CHAPTER 10

Disorders of the Neck

Cervical Spine

Conditions involving the cervical spine in children traditionally are discussed according to the anatomic structure or segment that is abnormal. Such a classification entails enumerating all possible abnormalities (e.g., C1–2 instability) affecting a particular area (e.g., occipitocervical).

A classification based on clinical presentation, however, may be more useful for understanding cervical spine pathologies and their relation to the brain stem, spinal cord, and vertebral artery system. Such a classification will also direct the evaluation and treatment. The clinical presentations of cervical spine abnormalities include *deformity*—torticollis, kyphosis, a shortened neck—and/or *symptoms* of instability or neurologic compromise due to a cervical spinal canal made stenotic. Thus, a patient who presents with a deformity but without neurologic symptoms or signs would be evaluated for one set of cervical abnormalities, while a patient presenting without a deformity but with neurologic symptoms or instability would be evaluated for another set of conditions. A classification system based on clinical presentation, then, promotes a unified understanding of cervical spine abnormalities and their pathomechanisms.

Congenital Torticollis

Torticollis (from the Latin meaning "twisted neck") is a *symptom* of cervical spine abnormality. Its differential diagnosis (Table 10–1) might seem complicated at first glance but can be simplified by determining whether the deformity is painful or not and whether the deformity was present at birth (congenital) or was acquired.

CONGENITAL MUSCULAR TORTICOLLIS

The most common form of congenital painless torticollis is congenital muscular torticollis, or wry neck. The deformity is usually obvious at birth or shortly afterward. The child's head is tilted toward the involved fibrotic sternocleidomastoid (SCM) muscle and the chin is rotated toward the contralateral shoulder, producing the "cock robin" appearance (Fig. 10–1A). The diagnosis is made on physical examination by detecting a mass or knot on the involved side of the neck in the body of the SCM muscle in the first 3 months of life (Fig. 10–1B).[85,102] The mass may regress after early infancy and be replaced by a readily palpable fibrous contracted band that can be followed from its origin on the mastoid to the sternum and clavicular insertions. Although Coventry and Harris reported this mass was undetected in 80 percent of patients,[21] the contracture is almost universally present after infancy.

Congenital muscular torticollis was first described in 1749 by Cheselden,[102] with a more detailed description provided by Anderson in 1893.[3] Its etiology remains unknown, but the condition likely results from local compartment syndrome or ischemia involving the neck, producing the fibrotic muscle.[28] It is also almost certainly a "packing" problem, based on the high prevalence of breech positioning and primiparous birth order in this condition.[85,89] It is hypothesized that the head becomes twisted and rotated in utero and, because of intrauterine crowding, the position is maintained for a period of time prior to birth, leading to ischemia, edema, and eventual fibrosis in the muscle. There is also evidence that progressive denervation of the muscle due to compression of the accessory nerve can exacerbate the fibrotic reaction.[127] Additional evidence of the lack of uterine space includes an increased incidence of congenital dislocation of the hip and of foot deformities (e.g., metatarsus adductus).[65,143] Although patients with torticollis may be at slightly greater risk for congenital dysplasia of the hip, we have not found anything more than routine neonatal examination and screening with ultrasonography to be appropriate. Prolonged observation for dysplasia in a child with torticollis does not appear warranted.[66]

The clinical presentation varies from a simple head tilt with slight rotation and minimal restriction of motion to a more severe plagiocephaly, which can be exacerbated by the positioning of the infant for sleep (Fig. 10–1C). Flattening of the face on the ipsilateral side of the SCM lesion can be worsened by the prone position when sleeping. The infant may also have a "bat" ear due to folding in utero. If infants are placed supine for sleeping, reverse modeling of the contralateral side of the skull can occur. Treatment with various helmets and headgear has been tried in an effort to mold the infant's head while it is still soft, with open fontanelles and sutures. Older children may be referred for scoliosis evaluation because of an apparent elevation of the ipsilateral shoulder (Fig. 10–2).

The differential diagnosis of congenital muscular torticol-

TABLE 10–1 **Torticollis**

Congenital
Congenital muscular
Vertebral anomalies
Failure of segmentation
Klippel-Feil
Occipitalization of C1
Failure of formation
Congenital hemiatlas
Combined failure of segmentation/formation
Ocular
Acquired—Painful
Traumatic
Atlantoaxial rotatory displacement
Os odontoideum
C1 fracture
Inflammatory
Atlantoaxial rotatory displacement (Grisel's)
Juvenile rheumatoid arthritis
Diskitis/osteomyelitis
Other infection in neck
Tumors
Eosinophilic granuloma
Osteoid osteoma/osteoblastoma
Calcified cervical disk
Sandifer's syndrome
Acquired—Nonpainful
Paroxysmal torticollis of infancy
Tumor of the CNS
Posterior fossa
Cervical cord
Acoustic neuroma
Syringomyelia
Hysterical
Oculogyric crisis (phenothiazine toxicity)
Associated with ligamentous laxity
Down syndrome
SED/MPS dysplasias

lis includes congenital bony abnormalities producing the deformity. Therefore, good-quality plain radiographs of the cervical spine are warranted if the typical SCM muscle contracture is absent. Because of difficulty in obtaining and interpreting such radiographs in a newborn or young infant, it is acceptable to forgo them if the clinical picture of an SCM mass and fibrosis is unmistakable, along with the plagiocephaly and other facial and ear abnormalities related to the packing problem. If, however, the deformity does not respond to the usual conservative measures, then radiographic evaluation is mandatory, along with ocular and central nervous system (CNS) evaluation.

Treatment. Excellent results with massage and a stretching program can be achieved in around 90 percent of patients.[9,16,18,66,102] This is the first treatment approach. At the time of diagnosis, the parents are instructed in the technique of stretching the contracted SCM muscle by rotating the infant's chin to the ipsilateral shoulder and simultaneously tilting the head toward the contralateral shoulder. The exercises should be done gently but with the goal of attaining full passive range of motion—both rotation and tilting—as quickly as possible (Fig. 10–3). Besides stretching, positioning toys and other maneuvers to solicit active rotation toward the involved side are important to actively overcome the fibrosis of the SCM muscle.

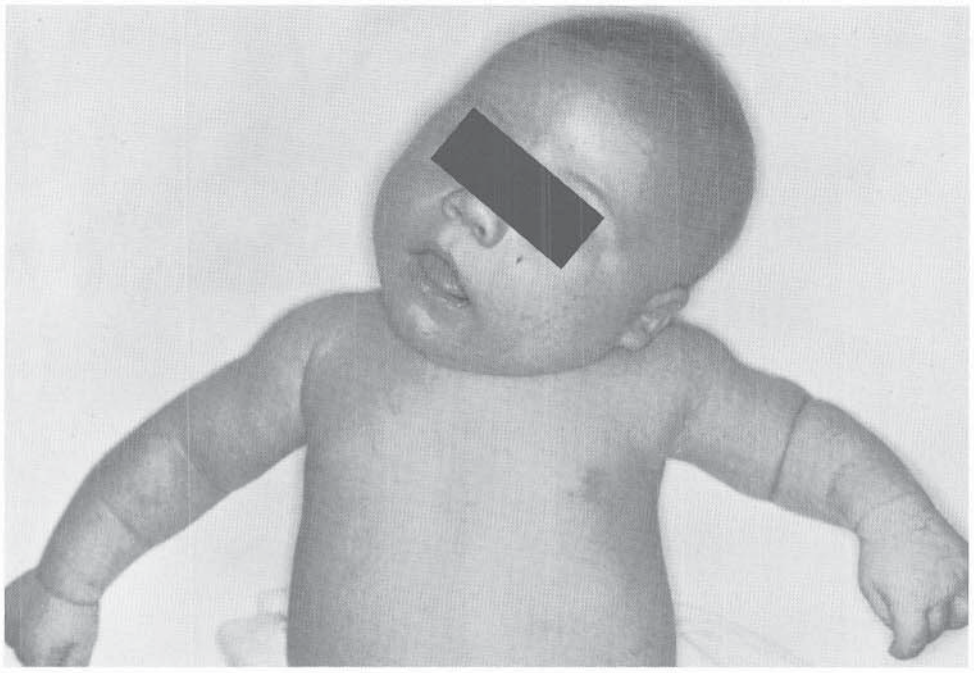
Early surgical treatment of congenital muscular torticollis is to be condemned. The natural history of the untreated deformity is benign, as more than 90 percent of patients eventually develop an adequate range of motion and an adequate cosmetic appearance. Less than 10 percent of cases come to surgery.[18] If a significant restriction of motion (lacking 30 degrees of full rotation or more) or facial asymmetry persists after the child achieves walking age, surgical intervention may be considered. However, there is little advantage and much disadvantage to surgical release in the young child, and we prefer to wait until just prior to school age before a decision on surgery is made.

The reasons for waiting are both technical and age-related. Operative procedures include subcutaneous tenotomy, open tenotomy of the lower SCM insertions, bipolar tenotomy, and excision of part or all of the muscle. Tenotomy or excision, while allowing an immediate increase in head excursion, are more likely to lead to muscle recontraction and a cosmetic deficit in the column of the neck due to the loss of muscle bulk. The earlier the surgery is performed, the more technically difficult is Z-plasty reconstruction of the muscle bulk (due to the diminutive size of the structures) and thus minimization of the cosmetic deficiency resulting from tenotomy or excision procedures.

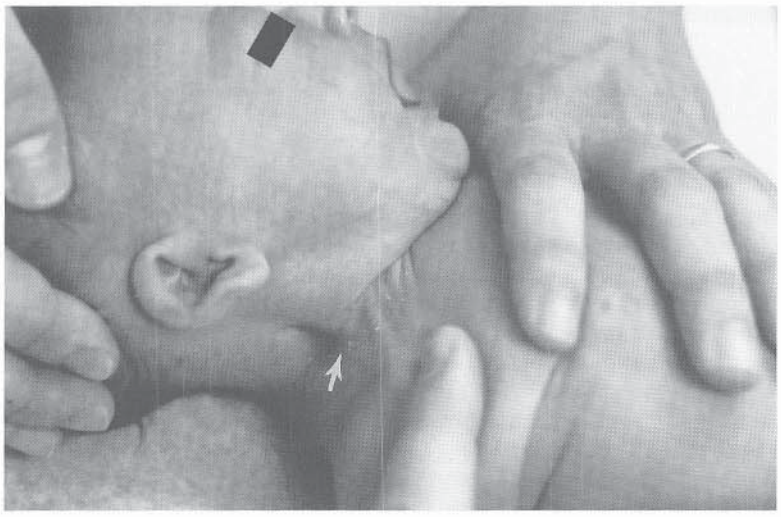
The complications of surgery in infancy are significant and include scar formation, recontracture with more severe fibrosis,[85] and, more important for this cosmetic deformity, an unacceptable cosmetic result due to removal of the SCM column of the neck line, which produces an unsightly "hole" at the distal insertion in the sternum and clavicle, reported in 40 to 90 percent of patients.[18,66,83,99] Because of the excellent results that can be obtained if surgery is delayed until an older age, there is simply no urgency for surgery in infancy for congenital muscular torticollis.

Most authors favor surgery, when indicated, up to 6 years of age.[66,85] Others have extended this upward to age 12[83] and beyond.[16,99] Functional outcome, as judged from range-of-motion evaluation, is not different for surgery performed between the ages of 1 and 6 years, and the disadvantages of early surgery (poor cosmesis, recurrence) decrease as the age of the child increases. Poorer results, primarily due to intractable facial asymmetry or some limitations of motion, are restricted to the most severe cases, although the benefits of later surgery in correcting head tilt and overall cosmesis are well established.[66,83,99,102]

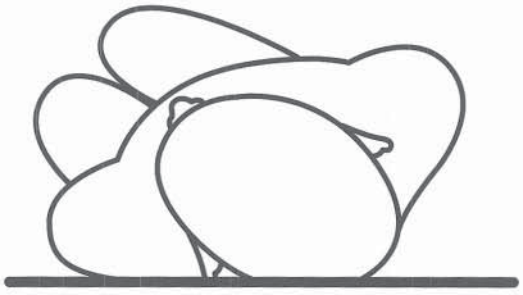
We prefer the bipolar lengthening technique of Ferkel and colleagues for patients needing surgery.[39] The release of the SCM muscle includes a careful reconstruction of the "column" of the SCM by either (1) performing Z-plasty of the clavicular insertion and releasing the sternal insertion from bone, or (2) transecting of the sternal portion of the muscle 1 to 2 cm proximal to its insertion, releasing the clavicular insertion from bone, and transferring the latter to the remaining distal sternal portion (Fig. 10–4). Such a Z-plasty reconstruction is technically difficult to perform in the infant or toddler, which explains why early release produces cosmetically unappealing results. Release at the mastoid process allows more vigorous and complete release of the patient's head, so that postoperative physical therapy can be more effective. The mastoid release should be performed at the bony insertion, to avoid possible injury to the spinal accessory nerve. Skin incisions should never be



A



B



C

FIGURE 10–1 **A,** Torticollis due to a contracted left sternocleidomastoid (SCM) muscle. **B,** Mass in right SCM (*arrow*) of a newborn. Note intrauterine folding deformity of the right ear. **C,** Flattening of the left occipital area and left ear deformation due to supine positioning of a child with right congenital torticollis.

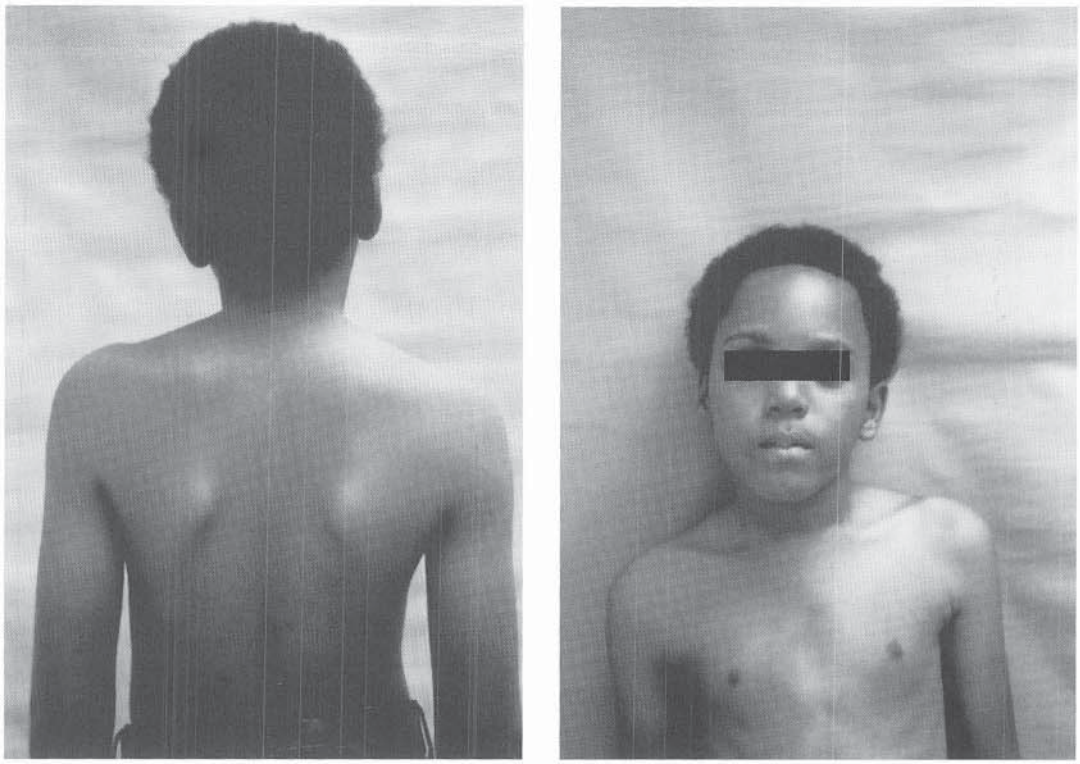
placed over the clavicle because of the unaesthetic scar spreading that will occur.

The results of surgical release in older children have also been satisfactory (Fig. 10–5). There is therefore little to criticize in benign neglect of the young child, with surgical release, if indicated, performed sometime between ages 5 and 12 years.

Postoperative care of the patient who has undergone bipolar release includes reinstitution of stretching exercises as soon as pain has abated and the surgical incisions have adequately healed.[18,66,99,102] Historically, postoperative treatment has included the use of all types of braces and cast correction,[138] but we have found that active range-of-motion exercises produce excellent results, and the use of postoperative immobilization is somewhat obsolete. Residual fascial bands[18,66,102] can lead to recurrence of deformity. These bands are best avoided by delaying the surgery until the recommended age of at least 5.

KLIPPEL-FEIL SYNDROME

A second form of congenital painless torticollis is associated with congenital osseous fusions (synostosis) and failure of segmentation of the cervical spine. Such fusions can involve the craniocervical junction (occiput-C2), the subaxial cervical spine, or both, and typically result in the appearance of a short, webbed neck combined with a low posterior hairline. An associated head tilt and loss of cervical motion complete



A B

FIGURE 10–2 **A** and **B,** Clinical appearance of a 10-year-old boy referred for presumed scoliosis because of left shoulder elevation. Contracture of the left SCM muscle (torticollis) produced the deformity.

the clinical triad commonly referred to as the Klippel-Feil syndrome (Fig. 10–6).[62,77,103] In practice, the term is used to describe any failure of segmentation in the cervical spine, and some series[54,62] report the full triad occurring in only half of patients with the diagnosis. The loss of motion, particularly rotation, associated with torticollis brings attention to the abnormality.

Frequently the neck webbing appears to produce the head tilt or deformity, thus making the torticollis appear secondary to the soft tissue abnormality. Simple failure of segmentation of the vertebral bodies or posterior elements may not produce a true head tilt or rotation, but there is frequently an asymmetry of fusions or an additional congenital unilateral fusion in the subaxial or cervicothoracic area that produces the torticollis. These fusions result from abnormal embryologic formation of the cervical vertebral mesenchymal anlagen. Not only does failure of normal segmentation of the cervical somites between the third and eighth week of gestation explain the cervical synostoses and anomalies but, owing to the scapular differentiation from mesenchymal tissue at the C3–4 level that occurs simultaneously, Sprengel's deformity, seen in up to 50 percent of Klippel-Feil patients, is also an expected anomaly accompanying the congenital fusions (Fig. 10–7).[1,62] The well-known omovertebral bone connecting the scapula and cervical spine in Sprengel's deformity is further evidence of a failure of segmentation underlying the entire process. The etiology of such failures of segmentation is believed to be either toxic or ischemic (anomalous vertebral artery development),[1] and because of the timing in embryologic development, the extent of the embryologic insult is also believed to result in abnormalities of other organ systems.

The identification of a family of genes (the homeobox or HOX family) that directs and regulates embryonic differentiation and segmentation along a craniocaudal axis has allowed speculation that mutations involving this family are responsible for the anomalies in humans, as is evident in murine homeobox genes and cervical abnormalities.[81,91,135] The development of the cervical region follows a pattern of craniocaudal resegmentation in which the eight pairs of embryonic cervical somites divide into a cranial and caudal segment.[81,112] These primitive mesenchymal segments separate, with each vertebral anlage then being formed by the caudal half-sclerotome of one somite and the cranial half of the next lower one. Variations at the occipitocervical junction occur: the cranial half of the first sclerotome remains as a half-segment (proatlas) between the occiput and the atlas proper, eventually becoming part of the occipital condyles and the tip of the odontoid. The atlas receives contributions from the lower occipital and first cervical somite (two posterior arches). The axis receives contributions from the primitive second cervical somite (posterior arches), the cranial half of the first (tip of the odontoid), and the primitive centrum of the second (atlas), which becomes the body of the odontoid.[110] Each segment from C3 to C7 is made up of a centrum (body) and two posterior arches that form from mesenchymal tissue migrating around each side of the neural tube. In light of this complex origin of the cartilaginous anlagen of the cervical vertebra, it is not difficult to understand how anomalous development in the form

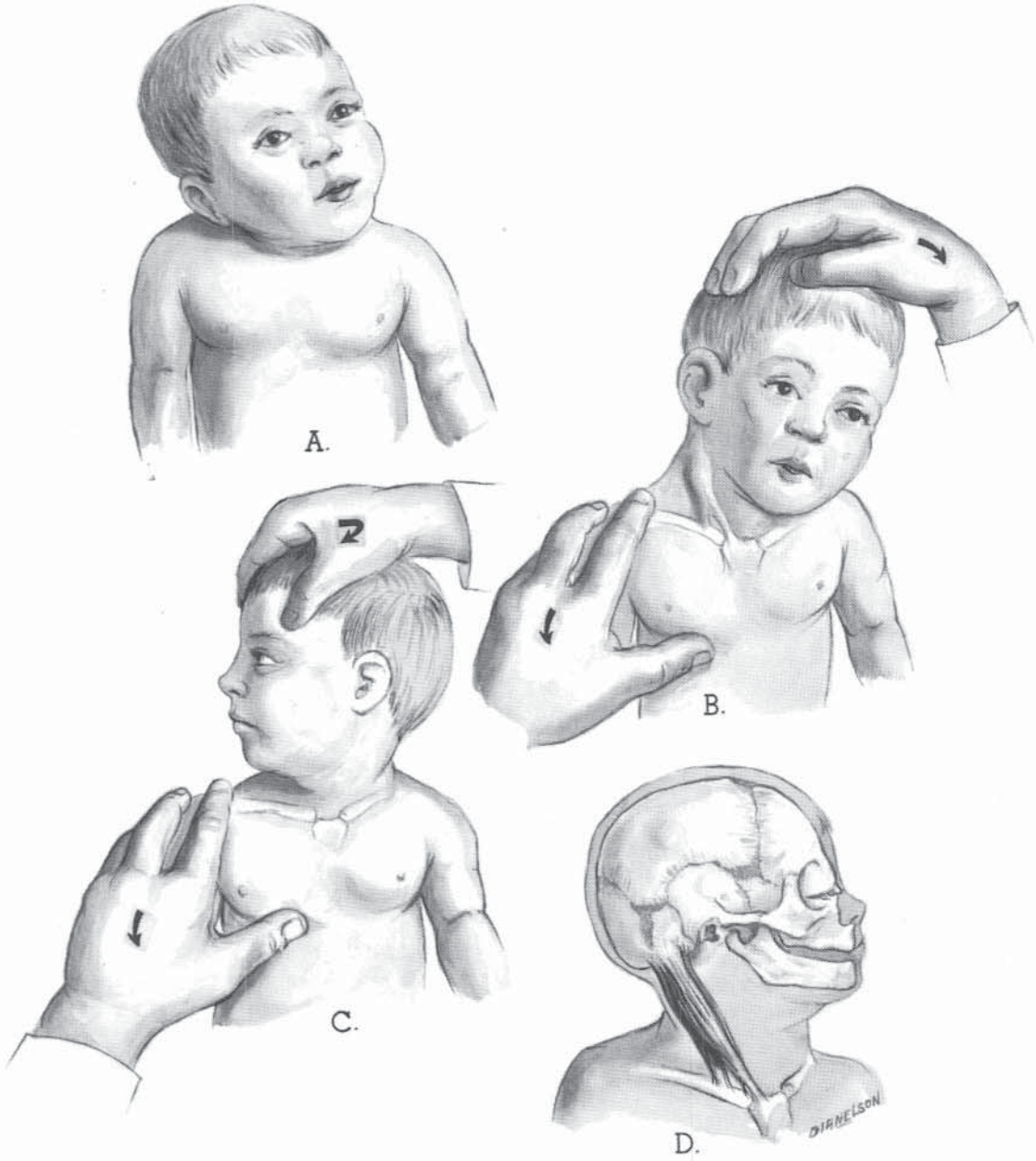


FIGURE 10–3 Passive stretching exercises for a right SCM contracture. **A,** The deformity. **B,** The head is bent laterally so that the ear of the left side touches the left shoulder. Note the tumor-like bulge in the SCM muscle. **C,** The head is rotated to the right so that the chin approaches the right shoulder. **D,** Anatomy of the SCM muscle.

of failure of segmentation and partial absences can occur, producing the deformities seen in Klippel-Feil syndrome.

As mentioned, other anomalies also appear, owing to the global nature and timing of the postulated fetal insult. Genitourinary anomalies are estimated to occur in 25 to 35 percent of children with Klippel-syndrome,[1,97,101] congenital heart disease in 14 to 29 percent,[1,109] deafness in 15 to 35 percent,[95,133] and synkinesis or mirror movements in 15 to 20 percent.[1,57]

Scoliosis, either congenital or idiopathic-like, occurs in 60 percent of Klippel-Feil patients,[1] and it is the congenital fusions involving the cervical and cervicothoracic junction that are most troublesome in producing deformity. Rib anomalies often accompany both congenital fusions and Sprengel's deformity (see Fig. 10–7). Syndromes producing all of the above anomalies include the VATER association, Goldenhar's syndrome,[124] and the fetal alcohol syndrome.

Clinical Features. The newborn or young infant with the classic triad of a low hairline, webbed neck, and limited motion with or without torticollis presents no problem in diagnosis (see Fig. 10–6). Patients with less obvious signs of classic Klippel-Feil anomalies are generally diagnosed on the basis of the restricted motion associated with vertebral fusions. The finding of an abnormal head position, true torticollis, and restricted range of motion, without an obvious SCM contracture, should prompt obtaining radiographs of the cervical spine. Screening for other vertebral anomalies is appropriate if any cervical fusions are found.

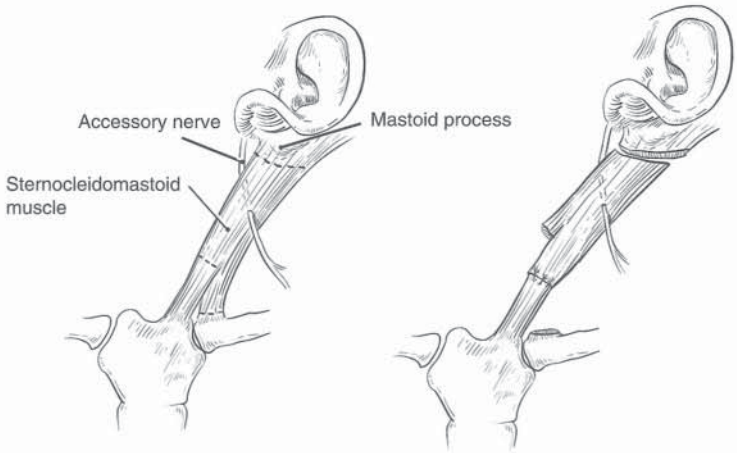


FIGURE 10–4 Bipolar lengthening of the SCM muscle. (Modified from Ferkel RD, Westin GW, Dawson EG, et al: Muscular torticollis: a modified surgical approach. J Bone Joint Surg 1983;65-A:894.)

Once vertebral fusions in the cervical spine are documented, a general pediatric evaluation should be undertaken to rule out congenital cardiac or other neurologic abnormalities. Renal ultrasonography is an appropriate screening test to diagnose genitourinary anomalies.[33] Magnetic resonance imaging (MRI) of the cervical cord and craniocervical junction is recommended whenever any orthopaedic procedure is contemplated, and certainly is indicated for evaluation of symptoms related to cord compression/stenosis or instability.

Patients with Klippel-Feil anomalies present at a young age for evaluation of deformity, which is managed in a fashion similar to the management of congenital scoliosis. Because there is no room in the cervical spine for a compensatory curve to develop to keep the head upright and compensated, any progression of a cervical scoliosis as the cause of a patient's head tilt and torticollis must be aggressively treated so that uncorrectable head tilt or a severe compensatory scoliosis that decompensates the trunk does not result from a progressive congenital deformity (Fig. 10–8).

Patients with Klippel-Feil anomalies may also present at an older age with pain, radiculopathy, or myelopathy due to cord compression in a congenitally anomalous, narrow canal or due to instability or hypermobility at unfused levels.[122,128] Torticollis may or may not be present, and may be recently *acquired* at the time of symptom appearance where it was previously absent. Patients with extensive vertebral fusions, often extending up to C3, may also have occipital-atlantal fusion, producing hypermobility at an unfused C1–2 or C2–3 level (see Fig. 10–7). Any unfused segment adjacent to an extensive synostosis may eventually become hypermobile, with or without neurologic symptoms.[58,82,124] Thus an adolescent with mild nonprogressive deformity may develop symptomatic hypermobility after years of being asymptomatic, although almost never before age 13.[124] Degenerative changes at the hypermobile segments may produce just enough cord or nerve root impingement in a young adult to produce radiculopathy and myelopathy. Degenerative stenosis without hypermobility may result in subaxial cervical segments when osteophytes and disk degeneration progress in adult life.

Depending on the site and type of stenosis (anterior or posterior) and anatomic level, motor and sensory deficits and reflex changes may occur, as well as paresthesias in the occiput, neck, and upper extremities. If the cerebellar tonsils are compressed or herniated (Arnold-Chiari malformation), neurologic findings may include ataxia, dizziness, and nystagmus. Cranial nerve changes from brain stem compression (difficulty swallowing, disturbed phonation) or hydrocephalus from obstruction of cerebrospinal fluid (CSF) flow (blurred vision, headache) by invagination of the odontoid into the foramen magnum (basilar impression) can be observed. Less commonly, vertebral artery involvement can produce syncope, seizures, or ataxia due to brain stem ischemia. Any of these varied neurologic signs and symptoms must be investigated in a patient with known cervical anomalies.

Radiographic Features. Imaging studies of the cervical spine, especially of the craniocervical junction, are crucial in the management of patients with Klippel-Feil anomalies. Besides defining the often bizarre, mixed anomalies, neuroradiologic evaluation is mandatory in patients who develop neurologic compromise. Positioning for imaging studies may be problematic because of the shortened neck and relative lack of motion. Overlapping shadows from the mandible and occiput can confound interpretation of plain radiographs. A lateral radiograph of the skull, rather than of the cervical spine itself, will best demonstrate the presence of occipitocervical bony abnormalities by eliminating some of the obliquity and rotational overlapping seen with torticollis.[146] If C1 has been assimilated into the occiput, the lateral skull film is also helpful in determining whether there is C1–2 pathology. Once anomalous osseous structures are visualized on a screening radiograph, further studies by computed tomography (CT) with or without three-dimensional reconstruction and MRI to evaluate the brain stem and cervical cord are recommended.

Besides symptoms of instability, patients with Klippel-Feil anomalies and neurologic symptoms must also be evaluated for basilar impression. A good-quality lateral radiograph will show the upward migration of atlantoaxial structures, particularly the odontoid, into the foramen magnum, and knowledge of the traditional radiographic lines (Chamberlain's, McRae's, and McGregor's) is useful in screening for the presence of basilar impression. McGregor's line, drawn from the upper surface of the posterior edge of the hard palate to the most caudal point of the occiput, is the

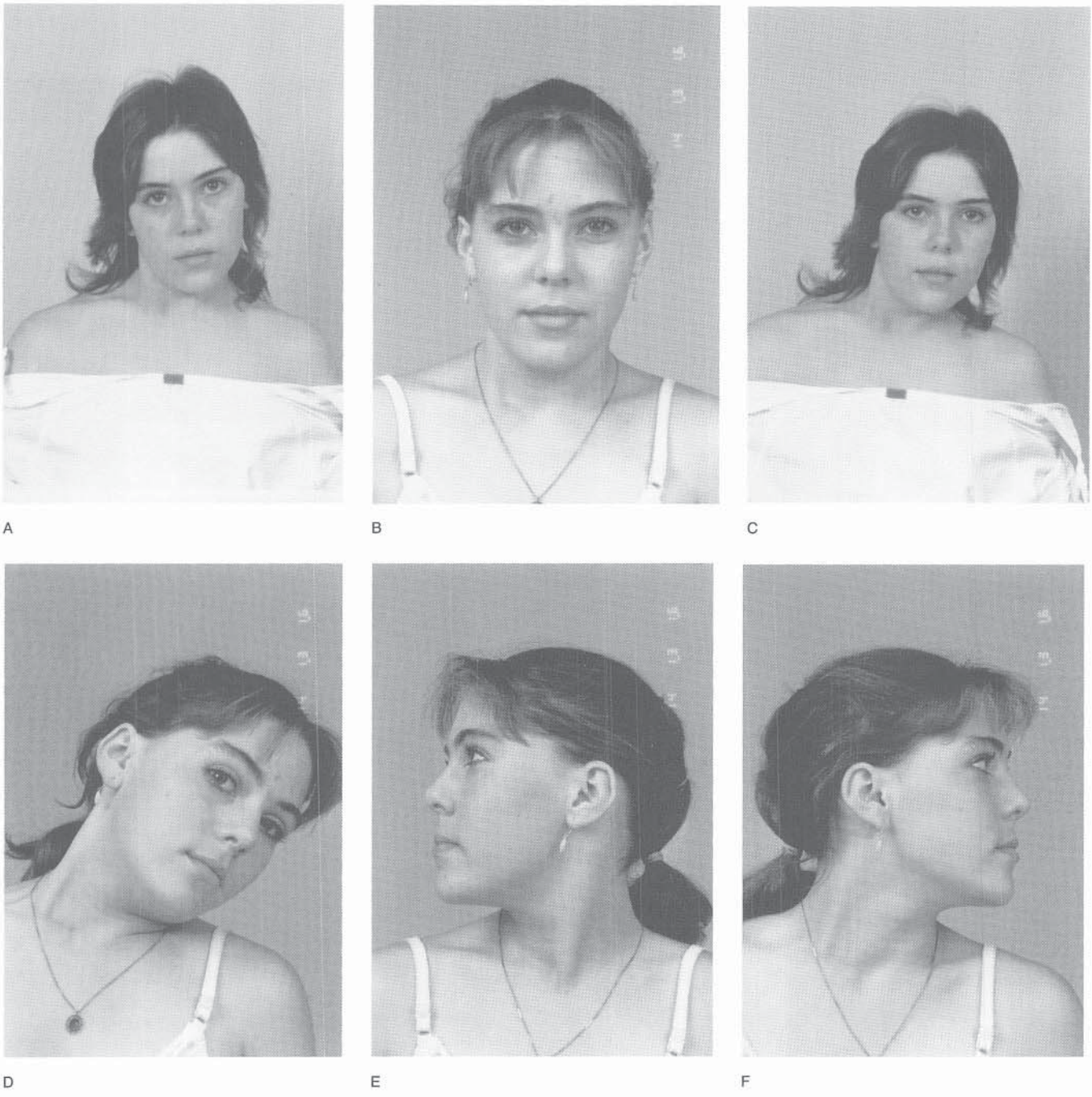


FIGURE 10–5 Torticollis release on the right in a 13-year-old child. **A** and **B,** Preoperative and postoperative appearance. **C** and **D,** Limitation on attempted tilting of the head to the left preoperatively, and improvement postoperatively. **E** and **F,** Full rotation to both sides postoperatively. Note the cosmetic maintenance of the anterior column of the neck provided by the lengthened SCM muscle, and the cosmesis of the mastoid incision.

best screening line because of the reproducibility and clarity of these radiographic landmarks (Fig. 10–9). McRae's line defines the opening of the foramen magnum and truly defines basilar impression, because the odontoid projects above this line in patients who are symptomatic.[32,87,96] Modern imaging studies, such as CT with sagittal or three-dimensional reconstruction, show the osseous relationships more clearly. If there is any question of neural impingement, MRI is the more revealing study.

Equally important is the determination of impending stenosis or cord impingement by evaluating the space available for the cord (SAC) and its corollary measurement at C1–2, the atlantis-dens interval (ADI) (see Figs. 10–7 and 10–9). These intervals are usually determined on lateral flexion-extension radiographs, generally obtained with the patient awake and voluntarily flexing the head. The SAC is measured as the distance between the posterior edge of the dens and the anterior edge of the posterior ring of the atlas

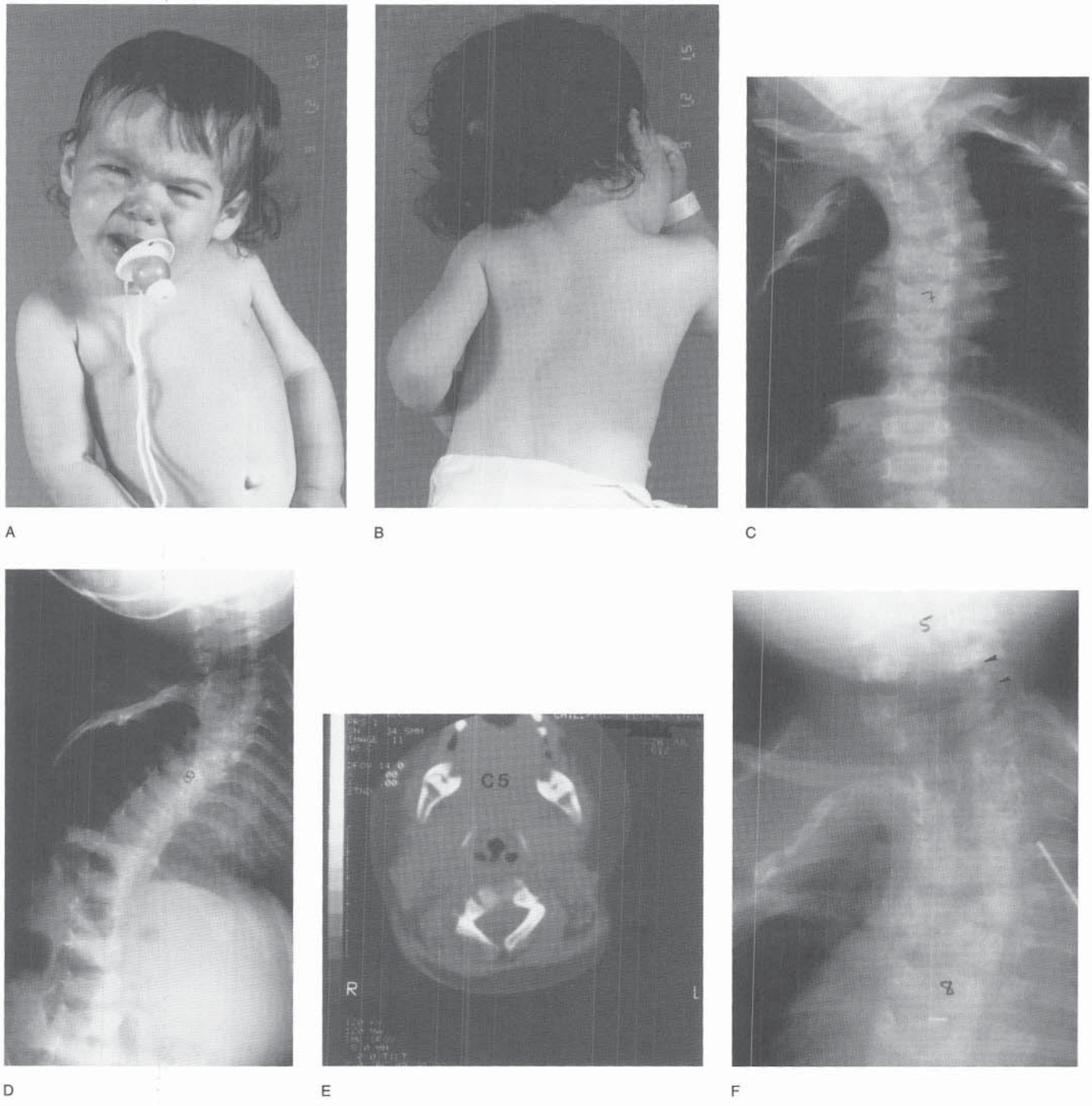
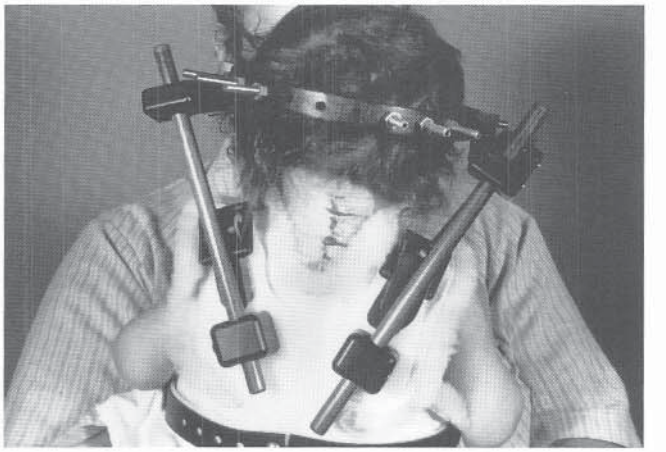


FIGURE 10–6 Klippel-Feil syndrome with torticollis. **A** and **B,** Clinical appearance of a 13-month-old toddler. Note the fixed head tilt, low hairline, and plagiocephaly. **C,** Initial radiograph obtained at age 5 months showing right convex scoliosis due to multiple congenital anomalies. **D,** Radiograph obtained at age 13 months showing progression of the deformity. A significant compensatory thoracolumbar curve has developed. **E,** The rotational deformity extended to C5. **F,** Anterior fusion was extended via a thoracotomy to the C6–7 disk through the chest (*arrows* point to the convex bone graft). Posterior fusion was extended to C5–T8.

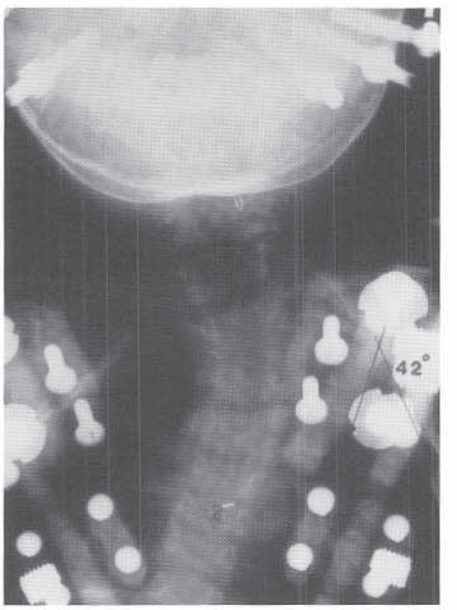
or the foramen magnum. An SAC of 13 mm or less is associated with neurologic compromise.[131] In patients with hypermobility, as suggested by an ADI of more than 5 mm between flexion and extension, measurement of the SAC gives a reasonable evaluation of how tenuous the neurologic situation may be. Up to 5 mm of motion at the ADI is considered normal in children.[86,114]

Normal range of motion at the occipital-atlantal joint is undefined. The occiput-C1 articulations are primarily saddle-shaped, elliptical surfaces that allow flexion-extension but little rotation or lateral flexion. Instability of this joint, being much less common than instability of C1–2, is not well described. Posterior subluxation of the occipital-atlantal joint in extension of more than 4 mm

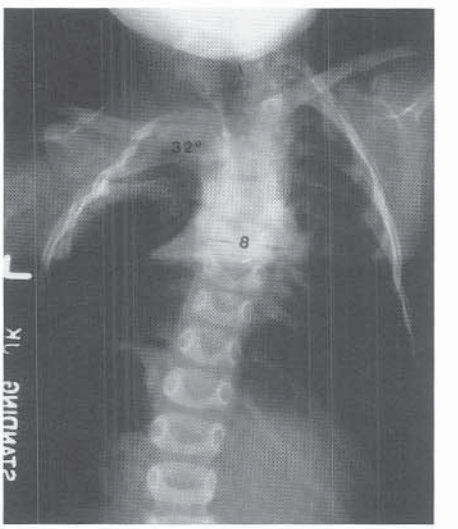


G

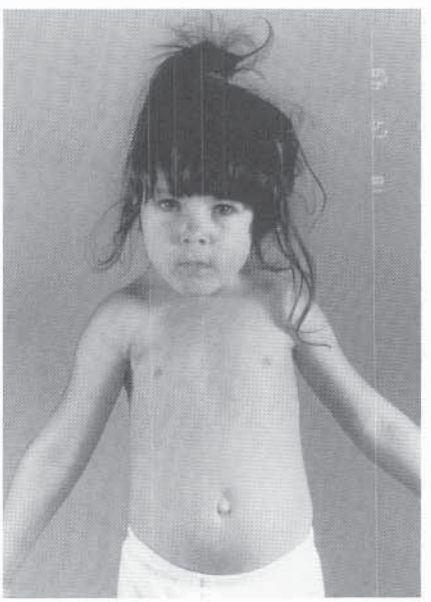
FIGURE 10–6 *Continued.* **G** and **H,** Postoperative immobilization in a halo-vest device. Head tilt and rotation were corrected. **I,** Radiograph obtained 2 years postoperatively showing stabilization of the deformity. **J** and **K,** Subsequent clinical appearance. There is no recurrence of the head tilt. **L,** Radiograph obtained 9 years postoperatively. Correction has been maintained by convex hemifusion anteriorly.



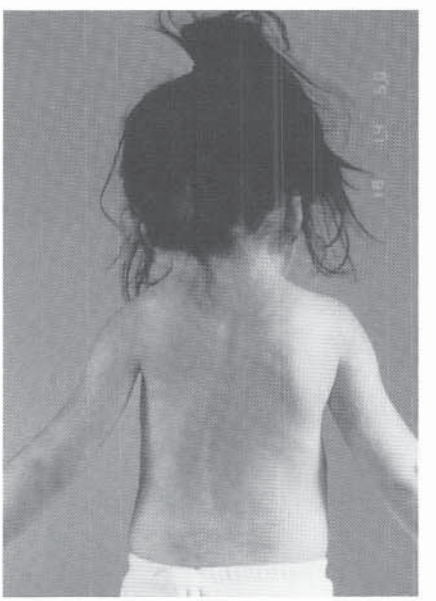
H



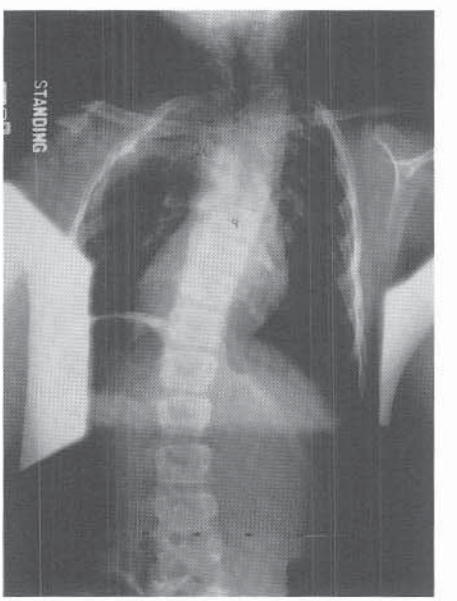
I



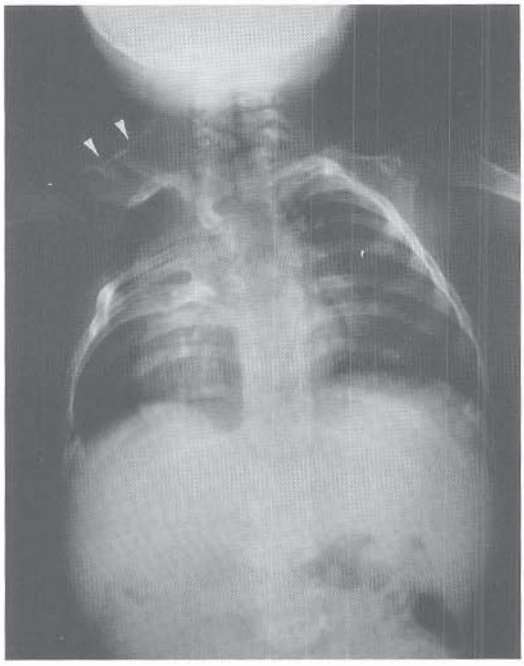
J



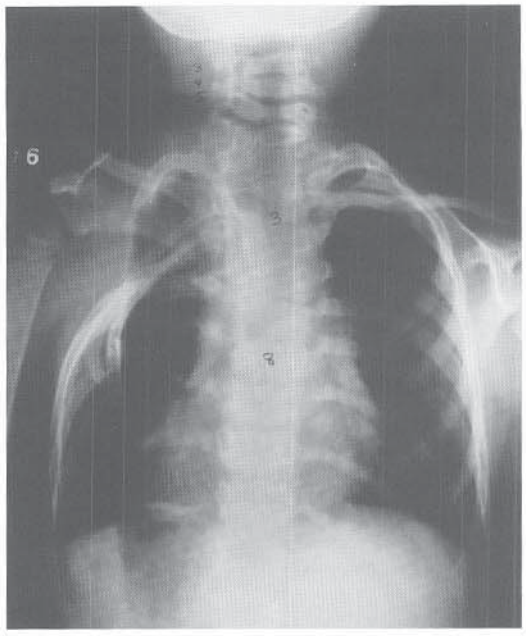
K



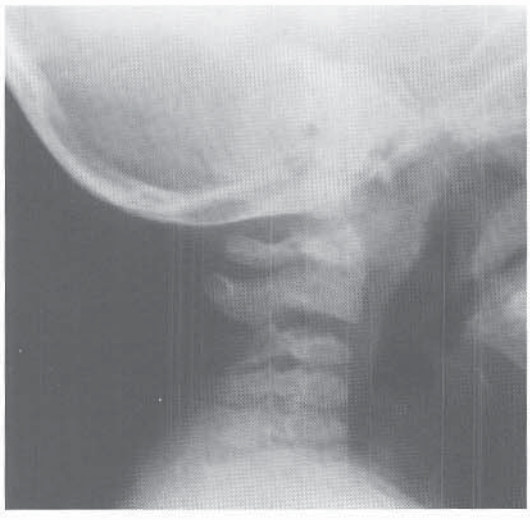
L



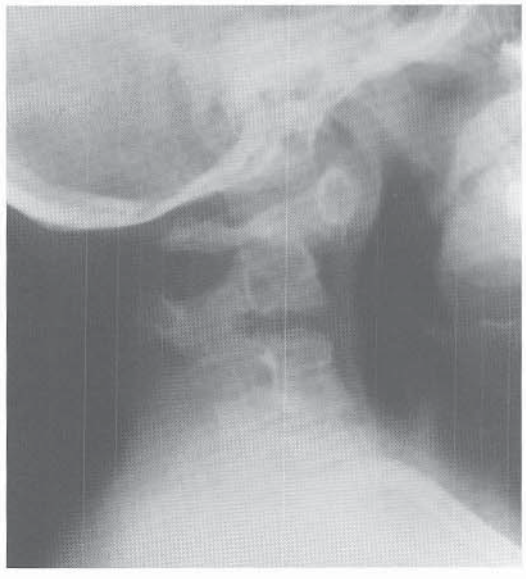
A



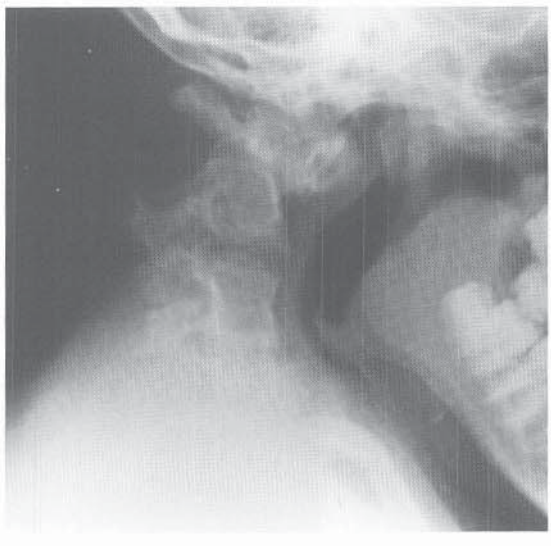
B



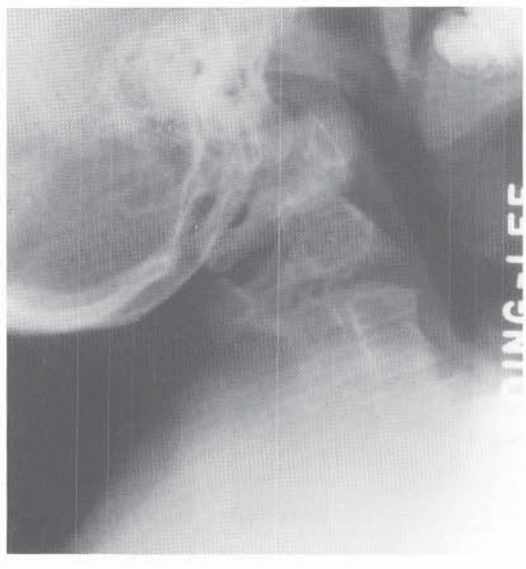
C



D

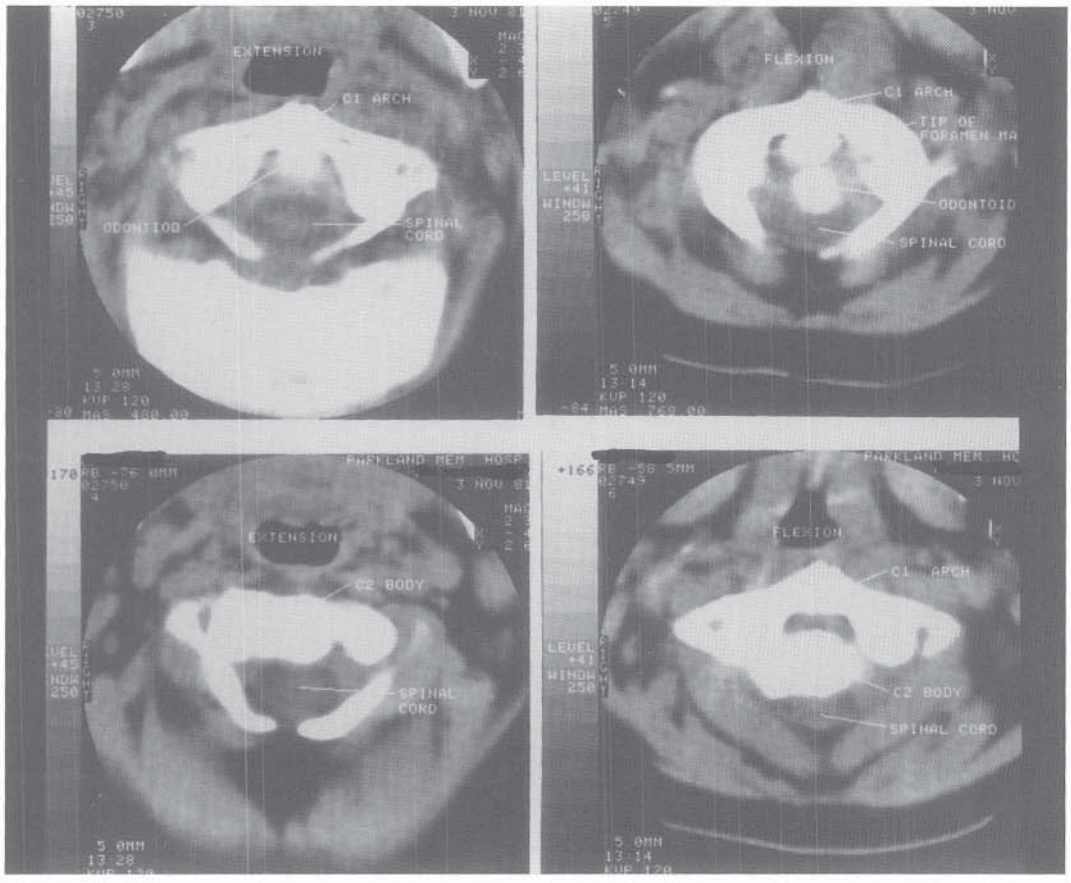


E

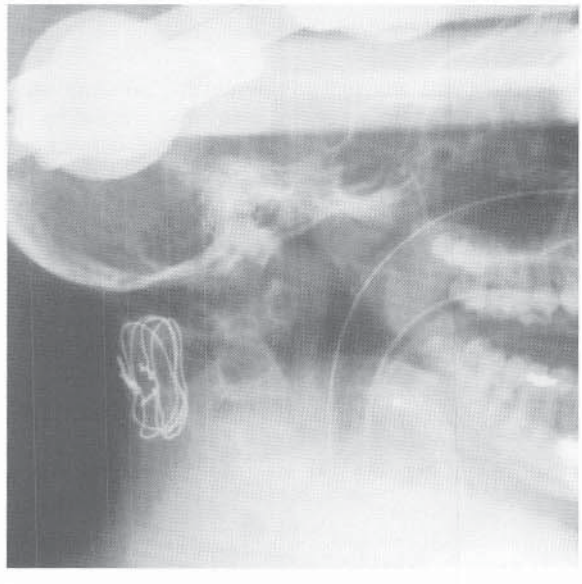


F

FIGURE 10–7 *See legend on opposite page*



G

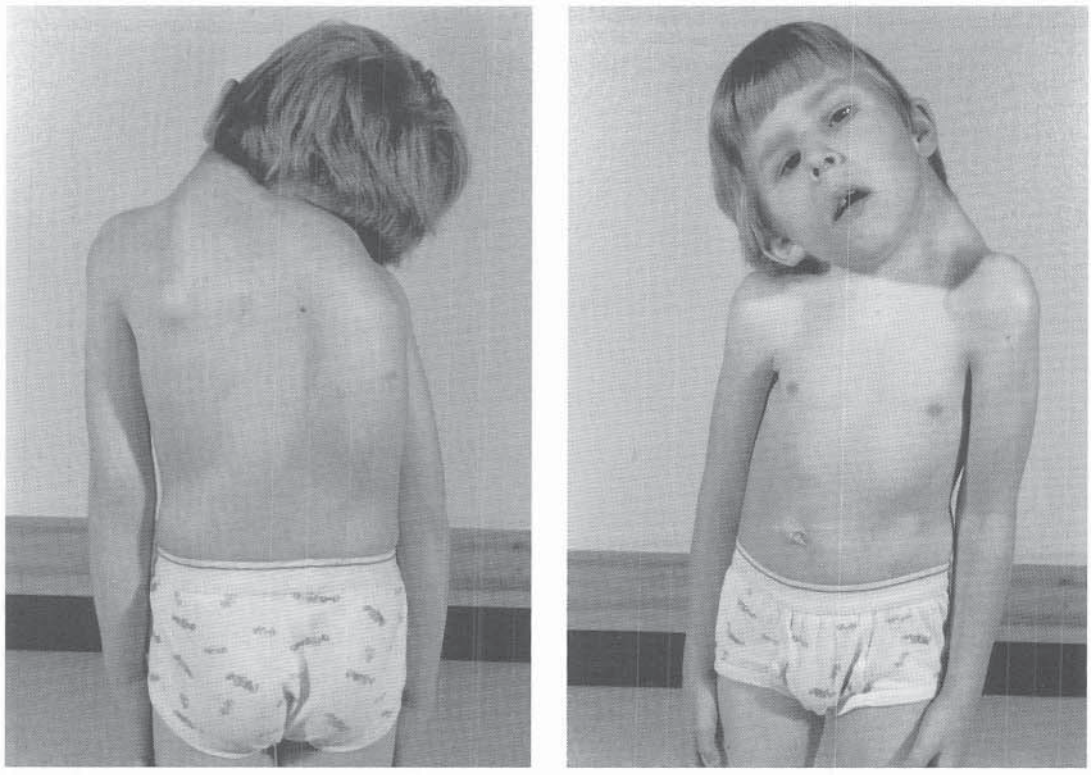


H

FIGURE 10–7 Klippel-Feil syndrome. **A,** AP spine radiograph of a 4-year-old boy with Klippel-Feil syndrome. Multiple congenital spinal anomalies and rib fusions are evident. A Sprengel's deformity is apparent on the left (*arrows*). **B,** Radiograph obtained 6 years later shows little change in the thoracic spine but a slight curve developing in the cervical spine owing to an unsegmented bar between C3 and C5. **C,** Lateral cervical radiograph obtained at age 2 years. C3–5 synostosis is suggested. The upper cervical spine appears normal. **D,** Radiograph obtained at age 6. The ADI at C1–2 is increasing. C2–3 now has a posterior synostosis. The patient is asymptomatic. **E** and **F,** Radiographs obtained at age 12. Flexion instability at C1–2, with ADI of 10 mm and a decrease in the SAC, is obvious, with a reduction in extension. The patient experienced neck pain and had several episodes of acute mild torticollis but was neurologically intact. There is minimal motion at C2–3 and complete synostosis below C3. **G,** CT scan in flexion showed significant impingement on the cord anteriorly by the odontoid. **H,** Despite a partially open ring of C1, the patient was able to undergo Brooks fusion at C1–2, to prevent further cord compromise. He is currently asymptomatic but has no cervical motion at all.

suggests instability.[139] This can be measured from the excursion of the basion or occipital condyles in relation to a fixed point, usually the posterior edge of the anterior ring of C1 during flexion-extension (Fig. 10–10).[147] Power's ratio identifies anterior occiput-C1 instability, but because most instabilities are more obvious in extension, this ratio may not be as useful. Normal occiput-C1 translation should be no more than 1 mm in adults,[147] and thus the importance of measuring the SAC on either plain radiographs or flexion-extension CT or MR images may be more critical at the occipital-atlantal joint.

Treatment. Treatment of Klippel-Feil syndrome and other synostotic anomalies must take into consideration both the



A B

FIGURE 10–8 **A** and **B,** Severe torticollis due to cervical dysraphism. The left cervicothoracic deformity produces uncompensated head tilt to the right, as there is no room in the cervical spine for a compensatory curve. A right lower thoracic scoliosis is developing to push up the right shoulder and rotate the head and neck counterclockwise as a compensatory mechanism. Ultimately this patient's compensatory scoliosis became untreatable because correction produced intolerable worsening of the fixed head tilt.

deformity and any neurologic deficit, potential or already present. Management of the deformity is considerably simplified if no neurologic deficit is present. The treatment of occipitocervical junctional abnormalities in the presence of neurologic deficit is fraught with the potential for significant morbidity and even mortality, owing to the proximity of the cervical cord and brain stem. In such a situation, the combined efforts of both orthopaedists and neurosurgeons may be required.

Deformity management involving head tilt or rotation

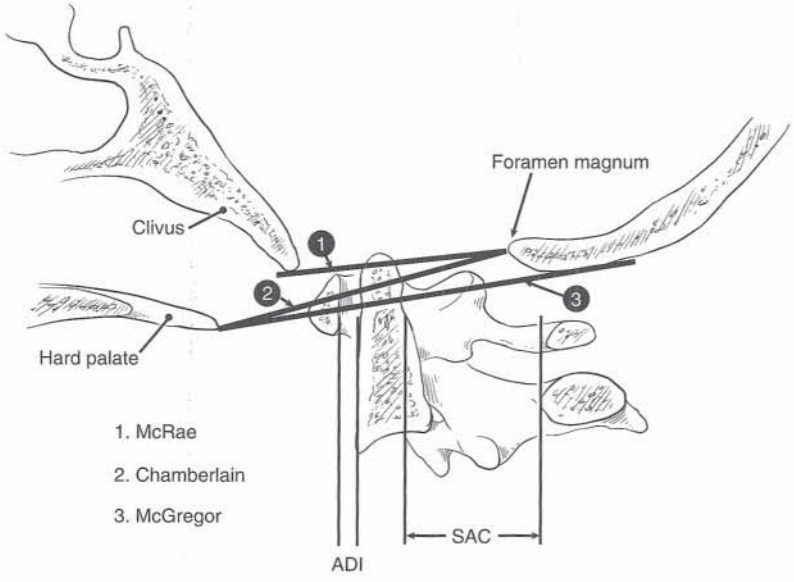
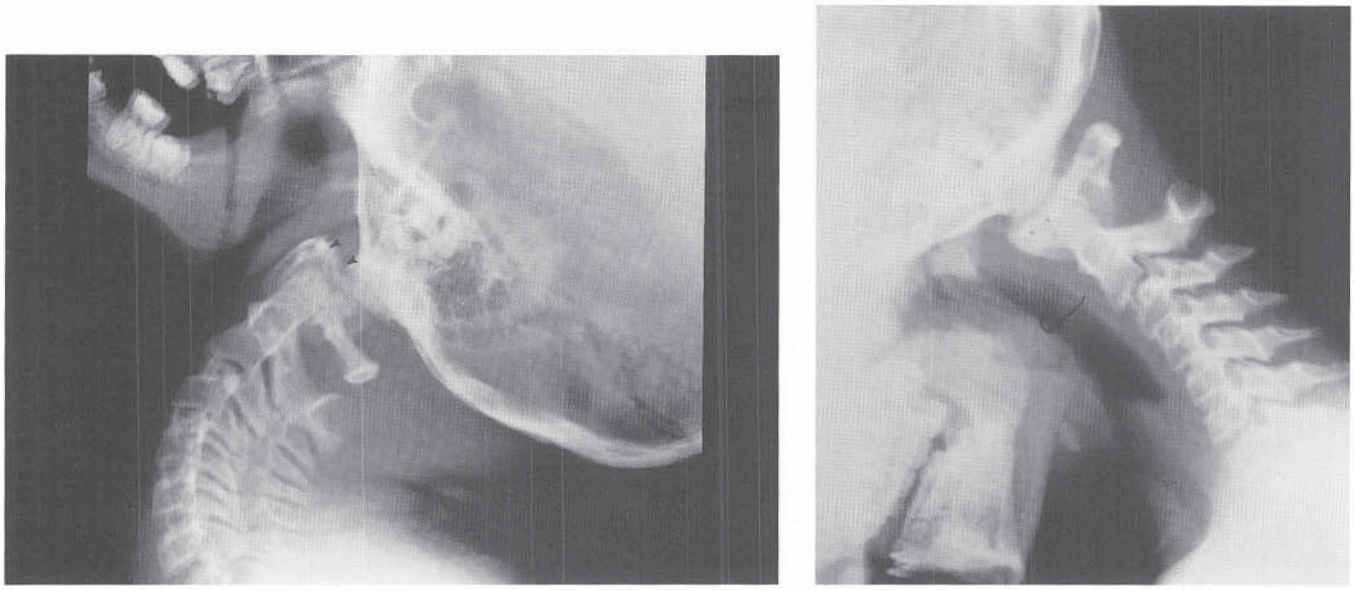


FIGURE 10–9 McRae's, Chamberlain's, and MacGregor's lines define basilar impressions on a lateral radiograph of the skull. C1–2 instability is determined by the space available for the cord (SAC) and the atlanto-dens interval (ADI).



A B

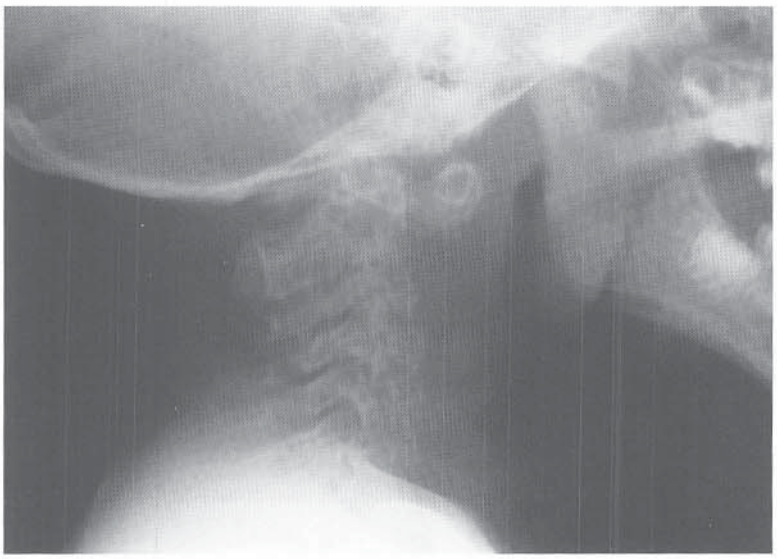
FIGURE 10–10 **A** and **B,** Occipital-atlantal instability in a patient with Down syndrome. There is 5 mm of posterior translation of the occipital condyles in extension (*arrows*), which reduces in flexion. However, in flexion the ADI is 6 mm, reducing to 1 mm in extension.

frequently requires the use of a halo to obtain and maintain correction. The halo is the one device that allows simultaneous correction and repositioning of the skull and upper cervical spine, and then provides the external immobilization necessary to protect a decompression and achieve spinal fusion. The device also has the advantage of avoiding skin complications around the mandible or occiput, the bane of most occipital-mandibular devices (Philadelphia collar, four-poster brace) when applied to children (see Fig. 10–6). Access to cervical incisions and freedom of the mandible for eating are important advantages, and because a halo-vest device usually does not need to be removed or adjusted once it has been applied, early mobilization of the patient is improved.

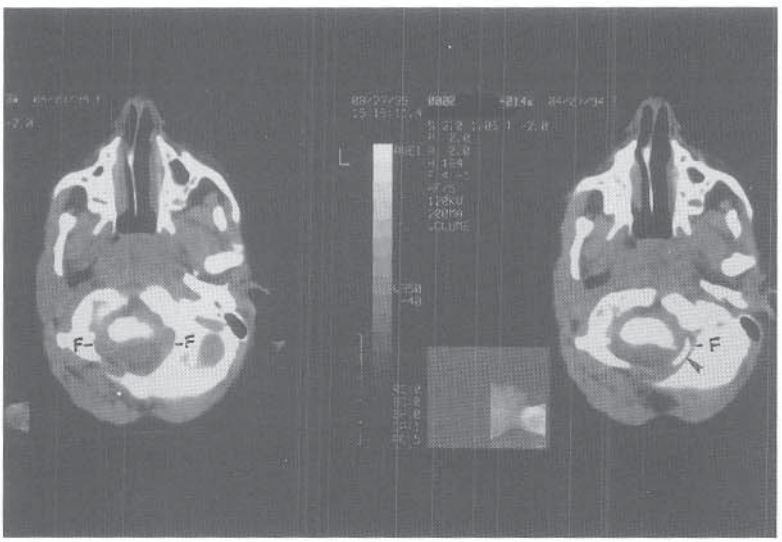
Reluctance to use halo fixation in young children stems from fear that the pins might penetrate the cranial vault, resulting in epidural abscess or osteomyelitis. In fact, loosening is a far more common problem.[6,30] Because of variation in skull thickness and suture formation, CT of the skull has been recommended,[84,151] but in practice we have not found that CT findings significantly alter intended pin placement. For immobilization of young children, halo pins are placed in areas of the skull that are palpated to have appropriate bone stock. These areas include the frontal areas near the anterior hairline (avoiding the temporal artery) and the posterior parietal and occipital areas. A decreased insertional torque is used, depending on the child's age, with a general rule of 1 pound of torque per year of age up to age 8.[32,105] We use every possible hole in the halo ring so that a maximum number of pins are placed, with the goal of at least six pins used in patients less than 6 years old. Placing pins as perpendicular to the skull as possible also increases pin stability.[19] With such a regimen, pin loosening and infection are manageable, with minimal morbidity. The major contraindication to the use of halo fixation in infants and young children is the presence of abnormally wide sutures or fontanelles, which allows the bones of the skull to move away from the tips of the pins during insertion, resulting in loss of fixation. The presence of a significant metabolic bone disease, such as renal osteodystrophy or osteogenesis imperfecta, is a relative contraindication to use of the halo device. A basilar impression may be secondary to the pathologic bone. Longitudinal traction via the halo device in an attempt to reduce such a basilar impression may not be possible with porotic or dysplastic skulls.

One indication for surgical treatment in patients with Klippel-Feil or other cervical anomalies is progression of head tilt or rotation that is not passively correctable by positioning. If the anomalies producing deformity (hemivertebra, unsegmented bar) are in the subaxial cervical spine or cervicothoracic junction, the patient will develop a head tilt owing to an insufficient number of cervical vertebrae cephalad to the deformity to develop a compensatory curve (see Fig. 10–8). Increasing rigidity and uncorrectability of the head tilt is a crucial indication of the need for surgical treatment, and the fusion should include all vertebrae that are involved in the primary curvature. The halo is used to maintain the correction of the head tilt during the healing of the fusion (see Fig. 10–6). Depending on the patient's age, both anterior and posterior fusion (the former to eliminate a possible crankshaft phenomenon) may be necessary to eliminate further growth of the anomalous vertebra. Because fusion to halt progression is almost always undertaken in young children who are developing a fixed and uncorrectable deformity, an anterior-posterior arthrodesis will be indicated in most instances.

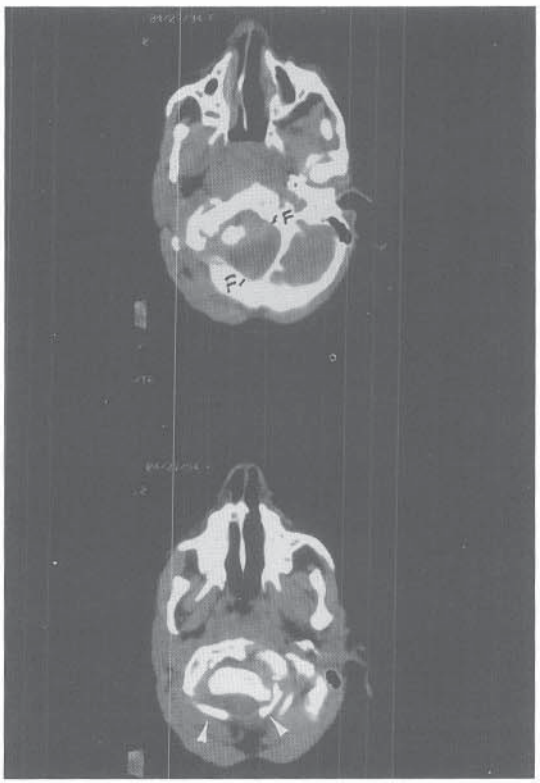
In the case of torticollis produced by upper cervical anomalies, posterior fusion alone, in association with halo correction of the deformity, is generally sufficient. In congenital unilateral absence of C1 (hemiatlas),[34] the deformity is present at birth and often progresses, and posterior fusion from the occiput to C2 is recommended between the ages



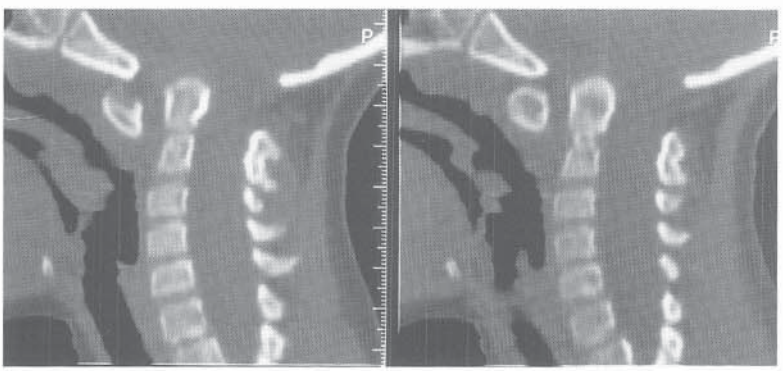
A



B

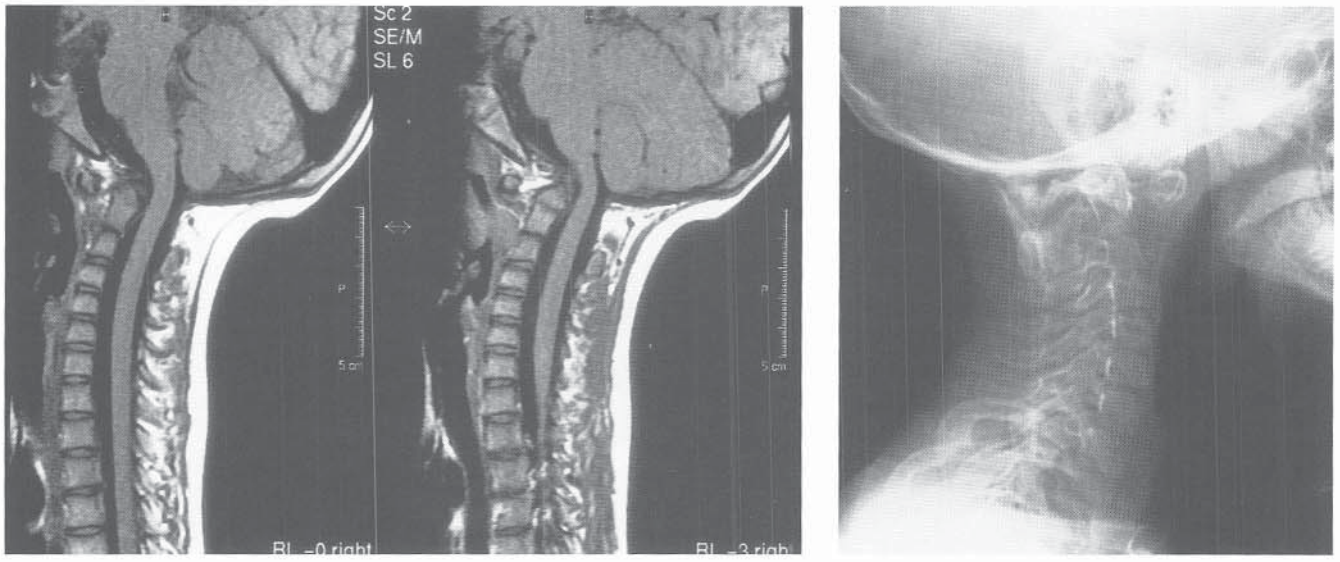


C



D

FIGURE 10–11 **A,** Occiput-C1 assimilation in a 5-year-old child with autism. There is a fixed C1–2 subluxation with an ADI of 7 mm. No neurologic deficit can be demonstrated except possible ataxia. **B** and **C,** Axial CT sections demonstrating an incomplete posterior ring of C1 and odontoid protrusion into the foramen magnum (*F*). The odontoid is seen on sections well cephalad, into the occiput. The ring of C1 (*arrows*) is invaginated up into the foramen magnum. **D,** Sagittal reconstruction confirming basilar impression.



E F

FIGURE 10–11 *Continued.* **E,** Anterior impingement of the lower brain stem due to basilar impression. **F,** Radiographic appearance 4 months after a posterior occiput-C3 fusion to prevent neurologic deficit.

of 5 and 8. Because of the limited amount of growth of the upper cervical vertebrae, recurrence due to the crankshaft phenomenon does not appear possible.

In patients with *congenital occipitocervical fusion* (synostosis), deformity (head tilt, torticollis) may be the only sign of neurologic compromise due to irritation of neural tissue, usually from either C1–2 instability or basilar impression. The occipital-atlantal assimilation may be a relatively isolated finding (Fig. 10–11) or may be part of a wide spectrum of congenital synostoses in a patient with obvious Klippel-Feil anomalies. In other situations, neck pain and frank neurologic deficit may develop as a result of encroachment of the odontoid into the foramen magnum or instability at the C1–2 articulation (see Fig. 10–7). Arnold-Chiari type I malformation is also associated.[14,26,32]

Treatment for neurologic compromise, either irritative (pain, deformity) or a frank deficit, invariably involves extension of the occipital-atlantal fusion to include the axis or perhaps C3, depending on whether or not the decompression is necessary. Transoral resection of the odontoid is the logical choice for anterior cord or brain stem impingement, while posterior craniectomy/C1 laminectomy are logical for posterior compression associated with anterior C1–2 instability.[14,93,143]

TECHNIQUES FOR UPPER CERVICAL FUSION. As delineated in the previous sections, occipitocervical arthrodesis is required to correct deformity or instability involving the craniocervical junction. Because of the significant forces producing movement between the skull and cervical spine, fixation of an upper cervical-to-occiput fusion is crucial to achieve arthrodesis. Immobilization with a halo device is strongly recommended, but additional internal fixation and specific bone grafting techniques are important to achieve the highest rate of fusion.[31,123] Bur holes can be placed near the foramen magnum, so that wire or heavy suture (in the case of young infants or children) can be passed either from bur hole to bur hole or from bur hole through the foramen magnum, and then fixed distally to the desired cervical level.[31] Equally important is the creation of a shaped corticocancellous bone graft, either one-piece or bilateral, that can be compressed against decorticated occipital bone and the cervical laminae (see Fig. 10–11). This graft is preferentially obtained from the posterior iliac crest. Alternatively, ribs can be harvested in young children. The graft is shaped to lie against the decorticated occipital surface, and in the case of a suboccipital craniectomy for decompression, it may be more convenient to cut the graft into two separate rectangular pieces to be placed on each side of the foramen magnum. The graft is fixed to the occipital bone by tightening wires over it, or a small hole may be placed in the graft so that wire or suture exiting a bur hole can be placed through the graft for fixation (Fig. 10–12). Caudally, the graft is compressed to the cervical laminae, either by passing wire sublaminarly (under the ring of C2, for example) or through the spinous process, and twisting or tying the wire or suture to fix the graft in position. The use of a threaded K-wire transversely through the base of the spinous process[27,123] adds additional stability to the caudal fixation for a wire or suture (see Fig. 10–12). Such an approach has resulted in a high rate of fusion.[31,32,123]

For atlantoaxial arthrodesis in children with C1–2 instability, standard techniques such as those of Gallie, Brooks, or Mageryl (Fig. 10–13) can be utilized, depending on the experience of the surgeon and the bony anatomy available. Transarticular screw fixation is biomechanically the stiffest of these methods and is recommended whenever possible because of the enhanced stability it procures in the setting of C1–2 instability.[100] For patients with incomplete posterior elements who are too small for transarticular screws, arthrodesis in situ with halo immobilization may be appropriate. Alternatively, in patients with incomplete or cartilaginous posterior spinous processes, Dewar's technique[27] using a K-wire across the posterior laminae above the dura is an excellent method for anchoring wire or suture that cannot

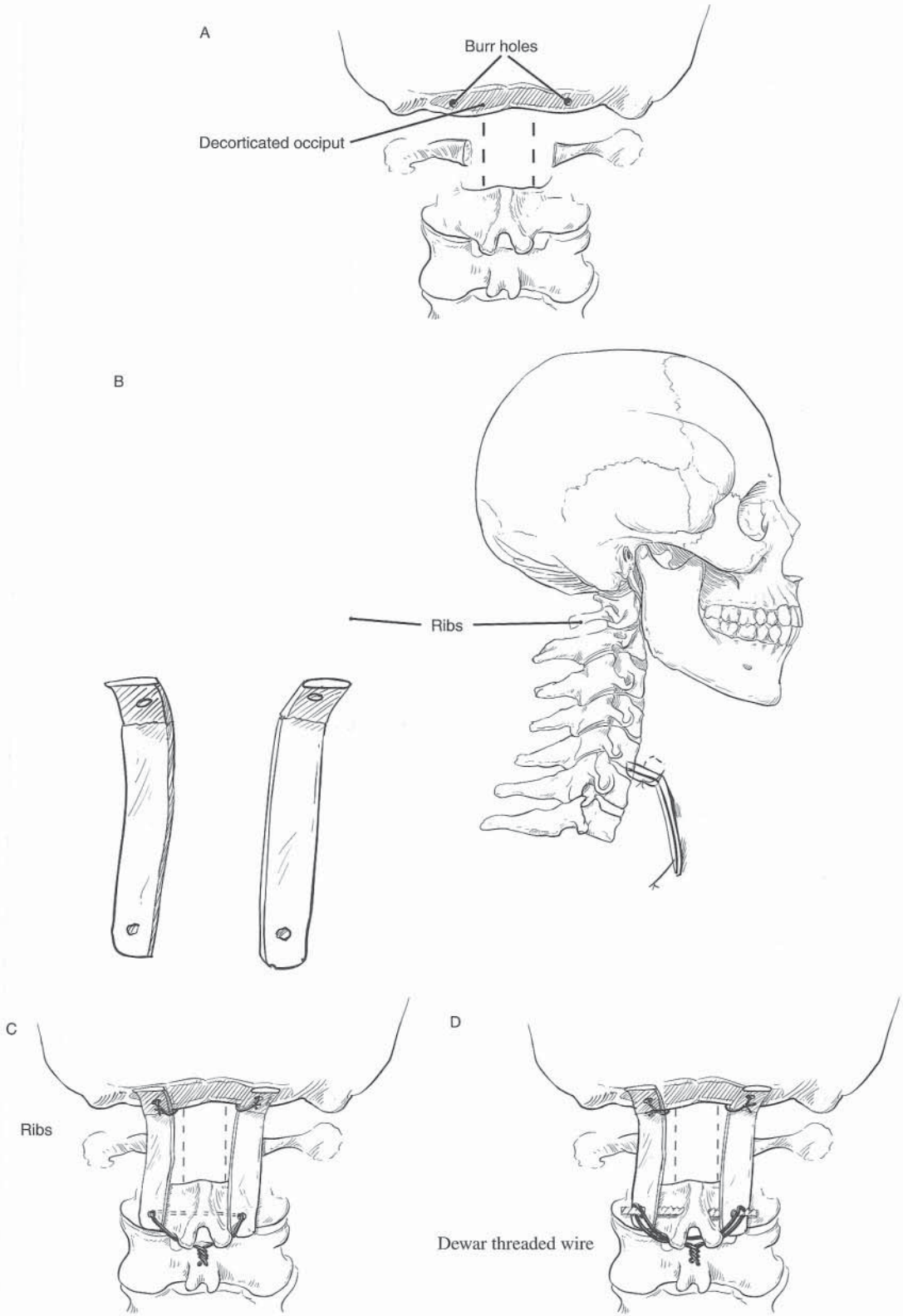


FIGURE 10–12 **A** to **C,** Scheme of occiput-C2 fusion following C1 decompression. **D,** The threaded wire technique of Dewar is useful when sublaminar passage of wire or suture is undesirable.

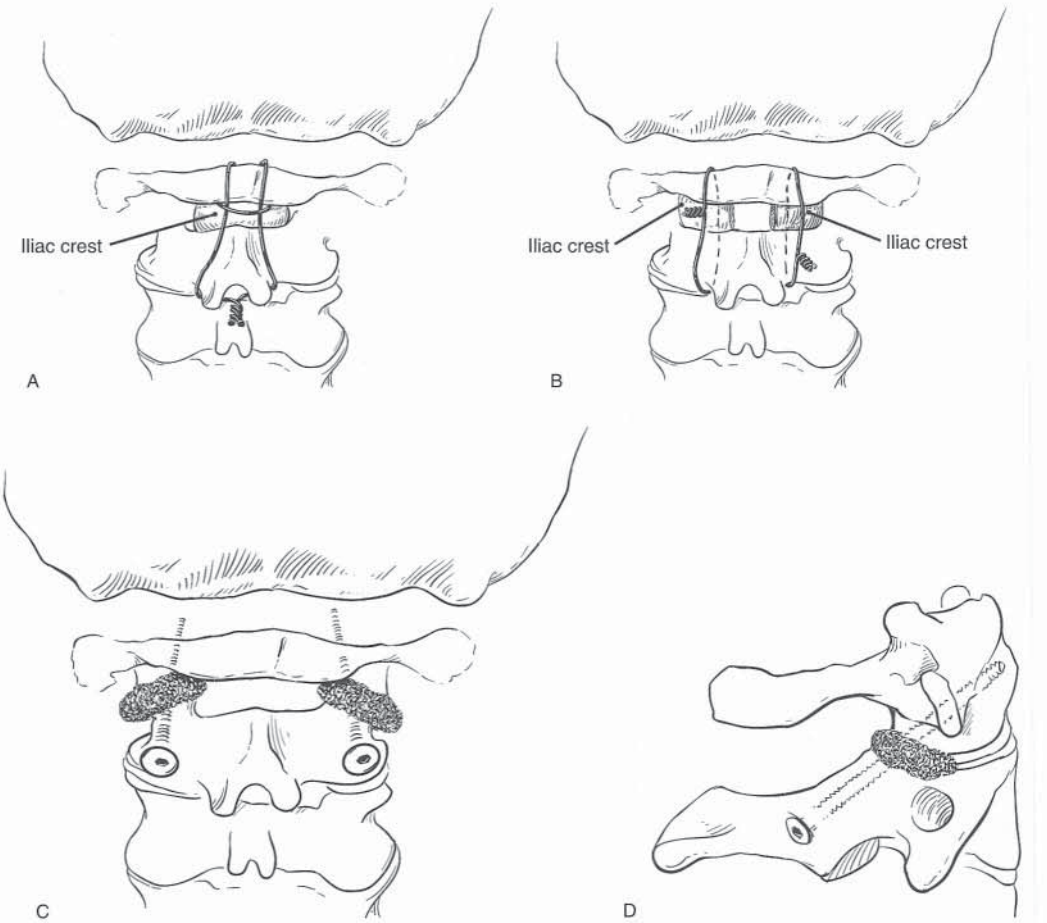


FIGURE 10–13 **A** to **D,** Gallie, Brooks, Mageryl techniques for atlantoaxial arthrodesis in children with C1–2 instability.

be passed sublaminarly (see Fig. 10–12D).[58] In patients less than 9 years old who are undergoing any posterior cervical fusion, this technique has been recommended to avoid wire pulling through a possible cartilaginous spinous process.[90]

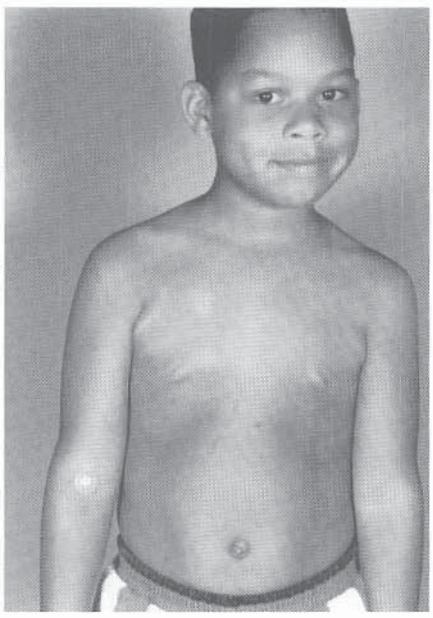
Acquired Torticollis

ATLANTOAXIAL ROTATORY DISPLACEMENT

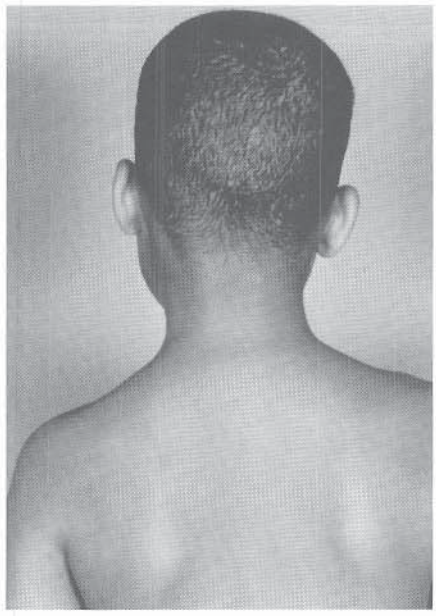
The most common condition producing *acquired, painful* torticollis is atlantoaxial rotatory displacement (AARD). Because of the relative frequency of upper respiratory infections, inflamed adenoids, and other oropharyngeal sources of bacteremia and the association of such sources of sepsis with Grisel's syndrome,[56,75,145] children frequently present with an acquired torticollis that, when manipulated to correct the deformity, elicits significant pain and resistance. The pathophysiology of spontaneous atlantoaxial displacement is probably inflammation of adjacent neck tissues resulting from a direct connection between the periodontoidal venous plexus and the pharyngovertebral veins of the posterosuperior pharynx.[113] This connection provides a route for hematogenous transport of peripharyngeal bacteria to the upper cervical spine region, leading to inflammatory hyperemia, which then produces ligamentous laxity at the atlantoaxial articulation. Coupled with regional lymphadenitis causing spasm and contracture of cervical musculature, the venous anatomy described by Parke and colleagues offers an explanation for the occurrence of AARD following upper respiratory infection, ENT procedures, and other forms of oropharyngeal surgery.[113] The rotatory laxity at C1–2 can then progress to a fixed position or torticollis.[92]

Other causes have also been proposed, most commonly trauma.[36] Meniscal-like folds of synovium in the occipital-atlantal and atlantoaxial joints, which can then be infolded during a sudden rotatory displacement (trauma), may actually prevent relocation of the atlantoaxial joints.[74]

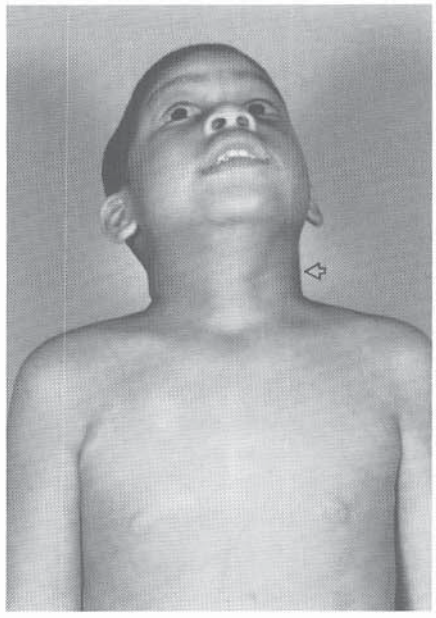
Fixed dislocation of the atlantoaxial joint (and thus rigid torticollis) is seen in only a small percentage of rotatory displacements. The milder forms of rotatory displacement probably resolve spontaneously without coming to medical attention, as the rotated displacement spontaneously reduces when the inflammatory process recedes. Fixed displacement is characterized by rigid torticollis, with the SCM muscle on the *contralateral* side, away from the head tilt, being in spasm and prominent, as if the muscle were trying to correct the deformity (Fig. 10–14). This is in contrast to the physi-



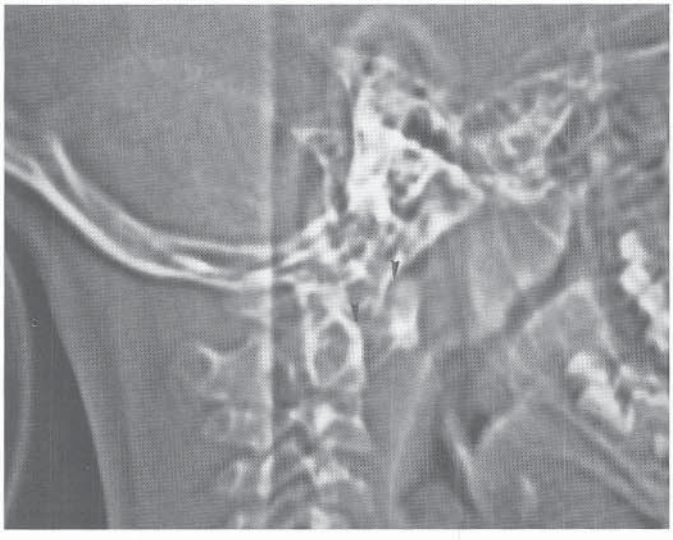
A



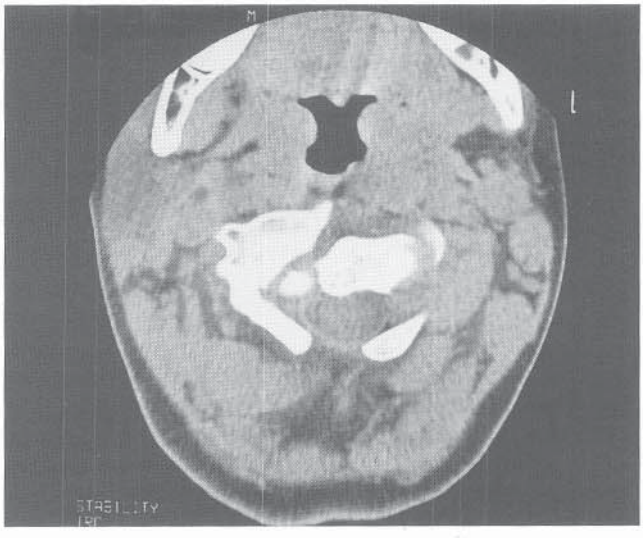
B

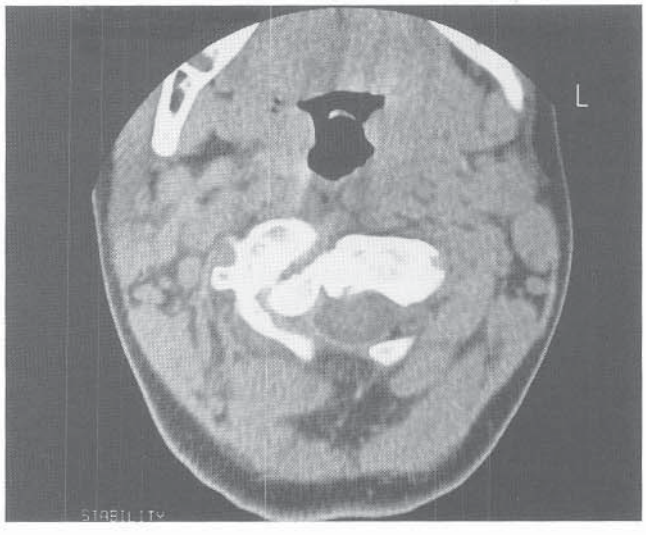


C



D





E

FIGURE 10–14 **A** and **B,** Clinical appearance of an 8-year-old boy with rigid torticollis. He had fallen off a jungle gym and hit his head several months earlier. **C,** Maximum extension. The left (contralateral) SCM is prominent (*arrow*). The patient had essentially no rotational movement. **D,** Lateral radiograph showing an increased ADI (*arrows*), indicative of persistent malreduction. Note residual tilt even with careful supervision of head position during fluoroscopy. **E,** CT scans showing rotatory displacement and minimal SAC.

cal findings in congenital muscular torticollis. Besides the "cock robin" tilt of the head and the finding of a prominent, contracted SCM muscle on the long side of the deformity, range of motion is markedly decreased, and the patient may experience pain at rest as well as increased pain with head manipulation. Plagiocephaly is usually not present unless the deformity has persisted for years. In posttraumatic cases, the inciting incident is often subtle and unknown to the parents, and in fact it may never be identified.[115]

Radiographic Findings. As with any torticollis, radiographs of the cervical spine and occipitocervical junction are often difficult to interpret. Malalignment of the head, along with the inability to comfortably position the patient, make it difficult to adequately assess this area, thus delaying the diagnosis.[40,41] Anteroposterior or open-mouth views of C1–2 are not useful, because it is impossible to differentiate the apparent facet subluxation seen in a normal child whose head is rotated from a fixed subluxation produced by AARD. The head tilt produces distortion of the normal appearance of the C1–2 joint on a routine lateral x-ray, and thus a true lateral view of the *skull*[146] is recommended. It is believed that the ring of C1 moves with the occiput.[111] Consequently, tilting of the head tilts C1, and a true lateral view of C1 is seen on a true lateral view of the skull. Such a radiograph will usually demonstrate an increased ADI due to the rotatory displacement (see Fig. 10–14D), thus giving the best plain radiographic evidence of AARD.

Cineradiography was used in the past to demonstrate the rotatory fixation,[40,41] but this older technique has been superceded by CT.[43] The diagnosis of rotatory fixation rests on the demonstration of a fixed rotation between C1 and C2 when the head is rotated maximally to the right and to the left and shows no motion or reduction of the rotatory displacement.

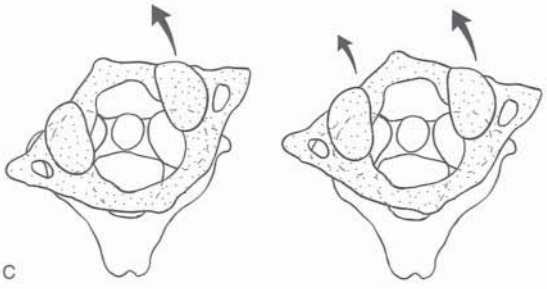
Rotatory displacement has been classified into four types:[41] type 1—a simple rotatory displacement without anterior shifting of C1; type 2—rotatory displacement and an anterior shift of 5 mm or less; type 3—rotatory displacement with an anterior shift of more than 5 mm; and type 4—rotatory displacement with a posterior shift. Anterior displacement of more than 3 mm in older children and more than 4 mm in younger children is considered pathologic.[43,86] Such displacement usually can be discerned on the true lateral view of the skull, but it is definitively seen on CT. Type 1 is by far the most frequent in the pediatric age group and is the most benign form, often resolving by spontaneous relocation of the facet joints. Types 2 and 3 (see Fig. 10–15C), in which some anterior shift is present, are the more severe, fixed rotatory displacements; and because of the decreased space available for the cord, these displacements raise the potential for neurologic compromise, which fortunately is rare in this condition, presumably because of the normally large diameter of the cervical canal.[145]

Treatment. The treatment of AARD is usually dictated by the duration of symptoms and deformity and proceeds in stepwise fashion from supportive analgesia and observation to surgical realignment and arthrodesis.[115] Patients with rotatory displacement of less than 1 week's duration can be treated with immobilization in a soft collar, combined with heat, rest, and analgesics. Full radiographic documentation is not necessary if the clinical presentation is typical. If spontaneous resolution does not occur and if more thorough imaging or evaluation does not suggest another cause (see below), then hospital admission for the use of halter traction, muscle relaxants, and stronger analgesics is recommended. Traction may be applied with the patient supine in bed, or it may be attempted with the patient sitting, using an overhead frame or traction apparatus over the top of a door. The use of other modalities (massage, deep heat, ultrasound, transcutaneous electrical nerve stimulation) to relieve spasm may also have value. Patients with rotatory displacement of more than 1 week's duration at presentation should probably be hospitalized directly for cervical traction and pharmacologic relaxation. If the cervical traction is unsuccessful or poorly tolerated (chin straps are not uncommonly discarded after a few days, at most), halo traction should be instituted. The halo allows both longitudinal and rotational forces to be applied to the head, thus increasing the chances for spontaneous reduction. Reduction should be confirmed by CT and held by a halo-vest apparatus for up to 3 months. In the author's experience, discontinuation of halo immobilization at 6 weeks allows resubluxation to occur, as the period of time required for atlantoaxial ligamentous healing probably exceeds the traditional 6 weeks of immobilization (Fig. 10–15). Other series[115] have also noted recurrence and resubluxation with periods of immobilization ranging from 2 to 8 weeks. In addition, an unstable reduction may not remain reduced in a halo vest, with resubluxation occurring and noted radiographically, usually within 1 week following immobilization in a vest (Fig. 10–16).

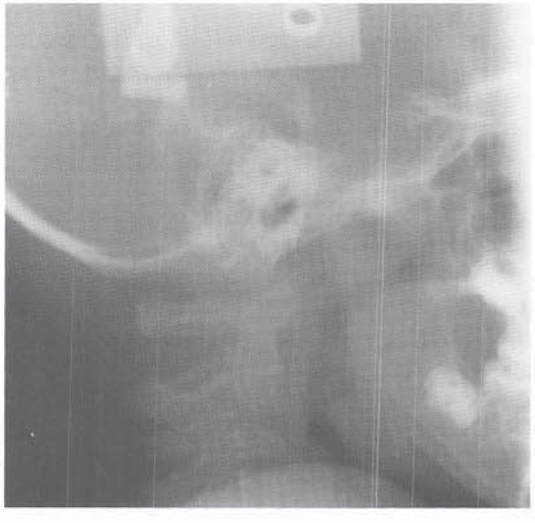
In patients presenting with rotatory displacement of more than 1 month's duration, the prognosis for successful closed reduction becomes guarded, and whereas traction is the appropriate first step, a halo device should probably be used from the outset to maximize the effects of the traction. Although traction has been recommended for up to 3 weeks,[87] failure to achieve reduction after 1 week probably indicates the need for closed reduction (repositioning) (see Fig. 10–15) and possible surgical fusion. Patients whose displacements do not reduce in traction can undergo *repositioning* under anesthesia and, with fluoroscopic control and neurologic surveillance (e.g., SSEP monitoring), can be placed in a halo-vest device in the slightly overreduced (overrotated) position.[13,36,72] Resubluxation may occur, and posterior C1–2 arthrodesis is then indicated (see Fig. 10–16). If the displacement cannot be reduced even by closed repositioning under anesthesia, the physician should proceed directly to surgical stabilization under the same anesthetic.

Posterior C1–2 fusion must be performed with maximal control of the patient's head, and thus a halo device should always be used for positioning. Fusion should be performed for any anterior subluxation, as this is potentially an unstable situation and can lead to a decreased SAC. Neurologic involvement preoperatively is rare; if present, it is an indication for immediate surgical reduction and/or decompression. In a patient with a longstanding unreducible AARD, an attempt at reduction by repositioning is not recommended because of possible neurologic injury. The C1–2 articulation should be stabilized in situ, and traditionally a Gallie-type fusion[41,48] has been recommended, with sublaminar wiring under the ring of C1 and around the spinous process of C2.

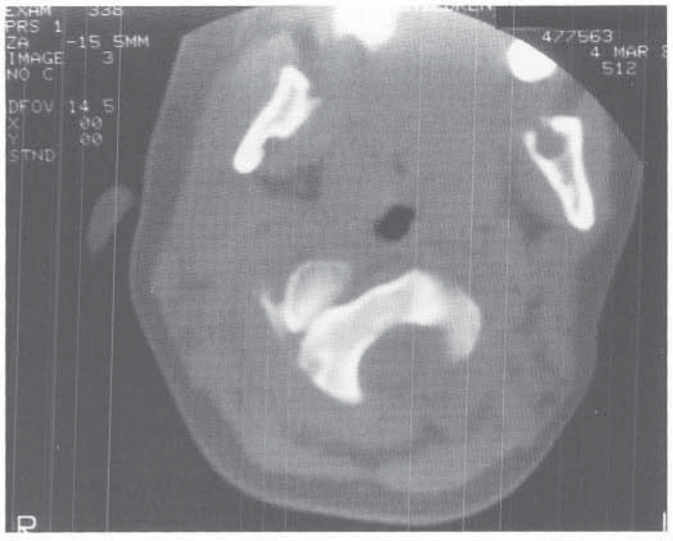
A

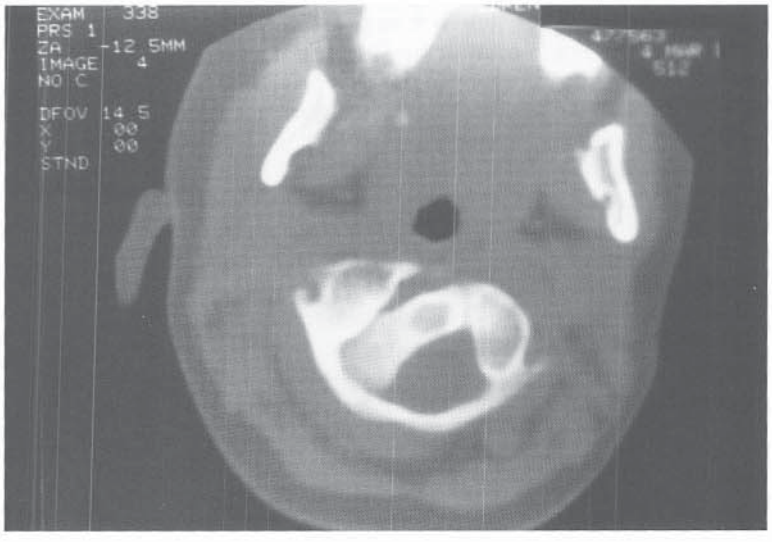


C

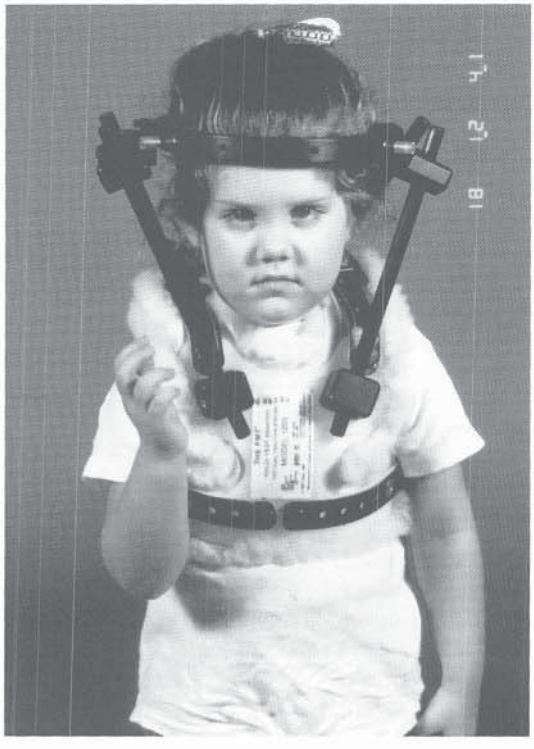


D



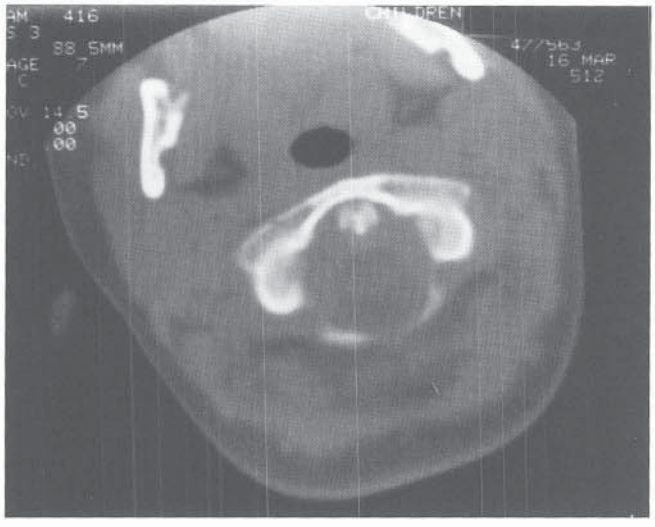


B

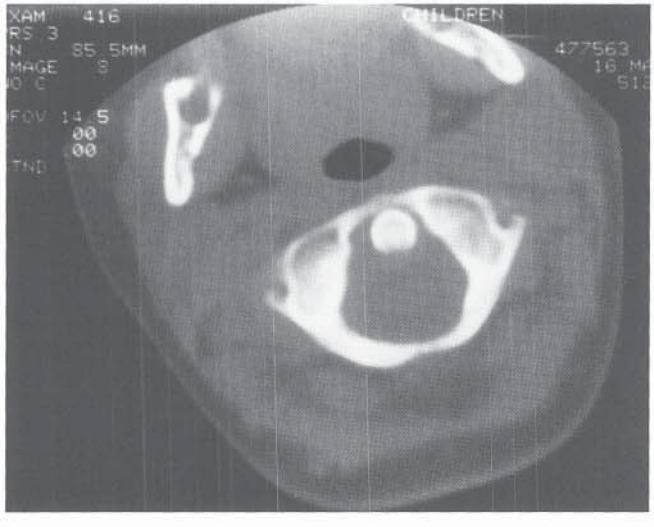


E

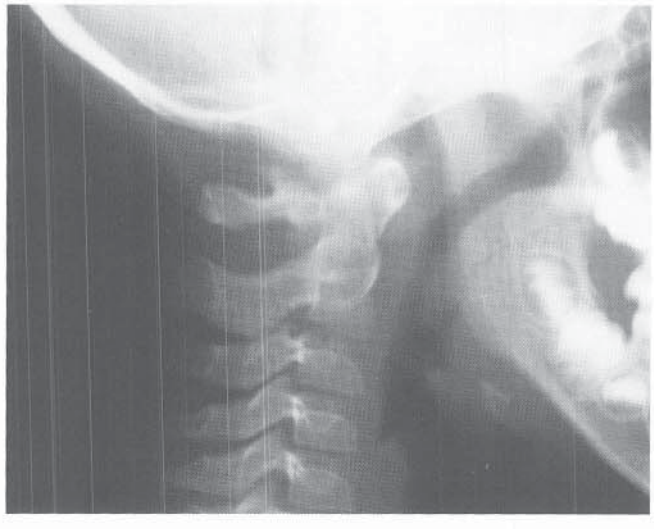
FIGURE 10–15 **A,** Painful torticollis 4 months after an unknown inciting incident in a 4-year-old child. **B,** CT scan showing type 2 atlantoaxial rotatory displacement (AARD). **C,** Types 2 and 3 AARD. (Modified from Fielding JW, Hawkins RJ: Atlanto-axial rotatory fixation [fixed rotatory subluxation of the atlanto-axial joint]. J Bone Joint Surg 1977;59-A:37.) **D** and **E,** Closed reduction and immobilization in halo vest after 1 week of traction.



G



F



H

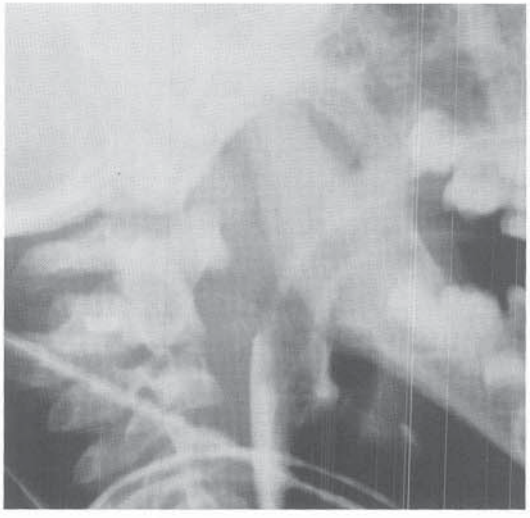
FIGURE 10–15 *Continued.* **F,** CT scan after reduction. The head has been overrotated to the right to maintain reduction. **G,** After 12 weeks, the reduction is maintained. **H,** CT apperance 1 year after reduction. Normal motion of the neck returned.

Sublaminar wire passage under C2, as in a Brooks fusion,[12] might jeopardize the dura in a narrowed SAC. The wiring is not intended to reduce the displacement but merely to stabilize it while arthrodesis occurs.

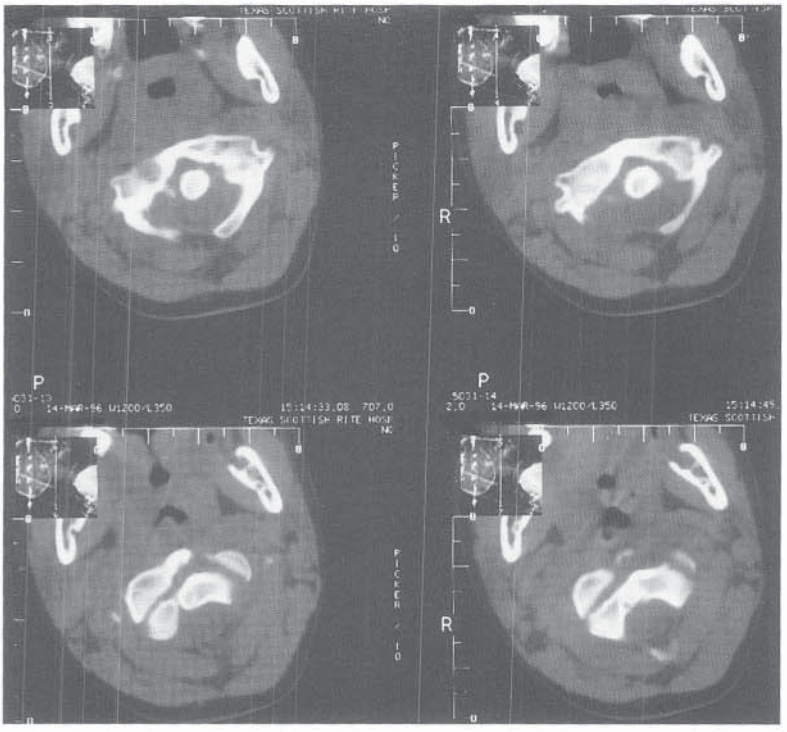
More recently, C1–2 transarticular screw fixation[68] has been an effective alternative, as it produces superior fixation,[100] and, if a lag screw technique is used, particularly in a type 2 AARD, to pull the anteriorly displaced side of the C1 ring posteriorly, it can actually reduce the rotatory displacement (see Fig. 10–16). The Mageryl technique does not require sublaminar passage of wires and thus does not invade the narrowed SAC, and it is invaluable should the posterior arch of C1 be incomplete, but yet another set of anatomic structures (e.g., the vertebral artery) must be avoided during screw placement. The use of cannulated screw systems is strongly recommended should the C1–2 transarticular screw technique be chosen. Due to the unreliability of young children, halo-vest immobilization is maintained in all patients with C1–2 fusions, regardless of the type of internal fixation used. With combined internal and external immobilization, rapid C1–2 fusion is usually noted in 6 to 8 weeks, at which time the halo is removed and the patient is allowed to resume normal activities, with the usual restrictions for patients who have undergone a cervical fusion. Although some rotation is lost due to the fusion of C1–2, significant recovery of motion, including rotation though the subaxial spine, occurs rapidly once the arthrodesis is solid and the displacement stable.

NEUROGENIC TORTICOLLIS

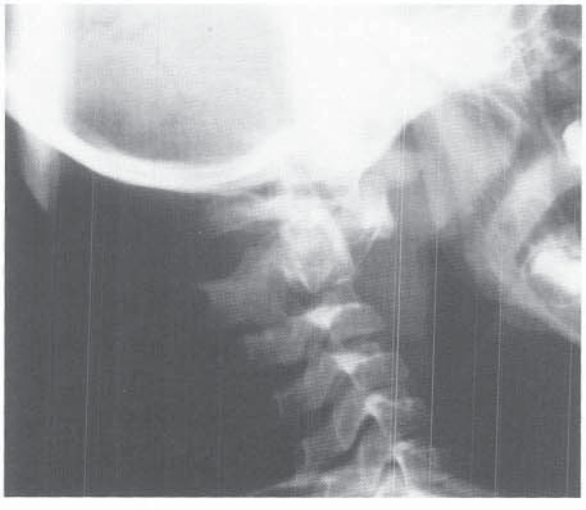
Rarer forms of torticollis must be considered when a deformity that is presumed to be of a congenital muscular type at initial presentation does not respond to the appropriate nonoperative measures or clearly becomes painful. The major neurologic causes include tumors of the CNS (posterior fossa or brain stem), Arnold-Chiari malformation, syringomyelia, and paroxysmal torticollis of infancy.



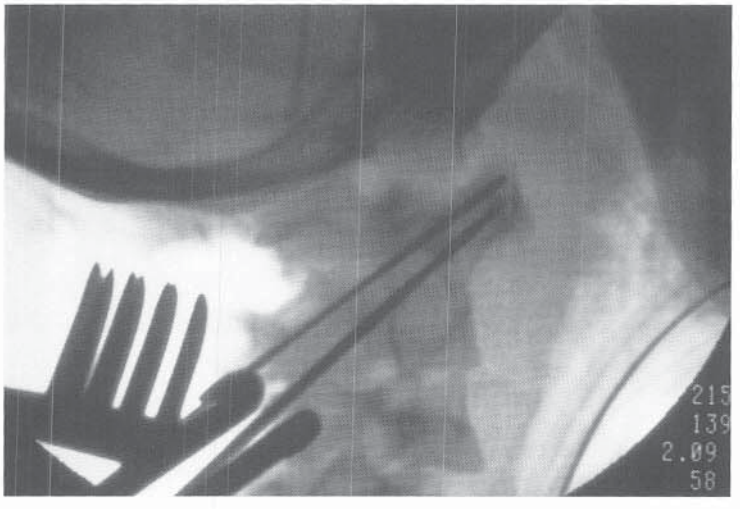
A



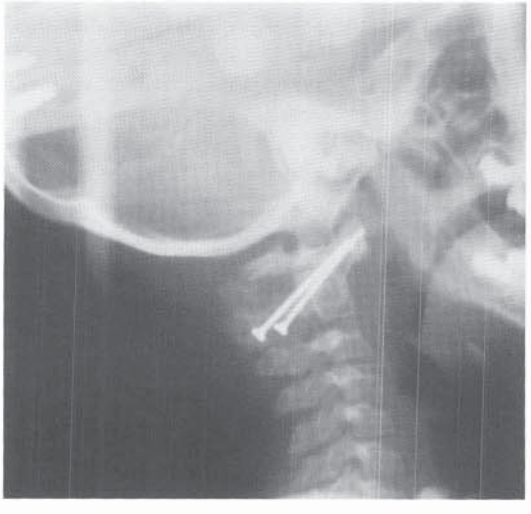
B



C



D



E

FIGURE 10–16 **A,** Closed reduction under anesthesia of the patient shown in Figure 10–14. He was placed in a halo-vest device. **B,** Incomplete reduction on CT scan, with some persistent anterior subluxation of the right facet. **C,** One week later, C1–2 is again displaced. **D,** Intraoperative position for cannulated screws for C1–2 fixation. **E,** Radiographic appearance 2 months postoperatively. The patient's halo immobilization was discontinued. Solid C1–2 arthrodesis is evident.

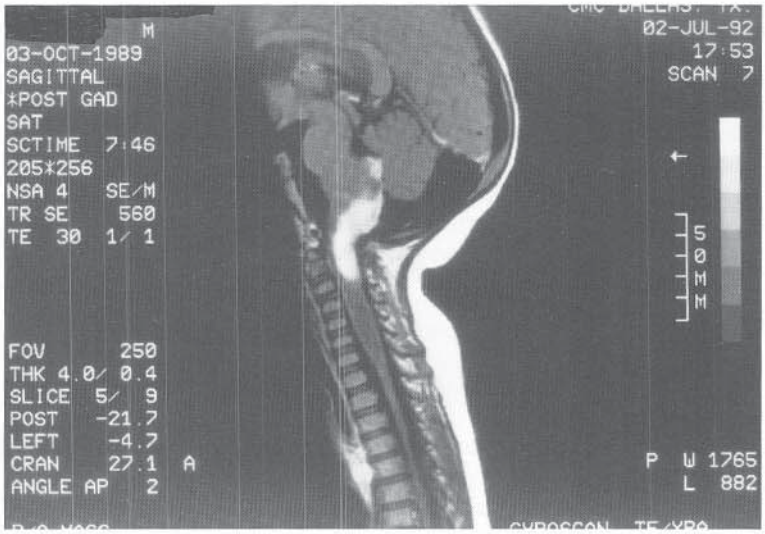


FIGURE 10–17 MRI of the brain stem, revealing the source of rigid torticollis associated with an ataxic gait.

A CNS tumor can manifest with essentially any neurologic sign—motor deficit, reflex changes (long tract signs), cranial nerve lesions, or signs of increased intracranial pressure. Additionally, owing to the mass effect of the tumor, the expansion of CNS tissue into the foramen magnum or cervical cord will generally produce restricted, painful motion of the head. Extraocular muscle paralysis, nystagmus, and papilledema are findings that should immediately raise the suspicion of a CNS tumor. Initially the torticollis may be diagnosed as congenital muscular torticollis, or the patient may be thought to have an obstetric palsy or cerebral palsy.[52,76] The rigidity of the deformity and irritability in a young child are additional indicators of the presence of a CNS tumor. The examiner's index of suspicion is crucial to the completeness of workup of the head tilt.

MRI of the head and cervical cord is the mainstay in diagnosing CNS tumors (Fig. 10–17). MRI should probably be the first study ordered if a neurogenic cause of torticollis is being considered, because it will accurately diagnose tumors, syrinx, and Arnold-Chiari malformations. Additionally, a bony inflammatory process such as diskitis, vertebral osteomyelitis, or an osteoid osteoma may also be identified during the search for the possible CNS tumor.

Because of the difficulty in confirming definite neurologic signs in young children, a delay in diagnosing an underlying CNS tumor can be significant, with the patient's overall health and survival sometimes at issue. Early recognition of an abnormal response in what is believed to be congenital muscular torticollis is probably the most important aspect of suspecting a noncongenital muscular cause. Consultation with a pediatric neurologist or neurosurgeon should be obtained whenever such an abnormal clinical response occurs.

Non-neoplastic causes of neurogenic torticollis primarily include the Arnold-Chiari malformation and associated expansion of the cervical cord from syringomyelia. An Arnold-Chiari type II malformation associated with myelomeningocele is routinely diagnosed, owing to the coexistence of the spinal and lower extremity deformities. The Chiari type I malformation, however, may be due to congenital deformities of the brain stem and cerebellum and is a not uncommon cause of an otherwise unexplained isolated torticollis associated with headaches and muscle spasm.[35]

A neurogenic torticollis is generally treated neurosurgically, so that the role of the pediatric orthopaedist becomes critical in identifying an underlying neurosurgical cause for the apparent orthopaedic deformity.

A rarer form of episodic torticollis, paroxysmal torticollis of infancy, is thought to be due to malfunction of the vestibular system.[113,129] Affected children are usually female and can present up to the age of $2\frac{1}{2}$ years, with episodic attacks of torticollis lasting from minutes to days, accompanied by lateral trunk curvature, ocular deviation, and alternating side of the torticollis. A family history of migraine is often associated, suggesting that the vestibular malfunction may be an infantile or pediatric response to a localized migraine-like episode. The diagnosis is one of exclusion, again requiring neurologic consultation. Fortunately, the condition appears to be self-limiting and does not require therapy.

INFLAMMATORY/SEPTIC CAUSES

Vertebral Osteomyelitis and Diskitis. In the radiographic evaluation of a patient with painful acquired torticollis, disk space narrowing or erosion of a vertebral body may be identified. When associated with fever and other signs and symptoms of infection, this vertebral osteomyelitis/diskitis is treated as any other pediatric spinal infection. Termed infectious spondylitis by Ring and colleagues,[121] the condition manifests with pain and difficulty in moving the head, and in approximately one-third of patients, *Staphylococcus aureus* can be isolated from either blood cultures or biopsy specimens of the involved disk space or vertebral body. However, with typical radiographic findings and the clinical syndrome, it is not necessary to wait for positive culture results before instituting treatment with antibiotics appropriate for gram-positive organisms. Disk and bone biopsy cultures are appropriate only if the clinical response to an initial course of antibiotics is unfavorable.

In a typical clinical response, the patient becomes afebrile and the pain resolves fairly rapidly with the institution of antistaphylococcal antibiotics. Nonsteroidal anti-inflam-

matory medicines are also useful in relieving localized neck pain and spasm. The intervertebral disk space may eventually reconstitute to varying degrees, but it rarely retains a normal height, and eventually spontaneous anterior vertebral fusion can occur. Surgical debridement for pathologic diagnosis and excision of the septic focus is necessary only when there is persistent evidence of osteomyelitis and the initial course of antibiotics is unsuccessful in resolving symptoms, such as in a case of fungal or microbacterial infection.

Tumor-like Conditions. Along with cervical inflammation due to infectious processes, several tumor-like conditions—eosinophilic granuloma, osteoid osteoma, osteoblastoma—can produce similar inflammatory or painful symptoms identical to the symptoms of a septic process. *Eosinophilic granuloma* usually produces vertebra plana, owing to involvement of the vertebral body, and its presence is usually obvious on plain radiographs.[55,128] However, should the lesion involve the upper cervical segments or base of skull, complex imaging may be needed to detect it.[11,29,55,140] Lesions of the upper cervical area or base of the skull may not be "hot" on bone scan[5,63] unless pathologic fracture has occurred, in which case instability as well as deformity may be the major clinical symptom. Maintaining a high index of suspicion—as in all types of painful, acquired torticollis—stimulates continued imaging until an occult lesion is found.

Treatment of eosinophilic granuloma usually requires little more than biopsy and curettage.[64] Most lesions of the spine resolve with minimal surgical intervention.[37] However, should neurologic deficit or instability occur, decompression and fusion may be required.[55,64]

Intervertebral Disk Calcification. This condition is diagnosed by the presence of calcified deposits delineating the nucleus pulposus on a lateral radiograph of the cervical spine. Over 100 cases have been reported,[38,63,130] with approximately one-fourth of the children presenting with torticollis. Movement is painful and limited, and, rarely, radicular signs or myelopathy are present. Because some one-fourth of children also present febrile, there is suspicion that this also represents a form of disk space infection, although trauma has also been proposed as a cause. MRI demonstrates inflammatory involvement of the vertebral body as well.[63] Mild disk protrusion is seen in symptomatic patients.

The cause of the calcification is obscure. There does not appear to be an accelerated aging process, as might be expected with the finding of calcific disk deposits in adults, because the calcific deposits regress and disappear in approximately 90 percent of children. Frank pyogenic spondylitis also appears to be ruled out, because the symptoms resolve rapidly in the majority of patients with simple symptomatic treatment. Analgesics, cervical collars, and bedrest with traction have all been successfully employed. Long-term sequelae, even with persistence of the calcification over the long term, do not appear to be significant, although degenerative changes may result.[150]

Juvenile Rheumatoid Arthritis. Patients with polyarticular or systemic onset juvenile rheumatoid arthritis (JRA) may demonstrate involvement of the cervical spine, usually early in the disease course, and usually present primarily with stiffness and loss of motion. Pain may also accompany the presentation.[61] Although patients with JRA may have a variety of cervical abnormalities, including erosion of the odontoid process, C1–2 subluxation, ankylosis of apophyseal joints, and subaxial subluxation due to rheumatoid involvement of the facet joints, a presentation that includes torticollis is rare, and additional diagnostic studies should be undertaken to rule out other causes of painful torticollis. The most frequent source of torticollis with JRA involvement is basilar impression due to erosion at the occipitocervical junction, which is rare in the pediatric age group.[47] Erosion of the lateral mass of the atlas with collapse can also cause a rigid, nonreducible head tilt.[59] As noted earlier, instability at either the upper cervical or subaxial levels due to bony erosion or subluxation is the main indication for treatment of the rheumatoid cervical spine. Positioning of the head with a cervical collar is appropriate nonoperative treatment for both torticollis and cervical instability unless there is neurologic involvement. In patients with systemic involvement, motion of the cervical spine is impaired, especially with apophyseal joints ankylosed early. Because of the tendency toward spontaneous ankylosis, instabilities tend to be self-limiting, but any patient with persistent neck pain and any sign of neurologic deficit must be evaluated by MRI or other imaging modalities to determine whether there is compression or stenosis secondary to basilar impression or subluxation. In these children, treatment of the instability or stenosis is performed as for any cervical anomaly, including decompression and fusion.

Sandifer's Syndrome. This syndrome, associated with gastroesophageal reflux, produces abnormal posturing of the head and neck[106,118] and is commonly seen in children with cerebral palsy or other conditions known to be associated with gastroesophageal reflux. The torticollis is believed to result from the child attempting to alleviate the pain of esophagitis from the reflux. Patients with significant gastroesophageal reflux may present with more obvious symptoms, such as vomiting, failure to thrive, and recurrent respiratory disease. In an infant, the differential diagnosis includes congenital muscular torticollis as well as the various dysplastic and congenital anomalies of the cervical spine. The patient usually is irritable from the esophagitis and associated respiratory discomforts, but range of motion of the neck is usually maintained, with no finding of SCM contracture. In this situation, by the process of exclusion, Sandifer's syndrome should be considered. The diagnosis is generally made by contrast studies demonstrating the reflux and appropriate pH studies of gastric contents confirming the esophagitis.[71] Treatment of the underlying gastroesophageal reflux cures the torticollis.

Ocular Torticollis. Ocular torticollis is a form of acquired torticollis, although the lesion that causes it is probably congenital. The diagnosis usually is not made until the child is approximately 9 months old, after head control and sitting balance are achieved. At this point, paralysis of the extraocular muscles—usually the superior oblique—produces strabismus and diplopia when the patient's head is level, and so the patient rotates the head to the uninvolved side in order to correct the diplopia.[10] The absence of SCM contracture in the setting of normal radiographic and neurologic findings should alert the examiner to the possibility of an ocular abnormality, with formal ophthalmologic consultation ob-

tained to confirm the diagnosis. Treatment of the extraocular muscle imbalance cures the torticollis.

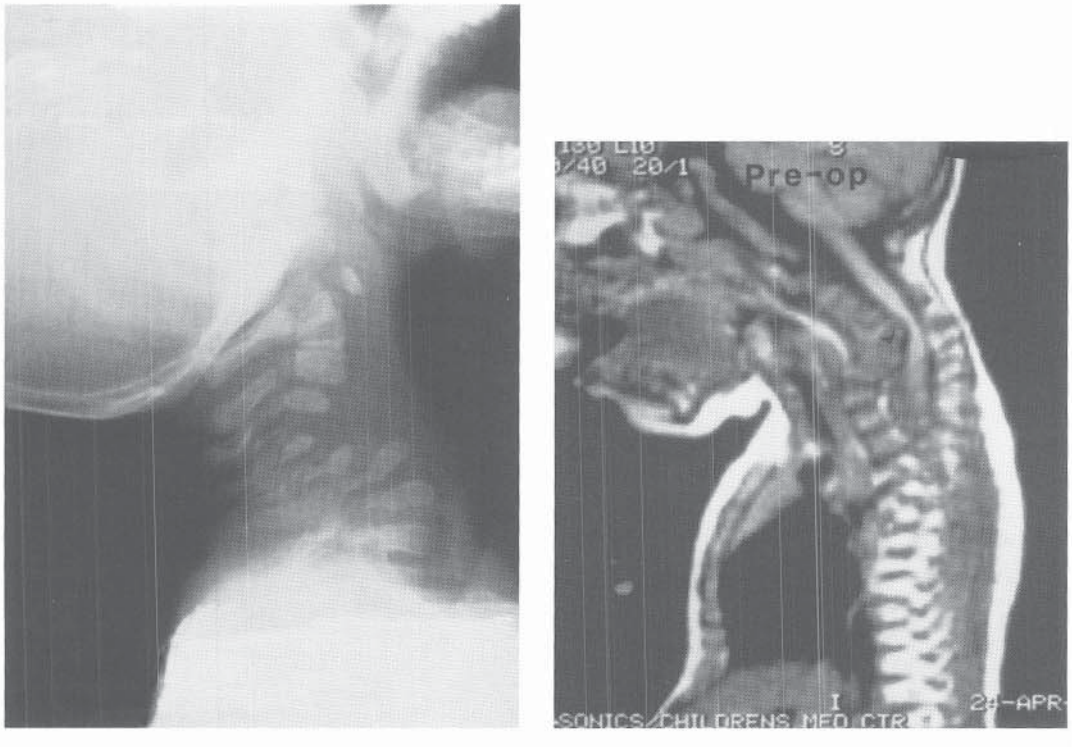
Cervical Kyphosis

Because the cervical spine is normally in lordosis, any kyphosis should be considered pathologic. Because of the relatively horizontal orientation of the cervical facet joints in a younger child,[40] one would expect a tendency for anterior translation and subluxation to produce kyphosis in the cervical spine, insofar as the weight of the child's head, being proportionately greater relative to the weight of the rest of the body at this age, produces a flexion moment. Nevertheless, except in specific instances in which dysplasia of an anterior vertebral body is present or in which the posterior cervical stability has been disrupted by laminectomy or disease, cervical spine kyphosis is relatively rare. It may be considered congenital or developmental when it is associated with syndromes or skeletal dysplasias, or acquired when it occurs after laminectomy.

Congenital or developmental cervical kyphosis is associated with Larsen's syndrome,[44,69,70,98] diastrophic dysplasia,[8,44,116,120] chondrodysplasia punctata (Conradi's syndrome),[136] camptomelic dysplasia,[20] and neurofibromatosis.[22–24,60,78,142,153] Because of the generalized skeletal involvement that may accompany any of these syndromes, the early diagnosis of cervical kyphosis depends heavily on the physician's index of suspicion. In addition, because of the early age at which the kyphotic deformity is present—at or shortly after birth, in most cases—the possibility of severe neurologic compromise developing in infancy necessitates early diagnosis and treatment. Although the syndromes associated with cervical kyphosis are rare, the devastating neurologic compromise resulting from failure to diagnose this deformity—a lifetime of paralysis, and even sudden death—has been well documented, especially for patients with Larsen's syndrome.[44,80,98,107]

CLINICAL FEATURES

In *Larsen's syndrome* the physician may be faced with one of the most challenging sets of orthopaedic deformities in existence. As originally noted by Larsen and colleagues in 1950, affected patients present with multiple joint dislocations (hips, knees, elbows) and frequently with various foot deformities (clubfoot, serpentine or "Z-foot," equinovalgus). Dysmorphic facial features include frontal bossing and a flattened nasal ridge. In the past, the multiple extremity deformities tended to distract attention from the cervical deformity, which was not emphasized in the original description of the syndrome[80] but is potentially the most serious and life-threatening manifestation of the syndrome because of impingement on the spinal cord at the apex of the kyphosis (Fig. 10–18). A typical presentation is that of an infant or young child who is nonambulatory due to "hypotonia," which traditionally was thought to be a part of the syndrome. Because of the multiple joint dislocations and foot deformities, more traditional methods of diagnosing spinal cord compression, such as tests to establish normal muscle strength or to identify pathologic reflexes, are difficult to perform or yield uninterpretable results. If cord com-



A B

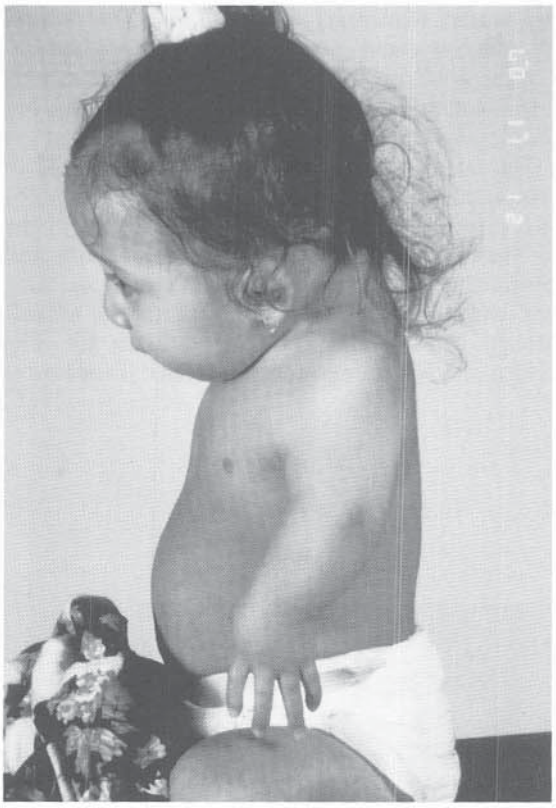
FIGURE 10–18 **A,** Lateral radiograph of a 10-month-old infant with Larsen's syndrome. **B,** MR image showing cord impingement and narrowing.

pression in an infant occurs prior to myelinization, the presence of hyperactive deep tendon reflexes or spasticity as a sign of upper motor neuron dysfunction will not be present; in fact, hypotonia is the more likely finding. Thus, as in any infant presenting with hypotonia, cervical cord compression must be considered during the neurologic evaluation.

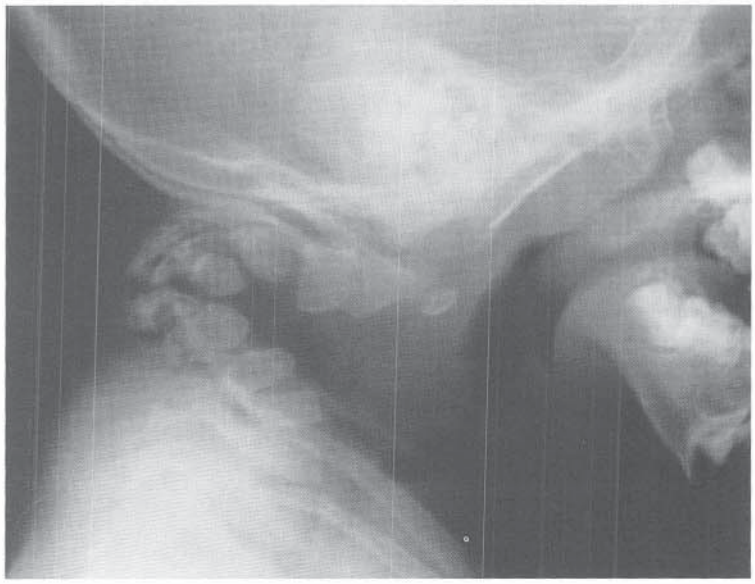
In *diastrophic dysplasia* the neurologic evaluation of such an infant or young child is further disturbed by the marked contractures and stiffness of the joints. Whereas in Larsen's syndrome laxity is the major finding, even though joints may be dislocated, the joints in diastrophic dysplasia show fixed contractures, often severe, and thus prevent an assessment of muscle strength or reflexes. The diagnosis is usually obvious, owing to the typical dwarfing of the extremities, the severe contractures, the typical "hitchhiker's thumb," rigid talipes equinovarus, and the characteristic chondritis of the ear cartilage, producing a pathognomonic ear deformity.[79,141] In a patient who has achieved sitting balance, the kyphosis of the cervical spine may be quite obvious, owing to the flexed position of the head (Fig. 10–19).

In Conradi's syndrome (*chondrodysplasia punctata*), the infant or young child will again appear hypotonic and will have characteristic facies, with an apparent dwarfing syndrome. Skin manifestations are an additional indication of the diagnosis, but the sine qua non is the finding of stippled calcification on radiographs, which may be present only in the earliest infant films and then gradually disappear over time (Fig. 10–20).[2,51]

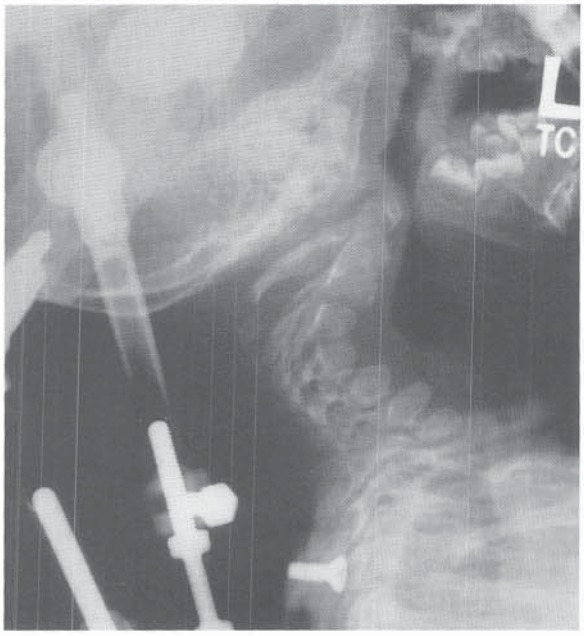
In *neurofibromatosis,* cervical kyphosis is more likely to



A

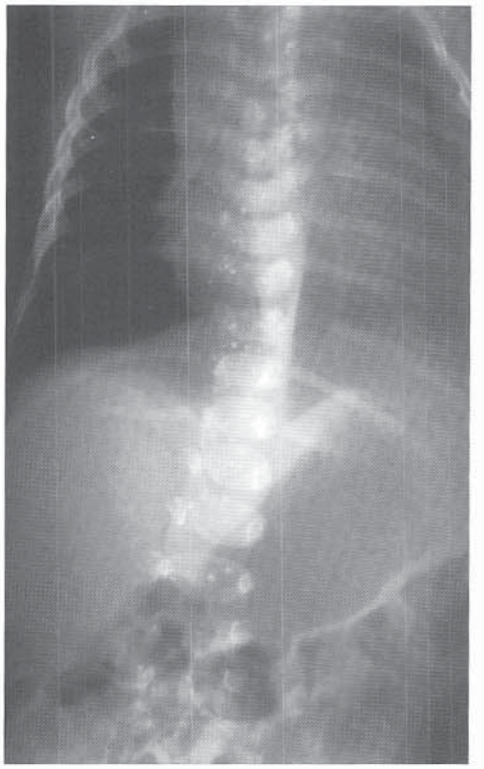


B

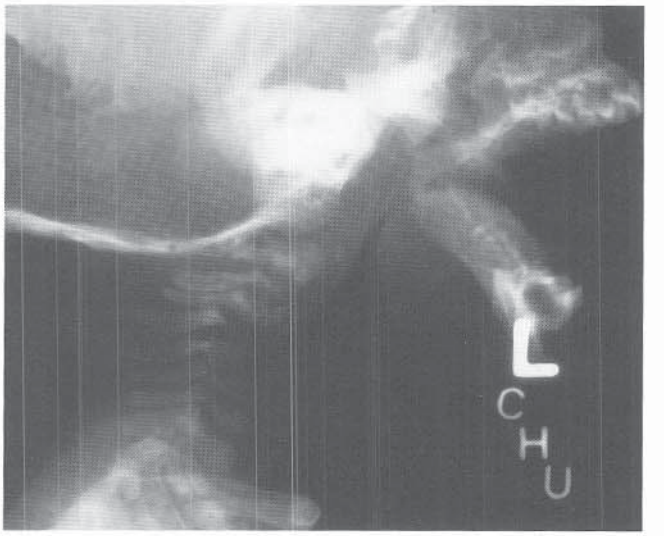


C

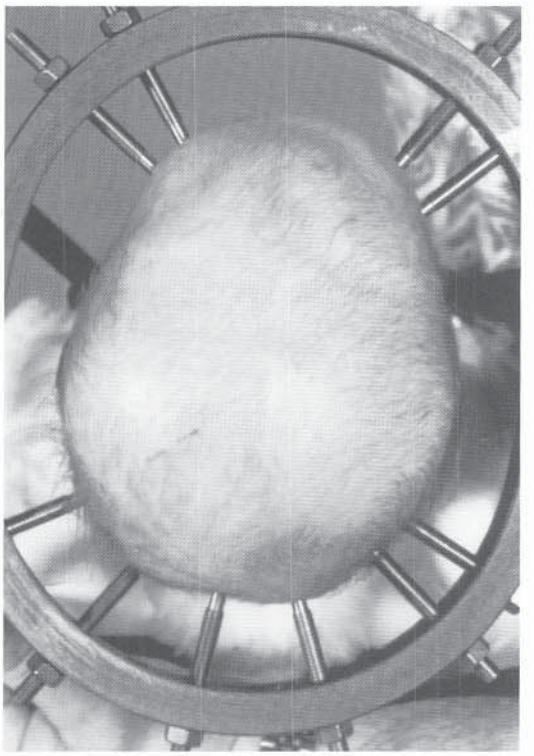
FIGURE 10–19 Diastrophic dysplasia. **A,** Clinical appearance. **B,** Lateral cervical radiograph. Because of the severe kyphosis, the odontoid is horizontal. **C,** Radiograph showing gradual correction by means of the halo-Ilizarov distraction technique prior to fusion (see Fig. 10–24).



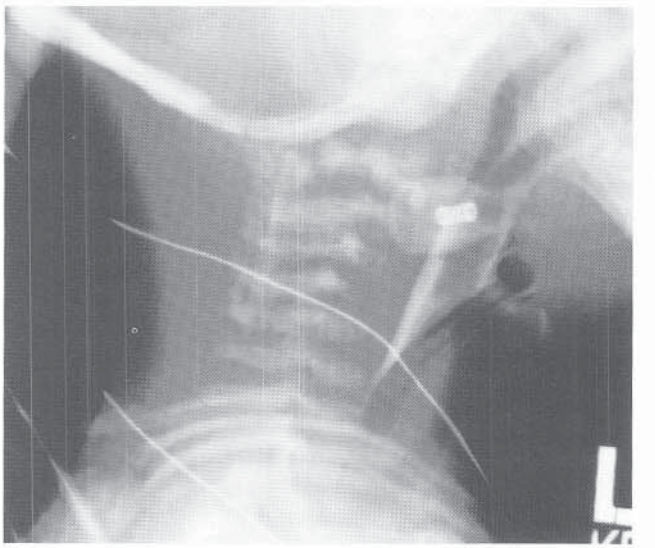
A



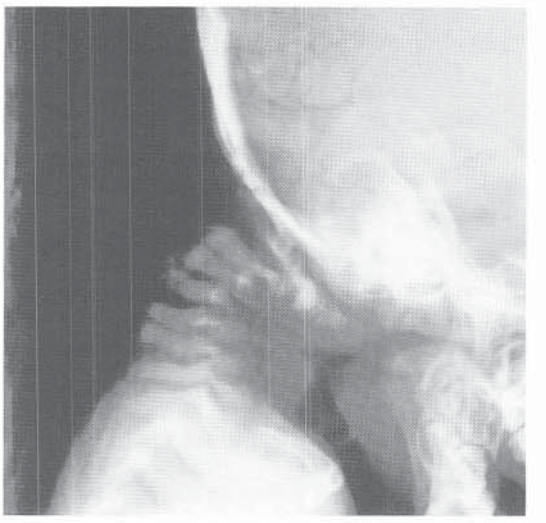
B



C



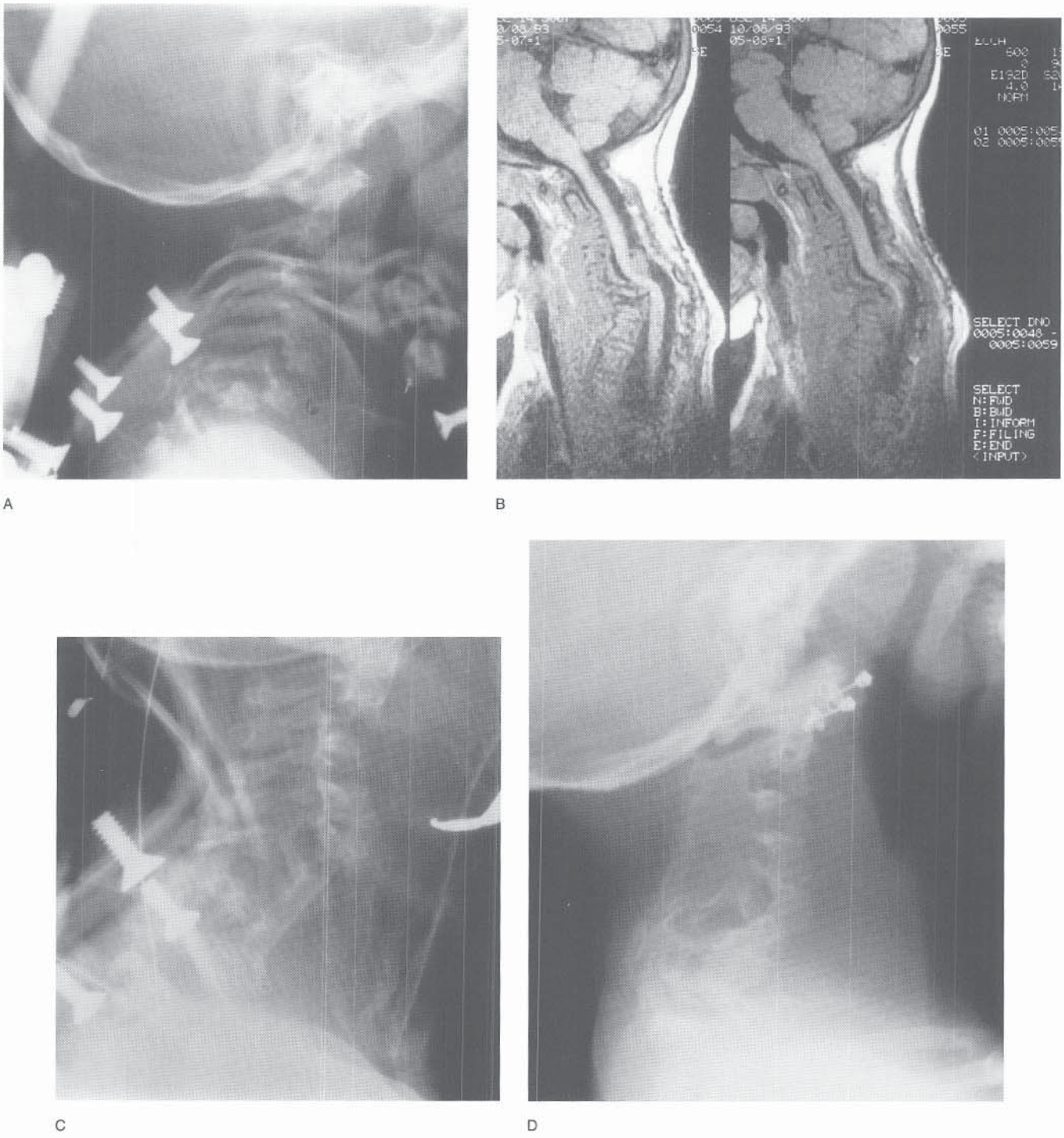
D



E

FIGURE 10–20 **A,** Stippled calcifications diagnostic of chondrodysplasia punctata. **B,** Upper cervical kyphosis in a 6-month-old infant with severe hypotonia and recurrent pneumonias. **C,** Deformation of the skull by screws because of soft bone and open cranial sutures. The most posterior screws loosened quickly. All screws were clinically loose at 6 weeks, but this was felt to be an adequate immobilization for such a young child. **D,** Posterior occiput-C5 fusion was performed to stabilize the kyphosis. **E,** Resorption of graft and pseudarthrosis following attempted occiput-C5 fusion at age 12 months.

occur in patients who are already known to have scoliosis or kyphoscoliosis.[22,153] The underlying neurofibromatosis will almost certainly have been diagnosed because of the skin manifestations, and in general, the cervical kyphosis may be found coincidentally after diagnosis of the spinal deformity. Actual neurologic compromise from kyphosis is unusual, although cord compression or dysfunction from intraspinal neurofibromas is well known (Fig. 10–21). The main issue with neurofibromatosis is to diagnose intraspinal masses before instituting traction (e.g., to treat a spinal deformity). The cervical spine abnormalities usually produce no symptoms whatsoever, with the only evidence being an abnormal lateral posture of the head and neck seen on clinical examination.



A B C D

FIGURE 10–21 Neurofibromatosis. **A,** Lateral cervical radiograph of a 3-year-old child who had transient weakness and neck pain following a roller-coaster ride. **B,** MR image showing marked compression due to a spondyloptosis of C6 on C7. The patient was placed in a halo vest. **C,** Anterior and posterior fusion was performed, owing to the risk of pseudarthrosis in neurofibromatosis. Posterior fusion was carried up to C2 because of the localized kyphosis at C2–3. **D,** Radiograph obtained 2 years postoperatively showing stabilization.

RADIOGRAPHIC FINDINGS

The cervical spine in Larsen's syndrome is characterized by multiple spondylolyses affecting several of the subaxial cervical vertebrae. The kyphosis usually develops because of hypoplasia of one or two midcervical vertebrae, most commonly C4 and C5 (see Fig. 10–18). A certain amount of flattening or platybasia of several of the vertebral bodies may also be present, and additionally, dysplasia of the posterior elements with incomplete formation of the spinous processes may be evident.[44,69] The actual kyphosis may range from 35 to as much as 100 degrees,[69,70] and the observation of relatively little neurologic compromise despite an alarming degree of kyphosis should not deter the physician from promptly stabilizing this deformity.

In diastrophic dysplasia, the presence of cervical kyphosis is better known than in Larsen's syndrome, having been reported to occur in 15 to 44 percent of patients.[8,44,116,120] The same hypoplastic vertebral bodies, especially C3, C4, and C5, associated with dysraphic posterior elements are noted. In addition, patients with diastrophic dysplasia usually exhibit a horizontal odontoid, thus requiring a significant occipital hyperlordosis in order to keep the head upright (see Fig. 10–19). In contradistinction to Larsen's syndrome, the kyphosis in diastrophic dysplasia has been reported to correct spontaneously.[8,44,116,120] The most current recommendation is to observe kyphoses less than 60 degrees that have no associated neurologic symptoms, with resolution on serial radiographs expected by age 7.[120] On the other hand, kyphoses greater than 60 degrees with apical vertebral wedging must be followed closely. Severe or progressive kyphosis can lead to quadriparesis and death.[8,73,120]

In Conradi's syndrome, kyphosis is again associated with hypoplastic or dysplastic vertebral bodies. Because of poor ossification and stippling (see Fig. 10–20), elucidation of bony detail may require other imaging studies, such as CT or MRI (see Fig. 10–20). Due to persistent dysplasia and hypoplastic growth of the cervical vertebra in Conradi's syndrome, correction of kyphosis by anterior growth in the presence of posterior fusion may be ineffective for this condition (see discussion under Treatment, below).

In neurofibromatosis, radiographic findings include dysplastic kyphosis at any location between C2 and C7. Scalloping of the vertebral bodies posteriorly as well as enlargement of cervical foramina, typical of neurofibromatosis involvement of the spine in other areas, is common, and the kyphosis, if sharply angular, may be associated with significant vertebral wedging. The degree of dysplasia of osseous structures tends to correlate with the severity of the deformity.[60,153] Instability is an additional factor that may exacerbate any cord impingement by kyphosis. Dural ectasia may produce marked laxity or pathologic dislocation at either the occipitocervical or subaxial levels.[67,134,142]

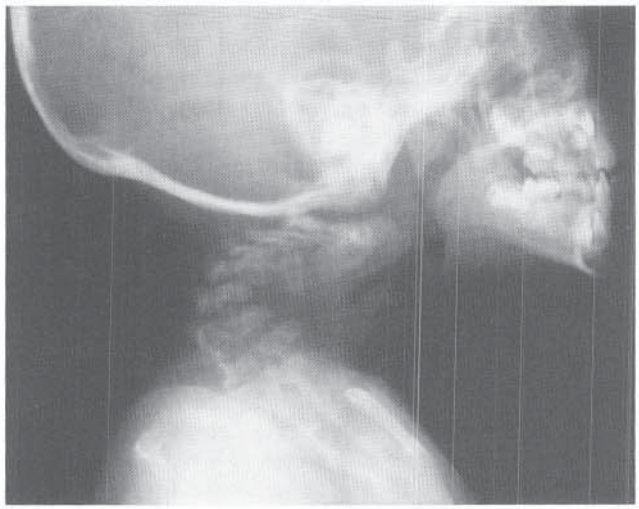
TREATMENT

The discovery of cervical kyphosis requires immediate assessment because of the threat of neurologic compromise. A baseline MRI to determine the narrowness of the cervical canal and the degree of anterior impingement is strongly recommended, even in a patient who appears to be neurologically intact. If the kyphosis is mild and no neurologic deficit can be determined, a period of observation may be appropriate, with the possibility of diastrophic dysplasia resolving sponteneously, as mentioned. Because the actual incidence of cervical kyphosis in these rare conditions is unknown, follow-up radiographs are probably appropriate for all of these diagnoses, even though the initial screening radiograph may show minimal or no deformity. There is no question that, in Larsen's syndrome at least, cervical kyphosis is probably underdiagnosed.[53,69]

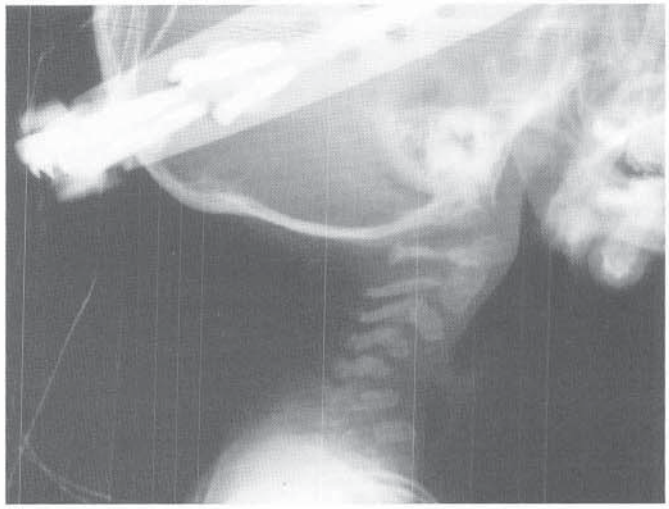
Orthotic management of cervical kyphosis may be appropriate in an attempt to buy time and allow growth before surgical stabilization is performed. There is no evidence that orthotic management alters the natural history of a cervical kyphosis,[120] but it may afford some protection until the prognosis can be determined. If the deformity is diagnosed before age 1 year and if there is no compelling neurologic deficit requiring immediate attention, surgical stabilization will be more technically feasible and solid arthrodesis more likely to occur if the surgical stabilization can be delayed to around 18 months of age. We have performed posterior fusion in children less than 1 year old, but, contrary to the traditional belief that posterior cervical fusion is almost assured in a young child, pseudarthrosis has occurred in patients with Larsen's syndrome and in those with Conradi's syndrome (see Fig. 10–20E). For this reason we currently recommend delaying posterior cervical fusion if at all possible until around age 18 months. Of course, neurologic deficit developing prior to this age eliminates not only the possibility of orthotic management but also any delay in stabilization or decompression.

Surgical options for *stabilization* include posterior cervical fusion, anterior cervical fusion, or both.[45,78,142] Decompression requires anterior vertebrectomy, as the source of the cord impingement is anterior, the apex of the kyphosis. In a young child with significant kyphosis, the surgeon faces a dilemma, in that anterior fusion in situ, an appropriate treatment for deformity without neurologic deficit, may in fact prove detrimental as growth proceeds and the spinal cord becomes chronically compressed against a thick, fused kyphosis. Anecdotal experience with anterior cervical fusion alone has been poor, as an early anterior fusion fixes the anterior column length, but if the posterior elements are allowed to continue to grow, the kyphosis may worsen, and the apical vertebral body anteriorly may gradually project further and further posteriorly into the spinal canal, producing late neurologic deficit (Fig. 10–22). Anterior cervical fusion is frequently recommended for a kyphotic deformity, because placing the anterior bone graft under compression achieves solid arthrodesis. However, in an infant or young child, the difference in growth between the anterior and posterior elements almost necessitates a posterior fusion to accompany any anterior fusion, to eliminate the possibility of increasing kyphosis with posterior element growth in the face of an anterior tether. Thus, in our hands, the main indication for an anterior fusion in young children with cervical kyphosis is to stabilize the anterior column following anterior *decompression,* or to salvage a posterior cervical fusion that has failed because of pseudarthrosis.[69]

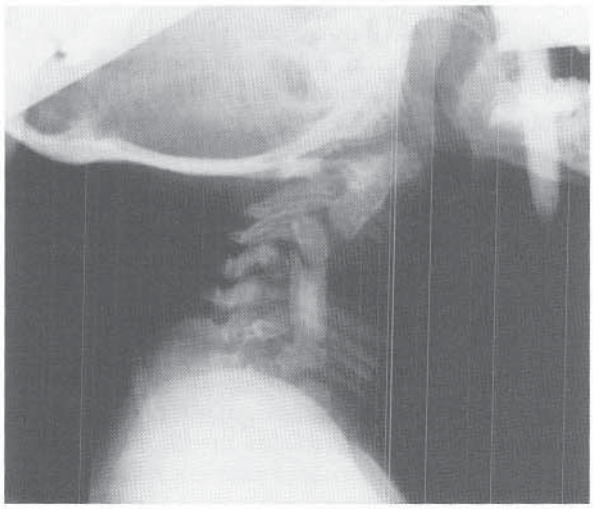
Indeed, the advantages of posterior cervical fusion in the young child are significant. First, a successful posterior fusion will tether posterior growth, and any anterior vertebral growth will then gradually correct the deformity and may



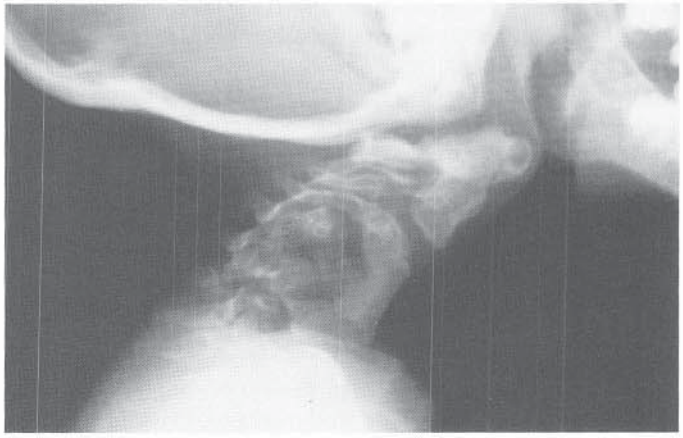
A



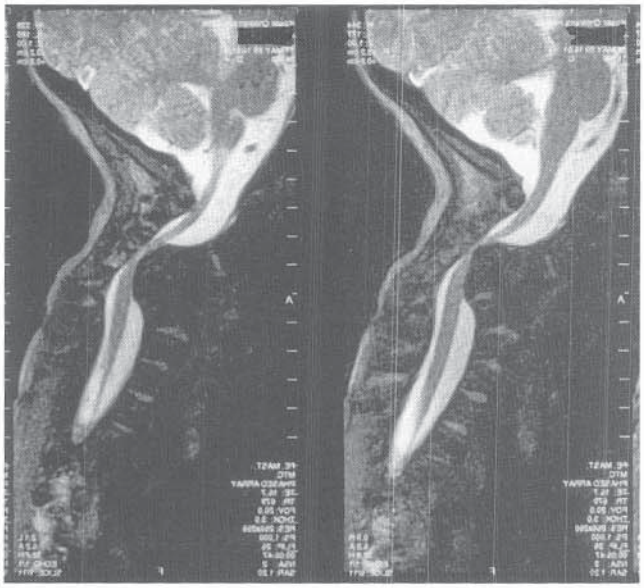
B



C

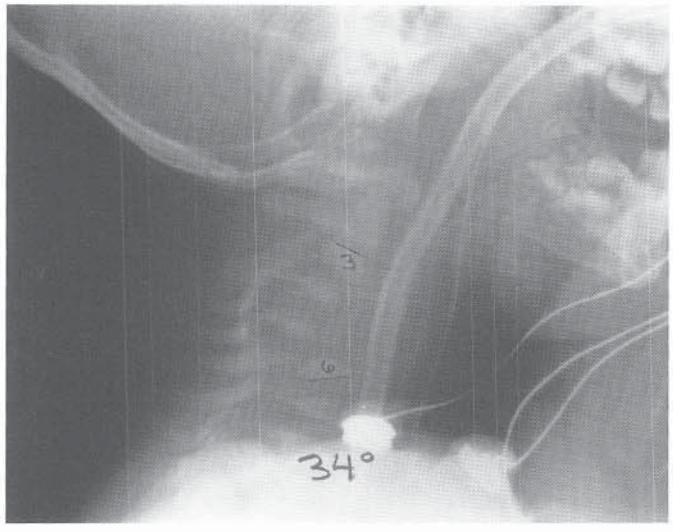


D

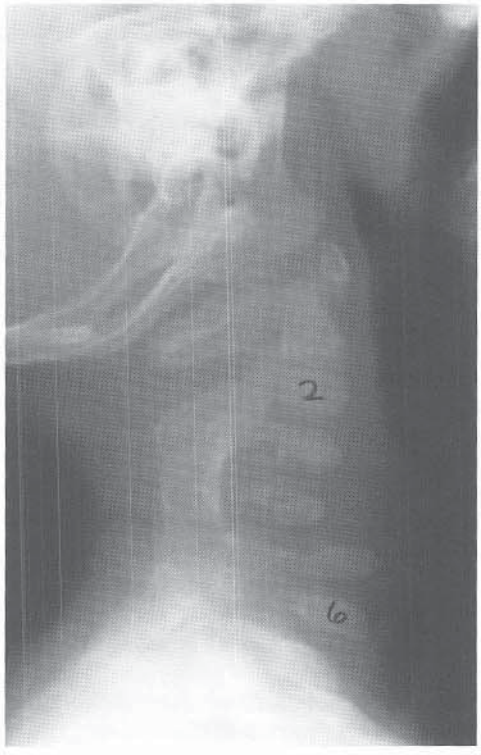


E

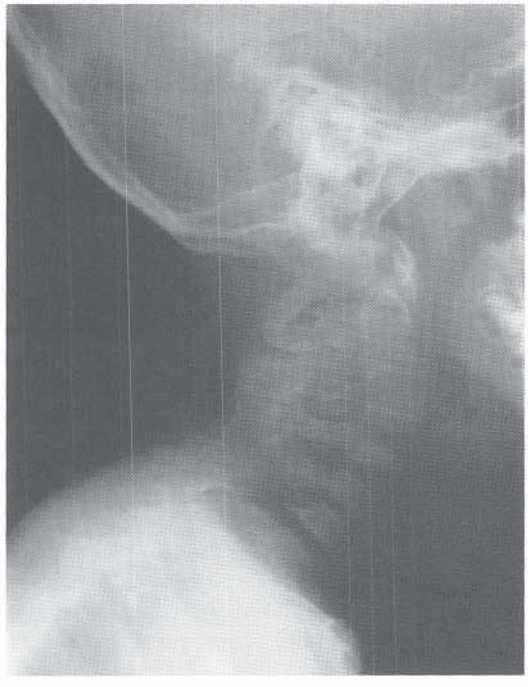
FIGURE 10–22 Larsen's syndrome in a 3-year-old child. **A,** Initial radiograph. At presentation, the child had difficulty walking because of weakness. **B,** Reduction of kyphosis in traction. **C,** Radiograph showing anterior strut grafting from C3 to C7. No posterior fusion was performed. **D,** Radiograph obtained 2 years later, after the child fell and became quadriplegic. Although there is solid anterior fusion, posterior growth has increased the kyphosis, and the body of C5 protrudes posteriorly into the canal. **E,** MR images demonstrating protrusion of the C5 body. The patient suffered respiratory arrest following emergency anterior decompression and died.



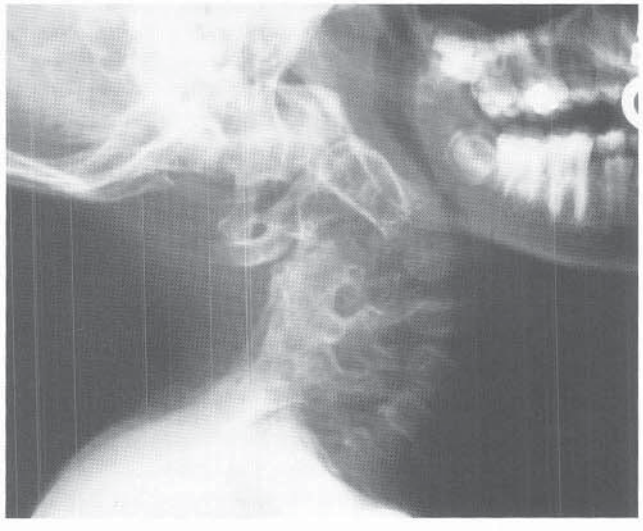
A



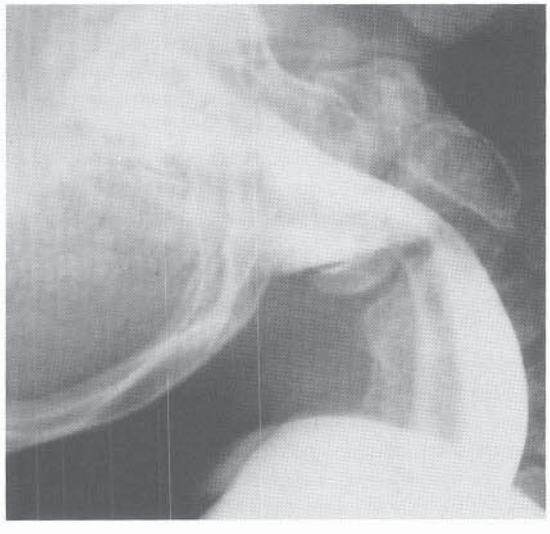
B



C

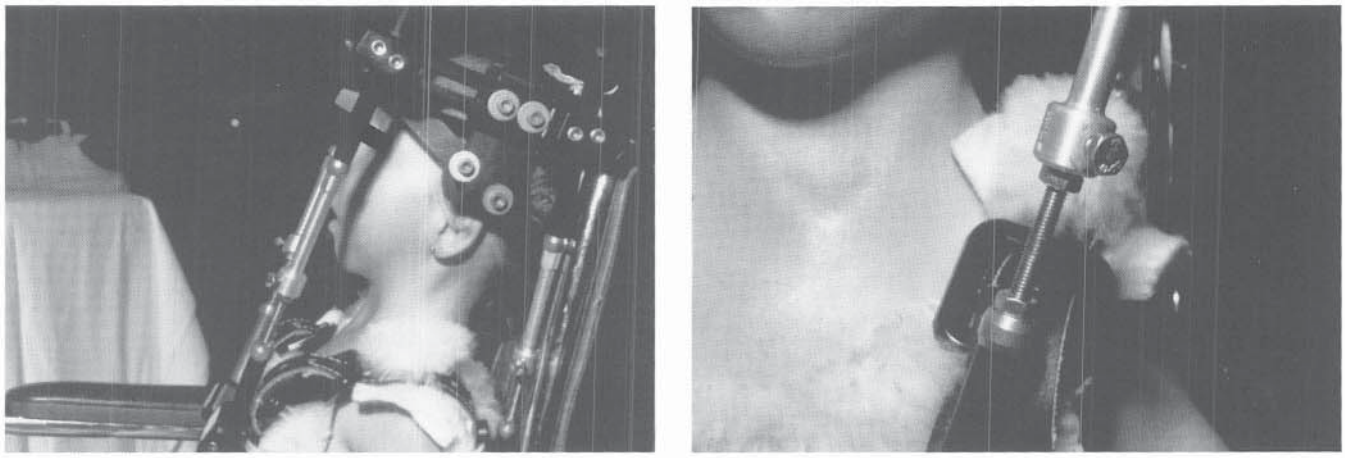


D



E

FIGURE 10–23 **A,** A 34-degree kyphosis in a 10-month-old child. **B,** Radiographic appearance 1 year postoperatively (posterior fusion of C2 to C6). **C,** Radiograph obtained 4 years postoperatively showing normal cervical lordosis. **D,** Radiographic appearance 10 years postoperatively. The kyphosis has progressed further, so that forward flexion is restricted. **E,** Radiographic appearance 11 years postoperatively. The patient had developed radicular pain. The dura now was impinged on posteriorly by the cephalad edge of the fusion mass, requiring decompression.



A B

FIGURE 10–24 **A** and **B,** Gradual distraction applied to a halo device by threaded rods in a patient with rigid kyphosis.

even reverse the kyphosis into a more normal cervical lordosis (Fig. 10–23). This is the basis for preferring posterior cervical fusion, if no neurologic deficit requires decompression. In addition, posterior cervical fusion is a technically simpler procedure in a small child and avoids possible respiratory and vascular complications, which are suspected causes of sudden death in patients undergoing anterior cervical fusion (see Fig. 10–22).[80,98] The use of a halo to obtain partial correction of the kyphosis is an important part of the stabilization by posterior fusion. In patients with Larsen's syndrome, mild traction achieved intraoperatively via a halo will usually produce some correction of the kyphosis, owing to the inherent laxity of the neck (see Fig. 10–22B). In diastrophic dysplasia, the deformity is usually more rigid, and we have used gradual extension-distraction of the neck postoperatively using threaded rods in the halo-vest apparatus (Fig. 10–24). With either method, a posterior arthrodesis can be gradually corrected or fixed in situ without concern about displacing an anterior strut graft. Finally, the only instances of pseudarthrosis of posterior cervical fusion in our experience occurred when the patient was not placed in the rigid immobilization provided by the halo-vest apparatus[69] or the patient was less than 1 year old. Autogenous graft should always be used in posterior arthrodesis because of the less favorable dysplastic conditions and incomplete elements.[132]

In patients with established neurologic deficit, either a chronic myelopathy or an acute condition related to trauma (Fig. 10–25), anterior decompression by vertebrectomy is indicated and should be stabilized by some form of anterior strut graft (usually rib or fibula). Posterior cervical fusion should also be performed to prevent increasing deformity from posterior element growth.

In older children there may not be enough growth remaining to expect correction of deformity by a fusion of one column (e.g., posterior). We have no experience in the use of, for example, posterior cervical fusion to correct kyphosis in a child as old as 10 years. It is unlikely that the remaining vertebral growth in a child of this age, especially one with a diagnosis of a syndrome or skeletal dysplasia, would be sufficient that tethering of the posterior column of the spine would produce a reversal of deformity. However, if reversal of a kyphosis has occurred by the age of 10, a symptomatic hyperlordosis may then develop, requiring posterior decompression and fusion mass resection.[70] Such a situation, occurring 11 years postoperatively from posterior cervical fusion, has been reported,[70] although anterior cervical fusion to prevent further lordosis from developing was unnecessary (see Fig. 10–23).

ACQUIRED POSTLAMINECTOMY KYPHOSIS

Conditions that render the posterior cervical facets, ligamentous structures, and bony elements (such as the spinous processes) incompetent or that require their removal can produce cervical kyphosis. The stability of the cervical spine is produced by the combined action of the bony and ligamentous structures to resist tensile forces. The posterior cervical musculature also actively resists kyphotic positioning of the cervical spine. Although localized kyphosis may develop as a result of hypoplasia or destruction of the anterior vertebral bodies (consequent on loss of resistance to compressive forces), loss of functional posterior cervical restraints renders the posterior column unstable, and a vicious cycle of kyphosis ensues as the weightbearing line of the skull translates more anteriorly (Fig. 10–26). The articular facets play an especially important role in preventing the development of cervical kyphosis. Removing as little as 50 percent of the bony facet joint can produce significant instability in flexion and torsion.[25,119,154,155] Finite element analysis has shown that simple resection of one or more spinous processes or posterior ligaments results in enough transfer of tensile forces to the facets to produce eventual failure and kyphosis.[126] Thus it is not surprising that cervical laminectomy performed for management of intraspinal neoplasms or other conditions is associated with an extremely high rate of postlaminectomy kyphosis, especially in children who have undergone cervical or cervicothoracic laminectomy.[7,45,46,152] Efforts to avoid facet injury during laminectomy for decompression of Chiari malformations have been shown to reduce kyphosis significantly.[94]

Normally the weightbearing axis for the cranium lies posterior to the vertebral bodies C2–7 (see Fig. 10–26). As soon as bony or ligamentous instability allows the line of

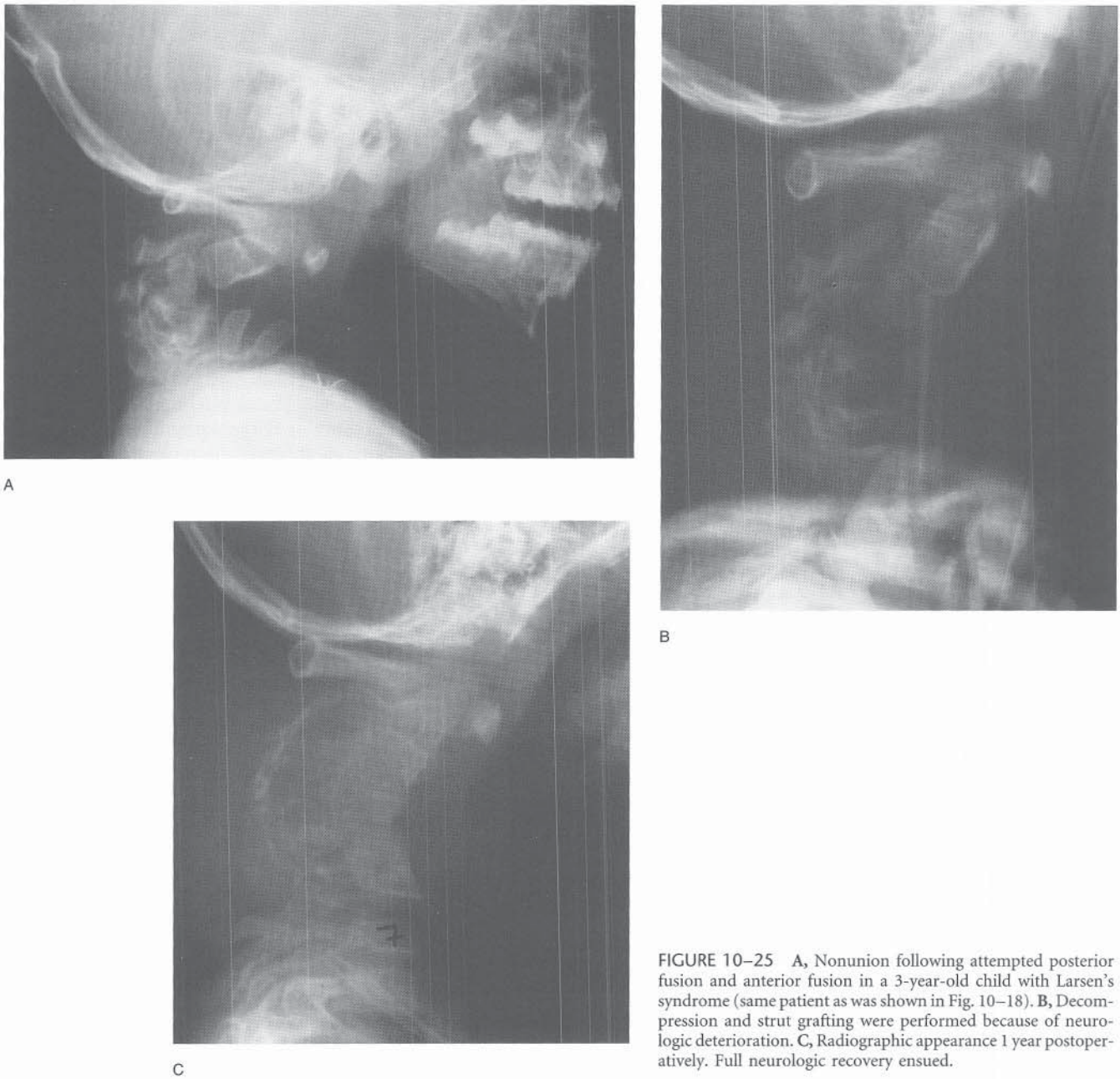


FIGURE 10–25 **A,** Nonunion following attempted posterior fusion and anterior fusion in a 3-year-old child with Larsen's syndrome (same patient as was shown in Fig. 10–18). **B,** Decompression and strut grafting were performed because of neurologic deterioration. **C,** Radiographic appearance 1 year postoperatively. Full neurologic recovery ensued.

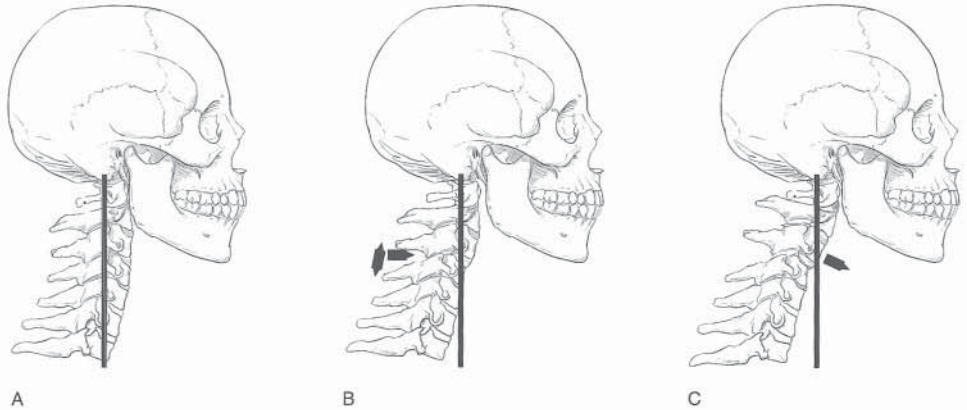


FIGURE 10–26 **A,** Normal cervical alignment. **B,** After destabilization at C3–4 of the posterior column, the weightbearing line shifts anteriorly. **C,** Progression of kyphosis due to continued anterior shift of the weightbearing axis.

cranial weightbearing to move forward, the posterior cervical musculature must act constantly to resist further forward flexion. If the posterior musculature has been denervated or fibrosed due to surgical exposure, the speed with which muscle failure allows progression of the kyphosis may increase. In children, any kyphosis associated with a cervical laminectomy can be rapidly progressive, owing to the combination of muscle weakness and growth. Compression of the spinal cord, due either to direct impingement over the apex of the kyphosis or to ischemia from impairment of the anterior spinal artery vasculature, can produce neurologic deficit. Decompression, when indicated, must be performed anteriorly to relieve these direct bony encroachments on neural tissue.

Cervical kyphosis in children may best be *prevented* by performing simultaneous posterior arthrodesis at the time of laminectomy.[4,15] Osteoplastic laminectomy, either by a trap-door technique or by en bloc removal of the required laminae and their immediate replacement at the end of the procedure, is an alternative to immediate arthrodesis.[117] Replacement of the lamina removed en bloc is undertaken with the intention of producing immediate stability, although long-term follow-up of this procedure is unavailable. If immediate arthrodesis of the laminectomized levels is performed, this can act as a posterior tether, and thus surveillance for the development of hyperlordosis must be carried out, depending on the age at which the laminectomy was performed and the extent of removal of the posterior elements and fusion.

Treatment. Orthotic management of postlaminectomy kyphosis, just as in developmental kyphosis in the cervical spine, is largely ineffective, primarily because the devices used to prevent forward movement of the head require extensive fixation to the thorax, mandible, and occiput and thus are probably tolerated for short periods of time only. Because the long-term management of cervical kyphosis by orthotic means is therefore neither feasible nor proved, the mainstay of treatment of cervical kyphosis is operative.

The first issue in operative stabilization is whether or not neural compression is already present, and if so, whether the compression must be relieved by anterior decompression or whether it is possible to decompress the spinal cord by simple realignment of the cervical spine (orthopaedic decompression). If the deformity is flexible, it may be possible to relieve spinal cord impingement by reducing the deformity. Thus, neurologic assessment of the patient is critical, and any *progression* of the neurologic deficit must be assessed. In the younger patient with a neurologic deficit due to the underlying intraspinal neoplasm or to some other mass lesion (abscess, cyst), it may be difficult to assess recovery by treatment of the primary condition or to document new neurologic deficits related to the kyphosis. Imaging of the cervical spinal cord in positions of flexion and extension (if possible) may show that orthopaedic decompression by realignment is possible.

If there is no clinical or radiographic evidence of spinal cord compression and if the kyphotic deformity has some flexibility, posterior cervical fusion is probably the standard operative procedure. Autograft is superior to allograft for posterior cervical fusion and is recommended whenever possible.[132] Use of a halo-vest device to align the cervical spine is most efficacious in children, and because of the frequent need for continued MRI surveillance of the cervical cord (owing to the original underlying cause), the use of internal fixation, especially stainless steel implants, is probably contraindicated (Fig. 10–27). Wiring techniques using titanium wire may make further MRI possible and can be used to secure onlay grafts or to perform facet wiring.[15,104] In adolescents in whom bone stock is considered adequate for internal fixation, plate fixation using titanium implants with lateral mass screws provides excellent stabilization, often obviating an extension of the fusion to an uninvolved normal level (as is required with onlay bone grafting) and the need for external immobilization postoperatively.[104] Titanium screw-plate implants appear to allow continued MRI with minimum artifact.[125]

If the kyphosis is relatively fixed, without spinal cord compression, anterior cervical release and fusion will be required to provide realignment possibility. This may require simple diskectomy or corpectomy if the bone deformity is extremely rigid, followed by reconstruction of the anterior column with interbody fusion or a strut graft. Either the release or the corpectomy must allow correction into lordosis with traction and/or disk space distraction at the time of surgery. Again, iliac crest autogenous bone graft is recommended to achieve the highest possible rate of union, and halo-vest immobilization is mandatory in smaller children and in adolescents (unless posterior internal fixation is utilized and thought to provide adequate internal fixation).

In the presence of spinal cord compression and neurologic deficit, anterior decompression is required. Typically, a corpectomy may be necessary to achieve full decompression, and, as in the case of kyphosis secondary to syndrome or dysplasia, this must be accompanied by structural realignment of the spine with a strut graft (see Fig. 10–25). In young children, because of the tethering effect of the anterior fusion, posterior fusion should also be performed, in most cases posterior fusion will already have been performed or will be performed during the same operation because of the postlaminectomy deformity. Posterior stabilization and fusion is also indicated in cases of postlaminectomy kyphosis, because of the high rate of pseudarthrosis associated with anterior fusion alone in adults.[154,155] Thus, in postlaminectomy kyphosis with cord compression, the typical scenario would involve anterior cervical decompression by corpectomy, anterior column reconstruction by strut grafting, posterior cervical fusion by onlay or cancellous bone grafting, and maintenance of position by halo-vest external immobilization, or, in the case of adolescents, by posterior cervical instrumentation for internal fixation.

Cervical Instability

Children without obvious deformity can develop symptoms and signs of instability requiring the same evaluations as the syndromes and deformities already discussed. The instability may be inherent to the underlying condition, as with Down syndrome or Klippel-Feil syndrome, or it may be present de novo in an otherwise healthy, unaffected child, as in os odontoideum. Neck pain may be the only symptom of instability. Alternatively, transient or gradually progressive myelopathy, syncope, or radiculopathy may be the neurologic signs provoking an evaluation. As in a workup for

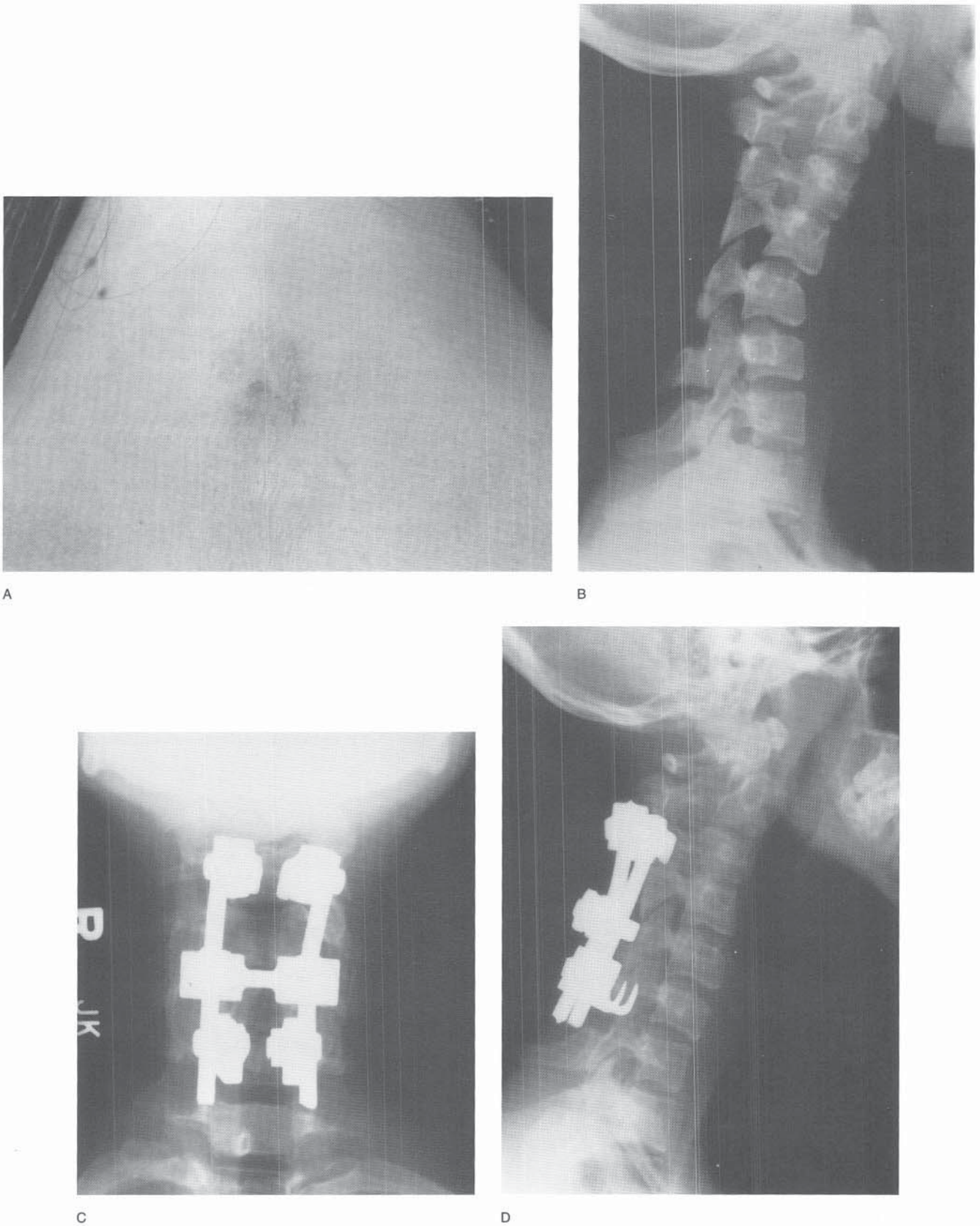


FIGURE 10–27 **A,** Small lesion found at the base of the neck in a 13-year-old girl who presented for scoliosis evaluation. MRI demonstrated a tether extending into the cervical cord. **B,** After C4–5 laminectomy and intradural resection of the fibrous band, she underwent scoliosis correction uneventfully. About 2 years later neck pain and radicular pain developed. **C** and **D,** Radiographic appearance following posterior instrumentation and fusion. Because of the stiffness of the hook-rod implant, no external immobilization was necessary. The girl's symptoms resolved, but the implant was later removed because of local discomfort. Although no further cord imaging was contemplated, MRI would have been technically unsatisfactory with these implants present.

deformity, plain radiography, including a flexion-extension series, is crucial to determine pathologic instability. In the upper cervical spine, excessive translation at the occiput-C1-C2 complex, as measured by the ADI, SAC, and occiput-C1 translation (see previous section on Klippel-Feil syndrome) must be ruled out, while in the subaxial spine, instability can be noted from anterior translation of vertebral bodies or from disruption of the posterior cervical line[137,148] associated with excessive interspinous motion if spinous processes are present (Fig. 10–28). However, awareness of pseudosubluxation at C2–3 and C3–4[17,137] must be maintained to avoid treating normal physiologic motion at these levels in children 7 years or younger. Once pathologic instability has been identified on plain radiographs, additional imaging, particularly flexion-extension MRI studies,[144] is indicated to identify exact points of compression and to select treatment.

OS ODONTOIDEUM

This anomaly, in which the upper portion of the odontoid is separated from the base by a gap resembling an un-united fracture, probably represents just that—an unrecognized fracture occurring at a young age that fails to heal because of lack of immobilization or interruption of blood supply to the upper segment.[42,149] It is often diagnosed following an episode of neck pain or, more rarely, an episode of paresis. It should always be considered in the differential diagnosis of cerebral palsy when a significant birth history is lacking (Fig. 10–29).

Radiographically the os resembles an ossicle with a smooth sclerotic border located at the normal position for the tip of the odontoid, with a gap wide enough to indicate an established nonunion. The possibility of a congenital anomaly cannot be ruled out, as there is rarely a reliable history of trauma, for example, to establish an etiology. CT or plain tomography may help delineate the lesion.

Neurologic symptoms, if any, result from cord impingement by posterior translation of the os in extension or anterior odontoid impingement in flexion. Thus, instability due to loss of integrity of the entire dens is required.

The natural history of os odontoideum is unknown. Usually, transient symptoms or neurologic signs resolve with cervical immobilization and avoidance of provocative activity. However, permanent nonstrenuous activity limitation in children usually is unacceptable and unenforceable, and the risk of catastrophic injury from either a seemingly trivial bump on the head or from whiplash in an automobile accident cannot realistically be determined. Thus, the long-term safety of such a patient may indicate a C1–C2 arthrodesis. Certainly an ADI of 10 mm or an SAC of 13 mm,[131] even in an asymptomatic patient, would be an indication for fusion, in addition to persistent symptoms or progressive neurologic signs.

Traditionally a Gallie-type posterior C1–C2 fusion has been recommended, with extra care to avoid total excessive C1 posterior translation during wire tightening (see Fig. 10–29). In small children, suture may be used. Halo immobilization for 6 to 12 weeks will increase the likelihood of successful fusion in otherwise uncooperative patients.

OCCIPITAL-ATLANTAL ANOMALIES

Patients with *typical* Klippel-Feil syndrome are easily recognized by the characteristic short, broad neck, low hairline, and restricted cervical motion. In a separate group are patients who lack the physical signs of Klippel-Feil syndrome but who have similar occipitocervical malformations, estimated to occur in as many as 25 percent of children (see Fig. 10–11).[88] A common pattern—assimilation of C1 into

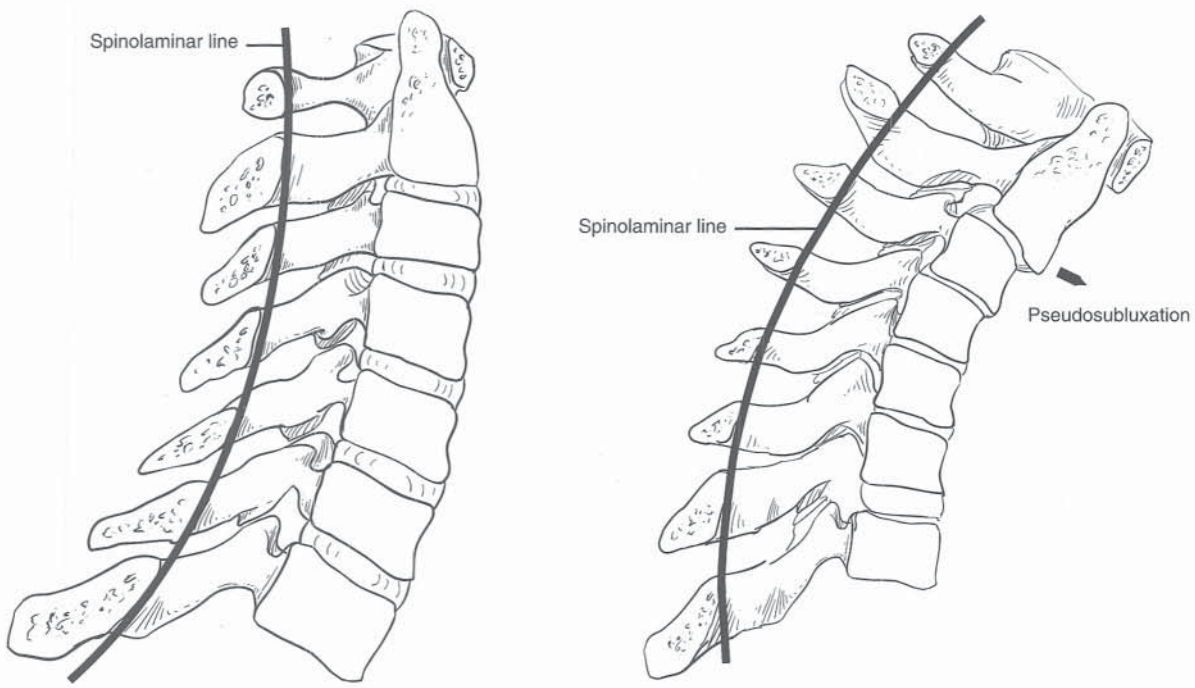
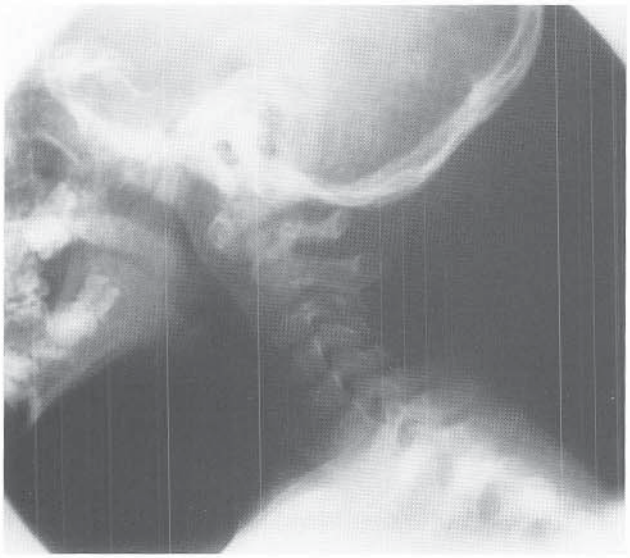
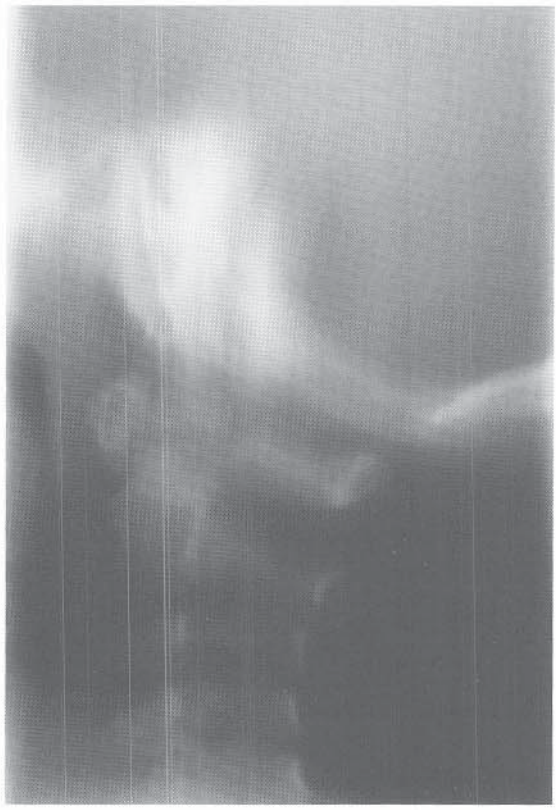


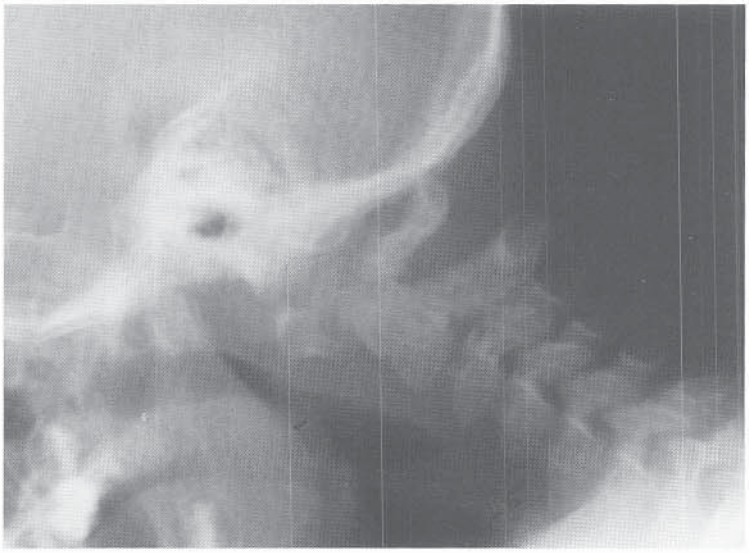
FIGURE 10–28 Pseudosubluxation of C2–3 (most common). Possible subluxation is eliminated because of the intact spinolaminar line at C2–3.



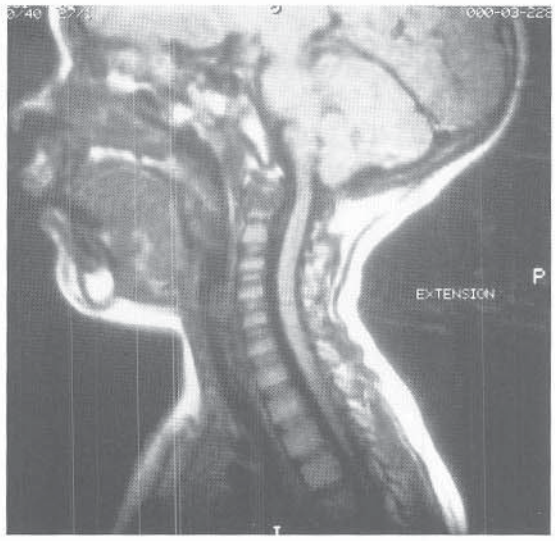
A



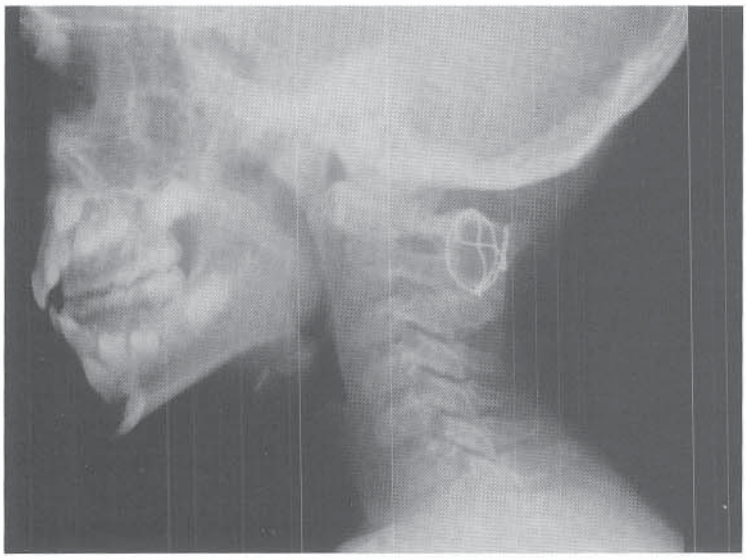
B



C



D



E

FIGURE 10–29 Os odontoideum. **A,** Radiographic appearance in a child age 4 years 8 months who presented for an AFO prescription for hemiparesis. She had no significant birth history and had walked at a normal age. Neck stiffness and pain were present. **B,** Tomograph demonstrating os odontoideum. **C,** Instability on flexion. **D,** MR image demonstrating lack of cord impingement in extension. **E,** Gallie fusion in extension was performed. There was no immediate change in the hemiparesis.

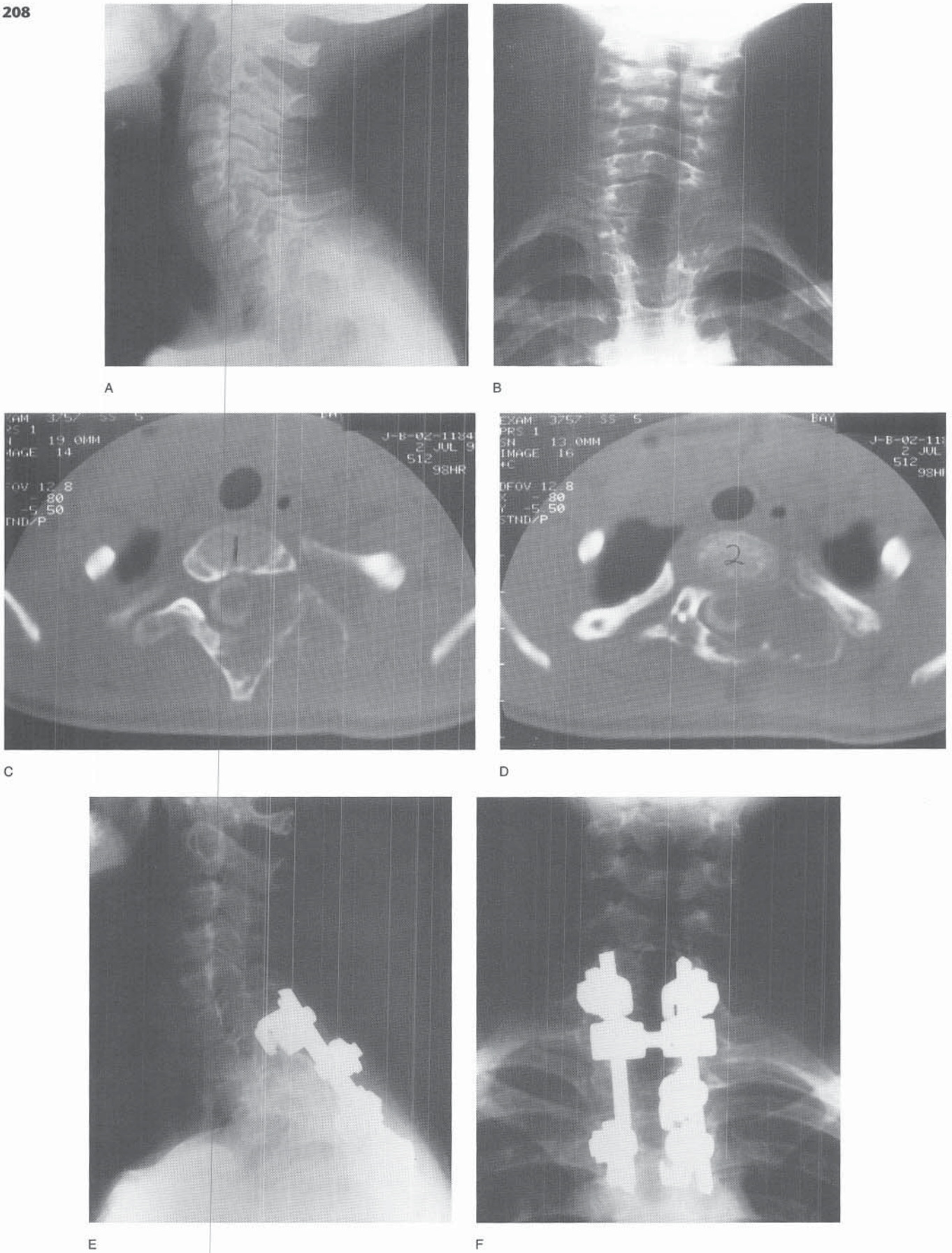


FIGURE 10–30 **A** and **B,** Radiographs of a 10-year-old girl with right shoulder pain and weakness. Expansile lesions of the spinous and transverse processes of T1 and T2 are present. **C** and **D,** CT-myelographs demonstrating expansile, lytic lesions of the T1 and T2 posterior elements on the right, encroaching on the canal. **E** and **F,** Aneurysmal bone cyst was diagnosed by biopsy. Decompression and excision of the right posterior elements (including pedicles) was performed, with simultaneous instrumentation and fusion. All symptoms resolved.

the occiput combined with a C2–3 synostosis—puts the patient at risk for C1–2 instability, as this joint may suffer gradual increased laxity due to lack of mobility of the segments immediately adjacent.[96,108]

Treatment for occipitocervical malformations depends on the amount of C1–C2 instability and the presence of neurologic signs. Excessive mobility without neurologic signs can be managed by education, decreased activity, and careful observation. Progression of instability or a decreasing SAC should probably be treated similar to os odontoideum, with fusion in extension, thus completing the patient's synostosis from the occiput to C3. If the ring of C1 is incompletely assimilated, formal fusion to the occiput by separate grafting should be included.

If neurologic deficit exists, decompression with stabilization is indicated. As in Klippel-Feil patients, decompression must address the site of cord impingement—anterior odontoid excision or posterior C1 laminectomy, as necessary.

ODONTOID ANOMALIES/LIGAMENTOUS INSTABILITY

Aplasia and hypoplasia of the odontoid occur routinely in a variety of skeletal dysplasias, most notably in the spondyloepiphyseal dysplasia group. Mucopolysaccharidosis storage diseases, such as Hunter's, Hurler's, Morquio's, or Maroteau-Lamy syndromes, have a similar degree of odontoid involvement. Every patient with one of these diagnoses should be evaluated every 1 to 2 years with plain radiography for C1–2 instability. As with all upper cervical instabilities, fusion prior to the development of neurologic signs must be considered, to avoid possible catastrophic injury. Decompression and fusion once myelopathy is present may stabilize but not reverse the neurologic deficit.

Ligamentous laxity associated with connective tissue disorders and Down syndrome is also well known to lead to upper cervical instability. In the latter condition, both occiput-C1 as well as C1–2 are known to be at risk. Specific management of upper cervical instability in Down syndrome is discussed in Chapter 30, Orthopaedic-Related Syndromes.

Instability may also accompany deformity in dysplastic bone conditions (osteogenesis imperfecta, neurofibromatosis). Such instability is investigated by flexion-extension radiographs, just as in any other condition.

NONTRAUMATIC OCCIPITAL-ATLANTAL INSTABILITY

Five cases of idiopathic occipital-C1 instability associated with a variety of neurologic signs and symptoms (vertigo, syncope, projectile vomiting) have been reported.[50] The symptoms were presumably due to vertebral artery insufficiency related to the occipital-C1 mobility. The instability was documented with cineradiography. Symptoms resolved with successful posterior occipital-atlantal fusion.

SUBAXIAL INSTABILITY

Probably the most common reason for instability in the lower cervical spine is a previous *laminectomy*. As was discussed for acquired kyphosis, the stability of the cervical spine can be significantly compromised by the simple resection of one or more spinous processes[126] and further destabilized by any facetectomy.[25,119,154,155] Cervical laminectomy for any reason can produce deformity in children.[7,45,46,152] Because instability precedes the development of deformity, the patient may present with symptoms (pain, radiculopathy, apprehension) before actual deformity occurs. The diagnosis is confirmed by flexion-extension radiographs.

Trauma is another cause of instability. In the subaxial spine, the typical presentation is an older child or adolescent with a history of a fall, a blow to the head, or other injury where symptoms resolved shortly after injury but then recurred. This scenario is covered in Chapter 40, Spinal Injuries.

Any treatment of *tumors* and *tumor-like conditions* in which bone must be resected or ligamentous stability removed during exposure is also likely to produce instability.

Aneurysmal bone cyst, osteoblastoma, or osteochondroma involving posterior elements are the tumors most likely to be encountered in the first and second decades of life, and surgical treatment because of pain or encroachment on the canal would then require stabilization. In general, the posterior structures of the spine can be considered as four separate functioning units—midline structures (spinous processes, laminae, interspinous ligaments), the left and right facet joints, and the posterior vertebral body wall and/or disk (the "middle" column).[49] Compromise of any two of these four structured units is an indication for stabilization by internal or external fixation, or both, as well as for performing fusion (Fig. 10–30).

ACKNOWLEDGMENTS

The author wishes to acknowledge the following colleagues who contributed to this chapter: Richard T. Holt, James R. Sackett.

REFERENCES

1. American Association of Orthopaedic Surgeons: Cervical Spine Deformities. Orthopaedic Knowledge Update 5, p 563. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.
2. Andersen PE Jr, Justesen P: Chondrodysplasia punctata: report of two cases. Skeletal Radiol 1987;16:223.
3. Anderson W: Clinical lecture on sternomastoid torticollis. Lancet 1893;1:9.
4. Aronson DD, Kahn RH, Canady A, et al: Instability of the cervical spine after decompression in patients who have Arnold-Chiari malformation. J Bone Joint Surg 1991;73-A:898.
5. Barber FA, Roach JW: Torticollis: a presentation of eosinophilic granuloma. A case report. Orthopedics 1986;9:1237.
6. Baum JA, Hanley EN Jr, Pullekines J: Comparison of halo complications in adults and children. Spine 1989;14:251.
7. Bell DF, Walker JL, O'Connor G, et al: Spinal deformity after multiple-level cervical laminectomy in children. Spine 1994;19:406.
8. Bethem D, Winter RB, Lutter L: Disorders of the spine in diastrophic dwarfism. J Bone Joint Surg 1980;62-A:529.
9. Binder H, Eng GD, Gaiser JF, et al: Congenital muscular torticollis: results of conservative management with long-term follow-up in 85 cases. Arch Phys Med Rehabil 1987;68:222.
10. Bixenman WW: Diagnosis of superior oblique palsy. J Clin Neuroophthalmol 1981;1:199.
11. Boisen E: Torticollis caused by an infratentorial tumour: three cases. Br J Psychiatry 1979;134:306.
12. Brooks AL, Jenkins EB: Atlanto-axial arthrodesis by the wedge compression method. J Bone Joint Surg 1978;60-A:279.
13. Burkus JK, Deponte RJ: Chronic atlantoaxial rotatory fixation correction by cervical traction, manipulation, and bracing. J Pediatr Orthop 1986;6:631.

14. Caetano de Barros M, Farias W, Ataide L, et al: Basilar impression and Arnold-Chiari malformation: a study of 66 cases. J Neurol Neurosurg Psychiatry 1968;31:596.
15. Callahan RA, Johnson RM, Margolis RN, et al: Cervical facet fusion for control of instability following laminectomy. J Bone Joint Surg 1977;59-A:991.
16. Canale ST, Griffin DW, Hubbard CN: Congenital muscular torticollis: a long-term follow-up. J Bone Joint Surg 1982;64-A:810.
17. Catell HS, Fittzer DL: Pseudosubluxation and other normal variations in the cervical spine in children. J Bone Joint Surg 1965;47-A:1295.
18. Cheng JC, Tang SP: Outcome of surgical treatment of congenital muscular torticollis. Clin Orthop 1999;362:190.
19. Copley LA, Pepe MD, Tan V, et al: A comparison of various angles of halo pin insertion in an immature skull model. Spine 1999;24:1777.
20. Coscia MF, Bassett GS, Bowen JR, et al: Spinal abnormalities in camptomelic dysplasia. J Pediatr Orthop 1989;9:6.
21. Coventry MB, Harris LE: Congenital muscular torticollis in infancy: some observations regarding treatment. J Bone Joint Surg 1959;41-A:815.
22. Craig JB, Govender S: Neurofibromatosis of the cervical spine: a report of eight cases. J Bone Joint Surg 1992;74-B:575.
23. Crawford AH, Schorry EK: Neurofibromatosis in children: the role of the orthopaedist. J Am Acad Orthop Surg 1999;7:217.
24. Curtis BH, Fisher RL, Butterfield WL, et al: Neurofibromatosis with paraplegia: report of eight cases. J Bone Joint Surg 1969;51-A:843.
25. Cusick JF, Yoganandan N, Pintar F, et al: Biomechanics of cervical spine facetectomy and fixation techniques. Spine 1988;13:808.
26. da Silva JA: Basilar impression and the Arnold-Chiari malformation: surgical findings in a series of 209 cases. Neurochirurgia 1992;35:189.
27. Davey JR, Rorabeck CH, Bailey SI, et al: A technique of posterior cervical fusion for instability of the cervical spine. Spine 1985;10:722.
28. Davids JR, Wenger DR, Mubarak SJ: Congenital muscular torticollis: sequela of intrauterine or perinatal compartment syndrome. J Pediatr Orthop 1993;13:141.
29. Davidson RI, Shillito J Jr: Eosinophilic granuloma of the cervical spine in children. Pediatrics 1970;45:746.
30. Dormans JP, Criscitiello AA, Drummond DS, et al: Complications in children managed with immobilization in a halo vest. J Bone Joint Surg 1995;77-A:1370.
31. Dormans JP, Drummond DS, Sutton LN, et al: Occipitocervical arthrodesis in children: a new technique and analysis of results. J Bone Joint Surg 1995;77-A:1234.
32. Drummond DS: Congenital anomalies of the pediatric cervical spine. In Bridwell KH, DeWald RL (eds): The Textbook of Spinal Surgery, p 951. Philadelphia, Lippincott-Raven, 1997.
33. Drvaric DM, Ruderman RJ, Conrad RW, et al: Congenital scoliosis and urinary tract abnormalities: are intravenous pyelograms necessary? J Pediatr Orthop 1987;7:441.
34. Dubousset J: Torticollis in children caused by congenital anomalies of the atlas. J Bone Joint Surg 1986;68-A:178.
35. Dure LS, Percy AK, Cheek WR, et al: Chiari type I malformation in children. J Pediatr 1989;115:573.
36. El-Khoury GY, Clark CR, Gravett AW: Acute traumatic rotatory atlanto-axial dislocation in children: a report of three cases. J Bone Joint Surg 1984;66-A:774.
37. Enneking WF: Musculoskeletal Tumor Surgery. New York, Churchill Livingstone, 1983.
38. Eyring EJ, Peterson CA, Bjornson DR: Intervertebral disc calcification in childhood. J Bone Joint Surg 1964;46-A:1432.
39. Ferkel RD, Westin GW, Dawson EG, et al: Muscular torticollis: a modified surgical approach. J Bone Joint Surg 1983;65-A:894.
40. Fielding JW: Normal and selected abnormal motion of the cervical spine from the second cervical vertebra to the seventh cervical vertebra based on cineroentgenography. J Bone Joint Surg 1964;46-A:1779.
41. Fielding JW, Hawkins RJ: Atlanto-axial rotatory fixation (fixed rotatory subluxation of the atlanto-axial joint). J Bone Joint Surg 1977;59-A:37.
42. Fielding JW, Hensinger RN, Hawkins RJ: Os odontoideum. J Bone Joint Surg 1980;62-A:376.
43. Fielding JW, Stillwell WT, Chynn KY, et al: Use of computed tomography for the diagnosis of atlanto-axial rotatory fixation: a case report. J Bone Joint Surg 1978;60-A:1102.
44. Forese LL, Berdon WE, Harcke HT, et al: Severe mid-cervical kyphosis with cord compression in Larsen's syndrome and diastrophic dysplasia: unrelated syndromes with similar radiologic findings and neurosurgical implications. Pediatr Radiol 1995;25:136.
45. Francis WR Jr, Noble DP: Treatment of cervical kyphosis in children. Spine 1988;13:883.
46. Fraser RD, Paterson DC, Simpson DA: Orthopaedic aspects of spinal tumors in children. J Bone Joint Surg 1977;59-B:143.
47. Fried JA, Athreya B, Gregg JR, et al: The cervical spine in juvenile rheumatoid arthritis. Clin Orthop 1983;179:102.
48. Gallie WE: Fractures and dislocations of the cervical spine. Am J Surg 1939;46:495.
49. Gelb DE, Bridwell KH: Benign tumors of the spine. In Bridwell KH, DeWald RL (eds): The Textbook of Spinal Surgery, p 1959. Philadelphia, Lippincott-Raven, 1997.
50. Georgopoulos G, Pizzutillo PD, Lee MS: Occipito-atlantal instability in children: a report of five cases and review of the literature. J Bone Joint Surg 1987;69-A:429.
51. Gilbert EF, Opitz JM, Spranger JW, et al: Chondrodysplasia punctata—rhizomelic form: pathologic and radiologic studies of three infants. Eur J Pediatr 1976;123:89.
52. Giuffre R, Di Lorenzo N, Fortuna A: Cervical tumors of infancy and childhood. J Neurosurg Sci 1981;25:259.
53. Goldberg MJ: The Dysmorphic Child: An Orthopedic Perspective. New York, Raven Press, 1987.
54. Gray SW, Romaine CB, Skandalakis JE: Congenital fusion of the cervical vertebrae. Surg Gynecol Obstet 1964:373.
55. Green NE, Robertson WW Jr, Kilroy AW: Eosinophilic granuloma of the spine with associated neural deficit: report of three cases. J Bone Joint Surg 1980;62-A:1198.
56. Grisel P: Enucleation de l'atlas et torticolis naso-pharyngien. Presse Med 1930:50.
57. Gunderson CH, Solitare GB: Mirror movements in patients with the Klippel-Feil syndrome: neuropathologic observations. Arch Neurol 1968;18:675.
58. Hall JE, Simmons ED, Danylchuk K, et al: Instability of the cervical spine and neurological involvement in Klippel-Feil syndrome: a case report. J Bone Joint Surg 1990;72-A:460.
59. Halla JT, Fallahi S, Hardin JG: Nonreducible rotational head tilt and atlantoaxial lateral mass collapse: clinical and roentgenographic features in patients with juvenile rheumatoid arthritis and ankylosing spondylitis. Arch Intern Med 1983;143:471.
60. Heard GE, Holt JF, Naylor B: Cervical vertebral deformity in von Recklinghausen's disease of the nervous system. J Bone Joint Surg 1962;44-B:880.
61. Hensinger RN, De Vito PD, Ragsdale CG: Changes in the cervical spine in juvenile rheumatoid arthritis. J Bone Joint Surg 1986;68-A:189.
62. Hensinger RN, Lang JE, MacEwen GD: Klippel-Feil syndrome: a constellation of associated anomalies. J Bone Joint Surg 1974;56-A:1246.
63. Herring JA, Hensinger RN: Cervical disc calcification. J Pediatr Orthop 1988;8:613.
64. Herring JA, Johnston CE II: Lytic lesion of C2. J Pediatr Orthop 1987;7:481.
65. Hummer CD, MacEwen GD: The coexistence of torticollis and congenital dysplasia of the hip. J Bone Joint Surg 1972;54-A:1255.
66. Ippolito E, Tudisco C, Massobrio M: Long-term results of open sternocleidomastoid tenotomy for idiopathic muscular torticollis. J Bone Joint Surg 1985;67-A:30.
67. Isu T, Miyasaka K, Abe H, et al: Atlantoaxial dislocation associated with neurofibromatosis: report of three cases. J Neurosurg 1983;58:451.
68. Jeanneret B, Magerl F: Primary posterior fusion of C1/2 in odontoid fractures: indications, technique, and results of transarticular screw fixation. J Spinal Disord 1992;5:464.
69. Johnston CE II, Birch JG, Daniels JL: Cervical kyphosis in patients who have Larsen's syndrome. J Bone Joint Surg 1996;78-A:538.
70. Johnston CE II, Schoenecker PL: Cervical kyphosis in patients who have Larsen syndrome [letter]. J Bone Joint Surg 1997;79-A:1590.
71. Jolley SG, Johnson DG, Herbst JJ, et al: An assessment of gastroesophageal reflux in children by extended pH monitoring of the distal esophagus. Surgery 1978;84:16.
72. Jones RN: Rotatory dislocation of both atlanto-axial joints. J Bone Joint Surg 1984;66-B:6.
73. Kash IJ, Sane SM, Samaha FJ, et al: Cervical cord compression in diastrophic dwarfism. J Pediatr 1974;84:862.

74. Kawabe N, Hirotani H, Tanaka O: Pathomechanism of atlantoaxial rotatory fixation in children. J Pediatr Orthop 1989;9:569.
75. Keuter EJ: Non-traumatic atlanto-axial dislocation associated with nasopharyngeal infections (Grisel's disease). Acta Neurochir (Wien) 1969;21:11.
76. Kiwak KJ, Deray MJ, Shields WD: Torticollis in three children with syringomyelia and spinal cord tumor. Neurology 1983;33:946.
77. Klippel M, Feil A: Un cas d'absence des vertebres cervicales, cage thoracique remontant jusquia la base du crane. Nouv Iconog Salpetrieve 1912;25:223.
78. Kokubun S, Ozawa H, Sakurai M, et al: One-stage anterior and posterior correction of severe kyphosis of the cervical spine in neurofibromatosis: a case report. Spine 1993;18:2332.
79. Lamy M, Maroteaux P: Le nanisine diastrophique. Presse Med 1960;68:1977.
80. Larsen LJ, Schottstaedt ER, Bost FC: Multiple congenital dislocations associated with characteristic facial abnormality. J Pediatr 1950;37:574.
81. Larsen WJ: Human Embryology. New York, Churchill Livingstone, 1993.
82. Lee CK, Weiss AB: Isolated congenital cervical block vertebrae below the axis with neurological symptoms. Spine 1981;6:118.
83. Lee EH, Kang YK, Bose K: Surgical correction of muscular torticollis in the older child. J Pediatr Orthop 1986;6:585.
84. Letts M, Kaylor D, Gouw G: A biomechanical analysis of halo fixation in children. J Bone Joint Surg 1988;70-B:277.
85. Ling CM: The influence of age on the results of open sternomastoid tenotomy in muscular torticollis. Clin Orthop 1976;116:142.
86. Locke GR, Gardner JI, Van Epps EF: Atlas-dens interval (ADI) in children: a survey based on 200 normal cervical spines. Am J Roentgenol Radium Ther Nucl Med 1966;97:135.
87. Loder RT: The cervical spine. In Morrissy RT, Weinstein SL (eds): Lovell and Winters Pediatric Orthopaedics, p 739. Philadelphia, Lippincott-Raven, 1996.
88. Macalister A: Notes on the development and variations of the atlas. J Anat Physiol 1983;27:519.
89. MacDonald C: Sternomastoid tumor and muscular torticollis. J Bone Joint Surg 1969;51-B:432.
90. Mah JY, Thometz J, Emans J, et al: Threaded K-wire spinous process fixation of the axis for modified Gallie fusion in children and adolescents. J Pediatr Orthop 1989;9:675.
91. Manak JR, Scott MP: A class act: conservation of homeodomain protein functions. Dev Suppl 1994:61.
92. Mathern GW, Batzdorf U: Grisel's syndrome: cervical spine clinical, pathologic, and neurologic manifestations. Clin Orthop 1989;244:131.
93. McAfee PC, Cassidy JR, Davis RF, et al: Fusion of the occiput to the upper cervical spine: a review of 37 cases. Spine 1991;16:S490.
94. McLaughlin MR, Wahlig JB, Pollack IF: Incidence of postlaminectomy kyphosis after Chiari decompression [published erratum appears in Spine 1997;22(11):1276]. Spine 1997;22:613.
95. McLay K, Maran AG: Deafness and the Klippel-Feil syndrome. J Laryngol Otol 1969;83:175.
96. McRae DL: Bony abnormalities in the region of the foramen magnum: correlation of the anatomic and neurologic findings. Acta Radiol 1953;40:335.
97. Mecklenburg RS, Krueger PM: Extensive genitourinary anomalies associated with Klippel-Feil syndrome. Am J Dis Child 1974;128:92.
98. Micheli LJ, Hall JE, Watts HG: Spinal instability in Larsen's syndrome: report of three cases. J Bone Joint Surg 1976;58-A:562.
99. Minamitani K, Inoue A, Okuno T: Results of surgical treatment of muscular torticollis for patients greater than 6 years of age. J Pediatr Orthop 1990;10:754.
100. Mitchell TC, Sadasivan KK, Ogden AL, et al: Biomechanical study of atlantoaxial arthrodesis: transarticular screw fixation versus modified Brooks posterior wiring. J Orthop Trauma 1999;13:483.
101. Moore WB, Matthews TJ, Rabinowitz R: Genitourinary anomalies associated with Klippel-Feil syndrome. J Bone Joint Surg 1975;57-A:355.
102. Morrison DL, MacEwen GD: Congenital muscular torticollis: observations regarding clinical findings, associated conditions, and results of treatment. J Pediatr Orthop 1982;2:500.
103. Morrison SG, Perry LW, Scott LP III: Congenital brevicollis (Klippel-Feil syndrome) and cardiovascular anomalies. Am J Dis Child 1968;115:614.
104. Moskovich R: Cervical instability (rheumatoid dwarfism, degenerative, others). In Bridwell KH, DeWald RL (eds): The Textbook of Spinal Surgery, p 969. Philadelphia, Lippincott-Raven, 1997.
105. Mubarak SJ, Camp JF, Vuletich W, et al: Halo application in the infant. J Pediatr Orthop 1989;9:612.
106. Murphy WJ, Gellis SS: Torticollis with hiatus hernia in infancy: Sandifer syndrome. Am J Dis Child 1977;131:564.
107. Muzumdar AS, Lowry RB, Robinson CE: Quadriplegia in Larsen syndrome. Birth Defects Orig Artic Ser 1977;13:202.
108. Nicholson JT, Sherk HH: Anomalies of the occipitocervical articulation. J Bone Joint Surg 1968;50-A:295.
109. Nora JJ, Cohen M, Maxwell GM: Klippel-Feil syndrome. Am J Dis Child 1961;102:110.
110. Ogden JA, Grogan DP: Prenatal growth and development of the musculoskeletal system. In Albright JA, Brand RA (eds): The Scientific Basis of Orthopaedics, p 47. New York, Appleton & Lange, 1987.
111. Ono K, Yonenobu K, Fuji T, et al: Atlantoaxial rotatory fixation: radiographic study of its mechanism. Spine 1985;10:602.
112. O'Rahilly R, Meyer DB: The timing and sequence of events in the development of the human vertebral column during the embryonic period proper. Anat Embryol (Berl) 1979;157:167.
113. Parke WW, Rothman RH, Brown MD: The pharyngovertebral veins: an anatomical rationale for Grisel's syndrome. J Bone Joint Surg 1984;66-A:568.
114. Pennecot GF, Leonard P, Peyrot Des Gachons S, et al: Traumatic ligamentous instability of the cervical spine in children. J Pediatr Orthop 1984;4:339.
115. Phillips WA, Hensinger RN: The management of rotatory atlantoaxial subluxation in children. J Bone Joint Surg 1989;71-A:664.
116. Poussa M, Merikanto J, Ryoppy S, et al: The spine in diastrophic dysplasia. Spine 1991;16:881.
117. Raimondi AJ, Gutierrez FA, Di Rocco C: Laminotomy and total reconstruction of the posterior spinal arch for spinal canal surgery in childhood. J Neurosurg 1976;45:555.
118. Ramenofsky ML, Buyse M, Goldberg MJ, et al: Gastroesophageal reflux and torticollis. J Bone Joint Surg 1978;60-A:1140.
119. Raynor RB, Pugh J, Shapiro I: Cervical facetectomy and its effect on spine strength. J Neurosurg 1985;63:278.
120. Remes V, Marttinen E, Poussa M, et al: Cervical kyphosis in diastrophic dysplasia. Spine 1999;24:1990.
121. Ring D, Johnston CE II, Wenger DR: Pyogenic infectious spondylitis in children: the convergence of discitis and vertebral osteomyelitis. J Pediatr Orthop 1995;15:652.
122. Ritterbusch JF, McGinty LD, Spar J, et al: Magnetic resonance imaging for stenosis and subluxation in Klippel-Feil syndrome. Spine 1991; 16:S539.
123. Rodgers WB, Coran DL, Emans JB, et al: Occipitocervical fusions in children: retrospective analysis and technical considerations. Clin Orthop 1999;364:125.
124. Rouvreau P, Glorion C, Langlais J, et al: Assessment and neurologic involvement of patients with cervical spine congenital synostosis as in Klippel-Feil syndrome: study of 19 cases. J Pediatr Orthop B 1998;7:179.
125. Rupp R, Ebraheim NA, Savolaine ER, et al: Magnetic resonance imaging evaluation of the spine with metal implants: general safety and superior imaging with titanium. Spine 1993;18:379.
126. Saito T, Yamamuro T, Shikata J, et al: Analysis and prevention of spinal column deformity following cervical laminectomy. I. Pathogenetic analysis of postlaminectomy deformities. Spine 1991;16:494.
127. Sarnat HB, Morrissy RT: Idiopathic torticollis: sternocleidomastoid myopathy and accessory neuropathy. Muscle Nerve 1981;4:374.
128. Sherk HH, Shut L, Chung S: Iniencephalic deformity of the cervical spine with Klippel-Feil anomalies and congenital elevation of the scapula: report of three cases. J Bone Joint Surg 1974;56-A:1254.
129. Snyder CH: Paroxysmal torticollis in infancy: a possible form of labyrinthitis. Am J Dis Child 1969;117:458.
130. Sonnabend DH, Taylor TK, Chapman GK: Intervertebral disc calcification syndromes in children. J Bone Joint Surg 1982;64-B:25.
131. Spierings EL, Braakman R: The management of os odontoideum: analysis of 37 cases. J Bone Joint Surg 1982;64-B:422.
132. Stabler CL, Eismont FJ, Brown MD, et al: Failure of posterior cervical fusions using cadaveric bone graft in children. J Bone Joint Surg 1985;67-A:371.
133. Stark EW, Borton TE: Hearing loss and the Klippel-Feil syndrome. Am J Dis Child 1972;123:233.
134. Stone JW, Bridwell KH, Shackelford GD, et al: Dural ectasia associated

with spontaneous dislocation of the upper part of the thoracic spine in neurofibromatosis: a case report and review of the literature [published erratum appears in J Bone Joint Surg 1988;70-A:312]. J Bone Joint Surg 1987;69-A:1079.
135. Subramanian V, Meyer BI, Gruss P: Disruption of the murine homeobox gene Cdx1 affects axial skeletal identities by altering the mesodermal expression domains of Hox genes. Cell 1995;83:641.
136. Svensson O, Aaro S: Cervical instability in skeletal dysplasia: report of 6 surgically fused cases. Acta Orthop Scand 1988;59:66.
137. Swischuk LE: Anterior displacement of C2 in children: physiologic or pathologic. Radiology 1977;122:759.
138. Tachdjian MO: Congenital muscular torticollis. In Tachdjian MO (ed): Tachdjian's Pediatric Orthopaedics, 2nd ed, p 112. Philadelphia, WB Saunders Co, 1990.
139. Tredwell SJ, Newman DE, Lockitch G: Instability of the upper cervical spine in Down syndrome. J Pediatr Orthop 1990;10:602.
140. Visudhiphan P, Chiemchanya S, Somburanasin R, et al: Torticollis as the presenting sign in cervical spine infection and tumor. Clin Pediatr (Phila) 1982;21:71.
141. Walker BA, Scott CI, Hall JG, et al: Diastrophic dwarfism. Medicine (Baltimore) 1972;51:41.
142. Ward BA, Harkey HL, Parent AD, et al: Severe cervical kyphotic deformities in patients with plexiform neurofibromas: case report. Neurosurgery 1994;35:960.
143. Weiner DS: Congenital dislocation of the hip associated with congenital muscular torticollis. Clin Orthop 1976;121:163.
144. Weng MS, Haynes RJ: Flexion and extension cervical MRI in a pediatric population. J Pediatr Orthop 1996;16:359.
145. Wetzel FT, La Rocca H: Grisel's syndrome. Clin Orthop 1989;240:141.
146. White AA, Punjabi MM: Clinical Biomechanics of the Spine. Philadelphia, JB Lippincott Co, 1978.
147. Wiesel SW, Rothman RH: Occipitoatlantal hypermobility. Spine 1979;4:187.
148. Williams CF, Bernstein TW, Jelenko CD: Essentiality of the lateral cervical spine radiograph. Ann Emerg Med 1981;10:198.
149. Wollin DG: The os odontoideum: separate odontoid process. J Bone Joint Surg 1963;45-A:1459.
150. Wong CC, Pereira B, Pho RW: Cervical disc calcification in children: a long-term review. Spine 1992;17:139.
151. Wong WB, Haynes RJ: Osteology of the pediatric skull: Considerations of halo pin placement. Spine 1994;19:1451.
152. Yasuoka S, Peterson HA, MacCarty CS: Incidence of spinal column deformity after multilevel laminectomy in children and adults. J Neurosurg 1982;57:441.
153. Yong-Hing K, Kalamchi A, MacEwen GD: Cervical spine abnormalities in neurofibromatosis. J Bone Joint Surg 1979;61-A:695.
154. Zdeblick TA, Abitbol JJ, Kunz DN, et al: Cervical stability after sequential capsule resection. Spine 1993;18:2005.
155. Zdeblick TA, Zou D, Warden KE, et al: Cervical stability after foraminotomy: a biomechanical in vitro analysis. J Bone Joint Surg 1992; 74-A:22.