AJR FOCUS ON:

Pediatric Craniocervical Junction Injuries

Joseph J. Junewick[1,2,3]

OBJECTIVE. The purpose of this article is to review pediatric craniocervical junction injuries in the context of embryology, developmental anatomy, and biomechanics.

CONCLUSION. The craniocervical junction is functionally and developmentally distinct from the rest of the spine, and mechanistic models often fail to explain these injuries. Various developmental features and complex anatomy likely contribute to injury in this region in children. Some of the injury patterns at the craniocervical junction in children are similar to adults, but many are unique.

Keywords: craniocervical junction injuries, CT, MRI, pediatrics

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[1]Department of Radiology, Helen DeVos Children's Hospital/Spectrum Health Hospitals, 100 Michigan Ave NE, Grand Rapids, MI 49503. Address correspondence to J. J. Junewick (jjunewick@advancedrad.com).

[2]Advanced Radiology Services, Grand Rapids, MI.

[3]Department of Radiology, Michigan State University, Grand Rapids, MI.

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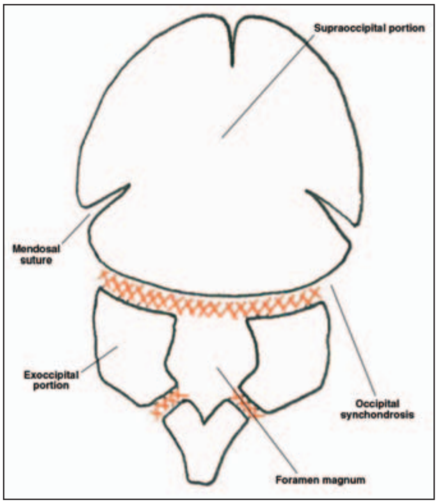
The craniocervical junction is functionally and developmentally distinct from the rest of the spine. Injuries in this region are difficult to understand in children and adults [1]. Mechanistic models often fail to explain these injuries, probably because forces are often multidirectional and may be sequential or simultaneous. Even though anatomic and curvature transitions are known to be zones of vulnerability in the spine, the craniocervical junction shows unique morphology and a dramatic transition between the occiput and the atlas. Various developmental features, such as lax ligaments, elastic boney matrix properties, decreased muscle tone, and shallow articulations, likely contribute to injury in this region in children. Some of the injury patterns at the craniocervical junction in children are similar to adults, but many are unique [2]. In this article, pediatric craniocervical junction injuries will be reviewed in the context of embryology, developmental anatomy, and biomechanics.

Anatomy

The craniocervical junction is a functional unit composed of the occiput, atlas, and axis (Fig. 1). Motion and a degree of stabilization are achieved by the paired atlantooccipital and atlantoaxial and the anterior atlantodental joints [1]. Ligaments provide the bulk of stabilization. The anterior longitudinal ligament extends cranially as the anterior atlantoaxial and atlantooccipital ligaments. The posterior longitudinal ligament continues cranially as the tectorial membrane. The posterior ligamentous complex is composed of the ligamentum flavum and interspinous ligaments. The cephalic extension of the ligamentum flavum is the posterior atlantooccipital ligament, whereas the cephalic extension of the interspinous ligament is the ligamentum nuchae. The transverse ligament of the atlas holds the dens against the anterior arch of C1; vertically oriented bands of this ligament extend to the anterior foramen magnum and to the posterior body of the axis to form the cruciform ligaments. The alar ligaments extend from the odontoid process to the lateral margins of the foramen magnum, limiting lateral rotation of the skull. The apical ligament extends from the tip of the odontoid process to the inner surface of the clivus but adds little structural support [3].

Embryology and Development

There are four occipital, eight cervical, 12 thoracic, five lumbar, five sacral, and 8–10 coccygeal sclerotomes; the occipital and first two cervical sclerotomes contribute to formation of the craniocervical junction [3–5]. The first two occipital sclerotomes form the basiocciput. The third sclerotome forms the exoccipital bone. The fourth occipital sclerotome forms the anterior tubercle of the clivus, anterior arch of the atlas, the os terminale of the dens; the alar, cruciate and the apical ligaments are also derived from the fourth occipital sclerotome. The fourth occipital sclerotome forms the anterior margin of the foramen magnum and the occipital condyles. The lateral masses and the neural



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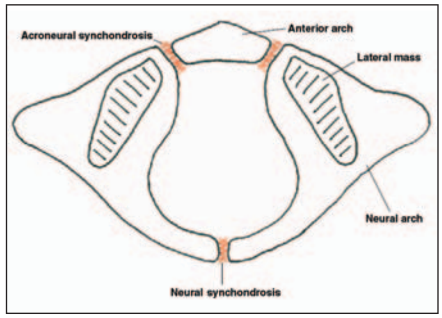

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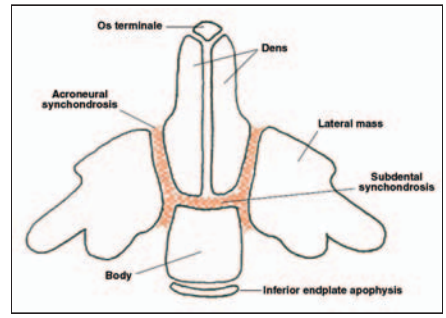

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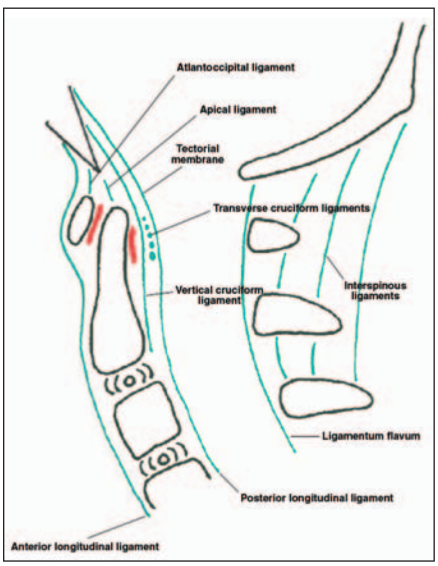


D

Fig. 1—Craniocervical junction anatomy.
A, Drawing shows axial rendering of occiput.
B, Drawing shows axial rendering of atlas.
C, Drawing shows coronal rendering of axis.
D, Drawing shows sagittal representation of ligamentous structures.

arches of the atlas are derived from the fourth occipital and first cervical sclerotomes. Portions of the first and second cervical sclerotomes fuse to form the odontoid process. The odontoid process is separated from the body of the axis vertebra by a vestigial disk that later becomes the subdental synchondrosis. The remainder of the axis is formed by the second cervical sclerotome [3–5].

The occipital bone is composed of the membranous supraoccipital and enchondral exoccipital portions. The pattern of membranous ossification gives rise to pseudosutures (mendosal sutures) at the skull base. A synchondrosis separates the supraoccipital and exoccipital segments, which fuses between 4 and 6 years of age [3–5]. The mendosal sutures and occipital synchondrosis can be confused with fractures.

The atlas is composed of three primary ossification centers. The anterior arch and right and left posterior neural arches are separated by synchondroses. The anterior arch is usually cartilaginous at birth. Ossification of the anterior arch is present in about one half of patients within the first year of life and increases with age. A single ossification center is present in most patients, although multiple ossification centers can be seen. Knowledge of these ossification patterns is necessary to confidently exclude injuries. There are no secondary ossification centers of the atlas. The synchondroses of the atlas are fused in most children by 8 years [3–6].

The five primary ossification centers of the axis include the two posterior neural arches, the vertebral body, and the right and left sides of the odontoid process. Synchondroses are present between the ossification centers. Occasionally, there may be two secondary ossification centers, the os terminale at the tip of the dens and the ring apophysis along the anterior-inferior vertebral body. The neural arches fuse posteriorly by 2–3 years of age and with the body of the odontoid process between 3 and 6 years of age [3–5].

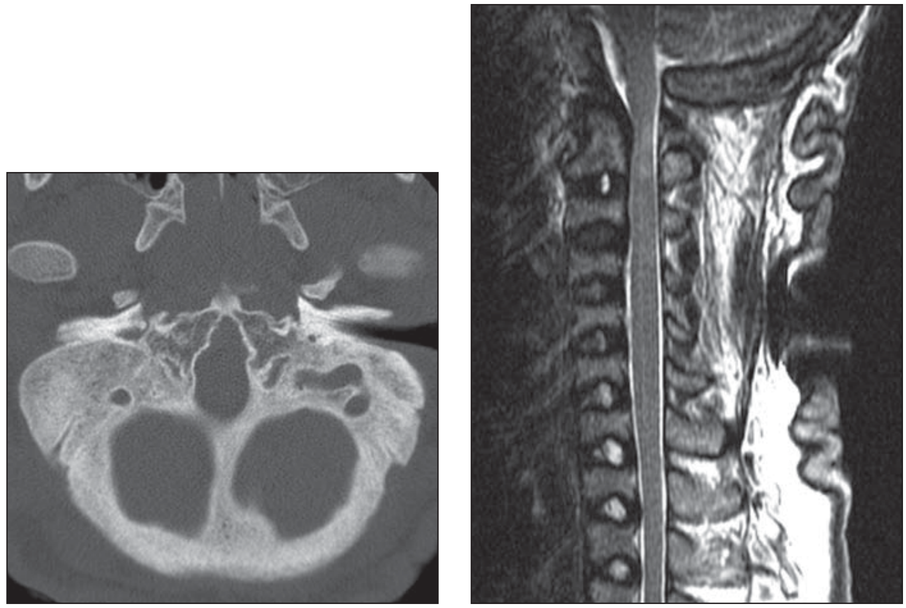
Imaging Strategy

Radiography is an inexpensive and readily available means to evaluate the craniocervical junction, although it offers relatively poor sensitivity and specificity in the detection of injury. Various measurements (e.g., Powers ratio, atlantodental interval, atlantooccipital interval, basion-axial interval, basion-dental interval) have been used to help diagnose craniocervical junction injuries but are difficult to apply in children because of age-dependent differences and other variations in ossification that make ratios and measurements inaccurate [7]. CT with multiplanar reformation offers significantly improved fracture detection compared with radiography [8–10]. MRI accurately depicts soft tissue

TABLE 1: Strategy for Evaluation of Potential Cervical Spine Injury

Situation	Imaging
If no focal cervical tenderness, no neurologic deficit, no distracting injury, and normal mental status	No imaging
If focal cervical tenderness, neurologic deficit, distracting injury, altered mental status	CT (1.0–1.25 mm with sagittal and coronal reformats)
If CT is negative with focal tenderness, neurologic deficit, or significant mechanism of injury	MRI (sagittal T1-weighted, fat-saturated T1-weighted, T2-weighted and STIR, and axial T1- and T2-weighted sequences)
If CT and MRI are negative with persistent focal tenderness, neurologic deficit, or significant mechanism of injury	Repeat MRI in 1 week
If CT is positive and management requires assessment of soft-tissue or cord integrity	MRI

Fig. 2—Unresponsive ejected unrestrained backseat teenage passenger involved in broadside motor vehicle accident. Coronal reformat image shows fracture of right occipital condyle with minimal medial displacement.



A B

Fig. 3—Teenager with thanatophoric dysplasia.
A, Axial CT image of skull base shows marked stenosis of foramen magnum.
B, Sagittal T2-weighted image shows diffuse cervical canal stenosis and central cord edema and gliosis.

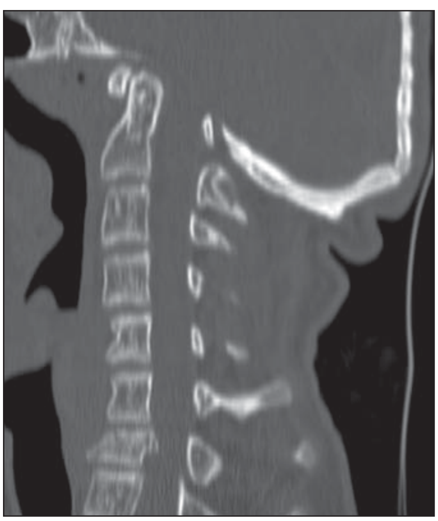


Fig. 4—9-year-old boy with osteogenesis imperfecta with platybasia. Note healing C7 burst fracture.

Fig. 5—Sagittal fat-suppressed T2-weighted image of cervical spine in child involved in motor vehicle accident shows precervical soft-tissue edema and hemorrhage, disrupted atlantoaxial and apical ligaments, elevation of tectorial membrane, fluid in atlantodental interval, and posterior paraspinal edema.

Fig. 6—Sagittal fat-suppressed T1-weighted MR image of cervical spine shows hyperintense subacute hemorrhage anteriorly at craniocervical junction related to inflicted injury in 9-month-old girl. Note dependent hemorrhage supratentorially and in posterior fossa.

injuries (disk annulus, ligament, muscle, and joint capsule) and cord integrity without radiation. MRI complements CT in evaluation of cervical spine injury and has an important role in surgical planning and clinical prognostication [11].

Imaging strategies in the evaluation of potential cervical spine injury vary. Probably the best pediatric data are drawn from the prospective multicenter National Emergency X-Radiography Utilization Study (NEXUS). NEXUS identified 30 fractures of the cervical spine in 3065 patients under the age of 18 years. In this cohort, 21 of 30 fractures were encountered with teenagers; no fractures were seen in patients under 2 years old, 4 fractures were seen between 2 and 9 years of age, and 5 were seen between 10 and 13 years of age. If there was absence of focal cervical tenderness, distracting injury, neurologic deficit, and altered mental status, no patient had a cervical spine fracture [12, 13]. In accordance with this information, the strategy used at Helen DeVos Children's Hospital is summarized in Table 1.

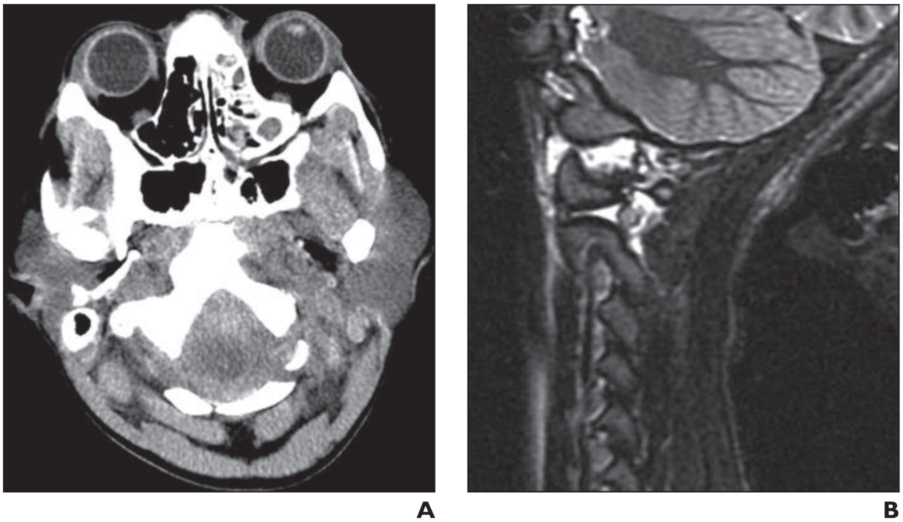


Fig. 7—12-year-old restrained backseat passenger in vehicle struck by snowplow who was agonal at scene of accident.
A, CT image of brain shows blood at foramen magnum.
B, Sagittal inversion recovery MR image of cervical spine shows anterior translation of occipital condyle relative to C1 lateral mass and posttraumatic joint effusion.

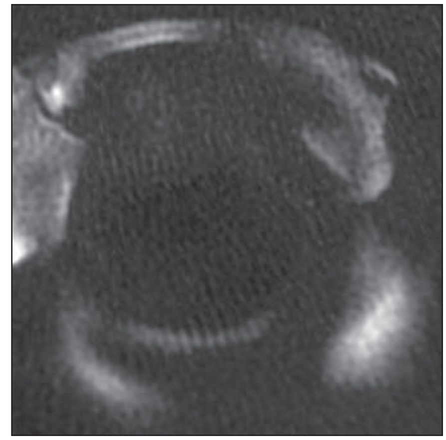


Fig. 8—Axial maximum-intensity-projection image in child shows impaction fracture arising from anterior lip of superior articular surface of left lateral mass of C1.

Fig. 9—4-year-old restrained backseat passenger in vehicle struck broadside. Sagittal reconstructed CT image shows avulsion fracture of inferior aspect of anterior arch of C1.

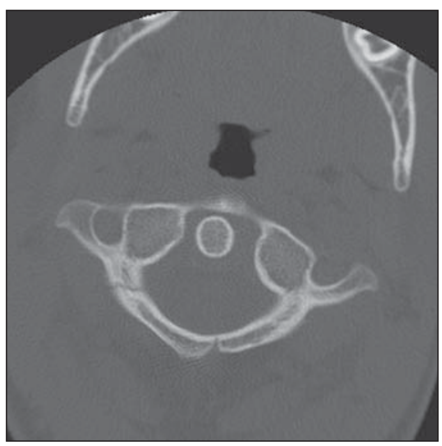


Fig. 10—Axial CT image in 12-year-old boy injured by falling tree limb shows fractures of posterior arch of C1.

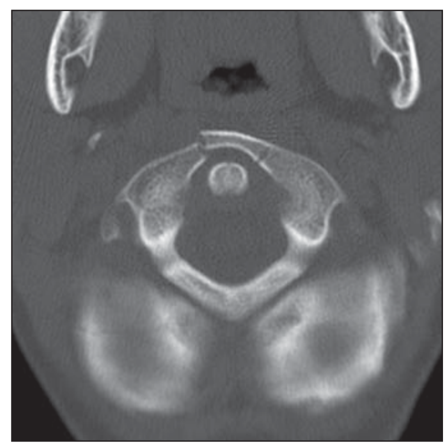


Fig. 11—Axial CT image in 6-year-old boy shows diastasis and malalignment of right synchondrosis of atlas.

Occipital and Skull Base Injuries

The configuration of the atlantooccipital joints allows flexion, extension, and lateral flexion but only limited rotation [1]. Small flat condyles and lax capsules in children result in increased mobility of these joints. Fractures of the occipital condyles related to axial loading and lateral flexion are usually stable; fragments are usually comminuted but minimally displaced (Fig. 2). Alar ligament avulsion infers instability related to rotational or translational forces and is evident by medial displacement of the condyle fragment [14]. The key to diagnosis of occipital condyle fractures is noticing blood at the foramen magnum on CT.

Many developmental anomalies predispose to injury at the skull base [15]. Congenital stenosis of the foramen magnum and spinal canal may also predispose to cord injury (Fig. 3). Patients with osteopetrosis, pycnodysostosis, and osteogenesis imperfecta are more prone to fractures because of defective mineralization [16] (Fig. 4).

Craniocervical Junction Injuries

Nonosseous injury of the craniocervical junction is much more common in children. MRI is crucial for evaluation; many children with normal CT examinations have MR evidence of craniocervical junction injury [17, 18]. Injuries range from minor soft-tissue injury to cervicomedullary contusion. Disproportionate head size, poor muscle tone, lax ligaments, and incompletely developed articulations make the craniocervical junction especially vulnerable in infants and young children (Fig. 5).

The craniocervical junction is a common site of injury in child abuse. Respiratory symptoms or neurologic depression may mask craniocervical junction injury. Much of the evidence regarding craniocervical junction injury in child abuse is drawn from autopsy reports. Intracranial subdural and epidural hemorrhage have been highly associated with known inflicted intracranial injury but also in patients with sudden infant death syndrome as well as other manifestations of child abuse (Fig. 6). Cervicomedullary contusion and edema, hangman's fracture, and fracture-dislocations have also been reported [19–22].

Craniocervical dissociation may be related to translational injury in the transverse plane, distraction, or a combination. The craniocervical junction is largely stabilized by ligaments and to a lesser extent by joints. Craniocervical injury may be manifested by subluxation or dislocation, although occasionally these spontaneously reduce (Fig. 7). Capsular and ligamentous avulsion fractures and impaction fractures may be evident (Fig. 8). Craniocervical dissociation is associated with cervicomedullary dysfunction, lower cranial nerve palsies (cranial nerves IX–XII), and vertebrobasilar arterial injury. Shallow atlantooccipital articulations and excessive ligament or capsule laxity can be present in trisomy 21 and other syndromes leading to increased movement at the craniocervical junction. Imaging may be necessary for screening these patients who are at risk for cervical spine injury or to define anatomic defects.

Atlas Fractures

Avulsion of the atlantoaxial ligament or longus colli muscle from hyperextension produces a transverse fracture through the tubercle of the anterior arch. This unusual injury is not associated with neurologic deficit or mechanical instability but can cause severe pain (Fig. 9). The presence of prevertebral soft-tissue swelling helps to distinguish fracture from enthesopathic or other chronic change in adults and variation of ossification in children [1].

Hyperextension can also result in compression of the posterior arch of the atlas between

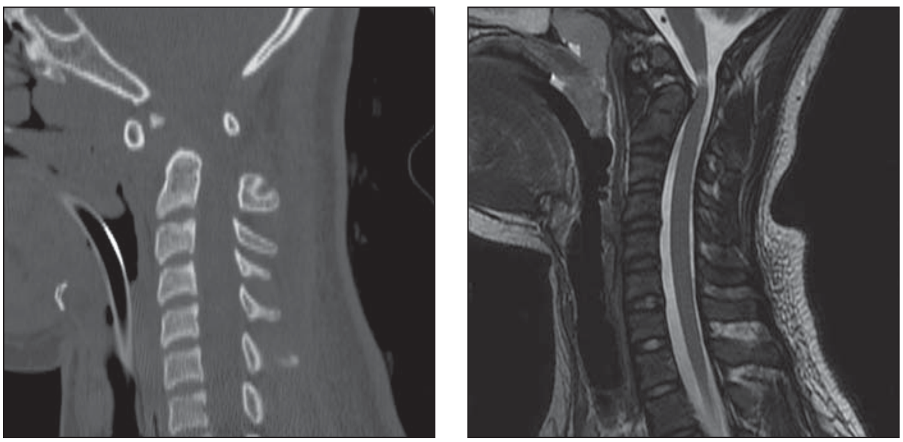


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Fig. 12—Teenager with trisomy 21.
A, Sagittal reconstructed CT image of cervical spine shows os odontoideum and narrowing of cervical canal at C1–C2.
B, Sagittal T2-weighted MR image again shows os odontoideum and also shows resultant myelomalacia.

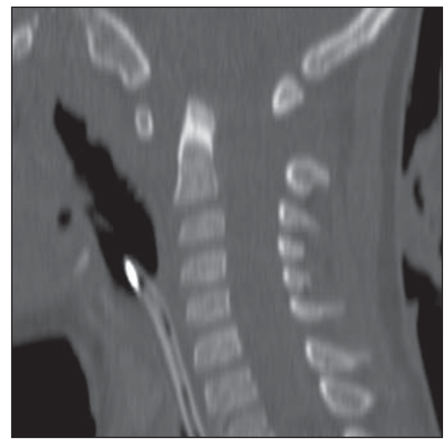

Fig. 13—Sagittal reformat CT image in child shows marked widening of predental space with discontinuous spinolaminar line indicating disruption of transverse atlantal ligament.

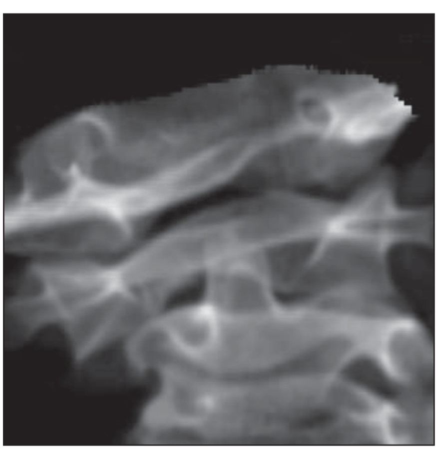

Fig. 14—Three-dimensional volume-rendered image in 5-year-old child shows atlantoaxial rotary subluxation.

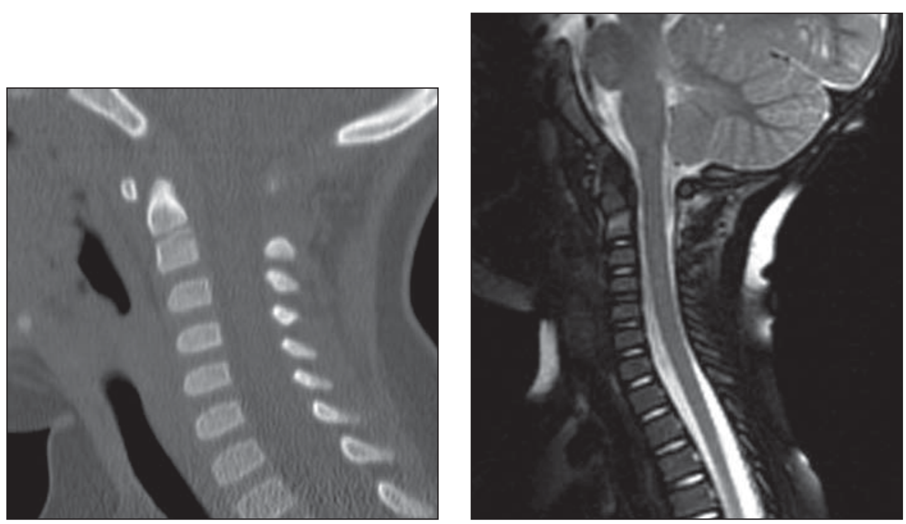

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Fig. 15—15-month-old backseat restrained (six-point harness booster chair) passenger involved in head-on motor vehicle collision.
A, Sagittal reconstructed CT image reveals subtle linear lucency in body of C2 just subjacent to subdental synchondrosis.
B, Sagittal fat-suppressed T2-weighted MR image confirms edema in C2 body, corresponding to CT abnormality.

the occiput and neural arch of the axis. This injury usually involves the right and left sides, although hyperextension combined with lateral flexion or rotation may result in asymmetric involvement (Fig. 10). These fractures are often nondisplaced and not associated myelopathy [1].

Jefferson fractures were thought to be quite rare in children [23–25]. With CT and MRI, this injury is more commonly recognized although still less common compared with adults because of the higher tensile strength and more elastic characteristics of the developing atlas. The mechanism of injury is compression, similar to adults. The posterior midline synchondrosis and the right and left anterior synchondroses of the atlas may participate in the Jefferson fracture. Synchondrosis involvement is diagnosed by malalignment or diastasis at the synchondrosis (Fig. 11).

Atlantoaxial Injuries

Atlantoaxial instability may be difficult to differentiate from the various forms of pseudosubluxation that occur at this level [26]. In hyperextension, the anterior arch of the atlas can be normally seen superior to the dens. The predental space can measure up to 5 mm. The lateral atlantodental interval may be normally asymmetric, giving the appearance of rotary subluxation or Jeffersonian burst; asymmetry of 1.39 mm (SD, 1.26 mm) was found in normal children (Borders H et al., unpublished data). Atlantoaxial instability related to ligamentous instability, incomplete segmentation, or hypoplasia is commonly seen in trisomy 21, type 2 collagenopathies, chondrodysplasia punctata (Conradi-Hünerman syndrome), diastrophic dysplasia, metatropic dysplasia, pseudoachondroplasia, and spondyloepimetaphyseal dysplasia among others [16] (Fig. 12).

The transverse atlantal ligament extends between the tubercles of the C1 lateral masses and cradles the posterior aspect of the odontoid process. Transverse atlantal ligament disruption is rare, and may be isolated or associated with a Jefferson burst fracture. The mechanism of injury is complex, likely related to a combination of flexion, lateral tilt, and axial compression. Diagnosis is suggested by a widened predental space and disruption of the spinolaminar line; the associated avulsion fractures are difficult to visualize even on CT (Fig. 13). The anterior-posterior translational movement of the atlas relative to the axis often results in cord injury [1, 25, 27].

Torticollis and acute rotary subluxation are abnormalities related to varying degrees of trauma resulting in rotation of the head in one direction and tilting of the head in the opposite direction (Fig. 14). The mechanism of injury is usually rotation and distraction. Usually the greater the degree of rotation of the lateral masses of C1 relative to C2, the greater the incidence of fixation and need for surgical reduction [1, 25, 28].

Axis Fractures

Fracture of the odontoid process is one of the most common fractures in young children, usually occurring through the subdental synchondrosis. The subdental (subchondral or neurocentral) synchondrosis of the axis usually fuses during adolescence but may variably persist into adulthood. The subdental synchondrosis is a potential weak region of the axis and prone to hyperflexion injury (Fig. 15). Because the sagittal fulcrum of motion in younger children is in the upper cervical region rather than the lower cervical spine as in adults, the vulnerability is potentiated. It is interesting to note that remnants of the synchondrosis persist in 87% of adults and may account for the increased incidence of low dens fractures and pseudoarthrosis [29].

Traumatic spondylolysis (hangman fracture) of the axis may be related to hyperextension with vertical compression of the posterior elements or hyperflexion with distraction [1]. Fractures are almost always bilateral but may involve different segments of the posterior elements (Fig. 16). Fractures most often occur between the pedicle and inferior articular facet. Traumatic spondylolysis is rare before fusion of the synchondroses and should not be confused with an unusual form of congenital spondylolysis (Fig. 17). Treatment of acute traumatic spondylolysis is largely dependent on the degree of translation and angular deformity of C2 relative to C3.

Isolated lamina fractures of the axis occur posterior to the pars interarticularis and are therefore distinct from the hangman fracture [1]. The C2 lamina fracture is similar to hy-

Fig. 16—Axial CT image of cervical spine shows coronal fractures through left pedicle-body and right pars interarticularis in 17-year-old restrained front seat passenger involved in high-speed head-on motor vehicle accident with no neurologic deficit.

Fig. 17—Lateral radiograph in 17-month-old boy who fell from couch shows smoothly marginated congenital pars interarticularis defect.

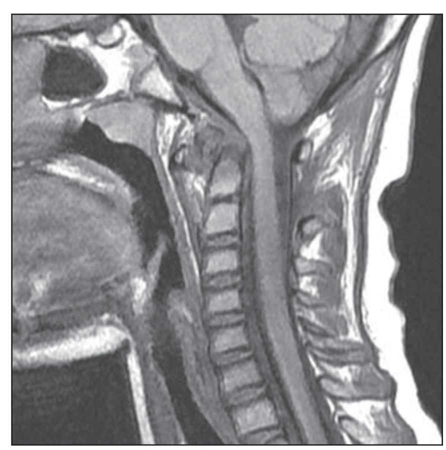

Fig. 18—6-year-old asymptomatic rear seat passenger involved in high-speed head-on motor vehicle accident. Sagittal T2-weighted MR image shows avulsion of tip of dens and atlantoaxial disruption.

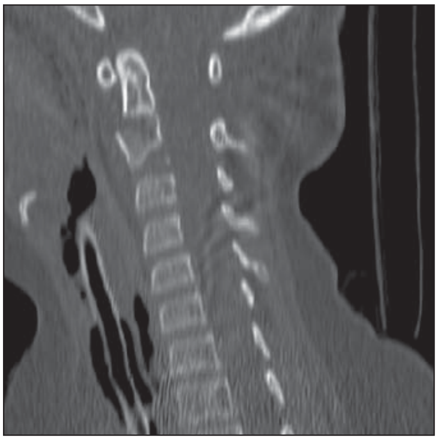


Fig. 19—10-year-old pedestrian with trisomy 21 who was hit by car traveling at 30 miles per hour.
A, Sagittal reconstructed CT image shows precervical soft-tissue swelling, low dens fracture, and fracture of posterior-inferior corner of C2 vertebral body.
B, Sagittal inversion recovery MR image shows disrupted posterior longitudinal ligament and annulus and interspinous edema at C1–C2 and C2–C3.

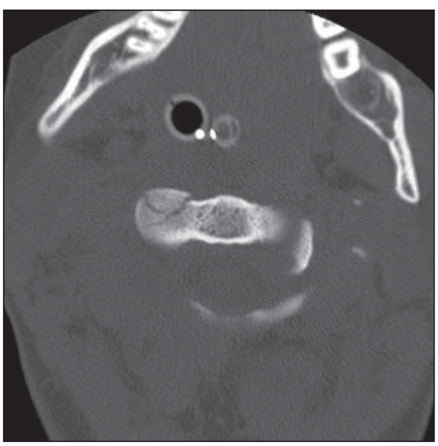

Fig. 20—17-year-old female driver found unresponsive next to vehicle after presumed high-speed vehicle impact against tree. Axial CT image reveals minimally comminuted fracture of right lateral mass of axis.

perextension fracture of the posterior arch of the atlas and to lamina fracture in the lower cervical spine. These fractures are mechanically stable although the fragments can potentially impinge on the cord.

After subdental synchondrosis closure, odontoid fracture patterns are similar to the adult. The mechanism of action is complex and often poorly understood. There is controversy over the classification of odontoid fractures. It is most practical to consider a fracture as "high" or "low." High fractures are limited to the odontoid process itself (Fig. 18). Low fractures extend into the body of the axis (Fig. 19). High fractures rarely occur in infants and children but are more common in teenagers. Because the odontoid is largely composed of cortical bone, nonunion is a common complication, especially if the fragment is displaced. Low fractures are mechanically unstable. Odontoid fractures are associated with fractures of the face, craniocervical dissociation, extension teardrop injuries, and Jefferson fractures [1, 30, 31].

Unilateral lateral mass fractures are relatively common. The mechanism is likely lateral flexion leading to compression of the pedicle and displacement of the overlying facet. Odontoid fracture is associated with this injury [32] (Fig. 20).

Axiocervical Injuries

The craniocervical junction ends at the C2–C3 disk space. The C2–C3 disk space represents a transition to conventional cervical vertebral morphology. As such, it is also susceptible to injury [33] (Fig. 18). Differentiation of posttraumatic subluxation of the cervical spine from "pseudosubluxation" may be problematic at this level. Sagittal plane pseudosubluxation is most common at C2–C3, occurring in 19% of children under 7 years old [26]. Nontraumatic pseudosubluxation can be confidently diagnosed if the anterior longitudinal line is offset by less than 3 mm with an intact spinolaminar line.

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