CHAPTER 21

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The Cervical Spine

SURGICAL PROCEDURE INDEX

Many of the diseases and congenital anomalies affecting the pediatric cervical spine are simply a reflection of aberrant growth and developmental processes. This chapter discusses these diseases and anomalies in this framework. A basic knowledge of the normal embryology, growth, and development of the pediatric cervical spine is necessary to understand these conditions. Most of the anomalies and diseases involving the pediatric cervical spine are easily divided into those of the upper (occiput, C1, C2) and lower (C3-C7) segments.

NORMAL EMBRYOLOGY, GROWTH, AND DEVELOPMENT

Embryology

Occipitoaxioatlas Complex. The occiput is formed from at least four or five somites. All definitive vertebrae develop from the caudal sclerotome half of one segment and the cranial sclerotome half of the succeeding segment (1). These areas of primitive mesenchyme separate from each other during fetal growth and then undergo chondrification and subsequent ossification. This chondrification and ossification is a passive process, following the blueprint laid down by the mesenchymal anlage. Because of this sequencing, the cranial half of the first cervical sclerotome remains as a half segment between the occipital and the atlantal rudiments and is known as the proatlas. The primitive centrum of this proatlas becomes the tip of the odontoid process, whereas its arch rudiments assist in the formation of the occipital condyles (2). The vertebral arch of the atlas separates from its respective centrum, becoming the ring of C1; the separated centrum fuses with the proatlas above and the centrum of C2 below, to become the odontoid process and body of C2. The axis forms from the second definitive cervical vertebral mesenchymal segment. The odontoid process is the fusion of the primitive centra of the atlas and the proatlas half segment. The posterior arches of C2 form from only the second definitive cervical segment.

Thus, the atlas is made up of three main components: the body and the two neural arches. The axis is made up of four main components: the body, two neural arches, and the odontoid (or five components if the proatlas rudiment is considered) (Figs. 21-1 and 21-2).

Vertebrae C3-C7. These vertebrae follow the normal formation schema of all vertebrae (3). A portion of the mesenchyme from the sclerotomal centrum creates two neural arches that migrate posteriorly and around the neural tube. This eventually forms the pedicles, the laminae, the spinous processes, and a very small portion of the body. The majority of the body is formed by the centrum. An ossification center develops in each of the two neural arches and one in the vertebral center, with a synchondrosis formed by the cartilage between the ossification centers.

Basic Science, Embryology, and Gene Expression. In the past decade, there has been an explosion of knowledge regarding the human genome and how it relates to normal developmental processes and pathologic conditions. Vertebral segmentation begins with clustering segments of the paraxial mesoderm, the somites. Segmentation of the mesoderm into somites is an important yet fundamental process that allows for spatial specialization in the organism and is under genetic control.

The homeobox is a highly conserved 160-base pair sequence found in the homeobox genes, termed *Hox* genes for short. These *Hox* genes encode a highly conserved family of transcription factors that play fundamental roles in morphogenesis during embryonic development. Vertebrate *Hox* genes help control developmental patterning in the embryo along the

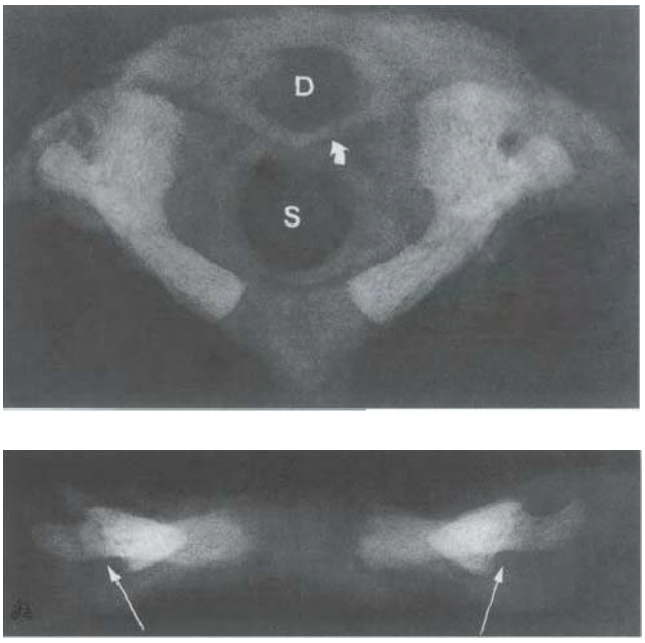


FIGURE 21-1. **A:** Cross-sectional radiograph of C1 in a full-term neonate. The posterior ossification centers are present. No ossification is present in the anterior cartilage. The transverse ligament (*arrow*) separates the dens (D) from the spinal canal (S). **B:** AP radiograph of C1 in a term neonate. (From Ogden JA. Radiology of postnatal skeletal development. XI. The first cervical vertebrae. *Skeletal Radiol* 1984;12:12–20, with permission.)

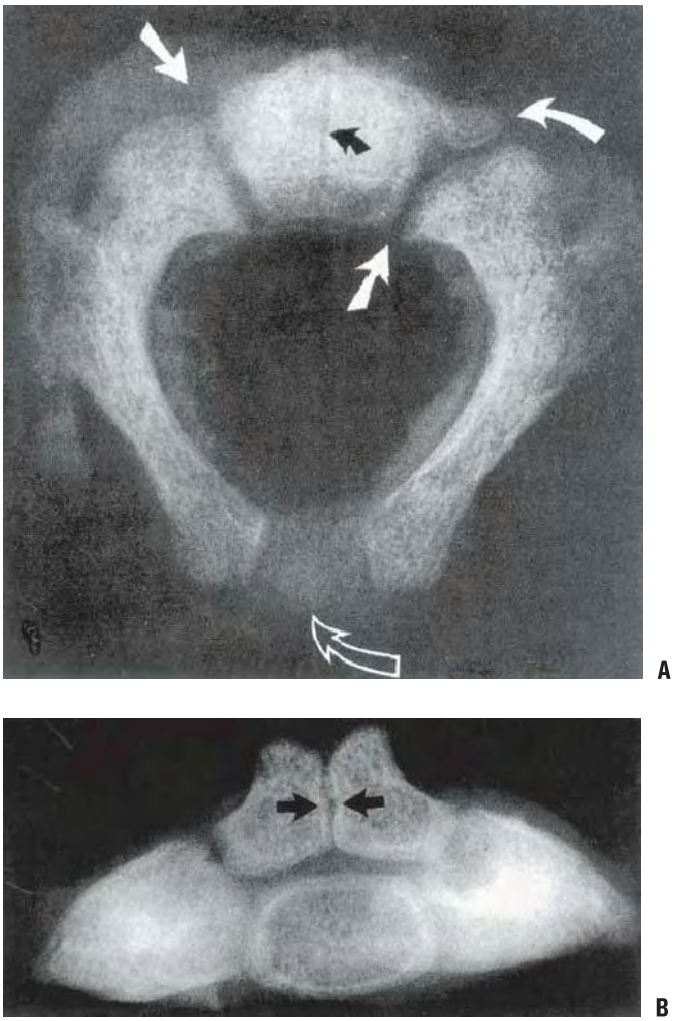


FIGURE 21-2. **A:** Cross-sectional radiograph of C2 in a neonate. The neurocentral (*solid arrows*) and posterior (*open arrows*) synchondroses are evident. A small area of accessory ossification is present in the right neurocentral synchondrosis anteriorly (*curved arrow*). Also note the central linear radiolucency (*black arrow*) indicating the synchondrosis between the dens ossificiation centers. The posterior ossification centers extend into the eventual vertebral body. **B:** The AP radiograph of C2 in a neonate. In this specimen, the dens ossification centers have not fused, leaving a midline synchondrosis (*arrows*) that extends from the chondrum terminale to the dentocentral synchondrosis. The superior margin of the eventual vertebral body is above the lower level of the dens. The neurocentral synchondroses are continuous with the "ring apophyseal" cartilage inferiorly, the facet cartilage inferiorly, and the dentocentral synchondrosis superiorly. (From Ogden JA. Radiology of postnatal skeletal development. XII. The second cervical vertebra. *Skeletal Radiol* 1984;12:169–177, with permission.)

primary (head-to-tail) and secondary (genital and limb bud) axes. There are 39 *Hox* genes in vertebrates that are organized into four clusters located on different chromosomes. In the human, these clusters are named HOXA, HOXB, HOXC, and HOXD, located on chromosomes 7p14, 17q21, 12q13, and 2q31, respectively (4). In animals, they are written in lower case (e.g., *Hoxc*); in the human, they are written in upper case (e.g., HOXC). Each cluster contains 9 to 11 genes, all oriented in the same 5′ to 3′ direction of transcription. There are 13 possible subsets of genes; no single cluster contains a representative from all 13 known numbered subsets (paralogous groups). The numbering of the genes in each cluster is based on their sequence similarity and relative positions, starting from that end of the complex that is expressed most anteriorly (cranially). The equivalent genes in each complex are called a paralogous group.

The expression domains of the HOX clusters display a nested arrangement. Along the body axis, the *Hox* genes are generally expressed with discrete rostral cutoffs that coincide with either existing or emergent anatomic landmarks. *Hox* genes at the end of the 3′ cluster (e.g., HOXA1) are generally expressed early, in anterior and proximal regions; *Hox* genes at the 5′ end (e.g., HOXA13) are generally expressed later, in more posterior and distal regions. Thus, the lower numbered *Hox* genes are involved in the development of the axial skeleton, and the higher numbered *Hox* genes are involved in the development of the limbs. There has been an explosion of knowledge regarding defects in the *Hox* genes and resultant congenital spinal anomalies in experimental animals, and to much lesser extent in humans (Table 21-1). The defect can be a distinct *Hox* gene mutation intentionally produced by the investigator or more random hits by teratogens (methanol, boric acid, retinoic acid, maternal hyperthermia) (13–17).

Another group of genes, the *Pax* genes, are also integrally involved in vertebral development. The *Pax* genes are a highly conserved family of developmental control genes that encode transcription factors containing a 128-amino acid DNA-binding domain (18, 19), called the paired box (20). To date, there are nine *PAX* genes (12, 21). The *Pax* gene family is broken down into four subgroups (*Pax1* and *Pax9*; *Pax2*, *Pax5*, and *Pax8*; *Pax3* and *Pax7*; *Pax4* and *Pax6*). *Pax1* and *Pax9* induce chondrogenic differentiation in the paraxial mesenchymal mesoderm of the sclerotome (19, 20, 22). *Pax1* expression is also seen in the posterior occiput, indicating that the basilar occiput from a developmental standpoint can be considered the uppermost vertebra (23). Thus, they are critically involved in vertebral formation. Abnormalities in the PAX1 sequence in humans have been associated in some patients with Klippel-Feil syndrome (12).

The Hedgehog family of proteins has also become increasingly recognized as crucial to axial skeletal development. The best known of these proteins is the sonic hedgehog (*shh*), which is expressed in the notochord (24, 25). *Shh* is believed to be the signal for induction of the ventral somite to differentiate into the sclerotome (25). In *shh* knockout mice, most sclerotomal derivatives are absent, in conjunction with reduced expression of *Pax1* (25). Thus, absence of *shh* leads to absence of *Pax1* expression and subsequent failure of the mesenchymal cells to chondrify. Defective *shh* signaling during embryogenesis in mice results in anomalies similar to those seen in the human VACTERL association (26).

Growth and Development

Atlas. Ossification is present only in the two neural arches at birth (27). These ossification centers extend posteriorly toward the rudimentary spinous process to form the posterior synchondrosis and anteriorly into the articular facet region to form all of the bone present in the facets. Anteromedial to each facet, the neurocentral synchondroses form, joining the neural arches and the body; this occurs on each side of the expanding anterior ossification center. The body starts to ossify between 6 months and 2 years, usually in a single center. By 4 to 6 years, the posterior synchondrosis fuses, followed by the anterior ones slightly thereafter. The final internal diameter of the pediatric C1 spinal canal is determined by 6 to 7 years of age. Further growth is obtained only by periosteal appositional growth on the external surface, which leads to thickening and an increased height, but without changing the size of the spinal canal. Thus, a spinal fusion after the age of 6 or 7 years has minimal impact on the internal canal diameter; when possible, surgical fusion should not be performed before this age due to the potential for later cervical stenosis.

TABLE 21.1 Axial Skeletal Malformations Due to Genetic Abnormalities

Involved gene	Gene abnormality	Phenotypic expression	Animal	Reference
Hoxb-4	Knockout	C2 becomes C1	Mouse	(5)
Hoxd-3	Homozygous	Atlas fused to occiput, C2 becomes more like C1	Mouse	(6)
Hoxa-4	Homozygous	Development of ribs on C7	Mouse	(7)
Bapx1	Knockout	Malformed basioccipital bone. Absence of anterior arch of atlas	Mouse	(8)
Uncx4.1	Homozygous	Absence of pedicles and transverse processes, cervical vertebrae	Mouse	(9)
Cdx1	Homozygous	Cranial transformations of cervical spine: absence of anterior arch of C1 C2 becomes like C1 C3 becomes like C2 Development of ribs on C7	Mouse	(10, 11)
PAX1	Sequence changes	Klippel-Feil syndrome	Human	(12)

Axis. The odontoid develops two primary ossification centers that usually coalesce within the first 3 months of life; these centers are separated from the C2 centrum by the dentocentral synchondrosis (28, 29). This synchondrosis is below the level of the C1-C2 facets and contributes to the overall height of the odontoid as well as to the body of C2. It is continuous with the vertebral body and facets, and it coalesces with the anterior neurocentral synchondroses and finally at the dentocentral synchondrosis. This closure occurs between 3 and 6 years of age. The tip of the dens comprises a cartilaginous region similar to an epiphysis, the chondrum terminale, which develops an ossification center between 5 and 8 years, becoming the ossiculum terminale. The ossiculum terminale fuses to the remainder of the odontoid between 10 and 13 years of age.

The posterior neural arches are partially ossified at birth, joined by the posterior synchondrosis. By 3 months of age, these arches, growing more posteriorly, form the rudimentary spinous process. By 1 year of age, ossification fills the spinous process, and by 3 years of age, the posterior synchondrosis has fused. Thus, both the posterior and the anterior synchondroses are closed by 6 years of age, and there is no further increase in spinal canal size after this age.

C3-C7. At birth, all three ossification centers are present. The anterior synchondrosis (i.e., neurocentral synchondrosis) is slightly anterior to the base of the pedicles; it usually closes between 3 and 6 years of age. The posterior synchondrosis is at the junction of the two neural arches; it usually closes by 2 to 4 years of age. In the neonate and the young child, the articular facets are horizontal but become more vertically oriented as the child ages and reaches the normal adult configuration. They are also more horizontal in the upper cervical spine than in the lower cervical spine. The vertebral bodies enlarge circumferentially by periosteal appositional growth, whereas they grow vertically by endochondral ossification. Secondary ossification centers develop at the tips of the spinous processes and the cartilaginous ring apophyses of the bodies around the time of puberty. These ring apophyses are involved in the vertical growth of the body. These secondary ossification centers fuse with the vertebral body around age 25 years. There is an overall increase in pedicle axis width but not length as the child grows; thus, pedicle screw fixation of the cervical spine in children is not considered safe (30).

Vertebral Body and Canal Diameter Changes with Growth. Due to the fact that the immature vertebrae are more cartilaginous compared to the mature adult vertebrae, there are significant differences between normal vertebral measurements in the child compared to the adult (31). As the child ages, the vertebral body height increases relative to the vertebral body depth. This is due to the activity of the apophyseal end plates that contribute proportionally more growth to the height of the vertebral body compared to the appositional growth that contributes to the depth of the vertebral body. The vertebral body height-to-depth ratio increases from approximately 0.5 in children <1 year of age to 0.8 to 0.9 in adults. This remains relatively constant for all vertebral bodies from C3 to C7. With these changes in the vertebral body height relative to depth, there are also changes in the sagittal diameter of the canal relative to vertebral body depth. The ratio of the sagittal canal diameter to vertebral body depth is stable at 1.4 in children from birth to 7 to 8 years of age and then gradually decreases to the normal 1.0 adult value (31). Knowledge of these normal growth parameters is important when determining the possibility of platyspondyly or spinal stenosis.

Normal Radiographic Parameters. Certain radiographic parameters that indicate pathology of the cervical spine in adults represent normal developmental processes in children (32). These parameters are the atlantooccipital motion and atlanto–dens interval (ADI), pseudosubluxation and pseudoinstability, variations in the curvature of the cervical spine that may resemble spasm and ligamentous injury, variations in the presence of skeletal growth and growth centers that may resemble fractures, and anterior soft-tissue widening. Normal cervical spine motion in children is also discussed.

Atlanto–Dens Interval and Atlantooccipital Motion. These intervals are determined on lateral flexion and extension views, which should be performed voluntarily with the patient awake. The ADI is the space between the anterior aspect of the dens and the posterior aspect of the anterior ring of the atlas (Fig. 21-3). An ADI of more than 5 mm on flexion and extension lateral radiographs indicates instability (33, 34). This is more than the 3-mm adult value because of the increased cartilage content of the odontoid and ring of the atlas in children as well as the increased ligamentous laxity in children. In extension, overriding of the anterior arch of the atlas on top of the odontoid also can be seen in up to 20% of children (35) (see Fig. 21-6B).

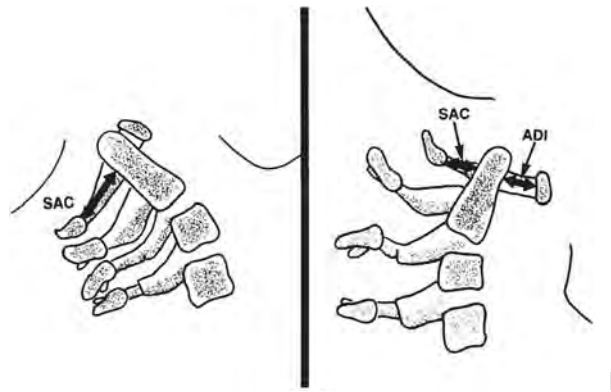


FIGURE 21-3. Lateral view of the atlantoaxial joint. The ADI is the distance between the anterior aspect of the dens and the posterior aspect of the anterior portion of the ring of the atlas. The SAC is the distance between the posterior aspect of the dens and the anterior aspect of the posterior portion of the ring of the atlas. In children, an ADI of 5 mm or larger is abnormal. In teenagers and adults, a SAC of 13 mm or smaller can be associated with canal compromise. In younger children, spinal cord impingement is imminent if the SAC is equal to or less than the transverse diameter of the odontoid. **A:** The relations in extension. **B:** The relations in flexion.

A mild increase in the ADI may indicate a subtle disruption of the transverse atlantal ligament. In adults, an ADI > 5 mm indicates ligament rupture (36). In chronic atlantoaxial conditions (e.g., rheumatoid arthritis, Down syndrome, congenital anomalies), the ADI is less useful. In children with these disorders who are frequently hypermobile but do not have ruptured transverse atlantal ligaments, the ADI is increased beyond the 3- to 5-mm range. The complement of the ADI, the space available for the cord (SAC), is a more useful measure in this situation. This space is the distance between the posterior aspect of the dens and the anterior aspect of the posterior ring of the atlas or the foramen magnum. A SAC of <13 mm may be associated with neurologic problems (37).

In patients in whom there is an attenuation of the transverse atlantal ligament without rupture, the alar ligament provides some stability. It acts like a checkrein (38), first tightening up in rotation and then becoming completely taut as the odontoid process continues to move posteriorly for a distance equivalent to its full transverse diameter. This safety zone between the anterior wall of the spinal canal of the atlas, the axis, and the neural structures is an anatomic constant equal to the transverse diameter of the odontoid. This constant defines Steel's rule of thirds: one-third cord, one-third odontoid, and one-third space. This rule remains constant throughout the growth of the cervical spine (39). The cord can move into this space (safe zone) when the odontoid moves posteriorly because of an attenuated transverse atlantal ligament. It is here that the alar ligament becomes taut, acting as a checkrein and secondary restraint, preventing further movement of the odontoid into the cord. In the chronic situation, it is important to recognize when this safe zone has been exceeded and the child enters the region of impending spinal cord compression. In the case of trauma, the alar ligament is insufficient to prevent a fatal cord injury in the event of another neck injury similar to the one that caused the initial interruption of the transverse atlantal ligament.

Normal ranges of motion at the atlantooccipital joint are not well defined. In a series of 40 normal college freshman, the tip of the odontoid remained directly below the basion of the skull in both flexion and extension (40). Thus, the joint should not allow any horizontal translation during flexion and extension. Tredwell et al. (41) believe that a posterior subluxation of the atlantooccipital relation in extension of more than 4 mm indicates instability (Fig. 21-4). This can be measured

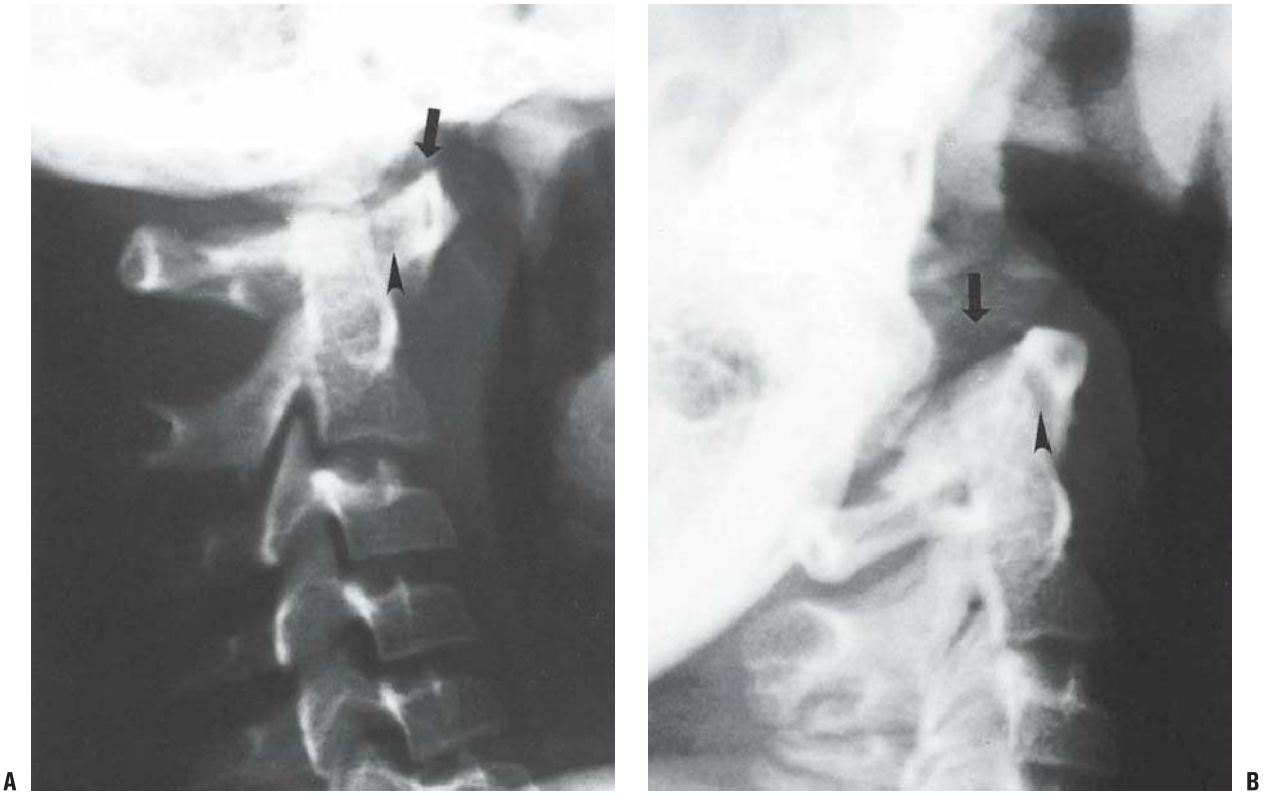


FIGURE 21-4. Lateral (**A**) flexion and (**B**) extension radiographs of an 11-year-old boy with Down syndrome. The child presented with loss of hand control when flexing his neck. Using the method of Tredwell et al. (41), the atlantooccipital distance is measured as the distance between the anterior margin of the condyles at the base of the skull and the sharp contour of the anterior aspect of the concave joint of the atlas. More than 4 mm of posterior translation is abnormal. The atlantooccipital distance (*arrows*) measured 10 mm in extension and 1 mm in flexion. The ADI was 1 mm in extension and 6 mm in flexion, for a total of 5 mm of motion (*arrowheads*). The SAC was 17 mm in flexion and 20 mm in extension. Both occipitoatlantal instability (more than 4 mm posterior translation) and atlanto–dens hypermobility (5 mm ADI in flexion) were present.

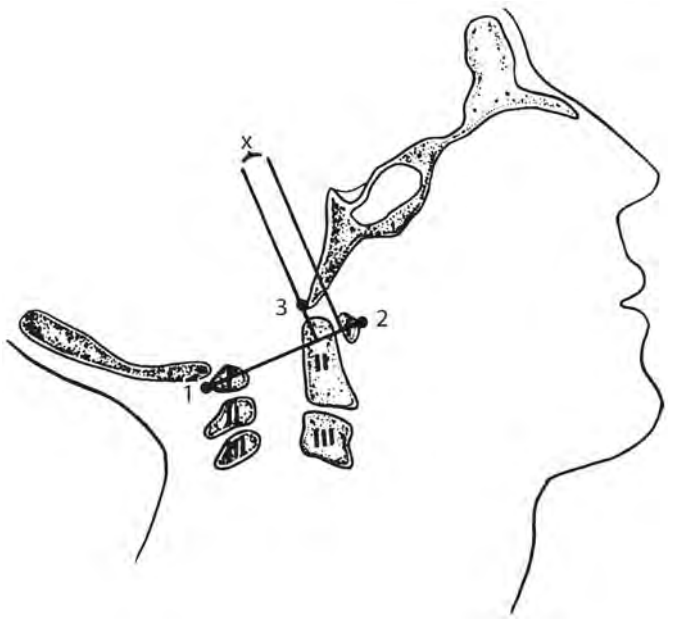


FIGURE 21-5. The method of measuring atlantooccipital instability according to Weisel and Rothman (42). The atlantal line joins points *1* and *2*. A perpendicular line to the atlantal line is made at the posterior margin of the anterior arch of the atlas. The distance (*x*) from the basion (*3*) to the perpendicular line is measured in flexion and extension. The difference between flexion and extension represents the AP translation at the occipitoatlantal joint; in normal adults, this translation should be no more than 1 mm. (From Gabriel KR, Mason DE, Carango P. Occipito-atlantal translation in Down's syndrome. *Spine* 1990;15:996–1002, with permission.)

as the distance between the anterior margin of the condyles at the base of the skull and the sharp contour of the anterior aspect of the concave joint of the atlas anteriorly, or as the distance between the occipital protuberance and the superior arch of the atlas posteriorly. Another method to measure posterior subluxation of the atlantooccipital joint is that of Wiesel and Rothman (42) (Fig. 21-5). With this technique, occiput-C1 translation from maximum flexion to maximum extension should be no more than 1 mm in normal adults. These norms in children have not yet been established.

Pseudosubluxation. The C2-C3, and to a lesser extent, the C3-C4 interspace in children, have a normal physiologic displacement. In a study of 161 children (35), marked anterior displacement of C2 on C3 was observed in 9% of children between 1 and 7 years old. In a more recent study, 22% of 108 polytrauma children demonstrated pseudosubluxation, and had no association with intubation status or injury severity (43). In some children, the anterior physiologic displacement of C2 on C3 is so pronounced that it appears pathologic (pseudosubluxation). To differentiate this from pathologic subluxation, Swischuk (44) has used the posterior cervical line (Fig. 21-6) drawn from the anterior cortex of the posterior arch of C1 to the anterior cortex of the posterior arch of C3. In physiologic displacement of C2 on C3, the posterior cervical line may pass through the cortex of the posterior arch of C2, touch the anterior aspect of the cortex of the posterior arch of C2, or come within 1 mm of the anterior cortex of the posterior arch of C2. In pathologic dislocation of C2 on C3, the posterior cervical line misses the posterior arch of C2 by 2 mm or more.

The planes of the articular facets change with growth. The lower cervical spine facets change from 55 to 70 degrees, whereas the upper facets (i.e., C2-C4) may have initial angles as low as 30 degrees, which gradually increase to 60 to 70 degrees. This variation in facet angulation, along with normal looseness of the soft tissues, intervertebral discs, and the relative increase in size and weight of the skull compared with the trunk, are the major factors responsible for this pseudosubluxation. No treatment is needed for this normal physiologic subluxation.

Variations in the Curvature and Growth of the Cervical Spine That Can Resemble Injury. In the classic study of Cattell and Filtzer (35), 16% of normal children showed a marked angulation at a single interspace, suggestive of injury to the interspinous or posterior longitudinal ligament; 14% showed an absence of the normal lordosis in the neutral position; and 16% showed an absence of the flexion curvature between the second and the seventh cervical vertebrae, which could be erroneously interpreted as splinting secondary to injury. These findings may occur in children up to 16 years of age.

Spina bifida of the posterior arch, or multiple ossification centers of the ring of C1, may mimic fractures. They can be delineated from fractures by their smooth cortical margins. In some children, the posterior ring of C1 remains cartilaginous, which is usually of no clinical significance (45, 46). Spina bifida also may occur at other cervical levels, and the overlapping lucent areas on anteroposterior (AP) radiographs when crossing a vertebral body, may mimic a vertical fracture of the body. Defects in the posterior arch of the atlas are present in 3.7% of the normal population (47).

The dentocentral synchondrosis of C2 begins to close between 5 and 7 years of age (28). However, it may be visible in vestigial forms up to 11 years of age (35) and may be erroneously interpreted as an undisplaced fracture. Similarly, the apical odontoid epiphysis (i.e., ossiculum terminale) may appear by 5 years of age, although it most typically appears around 8 years of age. This also can be misinterpreted as an odontoid tip fracture.

Wedging of the C3 vertebral body is a normal radiographic finding in 7% of younger children (Fig. 21-6B); the wedging corrects as the child matures and is extremely rare after age 13 (48). If it is unclear whether the wedging is a normal variation or a true compression fracture in the face of a traumatic history, a computed tomography (CT) scan will demonstrate fracture lines through the body if a fracture is present. In the lower cervical levels, secondary centers of ossification of the spinous processes may resemble avulsion fractures (35).

Normal Lower Cervical Spine Motion. Generally, the interspinous distances increase with increasing age, being the smallest at C4-C5 and the largest at C6-C7, until 15 years

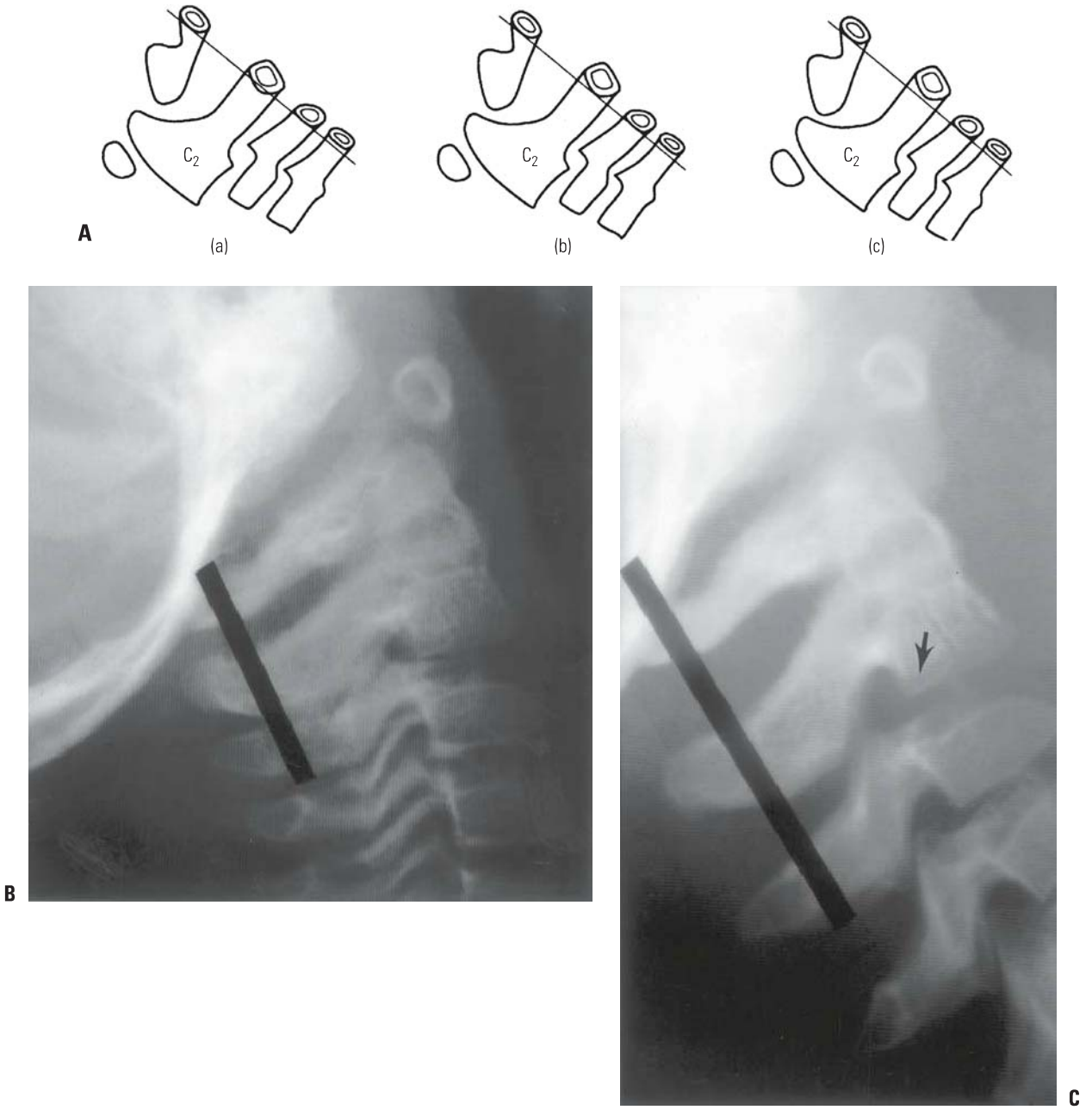


FIGURE 21-6. **A:** The posterior cervical line of Swischuk. In C2-C3 pseudosubluxation, the posterior cervical line may pass through *(a)*, touch *(b)*, or lie 1 mm in front of the cortex of the posterior arch of C2. (From Shaw M, Burnett H, Wilson A, et al. Pseudosubluxation of C2 on C3 in polytraumatized children—prevalence and significance. *Clin Radiol* 1999;54:377–380, with permission). **B,C:** Lateral cervical radiograph of a 2-year, 6-month-old child with pseudosubluxation at C2-C3. The radiograph in extension (B) demonstrates no step-off at C2-C3, whereas the radiograph in flexion (C) demonstrates a step-off at C2-C3 (*arrow*), but with a normal posterior cervical line (*solid line*). Also note the anterior wedging of the C3 vertebral body. Similarly, note the overriding of the anterior arch of the atlas on the tip of the odontoid in extension.

of age, when this distance is largest at C5-C6 (34). The AP displacement, from hyperflexion to hyperextension, decreases from C2-C3 to C6-C7. The angular displacement is greatest (15 degrees) at C3-C4 and C4-C5 for children 3 to 8 years of age, is greatest (17 degrees) at C4-C5 for children 9 to 11 years of age, and is greatest (15 degrees) at C5-C6 for children 12 to 15 years of age.

CONGENITAL AND DEVELOPMENTAL PROBLEMS

Torticollis. Torticollis is a combined head tilt and rotatory deformity. Torticollis indicates a problem at C1-C2 because 50% of the cervical spine rotation occurs at this joint. A head tilt alone indicates a more generalized problem in the cervical spine.

The differential diagnosis of torticollis is large and can be divided into osseous and nonosseous types. In a recent large series from a tertiary care pediatric orthopaedic center (49), a nonmuscular etiology of torticollis was found in 18% of patients, most frequently the Klippel-Feil syndrome or a neurologic disorder (ocular pathology, or central nervous system lesion).

Osseous Types. Occipitocervical synostosis, basilar impression, and odontoid anomalies are the most common congenital/developmental malformations of the occipitovertebral junction, with an incidence of 1.4 to 2.5 per 100 children (50). These lesions arise from a malformation of the mesenchymal anlages at the occipitovertebral junction.

Basilar Impression. Basilar impression is an indentation of the skull floor by the upper cervical spine. The tip of the dens is more cephalad and sometimes protrudes into the opening of the foramen magnum. This may encroach on the brain stem, risking neurologic damage from direct injury, vascular compromise, or alterations in cerebrospinal fluid flow (51).

Basilar impression can be primary or secondary. Primary basilar impression, the most common type, is a congenital abnormality often associated with other vertebral defects (e.g., Klippel-Feil syndrome, odontoid abnormalities, atlantooccipital fusion, and atlas hypoplasia). The incidence of primary basilar impression in the general population is 1% (52).

Secondary basilar impression is a developmental condition attributed to softening of the osseous structures at the base of the skull. Any disorder of osseous softening can lead to secondary basilar impression. These include metabolic bone diseases [e.g., Paget disease (53), renal osteodystrophy, rickets, osteomalacia (54, 55)] bone dysplasias and mesenchymal syndromes [e.g., osteogenesis imperfecta (56–62), achondroplasia (63), hypochondroplasia (64), neurofibromatosis (65), and rheumatologic disorders, e.g., rheumatoid arthritis, ankylosing spondylitis]. The softening allows the odontoid to migrate cephalad and into the foramen magnum.

These patients typically present with a short neck (78% in one series) (66). This shortening is only an apparent deformity because of the basilar impression. Asymmetry of the skull and face (68%), painful cervical motion (53%), and torticollis (15%) can also occur. Neurologic signs and symptoms are often present (67). Many children will have acute onset of symptoms precipitated by minor trauma (68). In cases of isolated basilar impression, the neurologic involvement is primarily a pyramidal syndrome associated with proprioceptive sensory disturbances (motor weakness, 85%; limb paresthesias, 85%). In cases of basilar impression associated with Arnold-Chiari malformations, the neurologic involvement is usually cerebellar, and symptoms include motor incoordination with ataxia, dizziness, and nystagmus. In both types, the patients may complain of neck pain and headache in the distribution of the greater occipital nerve and cranial nerve involvement, particularly those that emerge from the medulla oblongata (trigeminal [V], glossopharyngeal [IX], vagus [X], and hypoglossal [XII]). Ataxia is a very common finding in children with basilar impression (68). Hydrocephalus may develop as a result of obstruction of the cerebrospinal fluid flow by obstruction of the foramen magnum from the odontoid.

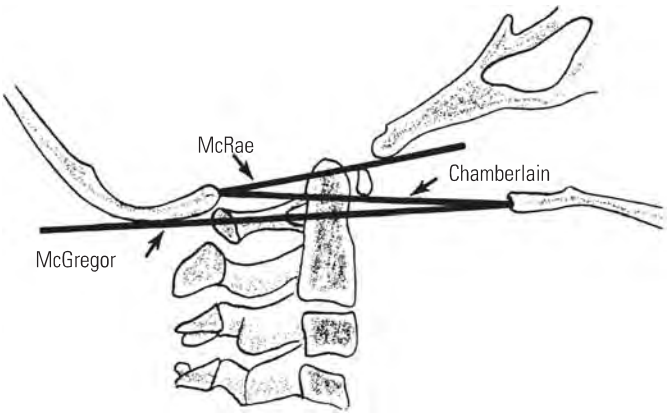


FIGURE 21-7. The landmarks on a lateral radiograph of the skull and the upper cervical spine used to assess basilar impression. McRae line defines the opening of the foramen magnum. Chamberlain line is drawn from the posterior lip of the foramen magnum to the dorsal margin of the hard palate. McGregor line is drawn from the upper surface of the posterior edge of the hard palate to the most caudal point of the occipital curve of the skull. McGregor line is the best line for screening because of the clarity of the radiographic landmarks in children of all ages.

Basilar impression is difficult to assess radiographically. The most commonly used lines are Chamberlain (69), McRae (70), and McGregor (71) (Fig. 21-7). McGregor line is the best line for screening because the landmarks can be clearly defined at all ages on a routine lateral radiograph. McRae line is helpful in assessing the clinical significance of basilar impression because it defines the opening of the foramen magnum; in patients who are symptomatic, the odontoid projects above this line. At present, CT with sagittal plane reconstructions can show the osseous relations at the occipitocervical junction more clearly, and magnetic resonance imaging (MRI) clearly delineates the neural anatomy. MRI and CT norms have been recently established (72, 73). Occasionally, vertebral angiography or MR angiogram (MRA) is needed (74).

Treatment of basilar impression can be difficult and requires a multidisciplinary approach (orthopaedic, neurosurgical, and neuroradiologic) (59, 61, 75–78). The symptoms rarely can be relieved with customized orthoses (79); the primary treatment is surgical. If the symptoms are caused by a hypermobile odontoid, surgical stabilization in extension at the occipitocervical junction is needed. Anterior excision of the odontoid is needed if it cannot be reduced (80), but this should be preceded by posterior stabilization and fusion. In some cases, the anterior decompression can be performed endoscopically (81). If the symptoms are from posterior impingement, suboccipital decompression and often upper cervical laminectomy are needed. The dura often needs to be opened to look for a tight posterior band (66, 82). Posterior stabilization also should be performed (83). In a recent series of 190 cases, decompression of the foramen magnum was appropriate for those without an

Arnold-Chiari malformation; transoral anterior decompression was reserved for those with an associated Arnold-Chiari malformation (84). These are general statements, and each case must be considered individually. Secondary basilar impression tends to progress despite arthrodesis (61).

Atlantooccipital Anomalies. Children with congenital bony anomalies of the atlantooccipital junction present with a wide spectrum of deformities (85). The anterior arch of C1 is commonly assimilated to the occiput, usually in association with a hypoplastic ring posteriorly (Fig. 21-8), as well as condylar hypoplasia (86). The height of C1 is variably decreased, allowing the odontoid to project upward into the foramen magnum (i.e., primary basilar impression). More distal cervical anomalies can also occur in association with the atlantooccipital anomaly. The odontoid may be misshapen or directed posteriorly more than normal. Up to 70% of children with this condition have a congenital fusion of C2-C3 (see Fig. 21-8). (Posterior congenital fusion of C2-C3 is a clue that occiput-C1 anomalies, or other more distal cervical fusions, may be present. These may be cartilaginous initially, and not appear on plain radiographs until the child becomes more mature.) The fusion of C1 to the occiput is classified by zones (86); zone 1 is a fused anterior arch, zone 2 a fusion of the lateral masses, zone 3 a fused posterior arch, and zone 4 a combination of zones. Zone 4 fusions are most common, but the highest prevalence of spinal canal encroachment is in zone 2 patients.

Clinically, these children resemble those with the Klippel-Feil syndrome: short, broad necks; restricted neck motion; low hairline; high scapula; and torticollis (Fig. 21-9) (82, 87–89). Recently, hemifacial microsomia has been noted to have associated atlantooccipital anomalies (90), as well as children with the 22q11.2 deletion syndrome (91). The skull may demonstrate a positional deformational plagiocephaly. They also may have other associated anomalies, including dwarfism, funnel chest, jaw anomalies, cleft palate, congenital ear deformities, hypospadias, genitourinary tract defects, and syndactyly. They can present with neurologic symptoms during childhood, but more often present between 40 to 50 years of age. These symptoms can be initiated by traumatic or inflammatory processes, and they progress slowly and relentlessly. Rarely do they present suddenly or dramatically, although they have been reported as a cause of sudden death. The most common signs and symptoms, in decreasing order of frequency, are neck and

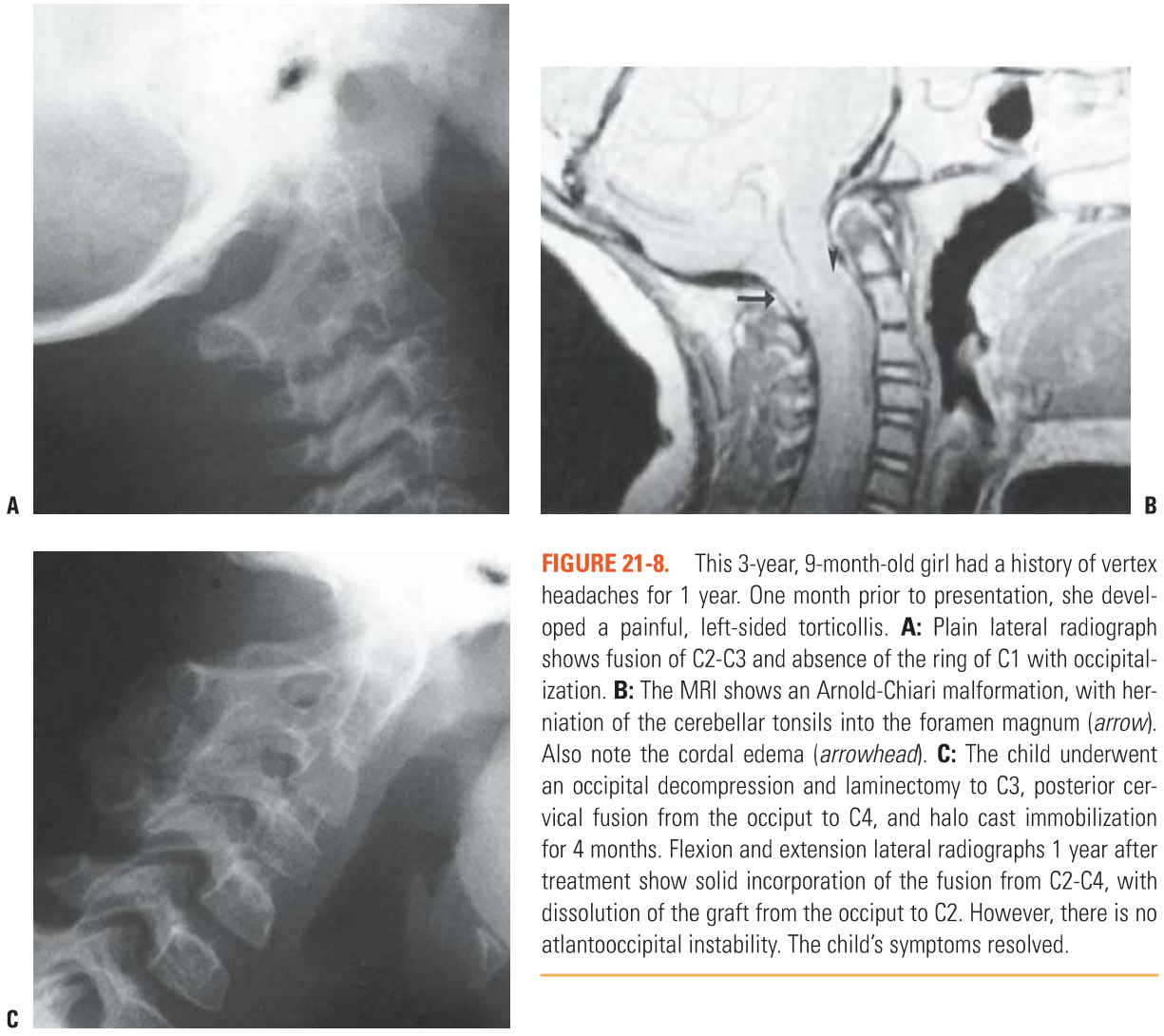


FIGURE 21-8. This 3-year, 9-month-old girl had a history of vertex headaches for 1 year. One month prior to presentation, she developed a painful, left-sided torticollis. **A:** Plain lateral radiograph shows fusion of C2-C3 and absence of the ring of C1 with occipitalization. **B:** The MRI shows an Arnold-Chiari malformation, with herniation of the cerebellar tonsils into the foramen magnum (*arrow*). Also note the cordal edema (*arrowhead*). **C:** The child underwent an occipital decompression and laminectomy to C3, posterior cervical fusion from the occiput to C4, and halo cast immobilization for 4 months. Flexion and extension lateral radiographs 1 year after treatment show solid incorporation of the fusion from C2-C4, with dissolution of the graft from the occiput to C2. However, there is no atlantooccipital instability. The child's symptoms resolved.

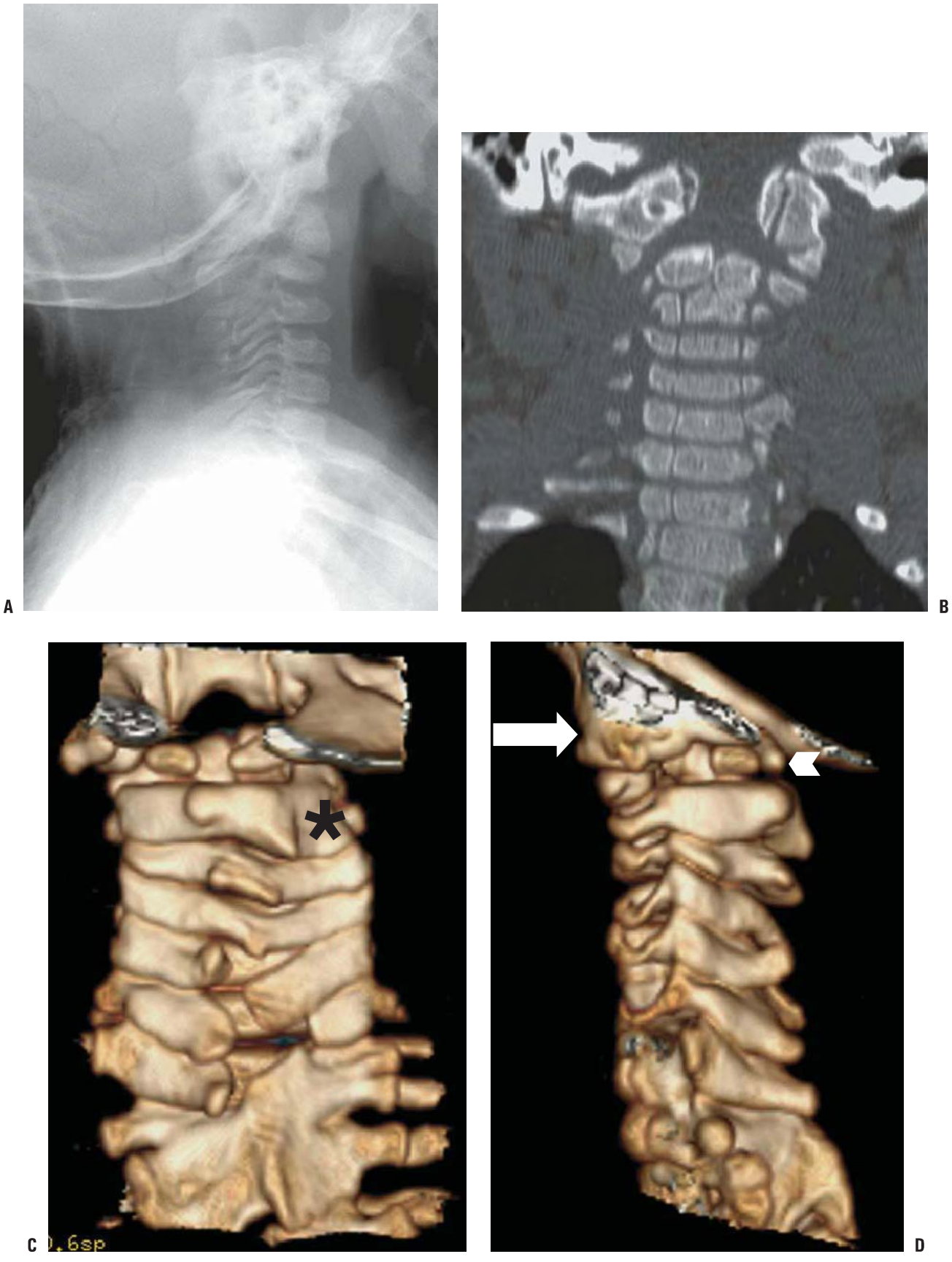


FIGURE 21-9. This 14-month-old girl presented with a history of torticollis since birth, with no response to physical therapy. Prenatal and birth history was unremarkable. Physical exam demonstrated a short neck with no sternocleidomastoid muscle contracture. **A:** A lateral cervical spine radiograph demonstrates overlap of the posterior elements and absence of the anterior portion of the C1 ring. **B:** A CT scan demonstrates multiple abnormal ossification centers of C2. **C:** A three-dimensional CT scan demonstrates fusion of the lateral masses and posterior elements of C2-C3 on the right (*asterisk*) and **D:** Fusion of the anterior atlas and skull base (*arrow*) with the posterior arch of C1 in a normal position (*arrowhead*).

occipital pain, vertigo, ataxia, limb paresis, paresthesias, speech disturbances, hoarseness, diplopia, syncope, auditory malfunction, and dysphagia (92, 93).

Standard radiographs are difficult to obtain because of fixed bony deformities and overlapping shadows from the mandible, occiput, and foramen magnum. An x-ray beam directed 90 degrees perpendicular to the skull (rather than the cervical spine) usually gives a satisfactory view of the occipitocervical junction. The anomaly usually is studied further with CT. In young children, the head-wag autotomography technique can be quite useful (94). This technique involves side-to-side rotation of the child's head, while a slow AP radiographic exposure of the upper cervical spine is performed. This rotation blurs the overlying head and mandibular structures, allowing for improved visualization of the occiput-C1-C2 complex.

The position of the odontoid relative to the opening of the foramen magnum has been described by measuring the distance from the posterior aspect of the odontoid to the posterior ring of C1 or the posterior lip of the foramen magnum, whichever is closer (87, 95). This should be determined in flexion because this position maximizes the reduction in the SAC. If this distance is <19 mm, a neurologic deficit is usually present. Lateral flexion and extension views of the upper cervical spine often show up to 12 mm of space between the odontoid and the C1 ring anteriorly (87); associated C1-C2 instability has been reported to develop eventually in 50% of these patients.

MRI is used to image the neural structures. Flexion-extension MRI is often necessary to fully evaluate the pathology (96). Compression of the brain stem or upper cervical cord anteriorly occurs from the backward-projecting odontoid. This produces a range of findings and symptoms, depending on the location and degree of compression. Pyramidal tract signs and symptoms (e.g., spasticity, hyperreflexia, muscle weakness, gait disturbances) are most common, although signs of cranial nerve involvement (e.g., diplopia, tinnitus, dysphagia, auditory disturbances) can be seen. Compression from the posterior lip of the foramen magnum or dural constricting band can disturb the posterior columns, with a loss of proprioception, vibration, and tactile senses. Nystagmus also occurs frequently as a result of posterior cerebellar compression. Vascular disturbances from vertebral artery involvement can result in brain stem ischemia, manifested by syncope, seizures, vertigo, and unsteady gait (97). Cerebellar tonsil herniation can occur. The altered mechanics of the cervical spine may result in a dull, aching pain in the posterior occiput and the neck with intermittent stiffness and torticollis. Irritation of the greater occipital nerve may cause tenderness in the posterior scalp.

The natural history of atlantooccipital anomalies is unknown. The neurologic symptoms may develop so late and progress so slowly because the frequently associated C1-C2 instability progresses with age, and the increased demands placed on the C1-C2 interval produce gradual spinal cord or vertebral artery compromise.

Treatment is difficult. Surgery for atlantooccipital anomalies is more risky than with isolated anomalies of the odontoid (82, 93). For this reason, nonoperative methods should be initially attempted. Cervical collars, braces, and traction often help for persistent complaints of head and neck pain, especially after minor trauma or infection. Immobilization may achieve only temporary relief if neurologic deficits are present. Patients with evidence of a compromised upper cervical area should take precautions not to expose themselves to undue trauma.

When symptoms and signs of C1-C2 instability are present, a posterior C1-C2 fusion is indicated. Preliminary traction to attempt reduction is used if necessary. If a reduction is possible and there are no neurologic signs, surgery has an improved prognosis (82, 92, 93). Posterior signs and symptoms may be an indication for posterior decompression depending on the evidence of dural or osseous compression. Results vary from complete resolution to increased deficits and death (82, 98). In the instance of no instability but only compressive pathology, the role of concomitant posterior fusion has not yet been determined. However, if decompression, whether anterior or posterior, can destabilize the spine, then concomitant posterior fusion should be considered (86, 99, 100).

If occiput-C1 instability is present, then occiput–atlanto–cervical fusion is indicated (Figs. 21-10 to 21-16). These cases are often associated with posterior decompressions/laminectomies.

Occipitoatlantocervical instability requiring occipitocervical fusion is often complicated by the fact that decompression of the base of the skull and the upper two cervical vertebrae is either required or the cause of the instability. These circumstances compromise the ability to achieve fusion because of the lack of bone surface to form a bed for the grafts, the large gap that must be bridged, and the instability (101). In addition, it has been demonstrated that the conventional cervical orthoses do not provide much immobilization for the upper cervical spine (102).

To circumvent these problems and to provide stability to the fusion area, surgeons have developed methods of internal fixation (103, 104). Since these reports, fixation using screws and plates has become more popular in many centers, but these techniques require a considerable learning curve and are often not amenable to the very small child with small anatomy or the dysplastic anatomy secondary to dysplasia/dwarfisms. In the small child, a useful technique that has proved effective is the use of rib or other cortical-cancellous graft to provide an element of stability, as well as to serve as the graft for the fusion. This is supplemented by the use of a halo orthosis. A combined team approach with neurosurgery is often helpful in these complicated cases.

Unilateral Absence of C1. This congenital malformation of the first cervical vertebra is, in essence, a hemiatlas or a congenital scoliosis of C1. Doubousset (105) described 17 patients with this absence. No definite population incidence is known. The problem often is associated with other anomalies common to children with congenital spine deformities (e.g., tracheoesophageal fistula).

Two-thirds of the children present at birth; the others develop torticollis and are noticed later. A lateral translation of the head on the trunk, with variable degrees of lateral tilt and rotation (best appreciated from the back) is the typical finding.

Text continued on page 838

Occipitocervical Facet Fusion After Laminectomy (Figs. 21-10 to 21-16).

FIGURE 21-10. The patient is positioned in the prone position. If the halo has not been applied to the skull, it is done before the patient is turned prone. The halo may be attached to the operating table by use of a specially designed attachment, or it may be attached to a traction, depending on the needs of the case.

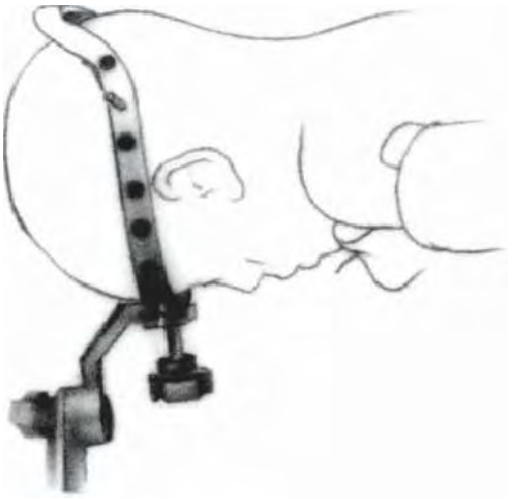
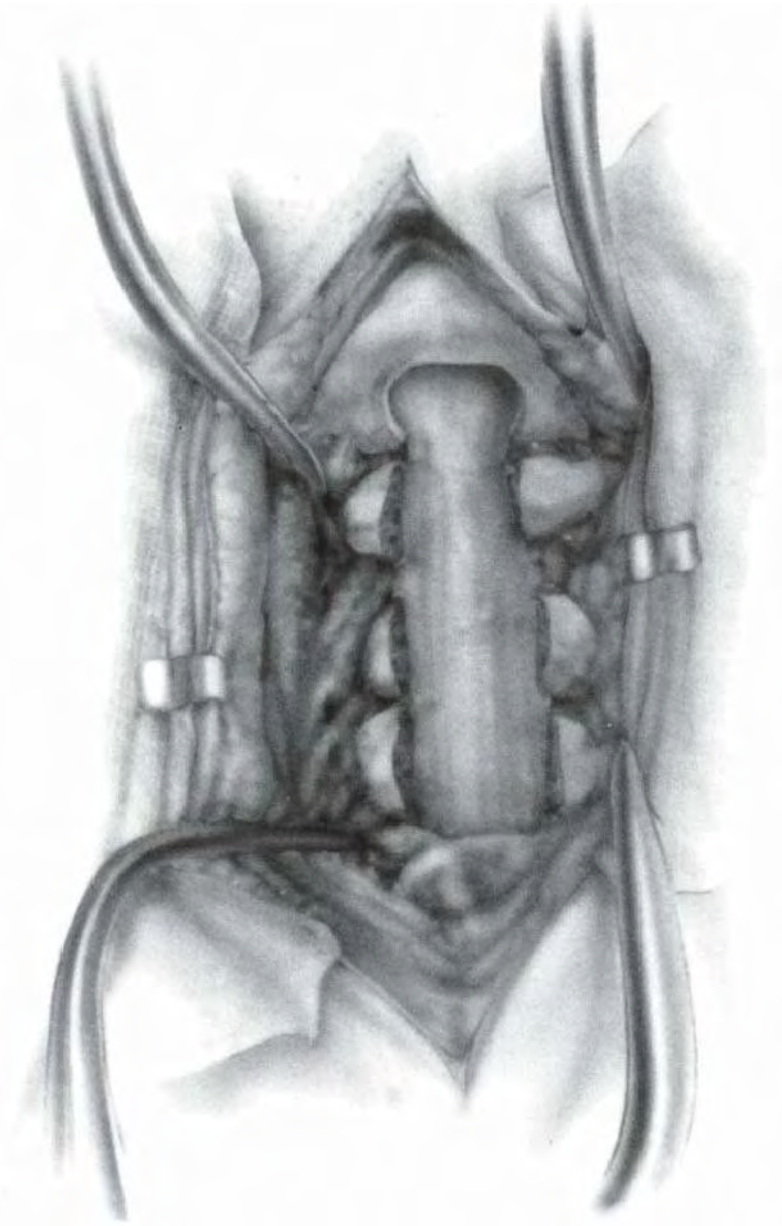


FIGURE 21-11. After the usual skin preparation and draping, a midline incision is used to expose the base of the skull and the cervical vertebrae to be fused. It is important to carry the subperiosteal dissection far enough laterally to have sufficient bone remaining after the laminectomy but not so far as to get into the venous plexus lateral to the facet joints or especially the vertebral artery at the C1 level. In the average-sized adult, the dissection should not proceed further than 1.5 cm lateral to the midline, to avoid the vertebral artery as it crosses the arch of C1. In the child, 1 cm is a safe limit to observe. The laminectomy and suboccipital decompressions are performed as indicated.



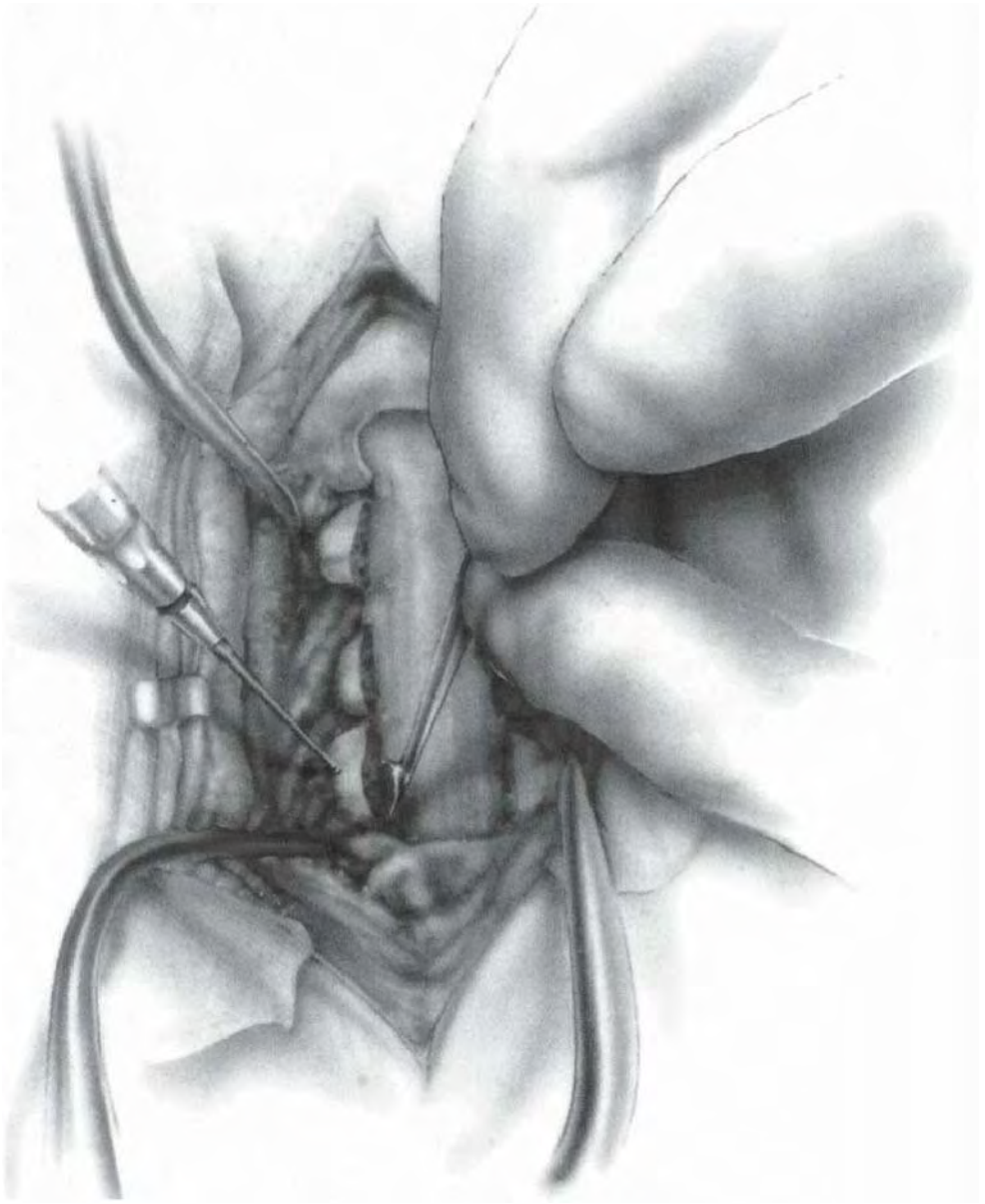


FIGURE 21-12. After completion of the laminectomy, a small periosteal elevator is used to dissect the periosteum off the underside of the remaining portions of the lamina. A small thin elevator is then placed under the lamina to protect the dura. Using a high-speed drill, a small hole is made in each lamina through which a wire is passed. Care must be taken to be certain that the elevator is under the portion of the lamina that the drill will penetrate. If the occiput is included in the fusion, holes are made in the occiput. It is extremely important that this is done by someone with expertise in this field who is familiar with the anatomy of the base of the skull because the large venous sinuses in this area must be avoided.

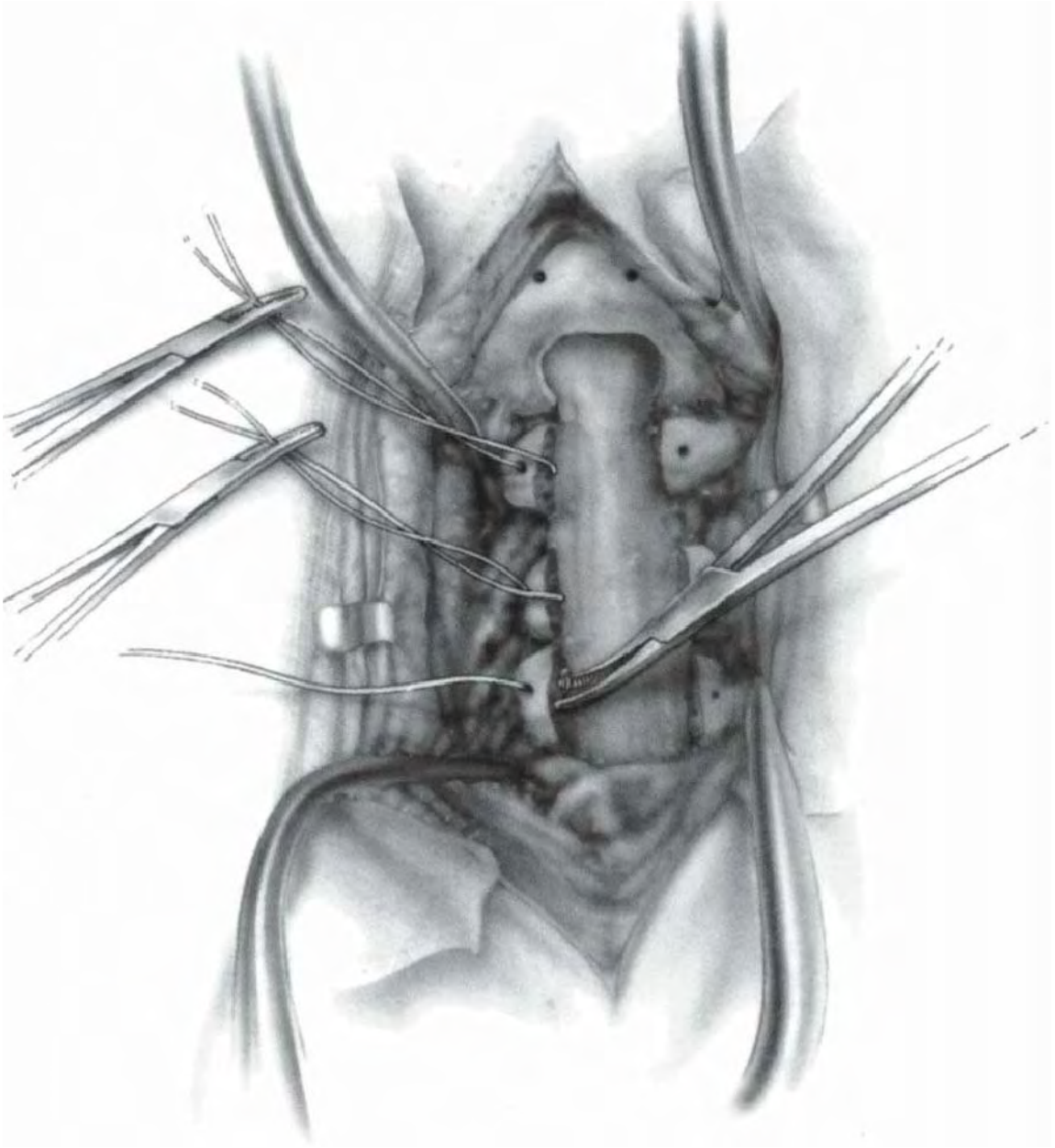


FIGURE 21-13. Depending on the size of the child and the lamina, a flexible wire (22-gauge), braided wire, or flexible cable is passed through each of the holes. If a wire is used, a small hemostat is used to reach under the lamina to grasp it as it comes through. After it is grasped, the wire is turned acutely so that it does not tear or puncture the dura, and it is then pulled through. If the child is large enough, braided cables are preferred because they are flexible and easier to use than wire.

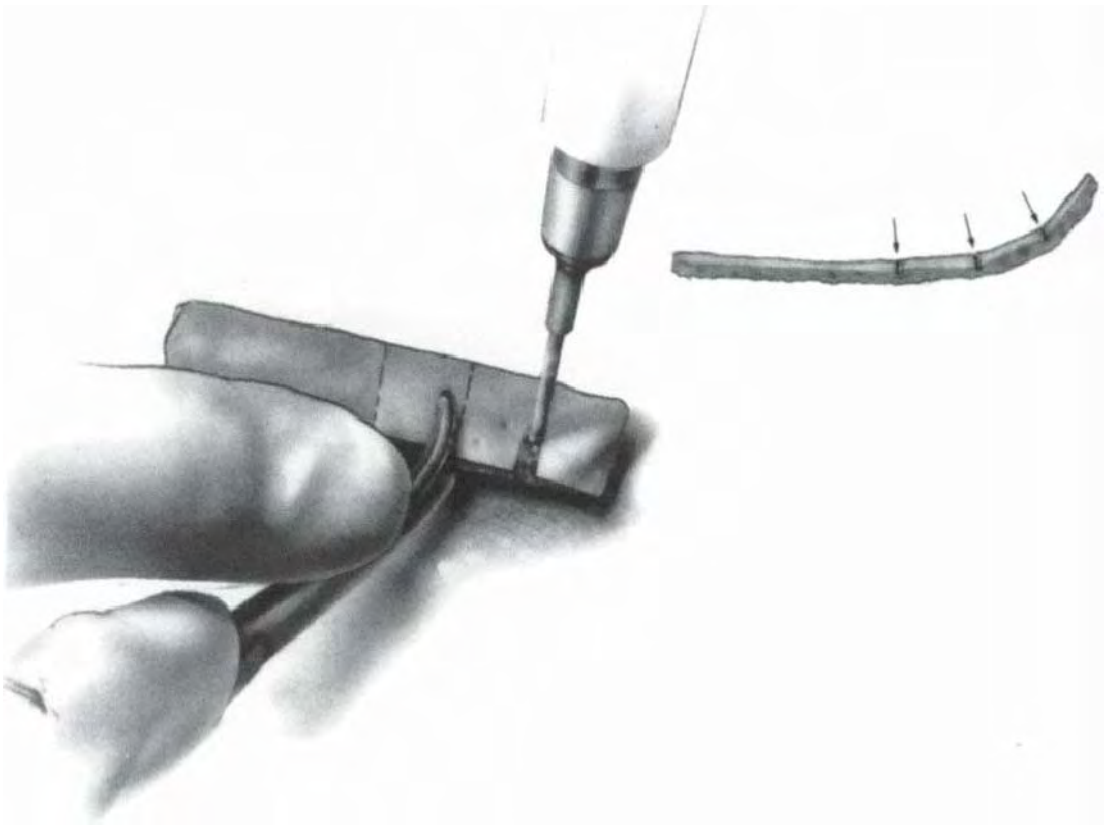


FIGURE 21-14. The bone graft can be either rib or corticocancellous bone from the iliac crest. The advantage of corticocancellous bone is that the thick portion of cancellous bone can be placed directly against the lamina. Rib graft has the advantage of strength, which imparts additional stability to the spine. Although either the rib or a portion of the iliac crest may have the general contour desired for the cervical spine, it is usually necessary to bend the graft so that more curve is present to bring the graft in contact with the C1 lamina before it turns up onto the skull. This may be accomplished by kerfing the graft. A *kerf* is a cut or channel made by a saw. This can be accomplished by use of the high-speed drill to cut through the cortex on the concave side of the curve in the graft. After the graft has been harvested, cut, and bent to the desired shape, holes are placed in it to correspond to the holes in the lamina over which it will lie. This can be done by holding the graft over the operative site while estimating the correct placement of the hole or by marking the site with a marking pen and then removing the graft to the back table to cut the holes.

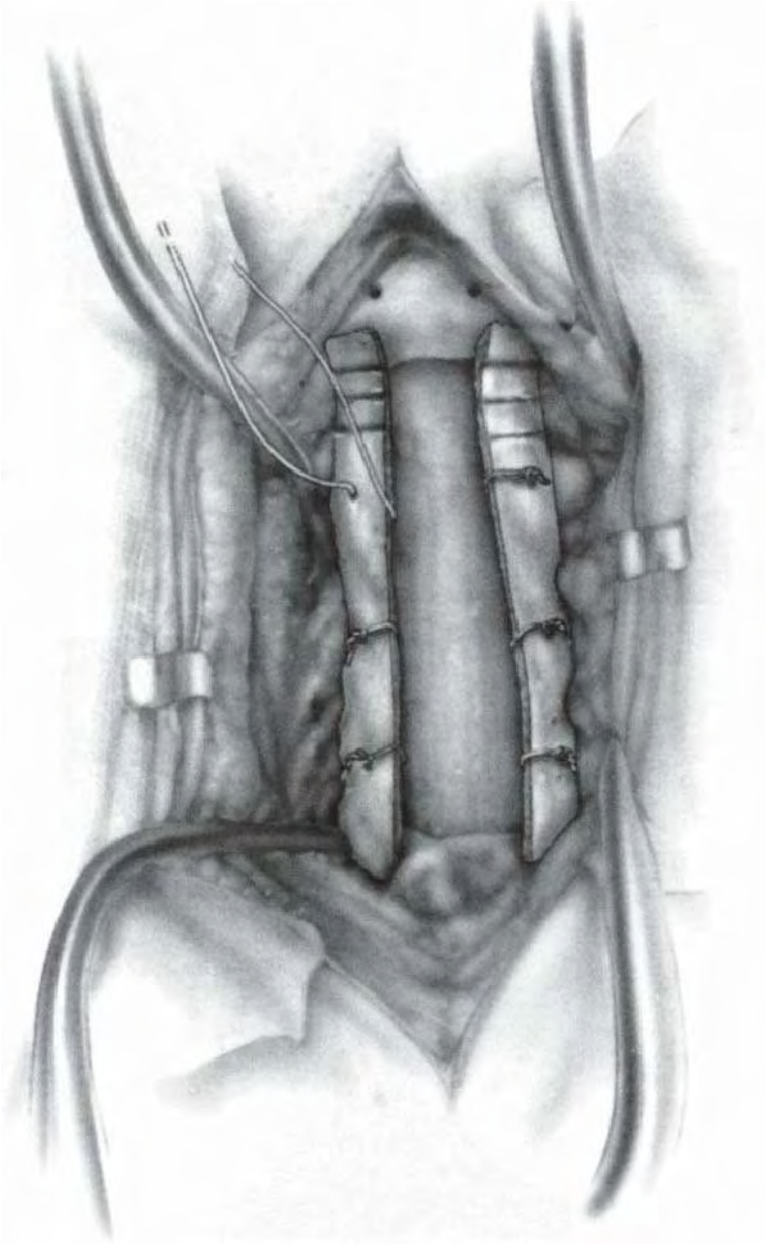


FIGURE 21-15. The portion of the wire or cable coming through the surface of the lamina is drawn through the bone graft. The segment of the wire coming from under the lamina is brought around the graft. This pushes the graft laterally and prevents it from coming to lie over the dura. If there is no additional internal fixation, the graft is held firmly against the lamina while the wires or cables are tightened.

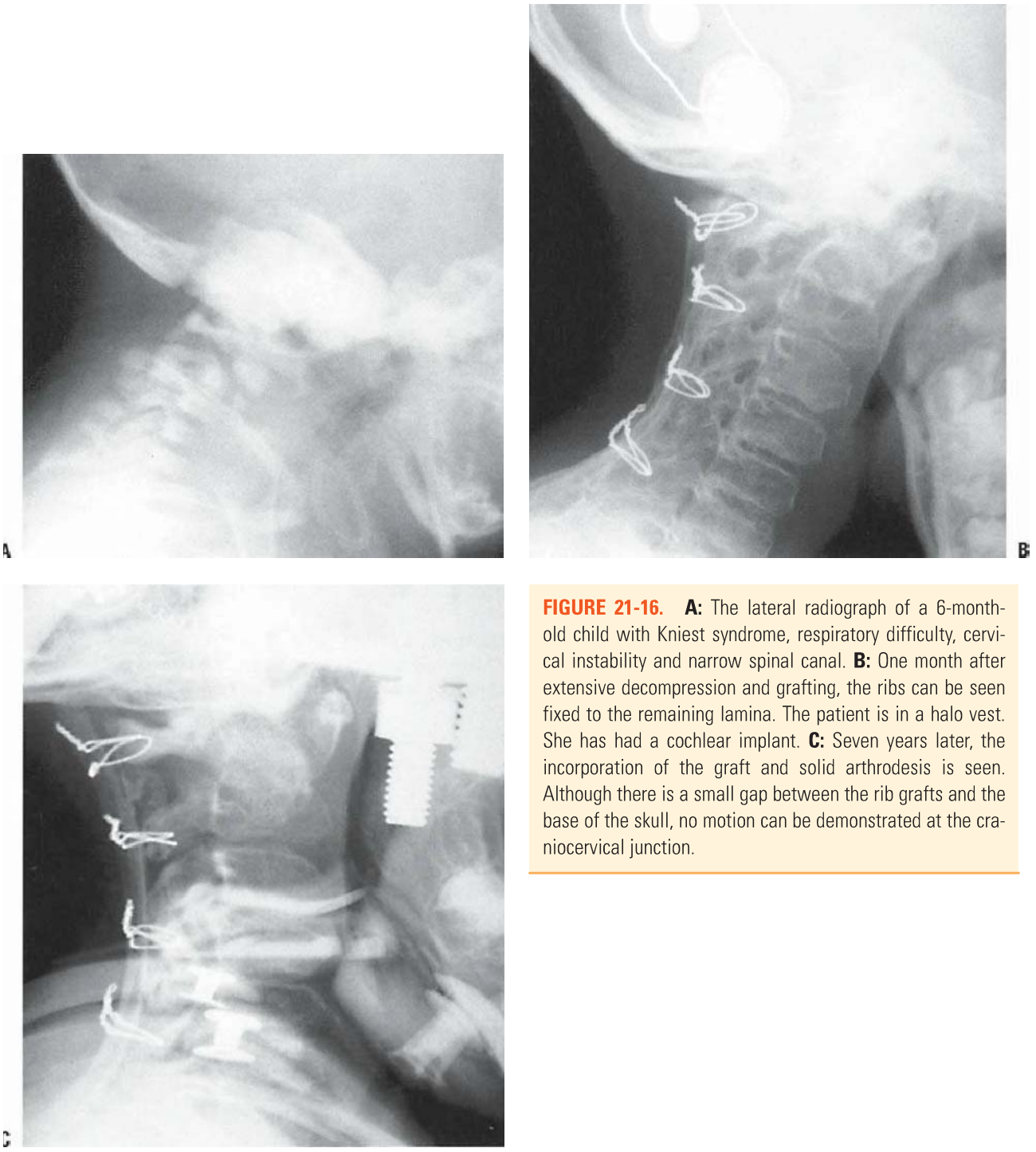


FIGURE 21-16. **A:** The lateral radiograph of a 6-month-old child with Kniest syndrome, respiratory difficulty, cervical instability and narrow spinal canal. **B:** One month after extensive decompression and grafting, the ribs can be seen fixed to the remaining lamina. The patient is in a halo vest. She has had a cochlear implant. **C:** Seven years later, the incorporation of the graft and solid arthrodesis is seen. Although there is a small gap between the rib grafts and the base of the skull, no motion can be demonstrated at the craniocervical junction.

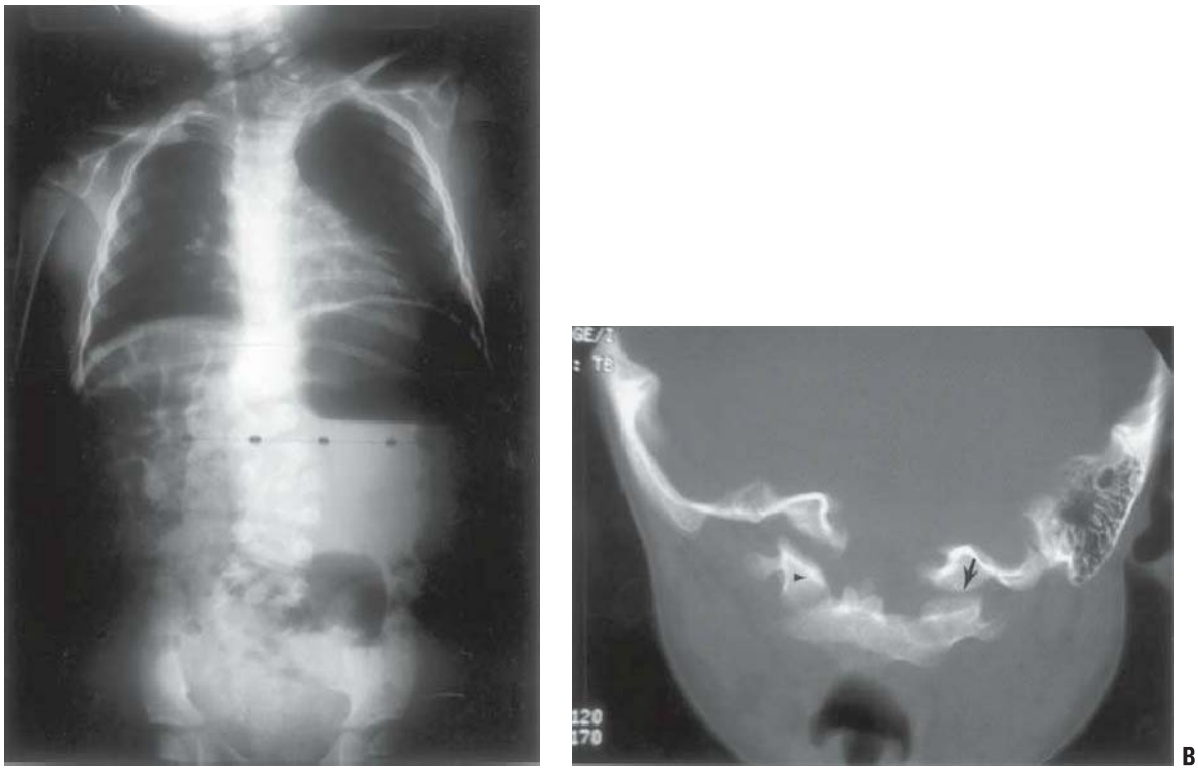
There also may be severe tilting of the eye line. The sternocleidomastoid muscle is not tight, although regional aplasia of the muscles in the nuchal concavity of the tilted side is noted. Neck flexibility is variable and decreases with age. The condition is not painful. Plagiocephaly can occur and increases as the deformity increases. Neurologic signs (e.g., headache, vertigo, myelopathy) are present in about one-fourth of the patients. The natural history is unknown.

Standard AP and lateral radiographs rarely give the diagnosis, although the open-mouth odontoid view may suggest it. Tomograms or CT scans usually are needed to see the anomaly (Fig. 21-17). The defect can range from a hypoplasia of the lateral mass to a complete hemiatlas with rotational instability and basilar impression. Occasionally, the atlas is occipitalized. There are three types of this disorder. Type I is an isolated hemiatlas. Type II is a partial or complete aplasia of one hemiatlas, with other associated anomalies of the cervical spine (e.g., fusion of C3-C4, congenital bars in the lower cervical vertebrae). Type III is a partial or complete atlantooccipital fusion and symmetric or asymmetric hemiatlas aplasia, with or without anomalies of the odontoid and the lower cervical vertebrae.

Once this malformation is diagnosed, entire spinal radiographs should be taken to rule out other congenital vertebral anomalies. Other imaging studies that may be needed are vertebral angiography and MRI. Angiography should be performed if operative intervention is undertaken, because arterial anomalies (e.g., multiple loops, vessels smaller than normal, abnormal routes between C1 and C2) often are found on the aplastic side. MRI also should be performed if operative intervention is undertaken, because many of these children will have stenosis of the foramen magnum, and occasionally an Arnold-Chiari malformation.

The deformity should be observed to document the presence or absence of progression. This observation is primarily clinical (e.g., photographs) because radiographic measurements are difficult if not impossible to obtain. Bracing does not halt progression of the deformity. Surgical intervention is recommended in those patients with severe deformities. A preoperative halo is used for gradual traction correction over 6 to 8 days. An ambulatory method of gradual cervical spine deformity correction has been described using the halo-Ilizarov technique (106). A posterior fusion from the occiput-C2 or -C3 is then performed, depending on the extent of the anomaly. Decompression of the spinal canal is necessary when the canal size is not ample, either at that time or if it is projected not to be able to fully accommodate the developed spinal cord. The ideal age for posterior fusion is between the ages of 5 and 8 years, corresponding to the age at which the canal size reaches adult proportions.

Familial Cervical Dysplasia. The epidemiology of this atlas deformity (107) is not known. Clinical presentation varies from an incidental finding to a passively correctable head tilt,



A B

FIGURE 21-17. This 4-year, 10-month-old boy presented with a torticollis. **A:** The standing AP radiograph of his entire spine documents the head tilt to the left, along with left sided hemivertebrae in the left lower cervical spine. Also note the multiple hemivertebrae in the thoracic and lumbar spine. **B:** A CT scan with frontal reconstruction clearly demonstrates an absent lateral mass of C1 (*arrow*) with a normal right lateral mass (*arrowhead*). This thus represents a Doubosset-type II C1 unilateral absence.

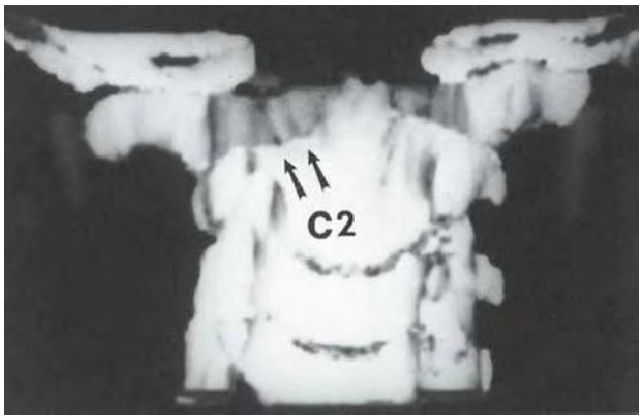


FIGURE 21-18. A three-dimensional computed tomographic scan of the upper cervical cord in a child with familial cervical dysplasia. The left superior facet of C2 is shallow and hypoplastic (*arrows*). (From Saltzman CL, Hensinger RN, Blane CE, et al. Familial cervical dysplasia. *J Bone Joint Surg Am* 1991;73-A:163–171, with permission.)

suboccipital pain, decreased cervical motion, or a clunking of the upper cervical spine.

Plain radiographs are difficult to interpret. Various anomalies of C1, most commonly a partial absence of the posterior ring of C1, typically are seen. Various anomalies of C2 also commonly exist, for example, a shallow hypoplastic left facet. Other dysplasias of the lateral masses, facets, and posterior elements and occasionally spondylolisthesis are seen. Occiput-C1 instability is frequently seen; C1-C2 instability rarely is seen. The delineation of this complex anatomy often is seen best with a CT scan and a three-dimensional reconstruction (Fig. 21-18). When symptoms of instability are present, MRI in flexion and extension is recommended to assess the presence and magnitude of neural compression. Occipitocervical junction instability due to the malformation may lead to neural compromise.

Nonsurgical treatment consists of observation every 6 to 12 months to ensure that instability does not develop, either clinically (e.g., progressive weakness and fatigue or objective signs of myelopathy), or radiographically, with lateral flexion and extension radiographs. Surgical intervention is recommended for persistent pain, torticollis, and neurologic symptoms. A posterior fusion from the occiput to C2 usually is required, with gradual preoperative reduction using an adjustable halo cast (106).

Atlantoaxial Rotary Displacement. Atlantoaxial rotary displacement is one of the most common causes of childhood torticollis. Rotary displacements are characteristically a pediatric problem, but they may occur in adults. There are several causes. Because the resultant radiographic findings and treatment regimens are the same for all pediatric causes, they are discussed as a unit and individual exceptions are noted where necessary.

The confusing terminology includes rotary dislocation, rotary deformity, rotational subluxation, rotary fixation, and spontaneous hyperemic dislocation (108, 109). "Atlantoaxial rotary subluxation" is probably the most accepted term used in describing the common childhood torticollis. "Subluxation" is misleading, however, because cases of "subluxation" usually present within the normal range of motion of the atlantoaxial joint. "Rotary displacement" is a more appropriate and descriptive term because it includes the entire range of pathology, from mild subluxation to complete dislocation. If the deformity persists, the children present with a resistant and unresolving torticollis that is best termed "atlantoaxial rotary fixation or fixed atlantoaxial displacement." Gradations exist between the very mild, easily correctable rotary displacement and the rigid fixation. Complete atlantoaxial rotary dislocation rarely has been reported in surviving patients.

The radiographic findings of rotary displacement are difficult to demonstrate (110). With rotary torticollis the lateral mass of C1 that has rotated anterior appears wider and closer to the midline (medial offset), whereas the opposite lateral mass is narrower and away from the midline (lateral offset). The facet joints may be obscured because of apparent overlapping. The lateral view shows the wedge-shaped lateral mass of the atlas lying anteriorly where the oval arch of the atlas normally lies, and the posterior arches fail to superimpose because of the head tilt (Fig. 21-19). These findings may suggest occipitalization of C1 because with the neck tilt the skull may obscure C1. The normal relation between the occiput and C1 is believed to be maintained in children with atlantoaxial rotary displacement. A lateral radiograph of the skull may demonstrate the relative positions of C1 and C2 more clearly than a lateral radiograph of the cervical spine. This is because tilting of the head also tilts C1, which creates overlapping shadows and makes interpretation of a lateral spinal radiograph difficult.

The difficulty with plain radiographs is differentiating the position of C1-C2 in a child with subluxation from that in a normal child whose head is rotated, since both give the same picture. Open-mouth views are difficult to obtain and interpret, and the lack of cooperation and the diminished motion on the part of the child often make it impossible to obtain these special views. Cineradiography has been recommended, but the radiation dose is high and patient cooperation may be difficult because of muscle spasms (110, 111). CT scans are helpful in this situation if it is done properly (112). A CT scan, when taken with the head in the torticollic position, may be interpreted by the casual observer as showing rotation of C1 on C2. If the rotation of C1 on C2 is within the normal range, as it usually is early on in this condition, the observer may attribute this rotation to patient positioning. A dynamic-rotation CT scan is helpful here. Views with the head maximally rotated to the right, and then to the left, will demonstrate atlantoaxial rotary fixation when there is a loss of normal rotation (Fig. 21-19). There are varying degrees of "locking" between the C1 and C2 vertebrae (113–115).

Rotary displacement can be classified into four types (Fig. 21-20) (108): type I is a simple rotary displacement without an anterior shift, type II is rotary displacement

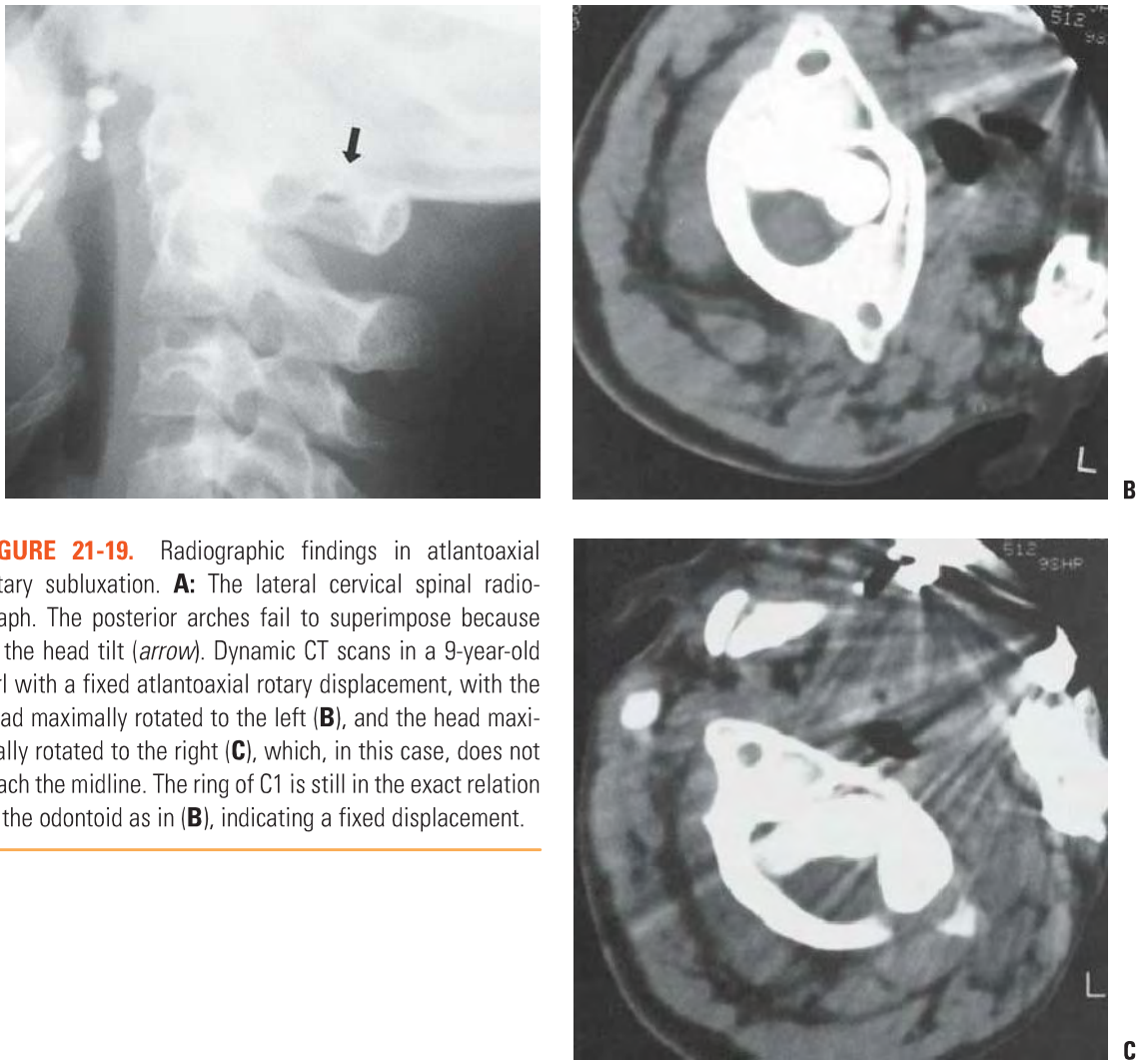


FIGURE 21-19. Radiographic findings in atlantoaxial rotary subluxation. **A:** The lateral cervical spinal radiograph. The posterior arches fail to superimpose because of the head tilt (*arrow*). Dynamic CT scans in a 9-year-old girl with a fixed atlantoaxial rotary displacement, with the head maximally rotated to the left (**B**), and the head maximally rotated to the right (**C**), which, in this case, does not reach the midline. The ring of C1 is still in the exact relation to the odontoid as in (**B**), indicating a fixed displacement.

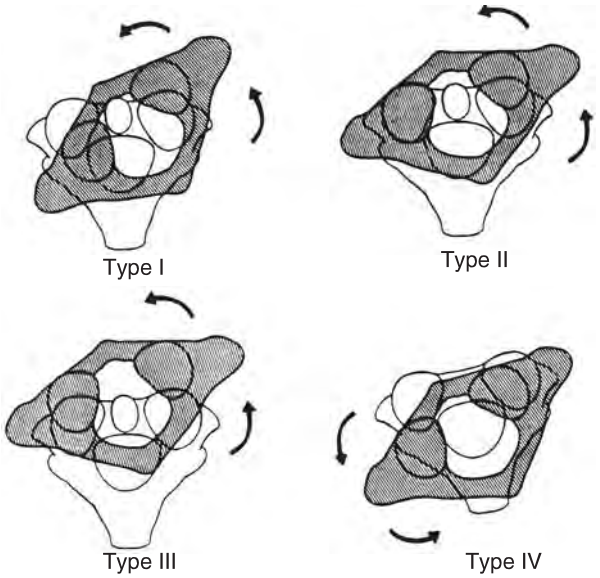


FIGURE 21-20. The four types of atlantoaxial rotary displacement. (From Fielding JW, Hawkins RJ. Atlanto-axial rotatory fixation. *J Bone Joint Surg Am* 1977;59-A:37–44, with permission.)

with an anterior shift of 5 mm or less, type III is rotary displacement with an anterior shift >5 mm, and type IV is rotary displacement with a posterior shift. The amount of anterior displacement considered to be pathologic is >3 mm in older children and adults and >4 mm in younger children (33). Flexion and extension lateral-stress radiographs are suggested to rule out the possibility of anterior displacement.

Type I is the most common pediatric type. It is usually benign and frequently resolves by itself. Type II deformity is potentially more dangerous. Types III and IV are very rare, but because of the potential for neurologic involvement and even instant death, management must be approached with great caution.

The etiology and pathoanatomy is not known completely (116). Several causative mechanisms are possible. Cervical spine fracture is a rare etiology. More commonly, atlantoaxial rotary displacement occurs following minor trauma [e.g., clavicle fractures (117)], after head and neck surgery including simple central line insertion (118), or after an upper respiratory infection. The children present with a "cocked-robin" torticollis and resist any attempt to move the head because of pain. The associated muscle spasm is noted on

the side of the long sternocleidomastoid muscle because the muscle is attempting to correct the deformity, unlike congenital muscular torticollis where the muscle causes the torticollis. If the deformity becomes fixed, the pain subsides but the torticollis persists, along with decreased neck motion. In long-standing cases, plagiocephaly and facial flattening may develop on the side of the tilt.

Spontaneous atlantoaxial subluxation with inflammation of adjacent neck tissues, also known as Grisel syndrome, is commonly seen in children after upper respiratory infections (Fig. 21-21). The children are frequently febrile (119). A direct connection exists between the pharyngovertebral veins and the periodontal venous plexus and suboccipital epidural sinuses (120). This may provide a route for hematogenous transport of peripharyngeal septic exudates to the upper cervical spine and an anatomic explanation for the atlantoaxial hyperemia of Grisel syndrome. In long-standing cases, soft-tissue abscesses or vertebral osteomyelitis may develop (121–123). Regional lymphadenitis is known to cause spastic contracture of the cervical muscles. This muscular spasm, in the presence of abnormally loose ligaments (hypothetically caused by the hyperemia of the pharyngovertebral vein drainage), could produce locking of the overlapping lateral joint edges of the articular facets. This prevents easy repositioning, resulting in atlantoaxial rotary displacement. The hyperemia after surgery of the oral pharynx, most frequently tonsillectomy and adenoidectomy, enhances the passage of the inflammatory products into the pharyngovertebral veins. It is known that patients may develop Grisel syndrome after otolaryngologic procedures (124), especially with monopolar electrocautery (125). Kawabe and Tang (126, 127) have demonstrated meniscus-like synovial folds in the atlantooccipital and lateral atlantoaxial joints of children, but not in those of adults, and have found that the dens–facet angle of the axis is steeper in children than in adults. They postulate that excessive C1-C2 rotation, caused by the steeper angle, compounded by ligament laxity from an underlying hyperemia, allows the meniscus-like synovial folds to become impinged in the lateral atlantoaxial joint, leading to rotary fixation. The predominance of this syndrome in childhood correlates with the predilection for the adenoids to be maximally hypertrophied and inflamed at this same time, and located in the area drained by the pharyngo-vertebral veins.

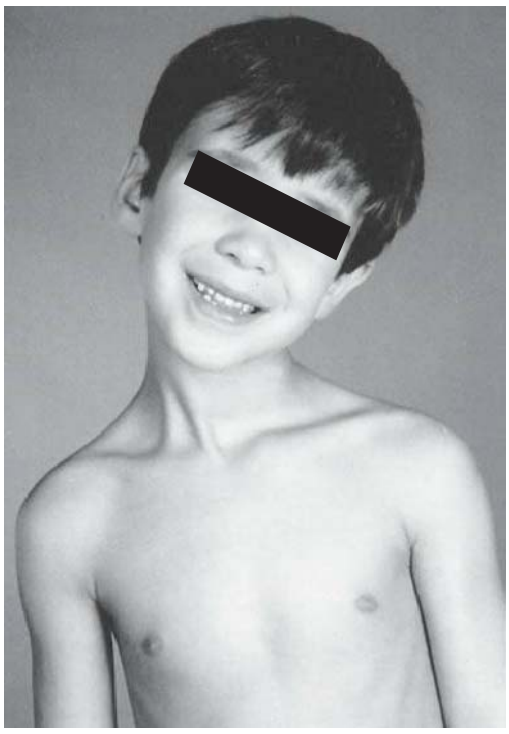


FIGURE 21-21. A 5-year-old boy developed an atlantoaxial rotary subluxation after an upper respiratory viral infection (Grisel syndrome). It rapidly resolved after treatment with a soft collar and mild doses of diazepam.

Most atlantoaxial rotary displacements resolve spontaneously. Rarely, however, the pain subsides and the torticollis becomes fixed. The duration of symptoms and deformity dictates the recommended treatment (128).

Patients with rotary subluxation of <1 week can be treated with immobilization in a soft cervical collar and rest for about 1 week. Close follow-up is mandatory. If spontaneous reduction does not occur with this initial treatment, hospitalization and the use of halter traction, muscle relaxants (e.g., diazepam), and analgesics is recommended next. Patients with rotary subluxation of >1 week but <1 month should be hospitalized immediately for cervical traction, relaxants, and analgesics. Gentle halo traction is occasionally needed to achieve reduction. The reduction is noted clinically and confirmed with a dynamic CT scan. If no anterior displacement is noted after reduction, cervical support should be continued only as long as symptoms persist. If there is anterior displacement, immobilization should be continued for 6 weeks to allow ligamentous healing to occur. In patients with rotary subluxation for more than 1 month, cervical traction (usually halo skeletal) can be tried for up to 3 weeks, but the prognosis is guarded. These children usually fall into two groups: those whose rotary subluxation can be reduced with halo traction but, despite a prolonged period of immobilization, resubluxate when the immobilization is stopped and those whose subluxation cannot be reduced, and is fixed. It has been recently shown that those patients with recurrence of deformity have a larger difference in the lateral mass–dens interval on the initial AP radiograph (129).

When the deformity is fixed, especially when anterior C1 displacement is present, the transverse atlantal ligament is compromised with a potential for catastrophe. In this situation, posterior C1-C2 fusion should be performed. The indications for fusion are neurologic involvement, anterior displacement, failure to achieve and maintain correction, a deformity that has been present for more than 3 months, and recurrence of deformity following an adequate trial of conservative management (at least 6 weeks of immobilization after reduction). Before surgical fusion halo traction for several days is used to obtain as much straightening of the head and neck

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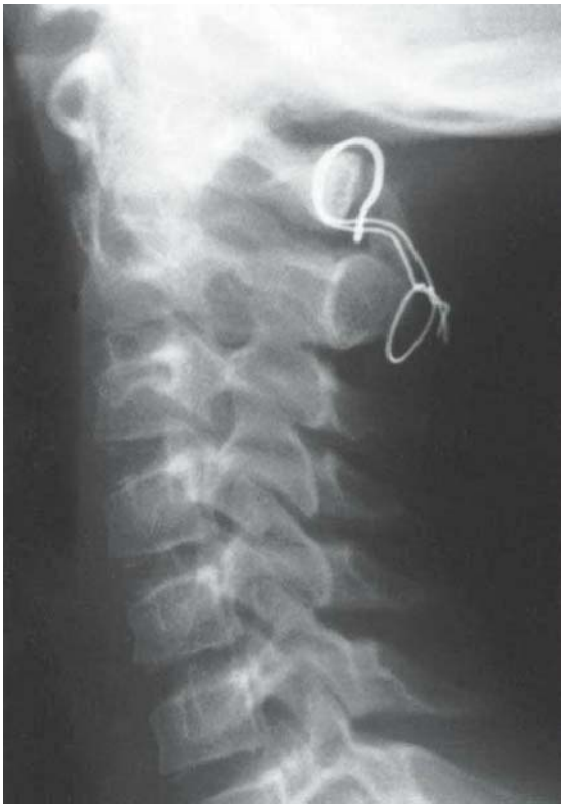
Gallie Fusion (Figs. 21-22 to 21-30)

FIGURE 21-22. The child in Figure 21-19 had a fixed deformity that occurred 6 months earlier, immediately after reconstructive maxillofacial surgery for Goldenhar syndrome. It did not respond to traction, including halo traction. She underwent a posterior C1-C2 (Gallie-type) fusion. A solid fusion was present 9 months later; clinically, the patient achieved 80 degrees of rotation to the left and 45 degrees of rotation to the right.



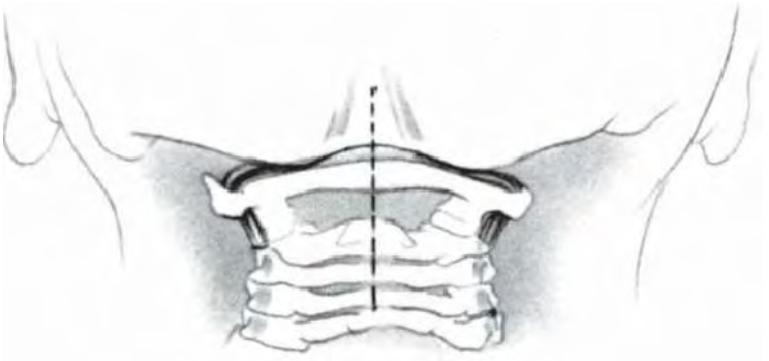


FIGURE 21-23. The most common arthrodesis of the cervical spine is between the axis and the atlas because of the numerous congenital and developmental problems that affect this region. Although several techniques have been advocated to achieve arthrodesis of these vertebrae, the technique attributed to Gallie (130) is the most reliable and the easiest to apply in children. In this technique, the wire not only helps to pull C1 back into position and hold it there but also holds the bone graft firmly in place (131, 132). Occasionally, the posterior arch of C1 is not formed completely, making this technique impossible; in these cases other techniques need to be used, such as only grafting with halo immobilization (104).

In cases in which there is a great deal of instability with chance for neurologic injury, it is preferred to place the patient in a halo vest or cast first. This can be done under local or general anesthesia, as needed. Reduction is achieved and confirmed by radiographs. If the halo was applied with the patient awake, anesthesia is then induced and the child turned prone for the posterior fusion. No head rest is necessary, and there is little danger of neurologic injury while carefully intubating and moving the patient with the halo vest in place.

The occipital region of the skull is shaved, and the posterocervical area and the posterior iliac crest are prepared and draped. The incision extends in the midline from the base of the skull to the spinous process of C4. Dissection is carried down to the tips of the spinous processes. At this point, a metal hub needle is placed in the spinous process of C2 and a lateral radiograph is taken. This is done to positively identify the correct vertebrae for exposure. In the young child, exposure of the base of the skull or any additional vertebrae may result in "creeping fusion."

FIGURE 21-24. After correct identification of the levels involved, the posterior arch of C1 and the lamina of C2 are exposed subperiosteally by a combination of sharp and blunt dissection. It is important to remember that the vertebral arteries are unprotected by the bony foramen at the C1 level just lateral to the facets. In small children this is approximately 1 cm from the midline; in bigger children it is approximately 1.5 cm from the midline. In the child with an unreduced atlantoaxial rotatory fixation, the lamina of C1 will not be parallel with the floor, but one hemilamina will be angled upward and the other downward. It is extremely important to remember this pathologic anatomy when exposing C1.

To prepare the arch of C1 for the passage of the wire beneath it, the periosteum must be separated from its anterior surface. This can be accomplished with a small, angulated, neural elevator. The spinal canal does not need to be opened. After this, a dental burr can be used to decorticate the exposed lamina of C1 and C2. This does not have to be as deep a decortication as is performed in scoliosis cases.

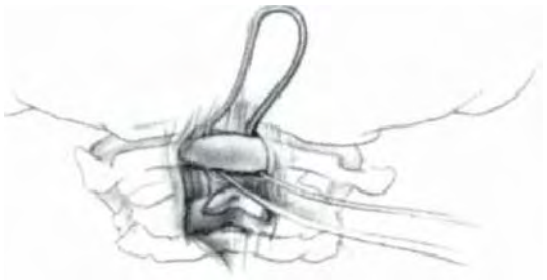


FIGURE 21-25. A double wire is passed under the arch of C1 from inferior to superior. The wire is bent back on itself, forming a smooth loop. Care should be taken not to introduce any sharp bends in the wire. The size of the wire depends on both the child's size and the surgeon's preference. Any size from 18- to 22-gauge can be used. Good-quality, fully annealed flexible wire allows a relatively larger size to be used because it pulls through easily without kinking.

FIGURE 21-26. The corticocancellous graft, which has previously been obtained and fashioned to fit over the lamina of C1 and C2, is now put in place. Small pieces of cancellous bone can be added beneath the corticocancellous graft. The loop of wire is pulled from under the arch of C1 over the graft and is placed around the spinous process of C2. A small notch cut in the base of the C2 spinous process helps to keep this in place.

FIGURE 21-27. The two ends of the wire that come out from under the arch of C1 inferiorly are pulled tight and brought around the sides and over the top of the graft. It is at this point, when the surgeon is pulling the wire tight, that the importance of a flexible wire that is not too large is realized. In working with the wire, it is best to keep it taut. This minimizes the possibility of the wire impinging on the spinal cord and makes tightening easier. After the wire is pulled tight, it can be secured with a wire twister.

FIGURE 21-28. In children, the spinous process of C2 is often small and does not provide much strength for fixation of the wire. The spinous process K-wire technique is an alternative technique (133). A threaded K wire of appropriate size is passed through a small stab wound on the side of the neck and through the paravertebral muscles and is drilled through the spinous process of C2. It is cut so that approximately 1 cm is protruding on each side.

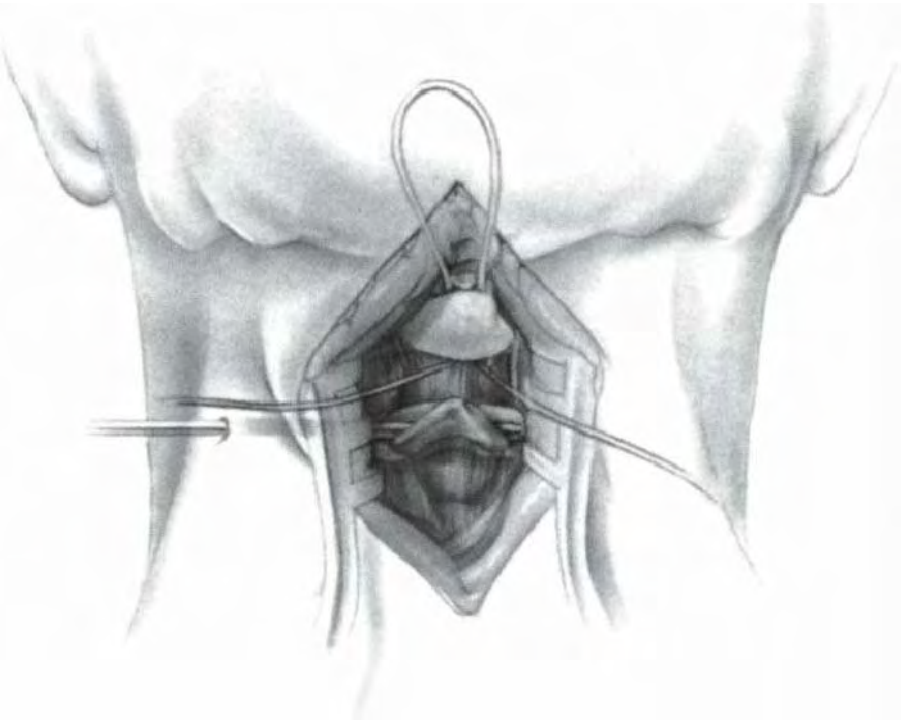
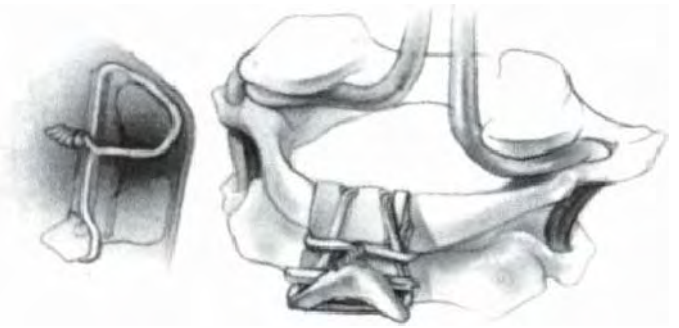


FIGURE 21-29. The corticocancellous graft is then put in place. It should fit under the K wire. The loop of wire that comes from under the arch of C1 is then drawn over the graft and looped around the spinous process of C2. The wire loop will be under the transverse Kirschner wire, however, which keeps it from slipping off the spinous process.



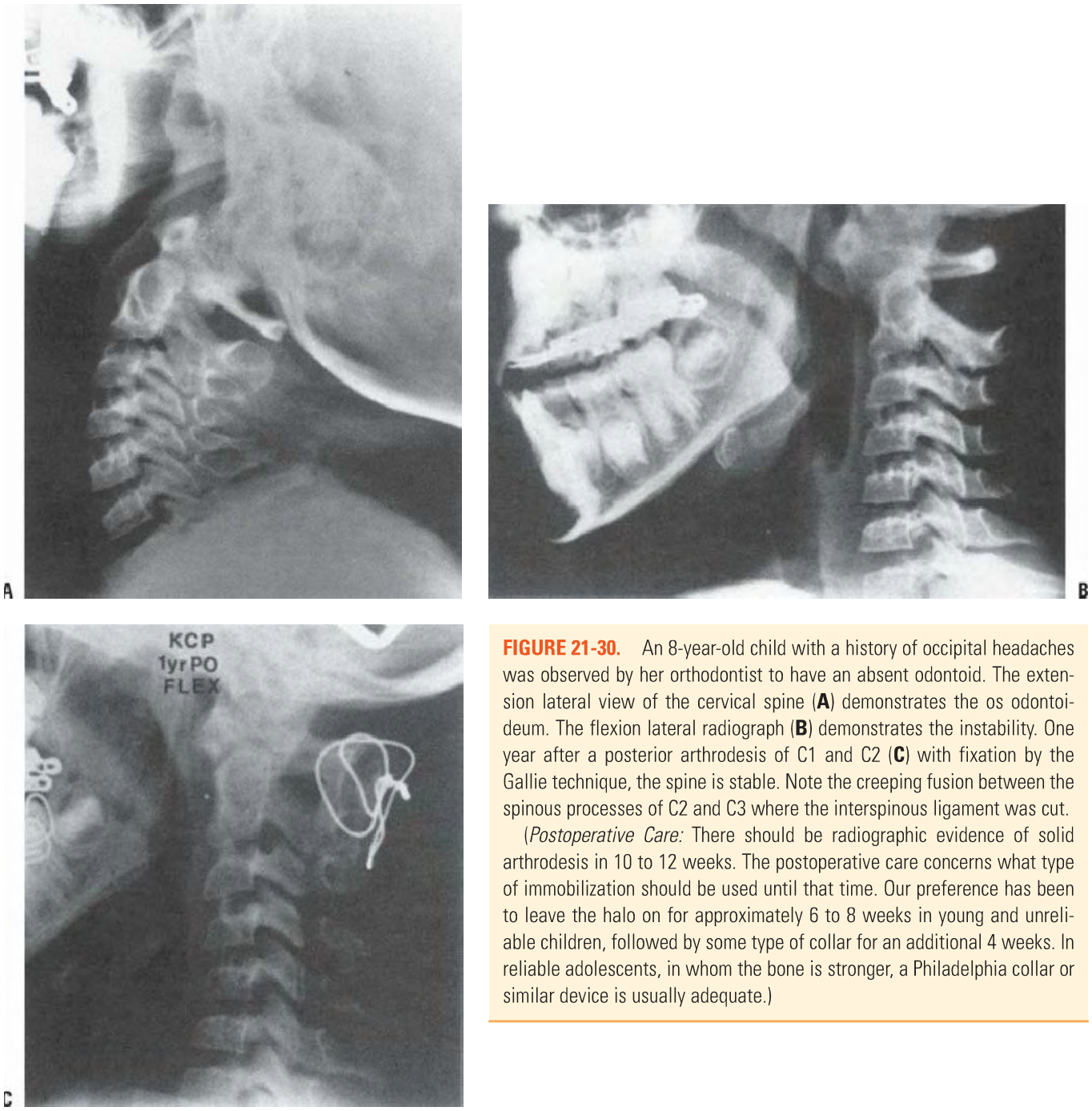


FIGURE 21-30. An 8-year-old child with a history of occipital headaches was observed by her orthodontist to have an absent odontoid. The extension lateral view of the cervical spine (**A**) demonstrates the os odontoideum. The flexion lateral radiograph (**B**) demonstrates the instability. One year after a posterior arthrodesis of C1 and C2 (**C**) with fixation by the Gallie technique, the spine is stable. Note the creeping fusion between the spinous processes of C2 and C3 where the interspinous ligament was cut.

(*Postoperative Care:* There should be radiographic evidence of solid arthrodesis in 10 to 12 weeks. The postoperative care concerns what type of immobilization should be used until that time. Our preference has been to leave the halo on for approximately 6 to 8 weeks in young and unreliable children, followed by some type of collar for an additional 4 weeks. In reliable adolescents, in whom the bone is stronger, a Philadelphia collar or similar device is usually adequate.)

as possible, a forceful or manipulative reduction should not be performed. Postoperatively, the child is simply positioned in a halo cast or vest in the straightened position obtained preoperatively; this usually obtains satisfactory alignment. A Gallie-type fusion with sublaminar wiring at the ring of C1 and through the spinous process of C2 is preferred to a Brooks-type fusion in which the wire is sublaminar at both C1 and C2. This is because of the decreased SAC at C2 with a higher risk of neurologic injury. This wiring does not reduce the displacement but simply provides some internal stability for the arthrodesis. The overall results for a Gallie fusion are very good (see Figs. 21-22 to 21-30) (131). Long-term results do not indicate any significant abnormalities of the sagittal profile (134). In the nonreduced rotatory fixation, transarticular C1-C2 screw fixation is contraindicated due to the pathologic anatomy from the rotational deformity (Figs. 21-31 to 21-37). A few surgeons advocate reduction of the deformity (135, 136); if a fusion is later needed and the deformity reduced, then transarticular C1-C2 screw fixation can be used if appropriate size screws are available.

Author's Preferred Treatment. Patients with rotary subluxation of <1 week are treated with immobilization in a soft cervical collar and rest for about 1 week. If spontaneous reduction does

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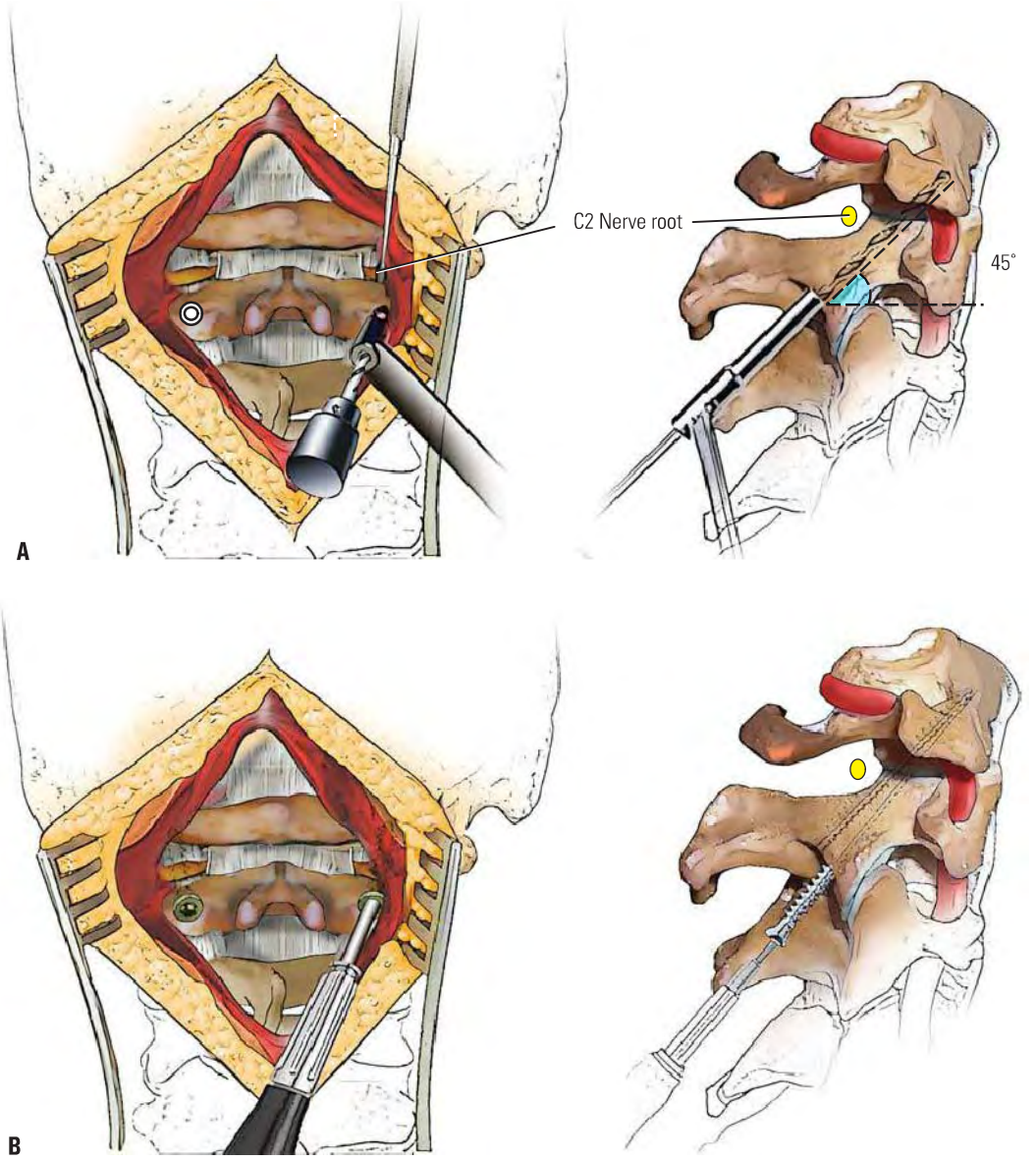


FIGURE 21-31. An alternative technique to the standard Gallie fusion uses either transarticular C1-C2 screws or lateral mass screws for both C1 and C2 with plate or rod connection. This technique can be used if there is an anatomic reduction of C1-C2 facets, but is not possible if there is atlantoaxial rotatory subluxation/fixation or anterior subluxation of the ring of C1 relative to the odontoid. The technique is the same through the exposure of the lamina of C1 and C2. The same precaution regarding the vertebral artery is necessary.

A: After exposure of the C1-C2 facet, transarticular screws may be placed. The C2-C3 facet capsule and upper portion of the C3 lamina needs to be clearly seen, and then the superior and medial aspect of the C2 lateral mass/pedicle is gently exposed with a Penfield dissector. The C2 nerve root exits laterally from the spinal canal overlying the C1-C2 joint ; it is undermined and mobilized with the Penfield dissector, and then the roof of the C2 pedicle is followed into the C1-C2 facet joint. The Penfield dissector is used to palpate the medial wall of the C2 pedicle. Under fluoroscopic guidance, the trajectory for the screw is confirmed. The typical trajectory is 15 degrees medial (to avoid the vertebral artery as the screw enters the C1 lateral mass) and 45 degrees cranial, crossing the C1-C2 facet and ending at the anterior arch of the atlas. The exact trajectory is confirmed with intraoperative fluoroscopy.

The C2 entry site is identified by first locating the inferomedial edge of the C2-C3 facet joint, without violating the joint or capsule. In an adult the entry site is ~3 mm superior and lateral to the inferomedial edge of the C2-C3 facet joint. This must be appropriately downsized in children. It is here that preoperative CT scans with sagittal and coronal reconstructions are necessary to determine appropriate trajectory, and screw diameter and length.

B: A high speed drill is used to pierce the outer cortex of the C2 lateral mass at this entry point, and then a guide wire from a cannulated screw set (if using cannulated screws) or an appropriate size drill (if using noncannulated screws) is directed up the C2 pedicle, across the C1-C2 facet joint, and into the lateral mass of C1, aiming for the anterior tubercle of C1.

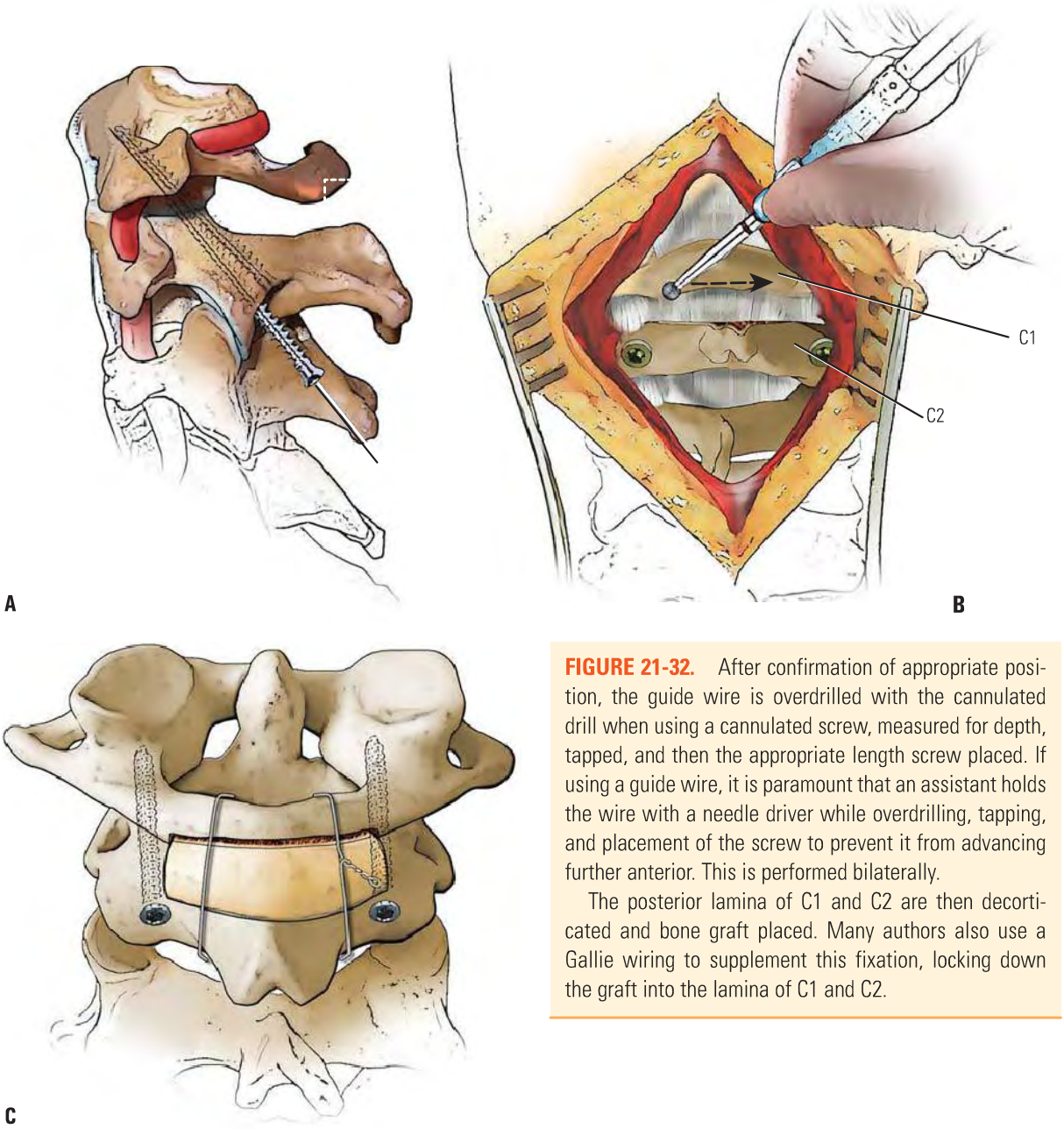


FIGURE 21-32. After confirmation of appropriate position, the guide wire is overdrilled with the cannulated drill when using a cannulated screw, measured for depth, tapped, and then the appropriate length screw placed. If using a guide wire, it is paramount that an assistant holds the wire with a needle driver while overdrilling, tapping, and placement of the screw to prevent it from advancing further anterior. This is performed bilaterally.

The posterior lamina of C1 and C2 are then decorticated and bone graft placed. Many authors also use a Gallie wiring to supplement this fixation, locking down the graft into the lamina of C1 and C2.

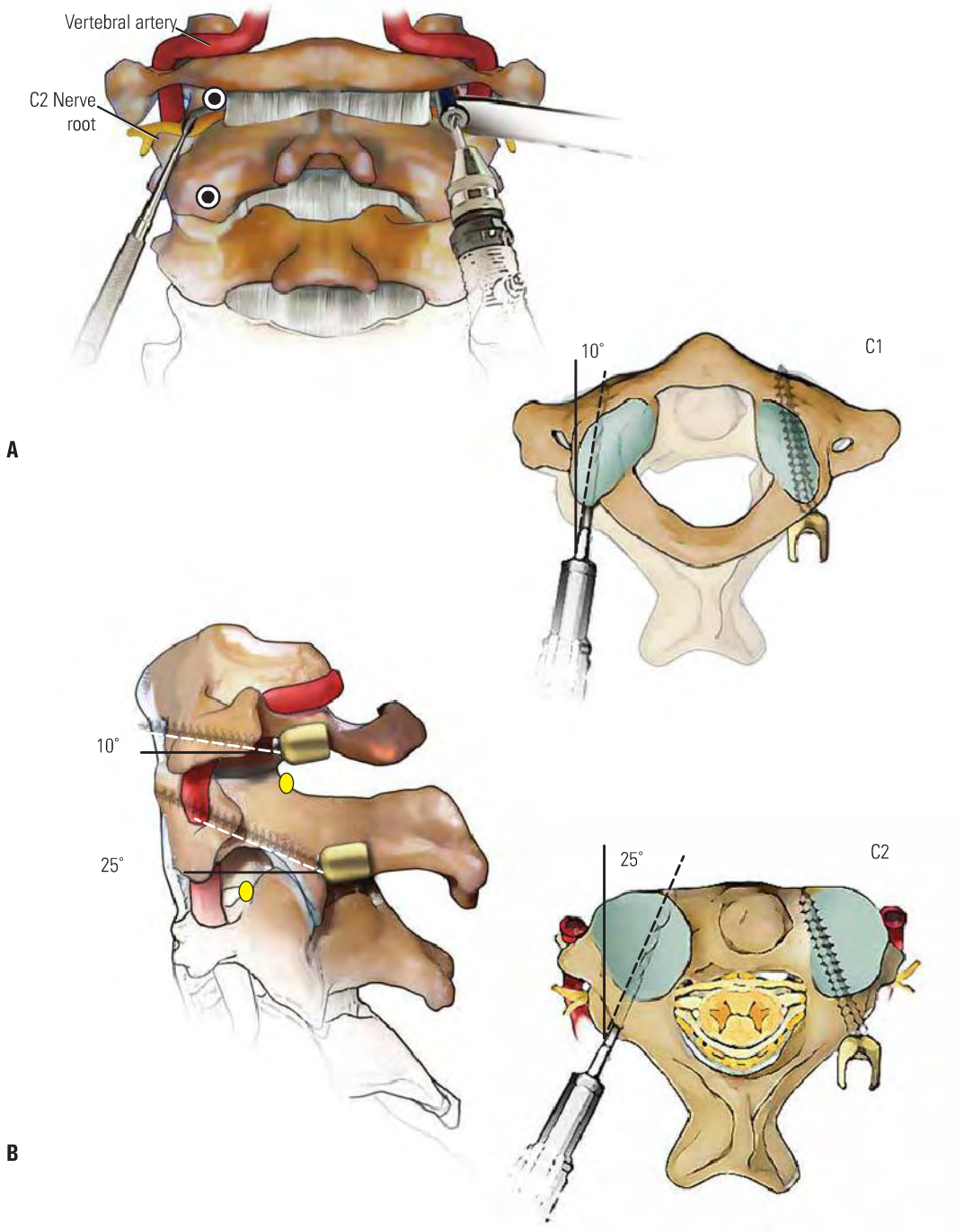
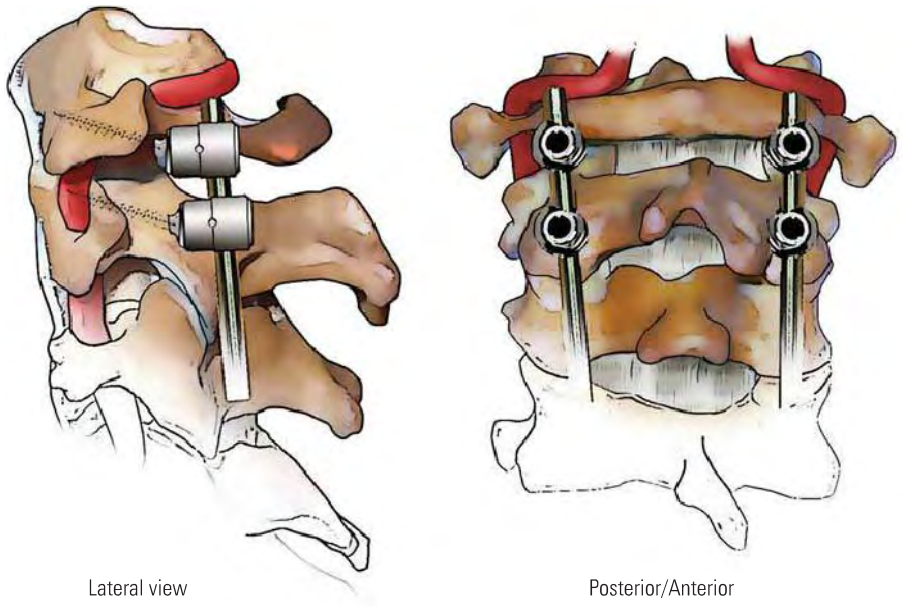


FIGURE 21-33. An alternative is to using lateral mass screws in both C1 and C2 linked with a rod. This entails the use of polyaxial screws. **A**: The entry point for the C1 lateral mass screw is at the midline of the lateral mass immediately inferior to the posterior arch of C1. Here again, the C2 nerve root exits laterally from the spinal canal overlying the C1-C2 joint and must be protected during screw placement. The high-speed drill is used to create the entry point, followed by placement of the screw after drilling. The trajectory for the drill is 0 to 10 degrees medial with intraoperative lateral fluoroscopy guiding the orientation of the drill into the lateral mass of C1 and engaging the anterior cortex. **B**: The hole is then tapped, measured, and a polyaxial screw placed, keeping the screw head above the C1 posterior arch. The C2 screw starting point is the same as for transarticular C1-C2 screw fixation, but the trajectory is more caudal and medial to avoid the transverse foramen.



Lateral view Posterior/Anterior

FIGURE 21-34. After all the screws have been placed, contoured longitudinal rods are placed into the screw heads and tightened in position, followed by a crosslink construct. Arthrodesis is the same, decorticating the C1 and C2 lamina and placement of bone graft, typically before the crosslink is placed.

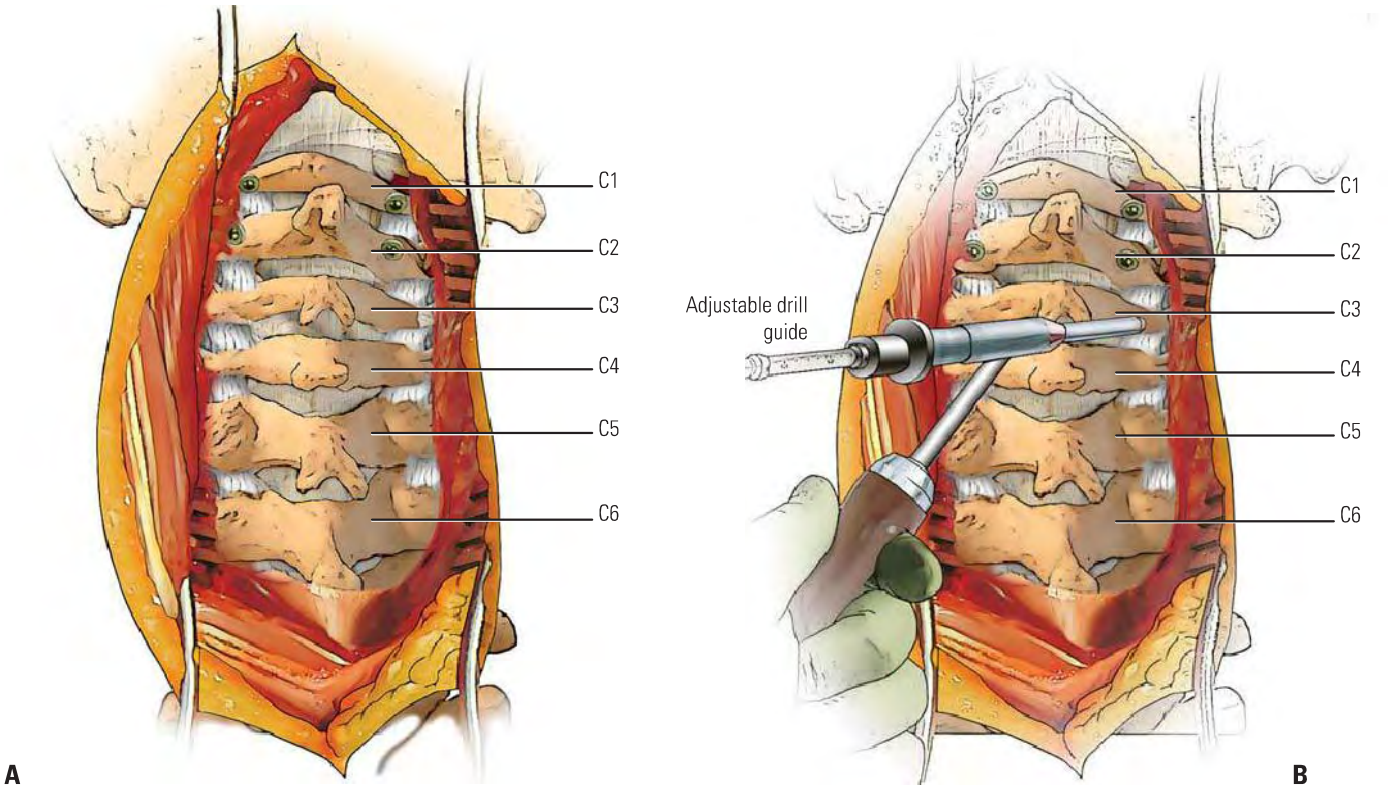
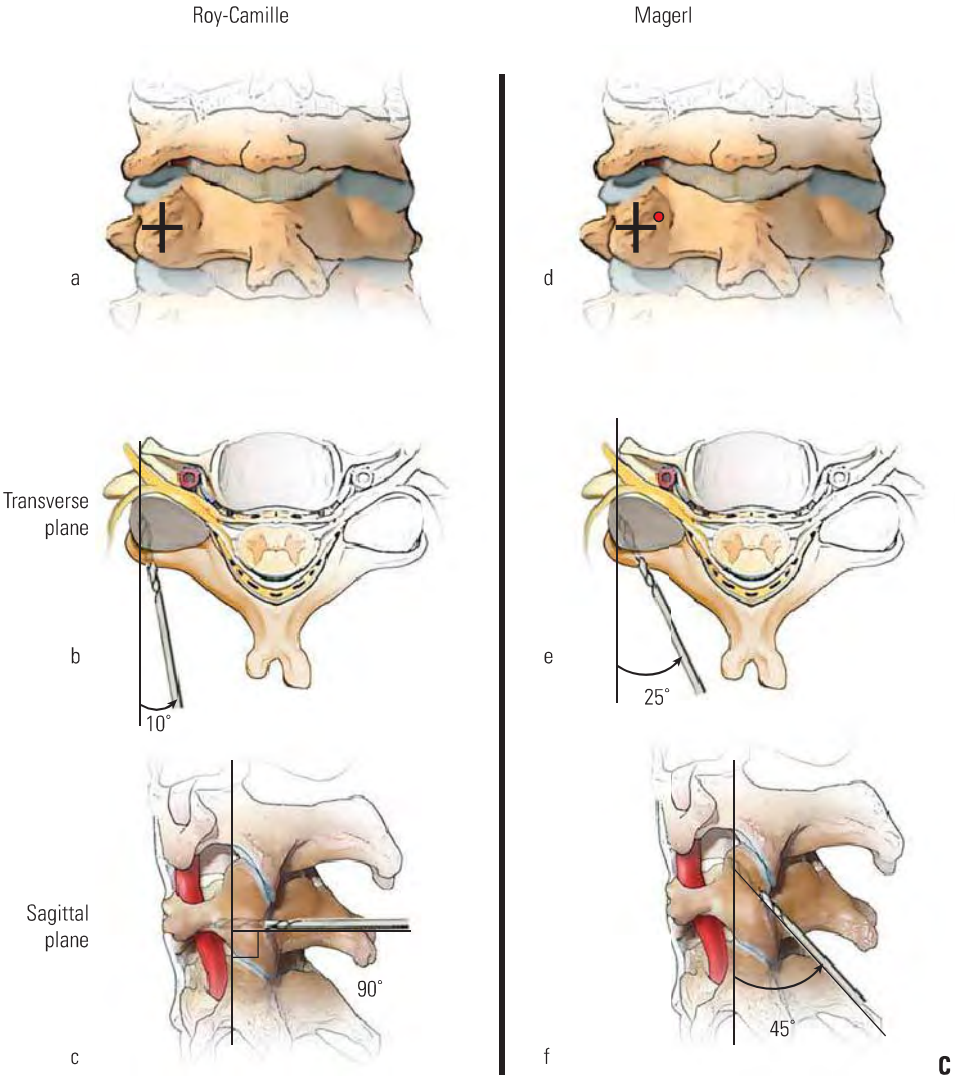


FIGURE 21-35. **A**: This technique can be extended into the subaxial spine, using polyaxial lateral mass screws or laminar hooks. **B,C**: There are two commonly used methods of lateral mass screw placement (Roy-Camille and Magerl). The Roy-Camille technique uses a starting point at the center of the lateral mass, with a trajectory directly perpendicular and 10 degrees lateral. The Magerl technique uses a starting point slightly medial and superior to the center of the lateral mass; in the sagittal plane the screw is oriented parallel to the surface of the superior articular facet (typically 15 to 30 degrees cranial) and 25 to 35 degrees lateral, with the screw tip engaging the superior and lateral portion of the ventral cortex of the superior facet. Fluoroscopic guidance is used to determine the exact trajectory in the sagittal plane.



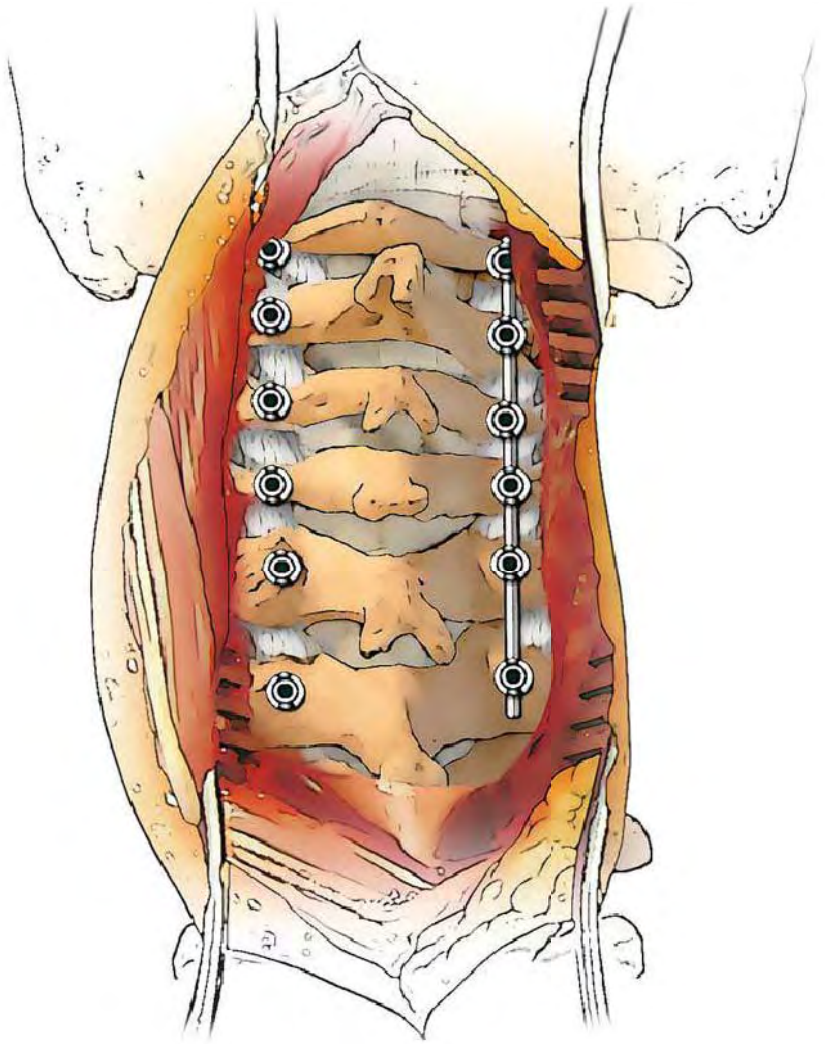


FIGURE 21-36. After bilateral placement of the screws, the rod is countered, placed into the polyaxial screws, and tightened in position. Standard decortication of the posterior elements and bone graft placement is performed for arthrodesis, followed by crosslink placement.

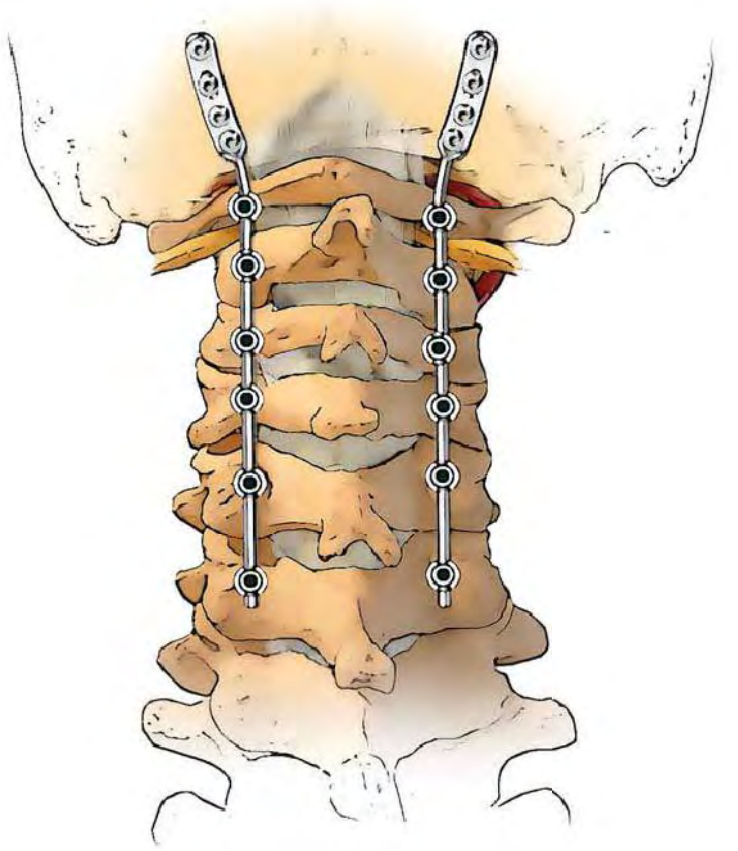


FIGURE 21-37. This technique can similarly be extended to the occiput. Many different manufacturers have occipital plates that change into rods for cervical spine fixation. When this technique is employed, it is typically best to place the C2 lateral mass screw first, followed by the subaxial lateral mass screws. Then the appropriate plate-rod construct is selected and contoured. The plate-rod construct is placed into the screw connectors. When the plate is against the occiput, drill holes in the outer cortex of the occiput are made, and appropriate-sized occipital screws are placed. A drill depth guide is strongly recommended to only drill the outer cortex. Again preoperative CT measurement of the occipital depth is useful. Standard arthrodesis is performed and then crosslinks placed.

not occur, halter traction, muscle relaxants (e.g., diazepam), and analgesics are prescribed. Patients with rotary subluxation of >1 week but <1 month should be hospitalized immediately for cervical traction, relaxants, and analgesics. Gentle halo traction is occasionally needed to achieve reduction. All reductions are confirmed with a dynamic CT scan. In patients with rotary subluxation for more than 1 month, cervical traction (usually halo skeletal) can be tried for up to 3 weeks, but the prognosis is guarded. If reduction cannot be achieved or maintained, then posterior C1-C2 arthrodesis is recommended (Fig. 21-23).

Nonosseous Types

Congenital Muscular Torticollis. Congenital muscular torticollis, or congenital wry neck, is the most common cause of torticollis in the infant and young child, presenting at a median age of 2 months (137). The incidence of soft-tissues abnormalities in the neck as documented by ultrasound was approximately 4% in a series of 1021 newborn infants (138). The deformity is caused by contracture of the sternocleidomastoid muscle, with the head tilted toward the involved side and the chin rotated toward the opposite shoulder. A disproportionate number of these children have a history of a primiparous birth or a breech or difficult delivery. However, it has been reported in children with normal births and those born by cesarean section (137, 139, 140).

The exact cause is not known and there are several theories. Because of the birth history, one theory is that of a compartment syndrome occurring from soft-tissue compression of the neck at the time of delivery (141). Surgical histopathologic sections suggest venous occlusion of the sternocleidomastoid muscle (142). This occlusion may result in a compartment syndrome, as manifested by edema, degeneration of muscle fibers, and muscle fibrosis. This fibrosis is variable, ranging from small amounts to the entire muscle. It has been suggested that the clinical deformity is related to the ratio of fibrosis to remaining functional muscle. If ample muscle remains, the sternocleidomastoid will probably stretch with growth, and the child will not develop torticollis; if fibrosis predominates, there is little elastic potential, and torticollis will develop. Another theory is *in utero* crowding, since three of four children have the lesion on the right side (143) and up to 20% have developmental hip dysplasia (144). The fact that this condition can occur in children with normal birth histories or in children born by cesarean section challenges the perinatal compartment syndrome theory, and supports the *in utero* crowding theory. The fact that it can occur in families (145–147) (supporting a genetic predisposition) also questions the compartment syndrome theory. A third theory is primarily neurogenic (148), supported by histopathologic evidence of denervation and reinnervation. The primary myopathy initially may be due to trauma, ischemia, or both, and unequally involves the two heads of the sternocleidomastoid muscle. With continuing fibrosis of the sternal head, the branch of the spinal accessory nerve to the clavicular head of the muscle can be entrapped, leading to a later progressive deformity (148).

The final theory concerns mesenchymal cells remaining in the sternocleidomastoid from fetal embryogenesis. Recent histopathologic studies have demonstrated the presence of both myoblasts and fibroblasts in the sternocleidomastoid tumor in varying stages of differentiation and degeneration (149). The source of these myoblasts and fibroblasts is unknown. After birth, environmental changes stimulate these cells to differentiate, and the sternocleidomastoid tumor develops. Hemorrhagic and inflammatory reactions would be expected if the tumor was a result of perinatal birth trauma or intrauterine positioning, yet these cells were not seen in the sternocleidomastoid histopathologic studies. The occurrence of torticollis depends on the fate of the myoblasts in the mass. If the myoblasts undergo normal development and differentiation, then no persistent torticollis will occur and conservative treatment will likely succeed. If the myoblasts mainly undergo degeneration, then the remaining fibroblasts produce large amounts of collagen, with a scar-like contraction of the sternocleidomastoid muscle and the typical torticollis.

There are three clinical subgroups; those with sternocleidomastoid tumor (43% of cases), those with muscular torticollis (31% of cases), and postural torticollis (22% of cases) (150). The clinical features of congenital muscular torticollis depend on the age of the child. It is often discovered in the first 6 to 8 weeks of life. If the child is examined during the first 4 weeks of life, a mass or a "tumor" may be palpable in the neck (139). Although the mass may be palpable, it is unrecognized up to 80% of the time (151). Characteristically, it is a nontender, soft enlargement beneath the skin, and is located within the sternocleidomastoid muscle belly. This tumor reaches its maximum size within the first 4 weeks of life then gradually regresses. After 4 to 6 months of life, the contracture and the torticollis are the only clinical findings. In some children, the deformity is not noticed until after 1 year of age, which raises questions about both the congenital nature of this entity and the perinatal compartment syndrome theory. Recent studies (152) indicate that the rate of associated hip dysplasia in children with congenital muscular torticollis is 8%, lower than the previously cited 20% (144). The sternocleidomastoid tumor subgroup, the most severe group, presents at an earlier age, is associated with a higher incidence of breech presentation (19%), difficult labor (56%), and hip dysplasia (6.8%) (150).

If the deformity is progressive, skull and face deformities can develop (plagiocephaly) (153, 154), often within the first year of life. The facial flattening occurs on the side of the contracted muscle and is probably caused by the sleeping position of the child (155). In the United States, children usually sleep prone, and in this position, it is more comfortable for them to lie with the affected side down. The face therefore remodels to conform to the bed. If the child sleeps supine, reverse modeling of the contralateral skull occurs. In the child who is untreated for many years, the level of the eyes and ears becomes unequal and can result in considerable cosmetic deformity.

Ultrasound can be quite helpful in differentiating congenital muscular torticollis from other pathologies in the

neck (138, 156). Radiographs of the cervical spine should be obtained to rule out associated congenital anomalies. Plain radiographs of the cervical spine in children with muscular torticollis are always normal, aside from the head tilt and rotation. If any suspicion exists about the status of the hips, appropriate imaging (e.g., ultrasonography, radiography) should be done, depending on the age of the child and the expertise of the ultrasonographer.

Research MRI studies demonstrate abnormal signals in the sternocleidomastoid muscle, but no discrete masses within the muscle (141, 157). The muscle diameter is increased two to four times that of the contralateral muscle. In older patients the signals are consistent with atrophy and fibrosis, similar to those encountered in compartment syndromes of the leg and forearm.

As the deficit in cervical rotation increases, the incidence of a previous sternocleidomastoid tumor, hip dysplasia, and the likelihood of needing surgery increases (158, 159). Treatment initially consists of conservative measures (139, 140, 151, 160, 161). Good results can be expected with stretching exercises alone, with one series reporting 90% success (160) and another 95% (159). Those children with a sternocleidomastoid tumor respond less favorably to conservative stretching exercises than those with a simple muscle torticollis; none of the children with postural torticollis need surgery (159). The extent of sternocleidomastoid fibrosis on ultrasound examination is also predictive of the need for surgery (162, 163). In those cases in which only the lower one-third of the muscle is involved with fibrosis, all responded to conservative therapy, and in those cases where the entire length of the muscle was involved with fibrosis, surgery was needed in 35% of the children (164).

The exercises are performed by the caregivers and guided by the physiotherapist. The ear opposite the contracted muscle should be positioned to the shoulder, and the chin should be positioned to touch the shoulder on the same side as the contracted muscle. When adequate stretching has occurred in the neutral position, the exercises should be graduated up to the extended position, which achieves maximum stretching and prevents residual contractures. Treatment measures to be used along with stretching consist of room modifications by modifying the child's toys and crib so that the neck is stretched when the infant is reaching for or looking at objects of interest. The exact role of the efficacy of these stretching measures, versus a natural history of spontaneous resolution, is not known (165); there are many anecdotal cases of spontaneous resolution. Occasionally, muscle stretching itself will result in partial or complete rupture of the sternocleidomastoid muscle (166). Recently, some have used botulinum toxin as an adjunct to assist in the stretching program, although dysphagia and neck weakness are significant side effects (167–169).

If stretching measures are unsuccessful after 1 year of age (159, 161, 165, 170), surgery is recommended. The child's neck and anatomic structures are larger, making surgery easier. Established facial deformity or a limitation of more than 30 degrees of motion usually precludes a good result, and surgery is required to prevent further facial flattening and further cosmetic deterioration (161). Asymmetry of the face and skull can improve as long as adequate growth potential remains after the deforming pull of the sternocleidomastoid is removed; good but not perfect results can be obtained as late as 12 years of life (151, 171, 172).

The best time for surgical release is between the ages of 1 and 4 years; (139, 173) for those treated surgically before the age of 3 years, excellent results can be expected in nearly all cases (171). Surgical treatments include a unipolar release (171) at the sternoclavicular or mastoid pole, bipolar release, middle third transection, and even complete resection. Although these surgical procedures are usually done open, endoscopic (174) and percutaneous (distal) approaches have been recently described (175). Bipolar release combined with a Z-plasty of the sternal attachment yielded 92% satisfactory results in one series, whereas only 15% satisfactory results were obtained with other procedures (170). Similar results, although not perfect, can even be achieved in older children with a bipolar release (172, 176). In a more recent series of surgical cases, excellent results were obtained with a unipolar release and aggressive postoperative stretching (171). Middle third transection has also been reported to give 90% satisfactory results (177). Z-plasty lengthening maintains the V-contour of the neck and cosmesis, which the middle third transection does not. Structures that can be injured from surgery are the spinal accessory nerve, the anterior and external jugular veins, the carotid vessels and sheath, and the facial nerve. Skin incisions should never be located directly over the clavicle because of cosmetically unacceptable scar spreading; rather, they should be made one finger breadth proximal to the medial end of the clavicle and sternal notch, and in line with the cervical skin creases. The postoperative protocol can vary from simple stretching exercises to cast immobilization. Some type of a bracing device to maintain alignment of the head and neck is probably a desirable part of the postoperative protocol (178).

Author's Preferred Treatment. For those children diagnosed before 1 year of age, a regimen of stretching exercises and room modifications is tried first. If this approach fails, or the child presents after 1 year of age, then a bipolar sternocleidomastoid release is performed (Figs. 21-38 to 21-41). Postoperative orthotic immobilization is employed along with frequent physiotherapy for at least 3 months after surgery.

Neurogenic Types. Although rare, these causes should be considered in the differential diagnosis of any atypical torticollis, especially when the condition is unresponsive or progressive in the face of therapy believed to be appropriate. The major neurogenic etiologies are central nervous system tumors (i.e., of the posterior fossa or the spinal cord), syringomyelia with or without cord tumor, Arnold-Chiari malformation, ocular dysfunction, and paroxysmal torticollis of infancy (179).

Text continued on page 862

Bipolar Sternocleidomastoid Release (Figs. 21-38 to 21-44)

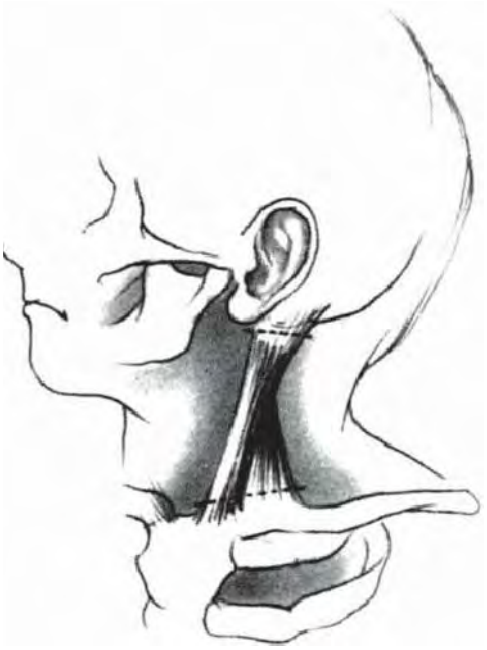
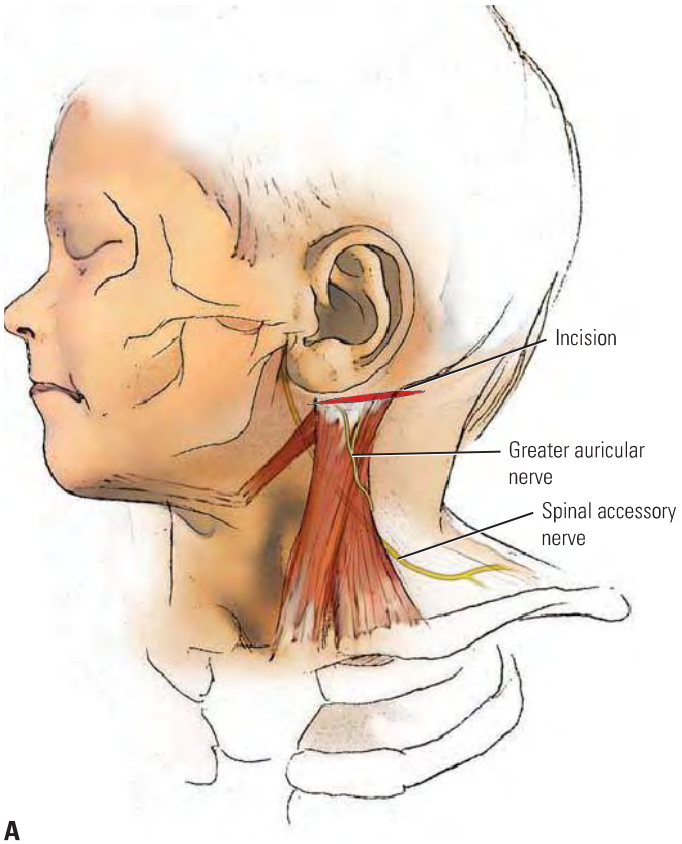
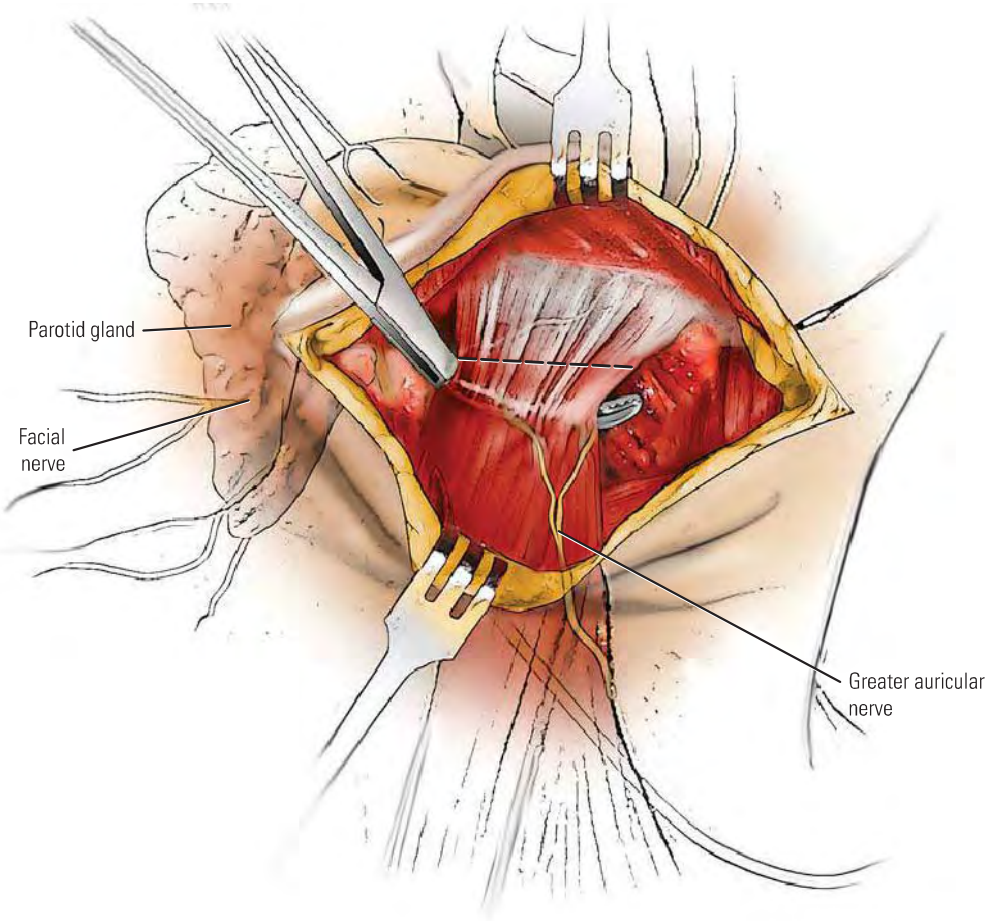


FIGURE 21-38. Bipolar release of the sternocleidomastoid for congenital muscular torticollis maintains the V contour of the neck and is cosmetically more acceptable than transection or resection of the muscle. The mastoid release is performed first.

After induction of anesthesia, the head is tilted away from the side involved, which increases the visibility of the tight muscle. The scalp and hair in the mastoid area are trimmed as needed. Folding the ear over a sponge and gently taping it anterior to the field allows for an easier exposure. Adherent plastic drapes are attached with adhesive to the skin to keep the field clear as well as prevent prepping solution from draining into the ears and eyes. A transverse incision is made just distal to the tip of the mastoid.



A



B

FIGURE 21-39. **A**: Dissection must not be too distal in order to avoid injury to the spinal accessory nerve. **B**: Similarly, dissecting too anterior places the facial nerve at risk where it exits from the parotid gland area. Once the mastoid origin is completely exposed, a careful transection is performed. Routine closure is performed.

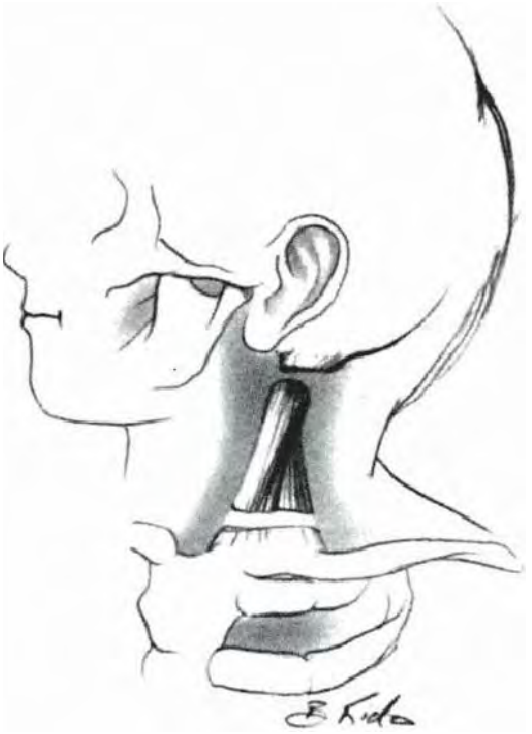
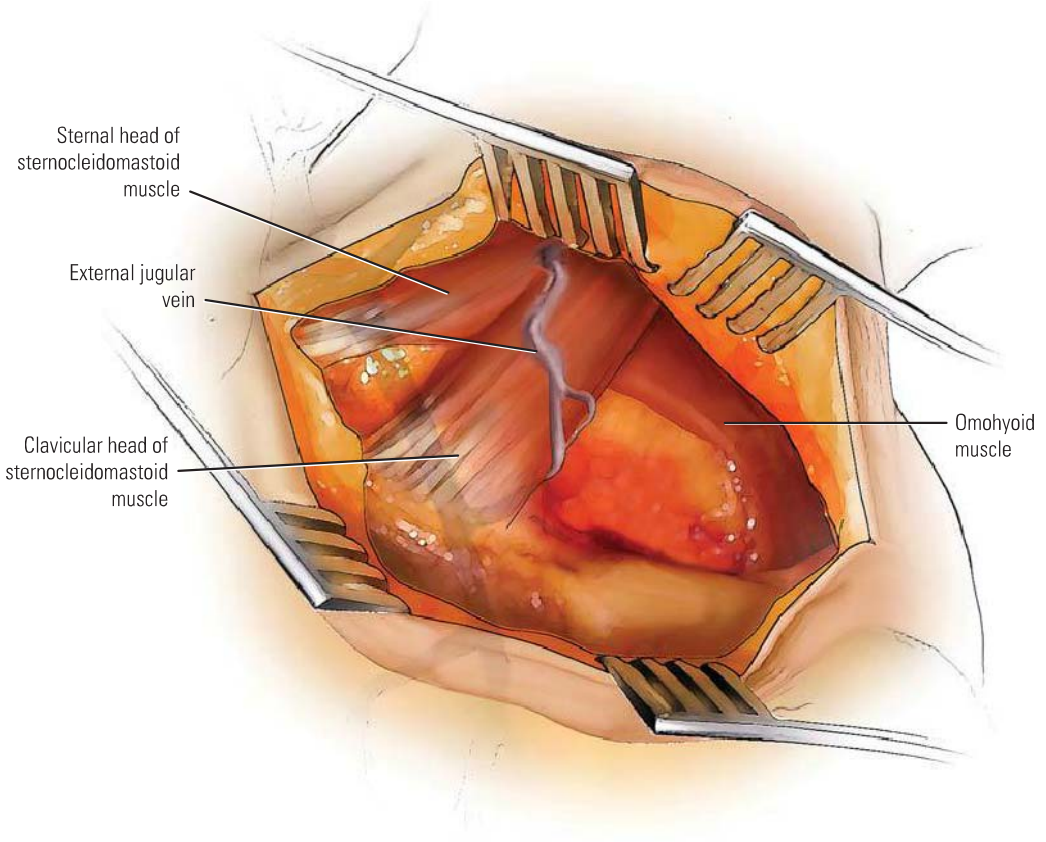
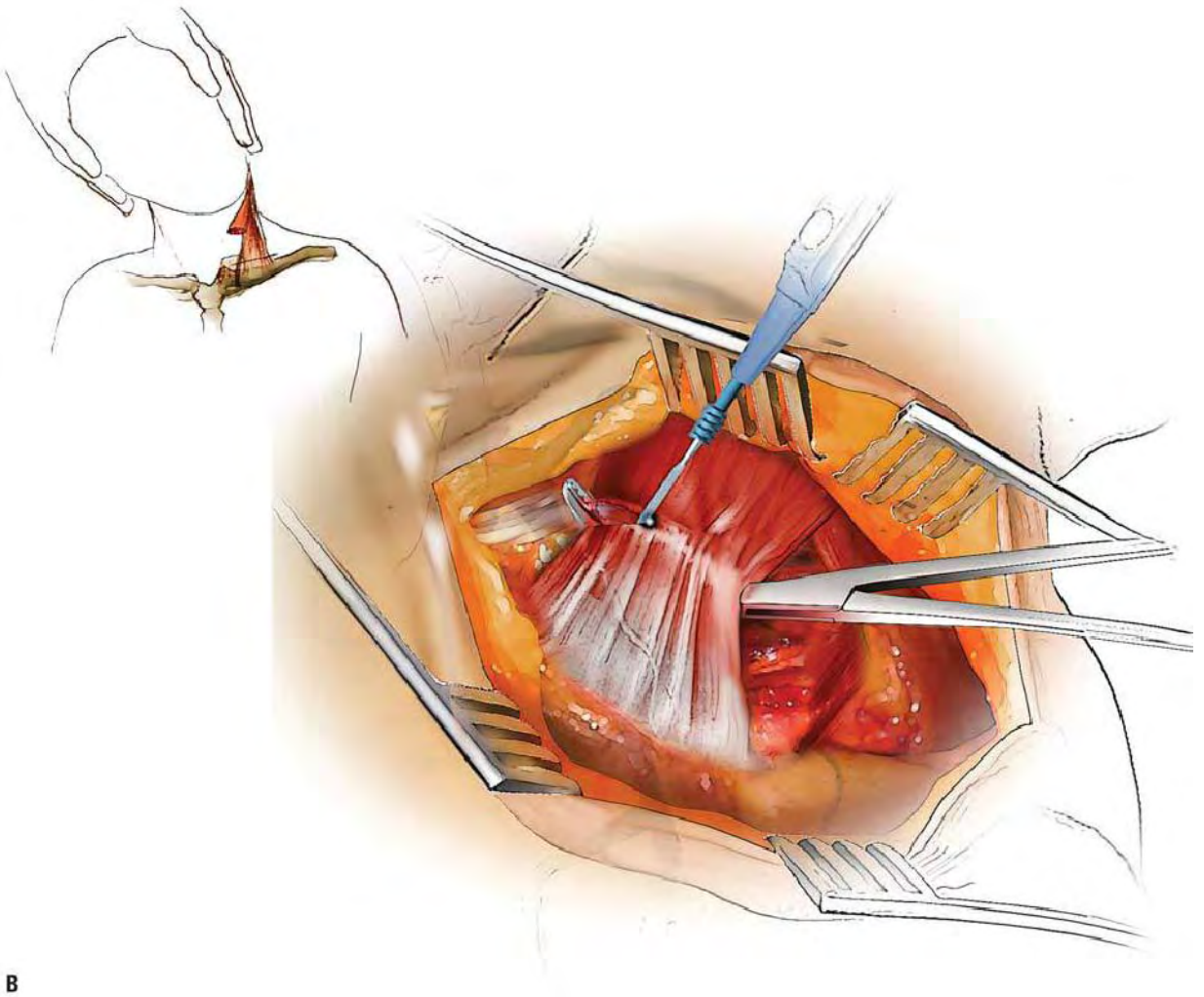


FIGURE 21-40. Attention is then directed to the distal release. The incision should be placed in a skin crease a short distance above the clavicle. Because the skin is mobile and can be moved (not stretched) from a medial to a lateral position, the incision can be small, running from the lateral border of the sternal head to the midportion of the clavicular head.


Sternal head of sternocleidomastoid muscle
External jugular vein
Clavicular head of sternocleidomastoid muscle
Omohyoid muscle

A



B

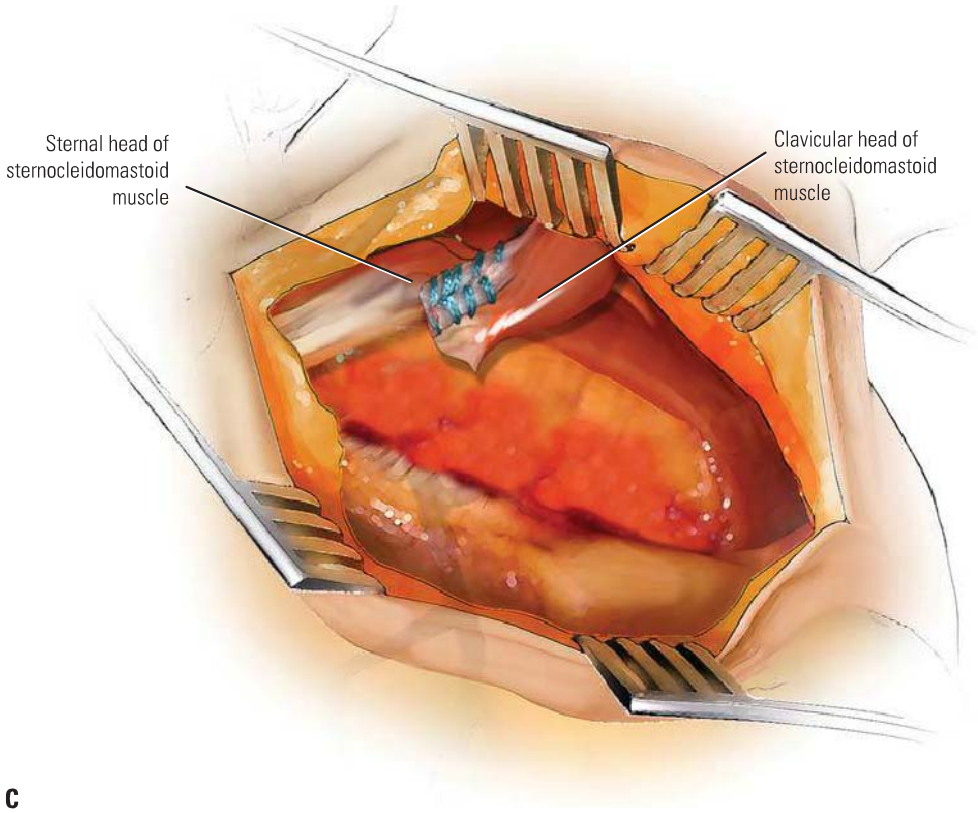
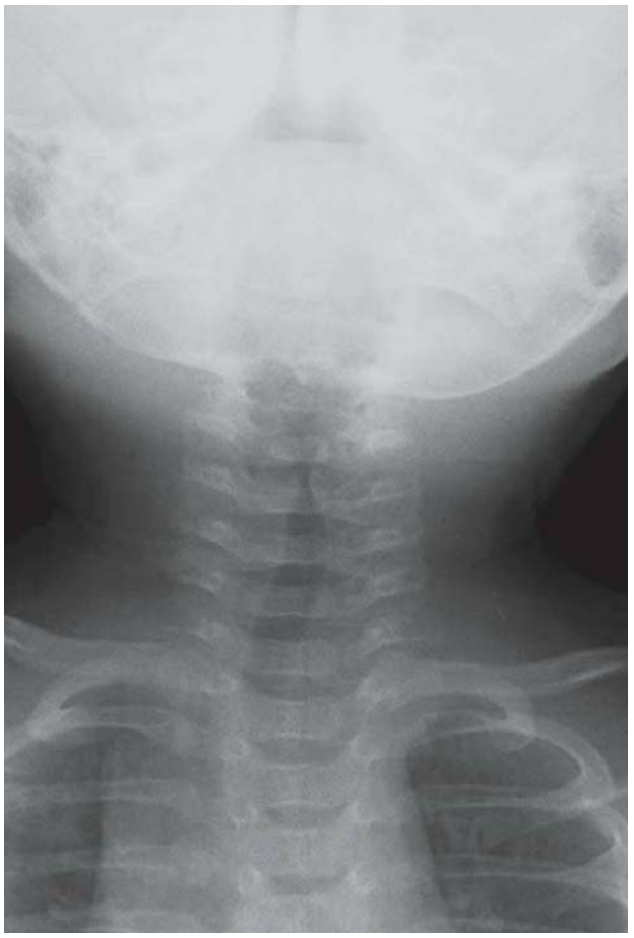
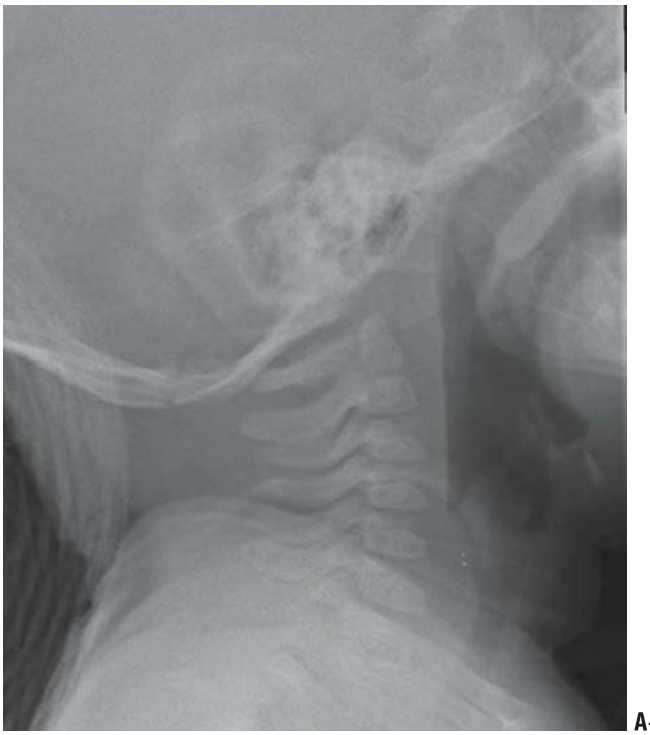


FIGURE 21-41. The platysma muscle should be identified as a separate and distinct layer so that it may be repaired at the time of closure. This helps preserve the contour of the neck and avoids an unsightly depression as a result of the released/lengthened muscle. **A**: Beneath the platysma muscle, the sternal and clavicular heads of the sternocleidomastoid muscle can usually be identified as distinct structures. In some cases, the clavicular head is thin and seems to be of little importance. Because the sternocleidomastoid muscle is adherent to the surrounding fascia, it should be dissected free for a distance of about 2 cm. In accomplishing this, the adherence of the muscle to the investing fascia is appreciated. If this dissection is not done, the muscular repairs lie in close proximity after the platysma muscle is repaired, and recurrence is likely. Although a fascial layer separates the sternocleidomastoid muscle from the deeper venous and arterial structures, the muscle is usually adherent and should be separated from this fascial layer with care. **B**: The clavicular head is first divided. After exposure, the muscle can be divided with a low cautery current close to the clavicle in its tendinous portion. If the anesthesiologist is asked to turn the head toward and away from the operative side, the tightness of this structure becomes apparent. Failure to divide the clavicular head usually produces disappointing results. **C**: Next, a z-plasty of the sternal head is performed, keeping the medial attachment of the sternal head preserved. After the z lengthening, the end-sand sutured together. The head is then moved and the operative area inspected and palpated for any remaining tight structures. Often deep fascial bands are identified; these should be divided. The wound is inspected for bleeding and irrigated; a small drain may be used if indicated. The wound is then closed in layers with particular attention to the platysma muscle.

(*Postoperative Care*: After 1 week, stretching exercises are resumed and continued for 3 months. Collars and orthotic devices are often helpful, especially in the older child in order to maintain correction during the healing phase.)

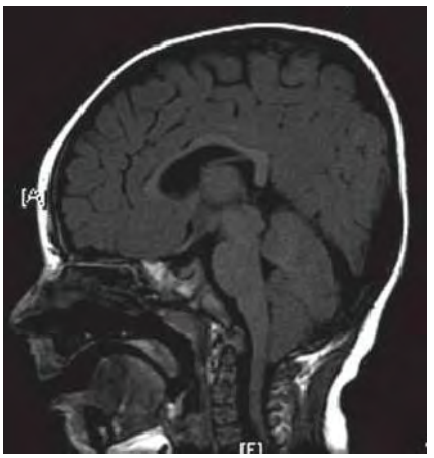


A-1



A-2

FIGURE 21-42. A 3-month-old boy presented with a 2-month history of right-sided torticollis. His prenatal and delivery history were unremarkable. Cervical rotation to both the right and the left was full, and there was no tightness of the sternocleidomastoid muscle. **A:** Cervical spine radiographs (**A-1**) and lateral (**A-2**) demonstrated only the tilt with no congenital vertebral anomalies. **B:** An MRI was obtained that demonstrated an Arnold Chiari malformation; the inferior tip of the cerebellar tonsils is at the level of the odontoid neurocentral synchondrosis.



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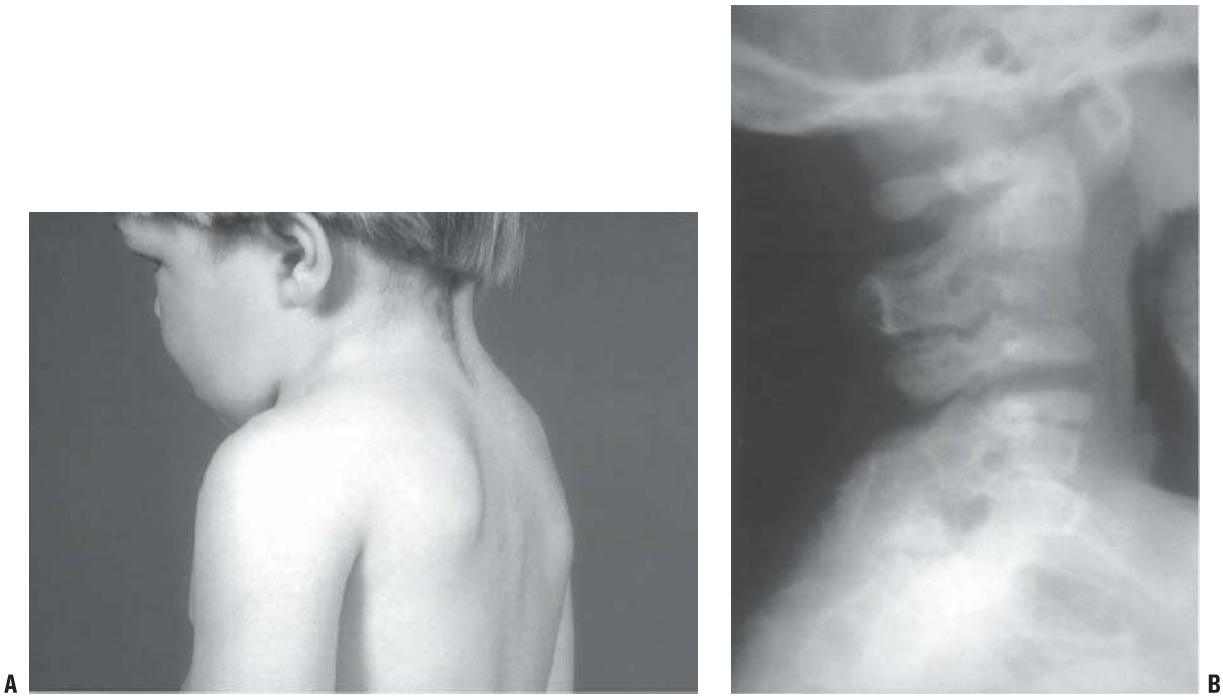
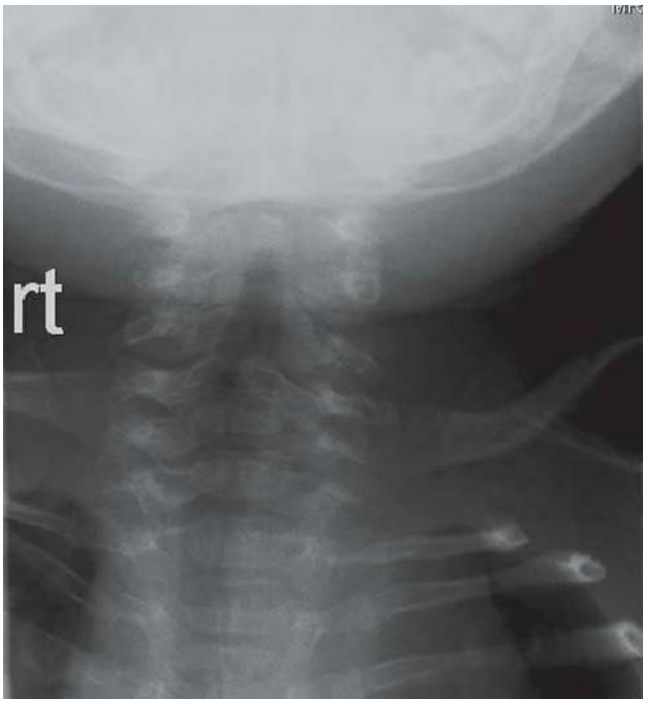
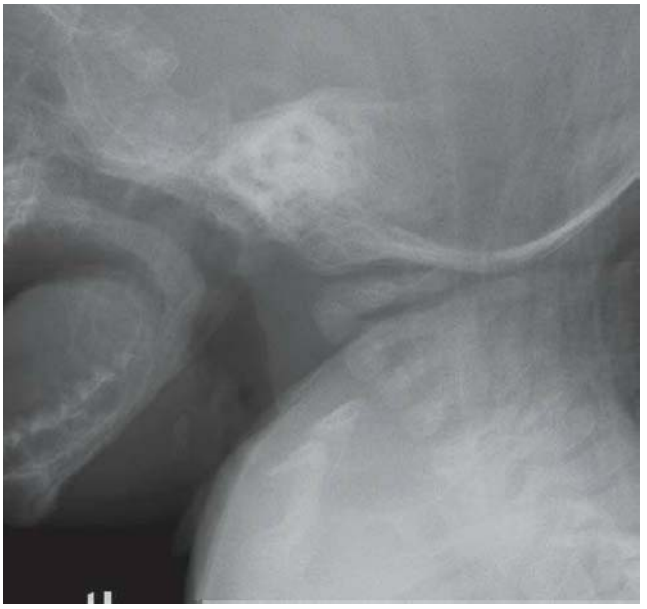


FIGURE 21-43. This 3-year, 6-month-old boy presented with a short neck and reduced motion. **A:** Note the short neck and low posterior hair line. **B:** The lateral cervical spine radiograph demonstrates complete fusion of the posterior elements of C2-C3, with reduced disc height anteriorly at C2-C3. Note the reduced space between C3 and C4, which most likely represents a cartilage fusion between C3-C4 and will likely later become an osseous fusion.



A



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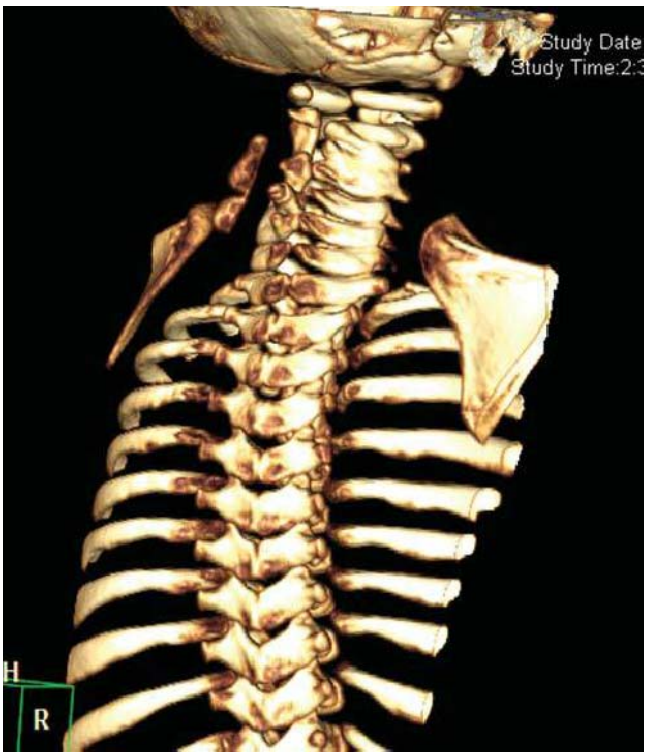


C

FIGURE 21-44. A 3-month-old girl presented for concerns of shoulder asymmetry. Physical examination demonstrated a short neck with a hypoplastic right ear. **A:** The AP radiograph demonstrates incomplete closure of the posterior arches. **B:** The lateral radiograph is concerning for fusion of the posterior elements. **C:** The three-dimensional CT reconstruction (viewed from the right posterior to left anterior) demonstrates an omovertebral bar between the spinous processes of the cervical spine and the scapula, as well as fusion of the posterior elements of C2 and C3 and incomplete fusion of the neural arch C2-C4 (spina bifida occulta).

Posterior fossa tumors can present with torticollis (180–182). The ophthalmologic literature (183) has described three children with torticollis, photophobia, and epiphora (tearing). In all three children, the diagnosis was delayed with an initial diagnosis of a local ocular inflammatory condition. The age at presentation ranged from 1 to 23 months. The delay in diagnosis ranged from 5 months to 4 years. The neoplastic diagnosis was not considered initially by the ophthalmologists because the primary signs of posterior fossa tumors are extraocular muscle paresis, nystagmus, and papilledema.

Cervical cord tumors can present with torticollis, often early in their course (184–186). Frequently the initial diagnosis is congenital torticollis, obstetric birth palsy, muscular dystrophy, or cerebral palsy (184). The peculiar, often-overlooked signs of the tumor are spinal rigidity, early spinal deformity, and spontaneous or induced vertebral pain. In young children, pain may be expressed as irritability and restlessness (187).

Imaging of a child with a potential central nervous system tumor should consist of plain radiographs of the skull and the cervical spine followed by CT and MRI scans. Vertebral angiography also may be needed, both diagnostically and in neurosurgical planning.

The Arnold-Chiari malformation (Fig. 21-42) is caudal displacement of the hindbrain, often with other congenital deformities of the brain stem and the cerebellum (188, 189). It may be associated with myelomeningocele (i.e., type II malformation). The Chiari type I malformation is a downward displacement of the medulla oblongata with extrusion of the cerebellar tonsils through the foramen magnum and is encountered in older children. Dure et al. (188) described 11 children with Chiari type I malformations; torticollis was the presenting complaint at 5 years of age in 1 of the 11 children. It also was associated with headaches and paracervical muscle spasm; the torticollis was left sided. As with tumors, the workup of a child with the potential for a Chiari malformation consists of plain radiographs of the skull and the cervical spine followed by an MRI scan (188). The treatment is neurosurgical.

Ocular pathology accounts for up to one-third of children with no obvious orthopaedic cause of torticollis (190). The torticollis is usually atypical (191). These children typically present around 1 year of age. The face can be turned about a vertical axis, the head can be tilted to one shoulder with the frontal plane of the face remaining coronal, the chin can be elevated or depressed, or a combination of any of these positions can occur. These abnormal head positions optimize visual acuity and maintain binocularity. An ocular cause is likely if the head is tilted but not rotated or if the tilt changes when the child is lying versus sitting or standing up. Children with ocular torticollis have a full range of cervical motion without the fibrotic sternocleidomastoid muscle seen in congenital muscular torticollis. Ophthalmologic evaluation is usually positive for paralytic squint or nystagmus. Detailed tests conducted by an experienced ophthalmologist are diagnostic. Treatment of ocular torticollis is usually surgical.

Paroxysmal torticollis of infancy is a rare, unusual episodic torticollis lasting for minutes to days with spontaneous recovery (192–194). The attacks usually occur in the morning, last from minutes to days, with a frequency from less than one episode per month to three to four episodes per month. The attacks can be associated with lateral trunk curvature, eye movements or deviations, and alternating sides of torticollis. The children are usually girls (71%), the average age of onset is 3 months (1 week to 30 months), and the recovery period is 24 months (6 months to 5 years). It has been suggested that paroxysmal torticollis of infancy is equivalent to a migraine headache (195, 196) because of positive family histories for migraines in 29%, or it is a forerunner of benign paroxysmal vertigo of childhood (193). Whatever the cause, it is usually self-limiting and does not require therapy. It may be linked to a mutation in the CACNA1A gene (196), the gene associated with familial hemiplegic migraine.

Sandifer Syndrome. This is a syndrome of gastroesophageal reflux, often from a hiatal hernia, and abnormal posturing of the neck and trunk, usually torticollis (197, 198). The torticollis is likely an attempt of the child to decrease esophageal discomfort resulting from the reflux. The abnormal posturing also may present as opisthotonos or neural tics and often mimics central nervous system disorders. The majority of patients present in infancy. The incidence of gastroesophageal reflux is high (up to 40% of infants) (199), with the principal symptoms being vomiting, failure to thrive, recurrent respiratory disease, dysphagia, various neural signs, torticollis, and respiratory arrest. The diagnosis of symptom-causing gastroesophageal reflux frequently is overlooked. On careful examination of these infants, the tight and short sternocleidomastoid muscle or its tumor is not seen, eliminating congenital muscular torticollis. Further workup excludes dysplasias and congenital anomalies of the cervical spine, and central nervous system disorders. In these situations, the physician should consider Sandifer syndrome in the differential diagnosis.

Plain radiographs of the cervical spine eliminate congenital anomalies or dysplasias; contrast studies of the upper gastrointestinal tract usually demonstrate the hiatal hernia and gastroesophageal reflux (200). Esophageal pH studies may be necessary; many children, both asymptomatic and symptomatic, show evidence of gastroesophageal reflux (201). Treatment begins with medical therapy. When this fails fundoplication can be considered, which is usually curative (202).

Klippel-Feil Syndrome. Klippel-Feil syndrome consists of congenital fusions of the cervical vertebrae clinically exhibited by the triad of a low posterior hairline, a short neck, and variably limited neck motion (Fig. 21-43A,B) (203). Its incidence is approximately 0.7% (204). Other associated anomalies are often present both in the musculoskeletal and other organ systems. The congenital fusions result from abnormal embryologic formation of the cervical vertebral mesenchymal anlages. This unknown embryologic insult is not limited to the cervical vertebrae and explains the other anomalies associated with the Klippel-Feil syndrome. In some instances, the Klippel-Feil syndrome is familial, indicating a genetic transmission (205–207).

These children may have an associated Sprengel deformity and/or omovertebral bar (Fig. 21-44) (203, 208). Other anomalies associated with the syndrome are scoliosis (both congenital and idiopathic like) (203), congenital limb deficiency (209), renal anomalies (210), deafness (211), synkinesis (mirror movements) (212), small spinal cords but with increased SAC (213, 214), pulmonary dysfunction (215), and congenital heart and vascular disease (216–218). Radiographs demonstrate a wide range of deformity, ranging from simple block vertebrae to multiple and bizarre anomalies. The fusions become more apparent as the child ages, and posterior fusions are more common than anterior fusions when the fusions are incomplete (219). Klippel-Feil syndrome can be divided into three types depending upon the extent of vertebral involvement;

type I involves the cervical and upper thoracic vertebra, type II only the cervical vertebra alone, and type III the cervical vertebra as well as lower thoracic or upper lumbar vertebra (220). Superior odontoid migration may also occur (221). Associated scoliosis makes interpretation of the radiographs even more difficult. Flexion and extension lateral radiographs are used to assess for instability, and should always be done prior to any general anesthetic. If instability is noted on the flexion and extension radiographs, the anesthesiologist should be so informed. The anesthesiologist may elect to undertake intubation differently (e.g., awake nasotracheal, fiberoptic-guided). Any segment adjacent to unfused segments may develop hypermobility and neurologic symptoms (222, 223). A common pattern is fusion of C1-C2 and C3-C4, leading to a high risk of instability at the unfused C2-C3 level (224). If the flexion and extension radiographs are difficult to interpret, a flexion and extension CT or MRI scan can be useful. CT is especially helpful at the C1-C2 level in assessing the SAC; sagittal MRI is more helpful at other levels.

All children with Klippel-Feil syndrome should be further evaluated for other organ system problems. A general pediatric evaluation should be undertaken by a qualified pediatrician to ensure that no congenital cardiac or other neurologic abnormalities exist. Renal imaging should be done in all children; simple renal ultrasonography is usually adequate for the initial evaluation (225). MRI should be performed whenever any concern for neurologic involvement exists on a clinical basis in order to define the site and cause of neurologic pathology. An MRI should also be performed before any orthopaedic spinal procedure; this is to rule out any other intraspinal pathology that might not be present clinically or radiographically (e.g., Arnold-Chiari malformation, tethered cord, nonosseous diastematomyelia) (226).

The natural history depends on the presence of renal or cardiac problems with the potential for organ system failure and death. Cervical spine instability (227–229) can develop with neurologic involvement, especially in the upper segments or in those with iniencephaly (227, 230). The more numerous the occipitoatlantal anomalies, the higher the neurologic risk (231). Degenerative joint and disc disease develops in those patients with lower segment instabilities. In adulthood, many patients with Klippel-Feil syndrome will complain of headaches, upper extremity weakness, or numbness and tingling. Subtle findings on neurologic examination can be seen in up to half of these adults. Those with mirror movement disorders are likely to have cervicomedullary neuroschisis (232). Degenerative disc disease, as seen on MRI scans, occurs in nearly 100% of these patients (233).

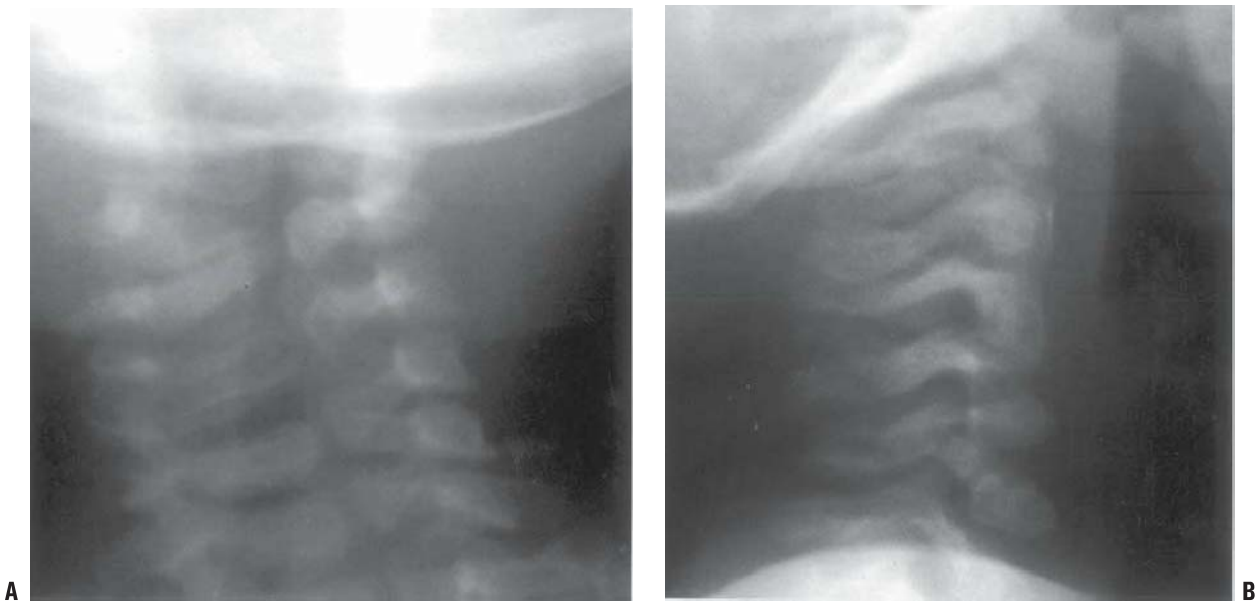
Because children with large fusion areas (Fig. 21-45) are at high risk for developing instabilities, strenuous activities should be avoided, especially contact sports (229, 234). Other nonsurgical methods of treatment are cervical traction, collars, and analgesics when mechanical symptoms appear, usually in the adolescent or the adult patient. Arthrodesis is needed for neurologic symptoms because of instability. Asymptomatic hypermobile segments pose a dilemma regarding stabilization. Unfortunately, no guidelines exist for this problem. The need for decompression at the time of stabilization depends on the exact anatomic circumstance, as will the need for combined anterior and posterior versus simple posterior fusions alone. Surgery for cosmesis alone is usually unwarranted and risky.

Author's Preferred Treatment. Once diagnosed, it is mandatory that imaging of the genitourinary system be performed if not already done. The patient is counseled against contact sports, especially collision sports (football, wrestling, ice hockey) and those sports that may place the cervical spine under stress (e.g., gymnastics, diving, basketball, soccer, volleyball). I typically ask the child what sports he/she likes to participate in, and then determine if that sport is likely to place the cervical spine under stress. If so, then that activity should be avoided. Arthrodesis is reserved for the very rare instance of symptomatic hypermobility.

Os Odontoideum. Os odontoideum is a rare anomaly where the tip of the odontoid process is divided by a wide transverse gap, leaving the apical segment without its basilar support (235). The exact incidence is not known. It most likely represents an unrecognized fracture at the base of the odontoid or damage to the epiphyseal plate during the first few years of life (235, 236). Either of these conditions can compromise the blood supply to the developing odontoid, resulting in the os odontoideum. MRI scans have further documented the presence nuchal cord changes consistent with trauma (237). A congenital etiology has also been proposed (238–241). It might represent an embryologic anomaly characterized by segmentation at the junction of the proximal 1½ somites of the 2½ somites from which the odontoid forms (242). Failure of segmentation of the dens from the anterior arch of the atlas (resulting in the so-called jig-saw sign) (240), with or without incomplete fusion of the atlas (the bipartite atlas) (239, 241). Sankar et al. (243) believe that the etiology is multifactorial and can be either congenital or posttraumatic.

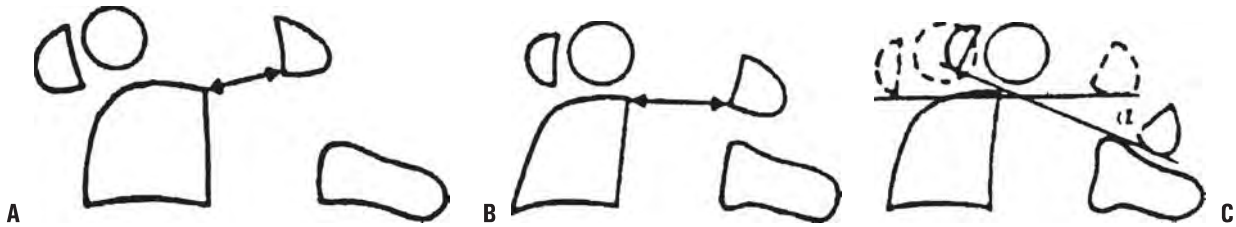
Local neck pain is the usual presentation; transitory episodes of paresis, myelopathy, or cerebral brain stem ischemia due to vertebral artery compression from the upper cervical instability are less common. Sudden death rarely occurs.

Radiographs demonstrate an oval or a round ossicle with a smooth sclerotic border of variable size, located in the position of the normal odontoid tip. It is occasionally located near the basioccipital bone in the foramen magnum area. There are three radiographic types of os odontoideum; round, cone, and blunt tooth (244). The base of the dens is usually hypoplastic. The gap between the os and the hypoplastic dens is wider than in a fracture, usually well above the level of the facets. However, it may be difficult to differentiate an os odontoideum from nonunion following a fracture. Tomograms and CT scans are useful to further delineate the bony anatomy and flexion and extension lateral radiographs to assess instability. The instability index and the sagittal plane rotation angle can be measured (Fig. 21-46) (245). The presence of



A B C

FIGURE 21-45. This 2-month-old girl presented with a left-sided torticollis. The AP (**A**) and lateral (**B**) cervical spine radiographs demonstrate congenital anomalies of the cervical spine at C1-C2. **C:** At age 7 years and 3 months, these have further ossified and matured, demonstrating massive congenital fusions of the cervical spine.



A B C

FIGURE 21-46. Radiographic parameters used to determine the instability index and sagittal plane rotation in os odontoideum. The minimum (**A**) and maximum (**B**) distances from the posterior border of the body of C2 to the posterior atlantal arch. The instability index = [(maximum distance – minimum distance)/maximum distance] × 100%. **C:** The change in the atlantoaxial angle between flexion and extension is the sagittal plane rotation. (From Watanabe M, Toyama Y, Fujimura Y. Atlantoaxial instability in os odontoideum with myelopathy. *Spine* 1996;21:1435–1439, with permission).

myelopathy is highly correlated with a sagittal plane rotation angle ≥ 20 degrees and an instability index ≥ 40%; it is also most common in the round type of os odontoideum (244). Myelopathy is also associated with cystic or fibrocartilaginous masses either behind the odontoid, within the transverse ligament, or at the level of the articulation between the os odontoideum and the remainder of the odontoid (238, 246–248); these typically regress after successful stabilization and arthrodesis (249).

The neurologic symptoms are due to cord compression from posterior translation of the os into the cord in extension or the odontoid into the cord in flexion. Hypermobility at the C1-C2 level may cause vertebral artery occlusion with ischemia of the brain stem and posterior fossa structures; this will result in seizures, syncope, vertigo, and visual disturbances.

Those with local pain or transient myelopathies often recover with immobilization. Subsequently, only nonstrenuous activities should be allowed, but curtailment of activities in the pediatric age group can be difficult. The risk of a small insult leading to catastrophic quadriplegia and death must be weighed. The long-term natural history is unknown.

Surgery is indicated when there is 10 mm or more of ADI, a SAC of 13 mm or less (37), neurologic involvement, progressive instability, or persistent neck pain. Surgery should also be strongly considered in asymptomatic patients with an instability index of >40% and/or a sagittal plane rotation angle of >20 degrees. A Gallie fusion is recommended. The surgeon must be careful when tightening the wire so that the os is not pulled back posteriorly into the canal and cord with disastrous consequences. In small children, the wire may be eliminated. In all children, a Minerva or a halo cast or vest also is used for at least 6 weeks, and often for 12 weeks. C1-C2 screw fixation has also been described in pediatric atlantoaxial instability (250) for those children older than 4 years of age. For all children undergoing C1-C2 posterior arthrodesis, care should be taken to avoid fixation of the C1-C2 segment in hyperlordosis, as that will lead to subaxial cervical kyphosis postoperatively (251).

Developmental and Acquired Stenoses and Instabilities

Down Syndrome. Because of underlying collagen defects in these children, cervical instabilities can develop at both the occiput-C1 and C1-C2 levels. The instability may occur at more than one level and in more than one plane (e.g., sagittal and rotary planes). With the advent of the Special Olympics, there has been much concern regarding the participation of children with Down syndrome, and much confusion regarding the appropriate approach to the problem of upper cervical instability in these children. Outlined below are the most recent recommendations regarding this problem.

The incidence of occiput-C1 instability has been reported to be as high as 60% in children (41) and 69% in adults (252). The vast majority are asymptomatic (253, 254). Measurement reproducibility is poor (255), but a Power's ratio of <0.55 is more likely to be associated with neurologic symptoms (256). Recent data also indicate that there are often underlying congenital differences in the shape of the occiput-C1 joint (lack of concavity of the superior surface of the lateral masses of C1) in Down children with occiput-C1 instability (257). No guidelines exist regarding the frequency of periodic screening or indications for surgery, with the exception of those for atlantooccipital fusion in the symptomatic child. Tredwell et al. (41) believe that treatment plans for these children should depend on the amount of room available for the cord rather than absolute values of displacement for both atlantoaxial and atlantooccipital instability.

Atlantoaxial instability in children with Down syndrome was first reported by Spitzer et al. in 1961 (252). Subsequently, there have been many reports on this instability. Despite these reports, there are none that document the true incidence of atlantoaxial dislocation (in contrast to instability), and there are no long-term studies regarding the natural history of this problem.

The incidence of atlantoaxial instability in children with Down syndrome has been estimated to range from 9% to 22% (41, 258–260). The incidence of symptomatic atlantoaxial instability is much less; it was reported to be 2.6% (258) in a series of 236 Down syndrome patients. Progressive instability and neurologic deficits are more likely to develop in boys older than 10 years (260). Children with Down syndrome have a significantly greater incidence of cervical skeletal anomalies, especially persistent synchondrosis and spina bifida occulta of C1, than do normal children (261). Also, children with both Down syndrome and atlantoaxial instability have an increased frequency of cervical spine anomalies, compared with other Down syndrome children without atlantoaxial instability (261). These spinal anomalies may be a contributing factor in the cause of atlantoaxial instability in these children.

The majority of children with atlantoaxial or occipitoatlantal hypermobility are asymptomatic. When symptoms occur, they are usually pyramidal tract symptoms, such as gait abnormalities, hyperreflexia, easy fatigability, and quadriparesis. Occasionally, local symptoms exist, such as head tilt, torticollis, neck pain, or limited neck mobility. The neurologic deficits are not necessarily attributable to hypermobility of the atlantoaxial or occipitoatlantal joints. Neurologic symptoms in one series of adult Down syndrome patients were equally as common in those with an increased ADI as those with a normal ADI (262). Further evaluation with flexion/extension CT or MRI scans to assess for cord compression is needed in this situation.

Rarely does sudden catastrophic death occur. Nearly all of the individuals who have experienced catastrophic injury to the spinal cord have had weeks to years of preceding, less severe neurologic abnormalities. In a review by the American Academy of Pediatrics, 41 cases of symptomatic atlantoaxial instability were compiled. In only 3 of these 41 children did the initiation or the worsening of symptoms of atlantoaxial instability occur after trauma during organized sports activities (259).

In the past, screening of Down syndrome patients with lateral flexion/extension radiographs had been recommended

(263). However, symptomatic atlantoaxial instability is very rare, and the chances of a sports-related catastrophic injury are even rarer. The reproducibility of radiographic screening for atlantoaxial and occipitoatlantal mobility is poor (254, 255, 264). Furthermore, the radiologic picture can change over time, most frequently from abnormal to normal (260). Because of all these factors, and the absence of any evidence that a screening program is effective in preventing symptomatic atlantoaxial and occipitoatlantal mobility (254), lateral cervical radiographs are of unproven value, and the previous recommendations for screening radiographs by the American Academy of Pediatrics have been retired (259).

The identification of patients with symptoms or signs consistent with symptomatic spinal cord injury is thus more important than radiographs. Neurologic examination is often difficult to perform and interpret in these children (41). Parental education as to the early signs of myelopathy is extremely important (e.g., increasing clumsiness and falling, worsening of upper extremity function). A thorough history and a neurologic examination are more important before participation in sports than are screening radiographs. However, further research is needed in this confusing matter, and because of persistent concerns, the Special Olympics does not plan to remove its requirement that all Down syndrome athletes have radiographs of the cervical spine before athletic participation.

Because of this requirement, spinal radiographs are often obtained without neurologic symptoms. When these are available, they should be reviewed to determine if there are any other associated anomalies, such as persistent synchondrosis of C2, spina bifida occulta of C1, ossiculum terminale, os odontoideum, or other less common anomalies. When the plain radiographs indicate atlantoaxial or atlantooccipital instability of 6 mm or more in an asymptomatic patient, CT and MRI scans in flexion and extension can determine the extent of neural encroachment and cord compression.

Once a Down syndrome patient presents with radiographic instability, what treatment should be instituted? Those with asymptomatic atlantoaxial or occipitoatlantal hypermobility should probably be followed up with repeat neurologic examinations; the role of repeat radiographs is more clouded as noted in the previous discussion. Because the risk of a catastrophic spinal cord injury is extremely low with organized sports in Down syndrome children without any neurologic findings, the avoidance of high-risk activities must be individualized. For those children with sudden-onset or recent progression of neurologic symptoms, immediate fusion should be undertaken if appropriate imaging confirms cord compromise. The most difficult question concerns the patient with upper cervical hypermobility with minimal or nonprogressive chronic symptoms. Before embarking upon arthrodesis, imaging with flexion/extension MRI (96) or CT should be undertaken to confirm cord compression from the hypermobility, and to eliminate other central nervous system causes of neurologic symptoms. CT is faster, reducing the need for sedation, which can be potentially dangerous in these children. CT also visualizes the C1-C2 relationships necessary to measure the SAC. MRI is more useful for evaluating other central nervous system lesions. Even if successfully stabilized, patients with chronic symptoms often show little symptomatic improvement after arthrodesis (265).

Posterior cervical fusion at the levels involved is the recommended surgical treatment. The classic technique for posterior C1-C2 fusion uses autogenous iliac crest bone graft with wiring and postoperative halo cast immobilization. Internal fixation with wiring and/or transarticular (266) screws provides protection against displacement, shortens the time of postoperative immobilization, permits the consideration of using less rigid forms of external immobilization, and is reported to aid in obtaining fusion (267–270). However, internal fixation with sublaminar wiring poses added risk. If the instability does not reduce on routine extension films, the patient is at high risk for development of iatrogenic quadriplegia with sublaminar wiring and acute manipulative reduction (271, 272). For this reason, it has been recommended that preoperative traction be used to effect the reduction. If reduction does not occur with traction, then only an onlay bone grafting should be performed without sublaminar wiring (271). Sublaminar wiring at C2 is not recommended regardless of the success of reduction because sublaminar wiring at C2 was associated with the only death in one series (273). If wiring is to be performed, pliable, smaller-caliber wires should be used. Satisfactory results can be obtained with onlay bone grafts and rigid external immobilization without internal fixation (274).

The Down syndrome patient is at higher risk for postoperative complications (neurologic and other) after fusion (275–277). Neurologic complications can range from complete quadriplegia and death to Brown-Sequard syndrome (273). Another potential cause of neurologic impairment is over reduction if an unstable os odontoideum is present (273). A posterior translation of the ring of C1 and the os fragment into the SAC can occur from this over reduction. In a study of the results of surgical fusion in 35 symptomatic Down syndrome children, 8 made a complete recovery, 14 showed improvement, 7 did not improve, 4 died, and the outcome for 2 is unknown (258). Patients with long-standing symptoms and marked neural damage showed no or little postoperative improvement, whereas patients with a more recent onset of symptoms usually made an excellent recovery. Other complications are loss of reduction despite halo cast immobilization and resorption of the bone graft with a stable fibrous union or unstable nonunion (274, 275).

The long-term results after cervical fusion are not yet known. Individuals with Down syndrome who undergo short cervical fusions are at risk for developing instability above the level of fusion, such as occiput-C1 after a C1-C2 fusion or C1-C2 after lower level fusions (278, 279). This later instability occurred in four of five children between 6 months and 7 years after surgery (278).

Author's Preferred Treatment. All children with Down syndrome should avoid collision sports (boxing, football, wrestling), even those with normal flexion/extension lateral radiographs. This

seems prudent in view of the known underlying ligamentous laxity and potential for development of cervical instability. Also, all children with Down syndrome should avoid any sports or activities that do or potentially may stress the cervical spine (boxing, football, wrestling, ice hockey, basketball, diving, gymnastics). Certainly any child with progressive instability yet who is neurologically intact should also not participate in any cervical spine–stressing activities. These children should also be followed closely from a clinical perspective to observe for the development of any neurologic symptoms or signs. Those children with neurologic signs or symptoms and cervical instability should undergo arthrodesis, usually posterior. Most instabilities correct with simple positioning. Internal fixation is advised, except for sublaminar wires at C2. If instability is present and does not reduce on routine extension films, the patient is at high risk for development of iatrogenic quadriplegia with sublaminar wiring and acute manipulative reduction. Preoperative traction should be used in such a situation to effect reduction. If reduction does not occur, then only an onlay bone grafting should be performed without internal fixation. The high complication rate associated with these procedures should be remembered and parents counseled accordingly.

Marfan Syndrome. Marfan syndrome affects ligamentous laxity and bone morphology. It is due to a mutation in the glycoprotein fibrillin, which has been mapped to the long arm of chromosome 15. Abnormalities regarding the cervical in this syndrome have only recently been described (280–282). These are primarily radiographic abnormalities but can be clinically significant instabilities (282). Focal cervical kyphosis involving at least three consecutive vertebrae occurs in 16%, with an average kyphosis of 22 degrees. The normal cervical lordosis is absent in 35%. Atlantoaxial hypermobility is common—54%. There is also an increased incidence of radiographic basilar impression (36%). Unlike Down syndrome, there is no increased incidence of cervical skeletal anomalies such as persistent synchondrosis and spina bifida occulta of C1. In spite of these radiographic abnormalities in those with Marfan syndrome, symptoms or neurologic compromise is rare. Neck pain is not increased compared to the general population. Patients with Marfan syndrome should be recommended to avoid sports with high-impact loading on the cervical spine; it does not appear necessary to routinely perform cervical spine radiographs for those undergoing general anesthesia. Atlantoaxial rotatory subluxation may be increased in those with Marfan syndrome, and this should be specially noted during surgical positioning.

Nontraumatic Occipitoatlantal Instability. Nontraumatic occipitoatlantal instability is rare in the absence of any underlying syndrome (e.g., Down syndrome). Georgopoulos et al. (283) have described pediatric nontraumatic atlantooccipital instability. Congenital enlargement of the occipital condyles may have been the cause by increasing motion at this joint. The presenting symptoms were severe vertigo in one 14-year-old boy and nausea with projectile vomiting in one 6-year-old girl. These symptoms are postulated to be a result of vertebrobasilar arterial insufficiency resulting from the hypermobility at the occiput-C1 junction. The diagnosis of instability is suggested by plain radiographs initially and confirmed by cineradiography. Both children were treated with a posterior occiput-C1 fusion with resolution of symptoms.

Cerebral Palsy. Cervical radiculopathy and myelopathy in cerebral palsy (284–287) was first described in the athetoid types and subsequently in the spastic types. Athetoid cerebral palsy patients, when compared with the normal population, develop cervical disc degeneration at a younger age. This degeneration progresses more rapidly and involves more levels. Angular and listhetic instabilities also are more frequent and appear at a younger age (288). The combination of disc degeneration and listhetic instability predisposes these patients to a relatively rapid, progressive neurologic deficit.

The symptoms are brachialgia and weakness of the upper extremity with decreased functional use or increased paraparesis or tetraparesis (285–287). In ambulatory patients, a loss of ambulatory ability is often a sign of presentation. Occasional loss of bowel and bladder control also occurs. Atlantoaxial instability has been recently described in patients with severe spastic quadriplegia; the symptoms are usually apnea, opisthotonos, torticollis, respiratory problems, muscle tone abnormalities and hyperreflexia, and bradycardia (289).

Radiographic findings (Fig. 21-47) are narrowing of the spinal canal and premature development of cervical spondylosis; malalignment of the cervical spine with localized kyphosis, increased lordosis, or both; and instability of the cervical spine manifested as spondylolisthesis. Flattening of the anterosuperior margins of the vertebral bodies and beak-like projections of the anteroinferior margins are radiographic findings of the spondylosis. Myelography demonstrates stenosis, disc protrusion, osteophyte projection, and blocks in dye flow, most commonly at the C3-C4 and C4-C5 levels.

The kyphosis, herniated discs, and osteophytes result in nerve root and cord compression. It is believed that the exaggerated flexion and extension of the neck in these young adults with cerebral palsy causes accelerated cervical degeneration and cervical stenosis earlier than in unaffected people, who develop stenosis in the late fourth and fifth decades of life. Exaggerated flexion and extension occurs in patients with athetosis and writhing movements. Difficulty with head control also can cause exaggerated flexion and extension in the spastic cerebral palsy patient.

Treatment is primarily surgical. Anterior discectomy, resection of osteophytes, and interbody fusion have been the most effective methods. A halo is best and is well tolerated in some patients with athetosis (286). However, postoperative immobilization can be a problem for some patients, and thus some authors also recommend a posterior wiring of the facets as well to minimize the amount of time postoperative immobilization is needed (285). Posterior laminectomy alone (286) is contraindicated in cerebral palsy patients with developmental cervical stenosis because this will increase the instability. Long-term

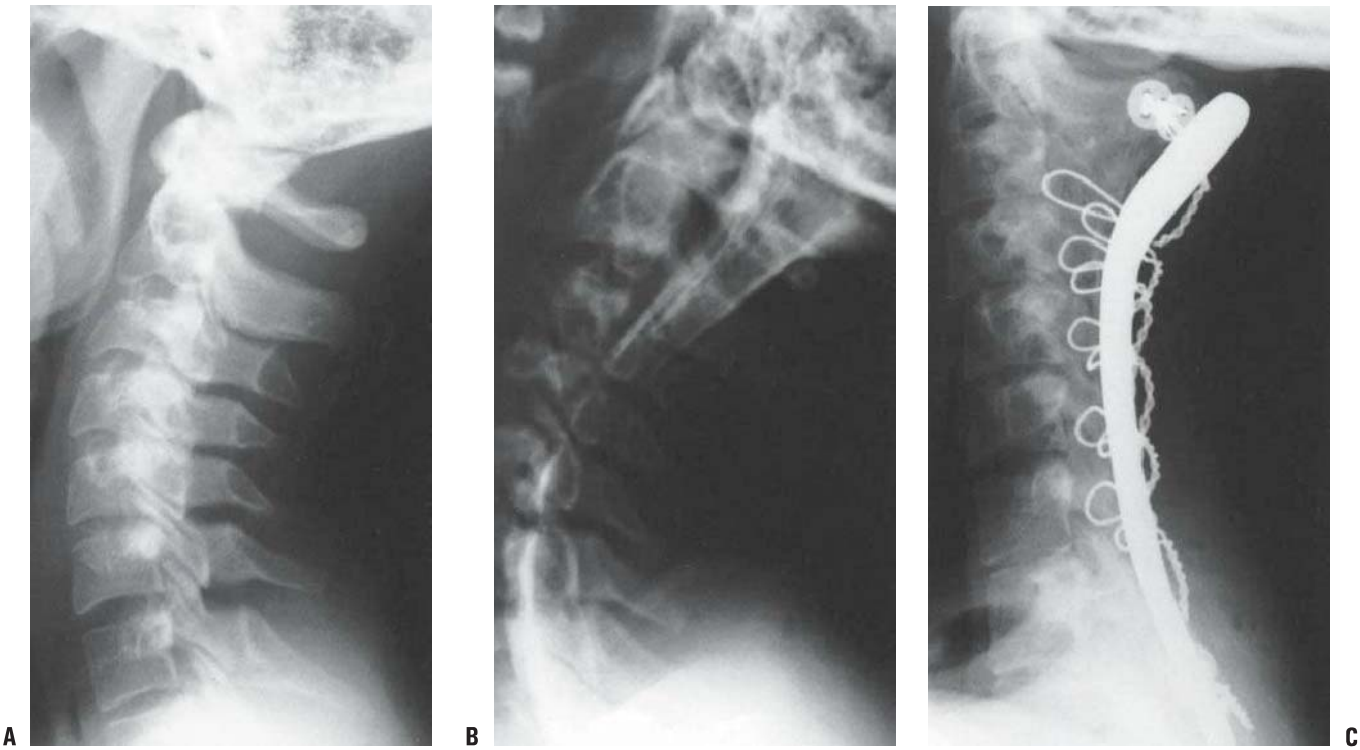


FIGURE 21-47. A 14-year-old girl with spastic quadriparesis showed progressive loss of upper extremity function with loss of ability to control her wheelchair and feed herself. She also complained of some mild neck pain. **A:** The lateral radiograph shows marked stenosis from C3-C6, as evidenced by a spinal canal-to-vertebral body ratio (Torg ratio) of <0.8. **B:** The myelogram shows near complete block of the dye column from C3-C5. This stenosis was treated by posterior laminectomy from C3-C7 and posterior cervical fusion from C2-T1 using Luque rectangle fixation with spinous process and facet wiring. **C:** Eight months postoperatively, there is stable fixation and solid facet joint fusion. The girl's upper extremity strength is improved, and she is able to feed herself. (From Loder RT, Hensinger RN. Developmental abnormalities of the cervical spine. In: Weinstein SL, ed. *The Pediatric Spine: Principles and Practice*, 1st ed. New York, NY: Raven Press, 1994, with permission.)

follow-up of surgically treated patients demonstrates late disc degeneration and increased range of motion at adjacent segments in those who underwent anterior arthrodesis (290).

Postlaminectomy Deformity. Cervical kyphosis is common after cervical laminectomy in children (291–299). This phenomenon is more likely in immature, growing children. It has been duplicated in animal models; a C3-C6 laminectomy in growing cats uniformly resulted in kyphosis; whereas normal cervical curves were maintained in adult cats (300). The natural history of postlaminectomy kyphosis is unknown; however, the incidence of kyphosis when extensive cervical laminectomies are performed in childhood varies from 33% to 100%, with an overall average of 70% (296). Postlaminectomy kyphosis is weakly age dependent (mean age at laminectomy of 10.5 years) and not dependent upon the total number of levels decompressed or the location of these levels (296) unless decompression spanning both the C1-C2 and C7-T1 levels is performed, which then increases the risk four fold (298). Postlaminectomy lordosis is less common and is strongly correlated with a peak age at decompression of 4 years (296). In one study, 12 of 15 children who had undergone a cervical or cervicothoracic laminectomy prior to 15 years of age developed kyphosis (295). The normal posterior muscular attachments to the spinous processes and laminae, as well as facet capsules, the ligamentum nuchae, and the ligamentum flavum, are violated by the laminectomy. This loss of posterior supporting structures allows for a progressive deformity, which, if kyphotic in nature, can eventually result in neurologic symptoms and deficits. Early radiographic features are a simple kyphosis; later, vertebral body wedging and anterior translations of one vertebral body on another can develop. A late, severe deformity is the swan neck deformity (294). Neurologic problems result from cord stretch and compression from the anterior kyphotic vertebral bodies. MRI is useful to delineate the extent of cord attenuation and compression.

Nonsurgical treatment starts with frequent radiographic follow-up studies after a laminectomy; the role of prophylactic bracing is not yet known. If only instability becomes present, then posterior cervical facet fusion can be performed (Figs. 21-10 to 21-16). After kyphotic deformities develop, anterior vertebral body fusion with halo cast or vest; or Minerva cast immobilization is recommended (293) (Fig. 21-48). The role of a prophylactic posterior fusion at the time of laminectomy is

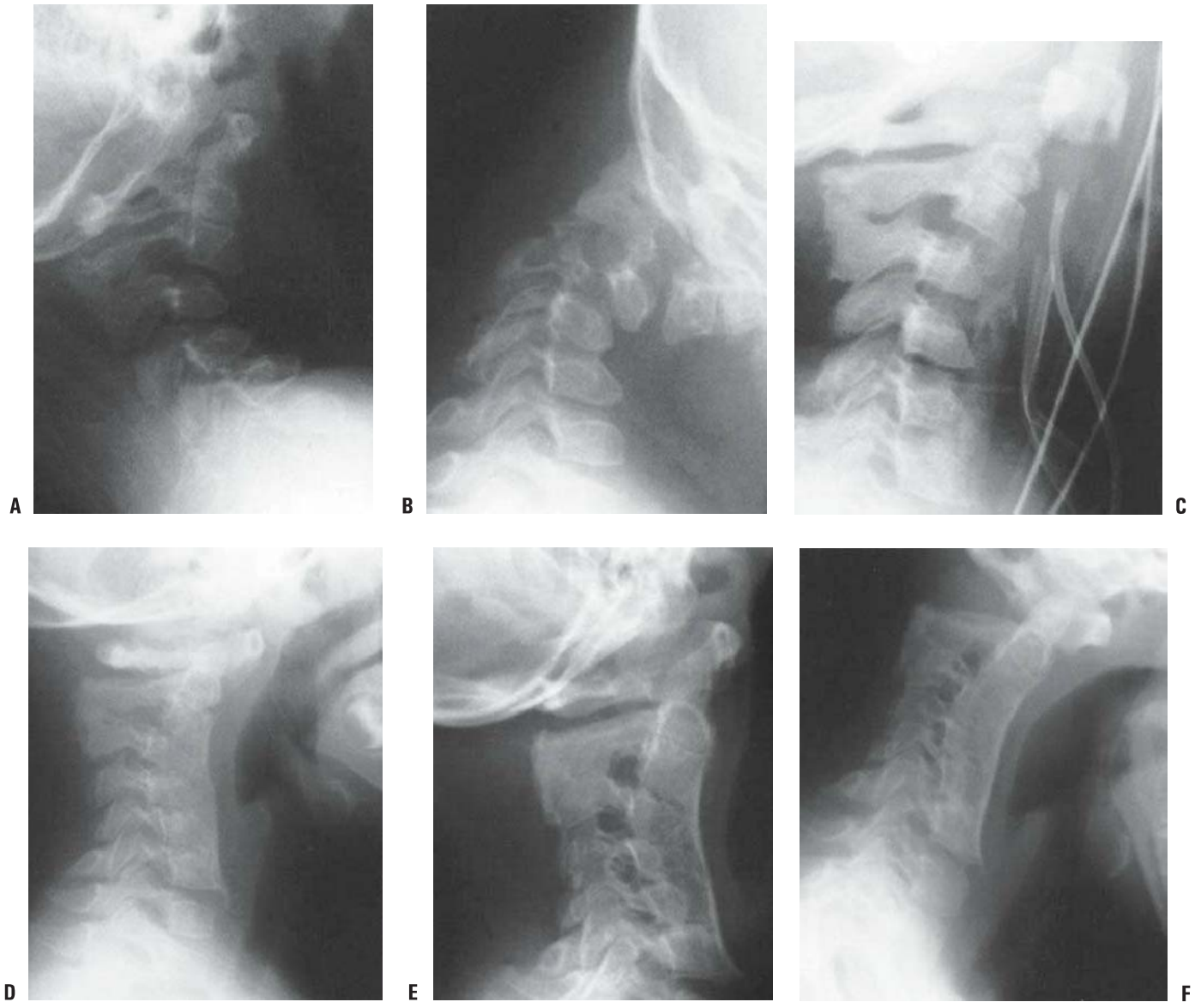


FIGURE 21-48. This girl underwent a cervical laminectomy from C2-C6 for a low-grade astrocytoma of the cervical cord. At 1 year and 7 months of age, she had a postlaminectomy kyphosis that was 45 degrees in extension (**A**) and 82 degrees in flexion (**B**). **C:** An anterior cervical discectomy and fusion from C2 to C6 was performed with autogenous iliac crest strut graft. Immediately postoperative, the kyphosis was corrected to 20 degrees. Halo-vest immobilization was used for 3 months. **D:** Solid incorporation of the fusion occurred by 6 months postoperatively. At 4-years and 7 months of age, flexion (**E**) and extension (**F**) lateral radiographs show maintenance of the correction, solid fusion, and no instability at the remaining levels.

not yet known (291), nor is the role of osteoplastic laminotomy instead of laminectomy (301), although this approach might not always be amenable to the primary pathology.

Other Syndromes

Fetal Alcohol Syndrome. Central nervous system dysfunctions, growth deficiencies, facial anomalies, and variable major and minor malformations are the characteristics of the fetal alcohol syndrome. The children present with developmental delay, especially in motor milestones, failure to thrive, mild-to-moderate retardation, mild microcephaly, distinct facies (hypoplasia of the facial bones and circumoral tissues), and congenital cardiovascular anomalies. The cervical findings are similar to those in the Klippel-Feil syndrome. Radiographically congenital fusion of two or more cervical vertebrae occurs in approximately half of the children, resembling Klippel-Feil syndrome (302). The major visceral anomaly in the Klippel-Feil syndrome is the genitourinary system, whereas in fetal alcohol syndrome, it is in the cardiovascular system (302).

The natural history is not known. Radiographic imaging and treatment recommendations regarding the cervical spine are the same as those for the Klippel-Feil syndrome.

Craniofacial Syndromes. Cleft lip and palate is the most common craniofacial anomaly. It can be a solitary finding, but more often associated with other syndromes and anomalies. Children with cleft anomalies have a 13% to 18% incidence of cervical spinal anomalies compared with the 0.8% incidence of children undergoing orthodontia care for other reasons (303, 304). This incidence is highest in patients with soft palate and submucous clefts (45%). These anomalies, usually spina bifida and vertebral body hypoplasia, are predominantly in the upper cervical spine. The potential for instability is unknown, as is the natural history. No documented information regarding treatment is available; however, the clinician should be aware of this association and make sound clinical judgments as needed. They also demonstrate a reduced cervical lordosis compared to those without cleft lip and/or palate (304).

Craniosynostosis Syndromes. The craniosynostosis syndromes—Crouzon, Pfeiffer, Apert, Goldenhaar, and Saethre-Chotzen—exhibit cervical spine fusions, atlantooccipital fusions, and butterfly vertebrae (305–310). Fusions are more common in Apert syndrome (71%) than in Crouzon syndrome (38%) (305). Upper cervical fusions are most common in Crouzon and Pfeiffer syndromes (307), whereas in Apert syndrome the fusions are more likely to be complex and involve C5-C6 (305). However, this syndrome variation is not accurate enough for syndromic differentiation. Congenital cervicothoracic scoliosis with rib fusions is seen in Goldenhar syndrome, usually from hemivertebrae (307, 311). C1-C2 instability in Goldenhar syndrome may be as high as 33%, and these children should be monitored carefully for this potential problem (311).

The cervical fusions are progressive with age; in younger children the vertebrae appear to be separated by intervertebral discs, but as the children grow older the vertebrae fuse together. There are no specific, standard recommendations for treatment. The author recommends following the same principles as in Klippel-Feil syndrome. The main concern is the potential difficulty with intubation in these children. Odontoid anomalies are rare; however, if any question exists regarding the stability of the cervical spine, lateral flexion and extension radiographs should be obtained. Children with Goldenhar syndrome have a high incidence of C1-C2 instability (312) and failures of segmentation (313). Children with Goldenhar syndrome have a much higher incidence of their mothers being diabetic; it has recently been suggested that children with Goldenhar syndrome should be assessed for maternal diabetes exposure, which should aid in counseling concerning cause and recurrence risk (314).

Skeletal Dysplasias. Skeletal dysplasias are discussed in detail in Chapter 4.

Combined Soft-tissue and Skeletal Dysplasias.

Neurofibromatosis. Neurofibromatosis is the most common single gene disorder in humans. The proportion of patients with neurofibromatosis and cervical spine involvement is difficult to assess: 30% of patients in the series of Yong-Hing et al. (315) and 44% of neurofibromatosis patients with scoliosis or kyphosis had cervical spine lesions. The cervical lesions are often asymptomatic (315). Symptoms, when they do occur, are diminished or painful neck motion, torticollis, dysphagia, deformity, and neurologic signs ranging from mild pain and weakness to paraparesis and quadriparesis (65, 316). Neck masses constituted 20% of presenting symptoms in one study of neurofibromatosis patients (317).

Radiographic features of neurofibromatosis in the cervical spine are vertebral body deficiencies and dysplasia or scalloping (Fig. 21-49) (315). This condition often is associated with kyphosis and foraminal enlargement (318). Lateral flexion and extension radiographs are recommended for all neurofibromatosis patients before general anesthesia or surgery (315). MRI is helpful for assessing the involvement of neural structures and dural ectasia. CT is useful for evaluating the upper cervical spine complex and bony definition of the neural foramen. The natural history regarding the cervical spine is unknown, but those with severe kyphosis often develop neurologic deterioration.

Surgical indications are cord or nerve root compression, C1-C2 rotary subluxation, pain, and neurofibroma removal (315, 316). Laminectomy alone without accompanying arthrodesis is contraindicated (319). A halo cast or vest is usually needed after fusion, with or without internal fixation, and is usually achieved with simple interspinous wiring. Kyphosis requires both anterior and posterior fusion. Pseudarthroses are frequent with isolated posterior fusions. Vascularized fibular grafts may be necessary to effect fusion in difficult cases (318, 320). If there are no indications for surgical treatment, then the patient should be followed closely.

Fibrodysplasia Ossificans Progressiva. Fibrodysplasia ossificans progressiva is an inherited, autosomal dominant disorder (321) of connective tissue with progressive soft-tissue ossification. The disorder itself is rare; most cases represent new spontaneous mutations. It likely represents overactivity of the BMP signaling pathway (322). Eventually all patients with this disorder develop cervical spine changes (323), often starting in childhood. These patients usually present with neck stiffness (324) within the first 5 years of life, and less commonly pain (325). No cases of neurologic compromise have been reported. Other general clinical features are big toe malformations, reduction defects of all digits, deafness, baldness, and mental retardation. Early in the course of the disease small, narrow vertebral bodies and large pedicles/posterior elements are seen radiographically. Occasionally, nuchal musculature ossification also is seen. Later, neural arch and facet fusions are seen (322). This factor reflects the progressive ossification of the cervical spinal musculature, ligament ossification, and spontaneous fusion of the cervical discs and apophyseal joints. No effective medical treatment is known. Surgical treatment of the cervical spine has not been necessary.

TRAUMA

Injuries to the cervical spine are rare in children and more common in boys than girls. In one study, the age- and gender-adjusted

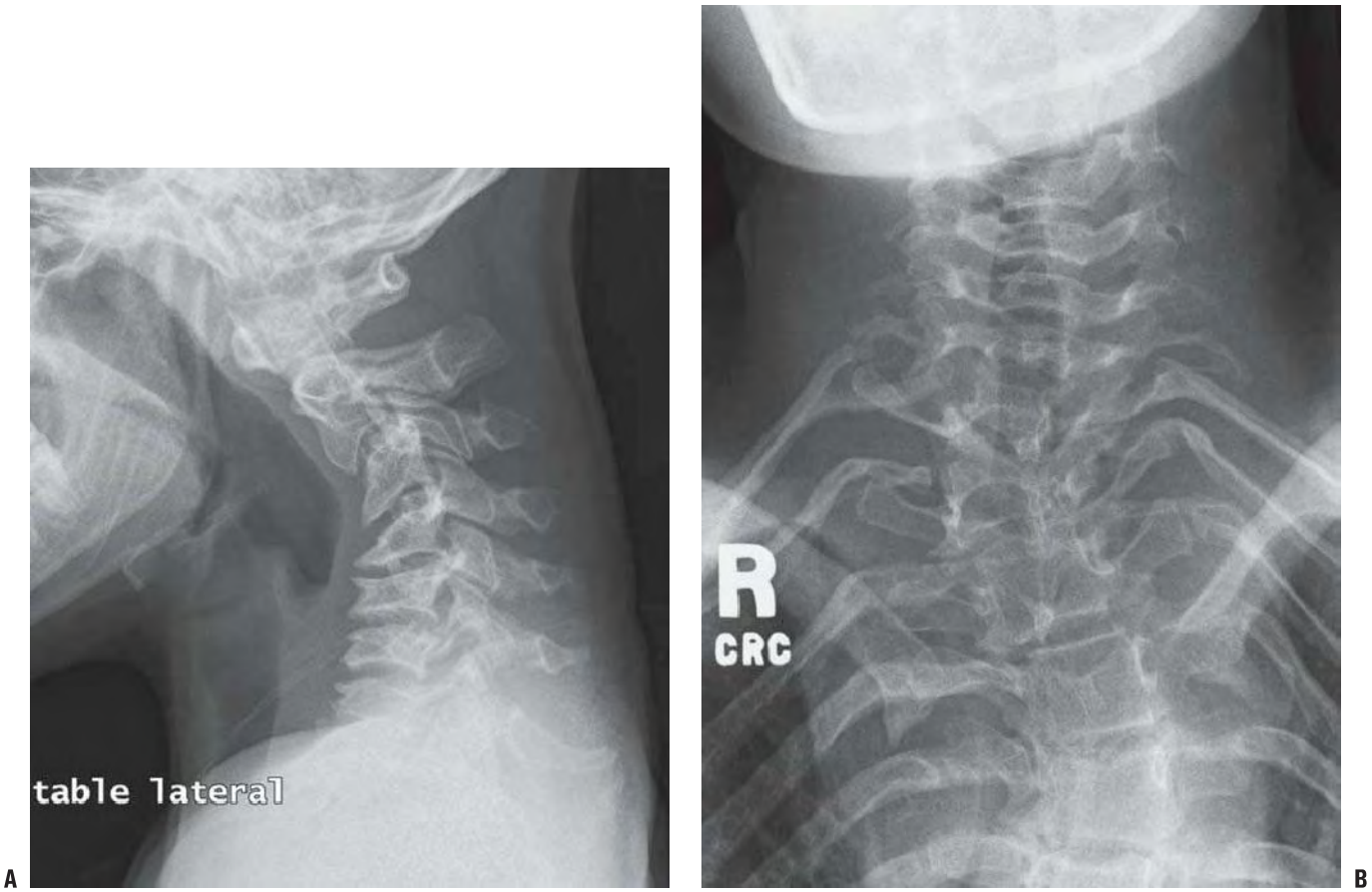


FIGURE 21-49. A 15-year-old girl with neurofibromatosis I has a painless deformity. **A:** Kyphosis on the lateral view. **B:** Notice the dysplastic changes in the vertebral bodies and the penciled ribs on the AP view.

incidence was 7.41 per 100,000 population per year (326); this incidence was much less in children (younger than 11 years of age, 1.19 per 100,000) compared with adolescents (older than 11 years of age, 13.24 per 100,00). The cause of the injury in children is frequently a fall, whereas in adolescents it is frequently sports, recreational activities, or motor vehicle crashes. Children involved in side impact crashes are more likely to have cervical spine injuries compared to those involved in frontal crashes (327). Unrestrained are more likely to sustain cervical spine injuries in motor vehicle crashes compared to restrained children (328, 329). In general, children (younger than 11 years of age) are more likely to sustain ligamentous injuries and injuries to the upper cervical spine, whereas adolescents are more likely to sustain fractures and injuries to the lower cervical spine (326). In a large series of 1098 children with cervical spine injury, upper spine injuries occurred in 52%, lower cervical spine injuries in 28%, and both upper and lower injuries in 7% (330). Upper cervical spine injuries carry a significantly higher mortality compared to lower cervical spine injuries (330, 331). By the age of 10 years, the bony cervical spine has reached adult configurations, and the injuries they sustain are essentially those of the adult. Therefore, the author will concentrate on those injuries sustained in the first decade of life.

Most children with potential cervical spine injuries have sustained polytrauma and frequently arrive immobilized on backboards and cervical collars. If the child is comatose or semiconscious, if there are external signs of head injury, or if the child complains of neck pain then cervical spine radiographs are needed. All children involved in motor vehicle crashes with head trauma and neck pain, or who have neurologic signs or symptoms, should have cervical spine radiographs (332, 333). The views recommended for this initial screening are the cross-table lateral and AP views. The need for an open-mouth odontoid is controversial, especially in children <5 years of age (334, 335). If the child is too critically ill to be positioned for all views, then the cross-table lateral view is adequate until a complete evaluation can be performed. Cervical spine precautions must be maintained until a complete evaluation has demonstrated no injury. Once a cervical injury has been identified, close scrutiny must be undertaken to ensure that there are no other injuries in the remainder of the axial skeleton.

The child arriving in the emergency suite is often on a standard backboard. Young children have a disproportionately large head, and positioning them on a standard backboard leads to a flexed posture of the neck (Fig. 21-50A) (336). This flexion can lead to further anterior angulation or translation of an unstable cervical spine injury and can also cause pseudosubluxation, which in itself in an injured child can be difficult to interpret. To prevent this undesirable cervical flexion in young children during emergency transport and radiography

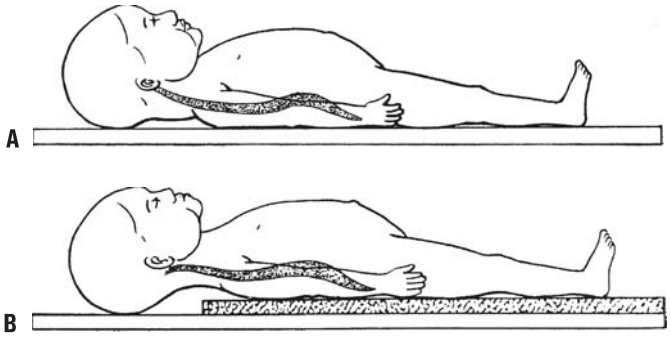


FIGURE 21-50. **A:** Positioning a young child on a standard backboard forces the neck into a kyphotic position because of the relatively large head. **B:** Positioning a young child on a double mattress, which raises the chest and torso and allows the head to translate posteriorly compensates for the relatively large head. This creates a normal alignment of the cervical spine. (From Herzenberg JE, Hensinger RN, Dedrick DK, et al. Emergency transport and positioning of young children who have an injury of the cervical spine. *J Bone Joint Surg Am* 1989;71-A:15–22, with permission).

modifications must be made by either creating a recess for the occiput of the larger head or using a double mattress to raise the chest (Fig. 21-50B). A simple clinical guideline is to align the external auditory meatus with the shoulder.

Flexion and extension lateral radiographs may be necessary to determine the stability of the cervical spine; hyperflexion ligamentous injuries may not be seen immediately, and flexion and extension views a few weeks later after the spasm has subsided may document instability. In one series of children with ligamentous injuries of the cervical spine, 8 of 11 children with lower cervical instability were diagnosed between 2 weeks and 4 months after the trauma (337).

Secondary signs of spinal injury in children often are seen before the actual injury or fracture itself. Malalignment of the spinous processes on the AP radiograph should be regarded as highly suspicious for a jumped facet. Widening of the posterior interspinous distances should be regarded as highly suspicious for a posterior ligamentous injury. In adults, an increase in the retropharyngeal soft-tissue space can indicate a hematoma in the setting of trauma and increase the suspicion on the part of the clinician that an upper cervical fracture exists. In children, however, the pharyngeal wall is close to the spine in inspiration, whereas there may be a large increase in this space with forced expiration, as in a crying child (338). This should be remembered when considering the significance of prevertebral pharyngeal soft tissue in the cervical spine radiographs of a frightened, crying child.

CT is useful to further assess the upper cervical spine, especially the occipital condyles, ring of the atlas, and occasionally the odontoid. As a rule, CT is not recommended for screening (339–342) but to further study suspicious areas on plain radiographs or for treatment planning (343). However, there is more interest in using CT as the initial screening study (344), but the concern for artifacts exists (345). It should be used to study all fractures of C1. MRI scans are useful to assess the spinal cord, discs, and interspinous ligaments (346). In an injured child, an MRI is the exam of choice to assess the pediatric cervical spine when (a) the child is obtunded and/or nonverbal and a cervical spine injury is suspected, (b) there are equivocal plain radiograph findings, (c) neurologic symptoms without radiographic findings are present, or (d) inability to clear the cervical spine in a timely manner (347).

Fractures and Ligamentous Injuries of the Occipital Complex to the C1-C2 Complex

Atlantooccipital Dislocation. Atlantooccipital dislocation is rare (348), and most of the children do not survive (349). Deployment of air bags has been recently associated with this injury in children (350–353). With the present rapid response to trauma victims and more aggressive field care, more of these children now survive. These children are usually polytrauma victims with severe head injuries and present with a range of clinical neurologic pictures (348, 349). In the past, those who survived had incomplete lesions, often demonstrating cranial nerve dysfunctions and varying degrees of quadriplegia. Many of the children who presently survive have complete loss of neurologic function below the brain stem and live only because of outpatient ventilatory support. Other presentations may be a responsive child with hypotension or tachycardia to a complete cardiac arrest. Occasionally, some patients present with normal neurologic examinations. As of 2001, there have been 29 children with atlantooccipital dislocation who have survived (354).

In severe cases the diagnosis is evident; however, some of the cases do not demonstrate marked radiographic displacement. In the past, a Power's ratio >1.0 (Fig. 21-51A) was used to indicate the presence of atlantooccipital dislocation (355). This criterion can cause the practitioner to miss isolated distraction injuries, anterior atlantooccipital dislocations that have spontaneously reduced after injury, and posterior atlantooccipital injuries (348). For this reason, the distance between the tip of the dens and the basion (Fig. 21-51B) has been used, in which a distance of more than 12.5 mm indicates the potential for

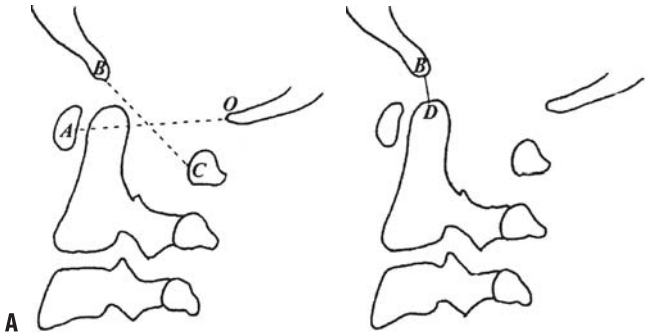


FIGURE 21-51. The *BC/OA* ratio (**A**) and the *DB* distance (**B**) are used to assess for traumatic atlantooccipital dislocation. (From Bulas DI, Fitz CR, Johnson DL. Traumatic atlanto-occipital dislocation in children. *Radiology* 1993;188:155–158, with permission.)

atlantooccipital dislocation. Recent studies have described the stabilizing nature of the tectorial membrane. When this membrane is disrupted, there is a high likelihood of atlantooccipital instability. If the C1-C2 to C2-C3 posterior interspinous ratio is >2.5, then there is a high chance of tectorial membrane disruption, and MRI evaluation is warranted (356).

The first obstacle in the treatment of this injury is its diagnosis. If the suspicion for craniocervical trauma is still present after inconclusive plain radiography, CT or MRI can be quite useful (Fig. 21-52). Subarachnoid hemorrhage at the craniocervical junction will be seen after atlantooccipital dislocation (357–359); CT can also assist in assessing osseous alignment

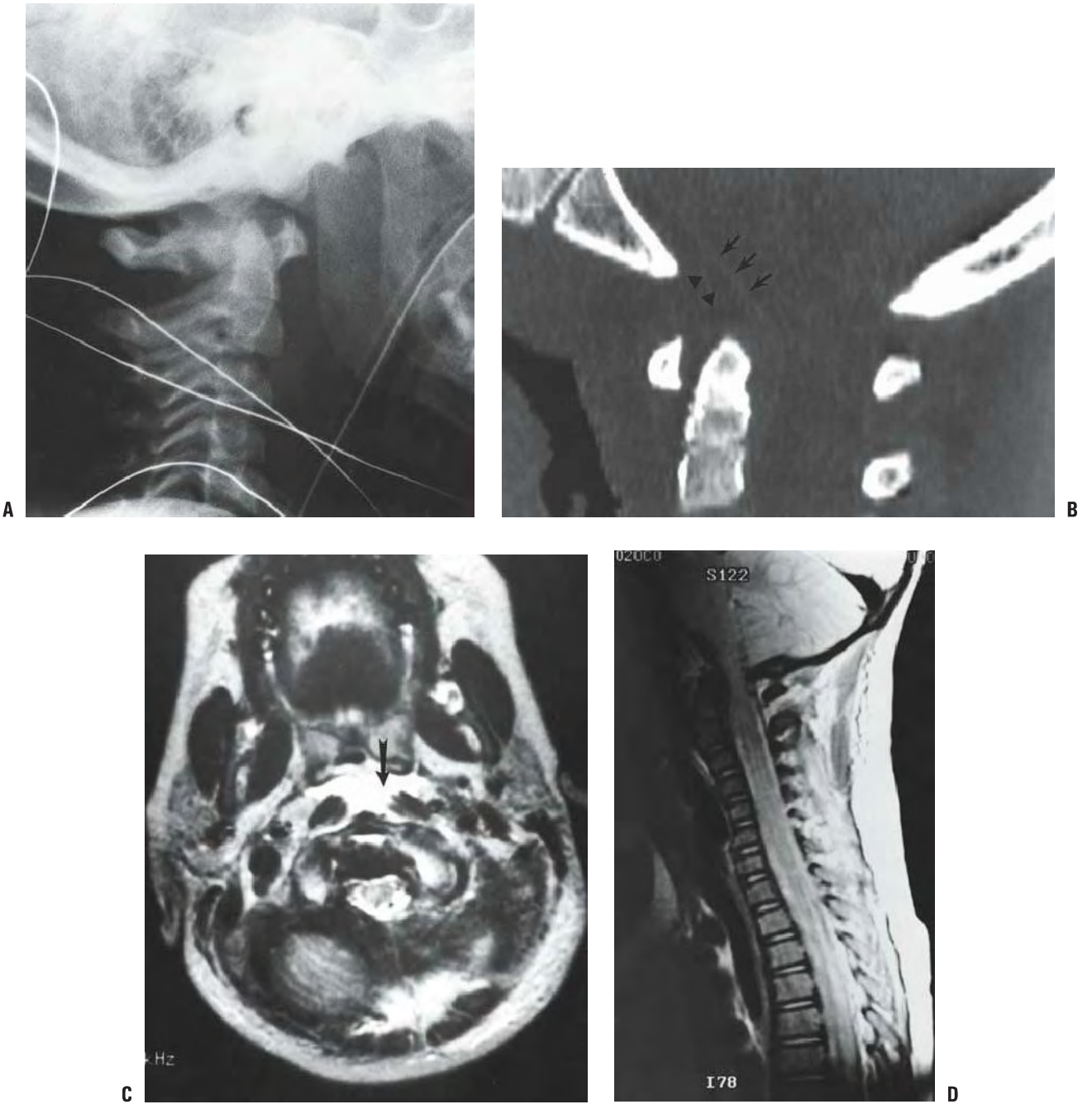


FIGURE 21-52. This 5-year, 6-month-old girl was hit by a van from behind and presented with bilateral palsies of cranial nerve VI. **A:** The lateral radiograph of the upper cervical spine demonstrates a rotational malalignment: the basion hemishadows fail to overlap while the C1 arches nearly superimpose upon each other, raising the concern for atlantooccipital dislocation. **B:** A CT scan with sagittal reconstruction demonstrates elevation of the periosteum at the caudal level of the clivus (*arrows*) and hemorrhage (*arrowheads*). **C:** An axial image from the MRI scan demonstrates abnormal fluid accumulation immediately anterior to the atlantooccipital junction (*arrow*). **D:** The MRI scan sagittal view demonstrates subarachnoid space narrowing at the level of the foramen magnum and atlantooccipital joint.