Pediatric Cervical Spine Instability

Editor:
Pedro K. Beredjiklian, MD
Associate Professor of Orthopaedic Surgery, Thomas Jefferson University Hospital, Chief, Division of Hand Surgery, The Rothman Institute, Philadelphia, PA

Harish S. Hosalkar MD, MBMS (Orth), FCPS (Orth), DN B (Orth)
Attending Orthopedic Surgeon, Clinical Associate Professor of Orthopedic Surgery, School of Medicine, University of California–San Diego; Co-Director of International Center for Pediatric and Adolescent Hip Disorders, Director Hip Research Program, Pediatric Hip and Trauma Specialist, AONA Faculty for Pediatric Orthopedic Trauma, Rady Children’s Hospital, University of California–San Diego

Denis S. Drummond, MD
Attending Surgeon and Emeritus Chief, Department of Orthopaedic Surgery, Children’s Hospital of Philadelphia; Professor of Orthopaedic Surgery, University of Pennsylvania School of Medicine, Philadelphia, PA

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NOTE FROM THE PUBLISHER:
This publication has been developed without involvement of or review by the American Board of Orthopaedic Surgery.
Introduction

Most pediatric spine injuries occur in the cervical spine, the majority of which involve the upper cervical spine. Although cervical spine trauma accounts for only about 1% of traumatic injuries in children,\(^1\,^2\) it can be life-threatening. Knowledge of the unique anatomy and biomechanics of the pediatric cervical spine forms the basis for accurate diagnosis and appropriate treatment.

Anatomy and Biomechanics

The pediatric cervical spine differs significantly from that of adults, and many of these unique features unfavorably affect spinal stability. The combination of an immature skeleton, ligamentous laxity, and a relatively large head size in children has a detrimental effect on cervical spine stability. Differences are notable in terms of injury as well: the effects of any cervical injury are magnified, particularly in the upper cervical spine, where the relatively large head of younger children increases acceleration forces applied to the spine.

Anatomy

The cervical spine in children can be divided into the upper cervical spine, which extends from the occiput to the C2–C3 disk space, and the lower cervical spine, which extends caudally from the same disk space. The embryology, development, and anatomy of the upper cervical spine differ from the lower cervical spine, which develops similarly to the rest of the spine. Compared with the lower cervical spine, the upper cervical spine has a greater dependence on the ligament complexes to provide stability. Also, the relatively larger head size of younger children negatively impacts stability and response to injury. This combination of factors creates a setting for ligament failure in the proximal cervical spine when large mechanical loads occur in high-impact injury. In addition, the dynamic stability of the pediatric cervical spine is challenged by weaker support from adjacent muscles that are smaller and less developed than in the adolescent or adult.\(^3\)

The anatomy of the pediatric cervical spine is constantly changing because of the growth, maturation, and ongoing ossification of vertebral bodies. The presence of the polar growth centers adjacent to the vertebral end plates and the many synchondroses of the immature skeleton determine the sites where vertebral injury is likely to occur. For example, the basilar synchondrosis caudal to the base of the dens is a common site of fracture displacement.
of the odontoid process in younger children. The facets and articulations of the spine of the younger child are shallow, making them less resistant to high loads and more susceptible to failure and instability. In addition to affecting cervical spine stability, the immature and changing anatomy complicates radiographic evaluation of children with traumatic injury or congenital vertebral anomalies.

**BIOMECHANICS**

The comparative ligamentous laxity in children has an adverse effect on stability of facet joints as well as on atlanto-occipital and atlanto-axial joint competency. Furthermore, the facet joints and condylar development of the atlanto-occipital articulation are relatively shallow in children. Accordingly, when acceleration and deceleration forces occur, children are more vulnerable to injury primarily to the spinal column and, secondarily, to the spinal cord, which is at risk for serious traction injury. Even when the spine appears normal on plain radiographs, with failure of supporting soft tissues the stability of the disks, facet joints, and synchondroses may fail. Although the pediatric spine can tolerate subluxation and distraction, the pediatric spinal cord cannot. When stretched beyond tolerance, myelopathy ensues. This phenomenon is termed *spinal cord injury without radiographic abnormality* (SCIWORA).

**DEFINITION OF INSTABILITY**

The parameters commonly used to define normal spine stability are slightly greater for children than for adults. Further, an acceptable increase of any measurement that defines stability is dependent not only on the measurement itself but also on the space available for the spinal cord. The measurement method and established normal value as observed on lateral flexion–extension radiographs follow:

- **Atlanto-occipital motion.** Instability in translation is best observed on the lateral extension view. Of the 3 commonly used parameters, the Powers ratio (Figure 1) and Harris technique best measure horizontal translation, whereas the Kauffmann method measures vertical displacement (Figure 2). The upper limit of normal for horizontal translation in children is 2 mm (versus 1 mm in adults); vertical displacement should not exceed 5 mm in both adults and children (> 5 mm is defined as atlanto-occipital dislocation [AOD]).

- **Atlanto-axial motion.** Atlanto-axial instability is best observed in flexion on the lateral radiograph, and the atlanto-dens interval is accepted as the standard method of measurement (see Figure 2). The accepted upper limit of normal for this index is usually 5 mm in children (versus 4 mm in adults).5

![Figure 1. Use of Power's ratio to determine atlanto-occipital subluxation. In patients without subluxation, BC : AO : 1. A = anterior arch of the atlas; B = basion; C = posterior arch of atlas; O = opisthion. (Adapted with permission from Drummond DS. Pediatric cervical instability: diagnosis and treatment concepts. Semin Spine Surg 1996;8:296.)](image-url)
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Subaxial motion. There is no accepted norm for subaxial translation to define instability in children, although some consider more than 2 mm as abnormal (versus 3.5 mm in adults). Subaxial translation is best appreciated radiographically on the lateral flexion view. Again, the available spinal cord space is an important consideration when evaluating children who have more than 2 mm of subaxial translation.

EVALUATION OF CERVICAL INSTABILITY

In the acute trauma situation, great care must be taken during evaluation, particularly in cases of head injury or with signs of neurologic impairment. With the alert and cooperative patient who shows no signs of spinal cord injury, stability can be confirmed with dynamic lateral flexion–extension radiography. For unconscious or obtunded patients, flexion–extension examination is unsafe. Neck immobilization by a firm fitted collar is critical, and computed tomography (CT) or magnetic resonance imaging (MRI) is used to evaluate the stability status.

Instability is frequently overdiagnosed in children younger than age 3 years because the incompletely ossified vertebrae may be perceived as translation or pseudosubluxation. Table 1 lists the normal vertebral observations unique to the immature cervical spine that can be mistaken for spinal pathology. By applying the spinolaminar line, the correct diagnosis of stability can be appreciated (Figure 3).

IMMOBILIZATION AND PHYSICAL EXAMINATION

Patients with cervical spine trauma or suspected spinal cord injury should be immobilized in the neutral position to prevent possible further neurologic damage. Younger children (age < 8 years) have disproportionately large heads compared with the

Table 1. Unique Features of the Normal Pediatric Cervical Spine

<table>
<thead>
<tr>
<th>General features involving the entire cervical spine</th>
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<tbody>
<tr>
<td>Secondary centers of ossification of the spinous processes may mimic fractures</td>
</tr>
<tr>
<td>Rounding of anterior vertebral body may give the appearance of a wedge compression fracture</td>
</tr>
<tr>
<td>Horizontal facets and ligamentous laxity allow greater intersegment mobility</td>
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<tr>
<td>Cervical lordosis is decreased</td>
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<tr>
<td>Wider prevertebral soft tissues that may mimic swelling</td>
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<table>
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<tr>
<th>Features of specific anatomic levels</th>
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<tr>
<td>C1 multiple ossification centers may mimic fractures</td>
</tr>
<tr>
<td>Absent ossification of anterior arch of C1 may be mistaken for C1–C2 instability</td>
</tr>
<tr>
<td>C1–C2 atlanto-dens interval may be up to 4.5 mm</td>
</tr>
<tr>
<td>C2 posterior angulation of odontoid (4% of children) may mimic fracture</td>
</tr>
<tr>
<td>Ossiculum terminale may mimic fracture</td>
</tr>
<tr>
<td>Basilar synchondrosis may mimic fracture</td>
</tr>
<tr>
<td>C2–C3 and C3–C4 pseudosubluxation may be mistaken for instability</td>
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</table>

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Figure 3. Four-line analysis of cervical spine radiographs: 1 = tips of the spinous processes, 2 = spinolaminar line, 3 = posterior margins, and 4 = anterior margins of the vertebral bodies. All lines should follow a smooth, even contour. (Adapted with permission from Copley LA, Dormans JP. Cervical spine disorders in infants and children. J Am Acad Orthop Surg 1998;6:207.)

After the patient has been safely immobilized and Advanced Trauma Life Support measures taken, a detailed physical examination should be conducted. Findings that correlate with cervical spine injury include head injury, neurologic deficit, torticollis, neck guarding, tenderness, palpable step-off between contiguous spinous processes, and multiple trauma. Skin abrasions over the neck area (shoulder-belt sign) may suggest underlying cervical spine or vertebral artery injury. Serial assessments may be necessary in children who have difficulty cooperating with the physical examination.

RADIOGRAPHIC EXAMINATION FOR STABILITY

With a high index of suspicion of a cervical spine injury, an anteroposterior (AP), lateral, and open-mouth odontoid view of the cervical spine should be obtained. In conscious and alert patients, voluntary flexion and extension views of the cervical spine may be performed to assess spine stability, provided that no abnormalities are seen on static films.

CT may serve as an adjunct to the radiographic evaluation of the bony cervical spine to reduce the risk of missing a fracture on plain radiographic evaluation alone. Helical CT is recommended in high-risk older children or adults who have sustained multisystem trauma after significant blunt injuries. Viccellio et al,7 in a prospective multicenter study of cervical spine injury in 3065 patients younger than 18 years, found that advanced cervical spine imaging may not be necessary in adolescents (age > 9 years) in the absence of high-risk criteria that include: midline cervical tenderness, altered level of alertness, evidence of intoxication, neurologic abnormality, and presence of painful distracting injury.

In unconscious patients, plain radiographs should be obtained but advanced imaging may be necessary to reduce the incidence of missed fractures. It is critical to note that lateral flexion–extension radiography is unsafe in the unconscious or very young child (age < 3 years). The relative ligamentous laxity in children may allow the cervical spine to deform more than the spinal cord can tolerate, and SCIWORA may occur. The flexion–extension maneuvers can also potentially lead to spontaneous reduction, and significant pathology could be missed. MRI is recommended in this situation. MRI can be highly effective...
Table 2. Causes and Subtypes of Cervical Instability

**Congenital causes**

Vertebral (bony anomalies)
- Cranio-occipital defects (occipital vertebrae, basilar impression, occipital dysplasias, condylar hypoplasia, occipitalized atlas)
- Atlanto-axial defects (aplasia of the atlas arch, aplasia of odontoid process)
- Subaxial anomalies (failure of segmentation and/or fusion, spina bifida, spondylolisthesis)

Ligamentous
- Osteochondrodysplasias (achondroplasia, spondyloepiphyseal dysplasia)
- Storage disorders (mucopolysaccharidoses)
- Combined anomalies (at birth as an element of somatogenic aberration)

Syndromic disorders/developmental dysplasias
- Down syndrome
- Klippel-Feil syndrome
- 22q11.2 deletion syndrome
- Marfan syndrome
- Ehlers-Danlos syndrome

**Acquired causes**

Trauma (Hangman fracture, odontoid fracture, atlanto-occipital dislocation)
- Iatrogenic (cervical laminectomy)
- Infection (pyogenic/granulomatous)
- Tumor (osteoid osteoma, osteoblastoma, aneurysmal bone cyst, Langerhans cell hystiocytosis)

Inflammatory conditions (juvenile rheumatoid arthritis)


Table 3. Common Types of Pediatric Cervical Spine Trauma

**Atlanto-occipital trauma**
- Atlanto-occipital dislocation
- Jefferson’s fracture
- Odontoid fracture

**Atlanto-axial trauma**
- Hangman’s fracture
- Traumatic spondylolisthesis
- Atlanto-axial subluxation
- Atlanto-axial rotational dislocation

**Subaxial trauma**
- Traumatic subluxation with or without fracture
- Traumatic fracture dislocations
- Anterior/ posterior/ lateral element fractures in isolation or combination

**Spinal cord injury without radiographic abnormality (SCIWORA)**


focuses on some of the most common conditions leading to instability.

**TRAUMATIC CAUSES**

Table 3 outlines common cervical spine injuries in children. Children younger than 8 years of age usually have upper cervical injuries, whereas older children, whose biomechanics more closely resemble those of adults, are prone to lower cervical injuries. Neonatal high cervical spinal cord injury may result from rotational forceps delivery exerting torsional forces that can cause quadriplegia, areflexia, and diaphragmatic paralysis. Other neonatal risk factors include breech position, which can place traction forces on the spine and cause injury, typically at the cervicothoracic junction.

Cervical spine trauma in children is more likely to be associated with concomitant head trauma,
a factor contributing to the higher mortality rates in children than in adults. Mortality is higher in patients with upper versus lower cervical spine injuries and is highest in patients with AOD. Children also more often present with cervical injuries that are ligamentous in nature, and they are more prone to SCIWORA.\(^8\)

**Atlanto-Occipital Trauma**

**Atlanto-occipital dislocation.** AOD is a rare but frequently fatal injury; however, timely and aggressive management may lead to survival.\(^9\) In a study of pediatric patients with AOD, those who could moan or phonate spontaneously and/or on demand at the time of intubation regained considerable neurologic function.\(^9\) This finding is thought to indicate incomplete brainstem injury with preservation of neurologic structures that support spontaneous ventilation. In these cases, it is important to stabilize the spine with a halo-vest, establish an airway, and administer intravenous steroids using the spinal cord injury protocol to control spinal cord edema.

**Jefferson’s fracture.** Atlas fractures are also rare in children and are best observed on CT axial images. When an atlas fracture does occur, only 1 fracture line may be apparent opposite to a plastically deformed synchondrosis. The finding of lateral masses more than 7 mm wide suggests transverse ligament rupture; thus halo-vest immobilization is urgently required. If the case is more stable (widening < 6 mm), an orthotic collar may suffice.

**Odontoid fracture.** Dens fractures are the most common instability-associated fractures of the pediatric cervical spine. These fractures occur most frequently in children younger than age 7 years through the basilar synchondrosis situated just distal to the base of the dens. Hosalkar et al\(^10\) recently proposed a new classification system for pediatric odontoid fractures based on location in the dens and degree of displacement. Of the odontoid fracture patterns observed, the type in which the fracture ran through the synchondrosis was most likely to be stable on reduction. In these cases, the anterior periosteum is usually partially intact and thus provides some stability and a hinge for reduction. A gentle reduction maneuver in extension against the anterior periosteal hinge followed by immobilization in a halo-vest usually yields a positive result and healing.

**Atlanto-Axial Trauma: Hangman’s Fracture**

Hangman’s fracture with spondylolisthesis at C2 is rare in children but should remain in the differential diagnosis following significant trauma, particularly trauma associated with seat belt injury. This fracture is best diagnosed on CT scans with sagittal and frontal formatting.

**SCIWORA**

SCIWORA is a diagnosis that must be considered in all cases of pediatric trauma. Unexplained neurologic signs, major head injury, and seat belt injuries are some of the related pathologies that should alert the physician to this possibility. MRI can confirm this injury. SCIWORA can be managed by immobilizing the patient in a halo-vest, stabilizing systemic problems, and administering systemic steroids using the spinal cord injury protocol. Spinal stability through the injured segment should be evaluated.

**CONGENITAL CAUSES**

The congenital causes of instability are complex. Table 4 lists anomalies classified by the region involved. These conditions are likely the product of a deficiency in embryogenesis that hinders spinal stability. In addition, reduction of the space
available for the spinal cord—caused by associated spinal stenosis at the unstable segments or encroachment on the space by an intrusive mass—can compound the instability. In Chiari type 1 malformation, for example, this encroachment at the brainstem level occurs as a caudal herniation of the cerebellar tonsils through the foramen magnum, which can cause brainstem-centered neurologic deficiency. Chiari type 1 malformation can also interrupt the normal flow of cerebrospinal fluid, leading to hydromyelia and scoliosis. These conditions can compound the associated instability that is likely in these cases.

Congenital anomalies of the cervical spine are more often multiple than singular. Therefore, if one anomaly is observed, the physician should search for other anomalies, not only in the cervical spine but in thoracic and lumbar levels as well. In such cases, congenital problems may also exist in other organ systems; thus, collaboration with other pediatric specialists for a thorough workup is often needed.5,11

### Canal Encroachment

Narrowing of the bony spinal canal (spinal cord ≤ 13 mm) has been historically described as canal stenosis.12–15 However, this term is inadequate to include all pathologies that decrease available spinal cord space and therefore the concept of canal encroachment is used. Encroachment of the spinal canal or spinal cord may be caused by static and/or dynamic factors (Figure 4).16 Static encroachment includes focal bony narrowing of the spinal canal (stenosis), a space-occupying mass such as a herniation of the cerebellar tonsils with the Chiari type 1 malformation, or odontoid intrusion through the foramen magnum with basilar invagination. Dynamic encroachment occurs when segmental translational instability encroaches on available space for the spinal cord or brainstem. Either condition alone can increase risk for neurologic injury; however, together these conditions are associated with a greater risk.16 Finally, with time and degenerative changes (by approximately the fourth to fifth decade), canal encroachment may result in a further neurologic deterioration. In complex pathologies of the cervical spine, the concept of canal encroachment enables physicians to better evalu-

<table>
<thead>
<tr>
<th>Table 4. Congenital Anomalies of the Upper Cervical Spine</th>
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<tr>
<td><strong>Bony anomalies of the occipitocervical region</strong></td>
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<tr>
<td>Malformations of occipital bone</td>
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<tr>
<td>Occipital vertebrae</td>
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<tr>
<td>Basilar impression</td>
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<tr>
<td>Occipital dysplasia</td>
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<tr>
<td>Condylar hypoplasia</td>
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<tr>
<td>Malformations of atlas</td>
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<tr>
<td>Occipitalization of atlas</td>
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<tr>
<td>Aplasia of the atlas arch</td>
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<tr>
<td>Clefts in the atlas arch</td>
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<tr>
<td>Malformations of axis</td>
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<tr>
<td>Persistent os terminale</td>
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<tr>
<td>Os odontoideum</td>
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<tr>
<td>Dysphasia, hypoplasia, and aplasia of dens</td>
</tr>
<tr>
<td>Spina bifida of the axis</td>
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<tr>
<td>Fusion of cervical vertebrae</td>
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<tr>
<td><strong>Bony anomalies of the subaxial region</strong></td>
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<tr>
<td>Primary failure of embryonic segmentation (Klippel-Feil syndrome and its variants)</td>
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<tr>
<td>Failures of fusion</td>
</tr>
<tr>
<td>Spina bifida</td>
</tr>
<tr>
<td>Spondylolisthesis</td>
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<tr>
<td><strong>Neurologic anomalies</strong></td>
</tr>
<tr>
<td>Chiari type 1 malformation</td>
</tr>
<tr>
<td>Syringomyelia</td>
</tr>
<tr>
<td>Meningomyelocele</td>
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<tr>
<td>Neurofibromatosis</td>
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Figure 4. (A) Schematic illustration showing the dynamic, static, and combined factors involved in canal encroachment. Some causative factors are outlined. (B) McGregor’s line extends from the superior surface and posterior edge of the hard palate to the most caudal point of the occiput. If the tip of the odontoid lies more than 4.5 mm above McGregor’s line, this finding is consistent with basilar impression. (Courtesy of Harish Hosalkar, MD.)

Figure 5. The 3 zones of occipitalization of the atlas vertebra. Z1 = anterior arch; Z2 = lateral mass; Z3 = posterior arch. (Adapted with permission from Gholve PA, Hosalkar HS, Ricchetti ET, et al. Occipitalization of the atlas in children. Morphologic classification, associations, and clinical relevance. J Bone Joint Surg Am 2007;89:572.)

Occipitalization of the Atlas

Occipitalization is defined as partial or complete congenital fusion of the occiput and atlas.17 The reported prevalence of occipitalization in the general (adult and pediatric) population ranges from 0.08% to 2.76%, with males and females equally affected.16 Occipitalization ranges from complete osseous fusion of the atlas and occiput to a partial osseous bridge uniting a focal area of the bones. The clinical spectrum of occipitalization varies from an asymptomatic condition to a condition causing neck pain and stiffness with varying degrees of neurologic deficit.

Pediatric atlas occipitalization is classified into zones based on whether segmentation failure occurs in the anterior arch, the lateral masses, or the posterior arch of the atlas (Figure 5).16 Zone 1 occipitalization represents fusion involving the anterior arch; zone 2 occipitalization corresponds to fusion involving the lateral masses; and zone 3 occipitalization represents fusion involving the posterior arch. Fusions involving more than 1 atlas zone are classified as a combination of zones. Within each zone, occipitalization can be partial or complete and unilateral or bilateral.
The risk of atlanto-axial instability is particularly high in cases of associated congenital C2–C3 fusion. Two-dimensional sagittal and coronal reformatted CT reconstructions and/or MRI can help to establish the diagnosis and allow categorization of occipitalization in 3 zones, each of which may have a different prognostic implication. Patients who are neurologically stable and have high activity level or decreased spinal cord space should undergo careful restoration of the atlanto-axial relationship in extension and occiput-cervical fusion at the appropriate level. In cases of neural impairment, decompression (suboccipital craniectomy and upper cervical laminectomy) with occiput-to-axis arthrodesis is usually required.

**Basilar Impression**

In this disorder, the axis is displaced cranially and the odontoid is displaced through the foramen magnum, resulting in brainstem encroachment. Basilar impression is commonly associated with other anomalies such as atlas occipitalization, hypoplastic atlas, and Klippel-Feil syndrome. Also, basilar impression may accompany systemic disorders such as achondroplasia, osteogenesis imperfecta, and Morquio-Brailsford syndrome. Rheumatoid arthritis of the upper cervical spine can cause acquired basilar impression.

Most cases of basilar impression remain asymptomatic until the second or third decade of life, when patients may present with headache, neck pain, and neurologic compromise. Motor and sensory loss are common in symptomatic cases. Cerbellar ataxia and lower cranial nerve involvement may occur with dysarthria, dysphagia, nystagmus, and respiratory center compression.

Evaluation should include plain radiographs and MRI. McGregor’s line (see Figure 4), drawn from the upper surface of the posterior edge of the hard palate to the most caudal point of the occipital curve, is a commonly used radiographic measurement. With suspicion of basilar impression on radiography, advanced imaging such as CT is recommended. Treatment may require neurosurgical assistance. Anterior impingement may require fusion in extension or anterior odontoid excision (transoral approach) and stabilization in extension. Posterior impingement may require suboccipital craniectomy and decompression of the posterior ring of C1 and possibly C2, followed by fusion at other appropriate levels.

**Hemi-Atlas**

Congenital unilateral absence of C1 can present at birth or later in childhood/adolescence as a secondary torticollis. CT, MRI, and myelography are the best imaging studies to evaluate this anomaly. Magnetic resonance angiography or plain angiography should be performed prior to surgical intervention, because hemi-atlas is commonly associated with vertebral artery anomalies. Management ranges from observation in subtle cases to surgical intervention in the form of decompression (if the spinal cord is inadequate) with posterior fusion, usually between ages 5 and 8 years.

**Odontoid Anomalies**

Odontoid anomalies have a varied spectrum of presentation from aplasia through different degrees of hypoplasia. These anomalies may lead to atlanto-axial instability because the odontoid is no longer a stabilizing structure. Os odontoideum, currently thought to be traumatic in origin (missed/unrecognized trauma), has features similar to aplasia or hypoplasia of odontoid. The clinical presentation of odontoid anomalies may vary from neck pain and discomfort to frank neural compromise and, occasionally, sudden quadriparesis or death.
resulting from minor trauma. Surgical stabilization is indicated for patients with neurologic involvement, instability on flexion–extension films (supervised), brainstem or spinal cord encroachment, or persistent neck pain.

**Syndromes Associated with Instability**

The most common associated syndromes are Down syndrome, Klippel-Feil syndrome, and 22q11.2 deletion syndrome, all of which can be diagnosed at birth or infancy.

**Down syndrome.** These patients exhibit significant ligamentous laxity, which in the cervical spine can lead to instability with translation of one spinal segment on the adjacent segment. Typically, instability occurs at the atlanto-occipital joint, the atlanto-axial joint, or both joints. Subaxial instability is less common but also occurs. The degree of translation depends on the severity of ligamentous laxity. The risk for neurologic complications depends on the degree of instability as well as the available space for the spinal cord. The translation at the atlanto-occipital and atlanto-axial joints may be significant in these patients, and the spinal cord should be evaluated with MRI in cases of cervical spine instability (> 5 mm translation of the atlanto-occipital articulation or > 7 mm translation of the atlanto-axial joint).

**Klippel-Feil syndrome.** Patients with Klippel-Feil syndrome can present with multiple congenital fusions of the vertebral bodies of the cervical spine. The classic presentation is a short, stiff neck with a low hairline. Instability can occur between 2 fused segments or at the interface of the fused and more normal vertebrae. Acute neurologic complications occur infrequently in children with this syndrome; more often, weakness and clinical signs have a delayed onset, often in middle age, and neurologic sequelae can occur insidiously and progressively. Also, the underlying spinal cord is smaller than normal in this syndrome. This finding might explain the insidious loss of neural function with the delayed recognition reported in some cases of Klippel-Feil syndrome and in other congenital cervical spine problems.

**22q11.2 Deletion syndrome.** This chromosome abnormality is one of the most common genetic syndromes and encompasses a wide spectrum of abnormalities including cardiac, palatal, and immunologic anomalies. In a recent series, upper cervical spine anomalies on plain radiographs were noted in 79 cases of this syndrome. At least 1 developmental variation of the occiput or cervical spine is observed in each case; occipital variations include platybasia and basilar impression. Atlas variations involve dysmorphic shape, an open posterior arch, and occipitalization, whereas axis variations occur as dysmorphic dens and dysmorphic spinous process. A range of cervical vertebral fusions was noted in these patients, most commonly C2–C3. Increased segmental motion as observed by translation in flexion or extension in the cervical spine was noted in more than 50% of patients with this syndrome, and more than one-third of patients have increased segmental motion suggestive of instability at multiple levels.

**POST-SURGICAL INSTABILITY**

Progressive deformity of the pediatric spine may occur with growth following multiple laminectomies. Lonstein et al have reported significant, severe, and progressive spinal deformities after laminectomy in as many as 50% of cases in their series. Risk factors for deformity are extensive laminectomy, neurologic involvement, and young age (usually < 10 years) at surgery. Therefore, pediatric laminectomy should be avoided or per-
Protocols for managing pediatric patients who have acute trauma with an established or potential spinal cord injury should include early treatment in the emergency department (ED) or intensive care unit. In addition, safe transport of the unconscious patient to the operating room and positioning for the surgical procedure must always be carefully considered. Table 5 and Table 6 outline the safe assumptions, diagnostic clues, and precautions for the early management of the pediatric patient with spinal injury.

**PRINCIPLES OF HALO MANAGEMENT**

Use of a halo-vest is a helpful adjunct to managing cervical spine instability. The halo itself can easily be applied in the ED under local anesthesia.
Once in place, the halo is readily connected to a halo-vest or other orthoses. Useful guidelines for halo placement follow.

First, the pediatric skull is thin and areas of maximal thickness differ among patients. Because no skull areas are sure to be safe, CT scans of the skull are useful in patients younger than age 3 years to choose the safest area for pin placement. The temporal artery should be palpated and avoided. Second, the pediatric skull is softer than that of the adolescent or adult. Accordingly, placement of at least 8 halo pins inserted at 2 to 4 in/lb is recommended for patients younger than age 6 years; placement of 4 halo pins inserted at 4 to 6 in/lb is recommended for patients age 6 to 8 years; and placement of 4 halo pins inserted at 6 to 8 in/lb is recommended for older children and adolescents. Complications include pin-tract loosening and infection.

**SURGICAL METHODS FOR STABILIZATION AND ARTHRODESI S**

In acute traumatic cases, initial halo stabilization is followed by surgical stabilization (often after 3–6 weeks), including decompression and/or arthrodesis likely with instrumentation. In cases of chronic instability, patients usually undergo elective arthrodesis with stabilization.

**Atlanto-Axial Arthrodesis and Patient Positioning**

For most of the arthrodesis techniques described below, the patient is positioned prone following application of a halo device. In cases of associated spinal instability, great care in patient transport is critical, and spinal cord monitoring should be used throughout this maneuver. Lateral radiographs confirm the reduction and position of the cervical spine. The halo should then be fixed to the Mayfield frame (type of skull fixation and traction device) or a similar device on the operating table, and lateral radiographs should again verify the reduction.

**Gallie technique.** Exposure is accomplished via a midline incision between the occiput and C3. The bifid spinous process of C2 and tubercle of the occiput are identified by palpation. It is important to avoid overexposure to prevent inadvertent fusion of adjacent levels. The posterior arch of the atlas is identified in the deeper layers, and the periosteum is incised and elevated for passage of sublaminar wires or cables. The vertebral artery can be avoided by taking care to not expose more than 1 cm lateral from the midline in small children and not more than 1.5 cm in adolescents and adults. An 18- or 20-gauge wire or cable can be passed around the arch at this point to secure the bone graft. A bicortical graft is then harvested from the posterior iliac crest and shaped to fit against the posterior aspect of the atlas and around the spinous process of the axis. With the graft in position, the wire loop is folded over the graft and around the spinous process of the axis. The free ends of the wire are then united from the lateral edge of the graft and twisted in the midline, thus securing it and providing cervical stability. Continuous spinal cord monitoring and intraoperative imaging to confirm the reduction and graft position are important. These authors prefer the routine use of a halo-vest to protect the fusion and fixation.

**Brooks arthrodesis.** Exposure for Brooks arthrodesis is the same as that for the Gallie technique. The Brooks fixation differs in that a double 18-gauge wire is passed around the arch of the atlas and lamina of the axis. These authors suggest using braided cable wire because it is soft and relatively safe to pass, resists fatigue well, and provides excellent fixation. Two rectangular grafts 1.25 x 3.5 cm (breadth and length, respectively) are harvested from the iliac crest, and the cancellous surface is prepared to fit into
the C1–C2 interval. The grafts help to prevent hyperextension and contribute to the stability of the Brooks procedure.

**Occipito-Cervical Stabilization and Arthrodesis**

**Stabilization.** Treatment of traumatic atlanto-occipital disruption consists of reduction using gentle skull traction and stabilization in a halo-vest. Care should be taken not to distract the spine. Decompression of large hematomas at the cervicomедullary junction may be necessary. Systemic steroids can be useful to reduce edema. Reduction should be confirmed radiographically and stability achieved by immobilization alone or by arthrodesis (see below).

In most patients, fusion can be restricted to extension from the occiput to C2, thus preserving as much motion as possible in the neurologically intact patient. Bone grafts may be taken from the rib or iliac crest. However, rib grafts can be readily contoured to span 2 or more segments, if that degree of span is needed. In late cases of unreduced dislocation, reduction should not be forcefully attempted; instead fusion in situ with a suboccipital craniectomy (to relieve posterior impingement) is usually sufficient.

**Arthrodesis.** Precautions in positioning are the same as described for occipito-cervical stabilization by halo. The exposure is an extension of that described for atlanto-axial arthrodesis—the midline incision extends from the occiput to the spinous process of C3. There are 2 techniques for arthrodesis of the occiput to C2 or lower. With both procedures, fixation wires or cables are passed through cranial burr holes created as 2 pairs on either side of the midline. The first technique, described in 1995, is based on a shaped structural graft harvested from the iliac crest, which is then fixed by wire or cable to a trough prepared at the base of the occiput and fit over the spinous process of the axis. With the second technique, described in 2001, the structural graft used is from the rib rather than the autogenous iliac crest. Paired autogenous rib grafts are harvested and fixed to the occiput and to the axis by sublaminar wires or braided cable. The rib’s natural curve has the advantage of fitting closer to the cervical spine anatomy and fusion bed. This procedure appears well tolerated and the stability is equivalent to that of the iliac onlay technique. The patient is immobilized in a halo-vest or a 4-post cervical orthosis postoperatively.

**Subaxial Injuries**

These injuries represent only 25% of all pediatric cervical spine injuries. Fractures and subluxations in the lower cervical spine are more common in older children and adolescents. Treatment follows the principles for adult injury. Despite a slightly improved prognosis for ligament healing in children, flexion–extension views should be used after immobilization for patients with subluxation and dislocation to verify healing and stability.

**Subaxial arthrodesis.** When internal fixation is limited by the patient’s size, the Dewar technique appears to work well. A modified procedure, using the button-wire implants developed for scoliosis correction, has been described. Patients are usually immobilized in a cervical collar for 6 to 8 weeks postoperatively.

**Other Surgical Techniques for Spinal Stabilization**

**Lateral mass plate fixation.** In adolescents and teenagers, fixation with lateral mass screws and plates can be considered. Posterior plate fixation may also include the C1–C2 motion segment. The advantage of this fixation is that prolonged immobilization in a halo-vest may not be necessary.
Transarticular fixation. This fixation for C1–2 fusion may be rarely indicated for failed arthrodesis or in case of defective posterior elements and may be performed via the posterior or lateral approach. Screw placement is critical, and good intraoperative imaging is necessary because the margin for error is very small.

Anterior cervical instrumentation. Anterior cervical plating is not routinely used in the pediatric spine. It may have a role in adolescents and teenagers in the presence of significant instability following vertebrectomy or corpectomy. One advantage is that posterior stabilization should not be required. Although there is little experience with this technique in the pediatric population, it is an option for the extraordinary situation.

SUMMARY

The evaluation of a child with a potential injury or instability of the cervical spine begins with the recognition of the unique anatomy and physiology in this age-group. Preventive efforts may reduce the incidence of pediatric spinal injury including: proper use of seat belts, education on safe participation in sports activities, and recognition of the signs of child abuse. Clinicians need to be aware of the common congenital and developmental conditions associated with the occurrence of cervical spine instability to be able to recognize them. Although advanced imaging techniques can be useful in the evaluation of subtle spine injuries and instability, a thorough knowledge of normal anatomic variants is essential for appropriate management.

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