Cervical Spine Disorders in Infants and Children

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Abstract

The evaluation of children with cervical spine disorders requires an understanding of the anatomic and developmental features that are particular to the pediatric spine. In this article, cervical spine developmental anatomy is briefly reviewed, along with common radiographic features of the pediatric cervical spine. The epidemiology, clinical presentation, and management of congenital cervical anomalies are considered. The evaluation and management of pediatric cervical trauma are also reviewed. Other disorders with common cervical spine involvement, such as skeletal dysplasias, connective tissue disorders, inflammatory arthritides, and storage disorders, are discussed.

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Congenital anomalies of the cervical spine, although rare, are worthy of attention because neurologic compromise from instability or stenosis may be prevented with early recognition and careful management of those at risk. Cervical anomalies may also herald the presence of other organ abnormalities, which may be detected with appropriate screening studies such as renal ultrasonography and echocardiography. Traumatic injuries to the cervical spine encompass a broad spectrum from minor soft-tissue injuries to severe fracture-dislocations with spinal cord injury. Safe immobilization techniques, an appropriate algorithm for evaluation, and correct early management of any recognized injuries are essential to optimal care.

Familiarity with the normal developmental anatomy and radiographic features, along with a knowledge of the common manifestations of hereditary and systemic diseases, is prerequisite to understanding the disorders that affect the pediatric cervical spine. Cervical abnormalities occur frequently in skeletal dysplasias, connective tissue disorders, inflammatory arthritides, and storage disorders (Table 1) and require knowledgeable evaluation and treatment.

Developmental Anatomy

Within the past decade, molecular genetic research has identified a family of genes responsible for the early complex differentiation processes of the axial and appendicular skeletons. These genes are referred to as the homeobox, or *Hox*, genes.[1] They direct and regulate the processes of embryonic differentiation and segmentation along the craniocaudal axis by means of activation and repression of DNA sequences that encode the transcription factors and proteins that affect the order and direction of development of the specific tissues of the axial skeleton.[1] Mutations involving the homeobox genes may be responsible for the common congenital anomalies of the cervical spine, as is evident in the murine homeobox gene *Cdx1* and *Hoxb-4 (Hox-2.6)*.[2]

The atlas develops from three ossification centers, one for each lateral mass and one for the body, which does not appear until 1 year of age (Fig. 1).[3] The posterior arches fuse by age 3 or 4 years, but the lateral masses do not fuse to the body until age 7 years.[4] The axis is derived from five primary ossification centers: the two neural arches or lateral masses, the two halves of the dens, and the body. There are two secondary centers: the ossiculum terminale and the inferior ring apophysis.[5] The two halves of the odontoid are generally fused at birth but may persist as two centers known as the dens bicornis.[5] The dens is separated from the body by a dentocentral, or basilar, synchondrosis, which lies well below the level of the superior articular facets, giving the appearance of a

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Table 1
Disorders With Common Involvement of the Cervical Spine

Skeletal dysplasia
Spondyloepiphyseal dysplasia
Achondroplasia
Osteogenesis imperfecta
Neurofibromatosis
Rickets
Diastrophic dysplasia
Connective tissue disorders
Marfan syndrome
Down syndrome
Inflammatory arthritis (juvenile rheumatoid arthritis)
Mucopolysaccharide storage disease (Morquio syndrome)

cork in a bottle on an open-mouth view of the axis, with the dens being the "cork" and the lateral masses and body together forming the "bottle."[3,5] The dentocentral synchondrosis of the axis remains open in all children up to age 3 years, is present in half by age 4 to 5 years, and is absent in most by age 6 years.[5] The tip of the dens is not ossified at birth but appears at age 3 years and fuses to the dens by age 12 years. Occasionally, it remains as a separate ossiculum terminale persistens and is of little clinical significance.[5]

The cervical spine approaches adult size and shape by age 8 years, as the vertebral bodies gradually lose their oval or wedge shape and become more rectangular.[6] There is increasing stability of the cervical spine with age, which explains the different prevalences of cervical trauma in children above and below 8 years of age. A recent study of 227 consecutively treated children with traumatic cervical spine fractures showed that 87% of children younger than 8 years had injuries at C3 or higher and had a higher risk of fatality from their injuries, whereas children older than 8 years had injury patterns similar to those seen in adults, with lesions predominantly below C4, and had no fatalities.[7]

Radiographic Evaluation

Because of the unusual radiographic variability in children, care must be taken in reviewing studies and in correlating the information obtained with the history and physical examination findings in the child. Hypermobility, unique vertebral configurations, incomplete ossification, and the presence of apophyses all contribute to the difficulty in interpreting radiographs of the pediatric cervical spine.[8] Because of the orientation of the facet joints and the relative laxity of the ligamentous and cartilaginous elements of the pediatric cervical spine, an increased amount of cervical vertebral mobility is noted in children less than 8 years of age. Pseudosubluxation is most common at C2-3 (19% of children aged 1 to 7 years) and is also seen at C3-4. The atlas overrides the odontoid in 20% of children aged 1 to 7 years. Absent lordosis (which occurs in 15% of children aged 1 to 16 years) and anterior angulation of the odontoid process (4% of children) are also seen.[8] Incomplete ossification accounts for increased measurements of the basion-dens interval and the atlantodens interval, as well as for the oval or wedge shape of vertebral bodies. The presence of apophyses may lead to the false impression of fracture of the base of the odontoid at the synchondrosis.[5]

Special studies can be used to supplement plain radiographs of the cervical spine in children. Supervised flexion-extension lateral radiographs are useful in the evaluation of instability (congenital or traumatic). Dynamic rotation computed tomographic (CT) scans are helpful in looking for atlantoaxial rotary displacement, and reconstructive CT is useful in evaluating congenital anomalies. Magnetic resonance (MR) imaging (either static or dynamic) is used to evaluate

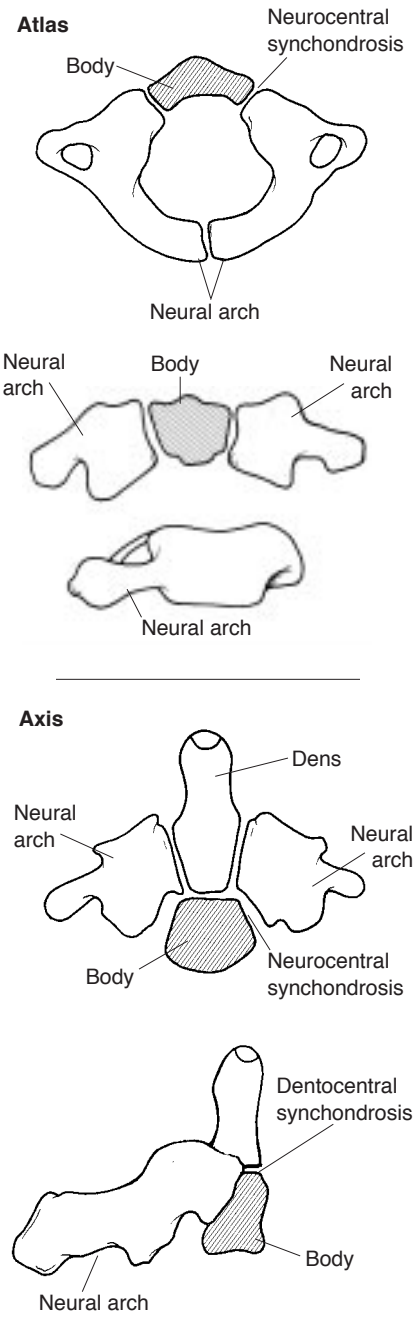

Fig. 1 Ossification centers of the atlas **(top)** and axis **(bottom)** during development.

patients with spinal cord injury without radiographic abnormality (SCIWORA)[9] or cord compression. Occasionally, myelography or CT myelography can demonstrate the presence of dural bands in cases of basilar impression.

Lateral static radiographs of the craniovertebral junction are useful in evaluating occipitocervical anomalies.[10-12] Because of the close relationship of the atlas to the occiput and their tendency to move together, care must be taken to align the x-ray beam perpendicular to the axis of the head, rather than the neck, in order to clearly visualize this relationship.[10] Several measurements are commonly used to demonstrate the relationship between the occiput (foramen magnum) and C1 or the odontoid: the McGregor, McRae, Chamberlain, Wackenheim, and Wiesel-Rothman lines and Power's ratio (Fig. 2).

Evaluation for the presence of instability at C1-2 in children utilizes the atlantodens interval (ADI) and the space available for the cord (SAC) (Fig. 2, A). The ADI should be less than 4 mm in children under 8 years (some consider 5 mm acceptable).[3] In older children and adults, the value should be 3 mm or less.[12] In chronic atlantoaxial instability due to skeletal dysplasia, connective tissue disorders, inflammatory arthritides, and storage disorders, the ADI may be increased at baseline.[13-16] In that case, attention should be directed to the SAC. A careful history and physical examination are necessary to look for neurologic signs and symptoms.

The SAC is roughly defined by the "rule of thirds" proposed by Steel.[12] At the level of the dens, one third of the area within the ring of C1 is occupied by the spinal cord and one third by the odontoid; the remaining third is space. The transverse ligament serves as the first line of defense, maintaining the ADI at 4 mm or less. The alar and apical ligaments have a checkrein effect, serving as the second line of defense (Fig. 3). When the ADI exceeds 10 to 12 mm, all ligaments have failed, the SAC is negligible, and cord compression occurs. These measurements can be correlated with the history and physical examination findings to determine the clinical relevance of the instability.

In pseudosubluxation, as much as 4 mm of stepoff anteriorly may

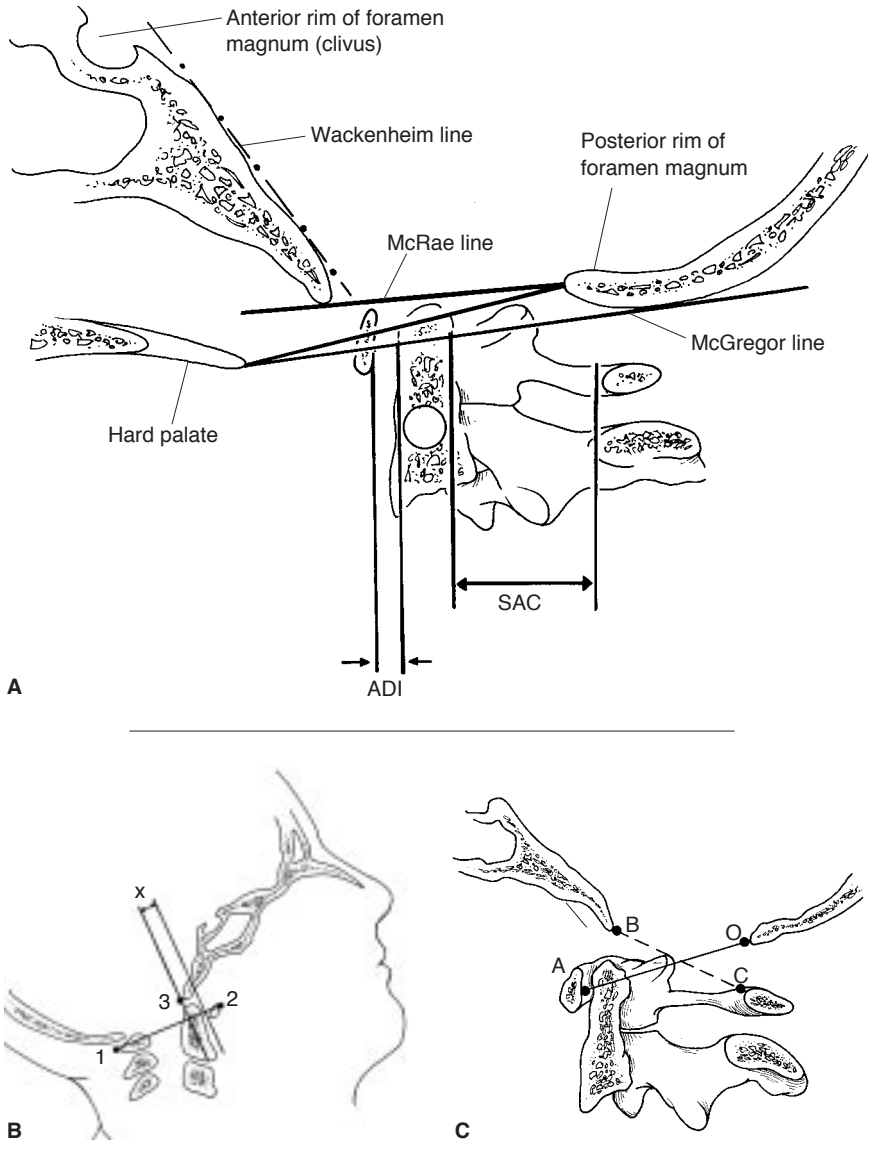

Fig. 2 Lateral craniometry. **A,** The lines commonly used to determine basilar impression and the measurements for determining atlantoaxial instability (ADI = atlantodens interval; SAC = space available for cord). **B,** Method of measuring atlanto-occipital instability according to Rothman and Wiesel. The atlantal line joins points 1 and 2. A line perpendicular to the atlantal line is made at the posterior margin of the anterior arch of the atlas. The distance (x) from the basion (3) to the perpendicular line should not vary more than 1 mm in flexion and extension. **C,** Power's ratio is determined by drawing a line from the basion **(B)** to the posterior arch of the atlas **(C)** and a second line from the opisthion **(O)** to the anterior arch of the atlas **(A)**. The length of line BC is divided by the length of line OA. A ratio greater than 1.0 is diagnostic of anterior occipitoatlantal dislocation.

be seen. The differentiation of this phenomenon from true injury can be facilitated by the use of Swischuk's line.[17] This is drawn along the posterior arch (spinolaminar line) from C1 to C3 (Fig. 4). The line should pass within 1.5 mm of the posterior arch of C2.[17]

The subaxial cervical spine can be evaluated on the basis of characteristics visualized on the lateral plain radiograph. Smooth lines should join the anterior vertebral body line, posterior vertebral body line, posterior arch line, and spinous processes from C1 to C7 (Fig. 5).[18] The articular facets should be parallel, and the interspinous distances and posterior disk spaces should be in balance.[18] The retropharyngeal space should be less than 7 mm, and the retrotracheal space should be less than 14 mm in children; however, these may be difficult to assess in a crying child.[19] The cervicothoracic junction should be visualized in every trauma patient with lateral plain films, a swimmer's view, or a CT study with fine sections through this portion of the spine.

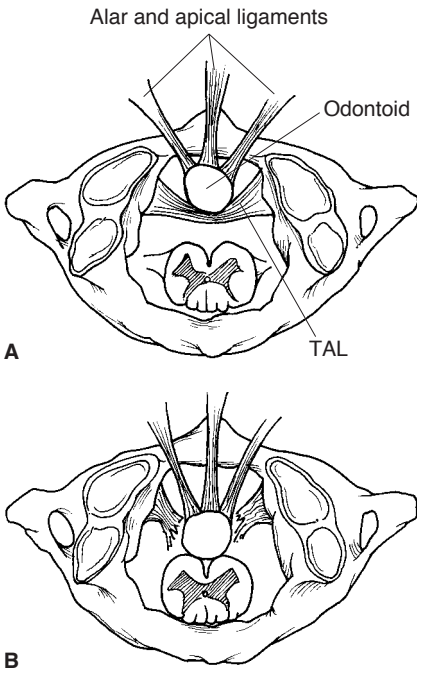

Fig. 3 **A,** Normal relationships of the alar and apical ligaments and the transverse atlantal ligament (TAL) in the atlantoaxial joint, as viewed from above. **B,** The checkrein effect of the alar ligaments prevents cord compression when the TAL ruptures.

Congenital Anomalies

Congenital anomalies range in severity from those that are benign and asymptomatic to those with the potential for fatal instability. Although these anomalies are rare, their recognition is important to prevent catastrophic paralysis due to injury, sports participation, or anesthetic manipulation.

Basilar Impression

Basilar impression and odontoid anomalies are among the most common developmental malformations of the proximal cervical spine.[10] Basilar impression is commonly associated with other anomalies, such as Klippel-Feil syndrome, hypoplasia of the atlas, bifid posterior arch of the atlas, and occipitocervical synostosis.[10] It is also commonly found in systemic disorders such as achondroplasia, osteogenesis imperfecta, Morquio syndrome, and spondyloepiphyseal dysplasia.

With basilar impression, the upper cervical spine encroaches on the brainstem and spinal cord as the base of the skull is displaced toward the cranial vault.[19] Motor and sensory disturbances are noted in 85% of individuals who are symptomatic.[19] However, most affected persons remain asymptomatic until the second or third decade, when they may present with headache, neck ache, and neurologic compromise (prevalence, 15%).[19] Some patients have been misdiagnosed as having multiple sclerosis, amyotrophic lateral sclerosis, posterior fossa tumors, or traumatic injury. Evaluation should include lateral craniocervical radiography followed by MR imaging. Myelography and/or CT with reconstruc-

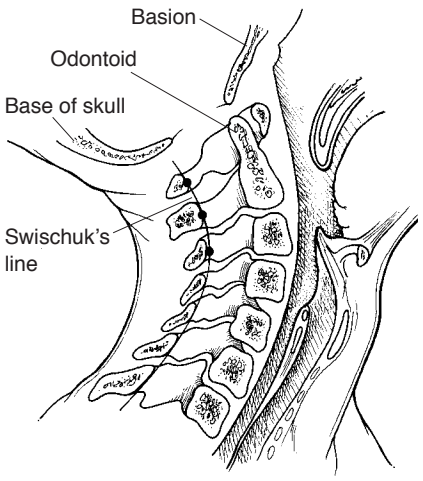

Fig. 4 The spinolaminar line (Swischuk's line) is used to determine the presence of pseudosubluxation of C2 on C3.

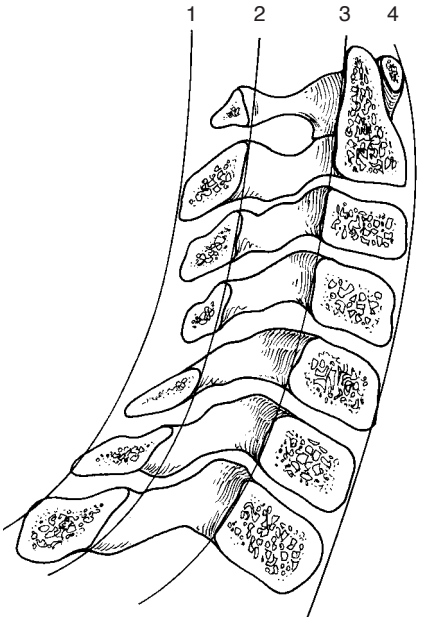

Fig. 5 Normal relationships in the lateral cervical spine: 1 = spinous processes; 2 = spinolaminar line; 3 = posterior vertebral body line; 4 = anterior vertebral body line.

tions may be helpful in some instances to further delineate the pathologic anatomy.

Treatment often requires the combined efforts of the orthopaedic surgeon and the neurosurgeon. Anterior impingement (by a hypermobile odontoid) may require fusion in extension (if reduction and decompression occur in extension) or anterior excision of the odontoid and stabilization in extension.[19] Posterior impingement may require suboccipital craniectomy and decompression of the posterior ring of C1 and possibly C2 with the release of tight dural bands. This is followed by fusion of the occiput to C2 or C3.

Occipitocervical Synostosis

Congenital occipitocervical synostosis ranges in severity from an asymptomatic anomaly to a more serious disorder with the potential for neurologic compromise after minor trauma. In one review,[20] a third of affected individuals were misdiagnosed as having a neurologic disorder. Associated findings of a short neck, low posterior hairline, and limited neck range of motion may be seen, and there is a common association of this disorder with C2-C3 fusion.

Because atlantoaxial instability develops in as many as 50% of patients with this condition, assessment is required.[20] Once instability has been demonstrated, treatment depends on the presence or absence of neurologic signs and symptoms. Neurologically stable individuals who are active in sports or who have a decreased SAC, suggestive of impending myelopathy, should undergo careful restoration of the atlantoaxial relationship in extension and fusion of the occiput-C1 complex to C2.[20] If there is neural impairment, consideration may be given to decompression by laminectomy, craniectomy, and release of posterior dural bands.

Unilateral Absence of C1

Congenital unilateral absence of the first cervical vertebra (hemiatlas) was recently described in a series of 17 individuals.[21] Either the condition presents at birth or torticollis later develops, with lateral translation of the head on the trunk and lateral tilt and rotation of the head. Other anomalies are commonly associated, such as tracheoesophageal fistula.

The deformity is best evaluated with CT, MR imaging, and myelography. Angiography or MR angiography should be done if operative intervention is undertaken, as vertebral artery anomalies are commonly found on the aplastic side.

Initial management involves observation to determine whether progressive head imbalance or loss of cervical range of motion has resulted. Because bracing is not effective in preventing progression, surgical intervention is recommended. Ideally, posterior fusion is done between the ages of 5 and 8 years. Decompression may be necessary if the SAC is inadequate.

Odontoid Anomalies

Anomalies of the odontoid occur on a continuum from aplasia (complete absence) through varying degrees of hypoplasia (Fig. 6). Os odontoideum, currently believed to be initiated by trauma, has clinical features similar to those of aplasia or hypoplasia of the odontoid.[22] These anomalies may lead to atlantoaxial instability with similar signs, symptoms, and treatment. The atlantoaxial joint becomes unstable because the odontoid is not a functional peg.[10]

The clinical presentation occurs at an average age of 19 years, with most patients having cervical pain and a spectrum of findings ranging from very mild discomfort to neurologic compromise and sudden death due to minor trauma.[22] Surgical stabilization is indicated if there is neurologic involvement, more than 10 mm of instability on supervised flexion-extension films, or persistent neck complaints. Prophylactic stabilization is controversial. However, it should be noted that good outcomes have been achieved with surgery, with low morbidity.[22]

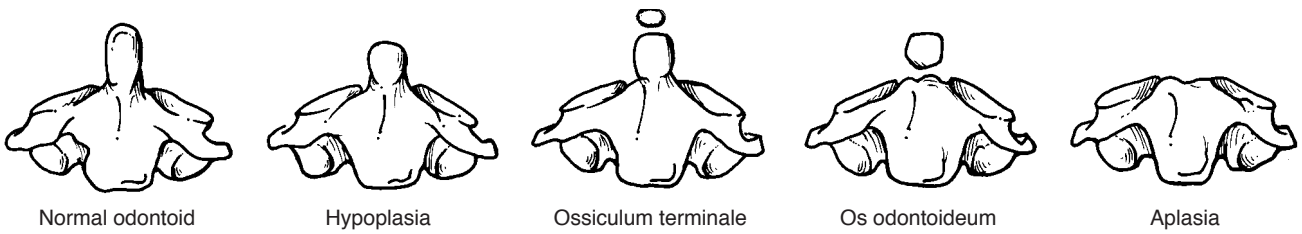

Fig. 6 Gradations of odontoid appearance.

Congenital Muscular Torticollis

Congenital muscular torticollis is the most common cause of a wryneck posture in the infant.[23] It is often discovered during the first 6 to 8 weeks of life. It is thought to be secondary to a "packing syndrome" and is therefore more common in first-born children. It has been associated with other similar conditions, such as developmental dysplasia of the hip (coexisting in 20% of cases) and metatarsus adductus (coexisting in 15% of cases).[10] There is evidence that the process is due to a compartment syndrome involving the sternocleidomastoid muscle, with subsequent intramuscular fibrosis. In nearly 75% of cases, the abnormally tight sternocleidomastoid is on the right side.[24]

Evaluation should include plain radiography of the cervical spine to rule out congenital skeletal anomalies. Care must be taken to consider other causes of this appearance, such as ocular dysfunction, tumors of the spinal cord and cerebellum, infection or inflammation, trauma, and atlantoaxial rotary displacement.[24]

Conservative management with standard passive stretching exercises (Fig. 7) yielded excellent results in 90% of children in one study.[24] However, if there is a limitation of motion of more than 30 degrees or if the condition persists beyond 1 year, surgical release of the sternocleidomastoid muscle may be necessary.[23] In one review of the data on 33 children,[25] distal open release of the sternocleidomastoid muscle led to good results in children between 6 and 12 years of age and acceptable results in children over 12 years.

Klippel-Feil Syndrome

Klippel-Feil syndrome is a congenital fusion of the cervical spine, ranging from a two-segment fusion to involvement of the entire cervical spine. This may be associated with the clinical triad of a short neck, a low posterior hairline, and marked limitation of range of motion of the neck. This condition may be associated with other congenital cervical anomalies, including occipitocervical synostosis, basilar impression, and anomalies of the odontoid.[26] Hensinger et al[26] have found that there are commonly additional associated disorders in these children, including congenital scoliosis, renal anomalies, Sprengel's deformity, impaired hearing, synkinesia, and congenital heart disease. The presentation of individuals with Klippel-Feil syndrome should initiate a thorough evaluation to detect these possibly underlying abnormalities, which may be serious enough to produce considerable morbidity.

Clinically, these individuals often are asymptomatic, and the disorder is discovered incidentally. Those who become symptomatic often present during the second or third decade, when the free segments adjacent to the fused segment become hypermobile, which leads to early degenerative arthritis or frank instability.[10] Generally, upper cervical instability occurs in younger individuals with a high prevalence of neurologic problems, whereas lower cervical hypermobility often occurs in older individuals as degenerative arthritis.[27]

There are three high-risk patterns that commonly produce instability and require early recognition: (1) fusion of C2 to C3 with occipitocervical synostosis; (2) long fusion with an abnormal occipitocervical junction; and (3) a single open interspace between two fused segments.[27] Each of these situations focuses motion at one level, with the abnormal biomechanics producing instability. Treatment by stabilization or modification of activity and bracing to prevent neurologic compromise with minor trauma may be necessary

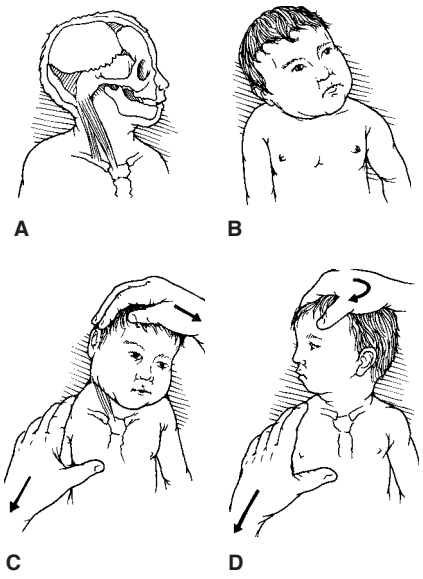

Fig. 7 **A,** Sternocleidomastoid anatomy. **B,** Congenital muscular torticollis. Passive stretching exercises include lateral head bend away from the affected side **(C)** and head rotation toward the affected side **(D)**.

The indications for and timing of stabilization are not clearly defined. However, neurologic compromise and sudden death have been reported with Klippel-Feil syndrome.[10] This risk must be weighed against the disadvantage of losing additional motion segments with a stabilization procedure. Conservative measures, such as activity modification, bracing, and traction, may reduce symptoms and delay surgery for these individuals.

Trauma

Traumatic injuries to the pediatric cervical spine are rare. In a series of 631 children with cervical spine injuries,[28] only 12 (1.9%) were less than 15 years of age. In that series, common causes of injury included motor-vehicle accidents (50%),

sports injuries and child abuse (20%), falls from a height (15%), and gunshot wounds (10%). In children less than age 8, most injuries occur at C3 or higher. Most fatalities due to cervical spine injury occur in this age group.

If injury to the cervical spine due to high-energy trauma is suspected, the child should be immobilized to prevent spinal cord injury. This may be accomplished either by using a spine board with an occipital recess or, more commonly, by placing a mattress beneath the shoulders and trunk of the child (Fig. 8).[29]

The cervical spine should be immobilized with a rigid cervical orthosis specifically designed for children. Sand bags should be used on each side of the head to prevent rotation. Movement should be minimized and done in a log-rolling fashion with gentle in-line cervical control until the spine is cleared of injury. Screening radiographs should include anteroposterior and lateral projections, as well as an open-mouth odontoid view. In a series of 300 patients with multiple trauma and cervical spine injury, 11 had initially unrecognized injuries, and neurologic deficit or death occurred due to inadequate immobilization during emergency management.[29]

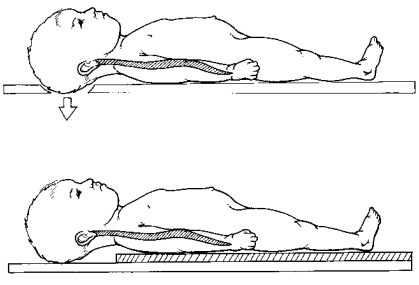

Fig. 8 Young children can be immobilized on a modified backboard with either an occipital recess (less practical) **(top)** or a mattress pad to raise the chest (more practical) **(bottom)**.

Physical examination of the spine may be performed with palpation and inspection to locate sites of tenderness, deformity, or ecchymosis. Range of motion should be evaluated only in conscious, cooperative children. If there is no tenderness and they have full range of motion of the spine, the cervical collar may be removed, and they can be taken off the spine board. If there is any tenderness or limitation of motion, voluntary flexion and extension lateral radiographs can be obtained to rule out instability due to ligamentous injury not detected on the initial radiographs. This should be accomplished only in children who are alert, oriented, neurologically intact, and of an appropriate age to cooperate with the examination. If the study is negative and tenderness persists, a soft collar is recommended for comfort.

In unconscious, uncooperative, or very young children and in children with SCIWORA, MR imaging may be useful for revealing spinal cord, soft-tissue, and bone injuries of the spinal column that are not visible on plain radiographs. If the study is negative, the cervical collar can usually be removed.

Spinal cord injury without radiographic abnormality occurs because of the greater flexibility of the pediatric spinal column compared with the spinal cord; the spinal column is capable of up to 2 inches of elongation, whereas the spinal cord will rupture with only 0.25 inch of stretch.[9] When SCIWORA does occur, careful neurologic evaluation is indicated to document the level of injury, to determine whether the cord injury is complete or incomplete, and to assess for the presence of spinal shock. Magnetic resonance imaging may be useful for identifying the nature of the injury. In children with associated head trauma, somatosensory evoked potentials have also been useful.[30]

Treatment of SCIWORA involves immobilization and frequent reevaluation over 6 to 8 weeks to look for resolution of the injury and to reassess the stability of the cervical spine. Laminectomy has not proved beneficial and may actually increase instability.[30]

Acutely, the use of corticosteroids is believed to improve outcomes in children with spinal cord injury.[31] The recommended dose of methylprednisolone is 30 mg/kg of body weight given as a bolus within the first 8 hours after the injury. This is followed by 5.4 mg/kg per hour for either 24 or 48 hours depending on the time when the treatment was initiated. (This recommendation is based on data from studies involving mostly adult patients.)

Atlantoaxial Rotary Displacement

Torticollis can be a common pathway for the manifestation of inflammation in the cervical spine due to a variety of causes, among them trauma, infection, rheumatologic conditions, and postsurgical inflammation. The spasm and inflammation from these causes may lead to the development of atlantoaxial rotary displacement (AARD), which represents a spectrum of pathologic changes ranging from mild displacement to severe displacement and eventually the fixation of C1 on C2 in a subluxated position.

The Fielding-Hawkins classification[32] is used to evaluate this disorder and guide management (Fig. 9). Expectant management with an initial trial of nonsteroidal anti-inflammatory medication and rest with a soft collar is indicated for up to 1 week after the onset of symptoms. Mild inflammation should resolve spontaneously within this period. A dynamic CT study is

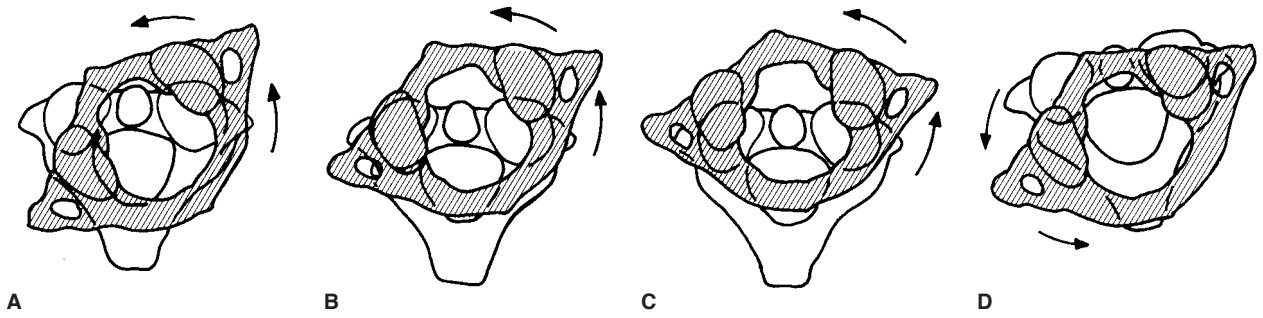

Fig. 9 Fielding-Hawkins classification of atlantoaxial rotary displacement. A = simple rotary displacement; B = rotary displacement with anterior shift of 5 mm or less; C = rotary displacement with anterior shift greater than 5 mm; D = rotary displacement with posterior shift.

obtained to confirm the diagnosis and rule out grade III or IV AARD or atlantoaxial rotary fixation. If symptoms persist, the child should usually be admitted to the hospital for soft halter traction.

Deformity that persists beyond 1 week can be classified as either reducible or fixed. A halo or rigid brace may be used for reducible AARD if the duration of signs and symptoms has been prolonged. Indications for surgical fusion of C1 to C2 include neurologic involvement and failure to obtain or maintain correction despite appropriate conservative measures.

Fractures and Dislocations

Fractures of the atlas are often caused by an axial compression force that is dissipated on the lateral masses and drives them apart. Spinal cord injury is rare in those who survive the injury. Immobilization for 6 to 8 weeks is followed by testing of the integrity of the transverse atlantal ligament as visualized on flexion and extension films. Fusion of C1 to C2 may be necessary in cases of residual instability.

Isolated rupture or avulsion of the transverse ligament may be demonstrated on lateral cervical spine flexion and extension radiographs, CT scans, or MR images. An ADI greater than 5 mm suggests instability of C1 on C2. Unless such an injury is associated with an avulsion fracture, conservative treatment is likely to fail, and surgical stabilization of C1 to C2 is indicated.[33]

Displaced fractures through the ring of C2 are treated with traction reduction and immobilization. Odontoid fractures are the most common pediatric cervical spine injury. Most odontoid fractures in children occur through the basilar synchondrosis and differ from the adult types. If displaced, they can usually be reduced with traction and will heal with halo immobilization for 6 to 8 weeks; supervised flexion and extension films should then be obtained to verify healing and stability. C1-C2 fusion is indicated for persistent nonunion at the fracture site.[34]

Subaxial cervical spine injuries include unilateral and bilateral facet subluxation or dislocation and a spectrum of fractures with varying potential for instability. Facet subluxation or dislocation often presents as 25% (unilateral) to 50% (bilateral) anterolisthesis of one vertebral body with respect to the next caudal level. The treatment of these injuries is dependent on the specific injury and is usually similar to the treatment for adults.[34] If displacement exists, traction may be used to obtain reduction; the type and amount of traction depend on the age and weight of the child. After traction, immobilization for 6 to 8 weeks usually obviates the need for surgery. However, a two-level posterior fusion with wires and bone graft may be needed if there is persistent instability.[34]

Other Disorders With Common Cervical Spine Involvement

Cervical spine problems are more common in persons with certain underlying disorders, such as skeletal dysplasia, connective tissue disorders, inflammatory arthritides, and storage disorders (Table 1).[13-16]

In children with Down syndrome, ligamentous laxity can occur at more than one level, including the occipitocervical level. Instability of C1 on C2 in excess of 5 mm occurs in 14% to 17% of patients, and hypoplasia of the posterior arch of C1 can occur in 26%.[13,35] This may amplify the risk of spinal cord injury following atlantoaxial subluxation because of the decreased SAC (Fig. 10).[35]

Recommendations for management of these children include a

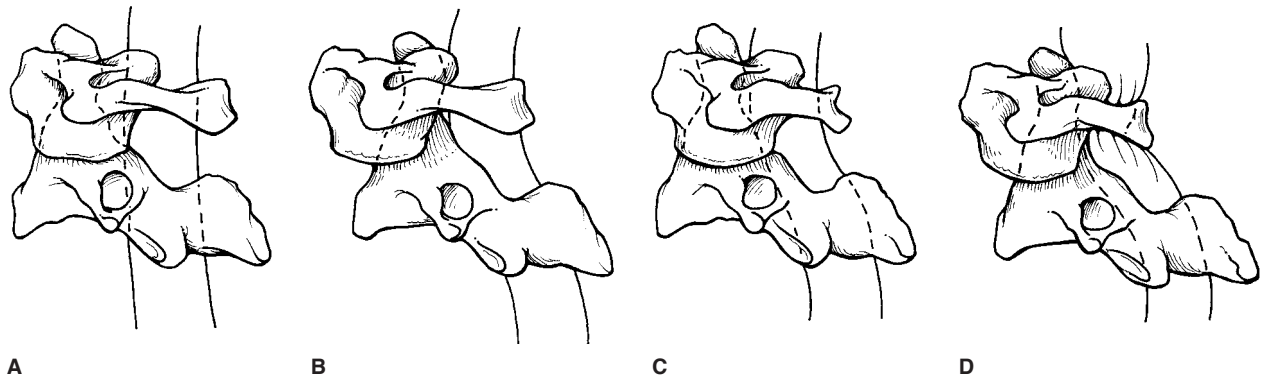

Fig. 10 **A,** Normal relationship of the atlas, axis, and spinal cord. **B,** Atlantoaxial subluxation with normal posterior arch of C1 and adequate SAC. **C,** Hypoplastic posterior arch of C1 with adequate SAC. **D,** Double jeopardy of hypoplastic posterior arch of C1 and atlantoaxial subluxation producing cord compression.

careful baseline radiographic examination, including flexion and extension films, and a careful neurologic examination. Although some have suggested that children with an ADI greater than 5 mm should be restricted from sports that involve a risk of stress to the head or neck, others recognize that fewer than 10% of those with more than 5 mm of instability will have signs and symptoms of cervical spine myelopathy.[14] Rather than restricting these children from all sports, some recommend that they avoid only sports that require stressful weight bearing on the head, such as gymnastics and diving.[14]

Prophylactic stabilization is not indicated. However, patients who become symptomatic or who demonstrate neurologic signs on physical examination should be evaluated more carefully, and consideration should be given to stabilization.[3] This is necessary for fewer than 10% of children with Down syndrome.[13] It should be noted that stabilization is frequently associated with complications, including nonunion.

In children with Morquio syndrome, atlantoaxial instability is more commonly due to hypoplasia of the odontoid, rather than incompetence of the transverse ligament.[15] Myelopathy due to instability is relatively common in these children. Fusion is indicated if there is more than 5 mm of instability, even in children who are asymptomatic. The same is true of children with spondyloepiphyseal dysplasia. Flexion and extension MR images are useful in the evaluation and treatment of these children.

In children with juvenile rheumatoid arthritis (JRA), the increased ADI (greater than 4.5 mm in 20%) is related to deficiency of the transverse ligament due to inflammation, but is not commonly associated with neurologic manifestations or pain.[16] Children with JRA who present with pain should be evaluated carefully for fracture or infection as an underlying cause.[16] The most frequent finding in the cervical spine in JRA is zygapophyseal fusion, which is often asymptomatic.[16] In children with a systemic or polyarticular onset of JRA, the odontoid waist may be eroded due to synovial hypertrophy. This may give an "apple core" appearance to the odontoid, making it more susceptible to fracture.[16]

Halo Immobilization

The halo ring and vest has been used increasingly for immobilization of the cervical spine in children. The advantages of the device include ease of application, superior immobilization and positioning of the cervical spine, fewer skin complications than with other orthoses, ease of access to wounds of the neck or scalp, freedom of motion of the mandible, and early mobilization of the patient, resulting in shorter hospitalization.

The technique of halo ring and vest application in children differs significantly from that in adults. Because of the variation in skull thickness and suture formation and closure, a CT study can be helpful before halo application on very young children. More halo pins (8 to 12) are used at lower insertional torques (1 to 5 ft-lb) compared with the standard values used in adults (4 pins at 6 to 8 ft-lb). Complication rates have been noted to be as high as 68% in children, with pin-site infections being the most common. However, it has been shown that the multiple-pin constructs used in children have a similar rate of complications when compared with four-pin constructs in children.[36]

Complications of Surgical Management

Progressive deformity in the pediatric cervical spine may take place after decompression with multiple laminectomies (postlaminectomy kyphosis) or after spinal cord injury. In one study,[37] significant spinal deformities, including kyphosis and swan-neck deformity, developed in as many as 50% of children who underwent laminectomy with or without suboccipital craniectomy for spinal cord tumors or myelomeningocoele. The likelihood of deformity increases with more extensive laminectomy, with neurologic involvement, and with younger age at surgery. Ideally, one should avoid laminectomy in children or should perform a fusion at the same time if laminectomy is necessary. The key to management if kyphosis does occur is early recognition, initial bracing, and prompt surgical management of progressive deformity.

Surgical treatment of postlaminectomy deformity includes preoperative traction with a single staged anterior release with strut graft (iliac crest or fibula). If the posterior elements are intact and fused, posterior osteotomies may be required initially along with intraoperative traction before release and fusion.[38]

Bone Graft and Posterior Arthrodesis

The efficacy of posterior arthrodesis and immobilization of the cervical spine in children has been well documented. Solid fusion is achieved in almost all cases in which delicate exposure of the desired levels of fusion is performed along with decortication and iliac-crest bone grafting.[39,40] The use of cadaveric bone has been far less successful. In one study,[41] failure of fusion was noted in all seven cases in which cadaveric bone was used, with eventual fusion after subsequent use of autogenous iliac-crest graft in all instances.[41]

Long-term follow-up results in children after posttraumatic cervical arthrodesis have been favorable. In a 17-year follow-up study, 90% of patients had excellent or good results with a low complication rate despite an overall increase in osteoarthrotic changes in unfused segments and a decrease in mobility.[42] The most common complication (38%) was spontaneous extension of the fusion mass.

Summary

Disorders of the cervical spine in children differ considerably from those in adults. Specific knowledge of the developmental anatomy and the common congenital anomalies and injury patterns in children is necessary to effectively evaluate and manage these disorders. Recent molecular genetic research is revealing more about the development of the axial skeleton. Improved imaging techniques are facilitating the radiologic evaluation of both congenital and acquired cervical spine disorders. More is being learned about the rare and unusual disorders that affect children and influence the stability of the cervical spine. With increasing awareness of the natural history of these disorders, it will be possible to more effectively manage the issues related to the cervical spine in children.

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