sweats, weight loss, and so on
7. Cervical lymphadenopathy and discharging sinuses
# Stages of Disease

Lifesø proposed three stages of CVJ TB:\textsuperscript{13}

Stage 1: minimal bony or ligamentous destruction; no AAD (Fig. 36.1)
Stage 2: minimal bony or ligamentous destruction; reducible or irreducible AAD present (Fig. 36.2)
Stage 3: significant bony or ligamentous destruction evident (Fig. 36.3)

Bhagwati et al. use the following grading system for various stages of the disease:\textsuperscript{14}

Grade I: merely inflammatory involvement of bony structures of the CVJ with formation of granulation tissue and destruction of bone
Grade II: formation of a large retropharyngeal abscess with bony changes
Grade III: associated subluxation of the atlantoaxial joint, by bony destruction and/or laxity of apical and transverse ligaments
Grade IV: formation of epidural abscess and compression of the cervicomедullary junction and the upper cervical cord, with neurological deficits that may be mild or severe

## Etiology

Discharged aerosolized pulmonary secretions containing tuberculous bacilli, consumption of unpasteurized milk infected with \textit{Mycobacterium bovis}, and inoculation with infected material may cause transmission of TB. The ability of TB bacilli to cause infection depends on the natural immunity and the antibacterial defenses of the person being infected. In 90% of infected persons, the organism remains dormant. The remaining 10% of individuals may present with early progressive disease within 5 years of exposure (5%) or with late recrudescent disease after several years of infection (5%).\textsuperscript{15}

Cell-mediated immunity and a delayed-type hypersensitivity immunological process develop in the host against \textit{Mycobacterium tuberculosis}. In cell-mediated immunity, the lymphocytes in the presence of tuberculous mycobacterial antigens produce local cytokines that attract monocytes and macrophages from the bloodstream into the lesion and activate them. Thus, it is a beneficial host response in which the activated macrophages produce reactive oxygen and nitrogen intermediates, lysosomal enzymes, and other products that kill and digest

## Pathophysiology

![Fig. 36.1a, b](a) Sagittal contrast-enhanced T1-weighted and (b) axial T2-weighted magnetic resonance imaging (MRI) showing a small amount of enhancing tubercular granulation tissue between the anterior arch of C1 and the odontoid process. There is a small intramedullary syrinx and an enhancing intramedullary lesion at the C2 level as well. The patient responded well to antituberculous therapy (ATT) and neck immobilization.
Fig. 36.2a–c

a Plain lateral radiograph of the craniovertebral junction (CVJ) showing fixed atlantoaxial dislocation (AAD).

b Contrast-enhanced T1-weighted image showing AAD with the granulation tissue between the anterior arch of C1 and the displaced odontoid, causing significant thecal compression. The granulation tissue extends beneath the posterior longitudinal ligament up to the subaxial spine.

c Plain lateral radiograph obtained during the follow-up examination after 5 years showing reduction of AAD with stable C1–C2 fusion performed by a modification of the fusion technique described by Brooks and Jenkins.


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Fig. 36.3a–d

a Plain lateral radiograph of the cervical spine showing destruction of the odontoid and the C2 body.

b Sagittal T2-weighted MRI showing fixed AAD, granulation tissue between the anterior arch of the atlas and the axis, and severe cervicomedullary compression.

c Computed tomography (CT) myelogram obtained at the follow-up examination after transoral surgery showing residual compression of the thecal sac at the upper border of the C3 vertebra.
the bacilli. The tissue-damaging delayed type of hypersensitivity reaction, by the same process as cell-mediated immunity, causes caseous necrosis, producing the death of local bacilli-laden nonactivated macrophages and nearby tissues.\textsuperscript{15,16}

The infection is divided into five stages. In stage 1, mature activated macrophages ingest and destroy the mycobacteria. In case the bacillus is not destroyed, it grows, multiplies, and destroys the activated macrophage. In stage 2, monocytes from the bloodstream reach the site of infection, convert to nonactivated macrophages, divide, and ingest the bacilli. The bacilli in turn multiply within the nonactivated macrophages. In stage 3, caseous necrosis occurs due to apoptosis produced by cytotoxic T cells and natural killer cells; anoxia due to thrombosis initiated by clotting factors produced by the macrophages; toxic cell products, such as free radicals, cytokines, tumor necrosis factor, hydrolytic enzymes, and complement; and toxic bacillary products, such as trehalose dimycolate. A delayed-type hypersensitivity reaction kills bacilli-laden macrophages. Thus, the center of the lesion is a solid caseous area where the extracellular bacilli (released due to macrophageal death) do not multiply. The nonactivated macrophages at the periphery permit intracellular bacilli multiplication. In this region, partially activated macrophages due to cell-mediated immunity also attempt to inhibit extracellular bacilli multiplication. In stage 4, if cell-mediated immunity is deficient, the bacilli at the periphery of the caseous necrosis multiply within peripherally situated nonactivated macrophages. A delayed hypersensitivity reaction kills these macrophages, causing enlargement of the caseous center and producing the clinically manifest disease. In cases with a good cell-mediated immunity, the activated macrophages ingest and destroy the bacilli, often arresting the lesion before it becomes clinically manifest. In stage 5, liquefaction of the caseous center occurs, and bacilli multiply extracellularly and overcome the cell-mediated immunity (as activated macrophages are ineffective in controlling extracellular multiplication of bacilli within a cavity).\textsuperscript{15,16}

Skeletal TB occurs due to the hematogenous spread of bacilli following primary infection. Lymphatic drainage from other organs in the vicinity of the spine, such as the pleura and kidneys, may spread to the periaortic lymph nodes with erosion within the spine. The lesion may be solitary or multiple. The anterior part of the vertebral body adjacent to the subchondral bony plate is most often affected. In children, the disk is vascularized; therefore, tuberculous diskitis may occur. In adults, diskitis usually follows a vertebral body infection. The narrowed disk space seen on radiology is often due to a collapsed vertebral end plate in adults. TB of the craniocervical region may cause neurological deterioration due to the formation of tuberculous abscess, granulation tissue, or vertebral body or disk sequestra that cause thecal compression; the destruction and incompetence of ligaments leading to AAD that may be irreducible (leading to a narrow spinal canal) or reducible (leading to recurrent cord injury due to repeated flexion and extension movements of the neck); or the collapse of anterior spinal elements, leading to a cervical kyphotic deformity. The angle of kyphosis depends on the loss of vertebral body volume. It progresses until the vertebral bodies meet anteriorly or

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{Fig_36.3d.png}
\caption{Axial CT images showing a skip tuberculous lesion at the T11 vertebra. The patient succumbed to septicemia following recurrence of CVJ and thoracic spinal tuberculosis (TB) leading to severe respiratory distress. (From Behari S, Nayak SR, Bhargava V, Banerji D, Chhabra DK, Jain VK. Craniocervical tuberculosis: protocol of surgical management. Neurosurgery 2003;52:72–81. Reprinted by permission.)}
\end{figure}
until the caseation and granulation calcify or ossify. An abscess may have subligamentous spread and move into adjacent ligaments along the subaxial spine or into soft tissue, producing a paraspinal abscess; may compress the pharynx, esophagus, or trachea; or even perforate a hollow viscus or body surface.

### Pathology

There are three types of tuberculous lesions:

1. **Exudative lesions**: there is migration of polymorphonuclear leukocytes, monocytes, and lymphocytes with vasodilation, edema, and a fibrinous exudate in the infected region.

2. **Proliferative lesions (tubercles)** (Fig. 36.4): these are granulomas containing numerous macrophages, epithelioid cells, and lymphocytes. The epithelioid cells within these tubercles are the activated macrophages with a vesicular nucleus. Langhans multinucleated giant cells are formed by fusion of two or more macrophages that surround caseous tissue too large for one cell to engulf.

3. **Composite lesions with both exudative and proliferative components**: within the center of the cellular reaction is a necrotic amorphous caseative necrosis. Fibrosis and calcification may surround the granuloma or occur in its center.

In patients with acquired immunodeficiency syndrome (AIDS), HIV reduces both the number and the functional capacity of CD4+ cells, which produce macrophage-activating factors. In patients with good CD4+ count levels, the pathology is similar to that of classic TB. When CD4+ counts decrease, granuloma formation decreases, and there may be a large number of bacilli and positive mycobacterial cultures. Atypical mycobacterial infection also increases.

### Diagnosis

#### Investigations

1. **Complete blood picture** shows a relative serum lymphocytosis and raised erythrocyte sedimentation rate (ESR).
2. **Acid fast staining**: the acid fast staining may be performed using (a) carbol-fuchsin methods, including the Zeihl-Neelsen and Kinyoun staining procedures, which stain the mycobacterial walls red against a methylene blue counterstain; and (b) the fluorochrome method using auramine O or auramine-rhodamine dyes, which stain the mycobacterium wall yellow to golden against a dark background. The Zeihl-Neelsen staining method uses heated carbol-fuchsin, and Kinyoun staining uses cold phenol to increase carbol-fuchsin penetration into the cell wall. The disadvantage of acid fast staining is the indiscriminate staining of both viable and nonviable mycobacteria.
3. **Culture**: mycobacteria are slow growing and have a generation time much longer than common bacterial and fungal flora. Thus, overgrowth of the latter may occur when the specimen is obtained in a nonsterile manner. The mycobacterial cell wall has high lipid content and is more resistant to strong acids and alkalis. Alkaline solutions such as benzalkonium chloride and 4% sodium hydroxide may be used to eliminate bacteria from the contaminated specimens. The culture media on which the mycobacteria may be inoculated include (a) egg-based media: these include whole eggs, potato flour, salts, glycerol, and malachite green, as well as Lowenstein-Jensen, Petragnani, and the American Thoracic Society media; (b) agar-based media, including the Middlebrook 7H10 and 7H11 media containing agar, organic compounds, salts, glycol, and albumin (7H11 also contains casein hydrolysate, which enhances growth of mycobacteria resistant to isoniazid); (c) selective media: Lowenstein-Jensen and Middlebrook selective 7H11 media are examples. Their base media include antimicrobial agents to inhibit contaminating bacteria. All media should be incubated for 6 to 8 weeks at 35°C within the first 3 to 4 weeks in an atmosphere of 5% to 10% CO₂. The colonies (rough, cauliflower-like, and colorless) should be subjected to an acid fast smear and subculture for identification. Nucleic acid probe testing may also be performed on them.

For faster detection of mycobacteria (in ~10–12 days), BACTEC 460TB (Becton, Dickinson, Franklin Lakes, New Jersey) with liquid Middlebrook 7H12 medium containing 14C-labeled palmitic acid may be used. The growth of mycobacterium releases 14C, which is detected by a sensor. Deoxyribonucleic acid (DNA) probe identification and antibiotic susceptibility may also be performed using this system.

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Fig. 36.4 Microphotograph showing bone TB with a characteristic granuloma with caseation, as well as epithelioid and foreign body giant cells (hematoxylin and eosin × 100).
4. **Chromatography**: with gas liquid chromatography and high-performance liquid chromatography, the long-chain fatty acids extracted from *M. tuberculosis* yield species-specific chromatographic peaks that may be used for the identification of mycobacteria. The results may be available within a few hours.

5. **Nucleic acid probes**: mycobacterial cells are lysed. An acridinium-labeled DNA binds with the ribosomal ribonucleic acid (RNA) of the mycobacterium. The resultant DNA-RNA complex exhibits a chemiluminescence that may be measured in a luminometer.

6. **Detection of drug-resistant *M. tuberculosis***: drug resistance may be seen in patients who have already been treated for TB, patients living in endemic areas, and contacts of patients with resistant TB. When the *M. tuberculosis* is inoculated in the agar medium or in the BACTEC medium, the failure of the organism to multiply or to produce ¹⁴CO₂, respectively, in the presence of antibiotic is indicative of antibiotic susceptibility. Detection of molecular mutations associated with specific drug resistance may also be studied.

7. **Nucleic acid amplification methods**: these facilitate early detection of *M. tuberculosis* by target, probe, or signal amplification systems. A transcription-based amplification system generates multiple copies of mycobacterial ribosomal RNA after the nucleic acids are released from the mycobacterial cells by sonication and their secondary structure has been disrupted by heat. Chemiluminescent-labeled DNA probes then detect these amplified ribosomal RNA. Another method uses a polymerase chain reaction–based assay that detects the ribosomal gene in the genome (DNA). The amplification is facilitated by oligonucleotide polymers, and the color developed by the horseradish peroxidase system detects the mycobacterium. The total time required with these methods is 5 to 7 hours. These tests, however, must be used in conjunction with conventional cultures and staining techniques or the diagnosis of TB.

8. **Tuberculin skin testing**: this is used to diagnose tuberculous infection and not tuberculous disease. Subcutaneous injection with *M. tuberculosis* with tuberculin, composed primarily of tuberculoprotein obtained from heat-sterilized filtered and concentrated cultures of tubercle bacilli, induces a delayed type of hypersensitivity reaction in the infected patient. The initial tuberculous infection brings about a period of 6 to 8 weeks of sensitization when the sensitized T lymphocytes developing in the regional lymph nodes enter circulation. Restimulation of these lymphocytes by tuberculin subcutaneous injection in the form of purified protein derivative (0.1 mL of 5 tuberculin units; the Mantoux test) causes an indurated skin reaction in infected patients that is maximal at 48 to 72 hours and lasts for more than 96 hours. A reaction size < 5 mm after 48 to 72 hours clearly separates the reactors and nonreactors. In the presence of HIV virus infection, a reaction > 5 mm to 5 tuberculin units of protein purified derivative is considered positive for tuberculous infection; a reaction size > 10 mm is considered positive in the high-risk population, such as those belonging to countries with a high tuberculosis prevalence, intravenous drug users, residents of long-term facilities, such as prisons and mental institutions, those having conditions where the risk of tuberculosis is increased, such as diabetes mellitus, those on immunosuppressive treatment, and those with hematological and other malignancies. A reaction > 15 mm is considered positive in other persons. A negative reaction to tuberculin testing does not rule out tuberculosis infection, as this may occur in renal failure, immunosuppression, AIDS, malignancies, fulminant bacterial and tuberculosis infection, and malnutrition. Positive reactions are obtained when patients have been infected with *M. tuberculosis* but do not have active disease, and when persons have been sensitized to nontuberculous mycobacteria or bacille Calmette-Guérin (BCG) vaccination (derived from *M. bovis*).

9. **Cytokine release assays**: QuantiFERON-TB (Cellestis, Carnegie, Victoria, Australia) is used for the diagnosis of latent tubercular infection in a population with low to moderate risk. The test’s performance will probably be enhanced by the use of antigens such as ESAT-6 and CFP-10 that are present in *M. tuberculosis* but absent from BCG strains and most nontuberculous mycobacteria.

10. Serological evaluation for HIV 1 and 2 in immunocompromised patients

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**Radiological Investigations**

Two types of TB are recognized. The caseous exudative type is characterized by more destruction, more exudation, and abscess formation. The granular type is less destructive, and abscess formation is rare. Generally, it is a dry lesion. The neurological deficits may be due to tuberculous abscess, granulation tissue, AAD, or cranial settling. Vascular compromise due to endarteritis associated with local meningitis may also be the cause of neurological deterioration in some cases. Lifeso and Shukla et al. suggested that CVJ TB results from infection that spreads from the retropharyngeal space to the osseous elements. A primary infection of the atlas or axis is rarely a cause of the disease. Lateral radiographs of the CVJ in neutral, flexion, and extension positions may show the presence of an AAD (atlantodental interval > 3 mm in adults and > 4 mm in children). This may be reducible, partially reducible, or irreducible in nature. If destruction of the anterior arch of the atlas and odontoid process by the tuberculous process leads to loss of conventional radiological markers used to diagnose AAD, such as the anterior arch of the atlas and the odontoid, the diagnosis...
of AAD and its reducibility may be made on the basis of the position of the posterior arch of the atlas relative to the spinolaminar line on lateral plain films of the CVJ.\textsuperscript{22,21} A prevertebral soft tissue shadow and bony destruction may also be visualized. An abnormal prevertebral soft tissue shadow \( > 7 \) mm measured at the lower border of the axis is often diagnostic.\textsuperscript{1} The other radiological features of CVJ TB are resorption of the dense margin of the end plate of the axis, C2–C3 disk space narrowing or obliteration, lytic destruction of the anterior portion of the vertebral body, increased anterior wedging and kyphosis, collapse of one vertebral body on another, and reactive sclerosis superimposed on a progressive lytic process. The plain film changes may lag behind the pathological alterations in TB spondylitis by 2 to 6 months because radiological evidence of bony destruction appears only after \( \sim 50\% \) of the vertebra is destroyed.\textsuperscript{22}

A computed tomography (CT) scan provides greater detail than plain radiographs. Lytic bony erosions, disk prolapse, end-plate sclerosis, and disruption of the cortical margin of vertebral bodies are well delineated. Contrast-enhanced images may show the soft tissue spread of granulation and cseation or of abscess formation. Sagittal reconstructed images may show the AAD and cervical kyphosis and bony destruction and cervical instability.

Magnetic resonance imaging (MRI) of the CVJ with gadolinium enhancement is the investigation of choice in these lesions. Patients often show skip lesions of the vertebral column. MRI is often useful in picking up extensions of C1–C2 TB lesions to the subaxial spine that may not be evident on plain film. Coronal images may show extensive paraspinal abscesses (Fig. 36.5). MRI of the active forms of CVJ TB may show low signal intensity on T1-weighted images and high signal intensity on T2-weighted images in the affected vertebral bodies because normal fat marrow is replaced by edematous tissue infiltration (Fig. 36.6). The limitation of T2-weighted imaging in TB is due to indiscriminate increase in the signal intensity from edema, abscesses, loculations, and tissue hyperemia.\textsuperscript{9} Other radiological features of TB are involvement of the posterior elements with complete sparing of the anterior elements, extradural spinal cord compression without radiological evidence of bony involvement, and destructive lesions of bone with subcutaneous masses (Fig. 36.3).\textsuperscript{23} Arachnoiditis and syringomyelic changes may occur in tuberculous myelomyelitis.\textsuperscript{1} CT and MRI of the CVJ in the early phase of tuberculosis may demonstrate bony fragmentation at the vertebral end plates and an associated obliteration of fat planes around the vertebral body as early signs of abscess formation.\textsuperscript{22} A multiloculated calcified abscess with a thick, irregular-enhancing rim in the presence of vertebral body fragmentation may occur. CT and MRI may also demonstrate disk space narrowing, a paravertebral soft tissue mass, vertebral involvement, and the presence of AAD. Contrast-enhanced images may reveal the spread of tubercular abscess and granulation tissue along the vertebral bodies under the longitudinal ligament and in the epidural plane with associated thecal sac compression. Other lesions at the CVJ, such as rheumatoid arthritis, sarcoidosis, brucellosis, fungal infections, lymphoma, and chordoma, may produce a radiological appearance similar to that of CVJ TB except that caseating necrosis leading to abscess formation only occurs in the latter condition and is virtually diagnostic of TB.\textsuperscript{22} Fat suppression techniques with gadolinium DTPA (diethylenetriamine pentaacetic acid) enhancement may prove to be a better option than T1-weighted enhanced studies because they potentially improve the contrast between the hyperemic osseous and soft tissue involvement and normal structures. Intramedullary tuberculosis may occur at the CVJ. MRI may reveal a rim-enhancing hypo- to isointense lesion on T1-weighted images and a
hypointense area with a variable center of hyperintensity reflecting caseating necrosis on T2-weighted images. Although intramedullary tuberculomas often respond to antituberculous therapy (ATT) alone, some patients may have progressive neurological deficits despite treatment, partially due to swelling of the lesion as the bacilli release irritative material and/or due to cord edema (Fig. 36.1).

In CVJ TB, serial follow-up T1-weighted MRI may be used to prognosticate the response to ATT. When the patient starts responding to ATT, T1-weighted images show a progressive increase in signal intensity within the previously affected vertebrae suggesting fatty marrow involvement. The corresponding T2-weighted MRI shows complete resolution of the altered signal pattern that was earlier present due to granulation tissue and pus formation correlating with clinical improvement. There may be a significant decrease in gadolinium contrast enhancement of the granulation tissue in patients responding to ATT (Fig. 36.7). Chest radiographs should also be done in these patients who may show either an active tuberculous lesion or evidence of healed TB.

### Treatment

#### Historical Aspects of Management

Historically, management of CVJ TB has varied from conservative management to radical surgical removal. Tuli reported 24 cases of TB CVJ in 1974 and treated them with medical therapy for 18 months, prolonged bed rest, and cranial traction with the neck held in extension. Lal et al. discussed six cases of tubercular AAD. Bhagwati reported three cases of CVJ TB with destruction of the occipital condyles and six cases of TB AAD seen over a period of 6 years. Fang et al. proposed a one-stage anterior cervical débridement and fusion. Lal et al. proposed a one-stage posterior decompression and internal fixation with the use of metal prosthesis and bone grafts followed by immobilization of the neck using a hard cervical collar. Lifeso and Edwards et al. proposed a two-stage procedure of transoral decompression with posterior fusion with an interval of 3 months between the two with the patient’s neck immobilized in a halo brace. This would allow sensitivity of mycobacterium to be determined to ensure that the patient is receiving appropriate ATT prior to stabilization.

#### Antituberculous Therapy

ATT forms the mainstay of therapy for all patients with CVJ TB. A four-drug oral regimen of ATT includes rifampicin, isoniazid, ethambutol, and pyrazinamide. Pyridoxine 20 mg/day is given along with isoniazid to prevent peripheral neuritis. Occasionally, intramuscular injections of streptomycin may be administered (a total of 90 injections). Table 36.1 gives a brief summary of common ATT.

#### Duration of ATT

The World Health Organization guidelines for national programs recommend treatment regimens for each diagnostic category of TB (Table 36.2). The duration of ATT recommended in these guidelines is much shorter than that reported for spinal TB in the neurosurgical literature. Most of the studies on CVJ TB recommend that pyrazinamide be continued for ~3 to 4 months, streptomycin (intramuscular injections) for 3 months, ethambutol for 1 year, and isoniazid and rifampicin for 15 to 18 months. If the patient develops liver dysfunction, isoniazid, rifampicin, and pyrazinamide are discontinued. Ethambutol is given along with streptomycin and ofloxacin until liver functions become normal. After that, isoniazid and rifampicin may be given for a total of 15 months as per the original protocol.
Second-line Antituberculous Therapy

There has been a growing apprehension about the increasing incidence of resistance of tuberculous infection to the primary line of drugs. Second-line ATT consists of the following drugs:

1. Quinolones: mode of action presumably is the prevention of DNA synthesis. Adverse effects are relatively uncommon, but fluoroquinolone-resistant TB is a source of growing concern. In children younger than 10 years, they may cause arthropathy.

2. Capreomycin: after streptomycin, capreomycin is the injectable drug of choice. It is similar to streptomycin in terms of dosing, mechanism of action, pharmacology, and toxicity. Cross-resistance with streptomycin is uncommon.

3. Rifabutin: recommended in place of rifampicin for treatment of HIV-positive patients who are also taking a protease inhibitor. Rifabutin is active against some strains of rifampicin-resistant *M. tuberculosis* and is more active than rifampicin against *M. avium* complex and other nontuberculous mycobacteria.

4. Amikacin: besides *M. tuberculosis*, is active against rapidly growing mycobacteria

5. Ethionamide: most useful in the treatment of multidrug-resistant TB. Ethionamide is bacteriostatic against metabolizing *M. tuberculosis* and some nontuberculous mycobacteria. It is widely distributed throughout the body, including the cerebrospinal fluid (CSF).

6. Para-aminosalicylic acid: rarely indicated

7. Cycloserine: has an excellent CSF penetration, but its use is limited by side effects such as psychosis, seizures, peripheral neuropathy, headaches, and somnolence

8. Rifapentine: as compared with rifampicin, is longer acting and administered in a dose of 600 mg once or twice a week, thereby increasing patient compliance. However, it is associated with significant side effects and must be used with caution.

Fig. 36.7a, b

a. Sagittal T2-weighted MRI showing tuberculous pus and granulation anterior to C1–C2 with subligamentous extension up to C4. There is no thecal compression.

b. Sagittal T2-weighted MRI after antituberculous therapy (ATT) and neck immobilization showing complete disappearance of the altered signal pattern due to pus and granulation.

Prophylaxis

Intracutaneous injection of BCG vaccination must be considered for uninfected persons when there is a high risk of being infected. Vaccination programs among infants and young children in endemic areas may help to decrease the infection rate. However, whether or not it is really protective against the development of frank TB in humans has not been established conclusively. In endemic areas, most persons become tuberculin reactors over the course of time. Depressed cellular immunity in AIDS and other immunosuppressive states are considered contraindications to vaccination with BCG.

Preventive therapy consists of oral administration of isoniazid in patients already infected where the risk of disease is high. It is given in the dose of 5 to 10 mg of isoniazid per kilogram of body weight, not exceeding a total dose of 300 mg/day in a single daily dose, for 6 to 12 months.

Management Protocol

The factors that influence the management protocol include the neurological grade of the patient, the extent of bony destruction, the degree of cord compression, the presence of associated AAD, and the clinical response to ATT.27 Goel et al have discussed the significance of site of involvement of tuberculosis in the lateral masses, mode of spread, degree of destruction, nature of progress of neurological symptoms and the response to drug treatment in the decision making regarding surgery.28

Patients with CVJ TB may be divided into two groups on the basis of severity of clinical presentation (Fig. 36.8):2

- Group A: those with minor deficits, including those with neck pain without pyramidal involvement, and those with minor disability not interfering with daily living
- Group B: those with major deficits, including those partially or completely dependent for activities of daily living

Patients with CVJ TB may fall into three categories: minor deficits (grades I and II), severe deficits (grades III and IV), and patients with CVJ TB with minor deficits after 3 months in whom conservative management under cover of ATT had been adopted (Figs. 36.8 and 36.9).

### Table 36.1 Summary of common antituberculous drugs with dosage and adverse reactions

<table>
<thead>
<tr>
<th>Drug</th>
<th>Mechanism of Action and CNS Penetration</th>
<th>Daily Dosage</th>
<th>Adverse Effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isoniazid</td>
<td>Inhibits cell wall (mycolic acid) biosynthesis; bactericidal for actively growing organisms; penetrates CNS</td>
<td></td>
<td>Fever, skin rash, hepatitis, peripheral neuropathy, agranulocytosis, thrombocytopenia</td>
</tr>
<tr>
<td>Rifampicin</td>
<td>Inhibits DNA-dependent RNA polymerase and interferes with RNA synthesis; effective against slow-growing and intracellular organisms; bactericidal with CNS penetration</td>
<td></td>
<td>Nausea, vomiting, dark urine, hepatitis</td>
</tr>
<tr>
<td>Pyrazinamide</td>
<td>Bactericidal to intracellular organisms in an acidic environment; used during first 3 months</td>
<td></td>
<td>Hepatitis, rash, gout</td>
</tr>
<tr>
<td>Ethambutol</td>
<td>Bacteriostatic; inhibits glucose incorporation into cell wall</td>
<td>15–25 mg/kg</td>
<td>Optic neuritis, gout</td>
</tr>
<tr>
<td>Streptomycin</td>
<td>Bactericidal against actively growing organisms; enters CNS only following meningeal inflammation</td>
<td>20–40 mg/kg</td>
<td>Ototoxicity, nephrotoxicity</td>
</tr>
<tr>
<td>Ofloxacin</td>
<td>Inhibits coiling action of DNA gyrase and disrupts DNA</td>
<td></td>
<td>Gastrointestinal upset, hypersensitivity, mild CNS reactions; in children and pregnancy, causes arthropathy</td>
</tr>
</tbody>
</table>

b.i.d., twice a day; CNS, central nervous system; DNA, deoxyribonucleic acid; IM, intramuscular; RNA, ribonucleic acid.
Table 36.2  WHO guidelines for recommended treatment regimens for each diagnostic category of tuberculosis

<table>
<thead>
<tr>
<th>Diagnostic Category</th>
<th>Patients</th>
<th>Initial Phase* (daily or 3 times weekly)</th>
<th>Continuation Phase* (daily or 3 times weekly)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>New smear-positive patients; new smear-negative pulmonary TB with extensive parenchymal involvement; severe concomitant HIV disease or severe forms of extrapulmonary TB</td>
<td>2HRZE</td>
<td>4HR or 6HE daily</td>
</tr>
<tr>
<td>2</td>
<td>Previously treated sputum smear-positive pulmonary TB; relapse; treatment after interruption; treatment failure</td>
<td>2HRZES/1HRZE</td>
<td>5HRE</td>
</tr>
<tr>
<td>3</td>
<td>New smear-negative pulmonary TB (other than in category 1); less severe forms of extrapulmonary TB</td>
<td>2HRZE</td>
<td>4HR or 6HE daily</td>
</tr>
<tr>
<td>4</td>
<td>Chronic and MDR TB (still sputum positive after supervised treatment)</td>
<td>Specially designed standardized or individualized regimens used</td>
<td>Specially designed standardized or individualized regimens used</td>
</tr>
</tbody>
</table>

*The number in front of the abbreviated names of the drugs represents the number of months for which the drugs are given. E, ethambutol; H: isoniazid; HIV, human immunodeficiency virus; MDR, multidrug-resistant; R, rifampicin; S, streptomycin; TB, tuberculosis; WHO, World Health Organization; Z, pyrazinamide.

Fig. 36.8b–c

b Protocol for management of patients with CVJ TB with minor deficits after 3 months in whom conservative management under cover of ATT had been adopted.

c Protocol for management of patients with CVJ TB with major deficits.

Fig. 36.9a–d  MRI showing extensive odontoid destruction due to tuberculous lesion at the CVJ in a neurologically intact patient. Despite the extensive radiological lesion that appeared to be causing significant thecal compression, the patient improved with conservative management.

If there is no demonstrable mobility at the CVJ, neck movements may be stabilized with a hard cervical collar, halo brace, or Minerva jacket, and ATT is continued. If there is an associated reducible AAD, either a posterior stabilization procedure is performed directly, or rigid immobilization of the neck with a halo brace or Minerva jacket is continued. If there is an associated large retropharyngeal abscess, translateral aspiration may be required before immobilization.

This protocol may be continued for 3 months, after which patients with minor deficits undergo a detailed neurological examination, dynamic translateral radiographs of the CVJ in flexion, extension, and neutral positions, and MRI. Generally, these patients demonstrate good clinical response in the form of disappearance of pain and systemic features of TB with neurological recovery. Radiological signs also improve in the form of decrease in granulation tissue and paraspinal abscesses, as well as the presence of new bone formation. Further management in these patients is continued by having the neck stabilized using a hard cervical collar.

After 3 months, if a reducible AAD is demonstrated on dynamic translateral radiographs of the CVJ (after removal of the external orthoses), direct posterior fusion is performed, after which the patient is continued on a hard cervical collar until a bony union is obtained. If the AAD remains irreducible at the evaluation following 3 months of ATT, there is significant canal compromise, and/or there is delayed neurological deterioration after an initial recovery, anterior decompression followed by posterior fusion is done. After surgery, neck movements are stabilized on a hard cervical collar until a bony union is obtained in ~3 to 4 months. If there is fixed AAD but no neurological involvement, the patient may be continued on a hard cervical collar.

If, after 3 months of ATT, no clinicoradiological response is seen or there is progressive neurological deterioration, a reassessment for immunocompromised status, diabetes mellitus, or some other diagnosis other than TB, as well as the possibility of nonresponsiveness or noncompliance to ATT, should be considered.2,3

Patients with severe deficits (Fig. 36.8c) after undergoing flexion, extension, and neutral translateral radiographs of the CVJ are usually not treated conservatively, as they are at high risk of developing the problems related to their recumbent status, such as respiratory infection, bedsores, and deep vein thrombosis (DVT).17,27 Thus, if there is anterior compression due to pus, granulation tissue, or destroyed bone, they should undergo anterior decompression with posterior fusion and then should be maintained on a hard cervical collar. Early fusion with implant imparts greater rigidity to the spine and has been shown to decrease progression of spinal infection...
and kyphotic deformity. If there is associated reducible AAD, a direct posterior stabilization procedure is done. If, however, the dislocation is irreducible, a trial of cervical traction is done to attempt reduction of dislocation. If the fixed AAD persists despite cervical traction, anterior decompression with posterior fusion is performed, and the patient is continued on a hard cervical collar. If after cervical traction the dislocation becomes reducible, direct posterior fusion is performed.2,7

### Rationale for the Current Protocol

The management plan incorporates both conservative and radical lines of management. Patients with CVJ TB usually respond very well to ATT, so that the granulation tissue resolves, and the destroyed bone heals. Thus, often the only treatment that is necessary is the institution of ATT and immobilization of the neck to relieve the patient’s neck pain and to promote healing. A good response to ATT may precipitate an intense fibrous reaction, which may obviate the need for additional craniovertebral stabilization. A clinicoradiological response takes 6 to 12 weeks to manifest after the institution of therapy. Patients with minor myelopathic features usually can tolerate conservative management without complications during the period in which the trial of ATT is performed, after which surgical intervention may be avoided altogether. In those patients who already have severe myelopathic features, however, it is always better to offer radical anterior decompression and posterior stabilization than to treat them with prolonged bed rest and immobilization, which carry well-known risks such as respiratory problems, bedsores, and deep vein thrombosis.2

### Is Tissue Diagnosis Always Necessary?

The first step in the management of CVJ TB is confirmation of the tissue diagnosis. It is mandatory to have a tissue diagnosis as radiological features are often not specific for TB. The specimen may be obtained by either CT-guided biopsy or transoral decompression.20,27 Edwards et al. emphasized that the radiological appearance of CVJ TB cannot be distinguished from other lesions, especially when a retropharyngeal abscess is not a common feature.12 The emergence of multidrug-resistant mycobacterial strains requires confirmation of sensitivity prior to therapy. However, a tissue diagnosis may not always be possible in CVJ TB. Serious complications have been reported while obtaining a histological sample via the transoral transpharyngeal route.21 The mycobacterial cultures may be negative. Valuable time may be lost while awaiting the results. The choice of antituberculous agents is limited and cannot be guided solely by the sensitivity patterns of the organism, because the second line of ATT has many adverse reactions; also, obtaining sensitivity to the chemotherapeutic agents is relatively expensive, particularly for people from low socioeconomic strata, who comprise the majority of patients with TB. Thus, obtaining a tissue diagnosis in all circumstances is not practical. A dramatic clinicoradiological response to ATT in a susceptible individual (immunocompromised, living in an endemic area, having an increased ESR or positive Mantoux test, TB at other sites, history of contact with a patient with TB, and/or history of TB) with the classic symptomatology (neck pain, neck movement restriction, and cervical myelopathy) is often a reliable guide to the presence of tuberculous infection of the CVJ.1,2

### Surgical Technique

Anterior decompression may involve either the transoral or the retropharyngeal approach. Transoral decompression is performed through the transpalatal, transpharyngeal route, and the anterior surfaces of the clivus, anterior arch of C1, odontoid, and body of C2 are drilled. The posterior longitudinal ligament and the tectorial membrane are incised, exposing the dura.

Following the transoral decompression, extensive osteocartilaginous excision requires a simultaneous posterior fusion, which may be performed under the same anesthesia to avoid the problems of instability and prolonged traction.29–31

In the anterior retropharyngeal approach, using a transverse submandibular incision, the submandibular gland, digastic and stylohyoid muscles, and hypoglossal nerves are mobilized upward. The suprahyoid dissection is continued in the retropharyngeal space between the carotid sheath laterally and the tracheoesophageal complex medially. The prevertebral fascia is incised for decompression of the epidural abscess, and the granulation tissue, along with destroyed vertebrae, is excised. The latter approach avoids the potentially contaminated oral cavity. The disadvantages include a wide anatomical dissection of the cervical fascial planes and a superior oblique trajectory that makes decompression of the ipsilateral epidural space difficult. There may be difficulty in gaining access to the rostral part of the dens and C1. Hence, if there is significant cranial settling due to tubercular destruction, transoral decompression may be preferable for complete decompression.32

For atlantoaxial posterior fusion, various techniques are available, including sublaminar wiring with strut and onlay bone graft; flexible multistrand, braided titanium cables; metallic rods and plates of various shapes; Halfix clamp stabilization; transarticular screw fixation; and lateral mass plate and screw fixation.33–35

Serious complications have been reported during positioning, biopsy, and surgery of the lesion. Knutti and Kaech described quadriplegia with respiratory arrest following transoral biopsy and attributed it to the perioperative hyperextension of the neck during positioning of the patient.36 Edwards et al. recommended fiberoptic intubation to be used in all patients in whom the cervicomedullary
juncion is at risk. Spinal cord monitoring using somatosensory evoked potentials is recommended during positioning and the procedure. A halo brace has been recommended prior to the simultaneous performance of transoral decompression and posterior fusion under the same anesthesia that is continued throughout both surgeries.

### Prognosis

There are no specific radiological predictors for outcome. Even patients with preoperative MR signal intensity changes in the cord on T2-weighted imaging may show a good response. The causes of such signal changes are myelomalacia, edema, ischemia, gliosis, and demyelination. In CVJ TB, these signal changes are often reversible and may not affect the final outcome. Long-term follow-up is recommended, and the importance of compliance to ATT must be emphasized to each patient. Repeat imaging at 6 and 12 months following completion of ATT should be performed to ensure that there is no evidence of recurrence of infection or failure of stabilization.

### Conclusion

The combination of ATT, decompression, and stabilization often results in marked improvement even in patients who are partially or completely dependent on others for their daily needs. Thus, this is one of the rare illnesses that have the potential to cause severe morbidity, even sudden death, and the likelihood of significant clinical improvement with the judicious use of therapeutic options in association with ATT.

### References

1. Tuli SM. Tuberculosis of the Skeletal System. 2nd ed. New Delhi: Jaypee Brothers; 1997
Tuberculous involvement of the craniovertebral junction (CVJ) is still seen frequently in India and other developing countries. Tuberculosis (TB) of the CVJ usually occurs secondary to a primary focus elsewhere in the body. Osteoligamentous destruction and deformities leading to a range of clinical presenting symptoms have been recorded. An understanding of the site of involvement of TB in the CVJ, the pattern of its spread, and the nature of its pathogenetic effects on the osteoligamentous assembly is crucial for defining the management strategy. This chapter covers the staging of the disease, indications for surgery, and long-term outcome.

### Natural Course of Disease

From our experience with the subject in general, and TB in particular, we identified the following stages of the disease, depending on the more commonly encountered pattern of bone and joint involvement. The cancellous part of the bone is most susceptible to TB. The cortical part is affected late, and the joints are secondarily involved.

#### Stage 1

In stage 1, there is unilateral involvement of the cancellous part of the facet of the atlas but no destructive deformation. Less frequently, there may be isolated and unilateral involvement of the cancellous part of the facets of the axis or of the odontoid process. Inflammatory granulomatous reaction is present, and caseous necrosis may be seen. Granulation tissue is usually located around the involved facet. The other parts of the atlas (or axis) bone and the contralateral facet are not involved (Fig. 37.1). In this stage, the patient experiences pain in the neck and restriction of neck movements. Systemic symptoms, such as loss of weight, loss of appetite, and fever, are usually present.

#### Stage 2

In stage 2, the disease progresses to involve the atlantoaxial joint by destructive necrosis and inflammation. The joint involvement is a result of extension of the inflammatory...
reaction. The destruction involves the atlantoaxial joint complex and extends to other parts of the atlas and/or axis bones. Tuberculous inflammation may extend widely. The contralateral joint is still unaffected in this stage. The incompetence of the joint and osseous destruction and the adjoining ligamentous disruption in such a situation have been known to result in atlantoaxial dislocation. The atlantoaxial dislocation is probably a result of the ineffectiveness of the alar and transverse ligaments, as their bone attachment site is destroyed. Because the contralateral atlantoaxial joint is normal, the atlantoaxial dislocation is of “fixed” and rotatory variety, and grossly mobile and reducible dislocation is seldom encountered. The facet of the atlas may be collapsed. Prevertebral or extradural spinal caseous necrosis or pus formation is usually encountered. On imaging, the joint space on the involved side is seen to be reduced or absent, whereas on the contralateral side, it is normal (Figs. 37.2, 37.3, and 37.4).
In this stage, the patient exhibits pain in the neck, neck muscle spasm, and severe restriction of neck movements. Torticollis is the characteristic and most prominent symptom. It appears to be a natural defense process, in which the neck turns to the contralateral side in an attempt to reduce all weight bearing by the affected lateral mass and the joint and protect the spinal cord from compression by infective granulation.

The patient may or may not have neurological symptoms or deficits.

**Stage 3**

In stage 3, the disease involves the contralateral atlantoaxial joint and other bones and joints in the region. Evidence of instability of the CVJ is usually seen (Figs. 37.5 and 37.6).

In this stage, the patient usually has a neurologic deficit.

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**Indications for Surgery**

Neurological deficits are notably delayed and less pronounced despite the aggressive destruction and deformation by TB. The disease is initially unilateral, and the contralateral atlantoaxial joint is spared until late in the disease process. Because of the relatively stable craniovertebral region, despite the unilateral facet destruction, and the effectiveness of current antituberculous drugs, surgery can be delayed or avoided.

The extent and nature of involvement of the joints are determining factors for any kind of surgery for stabilization of the region. Surgery for evacuation of pus or granulation tissue is seldom required. Despite the extentiveness of the bone destruction and inflammation, the patient is usually neurologically quite stable, and the systemic symptoms are not pronounced, until late in the
Fig. 37.4a–c A 20-year-old man presented with mild quadriplegia, neck pain, and fever. He was already improving on conservative drug therapy.

a  Sagittal view showing destruction of C2 and the lateral masses. Caseous necrosis and bone destruction can be seen.

b  MRI showing evidence of “fixed” atlantoaxial dislocation.

c  Coronal image of MRI showing unilateral destruction of the lateral masses of the atlantoaxial joint and adjoining inflammation.

Fig. 37.5a–c A 58-year-old woman had severe neck pain and quadriplegia. She was treated successfully with antituberculous drugs.

a  Axial CT showing extensive destruction of the body and lateral masses of the axis vertebra.

b  Sagittal MRI showing destruction and inflammation of the body of C2.

c  Coronal MRI showing bilateral involvement of the facets.
A 65-year-old male patient presented with severe quadriplegia. Following surgery that involved posterior fixation, he showed remarkable clinical improvement.

- **a** Axial CT scan showing extensive bilateral destruction of the atlas bone.
- **b** Sagittal CT showing pathological fracture of the odontoid process.
- **c** Sagittal T1-weighted MRI showing extensive tuberculous destruction and inflammation of the CVJ.
- **d** Sagittal T2-weighted MRI showing destruction and inflammation.
- **e** Postoperative radiograph with the neck in flexion showing lateral mass plate and screw fixation of the atlantoaxial joint.
disease process. In the absence of progressive neurological symptoms and in the presence of an intact contralateral atlantoaxial joint, surgery can be avoided. The patient needs to be placed on antituberculous drugs. In addition, the patient will need a firm four-poster cervical collar, and all activities related to neck movements and weight bearing should be avoided for at least 3 months or until there is evidence of bone healing and regression of symptoms.

Surgery is indicated less frequently and may be required in stages 2 and 3 of the disease. The most positive indication for surgery is the progression of neurological deficits. Radiological deformity in the presence of an intact neurological condition may be only a marginal indication for surgery. The more crucial issues are to identify and relate the problem to TB and to differentiate the lesion from several tumorlike pathological situations.

If the disease is localized to the atlantoaxial region, surgery can involve fixation of the contralateral atlantoaxial joint by lateral mass plate and screw fixation. Our experience suggests that even if fixation is done unilaterally, it is strong and provides a stable CVJ. Incorporation of the occipital bone in the fixation procedure is seldom necessary. However, in the presence of gross and bilateral destructive disease, occipitocervical fixation can be a useful alternative.

### Long-term Outcome after Conservative Treatment

The effectiveness of antituberculous drugs usually begins ~3 weeks after commencement of treatment. Pain eases, and the systemic symptoms begin to abate. Over the same period, patients recover in terms of neurological symptoms and deficits to varying degrees. Fibrous reunion of the region and of the joint usually occurs, and normalcy of the joint movements is at least partially restored. Patients who are nonresponsive to drug

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**Case Illustration**

A 63-year-old man, not a known diabetic and not immunocompromised, had multiple large and matted cervical lymph nodes for ~5 months. The infection resulted in multiple sinuses that drained pus over the skin of the neck. Investigation confirmed that the lesions were tuberculous in nature; accordingly, antituberculous drugs were started. Following the drug treatment, some of the sinuses healed, and the cervical lymphadenitis resolved. Although the patient had pain in the neck for several months, he and the physicians related it to cervical lymph node infection. For ~1 month, the pain in the neck exacerbated, and he had progressive weakness of all four limbs. The weakness was more pronounced on the right side. Over a period of 10 days, he worsened to such an extent that he was bedridden and could not perform useful activity with any of his limbs. He had to be fed and clothed. For urinary retention, he had to be catheterized. He needed an enema for stool evacuation.

Investigations revealed extensive destruction of the facets of the atlas and axis and the occipital condyle on the right side. Additionally, there was destruction of part of the anterior and posterior arch of the atlas and the odontoid process of the axis (Fig. 37.7). Adjoining the region of bone destruction, there was granulomatous soft tissue swelling. Magnetic resonance imaging showed altered cord signal intensity at the level of the CVJ; however, no evidence of direct neural compression by bone or soft tissue was evident. Dynamic flexion-extension plain radiology did not show any clear evidence of abnormal mobility in the region and of the odontoid process in relation to the anterior arch of the atlas. Computed tomography showed evidence of lateral dislocation of the facet of the atlas over the facet of the axis on the left side. The atlantoaxial joint space had collapsed, and the occipital bone, atlas, and axis bones appeared to be crowded together. The occipitoaxial joints and the subaxial joints were preserved. It was concluded that the lateral dislocation of the facet of the atlas over the facet of the axis was a result of incompetence of the ligamentous assembly, more importantly, the alar and transverse ligaments, due to destruction of the atlas facet on the contralateral side. Considering the rapid progression of neurological worsening, surgery for stabilization of the region was contemplated. The left-sided atlantoaxial joint was exposed. The region appeared moderately unstable. The articular cartilage was denuded, and bone graft pieces harvested from the iliac crest were stuffed into the joint space. Additionally, a titanium spacer measuring 3 mm in height, 11 mm in length, and 8 mm in width was impacted in the joint space after its distraction. Lateral mass plate and screw fixation of the atlantoaxial joint was subsequently done with the technique described by us. Bone graft pieces were also placed over the midline in the region of the arch of the atlas, the lamina, and the spinous process of the axis, after appropriately preparing the bed. Postoperatively, the patient was placed in a four-poster cervical collar, and all movements related to the neck were restricted for a period of 3 months.

The patient’s clinical symptoms improved remarkably immediately following surgery. He was placed on second-line antituberculous drugs for a total of 18 months. At follow-up at 33 months, he walked independently and could carry out his routine household activities unaided.
Fig. 37.7a–g  A 63-year-old male patient presented with neck pain and progressive quadriparesis. The patient was managed successfully with unilateral fixation of the atlantoaxial joint.

a  CT scan shows basilar impression, “fixed” atlantoaxial dislocation, and evidence of destruction of the odontoid process.

b  Sagittal view of CT scan showing destruction of the lateral mass of the atlas and axis.

c  Coronal view of CT scan showing lateral dislocation of the facet of the atlas over the facet of the axis.

d  T2-weighted MRI showing evidence of cord changes at the level of the foramen magnum.

e  Postoperative CT scan shows reduction of atlantoaxial dislocation and of basilar impression.
This patient developed tuberculous affection of the craniovertebral region while he was already on antituberculous drugs. He had rapid progression of neurological worsening, despite the fact that no definite abnormal mobility was observed on dynamic radiological imaging of the CVJ. The exact mode of involvement of the cord is unclear. However, it appeared from the radiological assessment that the lateral dislocation of the contralateral facet of the atlas over the facet of the axis and collapse of the atlantoaxial joint space height and reduced height of the spine in the region probably resulted in kinking of the cord.

In the presence of destruction of the facets of the atlas and axis on one side, it appeared that the alar and transverse ligaments became unilaterally incompetent. The shift of balance on the contralateral side and the obliquity of the inclination of the facet of the atlas in the atlantoaxial joint probably resulted in its lateral dislocation over the facet of the axis. Investigations to assess mobility and reducibility of the lateral dislocation were not completed. However, during surgery, the region appeared moderately unstable. Our patient recovered remarkably following surgery that involved distraction of the facets of the atlas and axis, restoration of the height of the atlantoaxial spine, and their fixation. This suggested the incompetence and collapse of the joint and instability of the atlantoaxial region. Considering the involvement of the posterior arch of the atlas, a midline wiring operation was not possible. Although fixation was done on only one side, lateral mass C1–C2 plate and screw fixation, jamming the joint movements, and distraction of the facets provided a stable fixation and ground for bone fusion. However, the need for inclusion of the occipital bone in the fixation procedure remains questionable.

**Conclusion**

Tuberculosis of the craniovertebral junction usually has a defined pattern of tissue involvement and of extension. Appropriate understanding of the pathogenesis of the disease and of its response to drug therapy, can lead to rationale surgical therapy.

**References**

## VI

### Tumors and Approaches

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The craniovertebral junction (CVJ) is an anatomical area that extends from the foramen magnum to the atlas and the axis. This transition zone between the cranium and cervical spine includes a portion of the medulla, the cervicomedullary junction, and the upper cervical spinal cord. Multiple ligaments and joints at the CVJ ensure stability while allowing a wide range of normal movements. Structural defects, acquired and congenital mechanical dysfunction, inflammatory lesions, and tumors affect this anatomically and biomechanically complex region. Tumors of this area can be hard to diagnose and difficult to treat. Early surgical treatments were risky; however, advances in the understanding of this region have generated new interest and new treatment options for previously difficult to treat lesions. We present here an overview of the clinical features of tumors that occur at the CVJ.

## Clinical Manifestations

Tumors of the CVJ are uncommon and can be difficult to diagnose because of the variety of symptoms they can produce and because of failure to consider this region in the differential diagnosis. The relatively large diameter of the spinal canal with respect to the spinal cord at the CVJ often permits tumors to grow to a large size prior to symptom development. Clinical findings can include any combination of dysfunction of the brainstem, lower cranial nerves, upper cervical cord, and upper cervical roots (Table 38.1). These symptoms can arise from direct compression, indirect compression, mechanical instability, and vascular disruption caused by the tumor. Tumors of the CVJ can be mechanically destabilizing, resulting in basilar impression, cranial settling, and atlanto-occipital instability. Symptoms can present as an insidious progression of a variety of symptoms and false localizing signs or as a rapid onset of sensorimotor symptoms and even sudden death. The symptoms may mimic other processes; thus, incorrect diagnosis and delay of treatment can result. The most common confounding diagnoses to consider are cervical spondylosis, carpal tunnel syndrome, and multiple sclerosis.

### Table 38.1 Clinical presentation of craniovertebral junction tumors

<table>
<thead>
<tr>
<th>Brainstem symptoms</th>
<th>Apnea, ataxia, dysmetria, nystagmus</th>
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<tbody>
<tr>
<td>Cranial nerve symptoms</td>
<td>Auditory changes, dysphagia, facial weakness, tongue atrophy and weakness, trapezius weakness</td>
</tr>
<tr>
<td>Myelopathy symptoms</td>
<td>Hemiparesis, paraparesis, quadriparesis, sensory changes, urinary issues</td>
</tr>
<tr>
<td>Pain symptoms</td>
<td>Neck, base of skull</td>
</tr>
<tr>
<td>Vascular symptoms</td>
<td>Syncope, vertigo, confusion, transient visual loss</td>
</tr>
</tbody>
</table>

## Radiological Evaluation

In the past, identification of CVJ tumors was very difficult. Plain films of the skull base are difficult to interpret. Secondary changes, such as bony erosion, sclerosis, bony expansion, calcifications, and mechanical instability, can suggest an abnormality. Pre- and postoperative dynamic plain radiographs are useful in determining instability before and after surgery. We now rely on computed tomography (CT) with or without myelography and magnetic resonance imaging (MRI) of the CVJ to diagnose tumors in this region. Current techniques permit...
excellent visualization of both the bony and soft tissues of the CVJ. CT is very useful to look for bony alterations secondary to tumor progression, to examine the region around the vertebral artery, and to assess the need for reconstruction after surgery. MRI is helpful to evaluate the nature of the tumor itself and the condition of the involved neural tissues and soft tissue. MR angiography (MRA) or CT angiography (CTA) can be useful for evaluating the vascular anatomy, although these methods can sometimes be limited by artifact. Cerebral angiography is another option for evaluating large vessel patency and tumor vascularity and allows for preoperative embolization, which can be useful in certain tumors. Finally, bone scans are useful in identifying and diagnosing several spinal lesions.

Craniovertebral Junction Tumors

Tumors of the CVJ arise from neural tissue (schwannomas, neurofibromas, astrocytomas, and ependymomas), their coverings (meningiomas and arachnoid cysts), or local bone and soft tissue (chordomas, osteomas, osteoblastomas, giant cell tumors, aneurysmal bone cysts, plasmacytomas, eosinophilic granulomas, and metastases).14 (Table 38.2). The most common tumors are intradural extramedullary tumors, including meningiomas, neurofibromas, and schwannomas.1 Extrudal tumors are the second most common tumors seen in the CVJ. The most common of these are metastases and chordomas. Intradural intramedullary lesions are the least common and include primary spinal cord tumors and tumors of the posterior fossa that descend downward, such as astrocytomas, ependymomas, cerebellar hemangioblastomas, medulloblastomas, and choroid plexus papillomas. Rare incidents of teratomas, lipomas, arachnoid cysts, paragangliomas, and dermoids have been reported.

Table 38.2 Tumors of the craniovertebral junction

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<tr>
<th>Intradural Extramedullary Tumors</th>
<th>Benign</th>
<th>Meningioma</th>
<th>Neurofibroma</th>
<th>Schwannoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malignant</td>
<td></td>
<td>Malignant peripheral nerve sheath tumor</td>
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</table>

<table>
<thead>
<tr>
<th>Intradural Intramedullary Tumors</th>
<th>Benign</th>
<th>Hemangioblastoma</th>
<th>Lipoma</th>
</tr>
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<tbody>
<tr>
<td>Malignant</td>
<td></td>
<td>Astrocytoma</td>
<td>Ependymoma</td>
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<table>
<thead>
<tr>
<th>Extrudal Tumors</th>
<th>Benign</th>
<th>Eosinophilic granuloma</th>
<th>Osteochondroma</th>
<th>Osteoid osteoma</th>
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<th>Giant cell tumor</th>
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<tbody>
<tr>
<td>Malignant</td>
<td></td>
<td>Metastases</td>
<td>Chordoma</td>
<td>Chondrosarcoma</td>
<td>Ewing sarcoma</td>
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Intradural Extramedullary Tumors

Benign

Meningioma

Meningiomas are the most common benign intradural tumors of the CVJ (Fig. 38.1). One study of 133 intradural extramedullary foramen magnum tumors reported 75% meningiomas and 13% neurofibromas.15 Meningiomas are slow-growing tumors that originate from arachnoid cap cells and commonly attach to the dural covering of the brain and spinal cord.16 The World Health Organization (WHO) classifies meningiomas as grade 1 (benign), grade 2 (atypical), and grade 3 (anaplastic), which account for 80%, 5% to 20%, and 1% to 2% of all meningiomas, respectively.17,18 In large clinical series, there is a strong association between outcome and grade.19 On MRI, meningiomas classically present as an intradural, extramedullary mass that is iso- or hypointense on T1-weighted images, hyperintense on T2-weighted imaging, and uniformly contrast enhancing with gadolinium. Other characteristic findings are an enhancing dural tail and hyperostosis of adjacent bone. Meningiomas often express hormonal receptors, which may explain the increased prevalence of meningiomas in women, where the overall ratio is 2:1 in the brain and up to 10:1 in the spine.20 The best established of these receptors is the progesterone receptor, which is found in more than two thirds of these tumors. Additionally, > 30% of meningiomas also express estrogen receptors, and ~40% express androgen receptors. Moreover, patients with meningiomas have been found to have a higher incidence of breast cancer, and breast cancer patients have a higher incidence of meningiomas.21 The most frequent genetic alteration seen in meningiomas is the loss of the neurofibromatosis type 2 (NF2) gene on chromosome 22q, which encodes a tumor suppressor called merlin (also known as schwannomin). Merlin is related to molecules of the protein 4.1 superfamily that are involved with cell growth and regulation.18,20

Gross total resection is the gold standard treatment for these tumors. Surgical resection generally leads to good outcomes and low recurrence rates in the spine,22 and
gross total resection of spinal meningiomas is achieved in 82 to 97% of cases. Interestingly, it has been reported that atypical and anaplastic meningiomas are found less frequently at the cranial base and spine, and the recurrence rate of spinal meningiomas is lower when compared with their intracranial counterparts. Chemo-therapy has not been shown to be effective in the treatment of meningiomas. Targeted therapies against the hormonal receptors have been disappointing, but other adjuvant therapies such as hydroxyurea and other targeted therapies are currently under evaluation. Radiation is reserved for poor surgical candidates, subtotal resection, unresectable tumors, or high-grade and recurrent tumors. When radiation was required in the past, spinal meningiomas were treated with traditional fractionated external beam radiotherapy, as frame-based stereotactic radiosurgery devices could not treat lesions below the foramen magnum. New advances with frameless radiosurgical techniques have overcome these shortcomings, and preliminary data report good control of spinal meningiomas after CyberKnife (Accuray, Sunnyvale, California) radiosurgical ablation.

**Neurofibroma**

These tumors are the second most common intradural extramedullary tumors arising in the CVJ and account for 15% of all nerve sheath tumors. They are described as elongated, lobulated masses that are well circumscribed though not encapsulated. Macroscopically, they exhibit intraneural growth (vs. extraneural growth of schwannomas) with functioning fascicles traversing the tumor. Neurofibromas may arise sporadically in some patients, but up to 60% are found in patients with neurofibromatosis type 1 (NF1). Mutations on chromosome 17q12 are seen in patients with NF1, and this mutation is also implicated in sporadic cases. Unlike in meningiomas, men and women are affected equally. The cell of origin is under debate; some believe that the tumor is mesenchymal in origin, whereas others believe that the Schwann cell is the cell of origin. The MRI signal characteristics of neurofibromas are similar to those of meningiomas except that they do not exhibit a dural tail. Larger tumors and tumors from patients with NF1 have about a 5 to 10% chance of malignant...
transformation, which usually presents as a rapid increase in the size of the tumor and pain. Pain, neurological deficit, cosmesis, rapid growth, and concern for malignant transformation are indications for treatment. If treatment is necessary, gross total resection with the assistance of electrophysiological monitoring is the treatment of choice. However, complete resection can be difficult without sacrificing the involved nerve root, which may result in neurological deficits. Preoperative counseling is critical in managing expectations of the patients with regards to complete versus subtotal resection and postoperative neurological deficits. Fortunately, pain relief is seen in > 85% of patients after surgery, even if only subtotal resection is achieved, and the recurrence rate after resection is ~12%.11,34

### Schwannoma

As the name implies, schwannomas are of Schwann cell origin and are the most common peripheral nerve sheath tumors. They occasionally occur in the CVJ (Fig. 38.2), are usually sporadic, but can sometimes be associated with NF2. The NF2 gene on chromosome 22q12 is thought to be a tumor suppressor and is also altered in > 50% of sporadic schwannomas.17 Schwannomatosis15 and Carney complex26 are two other conditions that predispose patients to schwannomas. Similar to neurofibromas, schwannomas occur equally in men and women. MRI characteristics of these tumors are similar to meningiomas except that they lack a dural tail and more frequently exhibit cystic changes and increased peripheral enhancement. Small to medium-size schwannomas can usually be resected en bloc by circumferential dissection of the passerby fascicles. In larger tumors, intratumoral debulking is often necessary prior to complete resection.32 The single nerve fascicle that gives rise to the tumor can often be isolated; electrophysiological monitoring can be used to confirm nonconducting. Complete resection can be achieved without any postoperative neurological deficit. If a functional fascicle cannot be dissected free, a small residual should be left to avoid postoperative neurological deficit.32,33 Even after subtotal resection, recurrence usually does not occur until many years later.37 At some centers, these tumors are treated with stereotactic radiosurgery if surgery is thought to be too risky or there is recurrence after prior resection. It has been found that patients with NF2 have a higher chance of recurrence with a 39% recurrence rate at 5 years when compared with an 11% recurrence rate at 5 years in patients with sporadic schwannomas.38

### Malignant

### Malignant Peripheral Nerve Sheath Tumor

Malignant peripheral nerve sheath tumors (MPNSTs) are a rare type of soft tissue sarcoma arising from Schwann cells or pluripotent neural crest cells of peripheral nerves and account for 5 to 10% of the 6000 soft tissue sarcomas diagnosed in the United States each year.19 They are associated with NF1, and ~4 to 10% of patients with NF1 will develop these tumors.40 The CVJ as the primary site is exceedingly rare but may be involved by secondary extension. There appears to be no racial or sex propensity, and these tumors can occur at any age, but mostly commonly occur in the seventh decade in sporadic cases and in the third or fourth decade in patients with NF1.41,42 There have been case reports of MPNST occurring many years after radiation exposure. Work-up includes careful neurological exam and imaging with MRI. These tumors are often positive on positron emission tomography (PET); however, fluorodeoxyglucose (FDG)-PET is unhelpful in determining histological grade.43 Any biopsy should be planned carefully, with the biopsy track amenable to resection at the time of definitive surgery. Unfortunately, the prognosis is poor in most patients. The mainstay of treatment is wide surgical resection followed by adjuvant radiotherapy. The goal of surgery is gross total resection with clear margins of ≥ 2 cm. Surgical resection is achievable in ~20% of paraspinal tumors and 95% of extremity tumors.40–48 Adjuvant radiotherapy improves local control in high-grade sarcomas and is recommended by the Oncology Group Consensus on MPNST.49 Chemotherapy has a modest activity in high-grade sarcomas, with a meta-analysis of high-grade sarcomas showing a nonsignificant 4% overall survival benefit. Local recurrence rates can be as high as 50%, and aggressive initial therapy affords the best chance for survival.50,51 Distant metastases develop in the lung, liver, brain, lymph nodes, skin, or soft tissue.44 Five-year survival rates for sporadic cases can be as high as 50% or as low as 10% for those with NF1.

![Fig. 38.2 MRI demonstrates a CVJ schwannoma. (From Levi AD. Neurological syndromes of the craniovertebral junction. In: Dickman CA, Spetzler RF, Sonntag VK, eds. Surgery of the Craniovertebral Junction. New York: Thieme; 1998:110, Fig. 5.5. Reprinted by permission.)](image-url)
Intradural Intramedullary Tumors

Benign

Hemangioblastoma

These tumors represent 3 to 11% of intramedullary spinal cord tumors, one third of which are associated with von Hippel-Lindau disease. They consist of thin-walled blood vessels intermixed with large, pale stromal cells and occur slightly more frequently in men. Hemangioblastomas strongly enhance after administration of gadolinium and are often associated with noncontrast-enhancing cysts that are frequently larger than the solid portion of the tumor (Fig. 38.3). The cysts can be hyperintense on T2-weighted imaging because of its proteinaceous contents. Patients with von Hippel-Lindau disease often have multiple lesions, especially in the posterior fossa. These tumors are generally located subpially on the dorsal surface and are well demarcated from normal spinal cord tissue. Therefore, gross total resection is often attainable after the tumor’s arterial supply is interrupted at the time of surgery. Subtotal resection often leads to recurrence. Significant hemorrhage can be encountered if the tumor is entered prior to coagulation of the blood supply, and some experts even recommend preoperative embolization, although this may be limited if there are many feeding vessels. Stereotactic radiosurgical ablation of recurrent, subtotally resected, or unresectable hemangioblastomas has been described.

Lipoma

These tumors rarely present in the spinal cord and are extremely rare in the CVJ (Fig. 38.4). When present, most are located near the cauda equina associated with a tethered cord. Their signal characteristics on MRI are similar to that of fat: noncontrast enhancing, hyperintense on T1-weighted imaging, and hypointense on T2-weighted images. These tumors are well demarcated from normal spinal cord but are adherent to its surrounding tissue; therefore, complete excision often leads to neurological deficit. Subtotal resection leaving residual at the interface with the spinal cord is recommended and often leads to pain relief. The intraoperative laser is an effective tool used for resection of these tumors. Adjuvant therapy is not recommended.

Malignant

Astrocytoma

These infiltrating tumors, which arise from transformed astrocytes, are the most common intramedullary tumors in the pediatric population and second only to ependymoma in the adult population. Unlike their intracranial counterparts, spinal astrocytomas are generally low grade. The cervical region is the most common location (Fig. 38.5), and there is a slight male predominance. On MRI, spinal astrocytomas are iso- to slightly hypointense on T1-weighted images and hyperintense on T2-weighted images. Despite being low grade, these tumors often
contrast enhance after gadolinium administration, though less so than ependymomas. Spinal astrocytomas are infiltrative and therefore are not well defined from surrounding normal tissue. Tumor-associated cysts are common. In children, pilocytic astrocytomas displace rather than infiltrate the surrounding tissue, allowing gross total resection in many cases. Fibrillary astrocytomas, unfortunately, are infiltrative and are difficult to distinguish from normal tissue intraoperatively. Gross total resection is often not possible without severe neurological deficit. Deficits may occur even when resection is performed exclusively within the tumor because viable axons can traverse the tumor. How aggressively one should resect a low-grade astrocytoma is controversial, as some researchers report excellent prognosis after radical resection, whereas others feel that gross total resection is impossible without damaging normal tissue and that the degree of resection does not alter the prognosis. Surgeons should be careful to spend extra time determining if the tumor is infiltrative or has a clear plane. Patients with ependymomas are often subtotally resected after an intraoperative frozen section reveals low-grade glioma and the tumor is deemed infiltrative. Some authors recommend radiation for all low-grade spinal astrocytomas despite the extent of resection, whereas others recommend adjuvant radiation for subtotal resections only. There are reports of reduced relapse rate after partial resection followed by radiation, whereas other reports did not find any consistent benefit of postoperative radiotherapy. Cooper et al. recommend MRI follow-up for subtotally resected low-grade spinal astrocytomas with radiation. Chemotherapy has not been shown to be effective for low-grade spinal astrocytomas. Despite being "low grade," the 5-year survival of one series where 17 of 21 tumors were grade 1 or 2 was 57%.

In high-grade spinal astrocytomas, the prognosis is uniformly poor and is not altered by surgery. Therefore, aggressive resection at the cost of postoperative neurological deficits is not warranted. Radiation also has not shown a clear benefit, although aggressive radiotherapy to doses that cause cordotomy has been reported to increase survival. This is an option for patients with high-grade spinal astrocytomas who already have poor neurological function. Chemotherapy is being used for this group of patients, but large studies still need to be performed to better define the role of adjuvant chemotherapy.

**Ependymoma**

These tumors (Fig. 38.6) arise from the ependymal lining of the central canal and grow slowly, with an average interval of 16 months between onset of symptoms and diagnosis. Pain at the level of the tumor is the most common...
Fig. 38.6a–e  Ependymoma. (a) MRI demonstrates a minimally contrast-enhancing CVJ lesion. Intraoperative images showing (b) spinal cord enlargement, (c) midline myelotomy and exposure of the intramedullary tumor, and (d) gross total resection. (e) Postoperative MRI revealing gross total resection of the tumor. (From Lustgarten JH, McCormick PC. Intramedullary lesions of the craniovertebral junction. In: Dickman CA, Spetzler RF, Sonntag VK, eds. Surgery of the Craniovertebral Junction. New York: Thieme; 1998:265, Fig. 12.6. Reprinted by permission.)
complaint, and severe neurological deficits are uncommon at presentation. When neurological signs are present, mild sensory loss or weakness is common, but bowel and bladder changes are rare from ependymomas at the CVJ. Ependymomas are classically isointense on T1-weighted MRI imaging, hyperintense on T2-weighted images, and enhance strongly after gadolinium.58 There may be mixed signal characteristics due to cystic changes within the tumor or hemorrhage. A hyperintense cap of hemosiderin on T1-weighted imaging is suggestive of an ependymoma and is likely due to prior hemorrhage. Ependymomas are better defined and have stronger contrast enhancement when compared with astrocytomas on MRI; however, these tumors are often indistinguishable on imaging.

Ependymomas grow from the center and push adjacent spinal cord away; as a result, there is often a plane between the tumor and normal spinal cord. This allows for gross total resection in many cases, which is usually curative for these tumors. If gross total resection is achieved, radiation therapy and chemotherapy are unnecessary, as recurrence is rare.52,59 In one study of 36 patients with a mean follow-up of 59 months, 39% were neurologically improved, 47% were stable, and 14% worsened.60 If there is recurrence on repeat imaging or residual tumor on postoperative imaging, reoperation is recommended. Radiation is only considered in tumors not amenable to complete resection.61

Extradural Tumors

Benign

Eosinophilic Granuloma (Langerhans Cell Histiocytosis)

These rare tumors are locally destructive but are most often self-limiting.62 They are the result of uncontrolled growth of the reticuloendothelial system characterized by infiltration of numerous histiocytes and eosinophils.53 The classic radiological appearance is that of a symmetrically flattened vertebral body in conjunction with disk space preservation, sparing of the posterior elements, and absence of kyphosis.63 Symptoms generally present in the first 2 decades of life, and this disease is characterized by both spontaneous regression and occasional reactivation. Therefore, treatment is usually conservative. For lesions in the spine, partial intralesional curettage, chemotherapy, and low-dose radiation therapy may be recommended. Optimal treatment includes pain control and external orthosis to maintain spinal alignment during the acute phase. Systemic disease is treated with methotrexate and vinblastin.62 Aggressive surgical resection is indicated only for progressive neurological decline or spinal instability.

Osteochondroma

These tumors account for 30 to 40% of all benign bone tumors, but only 4% occur in the spine.64 Over 50% of vertebral osteochondromas, however, arise in the cervical spine, particularly C2. In the spine, they are generally isolated lesions unless part of an autosomal dominant disorder called hereditary multiple exostoses (HME).65 Osteochondromas are cartilaginous tumors that are thought to arise during development when cartilaginous tissue is trapped outside an advancing epiphyseal growth plate.63 They have a > 2:1 male predominance and usually present before the age of 30.64 Patients with HME present on average 10 years earlier than sporadic cases.65 Osteochondromas generally affect the posterior elements and can transform (rarely) into chondrosarcomas.66 Marrow and cortical continuity of the lesion with the underlying bone is pathognomonic for this lesion on thin-cut CT. On MRI, the hyaline cartilaginous cap of the tumor has intermediate signal on T1-weighted imaging and is hyperintense on T2-weighted imaging. In an adult, a cartilaginous cap > 1.5 cm suggests malignant transformation to a chondrosarcoma.65 Asymptomatic lesions can be observed. Patients who need a pathological diagnosis or who present with pain or neurological deficit should undergo gross total resection, which is curative. Recurrence is rare, although there are some reports of recurrence after subtotal resection.64

Osteoid Osteoma

These tumors are small, benign, self-limiting, bone-producing lesions that contain osteoblasts. They predominantly occur in the posterior elements, with a slight 2 to 4:1 male predominance.67 Osteoid osteomas usually present in the second decade of life with localized pain. This pain is classically nocturnal or accompanies activity and responds to aspirin or nonsteroidal anti-inflammatory drugs (NSAIDs).68 Some patients have associated scoliosis with these lesions, although in the cervical spine, patients will present with painful torticollis and reduced cervical range.67 Patients with osteoid osteomas very rarely present with neurological symptoms. On imaging, these lesions characteristically have a discrete radiolucent nidus surrounded by a variable degree of sclerosis.33 Technetium bone scan shows intense uptake within the osteoid osteoma and only one false-negative has been reported.69 These scans are particularly useful when the lesion is not seen on CT or MRI, and some investigators have reported the use of intraoperative bone scanning to ensure complete resection.70 Medical treatment with aspirin and NSAIDs is effective at relieving pain, especially nocturnal pain. However, if the pain becomes medication resistant, complete surgical excision by intralesional curettage is curative.33,62,67 Localizing the lesion intraoperatively can be challenging, as osteoid osteoma looks like normal bone. Continued pain is indicative of residual tumor and requires further investigation. Recurrence after complete resection is rare.

Osteoblastoma

These tumors are similar to osteoid osteomas, except that they tend to be larger, more aggressive, and can undergo malignant transformation (Fig. 38.7). They also generally...
Osteoblastomas present in young patients, with the majority being diagnosed before the age of 30. They often present with pain, but unlike osteoid osteomas, this pain is not often relieved by aspirin or NSAIDs. A large percent of these patients also present with neurological deficit secondary to soft tissue extension, neuroforaminal encroachment, and epidural compression by the osteoblastoma. Radiographically, these lesions can have a radiolucent nidus with a sclerotic rim like osteoid osteomas, but they are more heterogeneous in appearance and appear as expansile lesions with or without calcifications. Bone scan remains the most sensitive test, and these lesions demonstrate intense uptake. Surgical resection is the mainstay of treatment; however, despite gross total resection, recurrence rates approach 10%. Chemotherapy and radiation have been used in recurrent or unresectable tumors.

### Aneurysmal Bone Cyst

These lesions are described as expanding lesions with blood-filled cavities separated by septa of trabecular bone or fibrous tissue containing osteoclast giant cells and comprise approximately 15% of all primary spine tumors. They are generally present in patients younger than 20 years but have been reported in patients from 2 to 67 years. Aneurysmal bone cysts are classified according to Enneking as type 1 (latent), type 2 (active), and type 3 (aggressive) lesions. They are also categorized as primary versus secondary, which are associated with other lesions. About one third of aneurysmal bone cysts are secondary and most commonly are associated with giant cell tumors, hemangiomas, osteoblastomas, chondroblastomas, and telangiectatic osteosarcomas. A solid variant that lacks the cavernous channels and spaces also exists and accounts for 3.5 to 7.4% of cases. The underlying pathogenesis of the tumor is unclear. Some believe that they are reactive lesions produced through a vascular disturbance that leads to increased venous and osseous pressure causing local distention of bone, whereas others believe that they are a result of aberrant bone repair after a traumatic insult. There are also reports that these lesions have an underlying genetic cause. Characteristic CT findings of this tumor include involvement of the posterior elements with associated vertebral body involvement with a “blown out” or “ballooned” expansile lytic lesion. The tumor will appear as a soft tissue mass surrounded by a “shell” of cortical bone. Fluid-filled levels are highly sensitive and specific.

The natural history of aneurysmal bone cysts is not well defined. There have been no deaths noted in large series, and spontaneous remission occasionally occurs, although surgical curettage without en bloc resection often leads to recurrence. Treatment choices include arterial embolization, percutaneous intralesional injection of...
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1. Open biopsy of the lesion when the diagnosis is uncertain after imaging with curettage at the time of surgery if the diagnosis in confirmed

2. Selective arterial embolization if the diagnosis is certain, there is no neurological compromise, there is no instability, and embolization is safe

3. Complete curettage, preferably preceded by embolization, if pathological fracture is a concern or if there is an anterior lesion with neurological compromise. Curettage is also recommended during recurrence or if embolization is not possible.

4. En bloc excisions for posterior element aneurysmal bone cysts

5. Bracing in children until skeletal maturity unless reconstruction is required at the time of surgery

Giant Cell Tumor

These tumors comprise < 5% of primary bone tumors and are osteolytic lesions that occur primarily in women in the third and fourth decades. Metastases have been reported, and rarely the tumor is multifocal. These lesions are iso- or hypointense to the spinal cord on MRI and enhance homogeneously after gadolinium administration. The tumor usually involves the vertebral body and may extend into the posterior elements with an expansile, lytic, nonsclerotic appearance (Fig. 38.9). CT is useful in evaluating the bony involvement, especially if surgery and stability are issues. A CT-guided core biopsy is recommended, as a needle biopsy may lead to incorrect diagnoses because of sampling error. Once the diagnosis is confirmed, preoperative tumor embolization followed by en bloc excision with wide margins is the optimal treatment, as these tumors have a propensity to recur. Because these lesions are often large, and complete resection at the CVJ is technically challenging, subtotal resection is often accompanied by tumor embolization and radiation therapy. However, despite combined embolization, subtotal resection, and radiation therapy, local recurrence...
rates remain high.\textsuperscript{52,63} Stereotactic radiosurgery is still being investigated as primary and adjuvant therapy for these locally aggressive benign tumors and may improve local control in these patients.\textsuperscript{60}

### Hemangioma

These are usually benign vascular tumors and are found in 10 to 12% of spines on autopsy.\textsuperscript{66} In most cases, they are asymptomatic and can be followed clinically; however, a small percentage may cause pain or even neurological deficit from bone expansion or pathological fracture. There are also reports of pregnancy-exacerbated cases, thought secondary to hormonal or hemodynamic influences.\textsuperscript{83} On imaging, these tumors are bright on T1-weighted MRI secondary to a high concentration of adipose tissue and bright on T2-weighted imaging secondary to slow flow through the vascular components of the tumor.\textsuperscript{35} CT demonstrates classic “honeycombing,” which represents vertically oriented vertebral lucenties separated by thickened trabecular bone.\textsuperscript{84} Treatment of symptomatic lesions can include radiation therapy, surgery, arterial embolization, percutaneous vertebroplasty, and transarterial particulate embolization or ethanol injection. Acosta et al. published recommended guidelines for treatment of these spinal tumors:\textsuperscript{84}

1. Asymptomatic lesions are treated with observation alone.
2. Symptomatic tumors with extraosseous tumor extension should be treated with transarterial embolization and aggressive resection.
3. Painful lesions that are limited to the vertebral body and/or posterior elements can be treated with percutaneous vertebroplasty or transarticular embolization. Recurrent pain can be treated with radiation therapy or aggressive resection.
4. Lesions limited to the vertebral body and/or posterior elements that cause neurological deficit should be treated with transarticular embolization and decompressive laminectomy. Patients with recurrent neurological deficit and/or pain after laminectomy can be treated with aggressive resection. Patients with residual or recurrent extravebral tumor without neurological deficit can be treated with radiation.

### Malignant

#### Metastases

Breast, lung, and prostate cancers are the most common primary tumors that metastasize to the spine, although hematological malignancies, such as multiple myeloma and Hodgkin disease, metastasize to the spine as well.\textsuperscript{85} The frequency of which spinal segment is affected is related to vertebral body number and volume; therefore, 70% of metastases to the spine involve the thoracic spine, 20% involve the lumbar spine, and only 10% involve the cervical spine. Most spine metastases are extradural; however, ~5% are intradural extramedullary and are considered to have leptomeningeal spread, and an even smaller percentage are intramedullary. Pain is the most common presenting symptom of metastases to the spine and often precedes neurological deficits by weeks to months.

Treatment modalities for metastatic tumors involving the spine include a combination of surgery, radiation, and chemotherapy and require a team of medical and radiation oncologists, as well as surgical oncologists. For highly vascular lesions such as metastatic renal cell carcinoma, preoperative embolization can be very helpful (Fig. 38.10). Pain medications, steroids, and bisphosphonates are a common part of treatment regimens for these patients. Early diagnosis and management of these metastases are important for reducing pain, preserving and improving neurological function, and improving quality of life. However, many factors, including the extent of disease, life expectancy, medical condition of the patient, sensitivity of the specific tumor to chemotherapy and radiation, extent of spinal cord or epidural compression, and patient’s signs and symptoms, need to be considered prior to choosing a treatment strategy.\textsuperscript{86} In the past, radiation was the first-line treatment, as laminectomy or laminectomy and radiation yielded poor results. More recently, Patchell et al. published results from a randomized trial showing improved outcome and survival rates after direct decompressive surgery plus postoperative radiotherapy when compared with radiotherapy alone for a certain group of patients with spinal cord compression caused by metastatic cancer.\textsuperscript{87} However, extensive resection/reconstruction is difficult in the CVJ. Moreover, patients who have extensive disease, multiple comorbidities, tumors that are particularly radiosensitive, or those who have been paralyzed for a prolonged period of time should not undergo surgery. Advances in radiation technology have also improved the treatment of metastases to the spine. Intraoperative radiation therapy, three-dimensional conformal radiotherapy, intensity modulated radiation therapy, and CyberKnife radiosurgery\textsuperscript{88} can increase the amount of radiation delivered to the tumor while limiting damage to the surrounding tissues.

### Chordoma

These are some of the most frequent extradural tumors seen in the CVJ (Fig. 38.11). They generally occur in adults, with a peak incidence in the fifth or sixth decade, and display a male/female ratio of 2:1 in the spine and 1:1 intracranially.\textsuperscript{89,90} Chordomas arise from notochordal elements within the vertebral body and can arise anywhere along the spinal axis, with a predilection for the clivus and sacrococcygeal regions.\textsuperscript{91} In the CVJ, they frequently present with sixth, seventh, or eighth nerve palsy, headaches, or signs of spinal cord and nerve root compression.\textsuperscript{89} CT
Fig. 38.10a–e  Metastatic renal cell carcinoma. (a) Axial and (b) coronal CT images demonstrate a lesion at C2. (c) Right vertebral artery and (d) right costocervical and thyrocervical trunk angiogram injections reveal vascularity. (e) Subclavian artery injection after glue embolization of the thyrocervical and costocervical trunks demonstrates no residual arterial supply to the tumor. (From Deshmukh VR, Fiorella D, Albuquerque FC, McDougall CG. Embolization techniques for neoplasms of the spine and spinal cord. In: Dickman CA, Fehlings MG, Gokaslan ZL, eds. Spinal Cord and Spinal Column Tumors. New York: Thieme; 2005:211, Fig. 9.3. Reprinted by permission.)

Fig. 38.11  Sagittal MRI demonstrates a chordoma. (From Obasi C, Johnson PJ, Hahn MS, Canalis RF. Tumors of the occipitocervical junction. In: Fessler RG, Sekhar L, eds. Atlas of Neurosurgical Techniques. New York: Thieme; 2006:28, Fig. 3.5. Reprinted by permission.)
Chondrosarcomas are similar to chordomas in their destructive nature, ability to invade nearby structures, and occasional incidence of metastases. However, they have a better prognosis when compared with chordomas and often are paramedian in location except when associated

with Ollier disease or Maffucci syndrome. Five-year survival rates correlate with the histological grade of the tumor. Grade I, II, and III lesions had, respectively, 90%, 81%, and 43% 5-year survival rates in one series.100 Overall, the 5-year survival rate is 90 to 99%, and the 10-year survival rate is 71 to 99%.99,105–107 Like chordomas, surgical series advocate radical resection when feasible. Postoperative proton irradiation and photon-based stereotactic radiosurgery are the most commonly used adjunctive treatments for recurrences, residual tumors, unresectable tumors, or tumors in patients who are not surgical candidates, as these tumors are resistant to standard external beam radiotherapy. In the CVJ, radical resection is often impossible, and tumors in this region require a multidisciplinary approach.

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**Ewing Sarcoma**

This is the most common primary malignant tumor of the spine in children.108 The male-to-female ratio is 3:2, and patients also present with low-grade fever, an elevated erythrocyte sedimentation rate, anemia, and leukocytosis.63 CVJ cases, however, are rare, and 20% of these patients have metastases on presentation, although this does not affect the overall survival.109 Bone scans reveal increased uptake, and MRI reveals a bony mass with a prominent soft tissue component that is intermediate signal on T1-weighted images and hyperintense on T2-weighted imaging.110 This tumor is sensitive to both radiation therapy and chemotherapy; therefore, aggressive resection is indicated only in patients with progressive neurological compromise or structural instability. Unfortunately, Ewing sarcoma is malignant, and 5-year survival rates range from 33 to 48% despite aggressive care.108,109

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**Osteogenic Sarcoma**

These lesions arise from pre-existing bony lesions such as Paget disease or osteoblastomas, or in previously irradiated bone. Spinal column lesions are more commonly secondary from another skeletal site.66 Radiographs often reveal an “ivory vertebra,” which is a result of the dense mineralization of these tumors. CT demonstrates foci of mineralized matrix throughout the lesion, and MRI reveals a lesion that is hypointense on T1-weighted images, hyperintense on T2-weighted images, and contrast enhancing after gadolinium administration. Pulmonary metastases are common. These patients should be treated with a combination of wide surgical resection, chemotherapy, and radiation therapy. Despite maximal therapy, median overall survival remains 23 months.111

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**Surgical Considerations**

Should surgery be warranted for the treatment of a CVJ tumor, careful planning should be done prior to surgery.
whether it is for biopsy, decompression, resection, or stabilization. The relationship of the tumor to neighboring structures will determine its resectability. Complete resection of the tumor is generally the goal of surgery; however, one must often attempt maximal resection while minimizing morbidity. In addition, extensive resection can result in instability; hence, instrumentation should be planned for in advance. Fortunately, advances in radiation therapy and chemotherapy allow subtotal resection. For instance, stereotactic radiosurgery can be applied to residual tumors. In fact, resecting tumor away from normal neural structures can significantly improve the safety and efficacy of stereotactic radiosurgery by increasing deliverable doses while staying within dose limits for critical structures.

There are five general directions of surgical approach to the CVJ: anterior, anterolateral, lateral, posterolateral, and posterior. The direct anterior approaches include the transoral approach and its variations. The anterolateral approaches include the mandibular swing transcervical and extrapharyngeal transcervical. The lateral transcervical is a direct lateral approach. Posterolateral approaches include the far/extreme lateral transcondylar and the posterolateral. Finally, the midline posterior approach can be used as a direct posterior approach. Anterior approaches are selected for extradural lesions situated at the anterior foramen magnum. They can be used to reach tumors of the inferior clivus and upper cervical vertebra. The transoral approaches are the most direct; however, their risk of meningitis, their limited lateral exposure, and their lack of vascular control limit their use to small midline extradural lesions. The extended transoral approaches can expand the surgical field of the standard transoral approach to the middle clivus or down to the low upper cervical vertebra. These are technically much more difficult and are associated with higher morbidity. All transoral approaches also carry a high likelihood of instability. In some cases, transcranial and transsphenoidal approaches may be better choices.

The anterolateral approaches may be useful for lesions that are anterior and in the midline but placed laterally, thus preventing the use of a transoral approach. The anterior extrapharyngeal approach also does not involve the oropharynx. Because they approach from a lateral angle, they can be used to treat anterior intradural lesions. The mandibular swing approach is technically challenging but provides a wide exposure. For lesions that involve the vertebral artery or are primarily located laterally, the lateral transcervical approach may be ideal. A drawback is that their approach to the foramen magnum requires extensive muscle dissection and detailed knowledge of the anatomy. In some cases of intradural anterior brainstem lesions, transcervical or posterolateral approaches may be more appropriate.

Most intradural tumors can be resected by posterior approaches. For lesions that affect the posterior and lateral CVJ, the midline posterior or posterolateral approach is commonly selected. These posterior approaches are preferred for most posterior to lateral intradural lesions. The posterolateral approach can treat some anterior intra- and extradural CVJ tumors. Extreme lateral and far lateral suboccipital approaches are used to approach more ventral and ventrolateral foramen magnum lesions with minimal retraction. If exposure around the brainstem is needed to view the distal vertebral artery and inferior clivus, they are ideal. These approaches are easily combined with craniotomies to expand the operative field.

### Conclusion

Tumors of the CVJ are both diagnostic and treatment challenges for the surgeon. Modern advances in tumor management and diagnostic imaging have brought a new interest in treating these lesions. As such, current surgical approaches can approach lesions at virtually any location. However, the selection of the treatment modality depends often on the lesion itself. A variety of lesions present in and around the CVJ, often with similar findings. Depending on their nature, surgery, chemotherapy, radiation, or a combination of the three is the best course of action. Surgery usually has an important role in not only treating the lesion but also diagnosing the lesion, resecting the lesion partially for further radiation, decompressing the neural elements, and stabilizing the CVJ. With the appropriate understanding of the diagnosis and prognosis of these lesions, coupled with a careful selection of approach and increased technical familiarity through practice, surgeons can successfully and safely treat some of these most difficult tumors.

### References

8. Symonds C, Meadows P. Compression of the spinal cord in the neighborhood of the foramen magnum. Brain 1937; 6:52–84
Peripheral nerve sheath tumors arising from the second cervical nerve root are relatively common and constitute \( \sim 15\% \) of all spinal peripheral nerve sheath tumors. In our series, out of the total spinal peripheral nerve sheath tumors seen during the period of 1992 to 2006, 24\% arose from the second cervical root.\(^1\) There are several isolated reports and some relatively large series of cases with C2 peripheral nerve sheath tumors published in the literature.\(^1\)–\(^5\) Spinal peripheral nerve sheath tumors are seen sporadically in the general population, but they are frequently associated with neurofibromatosis types 1 and 2 (NF1 and NF2).\(^5,6\) An intraspinal location is the most frequent form, whereas an extraspinal component accounts for 15\% of all cervical peripheral nerve sheath tumors. Some authors have noted a predominance of “extradural” location of peripheral nerve sheath tumors at the C2 vertebral level.

Despite their critical location, with extension of the tumor anterior or anterolateral to the cord and proximity to vital neural structures and the vertebral artery, surgery on C2 peripheral nerve sheath tumors constitutes one of the most gratifying experiences. If the anatomy of the tumor and its relationship to normal structures in the vicinity is appropriately understood, the surgery on these formidable-looking tumors is relatively straightforward.\(^1,7\)–\(^9\) The majority of peripheral nerve sheath tumors located at the level of C2 probably arise from the large C2 ganglion and are limited within the dural confines or are “interdural” in nature. In contrast to other spinal peripheral nerve sheath tumors, the location of C2–peripheral nerve sheath tumors is, in most cases, posterior to the lateral mass of the atlas and axis and the atlantoaxial joint and is exposed to the posterior without any bony cover (Figs. 39.1 and 39.2). Radical tumor resection is safe, resolution of clinical symptoms is rapid, and recurrence rates are extremely low. In select cases, bone work for tumor exposure and resection can be entirely avoided.

Multiplicity of peripheral nerve sheath tumors is frequently seen in patients with von Recklinghausen disease. George and Lot reported 4\% multiplicity of peripheral nerve sheath tumors at any spinal level.\(^4\) The dural anatomy of tumors associated with von Recklinghausen disease is not different from other C2 peripheral nerve sheath tumors, but the tumors in general are more firm in consistency and well defined in nature.

Fig. 39.1a, b
a Cadaveric dissection specimen of the C1–C2 region, showing the C2 ganglion and its relationships with the C1–C2 joint, vertebral artery, and dural tube. The vertebral artery on the side of ganglion is rudimentary in nature. The ganglion on the right side has been sectioned off to show the joint.

b Drawing showing the C2 ganglion and its relationships.
Clinical Features

We evaluated our series of 60 patients with C2 peripheral nerve sheath tumors. Symptoms and signs and radiological characteristics were usually diagnostic in nature. However, unusual symptoms of syncopal attacks, migrainous headaches, and unrelated motor and sensory symptoms can lead to misdiagnosis. The mean age of the patients was 28 years (range 6–62 years). There were 38 men and 22 women. The mean duration of symptoms at the time of presentation was 27 months (range 4 days–5 years). Mild to moderate myelopathy was present in 52 cases. None of the patients presented with severe myelopathy. All patients were able to walk unaided at the time of admission. The rest of the patients had either local symptoms, such as neck pain, or symptoms essentially unrelated to C2 peripheral nerve sheath tumor. In two of our patients with von Recklinghausen disease, the bilateral C2 peripheral nerve sheath tumors were large and had caused severe cord compression, but the presenting symptoms seemed unrelated to the C2 peripheral nerve sheath tumors and were related instead to other tumors in the spine or the brain.

Anatomical Features of C2 Peripheral Nerve Sheath Tumors

C2 is the largest ganglion of all the spinal roots. Its location is unique, as it is found outside the spinal canal exposed over the lateral mass of the axis and under the lateral aspect of the posterior arch of the atlas. Tumors located there are slow growing and usually acquire a large size before becoming symptomatic. Characteristic dumbbell-shaped C2 peripheral nerve sheath tumors having both intraspinal and paraspinous components have been reported. Dumbbell tumors located elsewhere in the spine are generally classified as intraspinal, foraminal, and extraforaminal. Depending on the anatomical location as identified during surgery, peripheral nerve sheath tumors of the C2 nerve are divided into three groups (Fig. 39.2). Those located within or extending into the spinal dural tube are called type A, those located within the dural tube of the C2 ganglion are labeled as type B, and tumors extending lateral into the paraspinous region are labeled as type C. Type C tumors are those that extend outside the limits of the spine and into the paraspinous spaces. The line of demarcation between type B and type C is set by an arbitrary vertical line drawn between the tips of the transverse processes of the atlas and axis. In the classification used in this series, type A was intradural, and type B and type C were interdural. Although it is possible to correlate the described classification with preoperative investigation in most cases, in some cases it may be impossible to decipher if the peripheral nerve sheath tumor has an intradural extension or not. Of the 60 patients with 64 C2 peripheral nerve sheath tumors, there were 5 pure type A tumors, 7 type A + B tumors, 31 type B tumors, and 21 type B + C tumors (Figs. 39.3, 39.4, 39.5, 39.6, 39.7, and 39.8). The tumors varied in size, ranging from 10 to 76 mm (average 31 mm). Sixteen tumors were >25 mm in its maximum diameter. Considering the location and extension, it appeared that the majority of peripheral nerve sheath tumors probably arise from the C2 ganglion and extend either toward the spinal dural tube or away from it into the paraspinous region. In the majority of cases, it was observed that...
although the tumors had a large intraspinal extension, they were within the interdural compartment and were not extending within the spinal dural tube. Although type B and C tumors were located outside the spinal dural tube, they had a dural or perineural sheath cover that was an extension of the spinal dura. This dural sheath was well defined and formed a firm limiting membrane that separated the tumor from the adjoining structures of the region. The dura was not transgressed by these tumors, and the vertebral artery and adjoining venous plexuses were displaced by the tumor mass. Although there was no pure type C tumor, inclusion of this group into the classification scheme assisted in categorization of the tumor regarding its physical size and nature and indicated the extent of surgical complexities. Infiltration of paraspinal muscles is rare and suggestive of malignant change and was not encountered.

Similarities of C2 Peripheral Nerve Sheath Tumors to Trigeminal Neurinomas

The gasserian ganglion is the largest and the C2 is the second-largest ganglion in the body (Fig. 39.9). Both the gasserian ganglion and the C2 ganglion are related closely to a major artery (the carotid artery in the case of the gasserian ganglion and the vertebral artery in the case of the C2 ganglion). Both are associated with large venous plexuses.
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The cavernous sinus in the case of the gasserian ganglion and the large paravertebral venous plexus in the case of the C2 ganglion). The dural and arachnoid relationships of the gasserian ganglion and the C2 ganglion have remarkable similarities. Moreover, the meningeal relationships of the trigeminal neurinoma are considerably similar to the meningeal relationship with C2 peripheral nerve sheath tumors. In our study on trigeminal neurinomas, we observed that these tumors were interdural in anatomical confines in relationship to the cavernous sinus. Transgressing of the dura and encasement of the nerves, carotid artery, and venous plexus of the cavernous sinus were not encountered.¹⁰⁻¹²

There are four classically described locations of trigeminal neurinomas. The tumor can be in the middle fossa (Type A), posterior fossa (Type B), dumbell-shaped (Type A+B), and those that have extracranial extension (Type C).¹² C2 peripheral nerve sheath tumors have similar extensions and meningeal relationships. We classified C2 peripheral nerve sheath tumors into types A, B, and C according to their extensions and dural relationships. Such anatomical features have considerable bearing on surgery of trigeminal neurinomas and C2 peripheral nerve sheath tumors. As in trigeminal neurinomas, it appears that working within the dural confines and respecting the dural cover of C2 peripheral

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**Fig. 39.6a, b**

*Coronal MRI showing a type B + C tumor.*

*Fig. 39.6a, b*  
*a* Coronal MRI showing a type B + C tumor.  
*b* Sagittal T2-weighted MRI showing the tumor.

*Fig. 39.7a, b*  
*a* T1-weighted MRI showing a type B + C tumor.  
*b* Contrast-enhanced scan showing the tumor.
nerve sheath tumors can lead to safe surgical tumor resection. Understanding the meningeal relationships will reduce the extent of exposure that would be necessary to resect these tumors and avoid the need for exposure and control of the vertebral artery during surgery. Peripheral nerve sheath tumors are slowly expanding in nature, and they remodel the confines of the surrounding structures as they grow. The tumors displace the soft tissues and never encase them. C2 peripheral nerve sheath tumors are located lateral, anterolateral, or anterior to the spinal cord.

Radiological Features

The pedicle of C2, the facets of C1 and C2, and the adjoining posterior arch of the atlas and lamina of C2 are uniformly eroded. The adjoining tissues such as the venous sinuses in the region and the vertebral artery are displaced and are never encased by the tumor bulk. Magnetic resonance imaging (MRI) was the principal investigation and was obtained in all of our patients both before and after the surgery. The tumors were predominantly isointense on T1-weighted images and hyperintense on T2-weighted sequences. The lesions, in general, had smooth contours and a homogeneous appearance, enhanced uniformly, and were well delineated on contrast administration. None of the tumors had any radiological evidence of hypervascularity. In none of the cases was the vertebral artery seen to be directly encased by the tumor mass (Figs. 39.3, 39.4, 39.5, 39.6, 39.7, and 39.8).

Surgical Approach

Several posterior, posterolateral, lateral, and anterolateral approaches have been described. From our experience in managing these cases, we conclude that the standard
midline posterior approach is suitable and most appropriate to resect almost all types of C2 peripheral nerve sheath tumors.

**Standard Midline Posterior Approach**

The neck is maintained in a slightly flexed position. A midline vertical incision is extended from the occipital protuberance up to the C4 spinous process. The C2 spinous process is widely exposed on the side of the tumor, and the paraspinal muscles are dissected subperiosteally to expose the C2 lamina and the pedicle and the lateral aspect of the arch of the atlas. At least part of the posterior surface of large type B tumors could be exposed without any bone resection in the region of the gutter over the lateral masses of the atlas and axis and the atlantoaxial joint. In most cases, C2 hemilaminectomy or resection of the superior quadrant of the ipsilateral half of the lamina was done and extended laterally to expose the tumor bulk and the spinal dural tube. Whenever required, additional bone resection that included the lateral aspect of the posterior arch of the atlas was performed. In several cases, the tumor could be identified and resected over the lateral aspect of the lamina and over the pedicle, without any bone removal. Once the posterior surface of the tumor was exposed, its posterior dural wall was incised, and intratumor debulking was performed. Most of the tumors were firm; some were

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**Fig. 39.9a–f**

- **a** Drawing showing the gasserian ganglion and its relationships (red, carotid artery; yellow, gasserian ganglion; blue, signifies CSF in Meckel’s dural cave).
- **b** Drawing showing the dural relationships of trigeminal neurinomas. Note the similarity of the tumor–meningeal relationship of this tumor with the C2 peripheral nerve sheath tumor (red, carotid artery; yellow, gasserian ganglion; green, trigeminal neurinoma).
- **c** Trigeminal neurinoma having a larger middle fossa and a small posterior cranial fossa extension. Note the similarity of this tumor to a type A + B tumor.
- **d** T1-weighted MRI showing a dumbbell-shaped trigeminal tumor. Note the similarity of this tumor to a type A + B tumor.
friable and were only moderately or minimally vascular in nature. The dura around the tumor mass formed a thick barrier that separated it from the large venous plexuses of the region and the laterally or anterolaterally displaced vertebral artery. The tumor extension into the paraspinal space is followed within the dural sheath. Preservation of the C2 nerve root bers was not pursued in any case. The facets of the atlas and axis and the atlantoaxial joint were eroded by the tumor, but the integrity and function of the joint were not compromised in any case.

We have used a similar posterior midline approach for resection of anterior or anterolaterally located foramen magnum meningiomas. Surgery for meningiomas of the region is significantly different from surgery on peripheral nerve sheath tumors, as the meningeal planes are much better defined in the latter. None of the tumors were highly vascular. Most of the tumors were firm and moderately vascular. The intradural part of the tumor was relatively simple to resect, as it had a well-defined arachnoid plane of dissection. Exposure of the posterior surface of the tumor and debulking within the dural confines led to safe tumor resection. The exposure is standard and quick, and there is no need for manipulation of any cranial nerves, blood vessels, or joint to affect exposure. It is sometimes difficult to resect type C peripheral nerve sheath tumors, as the lateral exposure from the midline can be incomplete. However, in none of the type A and B cases was the exposure inadequate, and radical resection of the tumor was safely possible.

Some authors have suggested proximal control of the vertebral artery prior to tumor resection. However, in none of our cases was proximal control of the vertebral artery obtained. There was no instance of vertebral artery laceration. From our experience, we can safely conclude that there is low risk of vertebral artery injury, and preoperative angiography or intraoperative proximal or distal control does not seem to be necessary.

The articular pillar of the atlas and axis were eroded, but the joint appeared functional and intact. Because the facet joints were anterior to the tumor from the posterior surgical approach, they were not exposed, manipulated, or resected. Immediate or delayed spinal instability was not encountered in any case. However, if there is any concern regarding postoperative spinal instability, lateral mass screw and plate fixation as described by us may be performed directly after tumor resection.

Successful tumor resection results in rapid and sustained neurological recovery. None of our patients worsened in neurological function after surgery, despite the fact that many of the operations were performed by junior consultants in our academic neurosurgical department. Radical and complete tumor resection is advocated by several authors to achieve cure from the disease. The significant chance of early tumor recurrence in a partially excised peripheral nerve sheath tumor is known, and every attempt should be made to remove them completely. In seven cases with large type C tumors, a partial resection was performed. However, none of the tumors has recurred or regrown. Except for patients with NF2 who were disabled by other tumors, the rest of the patients resumed their normal lifestyle. There has been no case with symptomatic tumor recurrence. On the basis of our experience, it appears that recurrence rates even after partial resection of these tumors are extremely low.

Conclusion

C2 peripheral nerve sheath tumors probably arise from the nerve sheath in the region of the C2 ganglion. They
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have a well-defined anatomical extension, and the majority are located outside the spinal dural tube and exposed over the posterior surface of the lateral masses of the atlas and axis. The surgical exposure can be minimized, and excessive bone work and soft tissue manipulation can be avoided if the characteristic clinical and radiological features of C2 peripheral nerve sheath tumors are appropriately understood on the basis of preoperative studies. In many patients, bone work for tumor exposure and resection can be entirely avoided. The standard midline posterior approach with unilateral dissection on the side of the lesion is safe and gives satisfactory exposure to resect the majority of peripheral nerve sheath tumors at this level. The vertebral artery is displaced over the surface of the tumor and can be avoided if the dural dissection plane is maintained during surgery.

References

Meningiomas in the region of the foramen magnum are relatively rare. Similar to meningiomas in other locations, meningiomas of the foramen magnum demonstrate a female predilection, with an estimated female-to-male ratio ranging from 2:1 to 3.6:1. A large proportion of foramen magnum meningiomas are located anterior to the brainstem in close relation to vital neural, vascular, and bone structures. Meningiomas with en plaque extension and extradural growth are rare. The clinical course is slowly progressive, leading to dysesthesia, asymmetrical motor weakness, gait ataxia, and, relatively less common, lower cranial nerve section. Misdiagnosis that results from uncommon symptoms, leading to wrong decisions and inappropriate treatment, has been observed frequently with these lesions. Magnetic resonance imaging (MRI) has provided a significant advancement in diagnosis. It clearly delineates the exact tumor size, location, site of dural attachment, and relation to vascular and neural structures; MRI also provides an opportunity to assess the consistency and vascularity of the tumor. Recovery after a successful tumor resection is almost instantaneous, and recurrence rates have been demonstrated as extremely low. However, injury to the brainstem, cranial nerves, or vertebral artery or its branches can lead to a disastrous outcome for the patient and the family.  

Surgery on meningiomas located in the region of the foramen magnum anterior or anterolateral to the brainstem constitutes a formidable challenge and has been studied by various authors for many years. Anterior transoral, anterolateral transcervical, extreme lateral, and several forms of lateral approaches have been advocated and preferred over the conventional posterior approach. Although lateral or anterior approaches to these lesions have certain obvious advantages, a posterior approach as used by various surgeons during the past century also has a distinct set of advantages that should be evaluated and considered. As discussed in this chapter, the approach that uses a midline skin incision is not outdated and continues to find favor with some authors. 

The meningiomas located in the region “anterior” to the brainstem, in the region of the foramen magnum, are only rarely strictly anterior. Despite their varied sizes, all the tumors in our series pointed laterally on one side of the midline, and the brainstem was displaced or twisted posterolaterally rather than posteriorly. According to the definition suggested by George et al., most of our cases would be categorized under the subgroup intermediate between anterior and lateral meningiomas. All of the cases were demonstrated to have an anterolateral rather than a strict midline location.

**Classification**

Foramen magnum meningiomas can be classified as follows:

1. Anterior/anterolateral
2. Lateral
3. Posterior

Depending on their vertical extension, the tumors are classified into:

1. Superior: more than half of the mass is above the level of the foramen magnum.
2. Inferior: more than half of the mass is below the level of the foramen magnum.

The meningiomas having lateral and posteriorly based dural attachment usually lend themselves to relatively straightforward surgical procedures. Posterior midline approaches are uniformly accepted as sufficient for such cases.

Meningiomas having an attachment to the anterior or anterolateral aspect of the dura of the foramen magnum are relatively complex due to the intimate relationship with the cervicomedullary cord, vertebral artery, and lower cranial nerves.

**Operative Strategy**

**Position**

Most patients in our series were operated on in a semi-sitting position, with the head in mild flexion. The position assisted in having the shoulders out of the way of the surgical approach and provided a clear surgical view to the region. It also allowed for a relatively bloodless field, with the blood washing out by gravity during surgery. Apart from anesthesia-related precautions, all the safety norms necessary while operating in a sitting position are mandatory. There are large venous lakes in the extradural
space around the foramen magnum, and generous packing of the region with Surgicel or Gelfoam is mandatory to avoid air embolism-related problems.

**Incision, Surgical Exposure, and Steps**

A straight midline incision is taken. The C2 spinous process forms the inferior landmark for exposure. The suboccipital bone, arch of the atlas, and C2 laminae are exposed in a subperiosteal plane. The exposure is widened on the side of the tumor using a self-retaining retractor. Such an approach allows wide lateral exposure up to the mastoid bone and to the condyle. Whenever necessary, the length of the incision can be increased to affect the lateral exposure. The incision and the subperiosteal bone exposure avoid the need for any muscle dissection or cutting into the muscles. The arch of the atlas is exposed widely. The subperiosteal dissection is done on the inferior aspect of the posterior surface of the arch of the atlas, and the exposure is extended laterally. The vertebral artery coursing over the posterior arch of the atlas is identified for proximal control. Large venous lakes inferior to the lateral aspect of the arch of the atlas in the region of the C2 ganglion may need packing with Surgicel. The bone exposure will depend on the nature and the extensions of the tumor. Usually a low suboccipital craniectomy on the side of the tumor that extends inferiorly up to the foramen magnum and laterally to the occipital condyle is done. Whenever indicated, condylar drilling is possible for a more lateral and anterior exposure. The arch of the atlas is removed until the groove for the vertebral artery. Vertebral artery exposure and control are obtained in cases where the tumor encases the intracranial vertebral artery. The C2 spinous process and the ipsilateral half of the lamina are resected depending on the inferior extent of the tumor. The dural incision extends from the spinal dura to the cerebellar dura. The dural flap is everted laterally. After the dura is opened, the tumor, cranial nerves, and presumed site of the vertebral artery course are identified (Fig. 40.1). The cerebellum is retracted superiorly to expose the tumor from a superior aspect. The cerebellar retraction is eased by arachnoidal sectioning that releases the vermis and tonsils from the medulla. On initial inspection, the tumor may appear formidable, with intimate relationships with blood vessels and nerves. However, as the dissection progresses, the tumor appears less complex, and resection becomes simpler. Dentate ligament is sectioned early in the operation to allow safer retraction of the cord. C1 and C2 roots must be differentiated from the spinal accessory nerve and other lower cranial nerves. Although sectioning of the C1 and C2 roots has almost negligible neurological sequelae, sectioning or even handling of the lower cranial nerves can result in devastating clinical deficits. Further dissection and tumor resection will depend on tumor-related factors. Consistency, vascularity, and the extent and nature of the dural attachment and relationship with the spinal cord, nerves, and blood vessels will ultimately determine the course of surgery. Most of the tumors are relatively soft and only moderately vascular. Tumors that encase the vertebral artery are usually softer.

Fig. 40.1a–c

a Drawing showing the extent of bone exposure. The ipsilateral half of the posterior arch of the atlas and the adjoining part of the suboccipital bone are removed.
b The cerebellum is elevated, and the tumor is exposed in the cerebellomedullary corridor.
c Axial drawing showing the tumor, bone work, and exposure.
in consistency. The tumor is first debulked progressively as much as possible. This procedure relaxes the region and allows more space to work around the tumor. Perforator injury is extremely dangerous for neural function and has to be prevented. After the initial tumor debulking, the site of tumor dural attachment is dealt with. The tumor is disconnected from its attachment, and bleeding sites are coagulated. In general, during surgery, coagulation is required only at the site of attachment. Coagulation within the confines or the bulk of the tumor is unnecessary. Coagulation outside the dome of the tumor is usually a technical error. The dissection of the tumor from the nerves and vessels is done in the subsequent stage of the surgery when the region is relaxed and the tumor is devascularized. This dissection is done with the use of meticulous and careful microsurgical techniques and appropriate angulation of the microscope. Resection of the part of the condyle can be done even during the tumor resection when an additional exposure is felt to be necessary. After resection of the tumor, the site of its attachment is coagulated, and the involved layer of dura is resected. No attempts were made in our series to resect the dura widely or to excise the involved bone.

Incomplete resection is usually due to wide tumor extensions, particularly when the tumor extends far beyond the midline. The portion of the tumor where dissection from perforators or nerve fibrils is difficult or dangerous can be safely left behind. Tumors that have calcification and those with elastic consistency are more difficult to resect. Recurrent tumors can sometimes be extremely difficult for dissection. In situations where dissection of the tumor from the cervicomedullary cord is difficult or where the pial plane is lost, it is safer to leave a large strip of the tumor in proximity of the cord. Any damage to the cord in this region can be extremely dangerous.

### Our Experience

A majority of the anterior or anterolaterally placed meningiomas in our series were relatively soft, were of moderate vascularity, and had a well-defined plane of arachnoid cleavage (Figs. 40.2, 40.3, 40.4, 40.5, 40.6, 40.7, and 40.8). All of these features made the resection easier even with a relatively small exposure. The tumors were resected after they were debulked initially, and the procedure was performed within the planes provided by tumor growth. Debulking of larger tumors provided more space anterior to the brainstem, making it unnecessary to operate from a more lateral angle. In both our patients in whom a lateral condylar resection was necessary to achieve adequate exposure, the tumor size was relatively small. In our series, incomplete tumor removal was never the result of an inadequate exposure provided by the approach. Total and radical removal of all tumors was attempted, but if the dissection of the tumor from the vertebral artery, its branches, or any cranial nerve could entail risk of damage, the dissection was discontinued and that portion of the tumor was left behind. Dural and wound closure with this approach is easy and safe, and it avoids the possible difficulties described in the lateral and anterior approaches. There were no postoperative cerebrospinal fluid fistulae in our series, as there have been in the reports by authors recommending lateral or transoral approaches. Although radiosurgery has been observed to be a useful adjunct for residual meningioma, this modality of treatment was not preferred in our cases.

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**Fig. 40.2a–g**

a) Contrast-enhanced scan showing an anteriorly placed meningioma in the foramen magnum.

b) Axial scan showing the tumor and its relationship to the basilar artery.

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Text continues on page 459
Fig. 40.2c–g

c  T1-weighted magnetic resonance imaging (MRI) showing the anteriorly placed tumor.
d  Sagittal view shows that the tumor is low clival and entirely above the level of the foramen magnum.
e  T1-weighted immediate postoperative scan showing the tumor resection.
f  T2-weighted axial scan showing the tumor resection.
g  T1-weighted MRI showing the tumor resection.
Fig. 40.3a–f
a  T2-weighted MRI showing the anterolaterally placed meningioma.
b  Contrast-enhanced MRI showing the tumor.
c  Contrast-enhanced coronal scan showing the tumor.
d  Contrast-enhanced sagittal scan.
Fig. 40.3e–f

*Postoperative T1-weighted MRI showing the tumor resection.*

Fig. 40.4a–e

*a T2-weighted MRI showing the anteriorly placed foramen magnum meningioma.

*b T1-weighted MRI showing the tumor.

*c Contrast-enhanced MRI showing the meningioma.

*Postoperative T2-weighted MRI showing the tumor resection.*
Fig. 40.4d–e

d Postoperative T1-weighted MRI showing the tumor resection.

e Sagittal scan showing the tumor resection.

Fig. 40.5a–e

a T2-weighted MRI showing a large anterior foramen magnum meningioma.

b T1-weighted MRI showing the tumor.
Fig. 40.5c–e
  
  c  Axial scan showing the tumor.
  d  Postoperative scan showing the tumor resection.
  e  Postoperative axial scan.

Fig. 40.6a–e
  
  a  T2-weighted MRI showing a large foramen magnum meningioma.
  b  Scan showing the encased vertebral artery.
**Fig. 40.6c–e**

- **c** Axial scan showing the relationship of the tumor to the spinal cord.
- **d** Immediate postoperative scan showing the tumor resection.
- **e** Axial scan showing the tumor resection.

**Fig. 40.7a–d**

- **a** Coronal contrast-enhanced MRI showing the tumor and its relationship to the vertebral artery.
- **b** Axial scan showing the tumor and the encased vertebral artery.
Figure 40.8a–f  A 35-year-old woman presented with complaints of occasional giddiness. There were no focal neurological deficits. The lesion was firm, ossified, encased the vertebral artery entirely along its intracranial course, and severely compromised the neural structures. These clinical and radiological issues made surgery on this tumor a formidable exercise.

- **a** CT (plain) axial scan shows an ossified lesion anterior to the medulla and occupying a large part of foramen magnum.
- **b** T1-weighted sagittal image showing the iso- to hypointense lesion anterior to the medulla displacing it markedly posteriorly.
- **c** T2-weighted axial MRI showing the hypointense lesion located anterior to the cervicomedullary cord and encasing the vertebral artery entirely.
- **d** Contrast-enhanced scan showing the lesion and the encasement of the vertebral artery.
- **e** Postoperative MRI sagittal image showing the resection of the tumor.
- **f** Postoperative coronal image showing the vertebral artery and the tumor resection region.
Advantages of the Posterior Midline Approach

The advantages of the posterior midline approach include the following:

1. The midline skin incision and exposure of the region of the foramen magnum, occipital bone, arch of the atlas, and axis are far easier, safer, quicker, and wider than those associated with any anterior or lateral approach.
2. Extensive drilling of the occipital condyle, lateral mass of the atlas, or anterior spinal elements is not needed, so the possibility of injury to the hypoglossal nerve and vertebral artery and spinal instability is avoided.
3. The vertebral artery is exposed easily in the region of the arch of the atlas at the site of its entry into the dura by a midline approach, and proximal control is possible. Exposure of the extradural vertebral artery and its manipulation and mobilization amid large venous plexuses, which are necessary in some lateral approaches, involve considerable effort and possible risks.
4. By using a slightly larger midline skin incision and additional retraction of muscles, significant lateral exposure is obtained.
5. The extent of additional bone removal necessary can be determined after the nature of the tumor is ascertained for consistency, vascularity, and the extent and site of dural attachment.
6. Additional drilling of the condyle is possible for a more lateral exposure with directly anterior meningiomas. Condylar drilling is safer because the vertebral artery and cranial and upper cervical nerves are already exposed. Condylar resection can be done even after tumor exposure. This is a variation of the predetermined bone drilling necessary for all lateral and anterior approaches.
7. The brainstem, cranial nerves, vertebral artery, and tumor are exposed in the same field, and dissection can be performed with visualization of all structures. Exposure can be improved after retraction of the cerebellum, and the operation can be performed in the cerebellomedullary angle from a lateral perspective by use of appropriate angulation of the microscope.
8. Exposure is wide, not deep, as compared with that obtained by transcervical, transoral, and even lateral approaches.
9. Exposure of the anterolateral and anterior foramen magnum and clival dura is direct. This makes coagulation and resection of the involved dura easy after tumor resection. Such a direct exposure is difficult or impossible by a strictly lateral approach and can be obtained only if the entire condyle is resected.

Determinants of the Course of Surgery

Determinants of the course of surgery include

1. Size of the tumor
2. Extent of dural attachment
3. Extent of lateral and anterolateral extension
4. Consistency of the tumor
5. Vascularity of the tumor
6. Extent of encasement/displacement of adjoining cranial nerves and blood vessels
7. Nature of presenting clinical symptoms: The more severe the clinical deficit, the more difficult the surgical procedure
8. Recurrent or residual lesions, which are more difficult to operate

Conclusion

On the basis of this experience, we think that the time-tested, posterior midline conventional approach to most anteriorly placed foramen magnum meningiomas remains a viable and possibly better option versus the alternative approaches described recently in the literature.

References

Surgical resection of lesions located at the level of the craniovertebral junction (CVJ), especially lesions located anteriorly, is particularly challenging. Indeed, many important neurological, vascular, and bony structures need to be preserved. Their exposure and control require particular techniques that have been developed over the past 20 years. These techniques use surgical approaches that have been given different names: far lateral, extreme lateral, transcondylar, and so on. In fact, all of these approaches may be grouped under the label lateral approaches as opposed to anterior (transoral) or posterior (standard posterior midline) approaches. These lateral approaches have an axis of work either anteriorly or posteriorly, and they can be subdivided into anterior and posterolateral approaches. Each of these approaches differs in the extent of bone resection and mobilization of the vertebral artery. Whatever the chosen technique, the general principles are the same, and each surgeon has to tailor the surgical technique according to each pathological case and his or her experience. Therefore, while dealing with a CVJ lesion, the surgeon must choose the best single or combined approach, namely, the anterior approach (transoral and derived approaches), posterior approach (standard midline posterior opening), or lateral approach (anterior or posterolateral approaches).

**Anatomy**

The lateral approaches are directed to the lateral wall of the CVJ (Fig. 41.1). This lateral wall is made up of the C1–C2 joint, the lateral mass of the atlas with the transverse process and the foramen of C1, the C0–C1 joint, the occipital condyle, and the jugular tubercle. It thus comprises important elements contributing to the stability of the CVJ. In fact, this wall is in a plane that is anterior to the neuraxis. It means that with an access flush to its posterior aspect, one may reach the subarachnoid space.
anterior to the spinal cord and the medulla oblongata. It must be noticed that there is no intervertebral foramen at the C0–C1 and C1–C2 levels, as the joints are anterior to the merging of the cervical nerve roots. The lateral routes can therefore take advantage of this wide free space situated behind the lateral wall of the CVJ.

The space posterior to the lateral wall of the CVJ is crossed by the vertebral artery, which is the key structure to control in lateral approaches. The vertebral artery runs vertically from the C2 to the C1 transverse foramina along the C1–C2 joint; it then runs horizontally in the groove of the posterior arch of the atlas behind the occipital condyle, then runs obliquely superiorly and medially to reach the dura at the level of the foramen magnum. The segment of the vertebral artery between the C2 transverse foramen and the foramen magnum dura is called the V3 or suboccipital segment.

The vertebral artery has a muscular and a radicular branch at each level. The muscular branches are connected to the muscular branches of the external carotid and subclavian arteries, making a network that can revascularize the distal vertebral artery in case of proximal occlusion. The radicular branches follow the second and first cervical nerve roots; they never give origin to the anterior radiculomedullary artery. The vascular supply of the dura mater of the anterior part of the foramen magnum comes from the radicular artery of the third interspace (C2–C3) (Fig. 41.2), which runs along the vertebral body of C2, then along the odontoid process (where it is called the anterior meningeal artery). It connects with the contralateral artery and with the ascending pharyngeal artery.

The anatomy of the CVJ and the vertebral artery is not fixed, and changes occur during movements of the head.

Fig. 41.2a–d  Branches of the vertebral artery.

a Anterior meningeal artery (arrow).

b Notice the blush of tumoral injection.

c Posterior inferior cerebellar artery (PICA) with extracranial (C1–C2) origin.

d Ascending pharyngeal artery (arrow) with tumoral injection of a foramen magnum tumor.
and neck, especially rotation. Because the atlas follows the rotation of the head, whereas the axis does not, the relations between the bony elements are changing, as are the relations between the vertebral artery and these bony elements; the vertebral artery is stretched on one side and compressed on the other. During surgery with the patient in the supine position and the head rotated toward the opposite side, the C1 transverse process is projected anteriorly, and the posterior arch of atlas is brought into view. The vertebral artery is stretched on both sides of the transverse foramen of C1, with the two vertebral artery parts (the C1–C2 portion and the part above C1) running almost parallel, with only the posterior arch of the atlas interposed between them. Consequently, for surgical approaches to lesions located anterior to the CVJ, care must be taken to not rotate too much of the head.

The variations and anomalies of the CVJ anatomy must be known and identified before contemplating surgery. First, in 40% of patients, both vertebral arteries are not of equal size; one is smaller and called minor or hypoplastic (atretic when it does not join the opposite vertebral artery and ends at the posteroinferior cerebellar artery (PICA) or at the occipital artery), and the other is bigger and called dominant. PICA may use part of the course of the radicular artery of the second interspace (C1–C2). When it is the vertebral artery, it corresponds to a duplication with the intradural course of the vertebral artery (Fig. 41.3); the normal extradural segment may persist or become atretic. When it is the PICA, it corresponds to an extracranial origin of this artery (20% of cases) (Fig. 41.2). Second, there may persist a congenital anastomosis between the carotid artery and

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**Fig. 41.3a–d** Anomalies of the vertebral artery.

- **a** Duplication at the level of the C2 transverse process.
- **b, c** Duplication with intradural course of a segment of the vertebral artery.
- **d** Proatlantal artery between the internal carotid artery (arrow) and the vertebral artery (arrow).
the vertebrobasilar system. Most of these anastomoses are intracranial (trigeminal, otic, or hypoglossal), but one of them, called the proatlantal artery, is located extracranially at the level of the CVJ ([Fig. 41.3]). Bony malformations are also frequent at the CVJ: fusion of various pieces of bone or supplementary bony elements may modify the mobility of the CVJ and the relations between anatomical structures. It may even cause pathological conditions, such as intermittent compression of the vertebral artery or of the neuraxis. Lastly, calcification or sometimes ossification of the occipitoatlantal ligament turns the groove of the vertebral artery in the posterior arch of the atlas into a tunnel.21,22

### Surgical Technique

Among the lateral approaches, the posterolateral is mainly designed for intradural lesions, especially foramen magnum meningiomas. Conversely, the anterolateral approach is best applied to extradural tumors, the most common being chordoma ([Fig. 41.4]).

#### Posterolateral Approach

This is a lateral extension of the standard midline approach ([Figs. 41.5, 41.6, 41.7, and 41.8]). It is best to start from the midline, where the anatomy is familiar. The patient’s position may be sitting, prone, or lateral, depending on the surgeon’s preference. Skin incision starts in the midline, extending vertically up from the C4–C5 level to the occipital protuberance, then curving laterally along the superior occipital line toward the mastoid process. The posterior muscles are cut together with the skin, leaving a cuff of tissue for further reattachment. The muscles are split from the bone to expose the posterior fossa and generally the posterior arch of the atlas and the lamina of C2.

Bone must be split subperiosteally, especially at the level of the arch of the atlas. Exposure progresses from medial to lateral, then from inferior to superior. The lamina of C2 is exposed up to the joint of C1–C2, the posterior arch of the atlas up to the transverse foramen. Between the arch of the atlas and the lamina of C2, the C2 nerve root is identified with venous plexuses on both sides.

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Fig. 41.6a–c
a, b Posterolateral approach (right side).
c Cadaver dissection with posterior arch of the atlas (A) and vertebral artery (star). The spatula separates the periosteal sheath of the vertebral artery from the bone of the vertebral artery groove. 2 lamina of C2. Sp spinous process of C2. N second cervical nerve root.

Fig. 41.7a–d Posterolateral approach (right side), cadaver dissection.
a The posterior arch is resected up to the vertebral artery groove.
b Schematic drawing after bone resection
Fig. 41.7c–d

c The vertebral artery groove is further resected with some drilling of the occipital condyle above the vertebral artery. Figure key for a and c as in Fig. 41.6. Dm dura mater, OC occipital bone.

d The dura mater has been opened. 2 intradural second cervical nerve root, Black circle dura mater and denticulate ligament, Black star intradural vertebral artery crossed by the accessory nerve, N extradural second cervical nerve root, White star vertebral artery. The spatula shows the PICA.

Fig. 41.8 Operative views of the posterolateral approach (left side), 2 laminae of C2, a lateral mass of the atlas, A posterior arch of atlas, c occipital condyle, Dm dura mater, N second cervical nerve root, O occipital bone, S sigmoid sinus, Sp spinous process of C2, Star vertebral artery, T jugular tubercle.

a Skin incision.
b, c Bone exposure.
d, e Bone opening.
Elevating the periosteum from the inferior edge of the arch of the atlas to the superior one allows the surgeon to free the groove and mobilize the vertebral artery out of it (Fig. 41.6). In fact, this technique preserves the periosteal sheath surrounding the vertebral artery and therefore avoids troublesome venous bleeding. The entire length of the vertebral artery groove can be exposed, thus allowing bone resection up to the C0–C1 joint (occipital condyle–lateral mass of the atlas) (Fig. 41.7). For resection of the occipital bone above the vertebral artery toward the occipital condyle, the superior aspect of the vertebral artery needs to be controlled. There is no landmark at this level, as the occipitoatlantal ligament continues with the periosteal sheath of the vertebral artery. Moreover, this ligament is sometimes calcified or even ossified. The bone opening thus obtained, without any drilling of the lateral mass of the atlas or of the occipital condyle, allows the surgeon to reach the intradural space anterior to the neuraxis.

The surgical approach is then decided according to tumor location, with consideration of essentially two parameters: whether the lesion is above or below the vertebral artery, and whether it is anterior or lateral to the neuraxis. The bony resection is extended above (toward the occipital condyle) or below (toward the lateral mass of the atlas), depending on the location of the tumor with respect to the vertebral artery. The difference between anterior and lateral tumors is the axis of displacement of the neuraxis. Lateral tumors displace the neuraxis laterally and therefore enlarge the access. Consequently, bone resection does not need to be extended at its maximum. In contrast, anterior tumors develop in front of the neuraxis, which is displaced straight posteriorly. Therefore, the space lateral to the neuraxis is not enlarged, and bone has to be drilled up to the C0–C1 joint. However, drilling of the occipital condyle must be exceptional and very limited.

Dura is opened by a curvilinear incision sometimes with a counter incision toward the vertebral artery (Fig. 41.7d), so that the space between the neuraxis and the dura is maximally opened.

In any case, the bone opening must be extended toward the opposite side beyond the midline so that the neuraxis cannot be compressed against the bone during tumor manipulation. It is also important to release the neuraxis as soon as possible after the bony and dural opening to give more space to the neuraxis. This is achieved by dividing the two first arches of the denticulate ligament and the first (sometimes also the second) cervical nerve root. The first arch gives passage to the vertebral artery and the first cervical nerve root. The nerve roots must be cut lateral to their connection with the accessory nerve. Indeed, sometimes proximal segments of the first and second cervical nerve roots are the main components of the accessory nerve.

In cases where the tumor is below the vertebral artery, the lower cranial nerves are pushed cranially and posteriorly by the tumor. These nerves will be found at the superior pole of the lesion at the end of surgery. The resection must start at the caudal aspect of the meningiomas, with the goal of releasing the dural attachment and controlling the vascular supply first; then the tumor is debulked in a dry surgical field with a sucker, an ultrasound aspirator, or a laser, according to the tumor consistency. When freeing the dural insertion, it is important to keep a small part of the base, at the side of the neuraxis, undetached, to avoid free movement of the lesion, which can cause inadvertent damage to the neuraxis during the remnant resection. When hollowing the tumor, a small layer is also kept with its capsule against the neuraxis. This part will be resected as the last surgical step and under better conditions when the meningioma is completely devascularized and the surgical field is widely open.

If the tumor is above the vertebral artery, two special points must be taken into consideration: the displacement of the lower cranial nerves and the dissection of the vertebral artery branches. Indeed, in this location, the displacement of the lower cranial nerves cannot be anticipated. To prevent damage, the rootlets must be under control on the side of the jugular foramen, then followed along their courses more or less adherent to the meningioma. With the lesion being progressively debulked, the nerve rootlets can be more easily mobilized, often inferiorly, to allow a more confident tumor resection at some distance from fragile nerve structures. The tumor dissection from the vertebral artery branches, especially the PICA, is another difficulty encountered with tumors located above the vertebral artery. Precise knowledge of the anatomy based on preoperative investigation is mandatory.

If the meningioma encases the vertebral artery, the technique is as described above. Special consideration is nevertheless required if the meningioma has its insertion on the dura surrounding the vertebral artery penetration. The dural resection is better achieved by progressing from the extradural side toward the intradural aspect, along the vertebral artery, because the vertebral artery invaginates into the dura with its periosteal sheath. This furrow can be resected as long as the vertebral artery adventitia is not invaded.

A watertight dural closure is required to prevent postoperative cerebrospinal fluid (CSF) leakage. The closure is generally easy with a curvilinear incision. If necessary, a dural patch using the suboccipital aponeurosis achieves a perfect closure. In any case, the muscular and aponeurotic layers must be tightly closed.

Anterolateral Approach

This is the standard technique used to expose the vertebral artery all along the neck but applied on the suboccipital (V3) segment (Figs. 41.9, 41.10, 41.11, 41.12, and 41.13). The general principles are the same whatever the segment but with some modifications in relation to the anatomy at this level.3,5

Text continues on page 469
Fig. 41.9a–f  Schemes of the different steps of the anterolateral approach.

a  Skin incision.
b  Opening between the internal jugular vein (IJV) and the sternomastoid muscle.
c  Retraction of cranial nerve (CN) IX.
d  Cutting of muscles on the transverse process of the atlas.
e  Exposure of the vertebral artery from C2 to the foramen magnum.
f  Opening of the jugular foramen.

Fig. 41.10a–c  Anterolateral approach, cadaver dissection. F fatty pad rolled around the accessory nerve, N accessory nerve, T transverse process of the atlas, V internal jugular vein.
Fig. 41.11a–d  Anterolateral approach, cadaver dissection. A posterior arch of the atlas, M mastoid process, N accessory nerve, Star vertebral artery, V internal jugular vein.

Fig. 41.12a–c  Anterolateral approach, cadaver dissection. In b, the vertebral artery is transposed out of the transverse foramen of the atlas with the occipital condyle (O). The blade retracts the IJV and the accessory nerve. 1 lateral mass of the atlas, 2 C1–C2 facet joint, B jugular bulb, Dm dura mater, S sigmoid sinus, Star vertebral artery, V IJV.
The patient is placed in the supine position with the head extended and more or less rotated toward the opposite side (Fig. 41.9). It must be kept in mind that the more the head is rotated, the more of the anterior arch of the atlas is projected away and the more the posterior arch is brought into view. Therefore, rotation must be chosen according to the part of the CVJ that has to be reached.

Skin incision follows the medial edge of the sternomastoid muscle for 10 cm, its upper part going up to the tip of the mastoid process and then along the mastoid process and the superior occipital crest toward the inion. The sternomastoid and posterior cervical muscles are detached from the occipital bone and mastoid process. The digastric muscle is separated from its groove and reflected anteriorly. Next, the internal jugular vein is exposed at the C3 level, and the space between the internal jugular vein and the sternomastoid is opened. This space is filled with fat and lymphatic elements. The accessory nerve (CN XI) must be identified and dissected free from its junction with the sternomastoid up to its crossing with the internal jugular vein and to the skull base. For easier identification, the patient must not receive any curare-like drugs so that working close to the nerve does not induce contraction of the sternomastoid. The fat pad surrounding the nerve is freed from the deep plane of muscles and rolled around the nerve for protection. A stitch is used to retract this fat pad as well as the nerve, both inferiorly and medially (Fig. 41.10).

Next, the small muscles inserted on the transverse process of the atlas are divided flush with the tip of this process. This transverse process is easily recognized, as it is projected anteriorly by rotation of the head. It can be felt with the finger 10 to 15 mm anterior and inferior to the tip of the mastoid process. Dividing all the muscles attached on the transverse process of the atlas brings into view the two segments of the vertebral artery (the C1–C2 segment and the part above C1) on both sides of the posterior arch of the atlas. Care must be taken to keep the periosteal sheath surrounding the vertebral artery intact to avoid any troublesome venous bleeding (Fig. 41.11).

Still working subperiosteally, the transverse foramen of the atlas can be unroofed with resection of the transverse foramen. This permits mobilization and, whenever necessary, transposition of the vertebral artery out of the
transverse foramen. When the vertebral artery is exposed from C2 to the end of the groove in the posterior arch of the atlas, any part of the CVJ on one side can be reached. The surgeon can work on the posterior part or move in front of the anterior arch of the atlas in the retropharyngeal space or work through the bone elements (Fig. 41.12). In fact, when instability already exists due to tumoral invasion or inflammatory process destroying the ligaments, drilling the lateral wall of the CVJ does not worsen the patient’s status and is an acceptable technique. Drilling the occipital condyle gives access to the tip of the clivus; drilling the lateral mass of the atlas gives access to the odontoid process and the contralateral mass of the atlas; and drilling the jugular tubercle leads to the jugular foramen (Figs. 41.13 and 41.14). The strategy must be adjusted according to the location and extent of tumor. The shortest way to reach a tumor is often the best, as it does not destroy important structures. As a rule, CVJ stability may be compromised by tumor development but not by surgical work.

Through the anterolateral approach, the surgeon can follow intradural extensions of a tumor which has its main bulk in the bone and the extradural space. However, this approach is certainly not well designed for intradural lesions.

### Personal Experience

Since we started using the lateral approaches in 1977, they were applied on different pathologies. The first indications were revascularizations of the distal vertebral artery; next we dealt with extradural tumors, then with intermittent VA compression, then with intradural foramen magnum tumors and finally with jugular foramen tumors.

Table 41.1 lists the different types of tumors that were treated by the postero- and anterolateral approaches at the CVJ level.

The posterolateral approach was used mostly on intradural meningiomas (Figs. 41.15 and 41.16). Few intradural meningiomas had some extradural extension that could be removed through the same approach (Fig. 41.17). Some very rare cases were entirely extradural and required an anterolateral approach. Neurinoma is the second most common type of intradural tumor. However, only 15% of them were strictly intradural, and 85% were extradural or intra- and extradural with an hourglass configuration. In fact, because neurinomas always have a lateral location, the posterolateral approach is used most frequently. The posterolateral approach is also more useful in cases of bilateral neurinomas because this approach can be performed on both sides in the same stage. Other pathologies are less common (Table 41.1).

### Table 41.1 Personal experience with tumors of the craniocervical junction

<table>
<thead>
<tr>
<th>Extradural tumors</th>
<th>144 (anterolateral approach)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Meningioma</td>
<td>3</td>
</tr>
<tr>
<td>Neurinoma</td>
<td>34</td>
</tr>
<tr>
<td>Chordoma</td>
<td>43</td>
</tr>
<tr>
<td>Sarcoma</td>
<td>19</td>
</tr>
<tr>
<td>Metastasis</td>
<td>17</td>
</tr>
<tr>
<td>Osteoid osteoma</td>
<td>7</td>
</tr>
<tr>
<td>Other primary bone</td>
<td>21</td>
</tr>
<tr>
<td>Pseudotumors</td>
<td>29 (anterolateral approach = 22; posterolateral approach = 7)</td>
</tr>
<tr>
<td>Intradural tumors</td>
<td>133</td>
</tr>
<tr>
<td>Meningioma</td>
<td>92 (86 anterior or anterolateral)</td>
</tr>
<tr>
<td>Neurinoma</td>
<td>27</td>
</tr>
<tr>
<td>Others</td>
<td>14 (12 anterior or anterolateral)</td>
</tr>
</tbody>
</table>

![Image of anatomical diagram](https://via.placeholder.com/150)
Fig. 41.15a–c  Lateral foramen magnum meningioma. MRI with a axial, b coronal, and c sagittal views. Notice the lateral displacement of the neuraxis not visible on the sagittal midline view at the tumor level.

Fig. 41.16a–d  Anterior foramen magnum meningioma. MRI with (a, c) axial and (b) sagittal preoperative views. (d) Sagittal postoperative view. Star vertebral artery. Notice the straight posterior displacement of the neuraxis visible on the sagittal midline view behind the tumor.
Lateral Approaches to Anteriorly Located Lesions of the Craniovertebral Junction

Posterolateral approach is more or less extended laterally depending on the exact location of each tumor. Menigiomas with a lateral location that displace the neuraxis laterally do not need a very lateral approach (Fig. 41.15). Conversely, the anterior location of a tumor that pushes the neuraxis straight posteriorly requires moving the medulla and thus a very lateral exposure (Fig. 41.16).

The anterolateral approach was essentially used for extradural and bony lesions, as listed in Table 41.1. The most common pathology was chordomas, followed by various types of primary bone tumors (Fig. 41.18). Because of their growth in many different directions, chordomas may necessitate the combination of several approaches in the same or in a different stage, including the anterolateral approach, for example, transoral and anterolateral approaches or bilateral anterolateral approaches. Radical resection with proton therapy seems to be very beneficial in terms of survival for patients with chordomas.

Besides tumors, several types of pseudotumors may be observed at the CVJ level, raising a question as to the goal of surgery: decompression or resection (Fig. 41.19). As a matter of fact, bony lesions at the CVJ often have mistaken features. Diagnosis of tumor or pseudotumor is often very difficult. Indeed, besides bone tumors with specific radiological features (e.g., osteoid osteoma or osteochondroma), many bone tumors demonstrate invasive features suggesting malignancy, although they are histologically quite benign.

Pseudotumors include inflammatory processes such as pannus associated with rheumatoid arthritis or synovial cysts, infectious pathologies, especially tuberculosis, degenerative changes, and bony malformations.

Finally, in cases of instability, a fixation procedure must be contemplated. As previously mentioned, instability could be related to tumor extension and therefore could be present before tumor resection. If present, correct positioning of the patient for the anterolateral approach may not be possible. The anterolateral approach can then be realized with the head straight without rotation, but this is much more difficult, especially if the tumor is located in the upper part of the CVJ. Nevertheless, patients with recurrent chordomas who have been stabilized can be reoperated on without removing the instrumentation. Most of the time CVJ stabilization is achieved by occipitocervical fixation through a posterior approach. However, in some cases, a fixation technique can be realized through the anterolateral approach (Fig. 41.20): an iliac bone graft is impacted between what is remaining...
of the occipital condyle and the C2 facet. Some authors advocate plating between the occipital bone and the C2 vertebra. In our experience, impaction of the bone graft using a vertebral spreader is sufficient. Obviously, this grafting technique can only be used in cases of small unilateral tumors.

Discussion

Yasargil et al. reviewed several series published from 1924 to 1976 and counted 114 cases of foramen magnum meningiomas. Since then, more than 400 cases have been reported. In 1993, we collected 106 meningiomas from 21 centers in France over 10 years. The recently published series are very heterogeneous and include pathologies other than meningiomas. Moreover, the exact location anterior or anterolateral to the neuraxis is not clearly and uniformly specified. Recurrent tumors account for up to 80% of some series.

Foramen magnum meningiomas are undoubtedly challenging tumors, requiring special considerations because of the vicinity of the brainstem, medulla oblongata, lower cranial nerves, and vertebral artery. Several approaches have been advocated. The goals are to achieve the greatest tumor removal and the lowest morbidity rate.

There is no discussion about the best surgical approach for posterior foramen magnum meningiomas. The posterior approach is the best option, as it is well known by neurosurgeons and is associated with a low morbidity rate. For anterior foramen magnum meningiomas, the transoral approach has been reported sporadically. Despite providing access to the anterior part of the CVJ, this approach has several drawbacks in case of intradural lesions: increased risk of CSF leak and meningitis after violation of the contaminated oral cavity; poor access to laterally extending tumors, resulting in a low rate of complete resection; and increased risk of postoperative instability and velopalatine insufficiency. The two main surgical approaches reported in the literature are the far lateral approach, also called the
posterolateral or lateral suboccipital approach, and the extreme lateral approach, also called the anterolateral approach. As detailed by Rhoton, the far lateral approach is a lateral suboccipital approach directed behind the sternocleidomastoid muscle and the vertebral artery and just medial to the occipital condyles and the atlanto-occipital joint. The extreme lateral approach is a direct lateral approach deep to the anterior part of the sternocleidomastoid muscle, behind the internal jugular vein and along the front of the vertebral artery. Both approaches permit drilling of the occipital condyle but provide a different exposure because of the differences in the direction of approach. Most of the time, a partial transcondylar technique is used in the extreme lateral transcondylar approach. During the partial transcondylar approach, the posterior one third of the occipital condyle and the superior facet of C1 are drilled away. Arnautović et al. published their experience in a series of 18 ventral foramen magnum meningiomas. They used the extreme lateral transcondylar approach. The condyle drilling ranged from approximately one third to one half of the condyle, without causing craniocervical instability. The extreme lateral transcondylar approach requires vertebral artery transposition to reach and drill the occipital condyle.

For intradural tumors, we advocate the posterolateral approach (far lateral approach) even for anterior intradural foramen magnum meningiomas. During this approach, the vertebral artery is controlled in the horizontal portion of the V3 segment, above the C1 posterior arch. The extent of foramen magnum lateral wall drilling is variable, and is

Fig. 41.20a–e Bone grafting through the anterolateral approach.

a–c CT scan showing histiocytosis invading the lateral mass of the atlas (arrow).

b–d CT scan after resection and iliac bone grafting (asterisk)

e Operative view showing the vertebral artery (star), bone graft (black circle), and accessory nerve (arrow). The blade retracts the IJV and digastric muscle.
in fact directly proportional to the tumor extension to the contralateral side. In the literature, occipital condyle resection varies from 0 to 66%. In a recent publication, Bassiouni et al. classified surgical approaches into two groups, transcendylar or retrocondylar, depending on whether the occipital condyle is drilled or not. In our review of the literature, we found nine series in which the retrocondylar approaches were used to reach anterior or anterolateral meningiomas. In four of these, a retrocondylar approach was not used in all cases. The five other series are homogeneous and included only patients treated through a retrocondylar approach. Of these, three studies reported complete resection in 100% of the cases. In the other two, complete resection was noted in 90% and 96% of cases, and the remaining lesions were resected subtotally. Surgical results were good and surgical morbidity and mortality rates low.

Extradural lesions can be treated by either the posterolateral or anterolateral approach (far lateral or extreme lateral approach). The extent of drilling during the procedure can be larger but is dictated by tumoral invasion and must be limited to the destroyed or invaded bone. Thus, the question of instability is only a preoperative concern. In our experience of tumors located at the CVJ, we have not observed any instability if less than half of the C0–C1 and C1–C2 joints were resected. Vertebral artery transposition was performed in select cases of foramen magnum meningiomas with extradural extension and never for intradural lesions.

The permanent morbidity rate in the literature is between 0% and 60%. This rate is lower for a far lateral approach, either transcendylar or retrocondylar, than for an extreme lateral transcendylar approach. Lower cranial nerve dysfunctions are the most frequently encountered preoperative deficits. These deficits have the propensity to recover postoperatively, except in cases of en plaque meningiomas or recurrent tumor.

Several factors make the surgical procedure still more difficult, thus negatively influencing the morbidity rate. These are anterior tumor location, tumor size (smaller intradural lesions are more difficult to resect), tumor invasiveness, extradural extension, vertebral artery encasement, absence of arachnoidal sheath, and adherence in recurrent lesions.

Based on a multicentric study of foramen magnum meningiomas from 21 hospitals, George et al. reported 77%, 16%, and 7%, of complete, subtotal, and partial removal, respectively. Over the past 20 years, most of the studies reported complete or subtotal removal of the tumor. Factors limiting complete resection are adherence of the lesion to vital structures, vertebral artery encasement, and invasiveness of the lesion. Adhesions are observed during repeat surgery and explain the lower rate of complete tumor resection (60–75% of Simpson grade I) in surgical series in which a high rate of recurrent foramen magnum meningiomas are included. In recurrent tumors, some authors advocate leaving a small tumor remnant to maintain a low morbidity rate. Vertebral artery encasement was noted in 38 to 59% in some series. This factor was recognized as an independent factor for incomplete removal. The location and extension of the meningioma into the extradural space reflect the tumor invasiveness. These tumors are less favorable for complete resection than pure intradural lesions. In the French cooperative study, the rate of complete removal of intradural, extradural, and intra/extradural meningiomas was 83%, 50%, and 45%, respectively.

Our experience was published in 1997 after the treatment of 40 foramen magnum meningiomas operated in the period 1980–1993. Thirty-four lesions were intradural, two were extradural, and four were intra/extradural. Eighteen were anterior, 21 lateral, and 1 posterior. The tumor was above the vertebral artery in 4 cases, below in 20 cases, and on both sides in 16 cases. The posterolateral approach was used in 31 cases, the anterolateral in 5, and the posterior midline in 4. The rates of complete resection for intradural and extradural lesions were, respectively, 94% and 50%. Postoperatively, the clinical condition improved in 90% of patients, remained stable in 2.5%, and worsened in 7.5%. Our present experience over the past 25 years is now based on 97 foramen magnum meningiomas (unpublished data). Complete removal was achieved in 86% and subtotal removal in 11%. Subtotal removals were due to extradural extension or to recurrent cases. The rate of complete removal increased to 94% when considering only intradural lesions treated at first presentation. We mainly use the posterolateral (far lateral retrocondylar) approach for intradural foramen magnum meningiomas. In some cases, the drilling of the foramen magnum lateral wall has to be performed for intradural anterior meningiomas but remains limited to the medial 20% of the foramen magnum lateral wall. The extent of drilling has to be tailored according to the tumor characteristics. Cadaveric study has demonstrated that increasing the bony drilling is not associated with a significant widening of the surgical corridor. In fact, resecting one third to one half of the occipital condyle increases the visibility, respectively, by 15.9 and 19.9°. Moreover, these studies did not consider the fact that in surgical approaches to anterior lesions, space-occupying lesions can enlarge the surgical corridor. Therefore, the supposed benefit in terms of exposure provided by the extreme transcondylar approach is counterbalanced by the risks associated with the accessory nerve dissection, the vertebral artery transposition, and the condylar drilling.

In contrast, bony tumors require a more or less extensive drilling of the bony structures. The anterolateral approach (extreme lateral approach) must be tailored according to the tumor. Drilling of the condyle and/or the lateral mass of the atlas and jugular tubercle may be needed. Therapeutic strategy may also require a combination of surgical approaches, such as bilateral lateral approaches or transoral and lateral approaches. It has been demonstrated that in cases of chordomas, aggressive surgery plus high-energy radiation therapy is most effective at first presentation. In such cases, survival rates of 80% and 65% and recurrence-free rates of 70% and 35%, respectively,
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at 5 and 10 years can be obtained. Conversely, the same strategy after recurrence gives only 50% and 0% survival at 5 and 10 years, respectively, and 0% recurrence-free rate at 3 years. Similarly, the delay before the first recurrence is as long as 43 months at first presentation versus 15 months in case of recurrence. The average number of recurrences per year is also much higher in cases of aggressive treatment after recurrence than at first presentation: 0.62 versus 0.15. Therefore, neurosurgeons dealing with chordomas or any other malignant tumors should be able to choose among all the different approaches to the CVJ, including the lateral approaches.

Case Illustration

A 62-year-old woman was complaining of cervical pain and stiffness of the neck for 6 months despite different medical treatments. At the neurological exam, a pyramidal syndrome was found but without sensory or motor deficit. MRI discovered a foramen magnum tumor located anteriorly. Neuraxis was displaced straight posteriorly and was entirely visible behind the tumor on the midline sagittal view. Complete removal could be achieved through a posterolateral approach without inducing any postoperative deficits (Fig. 41.16).

Case Illustration

A 48-year-old woman had cervical pain, limitation of head rotation, and paresthesias in the left arm. She also had some swallowing difficulties. Neurological exam showed a pyramidal syndrome on the left side. On MRI, a mass with features of meningioma was demonstrated in the anterior part of the foramen magnum. The neuraxis was displaced to the left by the tumor, which therefore originated from the anterolateral dura of the foramen magnum. In fact, the tumor had a dumbbell shape as it extended extradurally into the clivus. Complete removal was achieved using a posterolateral approach, and the extradural cavity was filled by a piece of cervical muscle. Postoperative recovery was uneventful (Fig. 41.17).

Case Illustration

A 43-year-old man was referred because of neck pain and stiffness severely limiting his head movements. CT and MRI studies showed a tumor involving the left anterolateral part of the CVJ, including the C1–C2 joints and part of the odontoid process. The vertebral artery was minor on the tumoral side and was completely encased by the tumor. Surgical resection was achieved through a left anterolateral approach with sacrifice of the left vertebral artery. The fat pad dissected in the approach was transposed into the cavity to fill the dead space. Surgical resection was immediately followed by posterior occipitocervical fixation. Pathology confirmed the diagnosis of chordoma. The patient was then treated by radiation and proton therapy. Follow-up was uneventful with no recurrence after 3 years (Fig. 41.21).

Fig. 41.21a–d  Chordoma. MRI (a) sagittal and (b) axial preoperative views. Arrow vertebral artery, T tumor.
A 46-year-old man had been suffering from neck pain and torticollis over the last year. He presented with left cranial nerve (CN) XII palsy with tongue atrophy. He had a first surgery through a transoral approach in another department. Histological diagnosis was chordoma. Only part of the tumor could be removed, and he was sent for resurgery. This was achieved through bilateral anterolateral approaches. A halo with traction was placed at the end of surgery and replaced by posterior occipitocervical fixation several days later. Then the patient underwent radiation and proton therapy. The left atrophy of the tongue persisted, but the patient is well, without recurrence after 2 years (Figs. 41.22 and 41.23).

Case Illustration

A 46-year-old man had been suffering from neck pain and torticollis over the last year. He presented with left cranial nerve (CN) XII palsy with tongue atrophy. He had a first surgery through a transoral approach in another department. Histological diagnosis was chordoma. Only part of the tumor could be removed, and he was sent for resurgery. This was achieved through bilateral anterolateral approaches. A halo with traction was placed at the end of surgery and replaced by posterior occipitocervical fixation several days later. Then the patient underwent radiation and proton therapy. The left atrophy of the tongue persisted, but the patient is well, without recurrence after 2 years (Figs. 41.22 and 41.23).

Fig. 41.22a–c  Chordoma. Preoperative MRI with a coronal, b sagittal, and c axial views. 2 vertebral body of C2, Arrow vertebral artery, T tumor.

Fig. 41.22c  Chordoma. Preoperative MRI with a coronal, b sagittal, and c axial views. 2 vertebral body of C2, Arrow vertebral artery, T tumor.
Fig. 41.23a–d  Chordoma (same case as in Fig. 41.22). CT with (a) sagittal, (b) coronal, and (c, d) axial postoperative views. Bilateral anterolateral approach was taken with fat graft used in the cavity. Arrow vertebral artery.
A 55-year-old man was referred with the diagnosis of malignant tumor of the right lateral mass of the atlas, including the C0–C1 and C1–C2 joints. He was complaining of severe neck pain with almost complete limitation of head and neck movement. He also had changes in his voice and more recently some swallowing difficulties. He had no objective symptom at the clinical examination and no relevant past medical history. Myeloma was eliminated from blood sampling. MRI and CT studies showed a large mass that enhanced markedly after contrast injection. Angiography demonstrated supply from vertebral artery branches, including the anterior meningeal artery. Surgical resection was achieved through a right anterolateral approach; the vertebral artery was exposed and controlled from C3 to the dura of the foramen magnum so that it could be preserved. In the same stage, posterior occipitocervical fusion was performed. Histological study demonstrated an undifferentiated metastatic carcinoma possibly of renal origin. Immediate follow-up was uneventful. Several months later, multiple metastases in the lung and the humerus were discovered, which led to the patient’s death (Figs. 41.24 and 41.25).

Fig. 41.24a–d Metastasis of a kidney carcinoma. Preoperative MRI with (a) coronal and (b, c) axial views. (d) Preoperative CT coronal view. T tumor. Star vertebral artery.

Fig. 41.25a–d Metastasis of a kidney carcinoma (same case as in Fig. 41.24).

a Preoperative MRI axial view. T tumor. Arrow vertebral artery.

b Angiography with high vascularization from vertebral artery branches (including the anterior meningeal artery).
**Conclusion**

Lateral approaches are complementary techniques to transoral and midline posterior approaches to the craniovertebral junction. They lead to the lateral wall of the CVJ following a posterior or anterior axis using, respectively, a lateral extension of the posterior midline approach or an upper extension of the cervical approach to the vertebral artery. The strategy must be defined according to the location of the tumor, with the aim of preservation of CVJ stability. Morbidity related to the surgical approach is very limited. Overstretching of the accessory nerve in the anterolateral approach must be avoided by control of the sternomastoid retraction. Vertebral artery injury is another possibility that should not occur if the surgeon respects the principle of vertebral artery extraperiosteal exposure. Encasement of the vertebral artery by tumors is another problem related to tumor extension and not to the approach itself. When the vertebral artery seems to be completely encased by a tumor, the periosteal sheath is rarely invaded, allowing the vertebral artery to be preserved. However, with radical resection, sacrifice of the vertebral artery may be contemplated in cases of chordomas or malignant tumors. This was done in our series of four cases.

**References**

A wide range of vascular, neoplastic, and degenerative disease processes involve the foramen magnum and the craniovertebral junction (CVJ). Different approaches to this region have been described, which are mostly an extension of the classic lateral suboccipital approach. Overall, the choice of one approach over another often depends on the degree of comfort of the individual surgeon with each approach, the nature of the pathology of the offending lesion, and the exact location of the lesion. Despite the introduction of lateral approaches to lesions located in the anterior aspect of the foramen magnum, some authors still find the conventional posterior midline approach suitable and most appropriate for surgery.

Improvements in skull base techniques and better understanding of the relevant anatomy have led to the development of several modifications of the lateral suboccipital approach. The so-called far lateral approach is the workhorse for a variety of lesions involving the anterolateral brainstem and upper cervical cord. The far lateral approach has served as the framework for the development of modifications with the dorsolateral-suboccipital-transcondylar approach, extreme lateral transcondylar approach, and extreme lateral inferior transtubercular exposure. In this chapter we describe step-by-step the far lateral approach, and at each step we describe the modifications useful to deal with specific pathologies.

### Positioning

Patient positioning depends primarily on the nature and location of the offending lesion. A prone position is used, as it avoids distortion of the upper cervical anatomy. Rotation of the table and/or the microscope is required to allow adequate visualization lateral and ventral to the cervicomedullary junction. Some authors prefer a lateral decubitus position without any flexion of the neck so as to not compromise the cervicomedullary junction, which is often already distorted by a preexisting compressive lesion. This position also avoids alteration of the course of the vertebral artery from C2 to its dural entrance. Some authors still prefer and recommend the sitting position for surgery.

For lesions located anterolateral to the cervicomedullary junction, a modified “park bench” position is generally preferred (Fig. 42.1). The contralateral arm and shoulder are kept in a dependent fashion below the level of the body to increase the amount of cranial flexion and rotation. The head of the patient is positioned so that the ipsilateral mastoid tip is the highest point of the surgical field. The ipsilateral shoulder is gently retracted or pulled to increase the distance between the patient’s head and the shoulder, thus increasing the degree of rotation of the microscope. Three key movements of the head are performed before it is fixed in a modified Mayfield head holder: (1) the head is flexed so that the chin is 1 cm from the sternum; (2) the head is then rotated contralateral to the lesion, thus increasing the angle between the atlas and the foramen magnum to the maximum; and (3) the head is laterally flexed to −30° toward the contralateral shoulder (Fig. 42.2).

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Fig. 42.1 Modified park bench position, superior view. (With permission of Barrow Neurological Institute.)
Incision

Although some authors use a straight vertical or “lazy S” incision, we prefer a “hockey stick” incision. The incision starts at the tip of the mastoid and is brought superiorly above the nuchal line if a wide exposure, including the ventrolateral pons, is required; otherwise, it can be extended just above the mastoid tip and curved medially to reach the midline. The incision is then extended inferiorly at the midline to the spinous processes of C2 and C3, depending on the inferior degree of exposure required (Fig. 42.3).

The skin flap is elevated while preserving the fascia of the neck muscles. Leaving a fascial cuff attached to the bone facilitates reconstruction of the fascial-muscular plane at the conclusion of the procedure and allows watertight closure of this plane, minimizing chances of cerebrospinal fluid (CSF) leak.

Muscle takedown is conducted as a single flap, which is then rotated anteriorly and retracted with fishhooks attached to rubber bands. The rubber bands are then attached under tension to a Leyla bar. Retraction offered by fishhooks is advantageous for many reasons, including

1. They provide “dynamic retraction,” which can be easily adjusted intraoperatively.
2. By applying retraction and at the same time downward tension on the skin flap, it is possible to flatten the edges of the wound, thus decreasing the depth of the operative field.

Exposure

While taking down the muscle flap, it is important to recognize several bony landmarks. The transverse process of C1 is an important landmark in estimating the extent of lateral exposure. The posterior arch of the atlas is followed laterally and superiorly all the way to the sulcus arteriosus.

Exposure of the C2 lamina as far laterally as possible facilitates adequate retraction of the paraspinous muscles and allows adequate visualization and lateral extension of the approach even in patients with a short, thick neck.

The extradural portion of the vertebral artery is an important landmark for any lateral approach to the foramen magnum. The vertebral artery is identified in its extradural portion at the level of the sulcus arteriosus. The degree of exposure and mobilization of the vertebral artery vary according to the type of pathology, degree of involvement of the artery itself, and need for proximal control in its extradural portion.

In most cases, isolation of the vertebral artery may not be necessary. More often it is sufficient to recognize and identify the artery at its extradural/intradural transition, so it can be protected during condylar drilling. Isolation and skeletonization of the vertebral artery are indicated when the artery is encased by tumor and significant manipulation of the vessel is anticipated. In such a situation, the surgeon must be aware of the generous venous plexus surrounding the artery, which can be a source of tedious bleeding during exposure (Fig. 42.4).

Depending on the inferior extent of exposure required, the ipsilateral portion of the posterior arch of the atlas can be removed with rongeurs or with a high-speed drill. Either a craniotomy or a craniectomy can then be performed. The superior extent of bone removal is dependent upon the degree of superior exposure required (Fig. 42.5).
Partial drilling of the occipital condyle increases the degree of lateral exposure (Fig. 42.6). In addition, partial removal of the posterior third of the condyle offers a flat exposure of the cervicomedullary junction, thus minimizing the need for retraction. The amount of condylar resection varies with the modification of the lateral approach being used. A summary of the different modifications of the far lateral approach variants and their key characteristics is given in Table 42.1.

In our experience, the majority of lesions affecting the intradural vertebral artery can be easily dealt with through limited drilling of the occipital condyle. Similarly, the majority of intradural mass lesions, such as meningiomas of the foramen magnum, can be easily resected without extensive condylar drilling by taking advantage of the additional space created during intratumoral debulking. Therefore, any unnecessary procedural steps, such as extensive condylar drilling (more than the posterior third) and exposure of the vertebral artery with coagulation of the venous plexus, should be avoided.18
Dural Incision

The dura mater is opened in a curvilinear fashion. The incision is begun at the superolateral aspect of the craniotomy, starting several millimeters below the sigmoid sinus, advancing toward the C2 lamina, and staying posterior to the dural entrance of the vertebral artery (Fig. 42.7). After the dura is opened, the exposure obtained encompasses the lower cranial nerves to C2 (Fig. 42.8).

If transposition of the vertebral artery is required, the dural incision may be extended anteriorly, in a T-shaped fashion, toward the dural entry point of the vertebral artery and advanced circumferentially to free its attachment. A dural cuff around the vessel is preserved for watertight dural closure.19 In cases where there is dural involvement by tumor or dural shrinkage at the end of the procedure, a fascia lata graft should be harvested and interposed. Alternatively, any of the commercial dural substitutes may be used. When the lesion encases the vertebral artery at its entry point, the ring of dural sheath has to be excised, and the closure must be augmented with a fascial graft sewn to the periadventitial tissues or the vertebral artery adventitia itself. Fibrin glue may be added to achieve a watertight seal.20

Assessing the Need for Occipital Condyle Drilling

Condylar resection is considered the key maneuver of the aforementioned approaches and represents the crucial step, allowing increased exposure of the ventrolateral surface of the brainstem and foramen magnum and minimizing brain retraction.

An increasing number of anatomical studies and quantitative analyses have been conducted with the aim of comparing the prospective angle and depth of the surgical field between the far lateral approach and its variants, versus the lateral suboccipital/retrosigmoid craniotomy.1,21–24 A partial condylectomy adds a few degrees to the viewing angle of the surgeon. It also provides a broader surgical path for more effective manipulation of surgical instruments in such a deep operative field. Drilling of the jugular tuberculum provides the most relevant increase in the viewing angle of the middle portion of the lower clivus.

Although some authors recommend a graded amount of condylar removal,1,20 others conclude that slight1,23 or no22,25 condylar resection is needed. The degree of further exposure gained through condylar resection is also dependent on individual variables:

- Length and orientation of the anteroposterior axis of the condyle
- Length of the anteroposterior axis of the foramen magnum

Table 42.1 Variants of the far lateral approach

<table>
<thead>
<tr>
<th>Variant</th>
<th>Key Characteristic</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>Retrocondylar</td>
<td>No condylar removal required</td>
<td>Mainly for intradural lesions of the cervicomедullary junction</td>
<td>Careful drilling around the CN XII canal; requires occipitocervical fusion</td>
</tr>
<tr>
<td>Partial transcondylar</td>
<td>Removal of the posterior third of the condyle and part of the superior facet of C2</td>
<td>Mainly for extradural lesions</td>
<td>Technically complex maneuver, not always completed extradurally</td>
</tr>
<tr>
<td>Complete transcondylar</td>
<td>Total removal of the condyle, also anterior to the hypoglossal canal</td>
<td>Better visualization of the vertebrobasilar junction</td>
<td></td>
</tr>
<tr>
<td>Transtubercular</td>
<td>Extrudal removal of the jugular tuberculum above the CN XII canal</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
• Posterior brainstem distortion by the tumor mass
• Cisterna magna decompression and distance between the ventral surface of the brainstem and the anterior edge of the foramen magnum
• Location of the lesion as compared with cranial nerve rootlets

Condylar resection appears to be helpful in reaching a small ventral lesion in a patient with large condyles and an elliptical foramen magnum with a long anteroposterior diameter and narrow width. A short distance between the pontomedullary ventral surface and the anterior edge of the foramen magnum and lower clivus also favors condylar resection. Therefore, in the case of a vascular lesion, each approach should be tailored to the individual patient. Careful preoperative planning based on neuroimaging should dictate the anticipated degree of exposure so that unnecessary bone removal is avoided.

Assessing the Need for Occipitocervical Fusion

The major structures providing stability to the CVJ complex include the occipital condyles and the lateral masses of the atlas, as well as the alar and transverse ligaments, which attach to the medial anterior third of the occipital condyle. Even when partial condylectomy is performed, it is usually done along the medial surface of the posterior third of the condyle; therefore, it does not compromise the integrity of the CVJ. It has been reported that patients undergoing up to 70% of condylar bone mass removal do not need further surgery in the form of occipitocervical fusion. On the other hand, extensive condylar resection and damage of the atlanto-occipital ligamentous apparatus may lead to instability of the CVJ; in these cases, occipitocervical fusion is indicated and usually performed a few days after the first operation.

Complications and Their Avoidance

Cranial nerve deficits as a result of surgical manipulation (CN VI, VII, VIII, IX, X, XI, and XII) are by far the most common complications immediately after surgery, with a 50% chance of complete recovery at 6 months’ follow-up. The hypoglossal nerve (CN XII) is the most frequently involved cranial nerve; it may be injured either during condylar drilling or during intradural manipulation of the offending lesion. Patients should be carefully monitored postoperatively for development of a lower cranial nerve dysfunction that may lead to significant swallowing dysfunction and ultimately aspiration pneumonia. If significant cranial nerve manipulation is anticipated intraoperatively, or in patients with preoperative lower cranial nerve dysfunction, elective preoperative tracheostomy and gastrostomy should be considered.

Vertebral artery injuries can occur at any time during the early phases of exposure, particularly in those patients undergoing mobilization of the artery at the C1 foramen transversarium; inadvertent lacerations may be controlled with temporary clipping and tamponade. Intraoperative vertebral artery thrombosis may also occur, probably as a result of excessive manipulation of the artery. Vertebral artery injury may lead to brainstem infarction, manifesting with quadriplegia, respiratory insufficiency, urinary incontinence, or even death. If vertebral artery injury is suspected, conventional cerebral angiography is required to rule out the possibility of pseudoaneurysm formation.
CSF leakage and pseudomeningocele formation may require temporary placement of a lumbar drain or even reoperation and wound revision. Postoperative meningitis may also occur, either as a consequence of contamination during surgery or as a complication of postoperative CSF fistula.

**Conclusion**

The far lateral approach along with its several variants is a useful route to a number of lesions located ventrolateral to the brainstem and the upper cervical cord, offering a suitable exposure with no or minimal retraction of such important neurovascular structures. Variations on this theme can be tailored to the nature and location that the lesion requires. The choice of the approach variant depends on the pathological feature and the degree of exposure required for an effective surgical treatment. Bone removal should be quantified for every single lesion. The approach should always be limited to the less aggressive steps and it still will be usually able to achieve enough exposure and surgical space. Therefore, the deep understanding of the microsurgical anatomy through the far lateral approach can improve the curative effectiveness of foramen magnum diseases’ surgical techniques and may reduce postoperative complications.

**References**


The extreme lateral approach is a skull-based procedure used to target a variety of pathologies at the craniovertebral junction (CVJ). It is a surgical technique that has evolved from classical posterolateral approaches to treat aneurysms as described by Roberto Heros and from Bernard George’s approach to vertebral artery lesions. Over the years, this approach has taken on many names. However, it is essentially one approach in which there are variations in the patient’s position, the skin incision, the muscle reflection, the transposition of the vertebral artery complex, the amount of condyle drilling, and the type of craniotomy.

Surgical approaches to the craniovertebral junction have historically been one of the most challenging and highly morbid procedures. The challenge of obtaining adequate surgical exposure in a small operative corridor coupled with the anatomical depth of surgery has historically limited many attempts at tumor resection. Tumors that are located anterior to the cervicomedullary junction and the upper cervical spine may be inadequately accessed through a purely anterior or posterior approach. The traditional anterior neurosurgical approaches can result in a more direct route to craniocervical lesions, but exposure is usually limited, resulting in inadequate tumor removal.

The anterior aspect of the CVJ consists of the lower clivus (beginning at the level of the jugular foramen) and C1. Many advances in the surgical approaches, imaging technologies, and better understanding of the surgical anatomy have improved the management of these tumors. The surgical approaches are influenced by several factors. These include tumor type, size, and precise location, as well as the experience and individual preferences of the surgeon. The approaches can be broadly divided into anterior, anterolateral, and posterolateral types, depending on the main trajectory of the approach. All of these approaches are useful, either individually or in combination with each other, as dictated by the individual case at hand. It is therefore important for the surgeon to be well versed with the entire spectrum of available approaches to tackle the specific problem. The focus of this chapter is to discuss the extreme lateral approach in skull base surgery and its indications.
masses of C1 articulate with the occipital condyles. The shape of the lateral masses of C1 allows for movement through their cup-shaped contour. This design primarily allows for flexion and extension. The transverse process of the C1 level projects anteriorly and laterally. Many important muscles attach to this transverse process, which include the superior and inferior oblique, levator scapulae, and rectus capitis. The longus capitis attaches to the lower clivus and the rectus capitis anterior to the front of the occipital condyle. The C1–C2 articular surfaces are flat to facilitate translational motion and, more importantly, rotational motion. Approximately 50% of axial rotation in the cervical spine occurs at the level of C1–C2. The adaptation that evolved to prevent AP and lateral translation of C1 on C2 is the odontoid peg, or the dens, and its associated ligaments. The cruciate, alar, and apical ligaments maintain the stability of the odontoid to the occiput. The alar ligament of the odontoid process attaches to the medial tubercle of the occipital condyle. The ligaments maintaining the stability between the odontoid and C1 are the transverse atlantal, atlantoalar, and atlantodental. This design is unique in the spinal bony architecture.

CN IX and X arise as multiple fascicles from the lateral aspect of the medulla, and CN XI arises from the medulla and the upper cervical spinal cord, traveling upward behind the dentate ligament. All then proceed to enter the jugular foramen medial to the jugular bulb. Anterior and superior to the occipital condyle is the hypoglossal canal, which transmits the hypoglossal nerve (CN XII). The third segment of the vertebral artery begins at the foramen transversarium of C2 and ascends through the C1 transverse process. Surrounded by a rich venous plexus, the vertebral artery proceeds above the arch of C1 and crosses medially behind the articular capsule of the atlanto-occipital joint to enter the dura in an oblique manner, traveling upward and anteriorly (Fig. 43.1). This segment of the vertebral artery above C1 runs in the anatomical suboccipital triangle. The triangle is delineated by three muscles: the superior oblique, extending from the occipital bone to the C1 transverse process; the inferior oblique, extending from the C1 transverse process to the C2 spine; and the rectus capitis posterior major, extending from the C2 transverse process to the occipital bone.

**Indications and Pathology**

The extreme lateral approach provides excellent exposure of the anterior foramen magnum (lower clivus below the jugular foramen down to C2) and allows for proximal control of the vertebral artery. The approach has evolved over recent decades from the initial lateral modifications of the suboccipital approach. The extreme lateral approach, as described by Sen and Sekhar and also by Bertalanffy and Seeger, incorporates medial mobilization of the vertebral artery from the dural entrance point to C2, and resection of the occipital condyle and the lateral mass of C1 (either partially or totally). This approach is suitable for both intra- and extradural lesions, allows control of the vertebral artery, permits dissection of the brainstem–tumor interface tangentially along this plane, and avoids traversing contaminated spaces. The extent of soft tissue dissection, bony resection, and vertebral artery manipulation is tailored to the type and extent of the tumor.

Specific preoperative studies are critical in surgical planning. Magnetic resonance venography/angiography (MRV/MRA) and computed tomography (CT) are routinely done. MRA/MRV provides information regarding the regional vascular anatomy, including the patency and dominance of the vertebral arteries and dural sinuses. We use only angiography for unresolved or aberrant findings on MRA/MRV.

**General Perioperative Clinical Evaluation and Neurophysiological Monitoring**

Once the decision has been made to proceed with surgery, all patients should undergo a detailed study of their neurological function, independent of their clinical neurological exam. The critical location of these tumors to vital neurological and vascular structures requires a more detailed cranial nerve study. Ophthalmological, audiometric, and electromyographic (EMG) studies can be supplemented to the preoperative assessment of the patient’s baseline function. Preoperative evaluation of CN IX through XII is especially important.

The importance of preserving lower cranial nerve function is crucial in the intraoperative procedure. We employ intraoperative neurophysiological monitoring to prevent injury to the cranial nerves. Somatosensory evoked potentials, motor evoked potentials, and brainstem
auditory evoked potentials are monitored bilaterally.\(^2\)
EMG monitoring of the vagus, accessory, and hypoglos-
sal nerves are done on the ipsilateral side (or bilaterally
if necessary). The vagus nerve can be monitored with an
EMG-electrode endotracheal tube or by a laryngeal surface
electrode that is placed after intubation. The accessory
and hypoglossal nerves are monitored directly with elec-
trodes placed on the ipsilateral trapezius muscle and the
inferior aspect of the tongue, respectively. The use of in-
traoperative neurophysiological monitoring avoids the use
of muscle relaxants during many stages of the operation.
We employ 24 hours of broad-spectrum intravenous anti-
biotics and perioperative steroids at the time of surgery.

**Description of Operative Technique**

The positioning of the patient is based on the location
and pathology of the tumor. A straight lateral position is
best for purely intradural tumors. For extradural tumors,
the supine position with the head turned is preferred,
especially when more temporal bone work is required.
Forward neck flexion is avoided to prevent compromise
of the brainstem by the tumor. A detailed understanding
of the suboccipital muscles (e.g., sternocleidomastoid,
splenius capitis, semispinalis and longissimus capitis, su-
peior and inferior obliques, and recti capitis major and
minor) is crucial in the procedure, much more so than in
a standard posterior midline approach.\(^5,6\) We use either
the inverted U-shaped incision or the C-shaped curvi-
linear incision. In the U-shaped incision, the skin flap is
reflected inferiorly with all the muscles attached as one
large flap. We have used the C-shaped incision more com-
monly (for extradural tumors), which begins in the ret-
roauricular area and extends into the cervical area along
the skin crease. The muscles of the posterior cervical tri-
gle are dissected in layers and detached from the cra-
nium and C1 transverse process, then reflected caudally.
The reasons for this detailed anatomical muscle dissec-
tion are the following:

1. Through this dissection, a wide, shallow exposure
   is created to better visualize the anterior portion of
   the foramen magnum.
2. It allows the surgeon to enter the foramen magnum
   from both caudad to cephalad directions, thereby
   minimizing trauma to the lower cranial nerves.
3. It allows identification and isolation of the extradu-
   ral segment of the vertebral artery above C1.
4. It reduces the depth of the surgical field.

The extreme lateral surgical approach essentially
involves the following general techniques. First, a C1
hemilaminectomy and suboccipital craniectomy (small,
extending 3 cm from the back of the occipital condyle)
is performed with unroofing of the sigmoid sinus. Next,
the extent of the drilling of the occipital condyle and C1
transverse process is determined by the type of tumor,
whether intradural or extradural.

Exposure and mobilization of the vertebral artery (in
the suboccipital triangle) above C1 or C2 depends on the
specific resection plan. The vertebral artery is surrounded
by a periosteal sheath, which is continuous and encloses
a venous plexus that is in communication with the epi-
dural plexus. Feeling for pulsation may be difficult; there-
fore, identification is best done by anatomical landmarks.
Between C1 and C2, the vertebral artery can be located
posterior to the upper attachment of the levator scapulae
and caudal to the inferior border of the inferior oblique
muscles. After emerging on the superior surface of the
C1 foramen, the vertebral artery turns posteriorly along
the upper surface of the C1 posterior arch, where it lies
deep to the superior oblique muscle. Keeping the head
and neck neutral is important during the dissection of
the vertebral artery. This is because the artery is redu-
dant above C1 and between C2 and C1. Once the foramen
transversarium of C1 is identified, it is rongeured open,
and the vertebral artery is displaced out of C1. This mobil-
ization is what facilitates anterior exposure of the C1–C2
region as well, particularly in extradural pathology.

**Intradural Tumors**

The prototypical intradural tumor that requires an ex-
treme lateral approach is a craniocebral meningioma.
In approaching meningiomas, the dura must be com-
pletely opened around the vertebral artery entry area,
so that the vessel may be mobilized posteriorly during
tumor resection. The condylar emissary vein connects
the jugular bulb with the epidural venous plexus sur-
rounding the vertebral artery.\(^16,22\) If a tumor extends into
the jugular bulb, the dominance and patency of the ve-
 nous drainage must be determined before entering the
bulb. If the jugular bulb is occluded (as is more often the
case in an extradural tumor), the sigmoid sinus is ligated
above and the internal jugular vein below, and the bulb
is opened directly to remove tumor from CN IX, X, and
XI. For purely intradural tumors, it is never necessary to
mobilize the vertebral artery at the C1 transverse fora-
men. After opening the dura, the arachnoid membrane of
the cerebellomedullary cistern is opened, and the intra-
dural tumor is often easily visualized.\(^13\) Resection of the
C1 intradural rootlets and the upper dentate ligaments
facilitates tumor exposure and helps in resectioning
(Figs. 43.2 and 43.3).

**Extradural Tumors**

For extradural tumors, the occipital condyle resection ex-
tends more anteriorly to excise the entire condyle. The
formal transcondylar approach for extradural tumors
allows resection of tumors from the following areas:
bony involvement inferior to the labyrinth, jugular fora-
men, hypoglossal canal, occipital condyle, lateral mass,
 anterior arch of C1, anterior part of C2, and lower clivus
up to the midline. If the internal carotid artery and the
jugular foramen are identified and dissected all the way up from the neck to the cranial base, then the ipsilateral retropharyngeal area is accessible as well (Figs. 43.4 and 43.5). Watertight dural closure is difficult, and a combination of dura graft and fibrin sealant is often used, with the possible addition of local muscle or fat.

Condylar Resection in the Extreme Lateral Approach

There have been differing opinions about the need for condylar resection in the extreme lateral approach for intradural tumors. Whereas many authors, such as Nanda et al. argue against a need for condylar resection, others,
such as Spetzler et al, believe that a partial condylar resection adds to the lateral view of anterior lesions, thus obviating the need for brain retraction.\textsuperscript{25,26} We believe that condylar resection is based on the need to mobilize the vertebral artery during resection of the anterior part of the tumor with its dural attachment. Releasing the vertebral artery from its dural tether and retracting it back creates a better work space with unimpended access to tumor dissection in an inferior-to-superior trajectory. This maneuver also reduces trauma to the lower cranial nerves. If the foramen magnum is deep, the tumor small, or the dural attachment hard to reach, condylar drilling is also helpful. In extradural tumors, condylar drilling is truly part of the tumor resection; therefore, the extent of drilling is dictated by tumor involvement.

If more than a partial resection of the occipital condyle is performed, stabilization will be needed. This occipitocervical fusion and instrumentation can be performed at the same sitting or as a different stage. Complications to this approach are pseudomeningocele, cerebrospinal fluid leakage, lower cranial nerve injury (especially involving CN IX, X, and XI), meningitis, vertebral artery injury/thrombosis, and unanticipated occipitocervical instability.

\textbf{Common Tumors at the Craniovertebral Junction}

The pathological differential diagnosis of tumors in the craniovertebral junction can be broken into intradural and extradural lesions. In many cases, the dura is involved by the extradural tumor, and/or a portion of the tumor is intradural. The differential diagnosis of extra-axial foramen magnum tumors include meningiomas as the most common, followed by chordomas, neurilemmomas, epidermoids, chondromas, chondrosarcomas, and metastatic lesions.\textsuperscript{27} Although meningiomas of the foramen magnum are the most common intradural tumor at this location, they are still very rare lesions when compared with all intracranial tumors. Lesions in the foramen magnum account for only 0.3% to 3.2% of all meningiomas and between 4% and 15% of all posterior fossa meningiomas combined.\textsuperscript{5,28} Benign soft tissue tumors include meningiomas, schwannomas of the lower cranial nerves, and neurofibromas. Meningiomas may cause hyperostosis and bone remodeling that is best visualized on CT bone window imaging. On MRI, meningiomas are isointense on T1 and T2 and enhance with gadolinium.

The radiological work-up of chordomas and chondrosarcomas consists of fine-cut CT imaging in which there are demonstrated variations in bone density and bone erosion with an outline of a destructive expansile lesion.\textsuperscript{5,29} Areas of punctate calcification can also be present in up to 50% of studies. On MRI, chordomas are low or intermediate in signal intensity on T1-weighted sequences and high signal on T2-weighted sequences. Also on MRI, areas of signal loss may be associated with calcification. The clivus can show areas of heterogeneous marrow signal. With contrast, chordomas can enhance variably, from low to high brightness. Differentiating chordomas from chondrosarcomas is difficult, though chordomas are traditionally described as occurring more in the midline compared with chondrosarcomas.

\textbf{Conclusion}

The advancement in neurosurgical techniques, detailed anatomical understanding, and anesthetic protocols, as well as the recent developments in skull base surgery, have dramatically reduced the once very high mortality associated with surgery for tumors located in the anterior foramen magnum. However, despite these advances, these tumors continue to remain some of the most challenging to treat in all of neurosurgery. Although the traditional posterior midline suboccipital approach is safe and useful in most tumors in the anterior foramen magnum, the additional benefits provided by the extreme lateral transcondylar approach far outweigh almost any other approach. In the extreme lateral approach, there exists a wider surgical area of exposure of the tumor–cord/brainstem interface, as well as the ventral dura, without the need for retraction of the neural structures. The value of the condylectomy alone both shortens the distance to the anterior foramen magnum and widens the lateral exposure for a better surgical angle for resection.\textsuperscript{17} Control of the involved vertebral artery is made possible, and devascularization of the tumor can be accomplished early in the operation. Although variations of the extreme lateral procedure already exist and have been reported, the final

\textit{Text continues on page 496}
Fig. 43.6a, b Axial T1-weighted MRI with contrast demonstrating an intradural meningioma (arrow) of the inferior clivus located anteriorly.

Fig. 43.7 Axial T2-weighted MRI showing the intradural meningioma (arrow) of the clivus with deformation of the brainstem.

Fig. 43.8 Postoperative T1-weighted MRI with contrast showing removal of the meningioma through the extreme lateral approach.
Fig. 43.9 Postoperative T1-weighted MRI with contrast showing removal of the inferior pole of the meningioma with no anterior residual tumor and improved brainstem compression via the extreme lateral approach.

Case Illustration

A 16-year-old male patient presented with bilateral upper extremity numbness and dysesthesia. MRI was significant for a cervicomedullary chordoma with involvement of the right vertebral artery (Fig. 43.10). There was also involvement of the retropharyngeal space with compromise of the pharyngeal cavity (Fig. 43.11). The right occipital condyle was already eroded by the tumor, as was the lower clivus. The patient underwent an extreme lateral
approach with occipital condylectomy and removal of the retropharyngeal portion of the tumor (Fig. 43.12). During the surgery, both vertebral arteries were dissected free and visualized (Fig. 43.13). The reconstruction was done with titanium cranioplasty, abdominal fat graft, and placement of a spinal drain. Postoperatively, the patient developed a right vertebral artery thrombosis, which required direct thromboembolectomy and arteriotomy with primary repair (Figs. 43.14 and 43.15). Because of the occipitocervical instability from the condylectomy, the patient also underwent occipitocervical fusion (Fig. 43.16). The patient tolerated these three surgeries well and was neurologically intact. Postoperative MRI after 2 months showed no residual tumor. No radiation was administered.
approach and its various modifications should always be tailored to the unique tumor presentation and the specific goals of surgery in the treatment of the individual patient.10

References

Craniovertebral junction (CVJ) anomalies occur in a small percentage of genetic and metabolic disorders usually as a result of hypoplasia or maldevelopment of the dens or ligamentous laxity. Most of them manifest in early childhood, and management of these conditions is challenging due to the associated systemic manifestations that influence their anesthetic management as well as long-term outcomes.

The principles of management in these patients follow the same algorithm as for any CVJ anomaly. The basic aims of management are (1) decompression of the cervico-medullary junction, (2) restoration of alignment at the CVJ, and (3) stabilization of the CVJ. Prior to achieving these goals, the diagnosis of the underlying primary pathology has to be ascertained through a detailed clinical and radiological examination complemented by molecular diagnostic tests when indicated. It is well recognized that the overall philosophy of managing CVJ anomalies in children with these inherited pathologies should be directed by their systemic manifestations and their ultimate impact on the long-term prognosis of the patient. A team consisting of a neurosurgeon, pediatrician, anesthetist, and physiatrist, each of whom understands the systemic and neurological manifestations of these diseases, should ideally handle the management of these conditions. A brief outline of the clinical and radiological evaluation recommended in these patients is given in Table 44.1. A list of metabolic and genetic disorders, in which CVJ anomalies can manifest, is given in Table 44.2.

In this chapter, we discuss the presentation, management, and outcome of CVJ anomalies in some of the more commonly encountered metabolic and genetic disorders.

### Down Syndrome

Of all the metabolic and genetic disorders associated with CVJ anomalies, those seen in patients with Down syndrome are the best characterized (Fig. 44.1). Down syndrome results from the most common chromosomal abnormality in humans, trisomy 21, occurring in ~1 in 700 births. It is well recognized by the clinical features of mongoloid facies, hypotonia, ligamentous laxity, mental retardation, and transverse palmar creases. Spitzer et al. is credited with the first description of the CVJ manifestations of Down syndrome in 1961, with a report on occipito-atlantal dislocations in nine patients. In 1965, Tishler and Martel reported atlantoaxial dislocation in Down syndrome, which is the most common anomaly seen in the CVJ of these patients.

### Natural History

Pueschel et al. reported the occurrence of atlantoaxial instability in 14.6% of 404 patients with Down syndrome who underwent dynamic cervical spine radiograph studies. Of these, only six (close to 1%) were symptomatic. Although the authors demonstrated an increase in the atlantodental interval in serial radiographs in a subgroup of patients at a follow-up of 3 to 10 years, none of them developed clinical manifestations. Burke et al. reported a progressive increase in the predental space in 6 out of 32 patients followed up over 13 years. Morton et al., however, reported on 67 patients with Down syndrome followed up for ~5 years and found that none developed de novo atlantoaxial instability. In other reviews, 7 to 40% of patients with Down syndrome have been reported to have atlantoaxial instability, with <1% being symptomatic. Ferguson et al. reported no statistical difference between

**Table 44.1 Evaluation of a patient with suspected skeletal dysplasia and craniovertebral junction anomaly**

<table>
<thead>
<tr>
<th><strong>Clinical evaluation</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Skull</td>
</tr>
<tr>
<td>Facial abnormalities, including oral cavity and palate</td>
</tr>
<tr>
<td>Shoulder deformity</td>
</tr>
<tr>
<td>Deformity of hip/knee</td>
</tr>
<tr>
<td>Deformities of the chest wall</td>
</tr>
<tr>
<td>Toes and fingers (syndactyly)</td>
</tr>
<tr>
<td>Deformities of the thoracic and lumbar spine</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Radiological evaluation</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Periodic screening radiographs of the craniovertebral junction</td>
</tr>
<tr>
<td>X-rays of the appendicular skeleton</td>
</tr>
<tr>
<td>X-rays of the cervical, thoracic, and lumbar spines</td>
</tr>
</tbody>
</table>
### Table 44.2  Inherited and metabolic disorders that can result in craniovertebral junction anomalies

<table>
<thead>
<tr>
<th>Metabolic Disorder</th>
<th>Metabolic/Enzyme Defect</th>
<th>Chromosomal Locus</th>
<th>Inheritance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morquio-Brailsford syndrome</td>
<td>N-acetylgalactosamine-6-sulfatase β-galactosidase</td>
<td>16q24.3 3p21-p14.2</td>
<td>Autosomal recessive</td>
</tr>
<tr>
<td>Nonkeratosulfate-excreting mucopolysaccharidosis</td>
<td>Defects in metabolism resulting in accumulation of chondroitin sulfate, dermatan sulfate, heparan sulfate</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pseudoachondroplasia</td>
<td></td>
<td>19</td>
<td>Autosomal dominant (type 1 and II), autosomal recessive (types III and IV)</td>
</tr>
<tr>
<td>Spondylometaphyseal dysplasia</td>
<td>Generic group of disorders</td>
<td></td>
<td>Autosomal dominant/recessive</td>
</tr>
<tr>
<td>Spondyloepiphyseal dysplasia congenita</td>
<td>Type II collagen</td>
<td>12 (gene locus Col2A1) Xp22</td>
<td>Autosomal dominant, sporadic</td>
</tr>
<tr>
<td>Spondyloepiphyseal dysplasia tarda</td>
<td></td>
<td></td>
<td>X-linked</td>
</tr>
<tr>
<td>Down syndrome</td>
<td></td>
<td>Trisomy 21</td>
<td>Autosomal dominant, sporadic mutation</td>
</tr>
<tr>
<td>Achondroplasia</td>
<td></td>
<td>4p16.3</td>
<td>Autosomal dominant (types I and IV), autosomal recessive (types II and III)</td>
</tr>
<tr>
<td>Osteogenesis imperfecta</td>
<td>Mutations in type I collagen</td>
<td></td>
<td>Autosomal dominant or recessive</td>
</tr>
<tr>
<td>Larsen syndrome</td>
<td>Connective tissue defect</td>
<td></td>
<td>Autosomal dominant</td>
</tr>
<tr>
<td>Diastrophic dysplasia</td>
<td>Probable type II collagen defect</td>
<td>5q31–34</td>
<td>Autosomal recessive</td>
</tr>
<tr>
<td>Metatropic dysplasia</td>
<td>Type II collagen defect</td>
<td></td>
<td>Autosomal dominant</td>
</tr>
<tr>
<td>Kniest dysplasia</td>
<td></td>
<td></td>
<td>Autosomal dominant</td>
</tr>
</tbody>
</table>


Fig. 44.1a–d  Plain lateral cervical spine radiographs of a 9-year-old boy with Down syndrome who presented with progressive spastic quadriparesis for 1 year with recurrent falls. He required support for ambulation and was Nurick grade 4 at initial presentation.

a Preoperative radiograph showing evidence of an atlantoaxial dislocation.
b Immediate postoperative radiograph following reduction of subluxation, excision of the posterior arch of C1, and occipitocervical fusion using a titanium loop and sublaminar wires with autologous rib graft showing the implant in situ. The patient was immobilized in a halo vest for 6 months.
c Follow-up radiograph at 1 year showing the implant in situ with evidence of bony union. There is minimal loss of the reduction obtained at the time of surgery. The patient had improved in his neurological status and was independently ambulant with a spastic gait (Nurick grade 2).
d Radiograph following removal of the implant and sublaminar wires at 1 year demonstrating bone formation between the occiput and C3.
the incidence of symptomatic myelopathy in the subluxator and nonsubluxator groups. Menezes\textsuperscript{8} commented that in his experience, the presence of atlantoaxial disease in children with Down syndrome does not have a benign relationship to neurological function.

The Committee on Sports Medicine of the American Academy of Pediatrics\textsuperscript{15} recommended routine screening of patients with Down syndrome with cervical spine radiographs prior to their participation in Special Olympics. The committee further recommended no routine follow-up radiographs in patients who do not have atlantoaxial instability in the initial radiograph. Most authors agree that asymptomatic patients with Down syndrome and atlantoaxial dislocation may be followed up clinically and radiologically.

\section*{Radiological Features}

The common radiological abnormalities of the CVJ in Down syndrome are atlantoaxial instability, occipitoatlantoaxial instability, rotary atlantoaxial or occipitoatlantal dislocation, basilar invagination, os odontoideum, and bifid or hypoplastic atlantal arches. The generalized ligamentous laxity associated with Down syndrome results in development of hypermobility that is in part responsible for development of some of the bony anomalies, such as os odontoideum.\textsuperscript{8–10}

\section*{Management}

The clinical presentation in these patients is akin to those with any CVJ anomaly, with neck pain, torticollis, and features of cervical cord compression. Sudden onset of neurological worsening associated with trauma, intubation procedures, or associated upper respiratory infection has also been documented.\textsuperscript{8}

The goal of treatment in symptomatic patients is reduction of the instability, neural decompression (by transoral decompression in irreducible atlantoaxial instability), and posterior stabilization of the CVJ. Nader-Sepahi et al.\textsuperscript{16} highlighted the importance of recognizing occipitoatlantal instability coexisting with atlantoaxial instability and recommended occipitocervical fusion in such cases. Incorporation of the occiput into the fusion has also been recommended in cases where there is an abnormality of the atlantal arch or where the atlantal arch has been removed to achieve neural decompression. Menezes\textsuperscript{8} commented that C1–C2 transarticular fixation may be used in isolated atlantoaxial arthrodesis, supplemented with bilateral interlaminar fusion.

Taggard et al.\textsuperscript{17} and Menezes\textsuperscript{8} demonstrated good results with bony fusion in 95% of 64 patients treated over 17 years. Nader-Sepahi et al.\textsuperscript{16} reported successful fusion in 7 of 12 patients after the first operation but ultimately achieved 100% fusion with repeat surgeries. However, others have reported less encouraging results. In a review of complications of upper cervical spine fusion in children, Smith et al.\textsuperscript{18} identified Down syndrome as one of the risk factors predisposing to incomplete fusion. Segal et al.\textsuperscript{19} reported that almost all of their patients with Down syndrome who underwent posterior arthrodesis had some complication, ranging from wound infection, incomplete reduction of the atlantoaxial dislocation, instability of the adjacent motion segment, to neurological deterioration. In their series of 10 patients, only 4 developed bony fusion. They commented that the following reasons could be responsible for graft resorption in patients with Down syndrome:

1. Patients with this syndrome have immune deficiencies resulting in decreased lymphocyte and monocyte function and may not be able to mount an initial inflammatory response that is essential for bone graft absorption.
2. Intrinsic defects in collagen could contribute to poor fusion.
3. Fibroblasts cultured from patients with Down syndrome have been documented to have increased expression of the β-interferon receptors (the genes that encode this receptor are located in chromosome 21), and this could enhance fibroblast activity, promoting release of collagenase and protease.\textsuperscript{20} This increased activity could contribute to graft resorption.

Taggard et al.\textsuperscript{17} found that inadequate postoperative immobilization, failure to recognize and appropriately treat ventral pathology, lack of consideration of bony anomalies, and inadequate bone grafting are the main causes of incomplete fusion. Halo vest immobilization would be ideal but difficult to maintain in these patients due to pin site morbidities in young children and the presence of mental retardation in several of these patients. Figure 44.1 illustrates the radiographs in a patient with Down syndrome and symptomatic atlantoaxial dislocation.

\section*{Morquio-Brailsford Syndrome}

Morquio-Brailsford syndrome, also known as type IV mucopolysaccharidosis (MPS), is a congenital connective tissue disorder caused by reduced activity of either N-acetylgalactosamine-6-sulfatase or β-galactosidase.\textsuperscript{1} The phenotype is a dysplastic dwarf, but less severe forms occur. Although most patients die in late childhood or early adulthood from neurological disability or valvular or myocardial dysfunction, longer survival has been reported.

\section*{Radiological Features}

Neurological disability due to the involvement of the CVJ has been reported by several authors.\textsuperscript{2–6} Dysplasia of the odontoid and anterior atlantoaxial subluxation is considered to be the main cause of spinal cord compression (Fig. 44.2). Thickening of the posterior longitudinal ligament, invagination of the posterior arch of the atlas into the foramen magnum, and stenosis of the foramen...
magnum have also been reported. Hughes et al. reported abnormal ossification of the dens with the presence of a soft tissue mass that was intermediate to low signal on T1- and T2-weighted images. Indentation by the posterior arch of the atlas was a contributing factor in all cases where compression was seen.

Stevens et al. reported on a series of 13 patients with type IV MPS with dynamic computed cervical myelograms. They observed midline ossification defects in the anterior and posterior arches of the atlas, presence of a dysplastic dens, posterior atlantoaxial subluxation (defined as present when any part of the anterior arch of the atlas overrode the body of the axis) in 11 cases and widened joint spaces at the atlantoaxial lateral mass articulation and atlanto-occipital articulation. They also observed the presence of thickened anterior extradural soft tissues in most patients. The soft tissue thickening indented the thecal sac, resulting in its compression. The site of maximal compression was opposite the basal part of the dens, and the compressing agents were soft tissue thickening anteriorly and unossified part of the neural arch of the atlas posteriorly.

**Pathophysiology**

According to Stevens et al., the underlying instability and hypermobility from ligament laxity creates abnormal shearing stresses at the cartilaginous stage of development. This in turn interferes with the normal ossification process, which extends from the centrum of C1 into the portion projecting above the articular surfaces of C2. Eventually, the ossification centers separate and the interposition of the transverse ligament and hypermobility results in established non-union. Hypermobility at the atlantoaxial complex causes repeated minor trauma, fracturing of the odontoid process, and its subsequent development into an os odontoideum, once it is pulled up by the apical and alar ligaments. It was also observed that following the occipitocervical fusion, the extradural tissue ossified, indicating that it probably represents unossified cartilage.

**Management**

It has been suggested that the optimal time for an elective MRI in these patients is between 3 and 8 years of age, as this is when skeletal development is usually complete, and little or no axial growth occurs thereafter. Based on a review of literature, Stevens et al. proposed that a reduction in measurable diameters of the spinal cord approaching 50% should be considered as an indication for surgery, regardless of the clinical status.

Bethem et al. suggested observation in patients who are asymptomatic. The management strategies in literature include external bracing or posterior occipitocervical fusion to stabilize the atlantoaxial subluxation and allow normal ossification of the dens to occur. Stevens et al. recommended performing posterior occipitocervical fusion. In their opinion, surgery should be considered only in those patients with marked thickening of the extradural soft tissues. In their experience, all patients tolerated the surgery well but had only limited improvement in clinical status, with no long-term follow-up reported. They opined that many patients with atlantoaxial subluxation survive into adult life without major spinal cord compression; hence, this alone should not be considered as a criterion for intervention.

**Achondroplasia**

Achondroplasia is a congenital, skeletal dysplasia that has been classified as an osteochondrodysplasia, with an incidence of 0.03% to 0.05% of live births. It is the most common form of short-limbed dwarfism. Familial cases are of autosomal dominant inheritance, with a high degree of mortality in infancy among homozygous chondroplasts (patients with two affected parents). Most reported cases are of heterozygous achondroplasts. Mental development is generally normal, although growth is slow and ceases early.

**Pathophysiology**

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**Pathophysiology**

Bones formed from cartilage are primarily affected. Short, thick, tubular bones are formed due to a decreased rate of enchondral ossification with normal periosteal bone
formation. Shortened extremities (rhizomelia), proportionally large trunk and head, frontal bossing, midfacial recession with coarse facial features, and exaggerated lumbar lordosis are common manifestations.\textsuperscript{22,23}

The neurological manifestations of achondroplasia are related to the abnormalities of the cranial base, compression of neurovascular structures, and hydrocephalus. Defects in enchondral bone formation in the cranial base result in a shortened basiocciput, shallow posterior fossa, shortened clivus, and abnormal basal spinal relationships. Foramen magnum stenosis is common and results from a combination of defective enchondral bone formation and abnormal placement and premature fusion of the basal synchondroses.\textsuperscript{22,24} This results in quadriplegia, feeding problems, respiratory difficulties,\textsuperscript{21,23} and sleep apnea.\textsuperscript{25} The presence of hyperreflexia is a common neurologic finding. Relative hypotonia, when present, is an indicator for surgical decompression.\textsuperscript{21}

### Radiological Features

The reported radiographic abnormalities of the CVJ in achondroplasia include foramen magnum stenosis with a teardrop shape and obliteration of the subarachnoid space. Ventral and dorsal cervicomedullary compression with or without basilar invagination and anterior indentation of the pons with upward displacement of the brainstem has been described. Other common manifestations are Chiari malformation, odontoid dysplasia, shortening of the clivus with thickening of the posterior rim of the foramen magnum, atlantoaxial subluxation, vertebrobasilar insufficiency, and decrease in the basal angle.\textsuperscript{21,22,26–28}

### Management

In a review of 186 patients with achondroplasia over a 13-year period, 6 patients were found to have symptoms or signs of cervicomedullary compression.\textsuperscript{21} Although surveillance of young patients with achondroplasia for the development of symptomatic cervicomedullary compression is essential, decompression of the CVJ is the recommended treatment in symptomatic patients. Ryken and Menezes\textsuperscript{21} recommended that patients should be evaluated for other causes of respiratory compromise, such as chest wall deformities and upper airway abnormalities.

The surgical decompression involves removal of the posterior rim of the foramen magnum up to the medial aspect of the occipital condyles bilaterally and excision of the posterior arch of the atlas. At surgery, a thickened fibrous band has been noted posteriorly, indenting the thecal sac. Dura may be opened after ultrasonographic assessment for the presence of cerebrospinal fluid (CSF) pulsations dorsal and ventral to the upper cervical spinal cord, following bone removal.\textsuperscript{21,22,25}

Bagley\textsuperscript{21} et al. reported a series of 42 symptomatic achondroplastic dwarfs with cervicomedullary compression. They observed a horizontal orientation of the foramen magnum rim with a thickened posterior lip and performed duraplasty in cases where CSF was not visualized on the ultrasound ventral and dorsal to the cervicomedullary junction following bony decompression. In five of these patients, there was evidence of symptomatic restenosis at follow-up that improved following repeat decompression. Arya\textsuperscript{25} et al. did not observe any restenosis at 19 months’ follow-up in 15 patients who underwent cervicomedullary decompression.

Several authors\textsuperscript{21,22,25} have commented on the presence of hydrocephalus secondary to the altered CSF dynamics and venous outflow obstruction due to the stenosis at the cranial base that could complicate the decompression surgery. While symptomatic patients underwent ventriculoperitoneal shunt, others were managed with an external ventricular drain placed at the time of the CVJ decompression surgery that could be removed in most patients.

### Osteogenesis Imperfecta

The term osteogenesis imperfecta (OI) covers a heterogeneous group of heritable bone disorders characterized by extreme bone fragility and an increased inclination to fracture.\textsuperscript{29,30} Sawin and Menezes\textsuperscript{31} reviewed the pathogenesis of basilar impression that occurs in a small subset of patients with OI. Recurrent microfractures in the region of the foramen magnum result in progressive infolding of the posterior skull base, thereby permitting upward translation of the rostral cervical spine into the posterior fossa. The authors suggested that ventral decompression followed by occipitocervical fusion resulted in functional improvement in 92% of patients. However, over a mean follow-up period of 5.9 years, basilar invagination progressed in 80% of their 25 patients, despite achieving solid bony fusion. Symptomatic recurrence occurred in ~30% of the patients. This was attributed to the further infolding of the squamo-occipital and petrous temporal bones. Patients who deteriorated clinically were managed with M-MLV. Ibrahim and Crocker\textsuperscript{22} reported long-term outcomes following ventral decompression and occipitocervical fusion in patients with OI and basilar impression. Fifteen percent of their patients developed symptomatic recurrence in the long term. In their analysis, the mean age of their patients was ~16 years older than that reported by Sawin and Menezes;\textsuperscript{31} this could explain the decreased incidence of recurrence. They also observed a high rate of postoperative upper airway problems and aspiration-related chest infection. The authors also commented that with the advent of bisphosphonates and their use in the treatment of OI, there was a decline in the number of cases with basilar impression and OI referred to their center.\textsuperscript{32}

### Other Causes

A large number of patients with dwarfism with congenital atlantoaxial dislocations have been described.\textsuperscript{2,4,26,30} Along with achondroplasia and Morquio syndrome, as discussed
above, pseudoachondroplasia, Scott’s syndrome, spondylometaphyseal dysplasia, congenital spondyloepiphyseal dysplasia (Fig. 44.3), cartilage hair dysplasia, diastrophic dwarfism, chondrodystrophic dwarfism, chondrodytrophic calcificans congenita, metatrophic dwarfism, and Kniest syndrome have been found to be associated with atlantoaxial dislocation. Maldevelopment of the odontoid has been observed in most of these cases.

Larsen syndrome is a rare inherited disorder of connective tissue formation associated with midcervical kyphosis and anteroposterior dissociation of the cervical spine. Atlantoaxial instability is a rare finding in this syndrome and may be observed with other abnormalities of the upper cervical spine, including occipitalization of the atlas and basilar impression.

Renal osteodystrophy has been reported to cause cervical myelopathy due to destructive spondyloarthropathy. In a few cases, involvement of the CVJ has been reported. The cause has been attributed to amyloid deposition as well as hyperparathyroidism.

### Anesthetic Management

There are several associated anomalies that can make anesthetic management of patients with CVJ anomalies due to inherited or metabolic disorders difficult. Aggravation of the cervicomedullary compression during intubation has been reported. The disorders could affect the larynx and the rib cage, necessitating precautions during intubation and maintaining ventilation during anesthesia. Patients with Larsen syndrome could have laryngomalacia, causing stridor in the postoperative period, and may need a tracheostomy. Obstructive sleep apnea has been reported in association with achondroplasia and could pose problems during induction and recovery from anesthesia. These disorders, along with the chronic hypoxic respiratory insufficiency seen in patients with severe quadriplegia resulting from cervicomedullary compression, can make the postoperative management of these patients very difficult. They are prone to developing atelectasis with pneumonia and respiratory distress and could require prophylactic tracheostomy. Excessively lax skin and soft tissues, as well as flexion deformities, could also make intravenous access difficult.

### Conclusion

CVJ anomalies secondary to metabolic and inherited disorders remain a challenge, even for the most experienced neurosurgeon. In addition to the technical expertise and skill required, prudent decision making by the surgical, pediatric, and anesthetic team in planning treatment is mandatory for obtaining a favorable outcome. As has been commented by Sawin and Menezes, all interventions for most of these disorders must be considered palliative until the fundamental molecular anomalies underlying these conditions can be addressed.

### References

Abnormalities of the odontoid process are relatively rare. Although most of the abnormalities of the odontoid process are congenital in nature due to its relatively complex embryological formation, the exact causative etiology of some cases remains unclear. Os odontoideum is among the more frequently encountered anomalies of the odontoid process.

Os odontoideum was first described by Giacomini in 1886. The term refers to an independent bone that lies above the axis and exists apart from a hypoplastic dens.

**Embryology**

The odontoid process is formed from the centrum of the first spinal sclerotome, with the centrum of the second spinal sclerotome forming the body of the axis. The tip of the dens is a derivative of the proatlas. The odontoid process at birth is separated from the body of the axis by a cartilaginous band that represents a vestigial disk. This is referred to as the neurocentral synchondrosis. The band lies below the level of the superior articular facets of the axis. In patients with os odontoideum, the gap between the os and the remaining body of the axis lies above the level of the superior articular facets of the axis. The tip of the dens that forms from the proatlas is not ossified at birth. Instead, it ossifies from a separate ossification center and fuses with the odontoid process by the age of 12 years. When it fails to fuse with the odontoid process, it is termed ossiculum terminale persistens.

**Pathogenesis**

The cause of os odontoideum is unclear. The pathogenesis has been explained by both congenital and traumatic theories. The posttraumatic theory is the more prevailing. Fielding et al. proposed that an unrecognized fracture in the region of the base of the dens or an acute ligamentous injury occurs in childhood with slight separation of the fracture fragments. This is followed by a contraction of the alar ligaments, causing distraction of the fracture fragments, thus pulling the fractured odontoid fragment closer to the occiput. This also disrupts the blood supply of the odontoid process and leads to poor healing and formation of an ossicle, the os odontoideum. The blood supply to the ossicle is taken over by the proximal arterial arcade, which also supplies the anterior arch of the atlas. This could account for the anterior atlantal arch hypertrophy that is seen in these patients. Several authors have shown that an os odontoideum may occur after a bony or ligamentous injury in early childhood, at which time a complete odontoid process has been demonstrated.

McRae believed that os odontoideum had an embryological origin but required the ongoing stresses of life to trigger instability and produce symptoms after childhood.

According to proponents of the congenital theory, os odontoideum results from a failure of fusion of the odontoid process with the body of the axis. This theory is supported by the increased incidence of associated congenital deformities in these patients (e.g., Klippel-Feil syndrome, basilar invagination, and assimilation of the atlas) and also by the increased incidence of os in patients with Down syndrome, multiple epiphyseal dysplasia, and other skeletal dysplasia. However, for this theory to be true, the gap between the os odontoideum and the axis should be located below the level of the superior axis facet, which has never been demonstrated. In addition, in all patients in whom an os odontoideum is present, a neurocentral synchondrosis is nearly always visible. Kirlew et al. proposed that os odontoideum results from an incomplete migration rather than an incomplete fusion, with the defect occurring at the site of migratory arrest. Hence, they said that the location of the defect that occurs at the anatomical base of the dens could be consistent with a congenital etiology.

**Classification**

There are two types of os odontoideum, depending on its position at the craniovertebral junction (CVJ). It is said to be orthotopic if it is present in the place of the normal dens and moves with the axis and atlas. A dystopic os is one that is attached to the clivus and moves with it and the anterior arch of the atlas.

**Indicators of Instability**

In the absence of a united odontoid process, the atlantoaxial movements appear to be supported only by ligaments. Several authors believe that os odontoideum represents
an unstable CVJ, and surgery is indicated irrespective of the clinical presentation. It appears that in cases where the odontoid process is not in continuity with the body of C2, the atlantoaxial joint is inherently unstable.

Radiological evidence of movement of the ossicle and C1 on dynamic films relative to the body of C2 indicates significant instability. The presence of cord signals on magnetic resonance imaging (MRI) in relationship to the superior edge of the body of C2 also indicates instability.

In most cases, the instability of the atlantoaxial joint is observed on flexion of the head (anterior subluxation). In rarer cases, instability may be observed on extension of the head (posterior subluxation). Thus, the motion dynamics may vary in each patient with os odontoideum. Lateral displacement has also been observed in patients with os odontoideum. This multidirectional instability at the articulation between the atlas and the axis is suggestive of defective cruciate and capsular ligaments.

### Clinical Presentation

Os odontoideum may be identified incidentally, and the symptoms in these cases are apparently unrelated to the odontoid abnormality. Worsening neurological symptoms may be of acute or gradual onset and progressive in nature. Neurological deficits can range from mild to severe motor and sensory abnormalities. Symptoms related to vertebral artery compromise have also been reported. Trauma may precipitate neurological symptoms in these patients. Neck pain and torticollis are prominent. Ataxia, limb weakness, limited neck movement, swallowing difficulties, and hoarseness of voice are a few of the presenting symptoms.

### Radiological Diagnosis

Os odontoideum is a radiographic diagnosis. In this abnormality, the odontoid process has a smooth, well-defined cortical margin that has no demonstrable bony continuation with the body of the C2. Os odontoideum is similar to type II odontoid fracture in its site of discontinuity from the body of C2. The gap between the os odontoideum and the body of the axis is always seen to be above the level of the superior facets of the axis vertebrae. There is often associated hypertrophy of the anterior arch of the atlas associated with os odontoideum. This helps distinguish an os odontoideum from an acute dens fracture.

Os terminale represents only the tip of the odontoid process that is free from the rest of the odontoid process. It does not represent an unstable anomaly.

Both os odontoideum and os terminale may move with the clivus or with the anterior arch of the atlas. The pattern of movement of the ossicle varies and has unique characteristics for every patient.

Degenerative arthritis of the atlantoaxial joint can lead to osteophyte formation at multiple places in the region. Degenerative osteophyte in relationship to the apical ligament can sometimes simulate an os-odontoideum.

### Management

#### Indications for Surgery

Probably because of the relative rarity of these cases, the exact treatment protocol and the indications and timing of surgery are not clearly defined in the literature. Our indications for surgery in patients with os odontoideum include the following:

1. Whenever os odontoideum is diagnosed in a case with significantly major trauma, it is difficult to differentiate it from fracture of base of odontoid process. In such cases, it may be worthwhile to treat both of the entities (os odontoideum and fracture base of odontoid process) in a similar manner. In cases where a patient develops neurological deficit following injury and then recovers, instability can be presumed to be present and treated accordingly.
2. Neck pain forms an important indication for surgery. Its presence indicates instability of the region.
3. Any indication of instability on dynamic imaging
4. Any radiological evidence of compromise of the canal diameter
5. Presence of neurological symptom(s) or deficit(s)
6. Surgery can be avoided in an incidentally detected os odontoideum. However, any clinical or radiological evidence of instability or cord compromise indicates the need for surgical treatment.

Following our experience in the region and safety of the operations of atlantoaxial fixation with our technique, we currently tend to favor surgery and would err in its favor.

### Surgical Management

Several types of surgical procedures have been recommended for treatment of instability related to os odontoideum. Anterior trans cervical screw fixation of the odontoid process under radiographic control has been seen to provide satisfactory fixation. The advantage of this technique is that the movements of the atlantoaxial joint are not hampered by the fixation. The disadvantages of the technique are that the fixation by itself is biomechanically weak, and failure rates are high. Occasionally, it can be difficult engaging the body and odontoid ossicle satisfactorily using a single or double screw. Various authors have recommended posterior fixation surgery using a range of described methods. In our experience, we have observed that our technique of C1 lateral mass and C2 pars interarticular fixation provides the biomechanically firmest and most rigid internal fixation. The possibility of denuding the articular cartilage of the atlantoaxial joint, stuffing bone graft within the joint, and manipulating the
Joint to reduce dislocation makes this technique versatile. Individual placement of screws in the C1 and C2 lateral masses makes the procedure safe as regards the vertebral artery. The procedure provides a segmental fixation; following the fixation procedure, the movements of the neck are nearly complete. In our series, we achieved 100% bone fusion, and all patients showed symptomatic and clinical neurological recovery (Figs. 45.1 and 45.2). There was no need for any kind of rigid external halo orthoses following surgery.\textsuperscript{10,11}

\section*{Os Odontoideum and “Fixed” Atlantoaxial Dislocation}

In some cases, os odontoideum may be associated with “fixed” atlantoaxial dislocation. Impaction of the transverse ligament between the odontoid process and the body of the C2 has been implicated in the development of fixed atlantoaxial dislocation. In our previous study on the subject, and with our technique of atlantoaxial joint manipulation, distraction and reduction of the dislocation, we observed that the joint in these cases is not entirely fixed. Subtle and persistent movements at this joint are the probable cause of clinical symptoms.\textsuperscript{12} Reduction of the dislocation is possible in these cases (Figs. 45.3 and 45.4).

\section*{Os Odontoideum and Basilar Invagination}

Several of our patients with basilar invagination had os odontoideum and ossiculum terminale persistens.\textsuperscript{13} It is unclear if os odontoideum by itself was the cause of long-standing instability and the subsequent development of basilar invagination. Direct distraction of the facets of the atlas and axis, reduction of the basilar invagination and atlantoaxial dislocation, and subsequent atlantoaxial fixation can most effectively treat this abnormality (Fig. 45.5).\textsuperscript{14}

Text continues on page 514
Fig. 45.2a–d Images of a 25-year-old man with a 2-month history of trauma.

a Radiograph with the neck in extension showing fracture of the odontoid at the base.

b Radiograph with the neck in flexion showing the dislocation.

c Magnetic resonance imaging (MRI) showing the odontoid fracture.

d Postoperative radiograph showing fixation of the region by our technique.

Fig. 45.3a–f Images of a 13-year-old girl who presented with neck pain. There was no history of trauma.

a Radiograph with the neck in flexion showing the os odontoideum and instability.

b Radiograph with the neck in extension showing reduction of the dislocation.
Fig. 45.3c–f

c  CT scan showing os odontoideum and instability.
d  Postoperative scan showing reduction of the instability.
e  CT scan with the cut passing through the facets showing the plate and screw fixation using our technique.
f  Postoperative radiograph showing the fixation.
Fig. 45.4a–h
a  Lateral radiograph of the craniovertebral junction (CVJ) in a flexed position showing atlantoaxial dislocation.
b  Radiograph with the head in extension showing irreducibility of the dislocation.
c  CT scan with the head in flexion showing os odontoideum and atlantoaxial dislocation.
d  CT scan with the head in extension showing os odontoideum and irreducible atlantoaxial dislocation.
e  T1-weighted MRI showing the dislocation and cord compression.
f  T2-weighted MRI showing the dislocation and cord compression.
Fig. 45.4g–h

**g** Postoperative scan showing reduction of the dislocation.

**h** Lateral radiograph in flexion showing lateral mass fixation using the plate and screw technique.

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Fig. 45.5a–c

**a** MRI showing basilar invagination and ossicle terminale.

**b** CT scan showing os odontoideum and basilar invagination.

**c** Postoperative CT scan showing reduction of the basilar invagination and realignment of the CVJ.
Conclusion

Os odontoideum is frequently identified with craniovertebral anomalies. Its presence is usually associated with instability of the region. However, presence of clinical and radiological evidence of instability alone should guide the need for surgical treatment.

References

6. McRae DL. The significance of abnormalities of the cervical spine. AJR 1960;84:3–25
Anomalies of the axis vertebra are common and involve primarily the odontoid process and, relatively less frequently, the vertebral body. Anomalies of the posterior elements are extremely rare. We encountered three cases where the posterior elements of the axis were absent, the body of the axis was dislocated anterior to the body of the C3 vertebra, and the posterosuperior lip of the C3 vertebral body caused severe compression of the spinal cord. The remarkable similarity of these three cases suggests that the absence of the posterior elements of the axis and spondyloptosis is a well-defined clinical entity and that treatment options in such cases need to be crystallized.

Incidence

Congenital absence of the posterior elements of the axis is extremely rare. In our three cases, there was complete absence of the posterior elements of the axis, and the body of the axis was dislocated and positioned anterior to the C3 cervical vertebra. The body of the C3 vertebra caused severe cord compression. We could locate only two additional reports of patients with partial defect in the posterior elements of the axis. Morizono et al. detected the defect in the spinous process and lamina of the second cervical vertebra during a roentgenographic study of the cervical spine for investigation of headache and neck pain following trauma. This patient responded to conservative therapy. Clinical details of the second case labeled as spina bifida of the axis are not mentioned in the report. In contrast, anomalies related to the defects in the development of the posterior arch of the atlas have a reported incidence of 3 in 1000 persons.

Development

Embryologically, the arch and the spinous process of the axis are formed from a pair of left and right primordial cartilages. The cause of a defect may be either malformation of the primordia or failure of ossification of the normally developed cartilage. Complete absence of both the cartilaginous and bony components of the posterior elements of the axis implies a very early fault in embryogenesis. O’Rahilly et al. studied the cervical spine of nine embryos. They found that by stage 23 of embryogenesis (8 postovulatory weeks), the neural arches of the axis extended laterally from the centrum and formed a complete or incomplete foramen transversarium. During the same period of embryogenesis, the neural arch proceeds directly backward to a variable extent, forming pedicles, articular facets, and part of the lamina. A developmental defect in our cases could have occurred during or before this stage of embryonic development. The exact cause of the defect in our patients cannot be known. There was no evidence of consanguinity or obvious teratogenic events in utero.

Clinical Features

The clinical and radiological features of all three cases encountered by us have remarkable similarities. There was no history of major trauma to the neck or head. In the first two patients, there was a partial reduction of the dislocation after the application of traction. However, our third patient did not tolerate traction and showed clinical symptomatic worsening.

Treatment

An attempt to fix the spine by interlaminar instrumentation after partial reduction of the dislocation with traction resulted in neurological worsening in our first patient. Transcervical decompression, which was followed by posterior fixation, resulted in satisfactory clinical improvement in the second patient. In the third, anterior transoral decompression and plate and screw fixation resulted in immediate and sustained clinical improvement. This case is illustrated below. Although there is the risk of infection following placement of a metal plate via a transoral route, the ease of exposure and decompression and fixation by this route can be employed to great advantage.

Conclusion

Further experience is mandatory to formulate the most appropriate treatment protocol for this complex anomaly and challenging therapeutic problem. We are not yet convinced whether a transoral or a transcervical route is superior. The best possible method of stabilization in such cases also needs to be identified.
A 31-year-old man presented with progressively worsening stiffness and weakness in all limbs for the past year. For 1 month, the disability worsened rapidly, and he needed support to walk or stand and could not perform his routine activities normally. In addition, for 1 month he developed tingling paresthesias in all four limbs. There was no urine or bowel problem. There was also no history of any trauma. His neck was relatively short since birth. Neurological examination revealed spastic quadriparesis, grade 4. Deep tendon reflexes were exaggerated. Plantar reflex was bilaterally extensor. The rest of the neurological examination was normal, including the lower cranial nerves and the sensory system examination. Cervical movements were restricted. Plain radiographs, magnetic resonance imaging, and computed tomography (CT) of the craniovertebral junction in flexion and extension revealed the complete absence of all posterior elements of the axis vertebra, namely, transverse processes, lateral masses, laminae, and spinous process (Fig. 46.1a–d). The body of the axis was dislocated anterior to the C3 body in the form of almost complete spondyloptosis. Large bony osteophytes were seen anterior to the body of the C2 and C4 vertebrae. The body of C3 severely compressed the adjacent cervical cord. There was no evidence of instability on dynamic films. Additionally, there was no anomaly of the odontoid process or any other bone in the region.

The patient was placed in traction, which had to be removed immediately, as the patient complained of diplopia and exaggerated paresthesias in the limbs. A transoral operation was performed. The indenting part of the C3 vertebral body was radically decompressed after exposing it through a large window obtained by drilling in the inferior aspect of the body of C2. A multiholed plate was used and was fixed by screws superiorly to the upper part of the body of the axis and inferiorly to the body of C4 (Fig. 46.1e, f). Bone graft was placed under the plate in the region of the resected body of C2. At follow-up after 6 months, the patient showed remarkable neurological recovery. The plate was in situ, and there was some evidence of bone fusion in the region of placement of the bone graft.

In such a case with a severe spinal deformity, the aim of surgery is to decompress the spinal cord widely and to do spinal fixation. The options for surgical approach to this patient were as follows:

1. Attempt to reduce the dislocation with the help of traction, then attempt fixation with the help of instrumentation in the maximally reduced dislocation
2. Decompress the spine from anteriorly through a transcervical approach and simultaneously fuse with instrumentation
3. Do transoral decompression, then perform a posterior fixation technique
Fig. 46.1a–f

a  Lateral plain radiograph of the craniovertebral region showing the axis vertebra dislocated anterior to the body of the C3 vertebra. The posterior elements of the axis are absent.

b  Sagittal magnetic resonance imaging of the craniovertebral region showing severe compression of the cord by the body of the C3 vertebra.

c  Axial computed tomography (CT) scan showing the anterior arch of the atlas, the body of the axis, and the C3 body lying in the same axial plane. All posterior elements of the axis are absent.

d  CT showing the anomaly. The body of the C3 vertebra is causing severe compression of the spinal cord.

e  Postoperative radiograph showing the anterior plate and screw fixation following transoral decompression.

f  CT scan showing the bone decompression of C3. Anterior fixation with plate and screws is seen. Bone graft is seen posterior to the plate.
References

Ossification of the ligamentum flavum is frequently seen in the lumbar spine and is more often secondary to a chronic degenerative spondylotic process. Congenital or posttraumatic ossification of the ligamentum flavum is rare, even in the lumbar spine. There are isolated case reports of ossification of the ligamentum flavum in the cervical spine. Although stenosis of the spinal canal in the atlantoaxial region in isolation and in association with the rest of the cervical spine has been reported, segmental calcification or ossification of the ligamentum flavum in the atlantoaxial region has not been. We identified one case of such ossification.1

Pathogenesis

The exact pathogenetic mechanism of ossification of the ligamentum flavum is unclear.1 Dynamic, chronic, and excessive stresses on the ligamentum flavum have been suggested as causative factors.1 Trauma to the ligamentum flavum has been identified as an etiological factor.1 Systemic hyperostosis, endocrine abnormality, and a generalized aging process may also be responsible.1

Histological examination helps to differentiate calcified ligamentum flavum, ossified ligamentum flavum, and calcium pyrophosphate dihydrate crystal deposition disease.2–5 In calcification of the ligamentum flavum, calcified granules are deposited within the degenerated ligament fibers, with no mature bone formed within the ligament, which is a feature of ossified ligamentum flavum.6

Differences exist between the ossification of the posterior longitudinal ligament and ossified ligamentum flavum, but similarities exist at the molecular and possibly the genetic levels.1 The clinical presentation may be that of myelopathy or radiculomyelopathy, of acute or insidious onset and progression. Based on the clinical presentation and radiological investigations, the differential diagnosis includes calcified ligamentum flavum, osteoid osteoma, and fracture or congenital anomalies of the lamina.1

Investigations

Computed tomography and magnetic resonance imaging provide definition of the extent, site, and nature of the disease.7–9 High-intensity intramedullary signal changes reflect edema or gliosis of the spinal cord and are generally associated with a poor prognosis.10–13 However, in patients with ossified ligamentum flavum, the high-intensity intramedullary signal may be due to reversible cord changes and are therefore not necessarily associated with a poor prognosis.2,14 In our patient, although the cord demonstrated a high signal intramedullary change on both preoperative and postoperative images, good clinical recovery was achieved.

Treatment

Myelopathy due to ossified ligamentum flavum responds dramatically to decompressive laminectomy.15–17 Meticulous microsurgical resection of the ossified mass is necessary to avoid damage to the dura mater, spinal cord, and nerve roots. The dura mater may be thinned or ossified due to long-term compression, and its separation from the ossified mass requires careful attention to avoid dural laceration.18

Case Illustration

A 30-year-old man presented with progressive paresthesias and weakness of all four limbs following a fall from a bicycle 6 months previously. Additionally, he had increased frequency of micturition and constipation. On admission, he was severely disabled and was even unable to sit or stand without support. Examination revealed spastic grade 3 quadriplegia and diminished spinothalamic and kinesthetic sensations below the second cervical dermatome. Investigations revealed unusual abnormal bony transformation of the entire ligamentum flavum between the lamina of the axis and the arch of the atlas, resulting in focal stenosis and severe cord compression. The rest of the cervical spine showed no significant abnormality (Fig. 47.1a–c).
The patient underwent posterior decompressive surgery. The bones of the atlas and axis were thick, sclerotic, and hard, and the intervening ligamentum flavum had been transformed into a block of hard bone. Laminectomy and excision of the ossified ligamentum flavum was completed using bone rongeurs and a high-speed drill. The dura was not adherent to the ligamentum flavum. Following surgery, the patient showed remarkable clinical motor and sensory recovery. At 6-month follow-up, he was almost asymptomatic. Histological examination of the surgical specimen of the abnormal bone showed endochondral ossification, lamellar bone structure, and marrow formation suggestive of ossification of the ligamentum flavum. Postoperative radiology demonstrated adequate decompression of the cervical cord and complete excision of the atlantoaxial ossified ligamentum flavum (Fig. 47.1d, e).
Conclusion

Ossified ligamentum flavum of the craniovertebral junction is a rare cause of cervical myelopathy. Posterior decompression can form a rational form of surgical treatment.

References

Several bony and soft tissue anomalies have been associated with anomalies in the craniovertebral region. In this chapter, we present a report of an unusual pathology of the atlas vertebra that involved unilateral facetal hypertrophy. We had reported a series of four such cases that presented with progressive symptoms of cord compression and torticollis and recently treated an additional fifth case. Additionally, we recently identified another case of atlantal facetal hypertrophy that appeared to be secondary to osseous changes related to acromegaly. Because the pathology is rare, the exact treatment protocol is not clear.

Hypertrophy of the facets of the lumbar and the dorsal spine has been reported frequently. Less commonly, hypertrophy of the facets of the subaxial cervical spine has been identified. Facetal hypertrophy is generally associated with spondylotic changes. Such hypertrophy usually results in spinal canal stenosis and related symptoms. Although isolated hypertrophy of a single facet has been reported, the hypertrophy is usually generalized or involves multiple spinal segments.

Unilateral hypertrophy of the facet of the atlas vertebra is extremely rare. The exact cause of the anomaly remains unclear. In none of our five cases was there a defined syndrome complex suggestive of a hereditary or genetic disorder. All of our patients had presented with long-standing torticollis and progressive symptoms of cord compression. The clinical progression of the symptoms is suggestive of either a congenital abnormality or a slow-growing pathology. The remarkable similarity of the symptoms and of the pathology in all our cases suggests a unique congenital abnormality. The presence of associated fusion of the C2–C3 vertebrae in one case also points toward a congenital anomaly complex. Investigations revealed hypertrophic abnormality of no other bone or its component. The maximum transverse dimension of the facet of the atlas ranged from 2.8 to 4.9 cm in size. In two cases, there was associated syringomyelia, suggesting chronicity of the problem. The bone appeared normal in its architecture and moderately brittle in consistency in two of our cases where drilling of the part impinging into the canal was done. Histological examination of the bone in this case did not reveal the presence of any bony tumor, such as fibrous dysplasia. At follow-up, there was no growth in the size of the lesion that could suggest a tumorlike pathology.

### Treatment

Several treatment options for spinal canal stenosis in the presence of facetal hypertrophy have been reported. A decompressive procedure that involves resection of the posterior arch of the atlas and adjoining parts of the foramen magnum and lamina of the axis was done in our first three cases (Figs. 48.1 and 48.2). In the fourth and fifth cases, direct decompression of the part of the bone compressing the cord was performed. Our familiarity with the surgical anatomy of the region of the facets of the atlas and axis and our techniques involving manipulation of the lateral masses of the atlas and axis probably influenced our surgical strategy in the latter case. Atlantoaxial fixation was done in the fourth case (Fig. 48.3), as the articular surface was manipulated during surgery, although the joint appeared intact and normal. In the fifth case, no atlantoaxial fixation was done (Fig. 48.4).

The technical ease of a posterior decompressive surgical procedure over direct manipulation and fixation of the region favors the former. However, it appears to us that direct decompression of the indenting part of the abnormal bone could be a more rational way of treatment. The more remarkable clinical recovery and improvement in torticollis in the patient undergoing the latter form of treatment are also suggestive of the superiority of the treatment modality. The need and effectiveness of unilateral fixation can be questioned. However, to define the best treatment protocol for this most unusual and uncommon anomaly, more experience appears to be mandatory.

### Acromegaly and Hypertrophy of the Atlas Facet

Recently, we encountered unilateral facetal hypertrophy in a known case of acromegaly (Fig. 48.5). Osteoarticular involvement is a featured sign in acromegaly leading to arthropathy, which is a major cause of morbidity. The spine is noticeably targeted by chronic growth hormone excess and presents with widened intervertebral spaces, vertebral enlargement, and osteophyte formation caused...
Fig. 48.1a, b
a  Computed tomography (CT) scan showing the large facet of the atlas.
b  Sagittal view showing the large facet. The corresponding articular surface is also relatively large. White arrow clivus, black arrow enlarged facet of atlas, arrowhead C2 facet adjoining the enlarged C1 facet.

Fig. 48.2a–d
a  CT scan showing the large facet of the atlas.
b  Three-dimensional CT scan showing the hypertrophic C1 facet.
c  T2-weighted axial magnetic resonance imaging (MRI) showing the hypertrophic C1 facet.
d  Coronal view showing the large facet of the atlas.
Unilateral Atlantal Facetal Hypertrophy

Fig. 48.3a–e

a  CT scan showing the large facet of the atlas.
b  Coronal view showing the large facet of the atlas.
c  MRI showing the indentation of the cord by the enlarged facet. Syringomyelia can be observed.
d  Postoperative view showing the drilling of the part of the facet indenting the cord.
e  Postoperative radiograph showing plate and screw fixation. C2–C3 fusion can be observed.
Fig. 48.4a–i

a CT scan showing hypertrophy of the facet of the atlas. The rest of the bones are normal.
b Axial cut of the CT scan showing hypertrophy.
c CT scan showing no evidence of atlantoaxial dislocation.
d T2-weighted MRI showing the hypertrophic C1 facet and indentation of the cord.
e T1-weighted MRI showing the hypertrophic C1 facet.
f Coronal section of the MRI showing the hypertrophic C1 facet.
g Three-dimensional CT scan showing the hypertrophic C1 facet.

Fig. 48.4 j–i  ▶
Fig. 48.4 h–i
h Postoperative CT scan showing resection of the hypertrophic part of the C1 facet.
i Axial CT cut showing resection of the hypertrophic cord indenting part of the facet.

Fig. 48.5a–h
a Preoperative T1-weighted MRI showing a relatively large pituitary tumor.
b Postoperative scan showing the residual tumor in the cavernous sinus.
c Axial CT scan showing unilateral hypertrophy of the facet of the atlas.
Fig. 48.5d–h

d  Sagittal view of the region showing hypertrophy of the atlantal facet.

e  T2-weighted MRI showing evidence of cord compression.

f  MRI showing cord compression.

\g  Axial CT scan showing the spacers within the joint and postoperative status of the facets of the atlas and the cord.

h  Radiograph showing the alignment of the region in flexion. Spacers are seen in both atlantoaxial joints.
by endochondral, marginal, and subligamentous growth of vertebral bone. In our case, the patient had marked symptoms of acromegaly and characteristic enlargement and thickening of several bones in the face and in the limbs. In the spine, investigations revealed hypertrophic abnormality of no other bone or its component. There was a suspicion of associated atlantoaxial instability during surgery, probably due to the incompetence of the facet joint. The bone appeared normal in its architecture and moderately brittle in consistency during drilling, but there was no histological evidence of any bony tumor. The part of the bone indenting into the cord was drilled, and atlantoaxial fixation surgery was done using the joint-jamming technique described elsewhere in this book (see Chapter 14). This patient exhibited clinical recovery following the surgery.

**Conclusion**

Unilateral enlargement is a rare but defined abnormality. Correct identification and appropriate treatment can result in a lasting cure.

**References**

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These relatively unusual cases are collected from the database of craniovertebral junction–related pathology from the Department of Neurosurgery at K.E.M. Hospital in Parel, Mumbai, India.

**Arachnoid Cyst and Basilar Invagination**

An 18-year-old male patient had torticollis ([Fig. 49.1](#)). There was no neurological deficit. Investigations showed basilar invagination and severe torticollis. An arachnoid cyst was seen on the side contralateral to the torticollis. The patient is under neurological observation.

**Case Illustration 1**

**Arachnoid Cyst and Basilar Invagination**

An 18-year-old male patient had torticollis ([Fig. 49.1](#)). There was no neurological deficit. Investigations showed basilar invagination and severe torticollis. An arachnoid cyst was seen on the side contralateral to the torticollis. The patient is under neurological observation.

**Fig. 49.1a–c**

- **a** Sagittal T2-weighted magnetic resonance imaging (MRI) showing basilar invagination and severe cord compression.
- **b** Coronal image showing marked torticollis.
- **c** MRI showing the presence of a large arachnoid cyst contralateral to the torticollis.
Bifid Anterior and Posterior Arches of the Atlas Associated with Mobile Reducible Atlantoaxial Dislocation

An 8-year-old girl presented with torticollis since birth, frequent falls, and progressive spastic quadriplegia (Fig. 49.2). Investigations showed mobile and reducible atlantoaxial dislocation. The cord was severely compressed. The anterior and posterior arches of the atlas were bifid. The patient underwent lateral mass plate and screw fixation. Following the surgery, she showed dramatic clinical recovery. The presence of bifid arches signifies a defect in the embryogenetic process. In presence of bifid posterior arch of atlas, midline wiring procedure may not have been possible.

**Case Illustration 2**

Fig. 49.2a–f

a Radiograph with the head in flexion showing atlantoaxial dislocation.
b Radiograph with the head in extension showing reduction of the dislocation.
c T2-weighted MRI showing severe cord compression.
d Postoperative axial computed tomography (CT) scan showing the bifid anterior and posterior arches of the atlas. Screws in the facet of the atlas can be seen.
e Postoperative CT scan showing the fixation in good alignment.
f Sagittal CT scan showing the screws in the facets of the atlas and axis.
Bilateral Lateral Dislocation

A 12-year-old boy had trauma to the head due to a traffic accident (Fig. 49.3). Apart from pain in the neck, there was no neurological deficit. The patient was treated conservatively. At 1-year follow-up, he was asymptomatic. However, the investigations showed persistent dislocation of the facets and fractures in the atlas and its ring. Lateral mass fixation procedure is possible in such a situation when midline wiring techniques may not be possible and may be technically more difficult and dangerous.

Fig. 49.3a, b

a CT scan showing bilateral lateral facetal dislocation.
b The anterior and posterior arches of the atlas are fractured, leading to disruption of the atlas ring.
c Sagittal image shows basilar invagination as a result of lateral facetal dislocation.
C2 Tuberculosis Radiologically Simulating a Chordoma

A 78-year-old woman who was diagnosed with breast cancer 8 years ago had progressive weakness of all four limbs and respiratory distress for about 1 month. Investigations revealed a lesion that destroyed a large part of the anterior elements of the C2 vertebra (Fig. 49.4). There was a prevertebral and retrovertebral soft tissue lesion. Initially, the lesion was believed to be a metastasis. The patient underwent anterior and posterior evacuation of purulent material and destroyed bone tissue. Histopathology revealed that the lesion was tuberculous in nature. The patient was subsequently treated with antituberculous drugs. She has shown progressive neurological recovery following this treatment. At a follow-up after 6 months, investigations showed resolution of the lesion.

Fig. 49.4a–e
a  Sagittal T1-weighted MRI showing the lesion involving the C2 vertebra and extending into the pre- and retrovertebral region, compressing the cord.

b  T2-weighted image showing the lesion.

c  Contrast-enhanced T1-weighted MRI showing the lesion.

d  Postoperative and following drug therapy T1-weighted MRI showing resolution of the lesion.

e  T2-weighted MRI showing the resolution of the lesion.
Chordoma Involving C2 Vertebra

Chordomas frequently involve the C2 vertebra and present a complex therapeutic challenge. A 60-year-old man presented with tingling, numbness in both upper limbs, and weakness of all four limbs. Examination revealed spastic quadriparesis. Investigations showed a lesion at the level of the C2 vertebra. It was isointense on T1-weighted images and hyperintense on T2-weighted images (Fig. 49.5). The lesion caused destruction of the vertebral body, the facet of the axis, and the odontoid process. The patient underwent transoral surgery with radical but incomplete resection of the tumor. Histopathology of the lesion revealed that it was a chordoma. Chordomas in this location are common. High recurrence rates and the relentlessly progressive course of the disease are hallmarks of chordoma in this region.

Fig. 49.5a–e
a  T1-weighted image showing extensive destruction of the C2 vertebral body and odontoid process. The tumor extends both in the prevertebral region and in front of the cord.
b  T2-weighted image showing the lesion.
c  Coronal CT scan showing the destruction of the body, odontoid process, and lateral mass of the C2 vertebra.
d  Sagittal image showing bone involvement.
e  Micrograph showing classic features of chordoma with physaliphorous cells and myxoid matrix.
Chordoma Involving C2 Vertebra

Chordomas frequently involve the C2 vertebra and present a complex therapeutic challenge. A 35-year-old man had surgery for a large clival chordoma (Fig. 49.6). Transcervical partial resection of the tumor and posterior occipitocervical fixation were done at that time. Eight months after surgery, the patient was referred to us in a poor neurological state. He had severe spastic grade 1 quadriparesis and needed ventilatory assistance for breathing. The tumor was massive and extended on both sides of the anteriorly displaced ororespiratory passages. The patient was reoperated. The anterior cervical approach was reopened, and the tumor was radically resected from the left side. The involved vertebral bone was removed, and the tumor extending to the right side was resected partially from underneath the esophagus and trachea. The soft nature of the tumor assisted in resection, and the planes of soft tissue made by tumor expansion assisted in radical tumor resection. Following the radical surgery on one side, and while waiting for surgery on the contralateral side, the patient showed dramatic clinical recovery. When seen after 1 month, he could walk unaided. He never returned for second-stage surgery for the tumor on the right side of the trachea.

**Case Illustration 6**

**Fig. 49.6a–d**

a Axial T1-weighted MRI shows a massive chordoma extending on both sides of the trachea after displacing it anteriorly. Artifacts related to metal implant can be seen.

b Coronal image showing the tumor.

c Postoperative scan (axial view) showing tumor resection on the left side.

d Postoperative scan (coronal view).
Circumferential Cord Tuberculous Granulations

A 49-year-old woman presented with a 7-month history of progressive quadripareis. At the time of admission, she had spastic grade 4 quadripareis. Both T1- and T2-weighted images revealed a hypointense lesion surrounding the upper cervical cord (Fig. 49.7). Computed tomography (CT) scan showed the mass to be hyperdense and contrast enhancing in nature. During surgery, a thick film of extradural granulation tissue was encountered that was partially removed. Histology revealed that the lesion was chronic granulation tissue. The patient was placed on empirical antituberculous drugs. She progressively recovered on drug treatment and when seen after 3 years was symptom free. Repeat investigations showed resolution of the granuloma.

**Fig. 49.7a–g**

a T2-weighted MRI showing the hypointense lesion in the craniovertebral region extending from the occipital region to the fifth cervical vertebra, located both anterior and posterior to the neural structures.

b T1-weighted MRI showing the lesion to be hypointense.

c Sagittal CT scan showing the hyperdense lesion.

**Fig. 49.7 d–g**

a T2-weighted MRI showing the hypointense lesion in the craniovertebral region extending from the occipital region to the fifth cervical vertebra, located both anterior and posterior to the neural structures.

b T1-weighted MRI showing the lesion to be hypointense.

c Sagittal CT scan showing the hyperdense lesion.
Clival Dysgenesis Associated with Chiari Malformation and Syringomyelia

An 11-year-old boy presented with the complaints of neck pain and difficulty in walking. He had nasality of voice since birth. Over a period of time, he had noticed that he was unable to run as fast as his friends and of late had a tendency to fall on uneven ground. On examination, he had a short neck and an abnormal shape of the occipital bone. He also had scoliosis of the dorsal spine with a convexity to the right. He had spasticity in all four limbs with hyperreflexia, but the power was grade 5 in all muscle groups. Sensory examination was normal. He had no bowel or bladder complaints. He was able to ambulate on his own. CT scan showed a rudimentary clivus with failure of fusion between the sphenoidal and occipital parts of the clivus and a shortened bone representing the clivus capping the dens. There was also evidence of C2–C3 fusion. There was basilar invagination, with the odontoid being 2.28 cm above the Chamberlain line. The atlantodental interval was normal, and there was no evidence of atlantoaxial instability. Magnetic resonance imaging (MRI) of the craniovertebral junction and cervical spine showed tonsillar herniation, as well as holocord syringomyelia with septations (Fig. 49.8). Dynamic flexion-extension images of the cervical spine did not show any instability. The patient was treated with foramen magnum decompression. Postoperatively, the patient improved in spasticity in all limbs and also noticed improvement in neck pain.

**Fig. 49.7d–g**

- d Axial CT scan (plain) showing the circumferential nature of the lesion.
- e Postoperative and post–drug treatment image showing marked resolution of the lesion.
- f T2-weighted scan showing resolution of the lesion.
- g Micrograph showing granuloma with epitheloid and Langhans giant cells.
The occipital bone is formed by the union of four primary cartilaginous centers and a fifth membranous element. The basioccipital, two exoccipital, and supraoccipital make up the cartilaginous components and the interparietal center the membranous component. The basiocciput is embryologically derived from fusion of four occipital sclerotomes. The clivus is formed from two components: the basisphenoid and the basiocciput. These two components fuse along the sphenooccipital synchondrosis. Ossification of this synchondrosis begins at 11 to 13 years in girls and 14 to 15 years in boys, and complete closure does not occur until 16 to 20 years. Sometimes, this sphenooccipital synchondrosis may persist into adult life, when it may be mistaken for a fracture. This fusion along the sphenooccipital synchondrosis is believed to be responsible for basicranial flexion, which develops in concert with the development of the upper airway and the ability to vocalize.

Hypoplasia of the basiocciput may occur to varying degrees depending on the number of occipital vertebrae affected. This results in clival shortening and basilar invagination.

The abnormality in our case was associated with a Chiari I malformation and syringomyelia. Persistence of the sphenooccipital synchondrosis is known, and there are few reports of basioccipital hypoplasia; however, our search of the literature did not find any case where there was basioccipital hypoplasia with a rudimentary clivus and an associated Chiari malformation with holocord syringomyelia.
A 46-year-old woman had swelling at the root of the nose since birth. She had a history of continuous, dull aching pain in the nape of the neck and left upper extremity for 8 years. There was slowly progressive weakness and numbness of the left upper limb over that period. For 1 year, she had facial asymmetry and bilateral hearing impairment. For a period of 3 months, she had progressive quadriparesis, the left side being weaker than the right. When admitted, she needed support to walk and preferred wheelchair ambulation.

On neurological examination, she had right lower motor neuron facial paresis and bilateral partial sensorineural hearing loss. There was hypotonia in the left upper limb and spasticity in the legs. No clear muscle wasting was seen. There was grade 4 hemiparesis on the left side. Also noticed was a dissociated cervicodorsal sensory loss in the left upper limb and left half of the trunk between C3 and T12 dermatomes. The joint position sense was impaired in all limbs. Romberg sign was positive. The patient had a broad-based gait and required bilateral support to walk. She had a characteristic facies with frontal bossing, prominent supraorbital ridges, broad nasal root, bilateral epicanthic folds, hypertelorism, hypoplastic maxillae, and small ear lobules (Fig. 49.9). She also had bilateral pseudophakia due to previous cataract surgery, retrognathia, and multiple hamartomas in the floor of the mouth. The swelling at the root of the nose was nontender, hard, and irreducible. Her neck showed pseudowebbing due to long clavicles. She also had pectus carinatum, scoliosis of the thoracic spine with convexity to the right, simian crease in the left hand, and cutaneous syndactyly of the right hand. Audiogram demonstrated bilateral mild sensorineural hearing loss.

The patient had two daughters, ages 23 (Figs. 49.10 and 49.11) and 21 years (Figs. 49.9 and 49.12), who had similar swellings at the root of the nose since birth. On examination, both daughters had prominent supraorbital ridges, broad nasal root, bilateral epicanthal folds, hypertelorism, hypoplastic maxillae, small ear lobules, pectus carinatum, and hamartomas in the floor of the mouth (Fig. 49.9). In addition, the elder daughter had thoracic scoliosis and a simian crease in the right hand, and her right scapula was higher than the left. Both daughters did not have any neurological symptom or deficit. The patient’s parents, grandparents, other siblings, and husband were normal.

Neuroradiological examination was performed of the entire family. The affected patient and her daughters had similar findings on CT scan of the cranium and face, which showed diffuse hyperostosis and sclerotic thickening of the calvaria and skull base, including both petrous bones and nasal and lacrimal bones, with hypoplasia of the

Fig. 49.9  A photograph of the affected family. The mother (the patient) and her daughters (front row) harbor craniofrontonasal dysplasia. The patient’s husband and mother are normal.
Fig. 49.10a–c Images of the affected patient (mother).

a Sagittal CT scan of the cranium, upper cervical spine, and face showing diffuse hyperostosis and sclerotic thickening of skull and nasal and lacrimal bones with hypoplasia of the maxillae and mandibles. Marked hyperostosis of the sub-occipital bone can be observed.

b Axial CT scan of the cranium showing diffuse hyperostosis of the skull bones, as well as nasal and frontal bones. Reduction in the posterior cranial fossa volume due to the hyperostosis of the bones can be clearly seen.

c T1-weighted sagittal MRI of the craniovertebral junction demonstrates the Chiari I malformation and syringomyelia.

Fig. 49.11a–c Images of the elder daughter.

a Axial craniofacial CT scan showing diffuse hyperostosis and thickening of the bones identical to the mother.
Fig. 49.11b–c
b  T1-weighted sagittal MRI showing Chiari I malformation and syringomyelia.

c  T2-weighted sagittal MRI shows no Chiari malformation.

Fig. 49.12a–b Images of the younger daughter.
a  Axial craniofacial CT scan showing diffuse hyperostosis and thickening of the bones.

b  T2-weighted sagittal MRI shows no Chiari malformation.
maxillae and mandibles (Figs. 49.10, 49.11, and 49.12). MRI of the patient and elder daughter revealed Chiari I malformation and cervicodorsal syringomyelia (Figs. 49.10c and 49.11b, c). The younger daughter did not have Chiari malformation (Fig. 49.12b). A skeletal survey was performed on all three women, which did not reveal any other abnormality. A genetic study documenting the mutated gene was not performed.

The patient underwent suboccipital, foramen magnum, and posterior arch of the atlas decompression. The bone was markedly thick and hard and moderately hypervascular. Postoperative recovery was uneventful. On 13-month follow-up, the patient reported significant improvement in the numbness of the left-sided limbs and relief of her neck pain. She was able to walk unaided. In the period of observation, the two daughters of the patient had developed no neurological symptom.

The term craniofrontonasal dysplasia was first introduced by Cohen in 1979 to describe a patient with coronal craniosynostosis, hypertelorism, limitation of shoulder movements, and digital abnormalities. It has also been referred to as craniofrontonasal syndrome. The typical manifestations of this syndrome in female patients are severe hypertelorism (frequently asymmetrical) with a central nasal groove, frontonasal dysplasia, and unilateral or bilateral coronal craniosynostosis (brachycephaly or plagiocephaly). The three female family members described in our case report had the above-mentioned characteristics and could be labeled as having this syndrome phenotypically. Male patients are usually affected mildly with hypertelorism only.

Our cases suggest that diffuse hyperostosis and sclerotic thickening of the skull base produce overcrowding of posterior fossa structures and promote hindbrain herniation through the foramen magnum. Early recognition of the syndrome is important for genetic counseling, as well as for detection of Chiari malformation and syringomyelia in an asymptomatic stage. This will permit close neurological monitoring and appropriate surgical intervention at an incipient stage.

Endodermal Cysts

Endodermal cysts are rare intracranial tumors, and only isolated cases are reported in the literature. Endodermal cysts are known by a variety of names, such as epithelial, enterogenous, neuenteric, bronchogenic, foregut, and respiratory cysts. Some studies suggest that colloid cyst, Rathke cleft, and enterogenous cyst all represent endodermal inclusion cysts, have a common histogenesis, and are named differently according to their respective location. Similar cysts are more frequently found in the lower cervical and upper dorsal spine and are lined by the presumed endodermal-derived epithelium. They most commonly occur in the posterior cranial fossa, and most of them have been located anterior or anterolateral to the brainstem. Frequently, they extend anterior to the cord at the cranio cervical junction. Tumors can be diagnosed on the basis of their characteristic radiological features. The image intensities on MRI are dependent on the viscosity of the cyst fluid. The cysts are large, iso- to hyperintense on T1-weighted images, and iso- to hypointense on T2-weighted images (Fig. 49.13). Usually there are associated small soft calcifications in proximity to the major arteries. During surgery, the cyst material usually has a puslike hue and consistency. The wall of the cyst is usually thin and “arachnoid-like.” In 2005, we proposed that evacuation of the cyst contents and safe partial resection of the cyst wall is sufficient for long-term control of these benign lesions. The cyst contents should be radically removed, and spillage into the subarachnoid space should be avoided, as such an intraoperative event can result in “chemical” meningitis.
An 8-year-old boy presented with progressively increasing swelling in the suboccipital region for 2 years. The patient had no stigmata of neurofibromatosis. On admission, the swelling measured $9 \times 9 \times 7$ cm. It extended from the external occipital protuberance to the midcervical region in vertical extent and to both the mastoid processes horizontally. The swelling was firm, solid, multinodular, and nontender. The overlying skin was smooth and shining but normal and free from the underlying mass. The patient had no neurological deficits. MRI revealed an extradural tumor that was isointense on T1-weighted images and hyperintense on T2-weighted images with homogeneous enhancement after contrast administration (Fig. 49.14). The tumor had widely destroyed the suboccipital bone and...
posterior elements of the atlas and axis. The vertebral artery was encased by the tumor along its course adjoining the arch of the atlas on the right side. Angiography revealed extensive tumor vascularity arising from the external carotid and vertebral arteries. Prior to surgery, the external carotid artery was embolized. At surgery, the tumor was found to be entirely extradural, firm, and moderately vascular. It was radically resected piecemeal. A small part of the tumor encasing the vertebral artery was left behind. The patient made an uneventful recovery. Postoperative MRI showed radical excision of the tumor with a small residue near the vertebral artery. Histological examination confirmed the diagnosis of a benign nerve sheath tumor. At 2-year follow-up, the patient was asymptomatic.

In this case, the vertebral artery was encased by the tumor in its course, suggesting that the tumor probably arose from the second cervical ganglion. Neurinomas arising from the second cervical ganglion are relatively common, but are usually dumbbell-shaped and seldom achieve such a massive size; they have never been reported to encase the vertebral artery. Radical resection of these tumors is usually advocated, and the long-term outcome after such treatment is excellent.11,12
Massive Cervicomedullary Intramedullary Spinal Cord Lipoma

A 17-year-old male patient presented with difficulty in walking, lifting heavy weights, and carrying out fine motor tasks, especially with the left hand, for 1 month. He did not have any bowel or bladder complaints. On examination, he had spastic quadriparesis. He was able to ambulate with support. There was no evidence of spinal dysraphism. MRI revealed an intramedullary lipoma on the posterior aspect of the cervical spinal cord extending from the craniovertebral junction to C6 (Fig. 49.15). The patient was managed surgically. A suboccipital craniectomy, excision of the arch of C1 and C2 to C6 laminoplasty, and subtotal excision were performed. A thick layer of tumor over the spinal cord was left behind, as no clear plane of dissection was available. The dura was closed primarily. Histological examination showed lipomatous differentiation, including mature fat and large signet ring adipocytes with compressed marginalized nuclei. The patient showed gradual improvement in his spasticity and power. Postoperative MRI revealed a small residual intramedullary tumor in the cervical cord. At 3-year follow-up, the patient was symptom free.

Fig. 49.15a–d
a Sagittal T1-weighted MRI showing the hyperintense lipoma.
 b T2-weighted image showing the hyperintense tumor. The presence of syrinx on the inferior end of the tumor suggests the long-standing presence of the tumor.
 c Postoperative scan showing the residual tumor and decompression of the cord.
 d Postoperative T2-weighted image.
Occipitocervical Pseudomeningocele

An 8-year-old boy sustained blunt injury when he fell on his back from a height of 5 m. He was injured at the nape of the neck by a large stone. Except for local skin contusion, there were no neurological sequelae. Subsequent to this, the patient noticed progressively increasing swelling over the nape of the neck that gradually extended over his shoulders. The swelling reached its maximum size in 3 to 4 hours. Over the next 3 to 4 days, the swelling regressed partially and became softer. A subcutaneous hematoma was suspected, and conservative management was advised. The size of the swelling did not regress any further over the next 3 weeks, and the boy also started complaining of headaches. Percutaneous aspiration revealed the contents to be cerebrospinal fluid (CSF). Imaging showed a massive CSF collection in the occipitocervical region with no suggestion of the site of the fistula with the subarachnoid spaces (Fig. 49.16). There was no occipital bone defect or fracture of the atlas or axis. The brain and the rest of the CSF-containing spaces were entirely normal. Lumbar puncture reduced the size of the swelling; a lumboperitoneal CSF shunt was placed, which resulted in complete regression of the swelling (Fig. 49.17). However, 4 weeks after surgery, the child was readmitted with reappearance of swelling of the same size. Three-dimensional CT scan with bone windows revealed the presence of a bony shell around the CSF collection (Fig. 49.18). Revision of the blocked lumboperitoneal shunt resulted in regression of the swelling. Once the swelling was decompressed, large areas of subcutaneous calcifications could be palpated. At 14-month follow-up, the child was asymptomatic.

Fig. 49.16a, b
a T1-weighted sagittal MRI showing a large pseudomeningocele in the suboccipital region. Note that the cyst fluid is in continuity with the fourth ventricle.

b T2-weighted axial MRI showing the pseudomeningocele and its communication with the spinal subarachnoid space.

Fig. 49.17a, b
a Postoperative T1-weighted MRI showing reduction of the size of the pseudomeningocele.

b T2-weighted MRI.
Massive Benign Osteoblastoma of the Clivus and Atlas

A 50-year-old man presented with pain in the neck and torticollis for 2 years. For 4 months, he had noticed progressively enlarging swelling in the right occipitocervical region. For 10 days prior to admission, he began developing a rapidly progressive neurological deficit. On examination, he had spastic quadriplegia, absent gag and palatal reflexes, and right hypoglossal nerve palsy. There was a swelling in the right occipitocervical region measuring 17 × 12 cm that appeared to be arising from the underlying bone. It was free from the overlying skin. Imaging showed a large multiloculated radiolucent mass overlying the occipital bone and upper cervical vertebrae (Fig. 49.19). An eggshell-like appearance was seen along the posterior suboccipital and right-sided walls. The central part of the overlying atlas, axis, and mastoid bones was irregular and densely sclerotic, suggesting new bone formation, calcification, or both. There was evidence of destruction of a large part of the atlas, odontoid process, lower clivus, and right lateral margin of the foramen magnum. MRI showed the massive tumor with extensions into the cervicomedullary junction and cerebellum, the former displaced to the left and the latter elevated. The tumor extended into the neck and parapharyngeal region. Vertebral angiography showed the right vertebral artery to be narrowed up to the basilar artery and revealed irregularity of its lumen from above C3. The tumor was vascular, deriving its blood supply from the muscular branches of the vertebral artery. Carotid angiography showed the common carotid arteries and their bifurcations displaced anteriorly by the mass. There was no contribution from the carotids. The patient was operated. The tumor had displaced and stretched the muscles of the region. It was moderately vascular and largely cystic. The cysts were filled with bloodstained yellow fluid. The walls of the cyst were made up of a tumorous bony shell. As the cysts were evacuated, a large exposure was obtained, enabling excision of the diseased bone around the craniocervical junction on the right side and anterior to the brainstem, including the rim of the foramen magnum, the lower one third of the clivus, the arch and lateral mass of the atlas, the lateral mass of the axis, and the odontoid process. Radical gross total excision of the extradural tumor was performed. Fusion of the grossly unstable craniocervical region was not performed at this stage, but a firm cervical collar was applied. The patient showed rapid neurological improvement following surgery. On the third day, when the patient was being turned to one side, he complained of a crackling sound in the neck and within a few minutes developed quadriplegia followed by respiratory arrest. The collar had been inadvertently removed at this time. He was placed on assisted ventilation but died 24 hours later.
Histopathology showed normal bony trabeculae destroyed by the tumor, which consisted of islands of osteoid tissue rimmed by osteoblasts with collagenous stroma. There were also large vascular spaces lined by endothelium and osteoblastic giant cells.

Case Illustration 15

Osteogenic Sarcoma of the Posterior Cranial Skull Base

A 62-year-old man was diagnosed as having a large ossified tumor involving the clivus, atlas, and occipital bone (Fig. 49.20). Both occipital condyles and lateral masses of the atlas were involved by the tumor. The tumor extended laterally to involve the petrous bones and the region of the jugular foramen. The patient presented with
symptoms of unilateral lower cranial nerve involvement and severe neck pain. Transoral biopsy revealed that the tumor was an osteogenic sarcoma. Occipitocervical fixation was done with screws in the occipital bone and in C1 and C2 lateral masses.

Osteogenic sarcomas are rare skull base tumors. The extensive destruction of the clivus, occipital bone, and atlas in our case is rare and a terminal feature.

**Case Illustration 16**

**Pancervical Vertebral Fusion and Craniovertebral Instability**

This 19-year-old female patient presented with complaints of short neck since birth. For ~4 months, she had severe neck pain. There was no neurological deficit. Investigations showed fusion of C2–C7 vertebrae (Fig. 49.21). Dynamic imaging showed evidence of moderate craniovertebral atlantoaxial instability. Considering the presence of short neck and possible surgical difficulties in surgery, as well as the relatively minor presenting symptoms, surgery to stabilize the region was not accepted by the patient and her family.
Fig. 49.21a–e
a  CT scan showing extensive fusion of the cervical vertebrae.
b  T1-weighted MRI showing cervical fusion and soft tissue alterations.
c  T2-weighted MRI.

Fig. 49.21d–e
Deformation of the Cord due to Posterior Longitudinal Ligament Ossification

A 52-year-old woman presented with a 2-year history of progressive quadriparesis. Images showed degenerative arthritis related to atlantoaxial dislocation and basilar invagination. Also seen were mobile atlantoaxial dislocation and persistent deformation of the cord in extension (Fig. 49.22). The cord deformation persisted in extension of the head. The patient underwent craniovertebral realignment surgery.

**Case Illustration 17**

**Deformation of the Cord due to Posterior Longitudinal Ligament Ossification**

A 52-year-old woman presented with a 2-year history of progressive quadriparesis. Images showed degenerative arthritis related to atlantoaxial dislocation and basilar invagination. Also seen were mobile atlantoaxial dislocation and persistent deformation of the cord in extension (Fig. 49.22). The cord deformation persisted in extension of the head. The patient underwent craniovertebral realignment surgery.

**Fig. 49.22a, b**
- **a** T2-weighted MRI showing atlantoaxial dislocation, basilar invagination, and Chiari malformation. Note the location of the posterior longitudinal ligament.
- **b** Image with the neck in extension. The deformation of the anterior surface of the cord by the posterior longitudinal ligament persists. Note the kinking and laxity of the posterior longitudinal ligament.
Posterior Fossa Arachnoid Cyst Manifesting as Chiari I Malformation and Syringomyelia

A 29-year-old man presented with headache and neck pain for 4 years. He had imbalance while walking and difficulty in performing activities with his hands for 6 months. There were no bowel or bladder complaints. On admission, there was hypotonia in both hands and wasting of the small muscles of the hands. There was mild spasticity in the legs. The patient exhibited decreased sensation of pain and temperature below the level of C4. MRI revealed a posterior fossa arachnoid cyst that herniated across the foramen magnum along with the cerebellar tonsils to result in Chiari I malformation (Fig. 49.23). In addition, there was a large syringomyelia that extended from C2 to D11. The patient was operated upon. Foramen magnum decompression with marsupialization of the posterior fossa arachnoid cyst was performed. Postoperatively, the patient's neck pain and headache improved. The power in both of his upper limbs also improved.

The association of arachnoid cyst and Chiari I malformation is rare.

Fig. 49.23a–d
a T1-weighted MRI showing a large posterior cranial fossa arachnoid cyst that herniated down across the foramen magnum along with the cerebellar tonsils. Large syringomyelia is seen.
b T2-weighted MRI showing the arachnoid cyst, Chiari I malformation, and syringomyelia.
c T1-weighted axial MRI showing the arachnoid cyst.
d T2-weighted image.
Posttraumatic Translatory Atlantoaxial Dislocation

A 28-year-old man fell from a height of 25 feet while trekking in a mountain range. He was brought to the hospital in a cervical collar by air ambulance. Investigations revealed type 2 odontoid fracture and posterior and left lateral translation of the atlas over the axis (Fig. 49.24). The facets of the atlas and axis were not in alignment, and their articular surfaces were not in direct contact with each other. Except for neck pain, there was no other symptom. Neurological examination did not reveal any deficit. Cervical traction was attempted, but the patient developed severe giddiness following its application, and the procedure was abandoned. For surgery, the patient was placed in the prone position. Considering that the patient did not tolerate traction, Gardner-Wells tongs were positioned, but weights were not applied. The basic surgical methods were the same as discussed in our earlier publications on the subject and are summarized here. The atlantoaxial region was exposed, and the dissection was extended laterally until the atlantoaxial joints on both sides were widely visualized. The C2 ganglion was sectioned to enhance the exposure. The posteriorly displaced atlas facets were identified. By extending the dissection on the undersurface of the facet of the atlas, the facet of the axis was exposed. At this stage, traction weights (5 kg) were applied. Traction visually assisted in partially realigning and distracting the facets. The facets of the atlas and axis were directly and manually distracted and realigned using an appropriate-size osteotome that was placed with its flat end into the joint and then turned 90° to effect distraction and reduction. Bone graft pieces were stuffed into the distracted joint. Plate and screw fixation was then performed. The C1 screws were inserted first and tightened. The C2 screw was then tightened to effect reduction in a manner similar to that described for reduction of spondylolisthesis. The patient tolerated surgery well. Postoperative images showed satisfactory realignment of the atlantoaxial joints (Fig. 49.25). At 28-month follow-up, the patient was asymptomatic.

Fig. 49.24a–c

a Sagittal cut of CT scan showing type 2 fracture of the odontoid process. The body of the axis is dislocated anteriorly in relationship to the fractured odontoid segment.

b Three-dimensional CT scan showing posterior and left lateral translatory dislocation of the atlas over the axis. The articular surfaces of the atlas and axis are not in direct alignment.

c Coronal CT scan showing the lateral and posterior translatory dislocation of the atlas over the axis.
In the presented case, the patient had translatory atlantoaxial dislocation, and the atlas and axis were malaligned following fracture of the odontoid process. Considering that the patient was in an intact neurological condition, the entire management was more complex. The facets of the atlas and axis were directly manipulated using the technique of facetal distraction. The method of screw insertion and tightening was modified to effect reduction of the translatory dislocation. A similar technique has been used for reduction of lumbosacral spondylolisthesis.\(^\text{19,20}\)

The screw in the atlas was tightened first. The subsequent tightening of the C2 screw over a taut plate assisted in further reduction of the translatory dislocation. The sequence of screw insertion and tightening was the reverse of what is done in cases with irreducible atlantoaxial dislocation, in which the atlas vertebra is dislocated anteriorly in relationship to the axis. The joint exposure in this case was relatively difficult due to the severe malalignment. Sectioning of the C2 root ganglion appeared necessary to provide a wide surgical field, and the entire manipulation procedure could be performed under direct vision.
Trifid Arches

A 4-year-old boy had a history of bronchopneumonia at the age of 6 months. He presented with progressive difficulty in walking, stiffness of all four limbs, and loss of bladder and bowel control for the past 9 months. At the time of admission, he was bedridden and unable to walk. On examination, he had spastic quadriparesis, with the left side being worse than the right. Power in the left-sided limbs was grade 3 and grade 4 in the right-sided limbs. Investigations revealed atlantoaxial dislocation, with trifid arch of the atlas (Fig. 49.26). The patient was operated, and C1–C2 lateral mass plate and screw fixation was performed. Postoperatively, the patient showed improvement in stiffness and the power of all four limbs. At 18-month follow-up, the patient was able to ambulate independently.

Fig. 49.26a–g

a Axial image showing the atlas bone and its trifid abnormal nature.

b Sagittal T1-weighted MRI showing atlantoaxial dislocation and syringomyelia.

c T2-weighted image.

d Radiograph with the head in flexion showing atlantoaxial dislocation.
Trigeminal Neuralgia in the Presence of Ectatic Basilar Artery and Basilar Invagination

A 65-year-old woman who was otherwise healthy and had no systemic illness presented with the complaints of trigeminal neuralgia for a period of 7 years. She had a short neck since birth. Drug trial with carbamazepine was successful for a period of ~2 years but subsequently failed. The neuralgic pain became progressively unbearable. In the interim, all the molars and premolars of the upper jaw on the ipsilateral side were removed, as they were considered to be causative of pain. When admitted, the pain was unbearable, and she refused to remove a blanket covering her face. Even a whiff of air over her face resulted in severe neuralgic pain in the distribution of V2 and V3 divisions of the trigeminal nerve. Neurological examination was conducted with difficulty, as the patient did not allow testing for sensations over the face. There was no neurological deficit. MRI and CT scan revealed an ectatic basilar artery that clearly indented into the root entry zone of the fifth nerve (Fig. 49.27). Multiple vertebral segments of the cervical spine were identified to be fused, the cervical lordosis was obliterated, the clivus had significant agenesis, and the odontoid tip reached almost to the sella. Considering the presence of marked craniovertebral anomaly and clear evidence of reduction in the posterior fossa volume, foramen magnum decompression was performed. During surgery, bone decompression was difficult due to the small posterior fossa, with the external occipital protuberance reaching almost to the horizontal level of the foramen magnum. The posterior rim of the foramen magnum had an unusual inward and deep curve, and the distance between the large and unusually shaped spinous process of the C2 vertebra was markedly reduced. Following surgery, there was immediate relief from the neuralgic pain. At 18-months follow-up, the patient had no recurrence of the neuralgia.

Microvascular decompression has been the accepted modality of treatment for trigeminal neuralgia. Even in cases of local or remote tumors, it has been speculated that vascular compression at the root entry zone ultimately causes the pain. Resection of the tumor, with or without direct manipulation of the vascular loop, has been associated with lasting relief from pain. Trigeminal neuralgia has only rarely been identified in the presence of clinical situations that result in an obvious reduction in the posterior cranial fossa volume. It is unclear from all the
reported cases if the reduction in the posterior cranial fossa volume ultimately results in the vascular loop indentation at the root entry zone of the fifth cranial nerve, or if the association of the two was an incidental observation. “Kinking” of normal-length arteries that have to adapt to a smaller posterior fossa in a case with basilar invagination associated with osteogenesis imperfecta has been identified earlier. Microvascular decompression has been seen to be successful in some of the reported cases where there was trigeminal neuralgia in the presence of a small posterior cranial fossa.

In this present case, basilar invagination was clearly associated with the presence of an ectatic basilar artery that resulted in fifth cranial nerve root entry zone compression. The therapeutic surgical option of either microvascular decompression or posterior fossa decompression was a philosophical dilemma. Because foramen magnum decompression resulted in lasting relief of symptoms, it does appear that the ectatic basilar artery was secondary to reduced posterior cranial fossa volume.
Unusual Bone Formation in the Anterior Rim of the Foramen Magnum: Cause, Effect, and Treatment

A 19-year-old female patient had traumatic neck injury when she fell on the back of her head while playing 3 years before presentation for treatment in our department. Following this fall, she developed weakness of the right-sided limbs and urinary retention. Her symptoms gradually improved completely in ~4 months. Three months prior to admission, she again had an accidental fall while walking. Following this, she developed quadriplegia. The weakness now progressed. When admitted, she was bedridden and could not perform any useful activities by herself. She also had urinary retention, for which she was catheterized. Clinical examination revealed spasticity in all four limbs. There were no definite sensory deficits. Investigations revealed an abnormal bone growth in the inferior end of the clivus that extended laterally. The tip of the odontoid process also had a posteriorly curving bone growth. The posterior arch of the atlas was not identified and was probably assimilated. The abnormal clival and odontoid bone and the posteriorly buckled tectorial membrane resulted in severe cord compression (Fig. 49.28). Transoral decompression that involved resection of the inferior end of the clivus and the abnormal bone of the odontoid process was performed. The abnormal bone was relatively soft. The patient was then turned prone, and posterior plate and screw fixation of the

Case Illustration 22

**Fig. 49.28a–c**

a CT scan reconstruction image showing the abnormal bone arising from the inferior end of the clivus and tip of the odontoid process. The posterior arch of the atlas is not identified.
b Sagittal T1-weighted MRI showing compression of the cord by the abnormal bone growth.
c T2-weighted MRI showing the abnormal bone and ligamentous mass, as well as neural compression.
C1 lateral mass and C2 pars was performed. Following surgery, the patient showed dramatic neurological recovery. At 23-month follow-up, she was able to walk unaided and had re-entered school (Fig. 49.29).

Although rare, proatlantal segmentation anomalies have been described. The hypocentrum of the fourth occipital sclerotome (proatlas) forms the anterior tubercle of the clivus. The centrum forms the apex of the dens. The rostral ventral neural arch contributes to the formation of the occipital condyles, third condyle, and anterior rim of the foramen magnum. The dorsal caudal part of the neural arch forms the posterior arch of the atlas and the lateral atlantal masses. The persistence of an anterior tubercle from the anterior margin of the foramen magnum represents the hypocentrum abnormality of the proatlas. Our patient had abnormalities of the posterior arch of the atlas in addition to the third condyle, signifying a segmentation anomaly of the neural arch of the proatlas.

The abnormality can be suspected when there is an abnormal bone growth in continuity with the inferior edge of the clivus. Although not done in this case, three-dimensional CT scan can show the anomaly clearly. The bone growth may not always be large, and neural compression may not be identified; some cases may be incidentally detected. Anterior transoral decompression followed by posterior fixation appears to be a rational method of treatment. Surgery results in rapid and sustained neurological recovery.

Fig. 49.29a, b
a Postoperative CT scan showing anterior decompression. Artifacts related to stainless steel implant are seen.

b Sagittal image through the lateral mass showing plate and screw fixation.

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