CHAPTER 19

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Kyphosis

Kyphosis is a curvature of the spine in the sagittal plane in which the convexity of the curve is directed posteriorly. Lordosis is a curvature of the spine in the sagittal plane in which the convexity of the curve is directed anteriorly. The thoracic spine and the sacrum normally are kyphotic, and the cervical spine and the lumbar spine normally are lordotic (1). Although several authors have tried to define normal kyphosis of the thoracic spine and normal lordosis of the lumbar spine, these studies have shown much variability in what is considered normal (2–8). The ranges of normal kyphosis and lordosis change with increasing age and vary according to the gender and the area of the spine involved (2–5). The degree of kyphosis or lordosis that is considered normal or abnormal depends on the location of the curvature and the age of the patient. For example, 30 degrees of kyphosis is normal in the thoracic spine but abnormal at the thoracolumbar junction.

The normal range of thoracic kyphosis is considered to be 19 to 45 degrees and that of lumbar lordosis, 30 to 60 degrees (9). Measurements of kyphosis and lordosis are made from standard scoliosis radiographs with the patient standing with his or her knees locked, feet shoulder width apart, elbows bent, and knuckles in the supraclavicular fossa bilaterally. This will place the patient's arms at approximately a 45-degree angle from the vertical axis of the body (10). Thoracic kyphosis is measured on a lateral radiograph as the angle between the superior end plate of T2 and the inferior end plate of T12. Proximal thoracic kyphosis is measured from the superior end plate of T2 to the inferior end plate of T5. Middle and lower thoracic kyphosis is measured from the superior end plate of T5 to the inferior end plate of T12. The apex of normal thoracic kyphosis is the T6–T7 disc space (11, 12). The thoracolumbar junction should have no kyphosis or lordosis (11). Lumbar lordosis begins at L1–L2 and increases gradually until the L3–L4 disc space. There is a reciprocal relationship between the orientation of the sacrum, sacral slope, and the pelvic incidence and the characteristics of lumbar lordosis and location of the apex of lumbar lordosis (Fig. 19-1) (13–16). A sacral slope of <35 degrees and a low pelvic incidence are associated with a relatively flat, short lumbar lordosis. A sacral slope of more than 45 degrees and a high pelvic incidence are associated with a long, curved lumbar lordosis (14).

Initially, during fetal and intrauterine development, the entire spine is kyphotic. During the neonatal period, the thoracic, lumbar, and sacral portions of the spine remain in a kyphotic posture. Cervical lordosis begins to develop when a child starts holding his or her head up. When an upright posture is assumed, the primary and secondary curves begin to develop. The primary curves are thoracic and sacral kyphosis, and the secondary or compensatory curves in the sagittal plane are cervical and lumbar lordosis. These curves balance each other so that the head is centered over the pelvis (2, 17, 18).

The ranges of normal thoracic kyphosis and lumbar lordosis are dynamic, progressing gradually with growth (19). During the juvenile and adolescent growth periods, thoracic kyphosis and lumbar lordosis become more pronounced and take on a more adult appearance. Mac-Thiong et al. (13) showed that pelvic incidence and tilt increased with growth but sacral slope remained stable.

Differences also exist between male and female spines (6), and thoracic kyphosis and spine mobility are different in boys and girls: during the juvenile and adolescent periods (ages 8 to 16 years), girls have less thoracic kyphosis and thoracic spinal mobility than do boys of the same age (3, 12). Thoracic kyphosis also tends to progress with age: from 30 to 70 years of age, women have a progressive increase in kyphosis, from a mean of 25 degrees to a mean of 40 degrees (19). Men also show a definite progression with age, but at a lower rate.

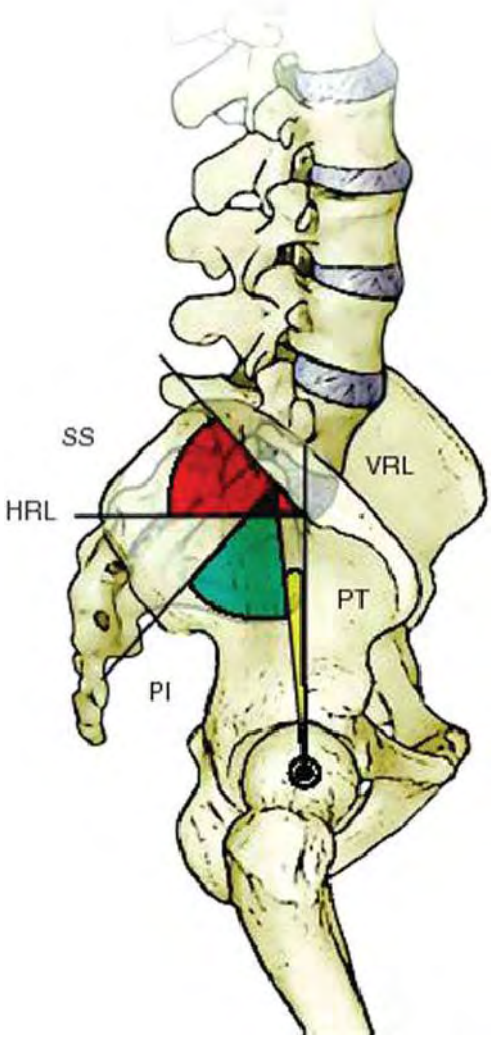


FIGURE 19-1. Radiographic measurements of pelvic incidence (*a*), sacral slope (*b*), and pelvic tilt (*c*). SS, sacral slope; HRL, horizontal reference line; PI, pelvic incidence; PT, pelvic tilt; VRL, vertical reference line. (From MF, Kuklo TR, Blanke KM, et al. *Radiographic measurement manual*: *Spinal Deformity Study Group* (*SDSG*). Memphis, TN: Medtronic Sofamor Danek, Fall 2004.)

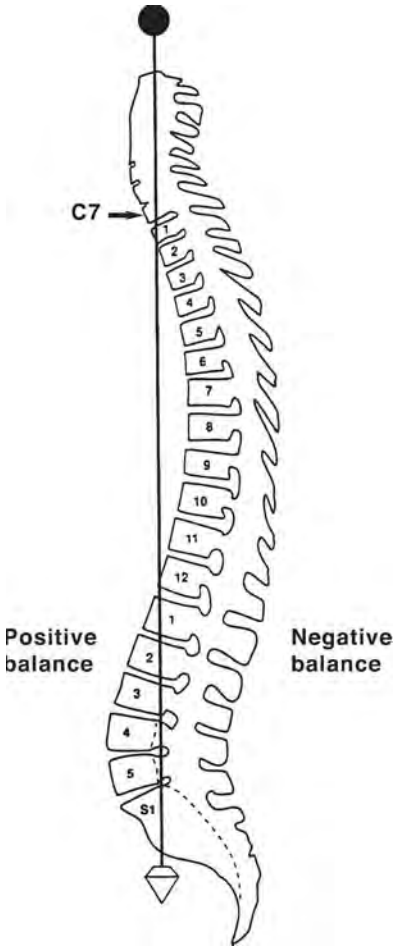


FIGURE 19-2. A plumb line is dropped from the middle of the C7 vertebral body to the posterosuperior corner of the S1 vertebral body. (From Bernhardt M. Normal spinal anatomy: normal sagittal plane alignment. In: Bridwell KH, DeWald RL, eds. *The textbook of spinal surgery*, 2nd ed. Philadelphia, PA: Lippincott-Raven, 1997:185.)

Normal sagittal balance is defined as a plumb line dropping from C7 and intersecting the posterosuperior corner of the S1 vertebral body (Fig. 19-2). Positive sagittal balance occurs when the plumb line falls in front of the sacrum, and negative sagittal balance occurs when the plumb line falls behind the sacrum (20).

Different forces are exerted on the spine, depending on the presence of kyphosis or lordosis. In the upright position, the spine is subjected to the forces of gravity, and several structures maintain its stability: the disc complex (nucleus pulposus and annulus), the ligaments (anterior longitudinal ligament, posterior longitudinal ligament, ligamentum flavum, apophyseal joint ligaments, and interspinous ligament), and the muscles (the long spinal muscles, the short intrinsic spinal muscles, and the abdominal muscles). Alteration in function resulting from paralysis, surgery, tumor, infection, or alteration in growth potentials can cause a progressive kyphotic deformity in a child (21). Both compressive and tensile forces are produced by the action of gravity on an upright spine (Fig. 19-3). In normal thoracic kyphosis, the compressive forces borne by the anterior elements are balanced by the tensile forces borne by the posterior elements. In a lordotic spine, the compressive forces are posterior and the tensile forces are anterior. These forces of compression and tension on the spinal physes can cause changes in normal growth, and a growth deformity can be added to a biomechanical deformity to cause a pathologic kyphosis (21, 22).

Voutsinas and MacEwen (23) suggested that relative differences in forces applied to the spine are reflected more accurately by the length and width of a kyphotic curve than by just the degree of the curve. For example, curves that are longer and wider (farther from the center of gravity) are more likely to cause deformity in an immature spine (Fig. 19-4). Winter

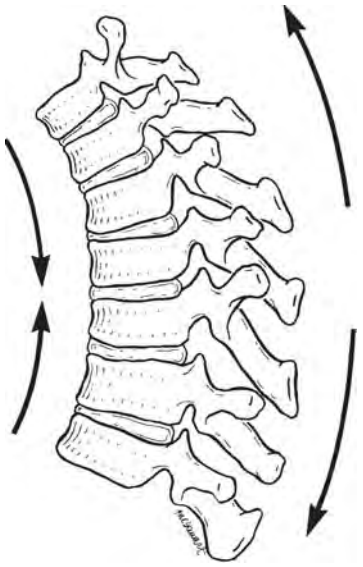


FIGURE 19-3. Forces that contribute to kyphotic deformity of the thoracic spine. The anterior vertebral bodies are in compression, and the posterior vertebral elements are in tension. (From White AA III, Panjabi MM. Practical biomechanics of scoliosis and kyphosis. In: White AA, Panjabi MM, eds. *Clinical biomechanics of the spine*. Philadelphia, PA: JB Lippincott, 1990:127.)

and Hall (24) classified disorders that result in kyphosis of the spine. Only the more common causes are presented in this chapter; the other causes are discussed elsewhere in this book (Table 19-1).

POSTURAL KYPHOSIS

Postural kyphosis is a flexible deformity of the spine and is common in juvenile and adolescent patients. Usually, the parents are more concerned about the postural roundback deformity than the adolescent is, and these parental concerns typically are what bring the patient to the physician's office. The physician's role in this situation is to rule out more serious causes of kyphosis. Postural kyphosis should be differentiated from pathologic types of kyphosis, such as Scheuermann disease, and from congenital kyphosis. When observed from the side, patients with postural roundback have a gentle rounding of the back while bending forward (Fig. 19-5). Patients with Scheuermann disease and congenital kyphosis have a sharp angular kyphosis or gibbus on forward bending when observed from the side. Radiographs usually are necessary to rule out pathologic types of kyphosis. Patients with postural kyphosis do not have radiographic vertebral-body changes, and the deformity is completely correctable by changes in position or posture. This deformity is common in patients who are taller than their peers and in young adolescent girls undergoing early breast development who tend to stoop because they are self-conscious about their bodies (25).

No active medical treatment is necessary. Bracing is not indicated. Exercises have been suggested and may help maintain better posture, but adherence to such a therapy program is difficult for juveniles and young adolescents. This problem is best treated by educating the patient and, more important, the parents and by observation (26).

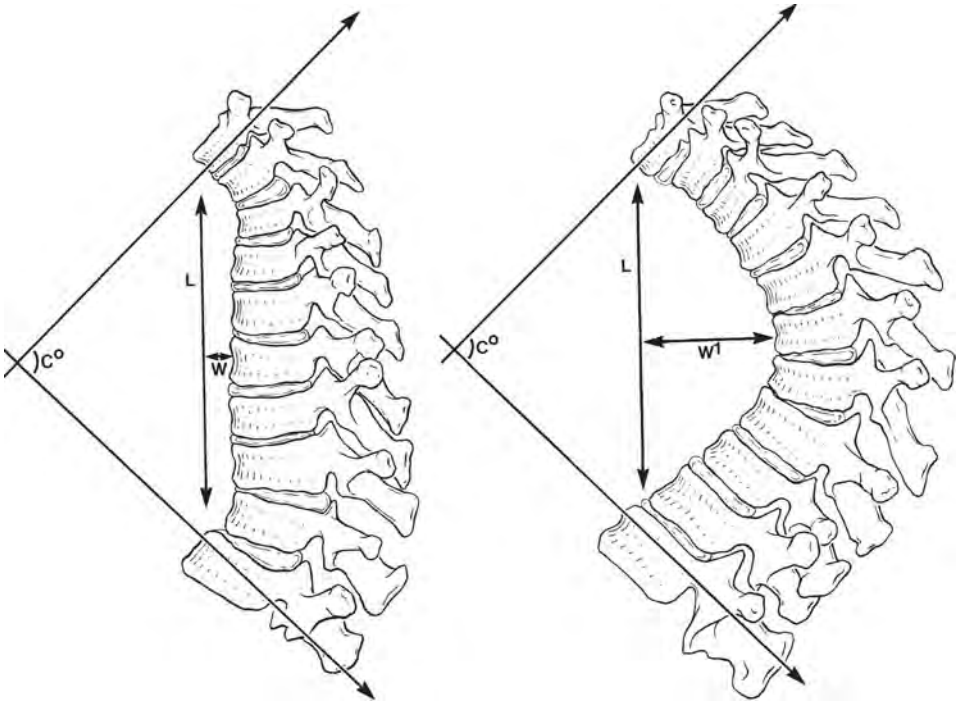


FIGURE 19-4. The two spinal curvatures represented by these drawings are different in magnitude; however, using cobb's method to measure the deformities, the degrees of curvature are identical. The differences in the curves are more accurately reflected when the length of the curves (*L*) and their respective widths (*W* and W^1) are taken into consideration. (From Voutsinas SA, MacEwen GD. Sagittal profiles of the spine. *Clin Orthop* 1986;210:235.)

TABLE 19-1 Disorders Affecting the Spine and Resulting in Kyphosis

I. Postural disorders
II. Scheuermann kyphosis
III. Congenital disorders
  a. Defect of formation
  b. Defect of segmentation
IV. Paralytic disorders
  a. Poliomyelitis
  b. Anterior horn cell disease
V. Myelomeningocele
VI. Posttraumatic
  a. Acute
  b. Chronic
  c. With/without cord damage
VII. Inflammatory
  a. Tuberculosis
  b. Other infection
VIII. Postsurgical
  a. Postlaminectomy
  b. Postbody (tumor) excision
IX. Inadequate fusion
  a. Too short
  b. Pseudoarthrosis
X. Postirradiation
  a. Neuroblastoma
  b. Wilms tumor
XI. Metabolic
  a. Osteoporosis
    1. Senile
    2. Juvenile
  b. Osteogenesis imperfecta
XII. Developmental
  a. Achondroplasia
  b. Mucopolysaccharidosis
  c. Other
XIII. Collagen disease (e.g., Marie-Strumpell)
XIV. Tumor
  a. Benign
  b. Malignant
XV. Neurofibromatosis

From Winter RB, Hall JE. Kyphosis in childhood and adolescence. *Spine* 1978;3:285.

TABLE 19-2 Winter's Classification of Congenital Deformity

Type	Description
I	Failure of formation of all or part of the vertebral body
II	Failure of segmentation of one or multiple vertebral levels
III	Mixed form, with elements of both failure of formation and failure of segmentation

CONGENITAL KYPHOSIS

Congenital kyphosis is an uncommon deformity, but, despite its rare occurrence, neurologic deficits resulting from this deformity are frequent.

Congenital kyphosis occurs because of abnormal development of the vertebrae, including a failure of developing segments of the spine to form or to separate properly (27). The spine may be either stable or unstable, or it may become unstable with growth (28). Spinal deformity in congenital kyphosis usually progresses with growth, and the amount of progression is directly proportional to the number of vertebrae involved, the type of involvement, and the amount of remaining normal growth in the affected vertebrae (28, 29).

Van Schrick in 1932 (30) and Lombard and LeGenissel in 1938 (31) initially described two basic types of congenital kyphosis: a failure of formation of part or all of the vertebral body and a failure of segmentation of part or all of the vertebral body. Winter et al. (27, 32) developed the most useful classification of congenital kyphosis, which divides the deformity into three types (Table 19-2). Type I is failure of formation of all or part of the vertebral body (Fig. 19-6A); type II is failure of segmentation of one or multiple vertebral levels (Fig. 19-6B); and type III is a mixed form, with elements of both failure of formation and failure of segmentation.

McMaster and Singh (33) further subdivided this classification into types of vertebral-body deformity. Defects of vertebral-body segmentation consist of a partial (anterior unsegmented bar) or a complete (block vertebrae) failure of segmentation. Defects of vertebral-body formation are divided into four types: (a) posterolateral quadrant vertebrae, (b) butterfly

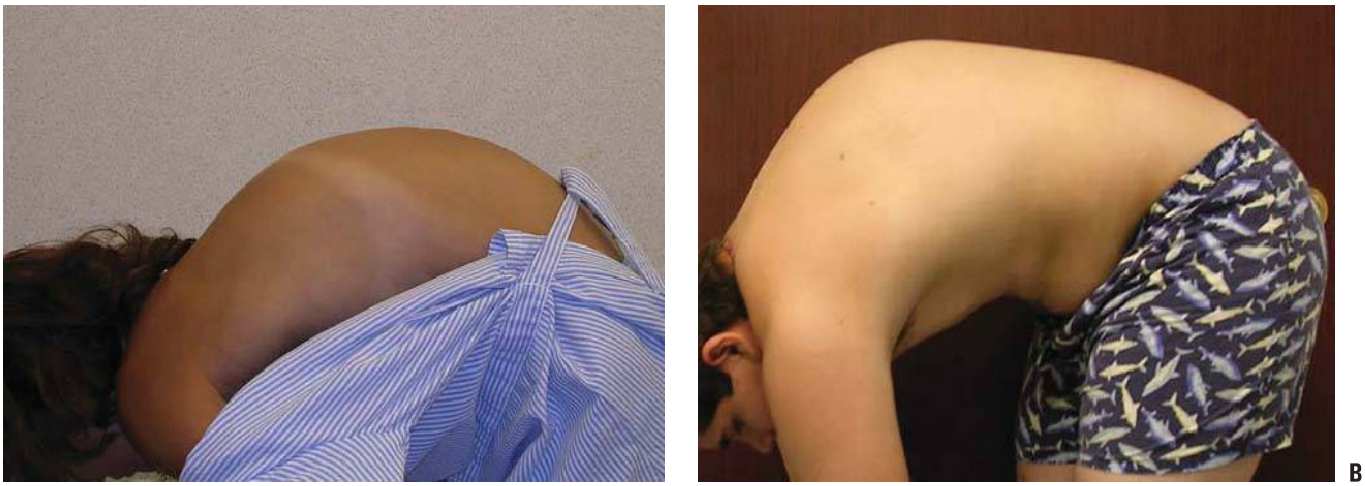


FIGURE 19-5. **A**: Lateral view of normal spinal contour on forward bending. **B**: Lateral view of a patient with Scheuermann disease on forward bending. Note the break in the normal contour and sharp angular nature of the spine.

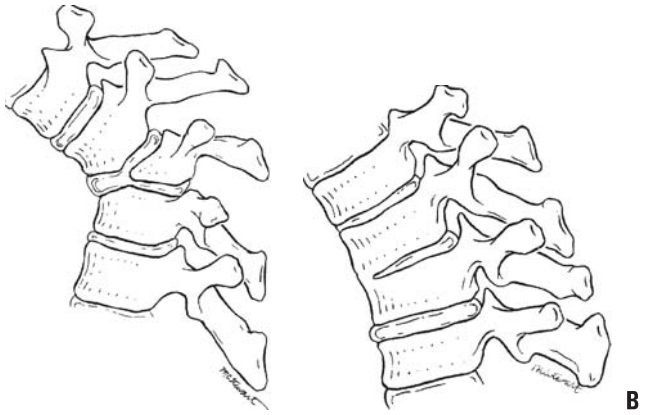


FIGURE 19-6. **A**: Congenital kyphosis caused by failure of formation of the vertebral body (type I). **B**: Congenital kyphosis caused by failure of segmentation (type II). (Courtesy of Robert Winter, MD, Minneapolis.)

vertebrae, (c) posterior hemivertebrae, and (d) wedged vertebrae (Fig. 19-7). Dubousset (34) and Zeller et al. (35) added a rotary dislocation of the spine, and Shapiro and Herring (36) further divided type III displacement into types A (sagittal plane only) and B (rotary, transverse, and sagittal planes). Any classification can be subdivided further into deformities with or without neurologic compromise; this is useful for making treatment decisions because each type of congenital kyphosis has a distinct natural history and risk of progression.

Most of the vertebral malformations that cause spinal deformity occur between the 19th and the 30th days of fetal development (28, 32, 37). The somatic mesoderm, which is devoted to the formation of the vertebral column and the rib cage, undergoes segmentation into 38 to 44 pairs of discrete, bilateral somites. The formation of a vertebra depends on contributions of cells from two separate and successive pairs of sclerotomes. This condensation of the paired sclerotomes occurs at approximately 5 weeks of gestation. If one side of the pair of sclerotomes fails to develop, a hemivertebra is formed, resulting in congenital scoliosis (38, 39).

Tsou (40) concluded that congenital kyphosis and congenital scoliosis occur during different periods of spinal development. He divided the development of the spine into an embryonic period (the first 56 days) and a fetal period (from day 57 to birth). During the embryonic period, failure of segmentation and aplasia of part of the vertebrae, resulting in hemivertebra formation, cause scoliosis, while congenital kyphosis occurs in the fetal period, during the cartilaginous phase of development (40). Failure of formation occurs in this phase when the cartilaginous centrum of the vertebral body forms a functionally inadequate growth cartilage.

Failure of formation varies from complete aplasia (which involves the pars and the facet joints and makes the spine unstable) to involvement of only the anterior one-third to one-half of the vertebral body. This abnormal development is

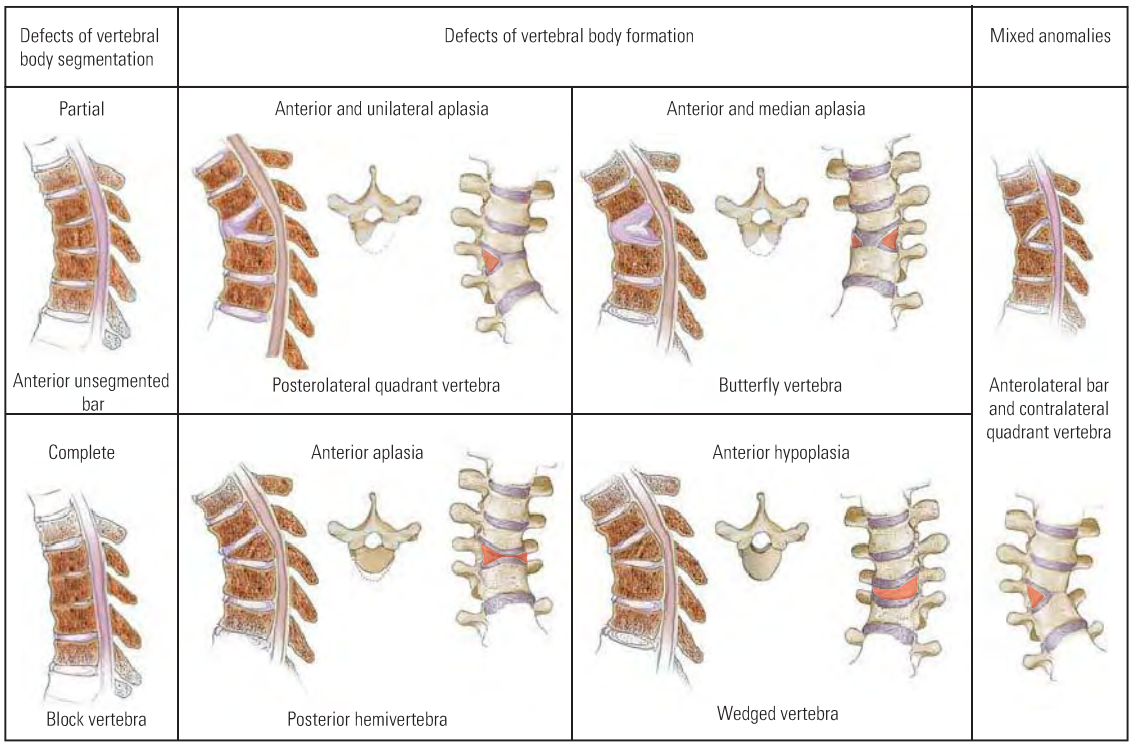


FIGURE 19-7. Drawings showing the different types of vertebral anomalies that produce congenital kyphosis or kyphoscoliosis. (From McMaster MJ, Singh H. Natural history of congenital kyphosis and kyphoscoliosis. *J Bone Joint Surg* 1999;81A:1367–1383.)

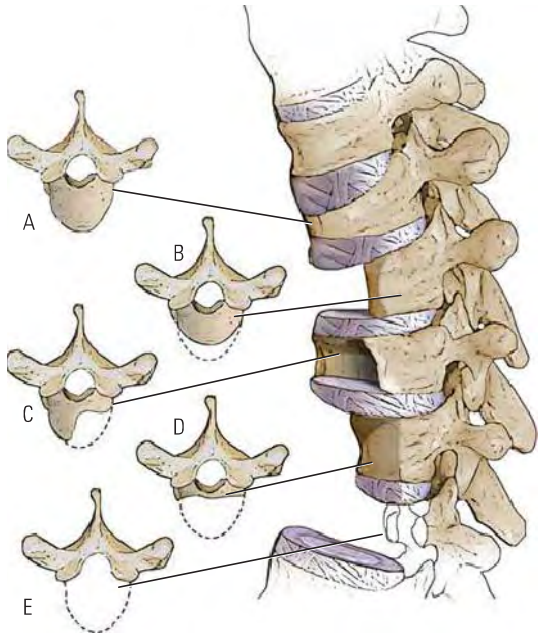


FIGURE 19-8. The five most common patterns of congenital vertebral hypoplasia and aplasia are illustrated in lateral and transverse views. Types *B* and *E* tend to produce pure congenital kyphosis. (From Tsou PM. Embryology of congenital kyphosis. *Clin Orthop* 1977;128:18.)

thought to be the result of inadequate vascularization of the vertebral body during the fetal period, leading to hypoplasia or aplasia of the anterior vertebral body. If one side of the vertebra is involved more than the other side, scoliosis also may occur (Fig. 19-8). Unlike hemivertebral anomalies that occur in the embryonic period because of maldevelopment of corresponding pairs of somites causing congenital scoliosis, posterior arch anomalies usually are absent in pure congenital kyphosis.

Failure of segmentation has been described as an osseous metaplasia of the annulus fibrosus (40, 41) that acts as a tether against normal growth and causing spinal deformity. The height of the vertebral bodies is relatively normal, but the depth of the ossification of the annulus fibrosus varies. Ossification may be delayed, with a period of normal growth followed by spontaneous ossification. Kyphosis caused by a "segmentation defect" is believed to represent a developmental defect of the perivertebral structures (the annulus fibrosis, the ring apophysis, and the anterior longitudinal ligament) rather than a true intervertebral bar (42).

The natural history of congenital kyphosis is well known and based on the type of kyphosis: failure of formation (type I), failure of segmentation (type II), or mixed anomalies (type III). Congenital kyphosis tends to be progressive, with the greatest rate of progression occurring during the time of most rapid growth of the spine (birth to 3 years of age) and during the adolescent growth spurt. Winter et al. (32) found that failure of formation (type I deformity) produces a much more severe kyphosis, with a rate of progression that averages 7 degrees per year, whereas type II deformities progress an average of 5 degrees per year. McMaster and Singh found the most rapid progression in type III kyphosis, followed by type I, because of involvement of posterolateral quadrant vertebrae. In their study, a type III kyphosis progressed at a rate of 5 degrees per year before 10 years of age and 8 degrees per year thereafter until the end of growth. Type I (failure of formation) kyphosis progressed 2.5 degrees per year before 10 years of age and 5 degrees per year thereafter (33). Type I and III deformities are associated with a much higher incidence of neurologic involvement and paraplegia than are type II deformities. Neurologic problems occur more frequently in patients with type I and III deformities because they tend to have an acute angular kyphosis over a short segment, which places the spinal cord at higher risk for compression at the level of acute angulation. Type II deformities (failure of segmentation) rarely result in neurologic problems because involvement of several segments produces a more gradual kyphosis, and vertebral-body height usually is maintained with little or no vertebral-body wedging. The most frequent location of congenital kyphosis is T10–L1 (32).

Patients with congenital kyphosis may have other anomalies. Intraspinal abnormalities have been reported to occur in 5% to 37% of patients with congenital kyphosis and congenital scoliosis (43–46). A study by Bradford et al. (47) indicated that this incidence may be even greater. They found that six of eight patients with congenital kyphosis had spinal cord abnormalities visible on magnetic resonance imaging (MRI). Although the proposed time of development of the deformity may be different from that of congenital scoliosis, other nonskeletal anomalies such as cardiac, pulmonary, renal, and auditory disorders or Klippel-Feil syndrome (48, 49) can be associated with congenital kyphosis. McMaster et al. (50) found an adverse effect on lung development and function caused by an increasing constriction of the rib cage and impairment of diaphragmatic movement. The more cranial the level of the congenital kyphosis, especially above T10, the more significant the effect on respiratory impairment.

Patient Presentation. The diagnosis of a congenital spine problem usually is made by a pediatrician before the patient is seen by an orthopaedist. The deformity may be detected before birth on prenatal ultrasonography (51) or noted as a clinical deformity in a newborn. If the deformity is mild, congenital kyphosis can be overlooked until a rapid growth spurt makes the condition more obvious. Some mild deformities are found by chance on radiographs that are obtained for other reasons. Clinical deformities seen in a newborn tend to have a worse prognosis than those discovered as incidental findings on plain radiographs. Physical examination usually reveals a kyphotic deformity at the thoracolumbar junction or in the lower thoracic spine. An attempt should be made to determine the rigidity of the deformity

by flexion and extension of the spine. A detailed neurologic examination should be done, looking for any subtle signs of neurologic compromise. Associated musculoskeletal and nonmusculoskeletal anomalies should be sought on physical examination.

High-quality, detailed anteroposterior and lateral radiographs provide most information in the evaluation of congenital kyphosis (Fig. 19-9). Failure of segmentation and the true extent of failure of formation may be difficult to detect on early films because of incomplete ossification. Flexion and extension lateral radiographs are helpful in determining the rigidity of the kyphosis and possible instability of the spine. Computerized tomography (CT) with three-dimensional reconstructions can identify the amount of vertebral-body involvement and can determine whether more kyphosis or scoliosis might be expected (Fig. 19-10). CT scans can identify only the nature of the bony deformity and the size of the cartilage anlage. They do not show the amount of growth potential in the cartilage anlage, and therefore only an estimate of possible progression can be made. MRI should be obtained in most cases because of the significant incidence of intraspinal abnormalities. In addition, the location of the spinal cord and any areas of spinal cord compression caused by the kyphosis can be seen on MRI. The cartilage anlage will be well defined by MRI in patients with failure of formation (Fig. 19-11); however, as with CT scans and plain radiographs, MRI cannot reveal how much growth potential is present in the cartilage anlage and can only help estimate the probability of a progressive deformity.

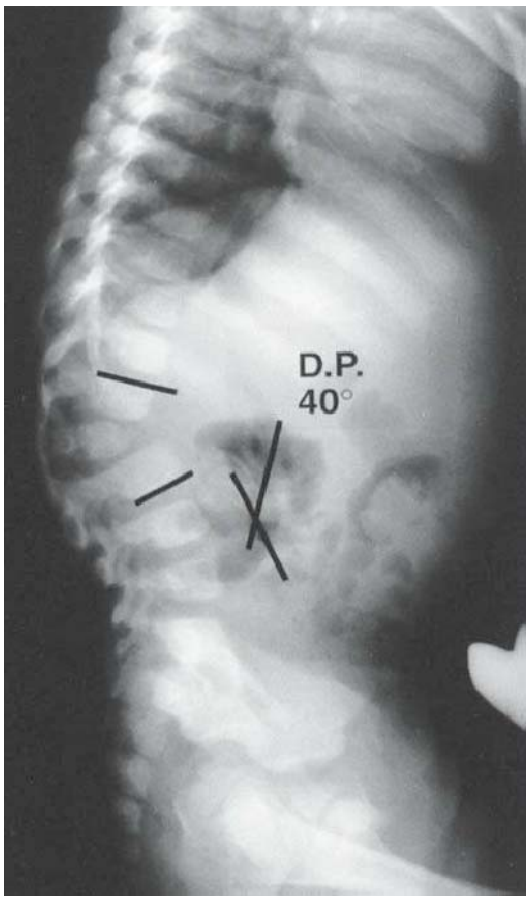


FIGURE 19-9. A 2-year-old child with type I congenital kyphosis measuring 40 degrees. Radiograph demonstrates failure of formation of the anterior portion of the first lumbar vertebra.

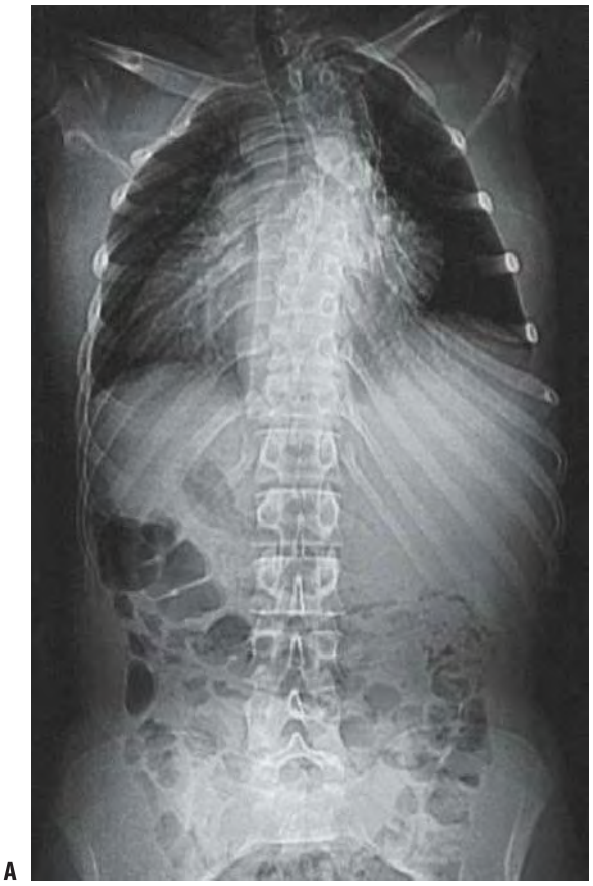
Congenital kyphosis, as well as associated renal problems, can be seen on routine prenatal ultrasonography as early as 19 weeks of gestation (51). Myelograms have been used for documenting spinal cord compression but have been mostly replaced by MRI. If myelography is used, images should be taken with the patient prone and supine. Myelograms obtained in only the prone position may miss information about spinal cord compression because of pooling of dye around the apex of the deformity. Myelography can be used in conjunction with CT scanning to add to the diagnostic information obtained.

Treatment. Because the natural history of this condition usually is one of continued progression with an increased risk of neurologic compromise, surgery usually is the preferred method of treatment (27). If the deformity is mild or if the diagnosis is uncertain, close observation may be a treatment option. However, observation of a congenital kyphotic deformity must be used with caution, and the physician must not be lulled into a false sense of security if the deformity progresses only 3 to 5 degrees over a 6-month period. If the deformity is observed over 2 to 3 years, it will have progressed 18 to 30 degrees and cannot thereafter be easily corrected. Bracing has no role in the treatment of congenital kyphosis, unless compensatory curves are being treated above or below the congenital kyphosis (27, 48, 52). Bracing a rigid structural deformity, such as congenital kyphosis, neither corrects the deformity nor stops the progression of kyphosis. To document that there has been a significant change in kyphosis, radiographs should be taken by a standardized method, and the same end vertebral bodies should be measured. This will ensure that any change that has occurred since the previous radiograph is accurately measured.

Surgery is the recommended treatment for congenital kyphosis. The type of surgery depends on the type and size of the deformity, the age of the patient, and the presence of neurologic deficits. Procedures can include posterior fusion, anterior fusion, both anterior and posterior fusions, and anterior osteotomy with posterior fusion. Fusion can be done with or without instrumentation.

Treatment of Type I Deformities. The treatment of type I deformities depends on the stage of the disease: early with mild deformity, late with moderate or severe deformity, and late with severe deformity and spinal cord compression.

Early Treatment of Mild Deformities. For type I deformities, the best treatment is early posterior fusion. If the deformity is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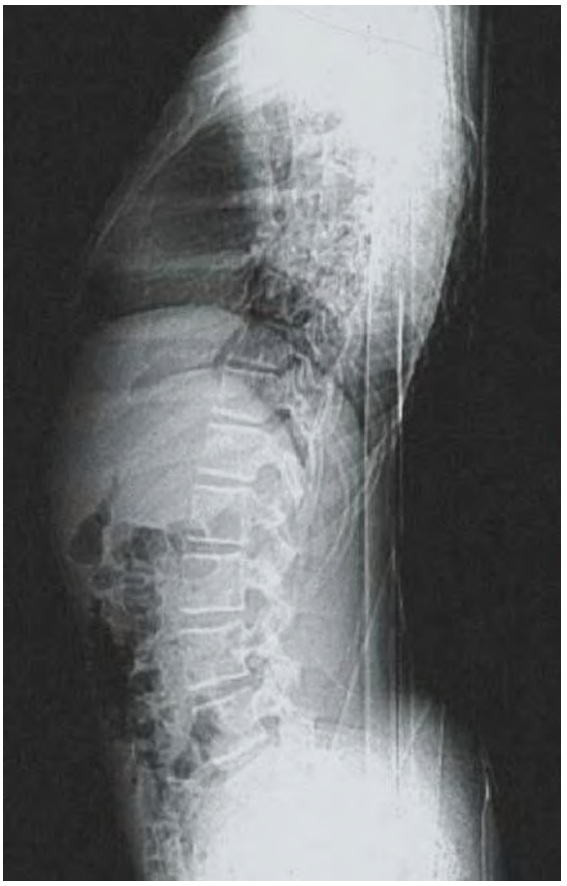
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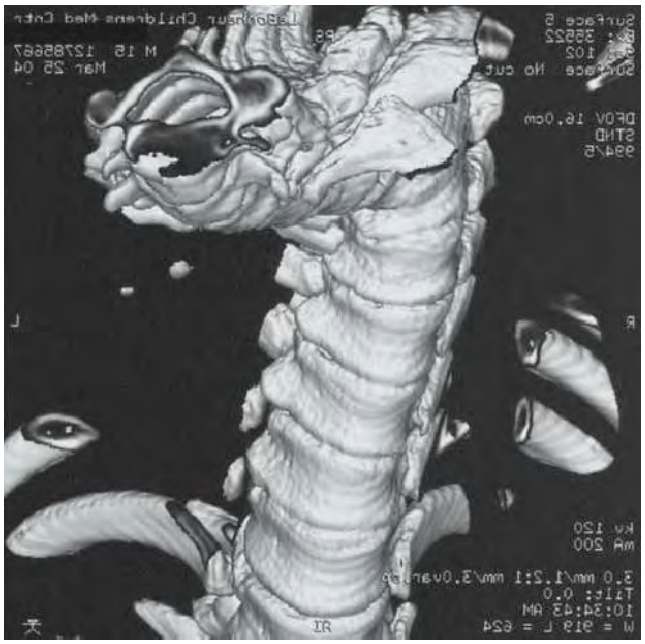
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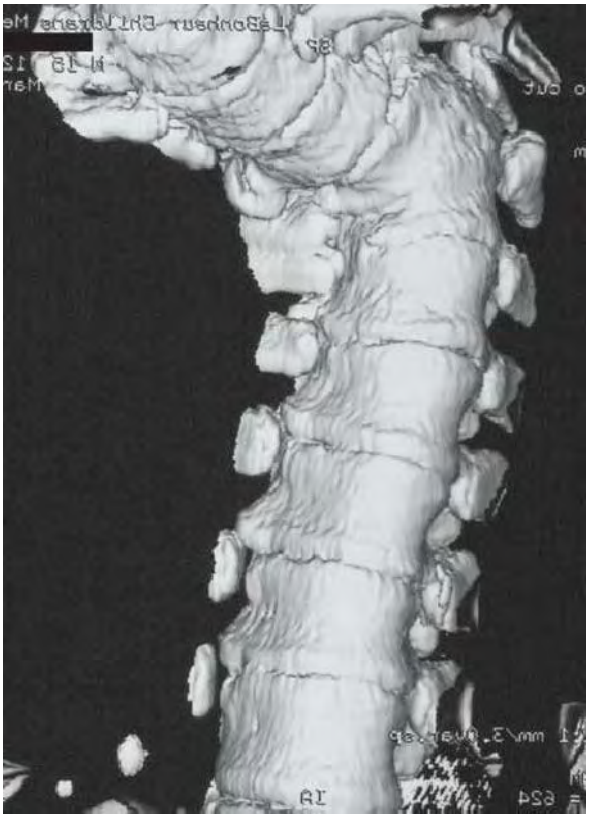
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FIGURE 19-10. Congenital kyphosis. **A**,**B**: Anteroposterior and lateral radiographs. Note inadequate detail of kyphosis on lateral radiograph of spine. **C–E**: CT three-dimensional reconstruction views that clearly demonstrate the bony anatomy of congenital kyphosis.

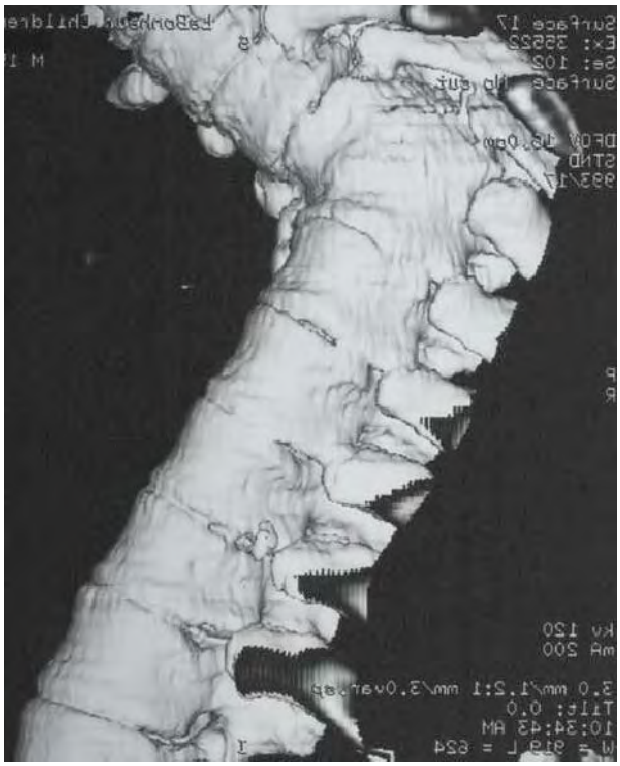

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FIGURE 19-10. *(Continued)*

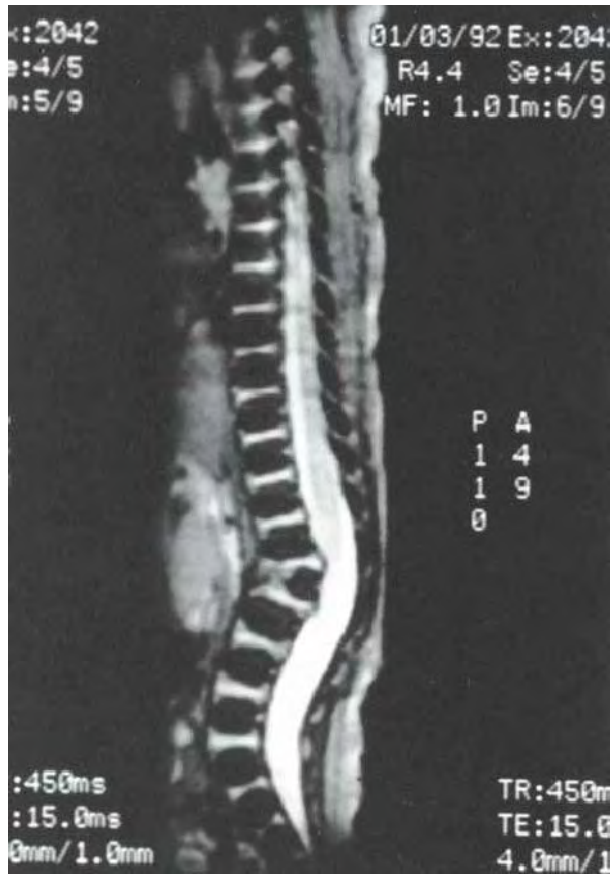

FIGURE 19-11. MRI of type I congenital kyphosis. Failure of formation of the anterior vertebral body is demonstrated, but the growth potential of the involved vertebra cannot be determined. Note the pressure on the dural sac.

<50 or 55 degrees and the patient is younger than 5 years of age, posterior fusion alone, extending from one level above the kyphotic deformity to one level below, is recommended (27, 32, 48, 53). This may allow for some improvement in the kyphotic deformity because of continued growth anteriorly from the anterior end plates of the vertebrae one level above and one level below the congenital kyphotic vertebrae that are included in the posterior fusion. McMaster and Singh (54) reported 15 degrees of correction in nine of 11 patients treated with this technique. Kim et al. (55) reported that after posterior fusion alone correction of kyphosis occurred with growth only in patients younger than 3 years of age and with type II and III deformities. Anterior and posterior spinal fusions at least one level above and one level below the congenital kyphosis are indicated in curves of more than 60 degrees (55). Anterior and posterior fusions predictably halt the progression of the kyphotic deformity but, because of ablation of the anterior physes (35, 53, 55), do not allow for the possibility of some correction of the deformity with growth.

Late Treatment of Moderate to Severe Deformities. In older patients with type I kyphotic deformities, posterior arthrodesis alone may be successful if the kyphosis is <50 to 55 degrees (32, 56). If the deformity is more than 55 degrees (which usually is the case in deformities detected late), anterior and posterior fusion produces more reliable results (32, 56). Anterior arthrodesis alone will not correct the deformity. Any correction of the deformity requires anterior strut grafting with temporary distraction and posterior fusion, with or without posterior compression instrumentation. The posterior instrumentation may allow for some correction of the kyphosis but should be regarded more as an internal stabilizer than as a correction device (27). Instrumentation has been reported to decrease the pseudoarthrosis rate (55). Correction by instrumentation should be used with caution in rigid, angular curves because of the high incidence of neurologic complications. If anterior strut grafting is done, the strut graft should be placed anteriorly under compression. If no correction is attempted and the goal of surgery is just to stop progression of the kyphosis, a simple anterior interbody fusion combined with a posterior fusion can be used. Smith et al. (57) described simultaneous anterior and posterior approaches through a costotransversectomy that allowed resection of the posterior hemivertebra and correction of the kyphosis with posterior compression instrumentation. Correction of the kyphosis could be obtained safely once the posterior hemivertebra was removed and the thecal sac could be observed during corrections. The use of skeletal traction (halo-pelvic, halo-femoral, or halo-gravity) to correct the deformity is tempting, but is not recommended because of the risk of paraplegia (58). In a patient with a rigid gibbus deformity, traction pulls the spinal cord against the apex of the rigid kyphosis and can lead to neurologic compromise (Fig. 19-12).

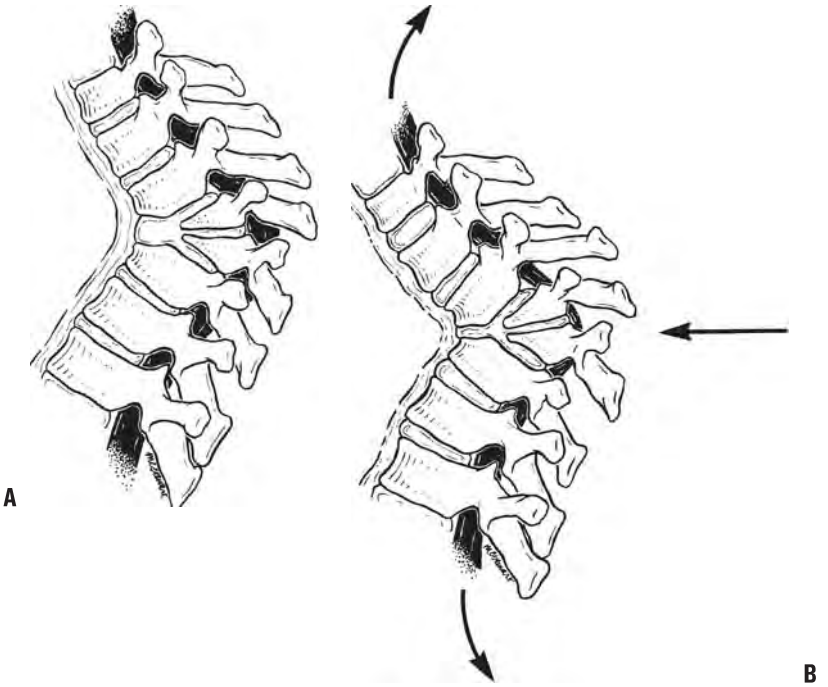

FIGURE 19-12. The effect of traction on a rigid congenital kyphosis. **A**: The apical area does not change with traction, but the adjacent spine is lengthened. **B**: As the spine lengthens, so does the spinal cord, producing increased tension in the cord and aggravating existing neurologic deficits. (From Lonstein JE, Winter RB, Moe JH, et al. Neurologic deficit secondary to spinal deformity. *Spine* 1980;5:331.)

ANTERIOR STRUT GRAFT FOR KYPHOSIS. Strut grafts are most often used for patients with severe kyphotic deformities with or without associated scoliosis. Strut grafts can also be used in cases of tumor resections and/or vertebral-body collapse secondary to infections and/or trauma. Anterior strut grafting with a free graft is generally used for a kyphosis without neurologic deficits in order to prevent further progression of the deformity (Figs. 19-13 to 19-17). It can be used either by an *in situ* fusion technique or fusion with minimal intraoperative correction. If intraoperative correction is anticipated, the kyphosis must be mobile at its apex. There must be no fusions posteriorly or, if there are, these must be released prior to any attempt at correction. Distraction of a rigid kyphosis carries a high risk of neurologic injury for the patient and therefore should never be attempted. The struts can be from the fibula or rib, or both. Vascularized or free vascularized grafts may also be used for these procedures. Nonvascularized fibular grafts are the most commonly used structural grafts for kyphotic deformities, as they provide the greatest structural support. The fibula is ideal because one can obtain a large length (up to 26 cm) in mature patients. The disadvantage, however, of using free fibula is that the graft may take a long time to incorporate (up to 2 years). These free fibular grafts are at their weakest point 6 months postoperatively (creeping substitution) and hence subject to fracture, particularly when the graft is placed more than 4 cm anterior to the apex of the kyphosis.

Although rib can be used, it is structurally a very weak graft. Rib is most often used to supplement a fibular autograft (58).

The strut graft generally extends from one end vertebra of the kyphosis to the other. It should lie as far anteriorly and as close to the midline as possible to allow for maximum structural support. Strut grafting has the advantage of being able to stabilize the kyphotic deformity by placing a compression arthrodesis in the line of axial stress of the spine. A vascularized rib graft is a useful alternative when rapid incorporation (6 to 8 weeks) is desired and ligation of segmental vessels is contraindicated. This may be used in kyphosis with preexisting neurologic lesions and especially in cases where fusion *in situ* is indicated to prevent further curve progression. Vascularized grafts are not subjected to creeping substitution. When a vascularized rib graft is planned, it is obtained by swinging a portion of the rib down with a vascular pedicle to provide an anterior strut for a kyphotic deformity. The rib selected is chosen to best fit with the level to be bridged. For kyphosis with an apex between T2 and T5, a rib two or three segments below the apex of the kyphosis is the ideal rib to use. If the apex of the kyphosis is at T6 or below, a rib two or three segments proximal to the kyphosis apex is the one to be selected. The rib is rotated on a muscle pedicle flap. This technique requires meticulous dissection with preservation of the vasculature. Firm incorporation is achieved within 6 to 8 weeks; hence, this technique is ideally suited for fusion *in situ* as the grafts have a low mechanical load capacity.

Strut grafting is contraindicated when a second procedure is planned that may produce enough correction to dislodge the anterior strut graft. If this is anticipated, the strut grafting should be done after the corrective posterior procedure is completed. Other contraindications include conditions where the bone is so soft that the strut graft may penetrate the posterior cortex and cause spinal cord injury.

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Anterior Strut Graft for Kyphosis (Figs. 19-13 to 19-17)

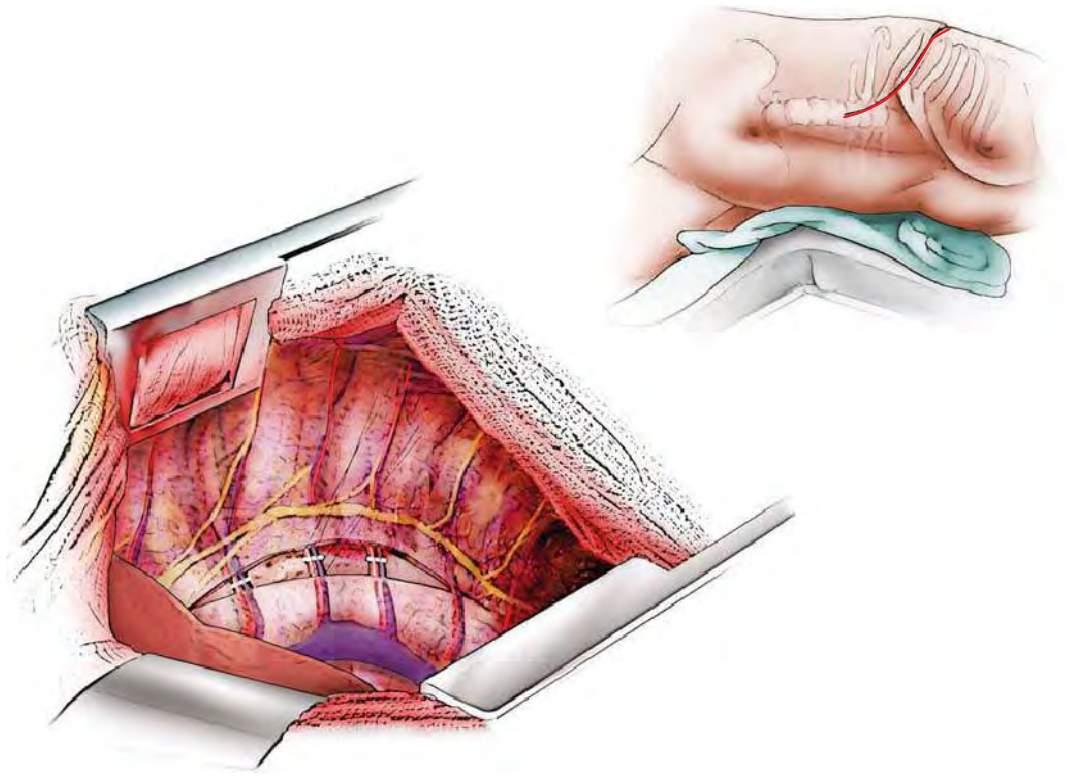
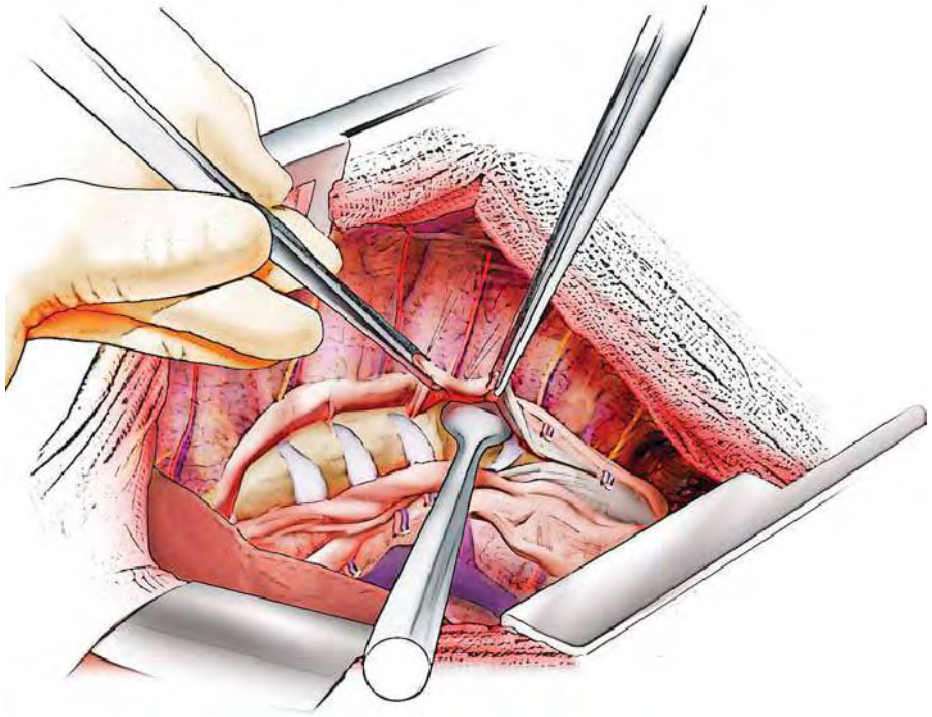


FIGURE 19-13. Anterior Strut Graft for Kyphosis. A standard thoracotomy or thoracoabdominal approach is used over the rib to be excised. The rib to be excised is at the upper end vertebra of the kyphotic deformity. Another way to select the appropriate rib is to identify the apex of the kyphosis and draw a line to the midaxillary line. This line will intersect the appropriate rib to be removed. The resected rib is excised subperiosteally and cut off at the costal transverse joint. Appropriate retractors are placed. The parietal pleura is incised and opened from the most proximal disc space to be exposed and then opened distally to the distal end vertebra. The segmental vessels are isolated in the midline and tied with interrupted 2-0 silk sutures. The vessels and the parietal pleura are bluntly dissected off the entire spine, exposing the entire spine throughout the length of the intended fusion. As the next step, we prefer to expose the spine subperiosteally by the development of a periosteal flap beginning at the rib head of the most proximal vertebra to be incorporated in the fusion and ending at the rib head (or transverse process in the lumbar spine) of the most distal included vertebra. This subperiosteal flap is developed as far to the opposite side of the spine as possible; this is especially important if a scoliotic deformity is present. In scoliosis, deformity exposure must include the concavity of the curve. This periosteal flap is protective of the soft tissues on the opposite side; it also provides an excellent bed for bony fusion. Some surgeons may prefer extraperiosteal dissection.

FIGURE 19-14. All exposed intervertebral discs are then removed back to the posterior longitudinal ligament with rongeurs and curettes of varying sizes and shapes. In congenital deformities, large amounts of cartilaginous material may be present in the apex of the kyphosis or the apex of the kyphoscoliosis. All this cartilaginous material and the vertebral end plates should be removed completely, as far back as the posterior longitudinal ligament, leaving this ligament intact.



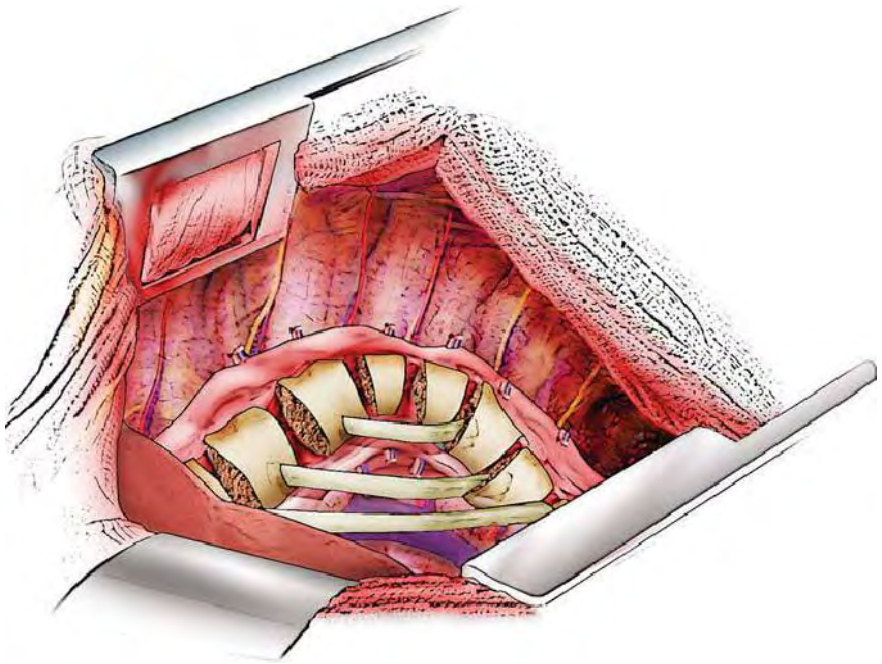


FIGURE 19-15. The fibular graft is harvested in the standard fashion. The fibular strut should lie as far anteriorly and as close to the midline as possible. A suture or a ruler can be used to measure the length of the most anterior strut. It is important not to overly shorten this strut prior to actual insertion. The anchoring holes for the fibular strut may be made in one of several ways. A burr can be used to develop anchoring holes in the anterior cortices of the vertebral body, or a trough can be prepared with curettes and gouges in the inferior aspect of the end vertebral body above and the superior aspect of the end vertebral body below. In this technique, a small notch is made in the cortex with a rongeur to allow the graft to be keyed into place during manual curve correction. For *in situ* fusion, mild correction will be obtained by use of manual compression over the apex of the kyphotic deformity. Manual pressure is applied over the apex of the kyphosis and the appropriate length graft keyed into position in one end vertebra and then the other. The end of the fibula may require some tapering, which is best done with a burr prior to insertion into the vertebral body. When the external pressure is relieved, the graft should be secure. If more than one structural graft is to be used, those closest to the apex must be inserted prior to insertion of the primary (most anterior) graft. Rib may be used for these secondary grafts and then morselized rib or iliac crest, or both, are packed into the intervening spaces.

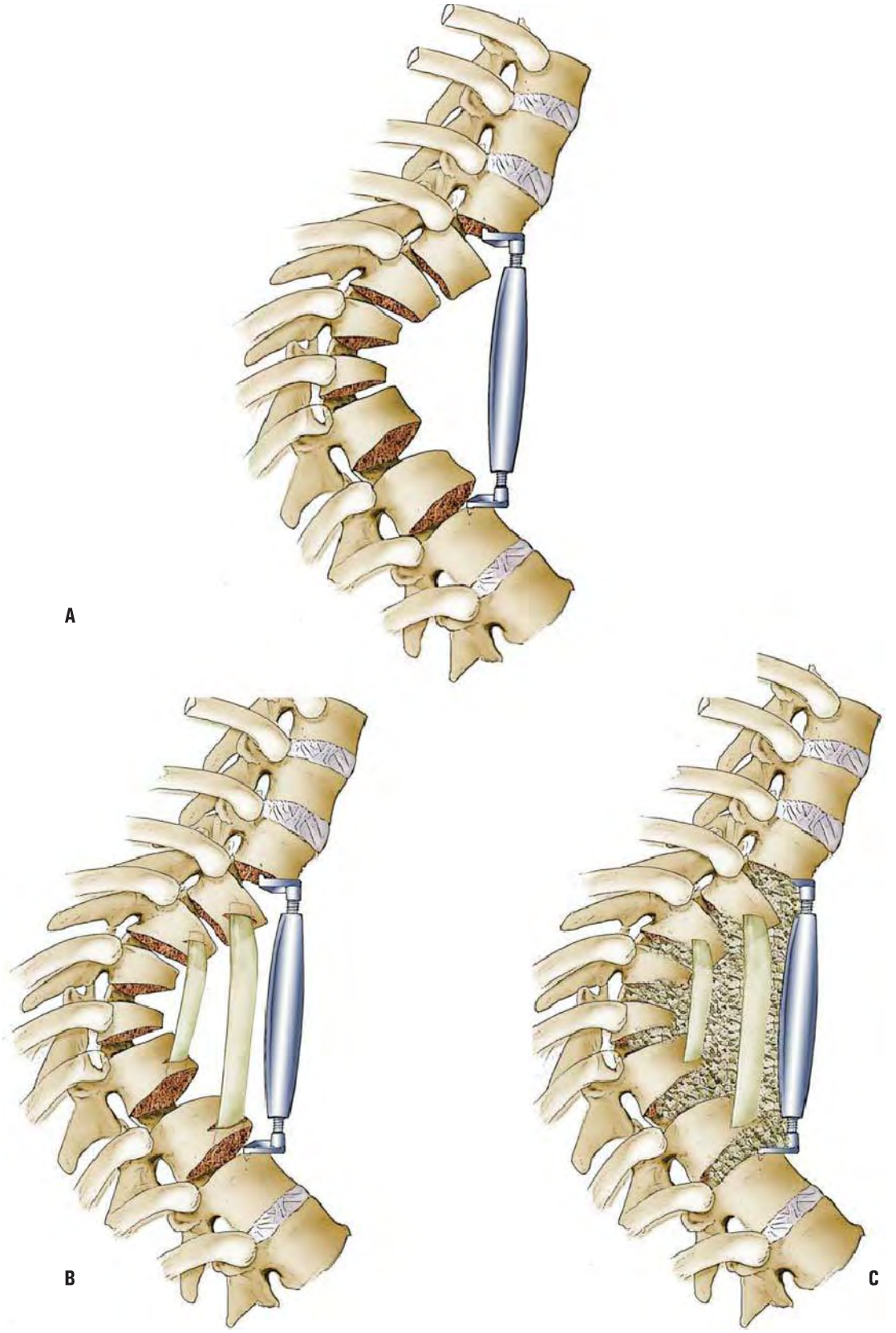


FIGURE 19-16. **A–C**: Although the procedure is not often done today, if correction is to be attempted anteriorly, one of several types of distractors must be placed in the furthest anterior position and gradually elongated. The distractor should be gradually spread over time with careful monitoring of spinal cord function by electrophysiologic (motor and sensory) monitoring. Once maximal distraction is obtained, grafts are inserted in a manner similar to the aforementioned technique.

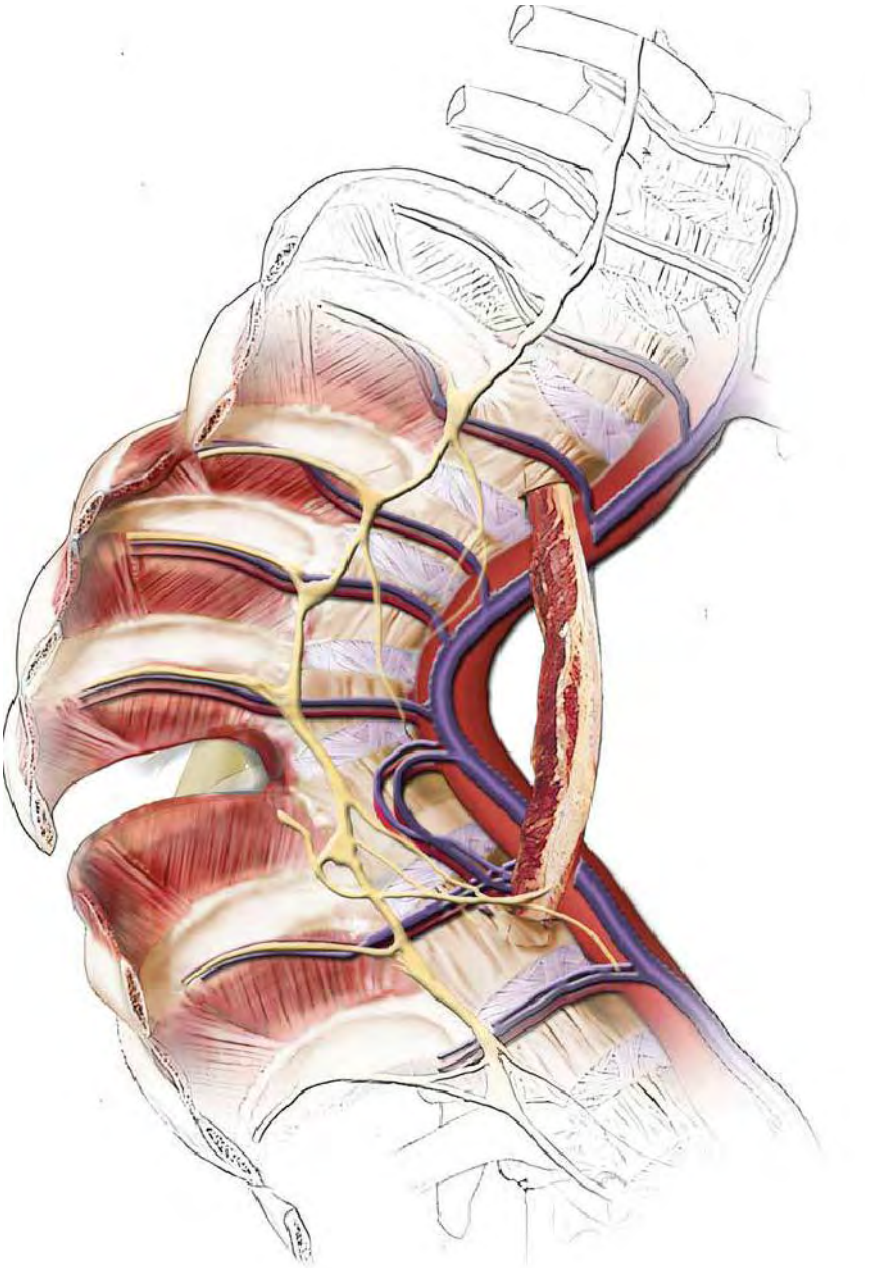


FIGURE 19-17. With the vascularized rib technique, the skin incision is made approximately at the level of the rib selected for the fusion. The intercostal musculature cranial to the rib is divided with the rib exposed subperiosteally at the costochondral margin and resected. The intercostal musculature is cut about 0.5 cm caudal to the selected rib leaving the neurovascular bundle intact. The appropriate length of rib is determined. The rib is subperiosteally exposed at the level of sectioning and the neurovascular bundle carefully ligated and sectioned. The intercostal musculature is then divided and the interspace opened further to allow exposure of the surgical area. The neurovascular bundle is then followed from the point of rib resection to the intervertebral foramen. The periosteum is freed on both ends of the ribs by approximately 1 cm to allow for insertion into the ends of the kyphotic segment. The fusion area can either be exposed as discussed above through subperiosteal dissection, or the graft can be keyed into position at the end vertebra as illustrated, without extensive subperiosteal dissection. I prefer to expose the bed for vascularized rib in the same fashion as for a free fibular graft.

Technique. The patient is placed in a standard lateral decubitus position. If the kyphotic deformity is associated with scoliosis, the kyphosis is approached from the convex side of the curvature. If, however, it is a pure kyphotic deformity, then in the thoracic region, a right-sided thoracotomy is preferred, whereas if the kyphosis apex is at the thoracolumbar junction, then a left-sided thoracotomy is preferred.

Late Treatment of Severe Deformities with Cord Compression. It is difficult to attempt late treatment of a severe congenital kyphotic deformity that is accompanied by spinal cord compression. If congenital kyphosis causes spinal cord compression, anterior decompression is indicated. The compression is created by bone or disc material pressing into the front of the spinal cord, and this can be decompressed only by an anterior procedure; laminectomy has no role in the treatment of this condition (24). If associated with scoliosis, the anterior approach for decompression may be on the concavity of the scoliosis to allow the spinal cord to move both forward and into the midline after decompression. After adequate decompression has been achieved, the vertebrae involved are fused with an anterior strut graft. This is followed by a posterior fusion, with or without posterior stabilizing instrumentation. Postoperative support with a cast, brace, or halo cast may be required.

Treatment of Type II Deformities.

Treatment of type II deformities can be divided into early treatment of mild deformities and late treatment of severe deformities as outlined by

Mayfield et al. (59). If a type II kyphosis is mild and detected early, posterior fusion with compression instrumentation can be done. The kyphosis should be <50 degrees for a posterior fusion alone to have a good chance of success. The posterior fusion should include all the involved vertebrae, plus one vertebra above and one vertebra below the congenital kyphosis.

Compression instrumentation can be used more safely in type II deformities, because the kyphosis is more rounded and affects several segments, instead of being sharply angular as in type I deformities. If the deformity is severe and detected late, correction can be obtained only with anterior osteotomies and fusion, followed by posterior fusion and compression instrumentation (59).

Complications of Treatment. Some of the more frequent complications of treatment of congenital kyphosis are pseudarthrosis, progression of kyphosis, and paralysis. Pseudarthrosis and progression of the kyphotic deformity can be minimized by using anterior and posterior fusions for deformities of more than 50 degrees. The posterior fusion should extend from one level above to one level below the involved vertebrae. This may allow for some correction with growth.

Paralysis is perhaps the most feared complication of spinal surgery. The risk of this complication can be lessened by not attempting to maximally correct the deformity with instrumentation. Instrumentation should be used only for stabilization of rigid deformities unless simultaneous anterior vertebral-body resection and posterior fusion and instrumentation are done through a costotransversectomy approach as described by Smith et al. (57). The use of halo traction in rigid congenital kyphotic deformities has been associated with an increased risk of neurologic compromise (58). Another long-term problem, occurring in approximately 38% of patients with kyphosis, is low back pain caused by increased lumbar lordosis, which is needed to compensate for the kyphotic deformity (60).

PROGRESSIVE ANTERIOR VERTEBRAL FUSION

Progressive anterior vertebral fusion (PAVF) is rare and is an uncommon cause of kyphosis in pediatric patients; however, if discovered late it may be confused with type II congenital kyphosis. Knutsson (61), in 1949, was the first to describe PAVF in the English-language literature, and fewer than 100 cases have since been reported (62–68). Because the largest reported series (26 patients) was from the University Hospital of Copenhagen (68), some have named this the Copenhagen syndrome. This condition is distinguishable from type II congenital kyphosis because the disc spaces and vertebral bodies are normal at birth and later become affected with an anterior fusion. Although the etiology is unknown, PAVF is probably a distinct clinical condition; however, it may represent a delayed type II congenital kyphosis.

Dubousset (34) suggested that certain forms of type II congenital kyphosis (failure of segmentation) may be inherited. The patients have a failure of segmentation, with delayed fusion of the anterior vertebral elements, which is not visible on radiographs until 8 or 10 years of age. He described one family in which three individuals had delayed ossification and congenital kyphosis, and another family in which the grandmother, mother, and two sisters had the deformity. Kharrat and Dubousset (62) also found this condition to be familial in 6 of 15 patients, and Van Buskirk et al. (63) reported associated anomalies in 7 of 15 patients, including heart defects, tibial agenesis, foot deformities, Klippel-Feil syndrome, Ito syndrome, pulmonary artery stenosis, and hemisacralization of L5.

Neurologic deficits are usually not seen in patients with PAVF, but Smith (63) reported one case of spinal cord compression resulting from an acutely angled kyphosis. Van Buskirk et al. (63) and Dubousset (28, 34) described five stages of PAVF: stage 1 is disc space narrowing, which occurs to a greater extent anteriorly than posteriorly; stage 2 is increased sclerosis of the vertebral end plates of the anterior and middle columns; stage 3 is fragmentation of the anterior vertebral end plates; stage 4 is fusion of the anterior and sometimes the middle columns; and stage 5 is development of a kyphotic deformity. Hughes and Saifuddin (67) described the MRI appearance of PAVF in three patients: early anterior disc narrowing (Fig. 19-18A), significant end-plate edema and fatty marrow changes (Fig. 19-18B), and finally multilevel anterior fusion and disc obliteration (Fig. 19-18C–E)

Kyphosis is the last stage in PAVF and is caused by the anterior disc space fusing while part of the posterior disc space remains open, allowing for continued growth in the posterior disc space and the posterior column. Bollini et al. (65) found that patients with thoracic PAVF had a relatively good prognosis, whereas those with lumbar involvement had a poor prognosis. Involvement of the thoracic spine is better tolerated by patients than is involvement of the lumbar area because of the normal kyphotic posture of the thoracic spine. Therefore, nonoperative treatment is recommended for most thoracic PAVF deformities. For PAVF in the lumbar spine, a posterior spinal fusion is indicated in stages 1, 2, and 3. In stages 4 and 5, the kyphotic deformity has already occurred in a normally lordotic lumbar spine. Posterior fusion will only stop progression of kyphotic deformity. If normal sagittal alignment is to be obtained, an anterior osteotomy followed by posterior fusion and instrumentation is recommended (61–68).

INFANTILE LUMBAR HYPOPLASIA

Campos et al. (69) reported thoracolumbar kyphosis secondary to lumbar hypoplasia in seven normal infants; the thoracolumbar kyphosis resolved spontaneously with growth. Patients presented with a clinically apparent kyphotic deformity in the first year of life. Radiographically, the patients had a relatively sharply angled kyphosis, with the apex at the affected vertebra (Fig. 19-19A). The affected vertebra had a wedge shape with an anterosuperior indentation, giving it a

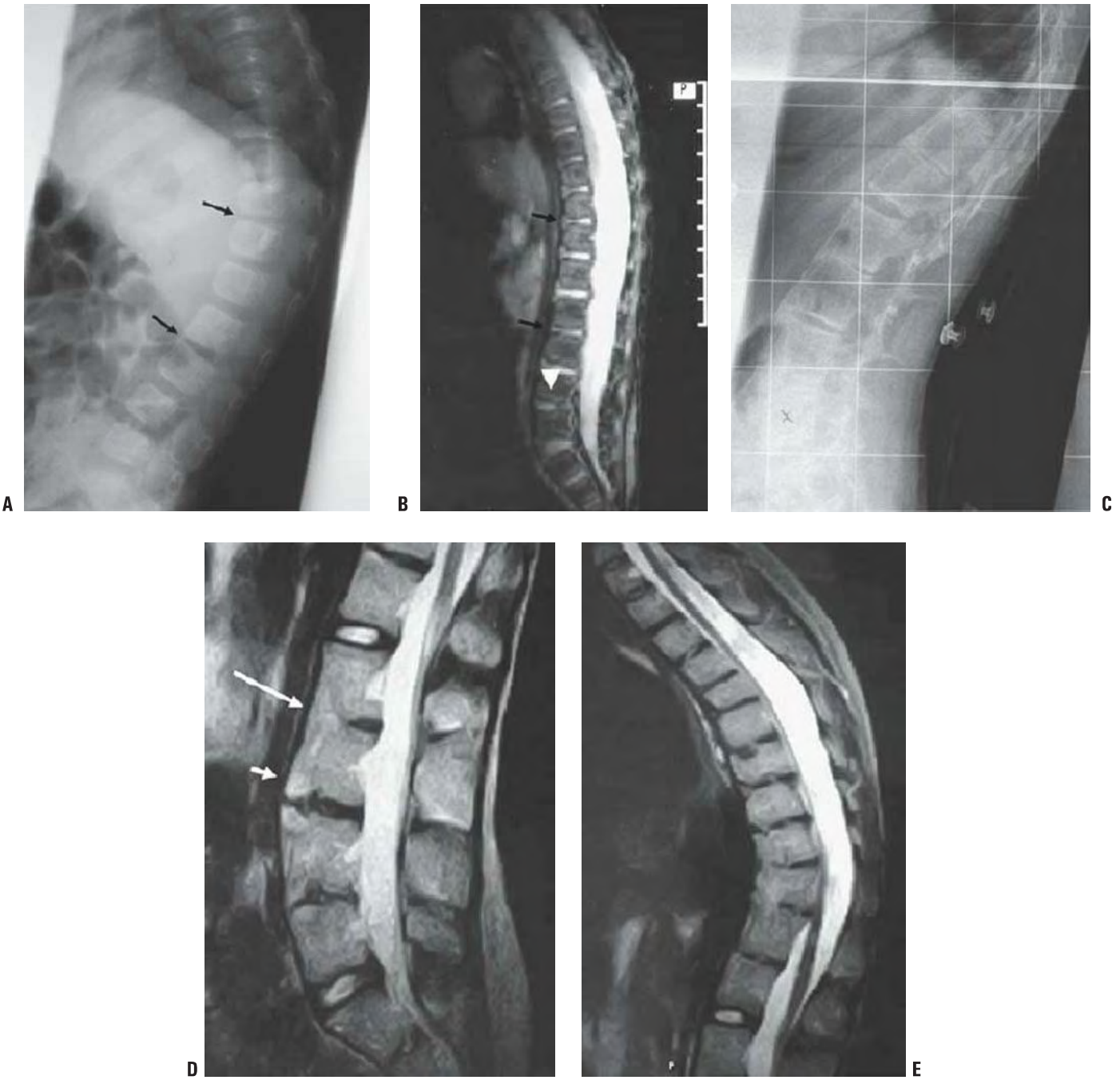


FIGURE 19-18. Progressive anterior vertebral fusion. **A**: Lateral radiograph of the thoracolumbar spine at age 12 months. Note narrowing at the T11/T12 and L2/L3 disc spaces anteriorly (*arrows*). **B**: MR imaging at age 12 months with sagittal STIR sequences through the thoracolumbar spine demonstrates early loss of anterior disc height at the T11/T12 and L2/L3 levels (*black arrows*). The horizontal high-signal intensity STIR abnormality (*white arrowhead*) at multiple end-plate levels is likely to represent normal physeal appearance at this age. **C**: Lateral radiograph (with gridlines for alignment) at age 12 years shows anterior fusion at multiple levels. **D**: MR image at age 12 years. Sagittal T2 FE sequences through the lumbar spine. Note solid fusion at the L2/L3 level (*long white arrow*), discovertebral anterior corner SI changes at the L3/L4 level (*short white arrow*), and fusion with the posterior elements. **E**: MR sagittal scanning through the thoracolumbar levels with T2 FSE sequences demonstrates T10/T11 and T11/T12 fusion and multilevel anterior disc space obliteration. (From Hughes RJ, Saifuddin A. Progressive non-infectious anterior vertebral fusion (Copenhagen syndrome) in three children: features on radiographs and MR imaging. *Skeletal Radiol* 1906;35:397–401.)

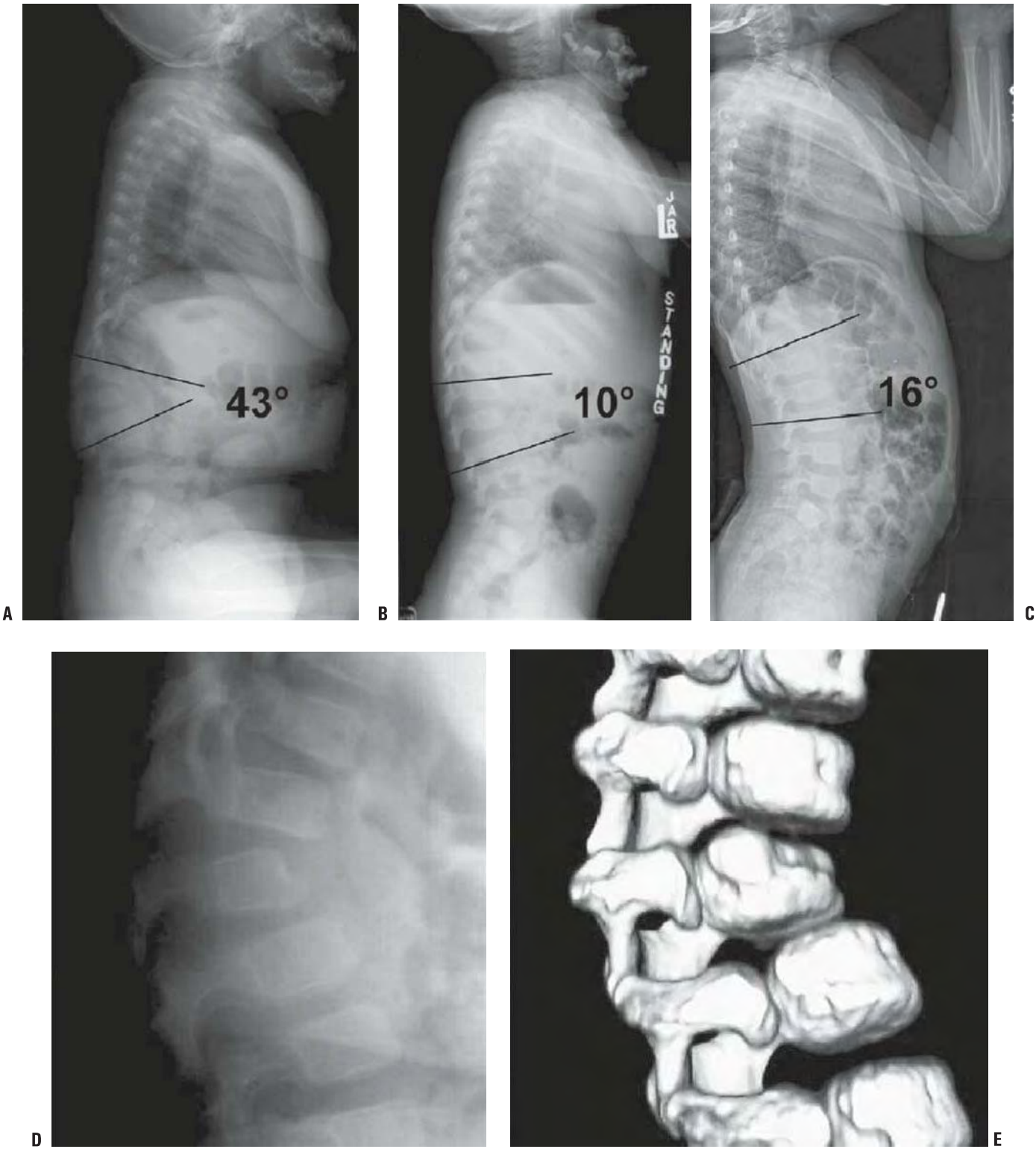


FIGURE 19-19. Spontaneous resolution of lumbar hypoplasia. Radiographs at 13 months of age **(A)**, 1 year and 11 months of age **(B)**, and 4 years and 6 months of age **(C)**. Radiograph **(D)** and computed tomographic three-dimensional reconstruction **(E)** show "beaked" L2 vertebra. (From Campos MA, Fernandes P, Dolan LA, et al. Infantile thoracolumbar kyphosis secondary to lumbar hypoplasia. *J Bone Joint Surg Am* 1908; 90:1726–1729.)

"beaked" appearance (Fig. 19-19B,C). Only one vertebra was involved in all seven infants, either at L1 or L2. The average initial kyphosis was 34 degrees. The kyphosis spontaneously improved after walking age and had corrected to normal by 6 years of age (Fig. 19-19D,E). Campos et al. recommended an initial period of observation for most patients with this type of congenital kyphosis to get a better assessment of the anomaly as ossification progresses and avoid overtreatment of lumbar hypoplasia that spontaneously improves with growth.

SEGMENTAL SPINAL DYSGENESIS

Segmental spinal dysgenesis is a congenital anomaly of the lumbar or thoracolumbar spine, consisting of focal agenesis or dysgenesis of the spine, and resulting in severe spinal stenosis and instability (70). A progressive kyphosis occurs at the site of segmental spinal dysgenesis. This condition often is confused with other spinal anomalies such as type I congenital kyphosis, sacral agenesis, lumbosacral agenesis, and lumbar agenesis. Faciszewski et al. (71) gave detailed radiographic and clinical definitions of this condition. Segmental spinal dysgenesis is characterized by severe focal stenosis of the spinal canal at the involved segment and is associated with significant narrowing of the thecal sac and absence of adjacent nerve roots. At the involved level, a ring of bone encircles the posteriorly positioned spinal canal, causing stenosis. The spinal canal is hourglass-shaped with no neurocentral junctions. There is limited potential for enlargement with growth because of the absence of neurocentral junctions, where growth occurs (Fig. 19-19). No pedicles or spinous or transverse processes are present at this level. Anterior to the bony ring is a fat-filled space. The distal bony anatomy and the spinal canal are usually normal, although spina bifida has been noted in a few cases (72). Neurologic function can range from normal to complete paraplegia. Associated anomalies are common, and there is a high incidence of neurogenic bladder (Fig. 19-20).

The etiology of segmental spinal dysgenesis is unknown. The diagnosis can be made on the basis of plain radiographs, but MRI and CT scans and three-dimensional reconstructions are usually needed to fully show the extent of this condition. Tortori-Donati et al. found that the patient's clinical status correlated with the amount of neural tissue seen on MRI at the level of the lesion (73). Progressive kyphosis occurs with this condition, and progressive neurologic deterioration was noted by Flynn et al. (74) and Faciszewski et al. (71). Early anterior and posterior fusions, with or without

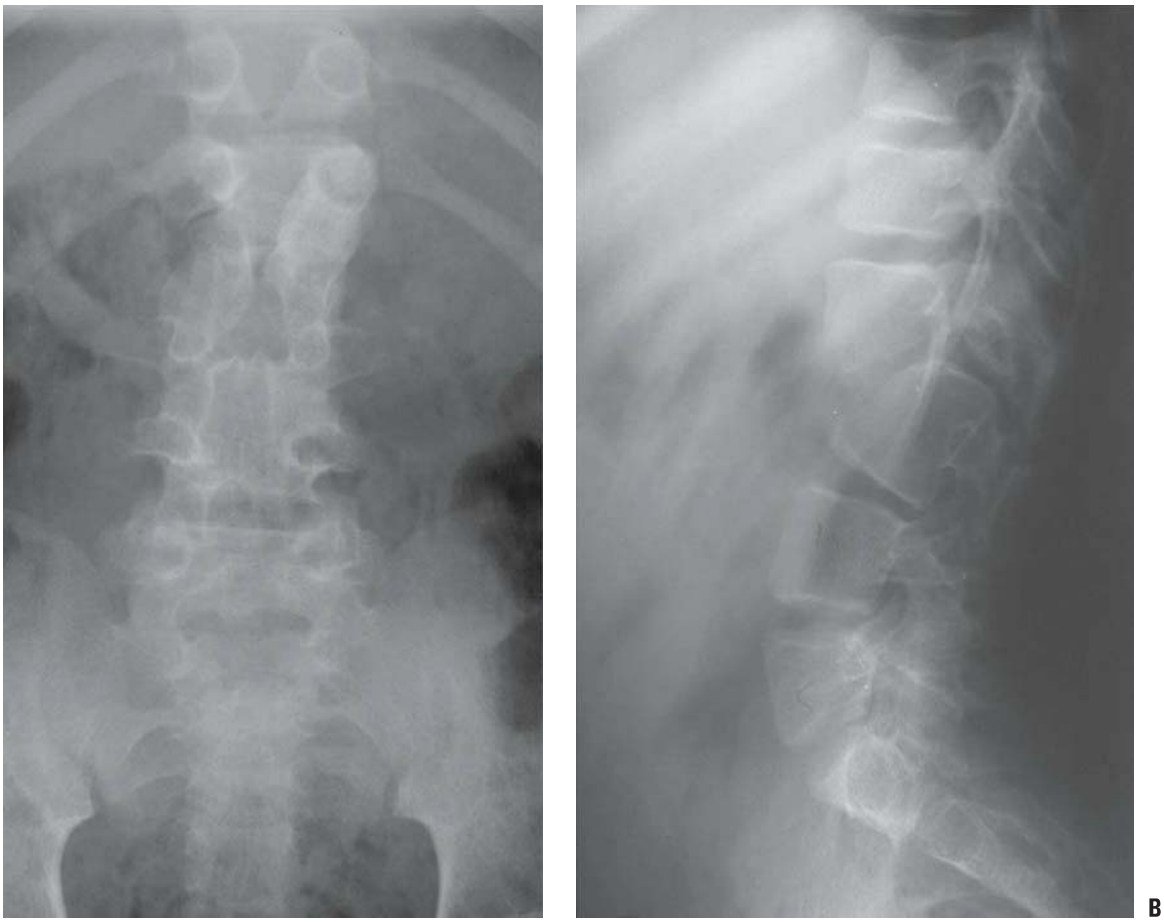


FIGURE 19-20. Segmental spinal dysgenesis. Anteroposterior **(A)** and lateral **(B)** radiographs show narrowing of spinal canal and absence of L1 and part of L2 vertebral bodies.

decompression, are recommended. The use of spinal instrumentation is controversial because of the small size of the patient. Hughes et al. (72) recommended that treatment be directed toward the establishment and maintenance of spinal stability first and toward decompression of the cord secondarily. Bristol et al. recommended rigid spinal immobilization for 12 to 18 months to allow growth and development before spinal fusion (75).

SACRAL AGENESIS

Sacral agenesis consists of a complete or partial absence of the sacrum (76–79). Rarely is it associated with absence of the most caudal segment of the lumbar spine. The association with maternal diabetes has been well documented (76–79). Kyphosis may occur with this condition, although it usually is not progressive and does not require treatment (80, 81).

SCHEUERMANN DISEASE

Scheuermann disease is a common cause of structural kyphosis in the thoracic, thoracolumbar, and lumbar spine. Scheuermann originally described this rigid juvenile kyphosis in 1919; it is characterized by vertebral-body wedging that is believed to be caused by a growth disturbance of the vertebral end plates (82, 83) (Fig. 19-21).

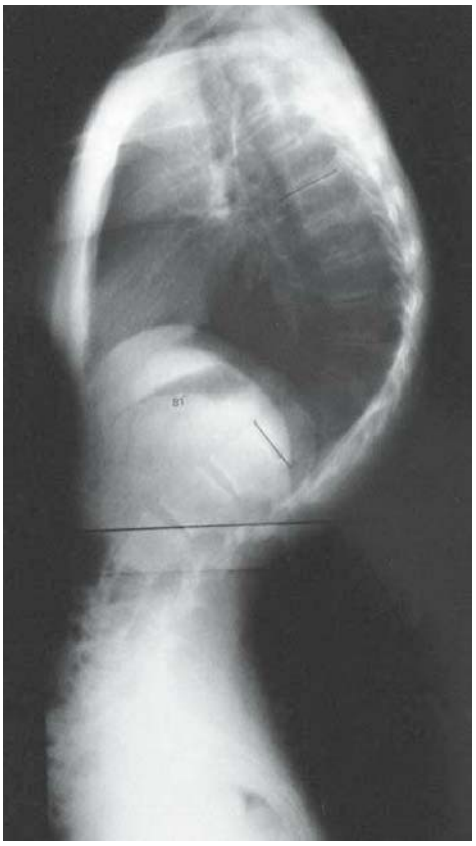


FIGURE 19-21. Lateral radiograph of a patient with Scheuermann disease and an 81-degree kyphotic deformity. Note the narrowing of the intervertebral disc spaces and the irregularity of the vertebral end plates. There is an associated increase in lumbar lordosis below the kyphotic deformity.

Classification. Scheuermann disease can be divided into two distinct groups: a typical form and an atypical form. These two types are determined by the location and natural history of the kyphosis, including symptoms occurring during adolescence and after growth is completed. Typical Scheuermann disease usually involves the thoracic spine, with a well-established natural history during adolescence and after skeletal maturity (84). In this classic form of Scheuermann kyphosis three or more consecutive vertebrae, each wedged 5 degrees or more (Sorensen criteria), produce a structural kyphosis. In contrast, atypical Scheuermann disease usually is located in the thoracolumbar junction or in the lumbar spine, and its natural history is well defined. The atypical type is characterized by vertebral end-plate changes, disc space narrowing, and anterior Schmorl nodes but does not necessarily fulfill Sorensen's criteria of three consecutively wedged vertebrae of 5 degrees. Thoracic Scheuermann is the more common form, with the atypical form less frequently seen.

Epidemiology. Typical Scheuermann disease consists of a rigid thoracic kyphosis in a juvenile or adolescent spine. The apex of kyphosis is located between T7 and T9 (11). The reported incidence of Scheuermann deformities in the general population ranges from 0.4% to 10% (85–89). Reported male-to-female ratios vary in the literature. Scheuermann originally reported a male preponderance of 88% (82). Most reports in the literature note either a slight male preponderance or an equal male-to-female ratio (87–92). Bradford et al. (86) have been the only ones to report an increased incidence of Scheuermann disease in women.

The age at onset of Scheuermann kyphosis is during the prepubertal growth spurt, between 10 and 12 years of age. Sorensen (88) described a Scheuermann prodrome in patients who had a lax, asthenic posture from the age of approximately 4 to 8 years, and in whom, within a few years, a fixed kyphosis developed. The clinical detection of Scheuermann disease occurs at approximately 10 to 12 years of age. Wedging of apical vertebrae has not been reported before 10 years of age (93). Radiographic evidence of Scheuermann disease usually is not detectable in patients younger than 10 years of age because the ring apophysis is not yet ossified. Until the ring apophysis ossifies, vertebral-body wedging and irregularity of the end plate are difficult to measure on radiographs.

Etiology. Many possible etiologies have been suggested for Scheuermann disease, but the true cause remains unknown. Genetic, vascular, hormonal, metabolic, and mechanical factors have been suggested as causes of Scheuermann kyphosis. Sorensen (88) noted a high familial predilection, and Halal et al. (94), in a study of five families, and McKenzie and

Sillence (95), in a study of 12 families, suggested that the disease may be inherited in an autosomal dominant fashion with a high degree of penetrance. Additional support for a genetic basis for this condition is provided by Carr et al. (96, 97) in a report of Scheuermann disease occurring in identical twins and by Damborg et al. (98), who found an almost 3% prevalence and 74% heritability in a large group of twins (over 35,000 individuals). Halal et al. (94), McKenzie and Sillence (95), and Carr et al. (97) reported possible autosomal dominant inheritance of Scheuermann kyphosis.

Scheuermann believed that the kyphosis was caused by a form of avascular necrosis of the ring apophysis, which led to a growth disturbance resulting in a progressive kyphosis with growth (82, 83). The problem with this theory is that the ring apophysis contributes little, if at all, to the longitudinal growth of the vertebrae (97, 99). Bick and Copel (99) demonstrated that the ring apophysis lies outside the true cartilaginous physis and contributes nothing to the longitudinal growth of the vertebral body. Therefore, a disturbance in the ring apophysis should not affect growth of the vertebrae or cause vertebral wedging.

Schmorl (100) described a herniation of disc material through the cartilaginous end plate, known as *Schmorl nodes*. He believed that the herniation of disc material occurred because of a weakened end plate. The disc herniation was thought to damage the anterior end plate, resulting in abnormal growth, which in turn caused the kyphosis. There is a definite increased incidence of Schmorl nodes in patients with Scheuermann kyphosis, but the problem with this theory is that Schmorl nodes are found outside the area of kyphosis and also are present in individuals who have asymptomatic, normal spines and do not have a kyphotic deformity.

Ferguson (101) suggested that persistence of an anterior vascular groove altered the anterior growth of the vertebral body, but Aufdermaur and Spycher (102, 103) and Ippolito and Ponseti (104) were unable to document growth disturbances around the anterior vascular groove and concluded that persistence of an anterior vascular groove is a sign of immaturity of the spine. Lambrinudi (105) postulated that Scheuermann disease resulted from upright posture and a tight anterior longitudinal ligament. The fact that no cases of Scheuermann disease have been found in quadruped animals lends support to this theory (106). This has led to the more popular belief that the anterior end-plate changes are caused by mechanical forces in response to Wolff's law or the Hueter-Volkmann principle. Compression forces in the anterior physis cause a decrease in growth in the area of the kyphosis. Indirect support for this argument can be found in the changes in the wedging of the involved vertebral bodies and the reversal of these changes when bracing or casting is used in the immature spine. Scoles et al. (106) also supported this theory by demonstrating disorganized endochondral ossification in the involved vertebrae, similar to that seen in Blount disease. They concluded that the changes in endochondral ossification resulted from increased pressure on the vertebral physis.

Ascani et al. (107, 108) found that patients who have Scheuermann disease tend to be taller than normal for their chronologic and skeletal ages with bone age more advanced than their chronologic age. Because they found increased growth hormone levels in these patients, they suggested that the increased height and the advanced skeletal age could be caused by the increased growth hormone. The increased height and the more rapid growth may make the vertebral end plates more susceptible to increased pressure and result in the changes seen in Scheuermann disease. The increased growth hormone levels noted by Ascani et al. may also lead to a relative osteoporosis of the spine, which, in turn, may predispose the spine to the development of Scheuermann disease.

Bradford et al. (85, 109), Burner et al. (110), and Lopez et al. (111) reported in the 1980s that Scheuermann kyphosis may be caused by a form of juvenile osteoporosis. However, using quantitative CT scans, Gilsanz et al. (112) found no evidence of osteoporosis in patients with Scheuermann kyphosis compared with normal research subjects. The authors suggested that the technique used to determine osteoporosis might account for the differences between their report and those that show osteoporosis. In a study using single-photon absorptiometric analysis of cadaver vertebrae from patients with Scheuermann kyphosis, Scoles et al. (106) also found no evidence of osteoporosis.

What is shown by the histologic studies of Ascani et al. (107), Ippolito et al. (104, 113), and Scoles et al. (106) is that an alteration in endochondral ossification occurs. Whether this altered endochondral ossification is the cause or result of kyphosis is not known. Ippolito and Ponseti (104) found a decrease in the number of collagen fibers, which were thinner than normal, and an increase in proteoglycan content. Some areas of the altered end plate showed direct bone formation from cartilage instead of the normal physeal sequences of ossification. These studies help support the belief that Scheuermann kyphosis is an underlying growth problem of the anterior vertebral end plates.

Atypical Scheuermann kyphosis, or thoracolumbar and lumbar kyphosis, is believed to be caused by trauma to the immature spine, resulting in irregularities of the end plate (114).

Natural History. Many early studies suggested an unfavorable natural history for Scheuermann disease and recommended early treatment to prevent severe deformity, pain, impaired social functioning, embarrassment about physical appearance, myelopathy, degeneration of the disc spaces, spondylolisthesis, and cardiopulmonary failure. Despite these reports, few long-term follow-up studies of Scheuermann disease were performed until that of Murray et al. (87). Findings by Travaglini and Conti (39, 115), Murray et al. (87), and Lowe (116) suggest that the natural history of the disease tends to be benign.

The kyphotic deformity progresses rapidly during the adolescent growth spurt. Bradford et al. (117) noted that, among the patients who required brace treatment, more than half had progression of their deformities during this growth spurt before brace treatment was begun. Little is known about progression of the kyphosis after growth is completed, and whether it is similar to that in scoliosis. It is not well documented whether the kyphosis will continue to progress beyond a certain degree during adulthood.

Travaglini and Conte (39) found that the kyphosis did progress during adulthood, but few patients developed severe deformities. What is known is that patients with Scheuermann kyphosis have more intense back pain, jobs that require relatively little physical activity, less range of motion of the trunk in extension, and different localization of back pain than the general population who do not have Scheuermann kyphosis (87). Even with these findings, when compared with normal individuals, patients with Scheuermann kyphosis have no significant differences in self-esteem, social limitations, or level of recreational activities. The number of days they miss from work because of back pain also is similar.

The data regarding the natural history of Scheuermann disease suggest that, although patients may have some functional limitations, their lives are not seriously restricted and they have few clinical or functional problems. Pulmonary function actually increases in these patients, probably because of the increased diameter of the chest cavity, until their kyphosis is more than 100 degrees. Patients with kyphosis of more than 100 degrees have restricted pulmonary function. Another finding in patients with Scheuermann kyphosis was that disc degeneration was five times more likely to be seen on MRI in patients with Scheuermann compared with controls (118). The clinical significance of this finding is not known (76).

Associated Conditions. Mild-to-moderate scoliosis is present in about one-third of patients with Scheuermann disease (116), but the curves tend to be small, approximately 10 to 19 degrees. Scoliosis associated with Scheuermann disease usually has a benign natural history. The scoliotic curve rarely is progressive and usually does not require treatment. Deacon et al. (118, 119, 120) divided scoliotic curves in patients with Scheuermann disease into two types, based on the location of the curve and the rotation of the vertebrae into or away from the concavity of the scoliotic curve. In the first type of curves, the apices of scoliosis and kyphosis are the same and the curve is rotated toward the convexity. The rotation of the scoliotic curve is opposite to that normally seen in idiopathic scoliosis. Deacon et al. (101, 119) suggested that the difference in direction of rotation is caused by scoliosis occurring in a kyphotic spine, instead of the hypokyphotic or the lordotic spine that is common in idiopathic scoliosis. In the second type of curves, the apex of the scoliosis is above or below the apex of the kyphosis and the scoliotic curve is rotated into the concavity of the scoliosis, more like idiopathic scoliosis. This type of scoliosis seen with Scheuermann kyphosis is the more common, and it rarely progresses or requires treatment.

Lumbar spondylolysis is a frequently associated finding in Scheuermann kyphosis (Fig. 19-22). The suggested reason

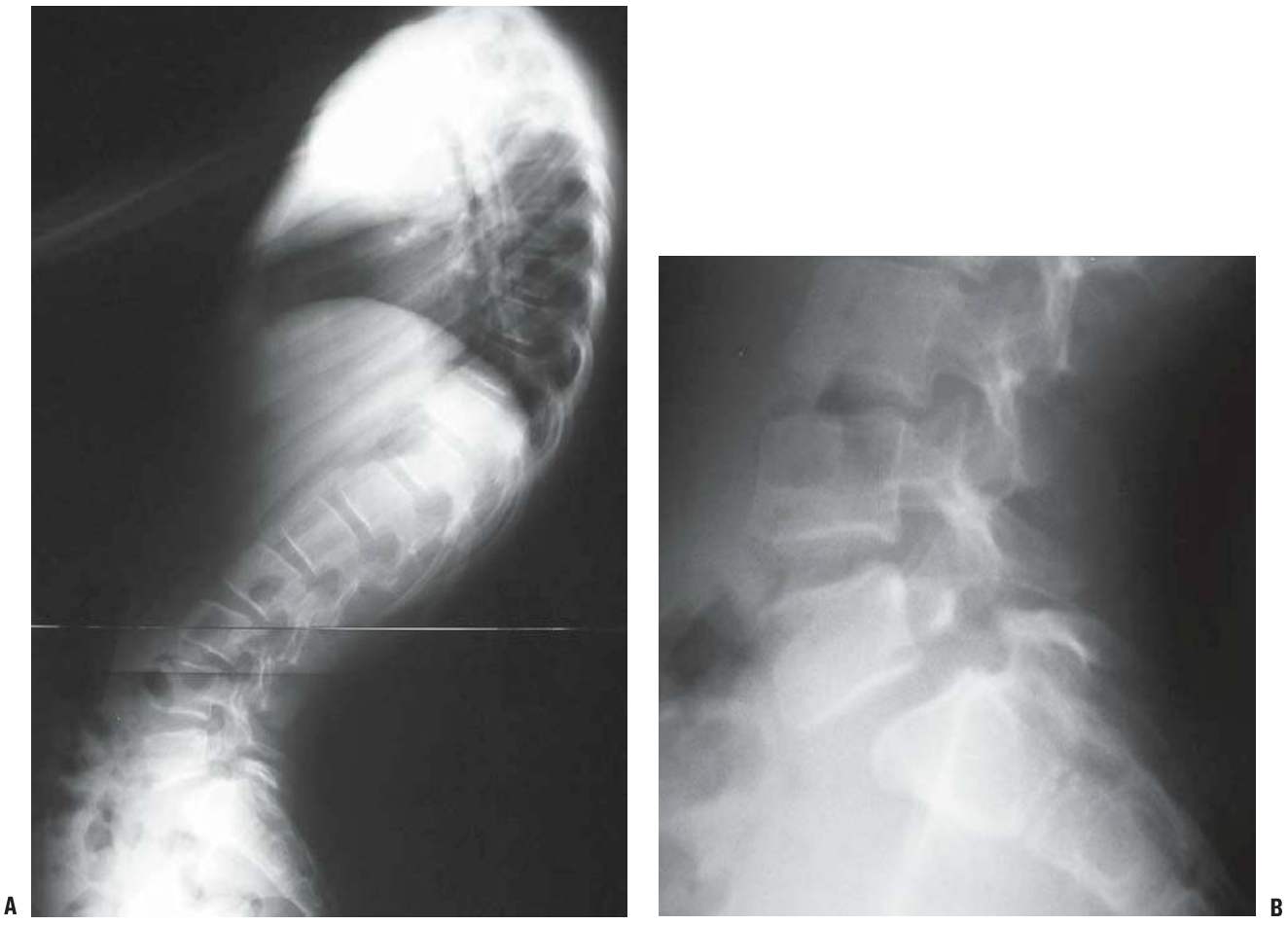


FIGURE 19-22. **A**,**B**: Lateral radiographs demonstrating spondylolisthesis with kyphosis.

for the increased incidence of spondylolysis is that increased stress is placed on the pars interarticularis because of the associated compensatory hyperlordosis of the lumbar spine in Scheuermann disease. This increased stress causes a fatigue fracture at the pars interarticularis, resulting in spondylolysis. Ogilvie and Sherman (121) found a 50% incidence of spondylolysis in the 18 patients they reviewed. Stoddard and Osborn reported a 54% incidence of spondylolysis in their patients with Scheuermann kyphosis (122).

Other conditions reported in patients with Scheuermann disease include endocrine abnormalities (123), hypovitaminosis (124), inflammatory disorders (122, 123), and dural cysts (106, 125).

Clinical Presentation. Clinical signs of Scheuermann disease occur around the time of puberty. The clinical feature that distinguishes postural kyphosis from Scheuermann kyphosis is rigidity. Often, mild Scheuermann disease is believed to be postural because the kyphosis may be more flexible in the early stages than in later stages. Usually, the patient seeks treatment because of a parent's concern about poor posture. Sometimes the poor posture has been present for several months or longer, or the parents may have noticed a recent change during a growth spurt. Attributing kyphotic deformity in a child to poor posture often causes a delay in diagnosis and treatment.

Pain may be the predominant clinical complaint rather than deformity. The pain generally is located over the area of the kyphotic deformity, but also occurs in the lower lumbar spine if compensatory lumbar lordosis is severe. Back pain usually is aggravated by standing, sitting, or physical activity. The distribution and intensity of the pain vary according to the age of the patient, the stage of the disease, the site of the kyphosis, and the severity of the deformity. Pain usually subsides with the cessation of growth, although pain in the thoracic spine can sometimes continue even after the patient is skeletally mature (87, 126). More commonly, after growth is completed patients complain of low back pain caused by the compensatory or exaggerated lumbar lordosis.

Most symptoms relating to Scheuermann disease occur during the rapid growth phase. During the growth spurt, pain is reported by 22% of patients, but as the end of the adolescent growth spurt approaches, this figure reaches 60%. Some authors believe that when growth is complete the pain recedes completely, except for well-circumscribed paraspinal discomfort (127–129). In adult patients with Scheuermann disease, pain may be located in and around the posterior iliac crest. This pain is thought to result from arthritic changes at T11 and T12, because the posterior crest is supplied by this dermatome. Stagnara (130) suggested that the mobile areas above and below the rigid segment are the source of pain.

Symptoms also depend on the apex of kyphosis. Murray et al. (87) noted that if the apex of kyphosis is in the upper thoracic spine, patients have more pain with everyday activities. The degree of kyphosis has also been correlated with symptoms. It seems logical that the larger the kyphosis, the more likely it is to be symptomatic, but Murray et al. found that curves between 65 and 85 degrees produced the most symptoms, whereas curves of more than 85 degrees and <65 degrees produced fewer symptoms. However, in patients with thoracolumbar or lumbar kyphosis (atypical Scheuermann disease), activity decreased as the degree of kyphosis increased.

Lumbar Scheuermann Disease. Patients with lumbar Scheuermann disease differ from those with thoracic deformity. These patients usually have low back pain but, unlike patients with the more common form of Scheuermann disease, their kyphotic deformity is not as noticeable. Pain is associated with spinal movement. Lumbar Scheuermann is especially common in men involved in competitive sports and in farm laborers, suggesting that the cause may be an injury to the vertebral physes from repeated trauma (131).

Physical Examination. In a patient with Scheuermann disease, a thorough examination of the back and a complete neurologic evaluation are essential. With the patient standing, the shoulders appear to be rounded and the head protrudes forward. The anterior bowing of the shoulders is caused by tight pectoralis muscles. Angular kyphosis is seen most clearly when the patient is viewed from a lateral position and is asked to bend forward. Normally, the back exhibits a gradual rounding with forward bending, but in patients with Scheuermann disease an acute increase is evident in the kyphosis of the thoracic spine or at the thoracolumbar junction. Stagnara et al. (132) found cutaneous pigmentation to be common at the most protruding spinous process at the apex of the kyphosis, probably the result of friction exerted by the backs of chairs and clothing. Compensatory lumbar and cervical lordosis, with forward protrusion of the head, further increases the anterior flexion of the trunk. Associated hamstring and hip flexor muscle tightness often is present.

The kyphotic deformity has some rigidity and will not correct completely with hyperextension. Larger degrees of kyphosis are not necessarily more rigid, and the amount of rigidity will vary with the age of the patient (87).

The neurologic evaluation usually is normal but must not be overlooked. Spinal cord compression has been reported occasionally in patients with Scheuermann disease (133–137). Three types of neural compression have been reported: ruptured thoracic disc (138), intraspinal extradural cyst, and mechanical cord compression at the apex of kyphosis; however, spinal cord compression and neurologic compromise are rare (139). Hughes et al. (67) found that only 1% of patients with a paralyzing disc herniation had Scheuermann disease. Ryan and Taylor (136) suggested that the factors influencing the onset of cord compression in patients whose cord compression is caused by the kyphosis alone are the angle of kyphosis, the number of segments involved, and the rate of change of the angle of kyphosis. This may be why neurologic findings are rare in Scheuermann kyphosis: the kyphosis occurs gradually, over several segments, and without acute angulation.

Radiographic Examination. The most important radiographic views are anteroposterior and lateral views of the spine with the patient standing. The amount of kyphosis present is determined by the Cobb method on a lateral radiograph of the spine. This is accomplished by selecting the cranial- and caudal-most tilted vertebrae in the kyphotic deformity. A line is drawn along the superior end plate of the most cranial vertebra and the inferior end plate of the most caudal vertebra. Lines are drawn perpendicular to the lines along the end plates, and the angle they form where they meet is the degree of kyphosis (140).

The criterion for diagnosis of Scheuermann disease on a lateral radiograph is more than 5 degrees of wedging of at least three adjacent vertebrae (88). The degree of wedging is determined by drawing one line parallel to the superior end plate and another line parallel to the inferior end plate of the vertebra, and measuring the angle formed by their intersection. Bradford and Garcia (141) suggested that three wedged vertebrae are not necessary for the diagnosis, but rather an abnormal, rigid kyphosis is indicative of Scheuermann disease.

The vertebral end plates are irregular, and the disc spaces are narrowed. The anteroposterior diameter of the apical vertebra frequently is increased (106) (Fig. 19-23). Associated Schmorl nodes often are seen in the vertebrae in the kyphosis.

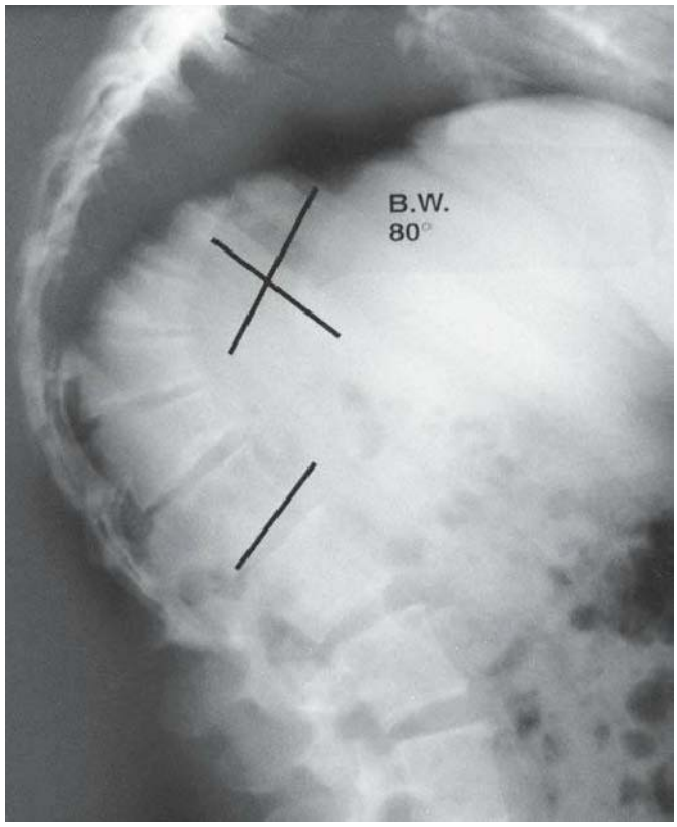


FIGURE 19-23. Lateral radiograph of a patient with Scheuermann disease demonstrates the kyphotic deformity seen in this disorder. Note the irregularity of the vertebral end plates and the anterior vertebral wedging.

Flexibility is determined by taking a lateral radiograph with the patient lying over a bolster placed at the apex of the deformity to hyperextend the spine and maximize the amount of correction seen on a hyperextension radiograph. On the lateral radiographs, most patients will be in negative sagittal balance (142). Sagittal balance is measured on the radiographs by dropping a plumb line from the center of the C7 vertebral body and measuring the distance from this line to the sacral promontory; a positive value indicates that the plumb line lies anterior to the promontory of the sacrum. Normal sagittal balance values are ±2 cm to the sacral promontory. On a lateral radiograph of lumbar Scheuermann kyphosis, irregular end plates, Schmorl nodes, and disc-space narrowing will be seen, but vertebral-body wedging is not as common. MRI and CT scans are necessary only if the patient has unusual symptoms or positive neurologic findings. An anteroposterior or a posteroanterior radiograph of the spine should be obtained to look for associated scoliosis or vertebral anomalies. The patient's skeletal maturity can be estimated from a radiograph of the left hand and wrist or from the Risser sign on the anteroposterior radiograph of the spine.

Treatment. The indications for the treatment of patients with Scheuermann kyphosis can be grouped into five general categories: pain, progression of deformity, neurologic compromise, cardiopulmonary compromise, and cosmesis.

Treatment options include observation, nonoperative methods, and surgery. Observation is an active form of treatment. If the deformity is mild and nonprogressive, the kyphosis can be observed every 4 to 6 months with lateral radiographs. The parents and the patient must understand the need for regular follow-up visits. If the deformity begins to progress, another form of treatment, such as bracing, casting, or surgery, may be indicated.

Nonoperative methods of treatment include exercise, physical therapy, bracing, and casting. Exercise and physical therapy alone will not permanently improve kyphosis that is caused by skeletal changes. The improvement seen with these methods is due to improved muscle tone and correction of bad posture. The goals of physical therapy are to increase flexibility of the spine, correct lumbar hyperlordosis, strengthen extensor muscles of the spine, and stretch tight hamstring and pectoralis muscles. The efficacy of this treatment method has not been proven, and although it may improve the postural component of Scheuermann disease, its effect on a rigid kyphosis is questionable.

Other nonoperative treatment methods can be divided into active correction systems (braces) and passive correction systems (casts). For either a brace or a cast to be effective, the kyphotic curve must be flexible enough to allow correction of at least 40% to 50% (93, 108, 143).

The Milwaukee brace is the brace recommended for the treatment of Scheuermann disease (144) (Fig. 19-24). The Milwaukee brace functions as a dynamic three-point orthosis that promotes extension of the thoracic spine. The neck ring maintains proper alignment of the upper thoracic spine,

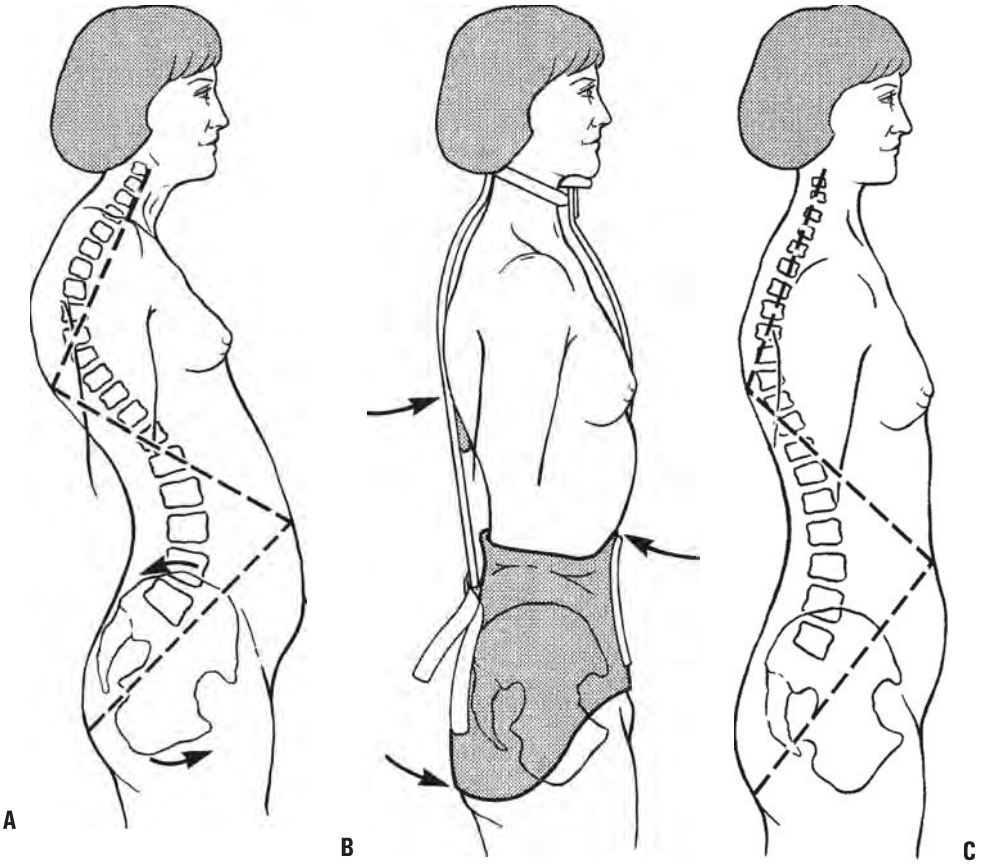


FIGURE 19-24. **A**: Patient with Scheuermann kyphosis has thoracic kyphosis, compensatory lumbar lordosis, anterior protrusion of the head, and rotation of the pelvis. **B**: Patient with Scheuermann kyphosis in a Milwaukee brace. The placement of the pelvic girdle, posterior thoracic pads, occipital pads, and neck ring encourages correction of the kyphosis. **C**: Correction of kyphosis after Milwaukee brace treatment. (Courtesy of Robert Winter, MD, Minneapolis, Minnesota.)

and the padded poster uprights apply pressure over the apex of the kyphosis. The pelvic girdle stabilizes the lumbar spine by flattening the lumbar lordosis. A low-profile brace, without a chin ring and with anterior shoulder pads, can be used for curves with an apex at the level of T9 or lower. The indications for brace treatment are an immature spine (at least 1 year of growth remaining in spine), some flexibility of the curve, and kyphosis of more than 50 degrees. The brace is initially worn full time for an average of 12 to 18 months. If the curve is stabilized and no progression is noted after this time, a part-time brace program can be used until skeletal maturity is reached. Gutowski and Renshaw (145) reported that part-time bracing (16 hours per day) was as effective as full-time bracing and was associated with improved patient compliance. In this study, a Boston lumbar orthosis was used to treat the kyphosis. The rationale for correction with this orthosis is that reduction of the lumbar lordosis causes the patient to dynamically straighten the thoracic kyphosis to maintain an upright posture. This presupposes a flexible thoracic kyphosis, a normal neurovestibular axis, and the absence of hip-flexion contractures.

Several orthopaedists have noted that, after initial improvement, there is a significant loss of correction after the discontinuation of brace treatment (56, 146). Montgomery and Erwin (92) stated that, if permanent correction of kyphosis is possible, a change in vertebral-body wedging should be seen before bracing is discontinued. Although some loss of correction can occur after bracing is discontinued, it still is effective in obtaining some correction of the kyphosis and possibly in reversing vertebral-body wedging, or at least preventing progression of the kyphotic deformity (92) (Fig. 19-25). Poor results with brace treatment have been reported in patients in whom the kyphosis exceeded 75 degrees or wedging of the vertebral bodies was more than 10 degrees and in patients near or past skeletal maturity (141).

Antigravity and localizer casts have been used extensively in Europe for nonoperative treatment of Scheuermann kyphosis, with good results (130, 147–149). De Mauroy and Stagnara (147) developed a therapeutic regimen that uses serial casts for correction. This method consists of three stages. First, a physical therapy program is started in preparation for the casts. Next, three sequential antigravity casts, changed at 45-day intervals, are applied to obtain gradual correction of the deformity. The third stage involves the use of a plastic maintenance brace that is worn until skeletal maturity is reached. With this regimen the deformity was reported to improve by 40%, and there was less loss of correction after this form of nonoperative treatment was discontinued (130, 148, 149).

The indications for surgical correction remain unclear because of various opinions about pain, disability, trunk deformity, and importance of cosmesis. Therefore, the decision for surgery must be made on an individual basis. The current indications for surgery are a progressive kyphosis of more than 75 degrees and significant kyphosis associated with pain that is not alleviated by nonoperative treatment methods. The biomechanical principles of correction of kyphosis secondary to Scheuermann disease include

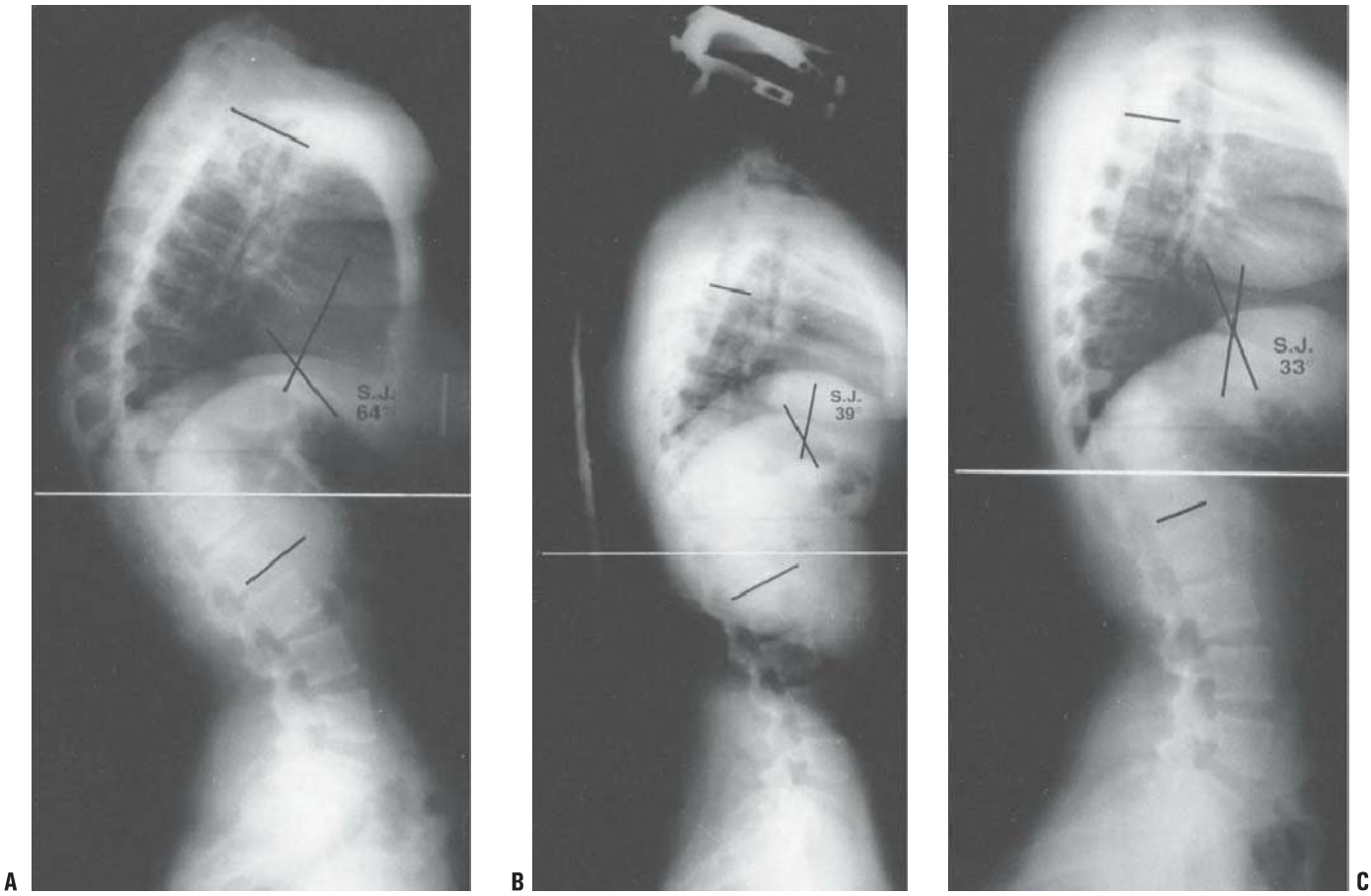


FIGURE 19-25. **A**: Lateral radiograph of a 15-year-old girl with a 64-degree thoracic kyphosis secondary to Scheuermann disease. **B**: Lateral radiograph of the patient in a Milwaukee brace with the kyphotic deformity improved to 39 degrees. **C**: Lateral radiograph obtained after the patient completed brace treatment; the kyphotic deformity has improved to 33 degrees.

lengthening the anterior column (anterior release), providing anterior support (interbody fusion), and shortening and stabilizing the posterior column (compression instrumentation and arthrodesis) (150). Surgical correction of kyphosis can be achieved by a posterior approach (Fig. 19-26), an anterior approach, or a combined anterior and posterior approach. The combined anterior and posterior approach has been the most frequently recommended and reported procedure (151–154). With the development of pedicle screw fixation and posterior spinal osteotomy techniques, such as the Ponte procedure (Fig. 19-27), there has been renewed interested in posterior-only surgery. A standard posterior procedure without osteotomy can be considered if the kyphosis can be corrected to, and maintained at, <50 degrees while a posterior fusion occurs (146–148, 155, 156). Historically, the use of Harrington compression rods was common, but these rarely are used now because of the frequent complications, including rod breakage, and the need for postoperative immobilization. Anterior instrumentation for Scheuermann disease was described by Kostuik (157); it consists of anterior interbody fusion and anterior instrumentation with a Harrington distraction system augmented by postoperative bracing. Although Kostuik reported good results with this technique, the anterior-only instrumentation approach for treatment of Scheuermann kyphosis is not widely used.

The Ponte osteotomy (Fig. 19-27) is performed using Kerrison rongeurs and begins by completely excising the ligamentum flavum, which allows for maximal mobilization of the spine. The osteotomy involves the complete resection of the inferior articular process of the cranial vertebra and the superior articular process of the caudal vertebra laterally to the neural foramen at each level. The osteotomies can be widened cranially and caudally at each level, typically in the range of 4 to 6 mm, depending on the amount of correction required. Typically, 5 to 10 degrees of correction at each level can be achieved (158).

When anterior and posterior surgeries together are used for Scheuermann disease, the anterior release and fusion are done first. The anterior release can be done through an open anterior exposure or by thoracoscopy. While thoracoscopic release may offer advantages over open thoracotomy, such as decreased postoperative pain, scarring, and impact on pulmonary function, the technique is technically challenging and has a high complication rate. Herrera-Soto et al. showed good sagittal correction, with no loss of

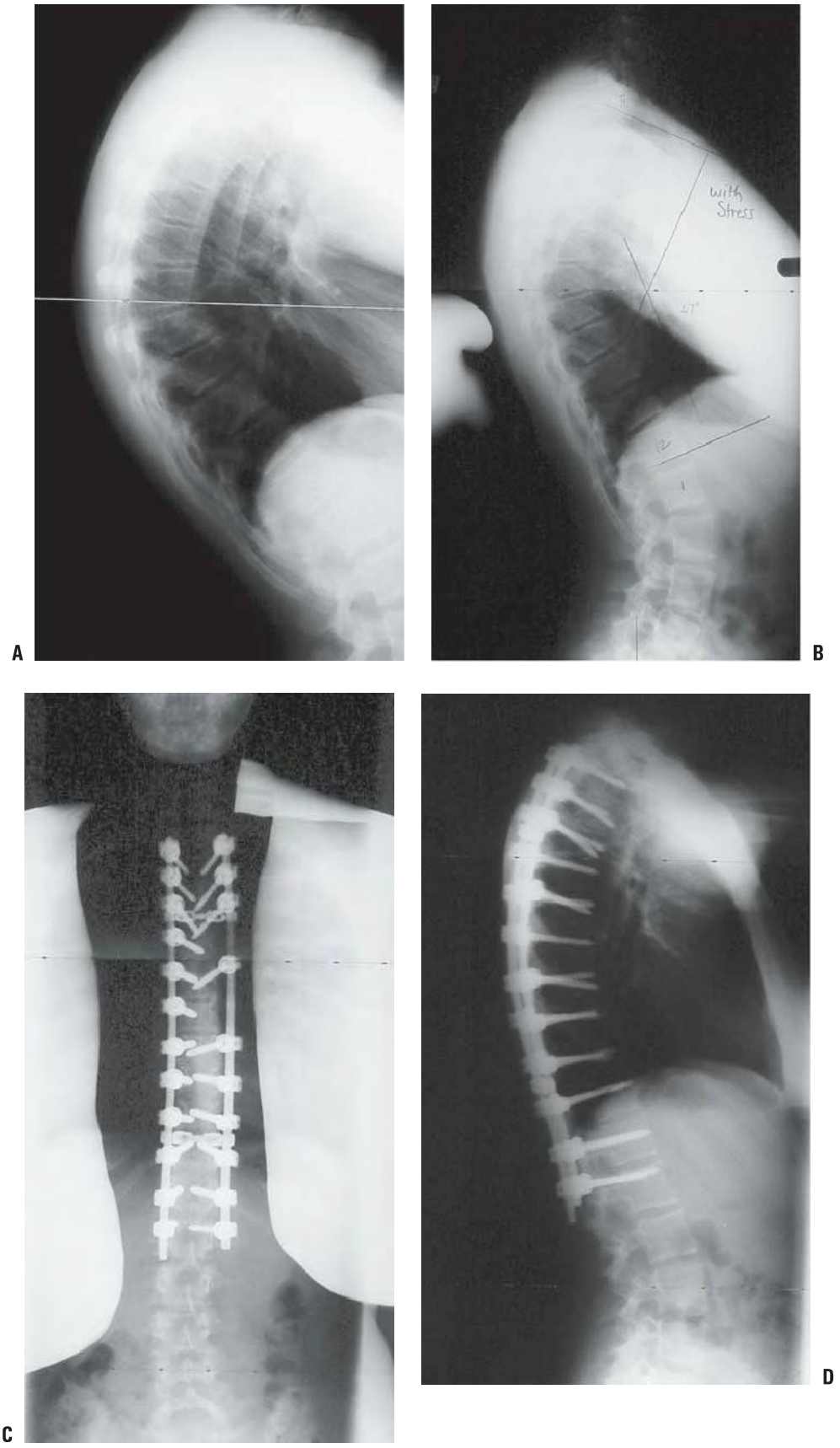


FIGURE 19-26. **A:** Thoracic Scheuermann kyphosis. **B:** After posterior fusion and instrumentation with pedicle screws. **C** and **D:** Postoperative status of posterior instrumentation and fusion with pedicle screws. (Courtesy of Dr. Anant Kumar.)

correction or junctional kyphosis using this technique; however, they had 11 complications in 9 of 19 patients, including 2 pneumothoraces and a deep venous thrombosis leading to a pulmonary embolus (159). Interbody cages have been used in an effort to improve sagittal correction (160, 161); however, Arun et al. (160) found no difference in outcomes between patients with anterior fusions using interbody cages compared to those with anterior fusions using autogenous rib graft.

The posterior fusion and instrumentation usually are done on the same day as the anterior release and fusion, but they can be done in a staged manner. For the posterior spinal fusion, a segmental instrumentation system using multiple hooks or pedicle screws or a hybrid of hooks and screws is used. Lowe (153) and Coscia et al. (162) reported high complication rates after using Luque rods and wires for posterior fixation, because this system does not allow for any compression. The use of posterior spinal osteotomies such as the Ponte

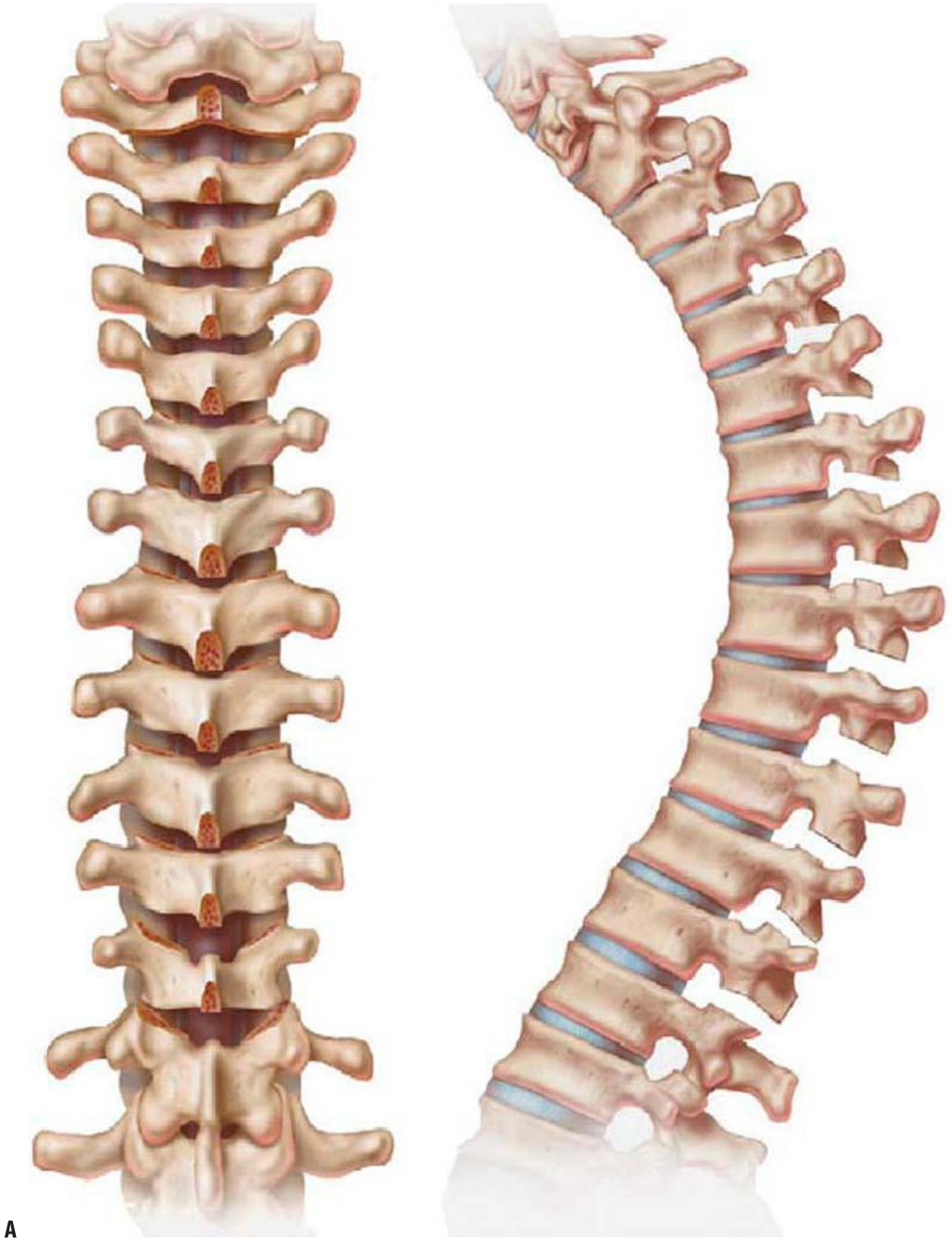


FIGURE 19-27. Ponte procedure: posterior-only osteotomies. **A**: Osteotomies. **B**: Anchor placement. **C**: Initial rod placement with proximal anchor compression. **D**: Final rod placement with final rod compression and deformity correction. (From Geck MJ, Macagno A, Ponte A, et al. The Ponte procedure: Posterior only treatment of Scheuermann's kyphosis using segmental posterior shortening and pedicle screw instrumentation. *J Spinal Disord Tech* 1907;19:586–593.)

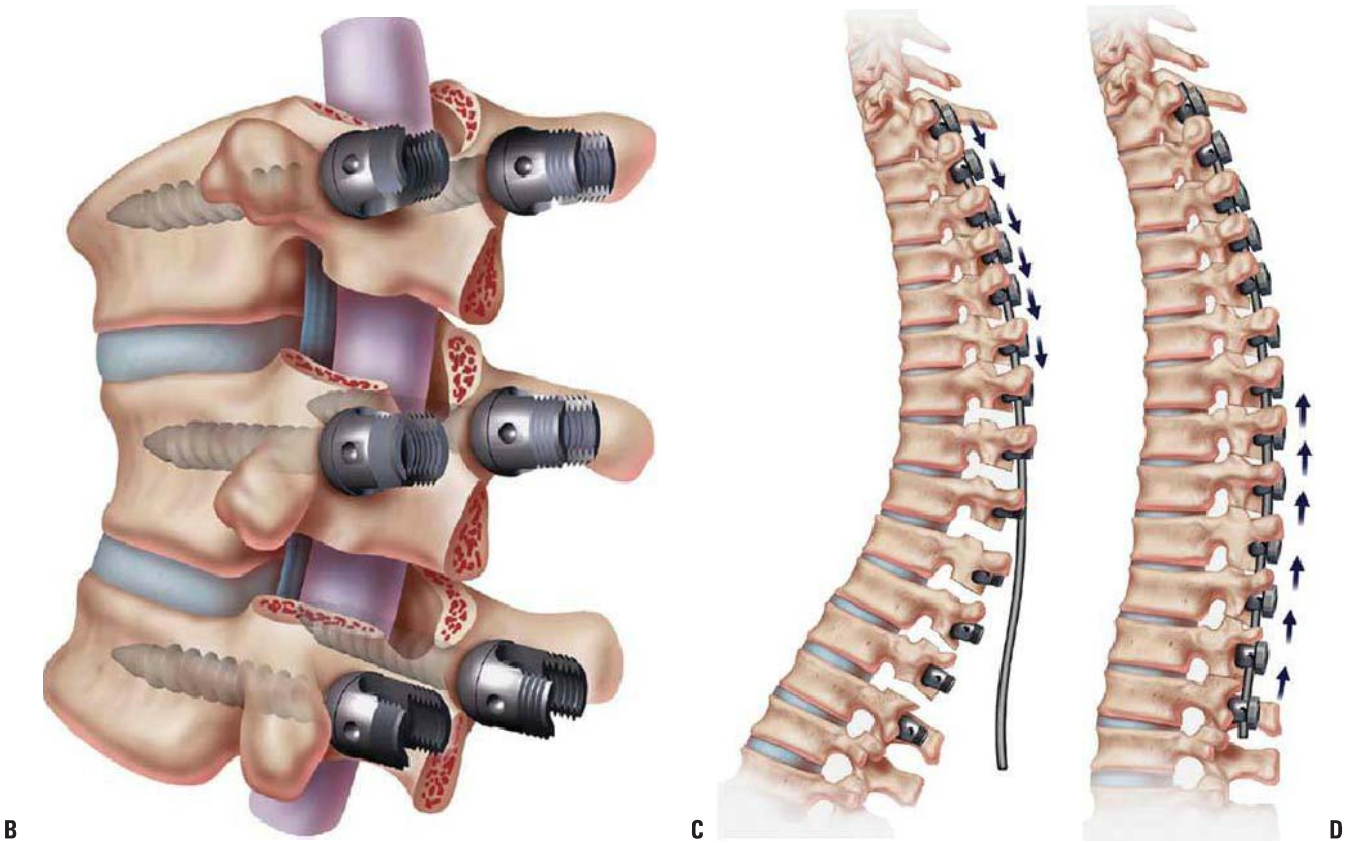


FIGURE 19-27 (*continued*)

osteotomy allows relative shortening of the posterior column, which allows greater correction of the kyphosis. Several studies have shown similar outcomes in terms of sagittal correction between patients with combined anterior–posterior release and those with posterior-only procedures with Ponte osteotomies (158, 163–165). In addition, patients with combined anterior–posterior release have more complications, longer surgical times, and more blood loss than those with posterior osteotomy and fusion alone (164, 165).

Regardless of the type of instrumentation used and whether or not other procedures such as anterior release or spinal osteotomy are done, posterior instrumentation should include at least three fixation points above the apex and at least two fixation points below the apex of the kyphosis. The fusion and instrumentation should include the proximal vertebra in the measured kyphotic deformity and the first lordotic disc distally (114, 142, 150, 166). If the fusion and instrumentation end in the kyphotic deformity, a junctional kyphosis at the end of the instrumentation is likely to develop (165).

POSTERIOR HOOK INSTRUMENTATION

This procedure has several attractive features for the treatment of kyphosis (Figs. 19-28 to 19-30). It is very rigid and requires no postoperative immobilization. In patients who have significant scoliosis in addition to kyphosis, it has the ability to correct both the kyphosis and the scoliosis. The hooks can be placed independent of the rod (unlike the classic Harrington compression rod); it is not a problem to insert the hooks on the other side of the spine after the first hooks have been placed and tightened.

There are also disadvantages. The rods must be contoured to the desired correction, which means that most of the correction is obtained at once. This makes it difficult to get the rod into the inferior hooks in closed-hook systems after they have been placed in the superior hooks. The use of pedicle screws at the bottom of the instrumentation or a hybrid construct obviates this to a large extent.

The area of the spine that is to be fused is exposed, and the hook sites are prepared.

Two methods of hook purchase can be used in the instrumentation of kyphosis. They differ in the method used to place the hooks on the thoracic vertebrae (Figs. 19-31).

Regardless of the technique used, junctional decompensation has been reported to occur in as many as 30% of patients (142, 165). Lowe (150, 153) emphasized that overcorrection of the deformity should be avoided to prevent junctional kyphosis. He recommended that no more than 50% of the preoperative kyphosis be corrected and that the final kyphosis should never be <40 degrees. He also found that patients with Scheuermann disease tend to be in negative sagittal balance and become further negatively balanced after surgery, which may predispose them to the development of junctional kyphosis (142). Lonner et al. (165) found

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Posterior Hook Instrumentation (Figs. 19-28 to 19-30)

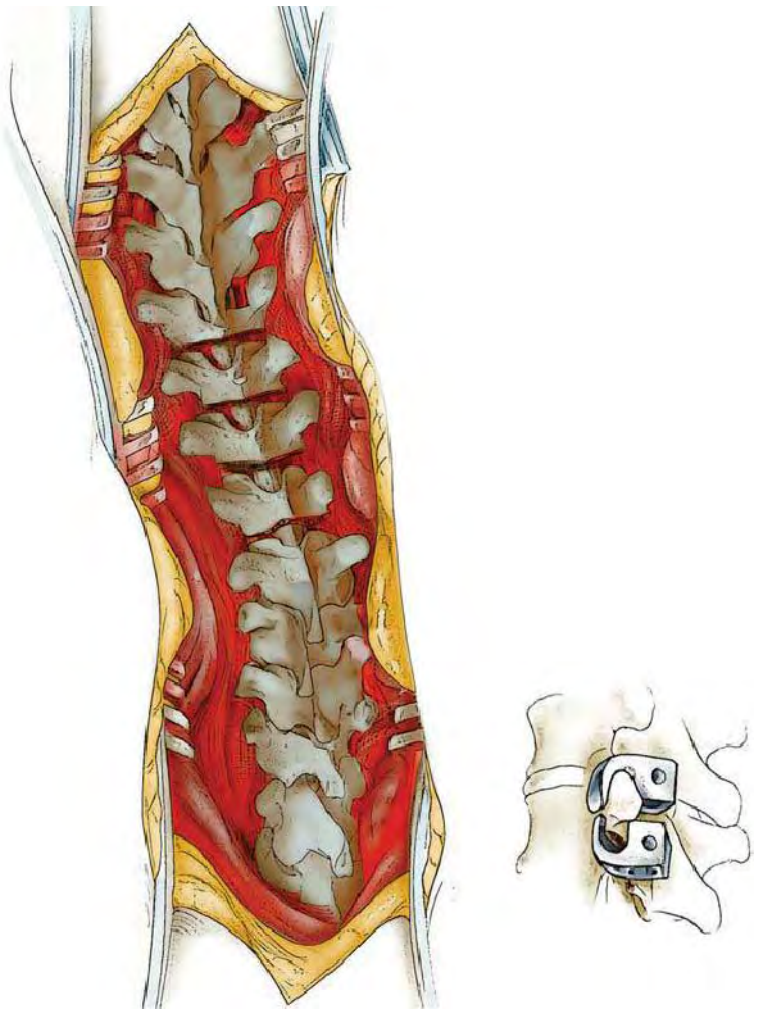


FIGURE 19-28. Posterior Hook Instrumentation. The method of hook purchase illustrated here uses the claw configuration on the thoracic vertebrae. On the cephalad side of the kyphosis, there should be at least three purchase sites on each side of the spine. These purchase sites may be of several combinations of claws, supralaminar hooks, and transverse process hooks, all depending on the bone strength, the rigidity of the curve, and the surgeon's choice. Some surgeons prefer to use supralaminar hooks as purchase sites, as opposed to the transverse processes. Others prefer to place the pedicle hook component of the claw one level distal. In this case, two claw configurations were used. The third hook was a simple transverse process hook. An alternative method on the cephalad portion of the kyphosis is the use of lamina hooks inserted into every other lamina. These can be staggered on either side of the spine. For example, a lamina hook may be inserted on the lamina of T3, T5, and T7 on one side of the spine and on the lamina of T4, T6, and T8 on the other side of the spine. These hooks are inserted on the cephalad aspect of the lamina to provide compression. Three hook sites should be prepared on each side of the spine inferior to the kyphosis. It is important when selecting levels to extend the instrumentation into the normal lordosis. These hook sites are prepared easily by removing the inferior edge of the lamina and then the ligamentum flavum to allow the lamina hook to be seated within the spinal canal. The hook sites should be prepared on both sides of the spine before any hooks or rods are placed. If this is not done, the closing of the interlaminar spaces as a result of placing the first rod makes it more difficult to prepare the sites on the opposite side. The use of pedicle screws at the lower end of the kyphosis makes insertion of the rod easier, although they may not make the correction any better. After this is completed, a radical facetectomy, with removal of a significant portion of the inferior part of the lamina, is performed in the area of the kyphosis to permit correction. This can be accomplished by entering the spinal canal in the midline and using a Kerrison rongeur to remove the bone. The bone that is removed includes the inferior portion of the lamina and the superior facet, as well as a portion of the inferior facet.

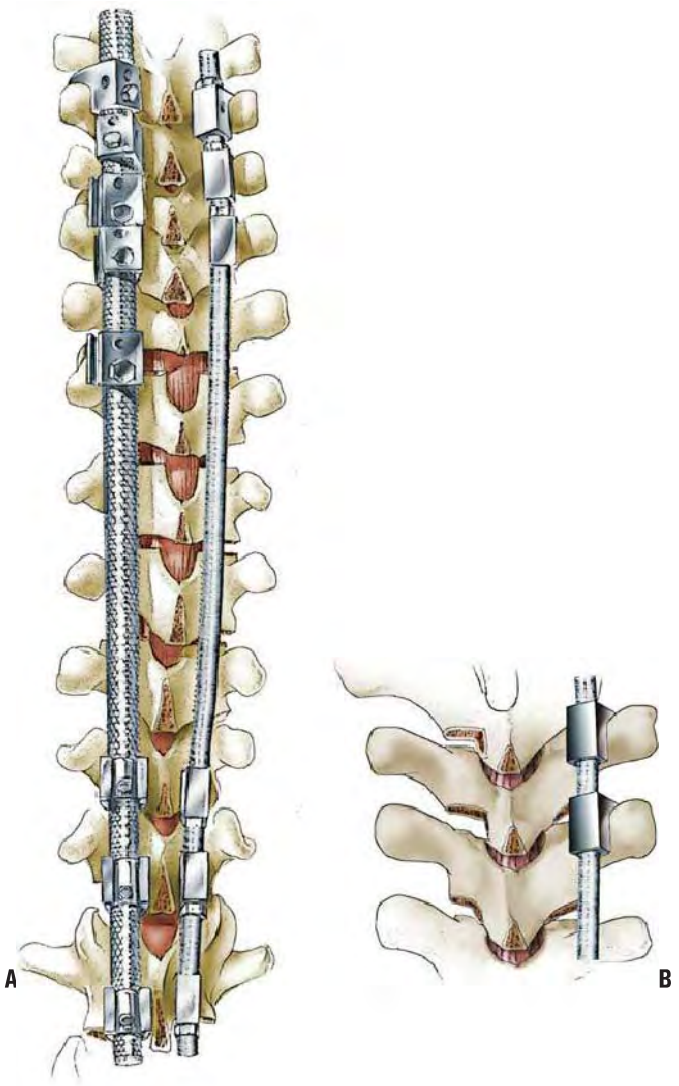


FIGURE 19-29. Now comes the most difficult part of this technique: placing the rods and the hooks. This is difficult because the rods must first be contoured to the desired final degree of correction; therefore, when they are inserted, most of the correction is gained at that time. If all the hooks and the rods are placed cephalad to the kyphosis, it is not easy to push them down into the caudal hooks. In a patient with severe kyphosis, the surgeon has the distinct impression that something will break with continued pushing. Several tricks have been suggested to deal with this problem, such as having an assistant push on the apex of the kyphosis, trying to lift the pelvis, or placing one rod in the cephalad hooks and one rod in the caudal hooks and pushing both down toward their corresponding empty hooks at the same time, as in a double-lever system. These methods may work in the case of flexible curves. Another method is to apply a small Harrington compression rod to one side, tighten it to gain correction, and then place the rigid rod system on the opposite side. The Harrington compression rod is then removed and replaced with the second rod **(A)**. In the thoracic region, the Harrington compression rod **(B)** can be placed on the transverse processes. These are usually strong enough for this temporary correction, and the hooks can be inserted rapidly. Below the kyphosis, the Harrington hooks can be placed in the holes that have been prepared for the hooks of the rigid rod system. With the newer top-opening systems, the rod can be secured in the hooks proximal to the kyphosis apex and then cantilevered into the hooks below. Compressive forces are then applied to continue the kyphosis correction.

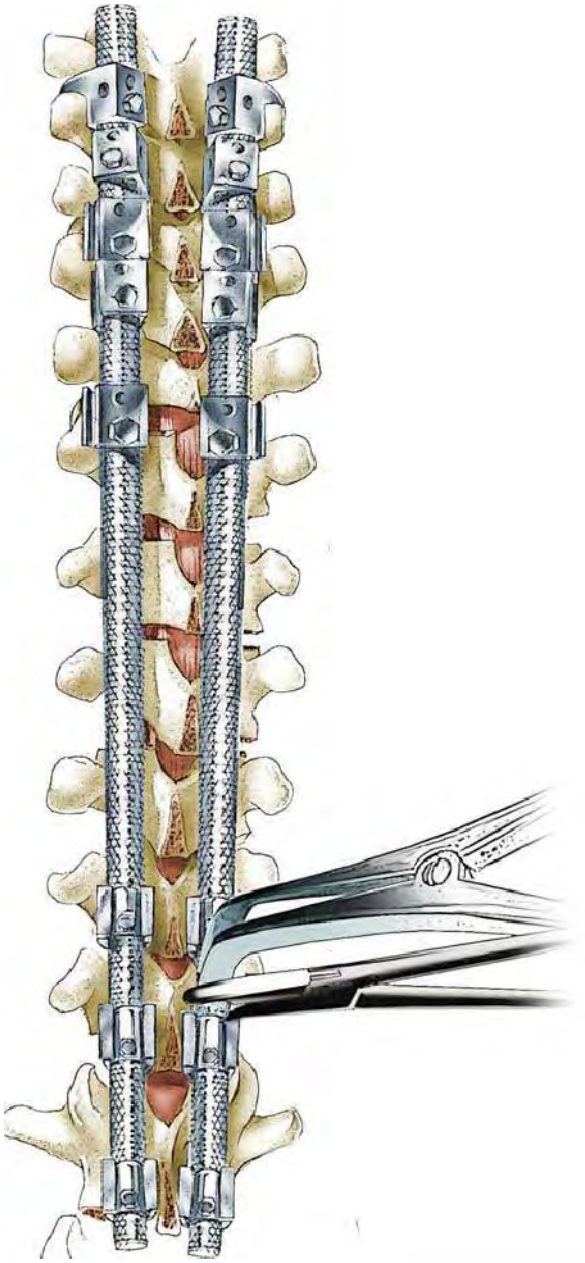


FIGURE 19-30. After both rods are placed, most of the correction would have been obtained if the rods were contoured correctly. Some additional correction may be obtained by tightening the hooks in compression, as was done with the Harrington compression rod, spreading between the hook and a rod holder clamped onto the rod. This has the additional advantage of tightening the hook against the bone and should be performed for each hook. To complete the operation, all possible decortication is accomplished and a large amount of bone graft is added.

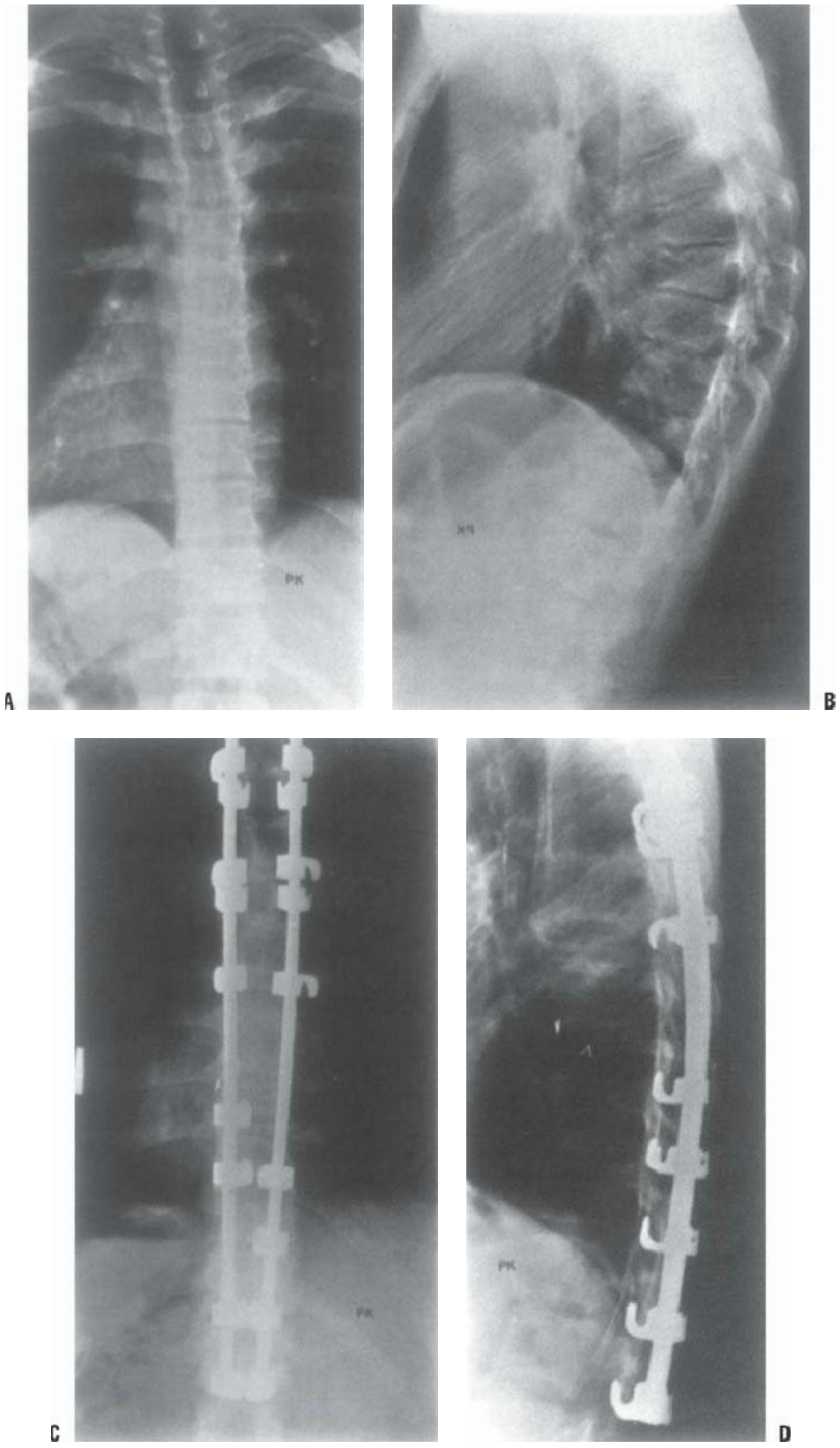


FIGURE 19-31. **A,B:** Anteroposterior and lateral radiographs of a 17-year-old boy with persisting pain secondary to Scheuermann kyphosis. **C,D:** Anteroposterior and lateral radiographs hook instrumentation in place. The upper hooks skipped a level to permit easier insertion, and the lower hooks were staggered to facilitate better decortication.

that pelvic incidence may be related to the amount of proximal junctional kyphosis and that distal junctional kyphosis was related to fusion that ended cranial to the neutral sagittal vertebra.

POSTLAMINECTOMY KYPHOSIS

A laminectomy or multiple laminectomies are needed most often in children for the diagnosis and treatment of spinal cord tumors, but also may be needed for other conditions such as neurofibromatosis, Arnold-Chiari malformation, and syringomyelia (167, 168). Although deformity after laminectomy is unusual in adults, it is common in children because of the unique and dynamic nature of the growing spine (138, 169–173). Younger age appears to be the most significant risk factor for the development of postlaminectomy cervical spine deformities (174). Postlaminectomy deformities usually result in kyphotic deformity, but a scoliotic deformity also may occur (170).

The pathophysiology of postlaminectomy kyphotic deformity can be multifactorial. Deformity of the spine after multiple laminectomies can be caused by (a) skeletal deficiencies (facet joint, laminae, and associated anterior column defects), (b) ligamentous deficiencies, (c) neuromuscular imbalance, (d) effects of gravity, and (e) progressive osseous deformity resulting from growth disturbances (167, 175). Panjabi et al. (176) showed that with loss of posterior stabilizing structures caused by removal of the interspinous ligaments, spinous processes, and laminae, the normal flexion forces placed on the spine will produce kyphosis. Gravity places a flexion moment on the spine, producing compression force on the anterior vertebrae and discs and a tensile force on the remaining posterior structures. This may explain why postlaminectomy deformities occur most often in the cervical and thoracic spine and less often in the lumbar spine. Gravity tends to cause a kyphosis in the cervical and thoracic spine, whereas it accentuates the usual lordosis of the lumbar spine.

Skeletal deficiencies also can produce deformity. An important factor influencing the development of postlaminectomy deformity is the integrity of the facet joint (170, 176–178). If the facet joint is removed or damaged during surgery, deformity is likely to develop. Raynor et al. (179) and Zdeblick et al. (180) found that, if more than 50% of the cervical facet was removed, instability and deformity of the cervical spine occurred. In addition, any secondary involvement of the anterior column, by tumor or surgical resection, adds to the risk of instability and deformity after laminectomy. Also, multiple laminectomies increase the risk of deformity when compared to single-level laminectomies (181, 182).

Insufficient soft-tissue restraints and paralysis of muscles that help stabilize the spine also can add to a postlaminectomy deformity. The spine is unable to resist the normal flexion forces placed on it by gravity and by the normal flexor muscles (183). Yasuoka et al. (184) noted increased wedging of the vertebrae and excessive motion after laminectomy in children, but not in adults. This increased wedging is caused by increased pressure on the cartilaginous end plates of the vertebral bodies. With time, the increased pressure causes a decrease in growth of the anterior portion of the vertebrae, according to the Hueter-Volkmann principle (Fig. 19-32). Excessive spinal motion in children after laminectomy can be attributed to the facet joint anatomy in the cervical spine and the greater ligamentous laxity of growing children. The orientation of the cervical facet joint in children is more horizontal than in adults. This horizontal orientation offers less resistance to forces that tend to cause kyphosis in the cervical spine.

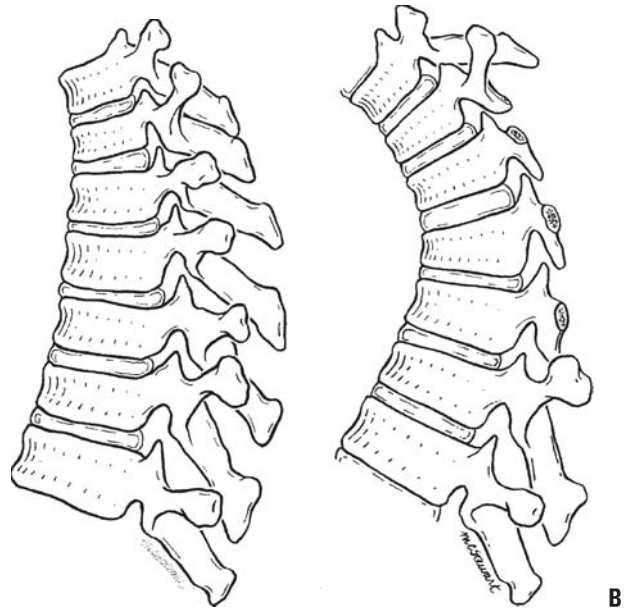


FIGURE 19-32. Drawings of the thoracic spine before and after repeated laminectomy demonstrate the effects on growth of the vertebral bodies. **A**: Before laminectomy, the anterior vertebral bodies are rectangular in configuration. **B**: The spine that has had multiple laminectomies will have increased compression anteriorly because of loss of posterior supporting structures. This compression results in less growth in the anterior portion of the vertebral body than in the posterior portion. In time, this will result in wedging of the vertebral bodies, causing a kyphotic deformity. (From Peterson HA. Iatrogenic spinal deformities. In: Weinstein SL, ed. *The pediatric spine: principles and practice*. New York, NY: Raven, 1994:651.)

Kyphosis is the most common deformity, although scoliosis also may occur, either as the primary deformity or in association with kyphosis. The incidence of postlaminectomy kyphotic deformity ranges from 33% to 100% (185), and depends on the age of the patient and the level of the laminectomy. Generally, the deformity is more likely in younger patients and after more cephalad laminectomy. For example, Yasuoka et al. (184) found that spinal deformity occurred in 46% of patients younger than 15 years of age, but in only 6% of patients 15 to 24 years of age. All the patients between 15 and 24 years of age in whom deformity developed were 18 years of age or younger. Yasuoka et al. (184) and Fraser et al. (186) found that higher levels of laminectomy were associated with a greater chance of deformity. In their studies, deformity

occurred after 100% of cervical spine laminectomies, after 36% of thoracic laminectomies, and in none of the lumbar laminectomies. Yeh et al. (182) and Papagelopoulos et al. (187) found that the greater the number of laminae removed, the greater the risk is for developing kyphosis.

Kyphosis in the cervical and thoracic spine is the most common postlaminectomy deformity (188). The lumbar spine is normally in lordosis, and this may protect it from developing kyphosis after multiple lumbar laminectomies. Papagelopoulos et al. (187) reported that hyperlordosis occurred in children who had lumbar laminectomies for intraspinal tumors. If the laminectomies extended into the thoracolumbar junction, kyphosis at the thoracolumbar junction occurred in 33% of their patients. Peter et al. (189) found that most of their patients did not develop a significant deformity after multiple lumbar laminectomies for selective posterior dorsal root rhizotomy; however, 9% developed spondylolysis. This may be the result of increased lordosis in this patient population (190).

Postlaminectomy deformity can occur early in the postoperative period or gradually over time. Kyphotic deformities have been reported to occur as late as 6 years after surgery (169, 190). Progression can be either sudden or gradual, or the deformity may progress significantly only during the adolescent growth spurt.

The natural history of postlaminectomy spinal deformity is varied and depends on the age of the patient at the time of surgery, the location of the laminectomy or laminectomies, and the integrity of the facet joint. Three types of postlaminectomy kyphosis have been described in children: (a) instability after facetectomy, (b) hypermobility between vertebral bodies associated with gradual rounding of the spine, and (c) wedging of vertebral bodies caused by growth disturbances (185).

Kyphosis from instability after facetectomy tends to be sharp and angular and usually occurs in the immediate or early postoperative period, causing associated loss of neurologic function (Fig. 19-33). Gradual rounding of the kyphotic deformity is seen more often when the facet joints are preserved. Kyphosis increases gradually over time because of the stress placed on the remaining posterior structures. If the spine is immature when the laminectomy is performed, the resulting kyphosis can inhibit the growth of the anterior physes of the involved vertebrae. Unequal growth results in wedge-shaped vertebrae and a progressive kyphotic deformity that is accelerated during the adolescent growth spurt.

Other associated conditions that also may add to or cause kyphotic deformities include persistent spinal cord tumors, neurologic deficits, intraspinal pathology (hydromyelia), and radiation therapy (191, 192).

Evaluation. The evaluation of a postlaminectomy deformity should focus on (a) the flexibility of the deformity, (b) loss of spinal structures, and (c) determination of future deformity with growth. The flexibility of a deformity can be estimated by flexion and extension lateral radiographs. If these cannot be obtained, a lateral traction film can be used. CT scans and three-dimensional reconstruction views may better delineate which bony elements are missing. MRI may be used but gives more information about the spinal cord, disc, and surrounding soft tissue than about the bony elements. To aid in preoperative planning, Lonstein (167) recommended drawing the spine preoperatively. The lines should represent the spinous processes and intact laminae and facet joints. This may aid in predicting progression of a postlaminectomy deformity.

Treatment. Treatment of postlaminectomy kyphosis is difficult, and it is best to prevent the deformity from occurring (193). The facet joints should be preserved whenever possible during laminectomy. Localized fusion at the time of facetectomy or laminectomy may help prevent progressive deformity (194). Because of the loss of bone mass posteriorly, however, localized fusion may not produce a large enough fusion mass to prevent kyphosis. Even so, this approach is advocated because it may produce enough bone mass posteriorly to stabilize what otherwise would be a severe progressive deformity.

The surgical technique of laminoplasty to expose the spinal cord may lessen the chance of progressive deformity. This approach involves suturing the laminae back in place after removal or removing just one side of the laminae and allowing them to hinge open like a book to expose the spinal cord, then suturing that side of the laminae back in place (195–197). This procedure may provide only a fibrous tether connecting the laminae to the spine, but studies have shown a decreased incidence of postlaminectomy kyphosis when it has been used (198, 199). Another technique is to hinge the laminae open in a lateral direction after dividing the laminae in the midline. This provides a lateral trough for the placement of bone graft for a lateral fusion (190, 191, 200, 201). The use of these techniques has been reported to decrease the incidence of postlaminectomy deformity, although there also have been reports of postlaminectomy deformity occurring even when laminoplasty was done (182, 192, 193, 202).

After surgery in which the laminae have been removed, bracing has been suggested to prevent deformity (194, 195, 203, 204), although no studies have documented the efficacy of this form of treatment. After the deformity has occurred and started to progress, bracing is ineffective in preventing further progression (167, 170).

For progressive or marked deformity, spinal fusion is recommended, although the patient's long-term prognosis should be considered before making definitive treatment plans. If the prognosis for survival is poor, spinal fusion may not be appropriate. However, given the availability of effective treatment protocols for tumors and the improved survival rates, fusion is usually indicated for progressive deformity. Combined anterior and posterior spinal fusion is preferred in most patients (196, 205) because the frequency of pseudarthrosis is greater if either procedure is done alone.

Lonstein (167) reported pseudarthrosis in 57% of patients after posterior fusion and in 15% of patients after anterior fusion. Anterior and posterior fusion can be done on the same day or as staged procedures. When the anterior procedure is done, care must be taken to remove all the physes back to the posterior longitudinal ligament. Leaving some of the physes in the vertebral body can cause an increase in

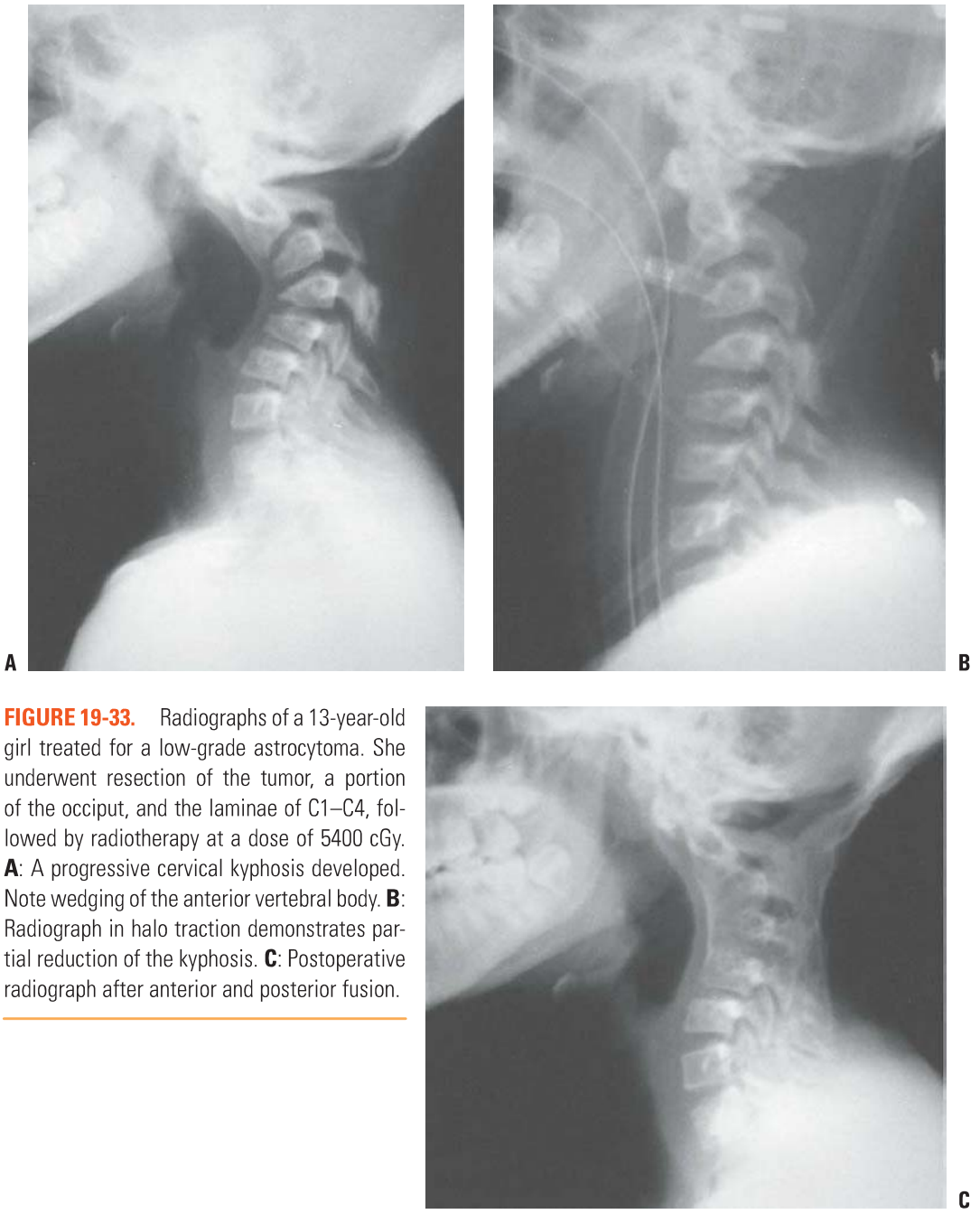


FIGURE 19-33. Radiographs of a 13-year-old girl treated for a low-grade astrocytoma. She underwent resection of the tumor, a portion of the occiput, and the laminae of C1–C4, followed by radiotherapy at a dose of 5400 cGy. **A**: A progressive cervical kyphosis developed. Note wedging of the anterior vertebral body. **B**: Radiograph in halo traction demonstrates partial reduction of the kyphosis. **C**: Postoperative radiograph after anterior and posterior fusion.

the deformity. When the posterior procedure is done, instrumentation of the involved spine is desirable, but not always possible, because of the absence of posterior elements. The development of pedicle screw fixation has been helpful in allowing the use of posterior instrumentation for postlaminectomy kyphosis. When it can be performed safely, this procedure provides secure fixation while the spinal fusion is maturing. Torpey et al. (206) recommended a posterior fusion using titanium rod instrumentation at the time of laminectomy. The instrumentation provides stability postoperatively, and the titanium rods allow for postoperative MRI to evaluate spinal cord tumors. In certain cases, anterior instrumentation with rod and bone screws or plates can be used to obtain stability and correction of the deformity (197). If the deformity is severe or long-standing, anterior release followed by halo traction or a halo cast with an Ilizarov device can be used for obtaining gradual correction (198, 199, 207, 208).

RADIATION KYPHOSIS

The relative radiosensitivity of growing cartilage was discovered by investigators during the 1940s, and animal studies documented radiation-induced growth inhibition in growing cartilage and bone (209–214). The longitudinal growth of a vertebral body takes place through normal endochondral ossification, similar to the longitudinal growth of the metaphyses of long bones. Bick and Copel (98, 99) demonstrated this on

histologic sections in fresh autopsy specimens of vertebral bodies taken from research subjects ranging in age from 14 weeks of fetal development to 23 years. This endochondral ossification at the physis is radiosensitive (98, 99, 209–211, 214, 215). Engel (209, 210) and Arkin and Simon (216) were able to produce spinal deformities in experimental animals using radiation. Arkin et al. (217) were the first to report spinal deformity in humans that was caused by radiation. After these reports, it has become clear that exposing an immature spine to radiation can produce spinal deformity, including scoliosis, kyphoscoliosis, lordoscoliosis, and kyphosis.

The three most common solid tumors of childhood for which radiation therapy is part of the treatment regimen, and in which the vertebral column is included in the radiation fields, are neuroblastoma, Wilms tumor, and medulloblastoma. Early in the history of radiation therapy, survival rates were poor and spinal deformities were not as prevalent. With improved treatment protocols and survival rates, the incidence of spinal deformities has increased. The degree of growth inhibition of the spine is related to the accumulated radiation dose and the age of the child when the spine is irradiated. Progression is directly dependent on the remaining growth potential in the irradiated vertebrae. The younger the child and the greater the accumulated radiation dose, the greater the chance of deformity (218–223). The most severe growth changes occur in patients who are 2 years of age or younger at the time of irradiation. Initial vertebral changes usually occur 6 months to 2 years after radiation exposure (224), but the deformity may not become apparent until years later, after a period of growth (218, 222).

Reports of radiation involving the spinal column show that an accumulated dose of <1000 cGy (centigray) does not produce a detectable inhibition of vertebral growth, whereas a dose of 1000 to 1900 cGy causes a temporary inhibiting effect on growth. Sometimes, this is manifested as a transverse growth arrest line in the vertebra, which gives the appearance of a bone within a bone. A dose of radiation between 1900 and 3000 cGy causes irregularity or scalloping of vertebral end plates and diminution of axial height and sometimes leads to a flattened, beaked vertebra (218, 220, 222, 224–228). A dose of 5000 cGy causes bone necrosis (192). The effect that radiation has on soft tissue also affects the progression of spinal deformity. The soft tissue anterior to the spine and the abdominal muscle can become fibrotic and act as a tether with growth, adding to the deformity of the spine as the child grows (229).

The incidence of spinal deformity after irradiation of the spine has been reported to range from 10% to 100% (218, 222, 223, 225, 230–233). These rates are decreasing because of shielding of growth centers, symmetric field selection, and decreased total accumulated radiation doses. The last of these changes has resulted from an increase in the use and effectiveness of chemotherapeutic regimens that reduce the need for large doses of radiation. Early reports showed an increased incidence of scoliotic deformities with the use of asymmetric radiation fields, and the incidence of kyphotic postirradiation deformities has increased with the use of symmetric radiation fields (234).

Any child who has received irradiation of the spine should be observed carefully for the development of spinal deformity. Because the development of deformity is related to the amount of disordered growth in the vertebral bodies that were affected by irradiation, it depends to a large extent on the amount of growth left in the spine when the irradiation was started and the amount of damage to the physes caused by irradiation (which correlates directly with the accumulated radiation dose). If the dose of radiation is large enough to cause permanent damage to the physes, the deformity will be progressive. Both postirradiation scoliosis and kyphosis progress more rapidly during times of rapid growth such as the adolescent growth period (222, 223, 226, 234). Before the adolescent growth spurt, the deformity may remain relatively stable or progress at a steady rate. Severe curves can continue to progress even after skeletal maturity, and these patients may require continued observation (Fig. 19-34).

Radiographic evaluation of a postirradiation deformity should include standard posteroanterior and lateral radiographs of the spine. Occasionally, CT scans with sagittal or coronal reconstruction are needed for better delineation of the vertebral-body deformities. The spinal cord and the surrounding soft tissue are evaluated best with MRI. Neuhauser et al. (220) described the radiographic changes seen in irradiated spines. The earliest changes were alterations in the vertebral bodies within the irradiated section of the spine caused by impairment of endochondral growth at the vertebral end plates. Growth arrest lines produced a bone-within-a-bone picture. This occurred in 28% of the 81 patients in the study by Riseborough et al. (222). Other radiographic changes were end-plate irregularity with an altered trabecular pattern and decreased vertebral-body height. This pattern was the most common radiographic change reported by Riseborough et al. (222) (83%). Contour abnormalities causing anterior narrowing and beaking of the vertebral bodies, much like those seen in patients with conditions that affect endochondral ossification (e.g., Morquio syndrome, achondroplasia), were the third type of radiographic change noted by Neuhauser et al. (220).

Treatment. Milwaukee brace treatment has been recommended for progressive curves, but generally has been ineffective for postirradiation kyphosis (222, 234), especially in patients with soft-tissue contractures contributing to the deformity. The irradiated skin also may be of poor quality, making long-term brace wear difficult. If progression occurs, spinal fusion, with or without instrumentation, should be done regardless of the age of the patient. Because bone quality is poor, fusion can be difficult to obtain after a single attempt. Anterior and posterior fusions are recommended and should extend at least one or two levels above and below the end of the kyphosis (185, 197, 222, 234, 236). The posterior fusion mass may require reexploration and repeated bone grafting after 6 months, and immobilization may need to be prolonged for 6 to 12 months. Posterior instrumentation should be used whenever feasible, because it adds increased stability while the fusion mass is maturing and may allow some limited correction of the kyphotic deformity (Fig. 19-34). Anterior instrumentation can be used in certain

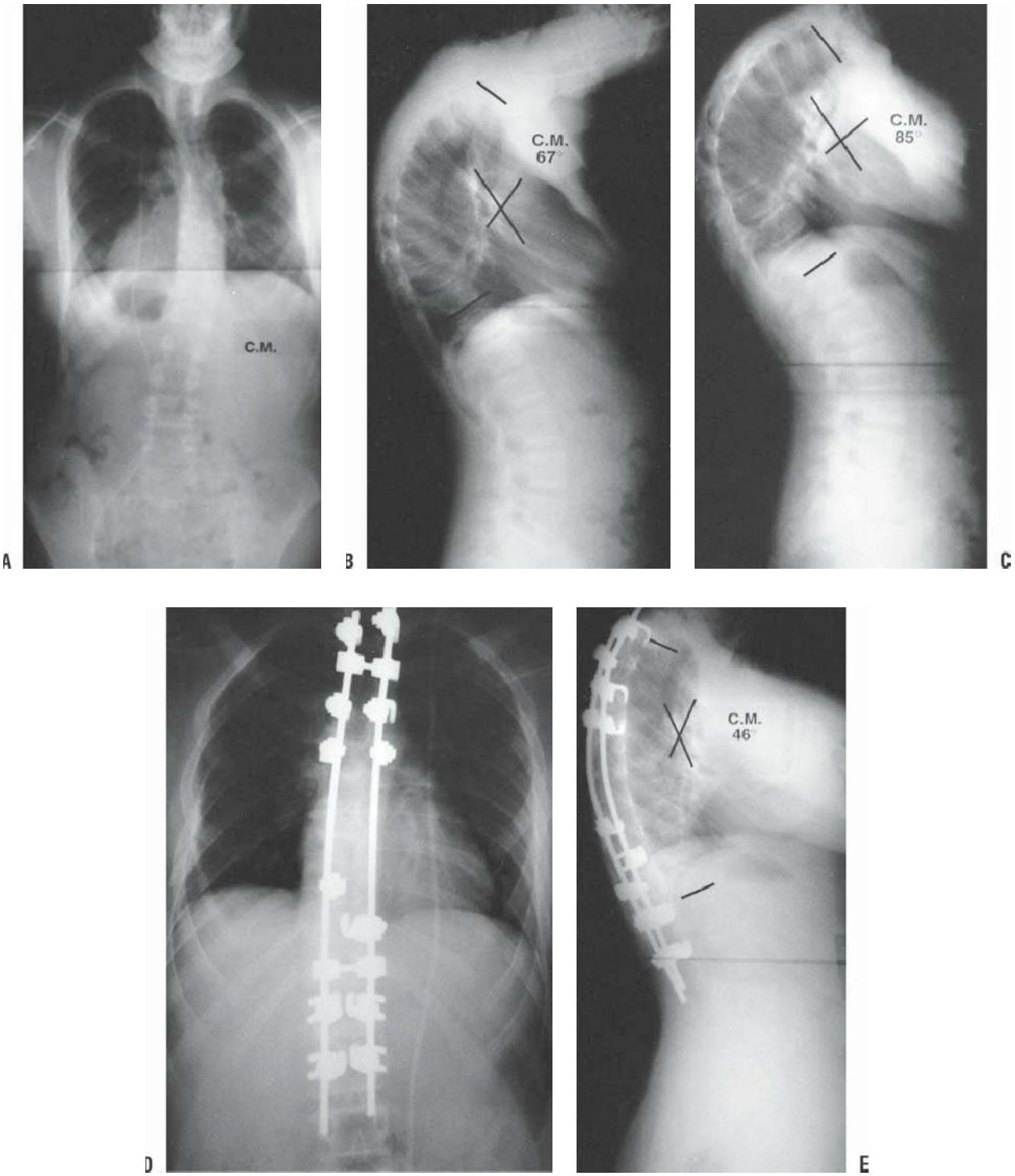


FIGURE 19-34. **A,B**: Anteroposterior and lateral radiographs of a 16-year-old child with a suprasellar germinoma treated with resection and 3400 cGy of radiation to the base of the skull and the entire spine. Radiographs demonstrate a 67-degree kyphosis with associated scoliosis. **C**: The kyphosis progressed to 85 degrees over 18 months despite bracing. **D,E**: Anteroposterior and lateral radiographs after anterior and posterior fusion with posterior instrumentation. The kyphosis has been corrected to 46 degrees.

cases; however, because of the radiation, the vertebral bodies usually remain in an infantile form, and instrumentation with bone screws may be difficult.

Correction of postirradiation kyphosis is difficult. Typically, these curves are rigid, and soft-tissue scarring and contractures often further hamper correction. Healing can be prolonged, and pseudarthrosis is common. Infection is a frequent complication in these patients because of poor vascularity of the irradiated tissue (222). Riseborough et al. (222) reported a pseudarthrosis rate of 37% and an infection rate of 23% in their patients after surgery. King and Stowe (234) also reported a high complication rate in patients who were treated surgically. Because viscera also can be damaged by irradiation, bowel obstruction, perforation, and fistula formation may occur after spinal fusion.

This can be difficult to differentiate from postoperative cast syndrome, and the treating physician should be aware of this complication (237). Radiation myelopathy also may occur in this patient population (238). King and Stowe (234) reported postoperative paraplegia in two of seven patients who had undergone radiation treatment for neuroblastoma and surgery for correction of their kyphotic spine deformity. King and Stowe concluded that these two patients had a subclinical form of radiation myelopathy and that spinal correction compromised what little vascular supply there was to the cord. Therefore, the surgeon should be aware of this possibility and try to avoid overcorrection.

MISCELLANEOUS CAUSES OF KYPHOTIC DEFORMITIES

Spinal deformity in the sagittal plane can occur in patients with skeletal dysplasia (239, 240). The natural history of spinal deformity varies with the type of deformity and the type of dysplasia. Some sagittal plane deformities that appear severe at birth or in infancy improve spontaneously with growth, whereas others continue to progress and eventually can cause paraplegia. A knowledge of the various skeletal dysplasias and the natural history of sagittal plane deformities in each is necessary to prevent overtreatment and undertreatment.

Achondroplasia. Treatment of spinal problems often is required in patients with achondroplasia. The most common sagittal plane deformity in achondroplastic dwarfs is thoracolumbar kyphosis (241–243). The kyphosis usually is detected at birth and is accentuated when the child is sitting because of the associated hypotonia in these infants (244) (Fig. 19-35). Ambulation is delayed until approximately 18 months of age, but after ambulation begins, the thoracolumbar kyphosis tends to improve. The kyphosis usually does not resolve in children who have more hypotonia. According to Lonstein (245), thoracolumbar kyphosis resolves in 70% of achondroplastic dwarfs and persists in 30%. In one-third of these patients, or 10% of achondroplastic dwarfs, the thoracolumbar kyphosis is progressive (245) (Fig. 19-36).

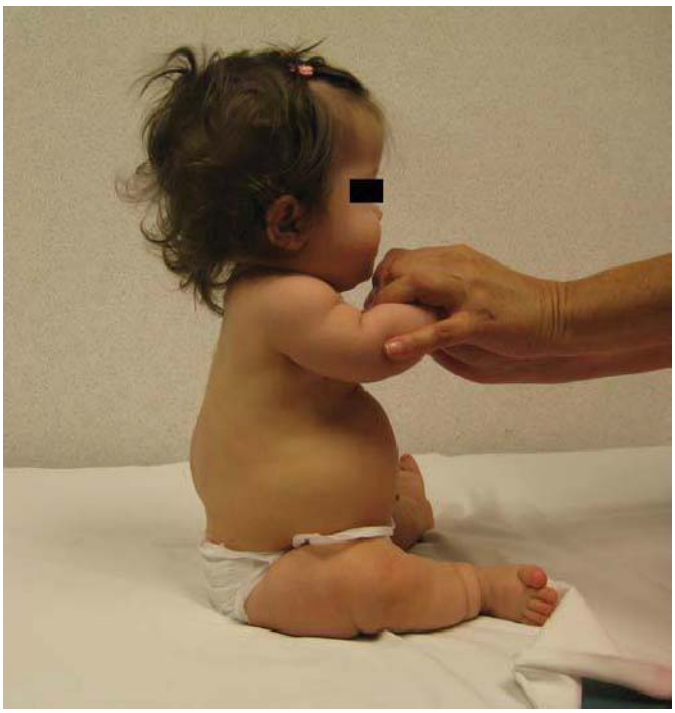


FIGURE 19-35. Achondroplastic dwarf with thoracolumbar kyphosis.

A lateral radiograph of the thoracolumbar spine during infancy shows anterior wedging of the vertebrae at the apex of the kyphosis (243). In patients whose thoracolumbar kyphosis resolves, the anterior vertebral-body wedging also improves. When the kyphosis is progressive, anterior vertebral-body wedging persists.

Sponseller listed three reasons why thoracolumbar kyphosis should be corrected: (a) it may cause pressure on the conus and result in neurologic symptoms; (b) it results in an increase in the compensatory lumbar lordosis, which can increase problems from an already stenotic lumbar spine; and (c) it may increase significantly if decompressive laminectomies are needed for lumbar stenosis in the future (44).

If no improvement in the thoracolumbar kyphosis is evident once a child begins walking, a thoracolumbosacral orthosis (TLSO) is recommended to try to prevent progression of the kyphosis (246–249). Early treatment to prevent the development of a progressive kyphosis was recommended by Pauli et al. (250). They developed an algorithm for treatment of young achondroplastic patients, first counseling the parents against unsupported sitting and continuing with close follow-up. If kyphosis develops and is >30 degrees, TLSO bracing is begun and continued until the child is walking independently and there is evidence of improvement in vertebral-body wedging and kyphosis. Using this form of early intervention, Pauli et al. (250) reported no occurrences of progressive kyphosis in 66 patients. Sponseller recommended serial hyperextension casting if the kyphosis does not respond to bracing. If there is a satisfactory response to serial casting (50% or more correction in 3 to 4 months), brace treatment can be resumed (244).

Indications for surgery are documented progression of a kyphotic deformity, kyphosis of more than 40 degrees in a child older than 5 or 6 years of age, and neurologic deficits relating to the spinal deformity (242, 244, 251). Distinguishing between neurologic deficits that result from a kyphotic deformity and those associated with lumbar stenosis (which is common in achondroplastic dwarfs) can be difficult. A thorough physical examination and diagnostic studies such as CT scan and MRI may be necessary to determine appropriate treatment. The infant should be evaluated for foramen magnum stenosis, because this may be the underlying cause for the hypotonia and delayed ambulation in achondroplastic patients with kyphosis. If present, the stenosis should be treated by decompression of the foramen magnum (252). Most patients with progressive thoracolumbar kyphosis require combined anterior and posterior fusion. Instrumentation that uses hooks or wires that go into the spinal canal is not recommended in these patients

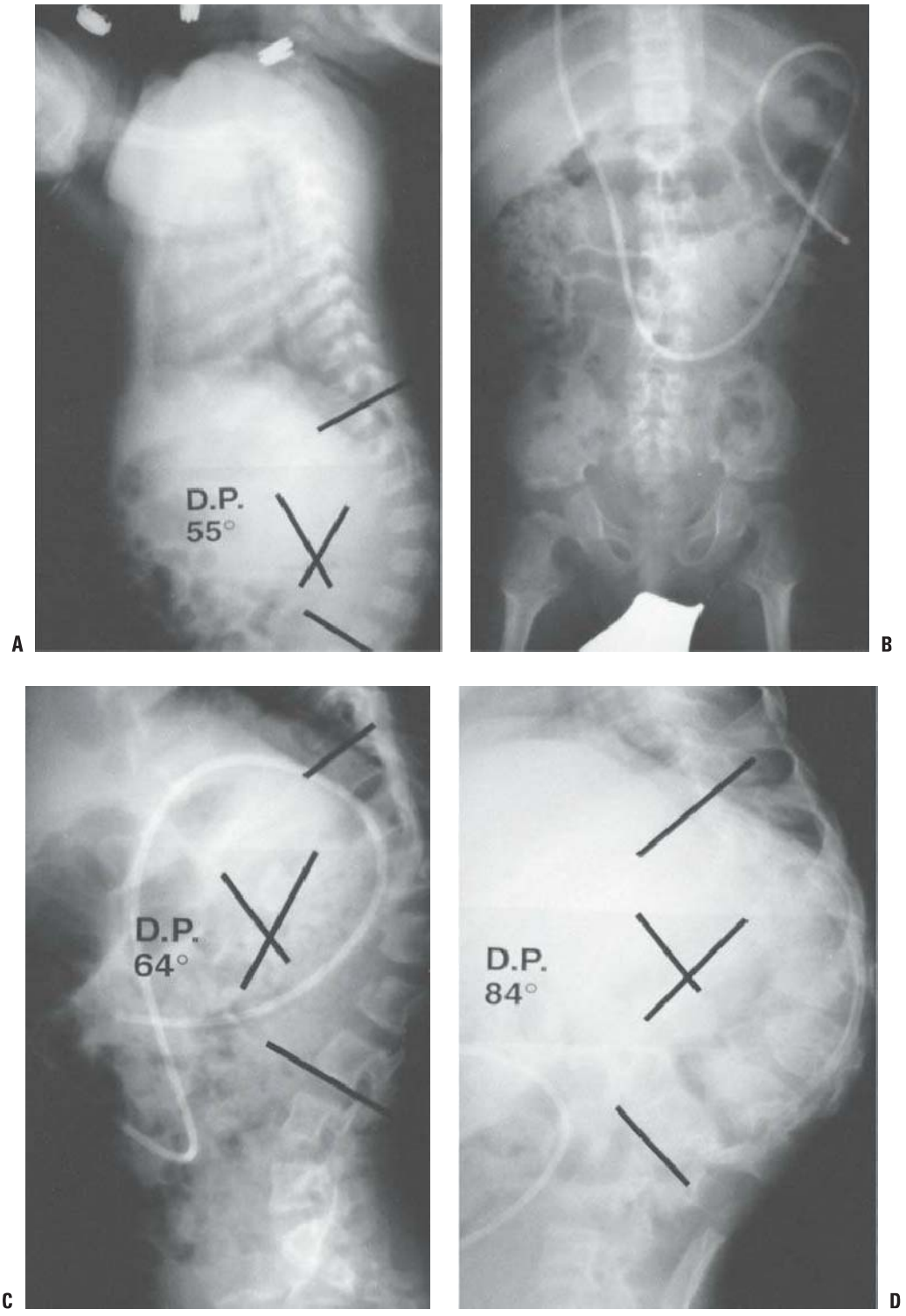


FIGURE 19-36. Achondroplastic dwarf with progressive thoracolumbar kyphosis. **A**: Lateral radiograph at 1 year of age shows a 55-degree thoracolumbar kyphosis. **B**: Anteroposterior radiograph at 5 years of age shows narrowing of the lumbar interpedicular distance characteristic of achondroplasia. **C**: Lateral radiograph at 5 years of age reveals a 64-degree kyphosis. **D**: Lateral radiograph at 9 years of age shows an 84-degree thoracolumbar kyphotic deformity.

because the small size of the spinal canal and the lack of epidural fat make instrumentation hazardous. If pedicle screws can be placed safely, this will allow for posterior instrumentation. This has the advantage of not entering an already stenotic spinal canal and giving secure fixation to aid in spinal fusion. Ain and Shirley reported (253) good results with anterior fusion and instrumentation combined with posterior fusion.

Postlaminectomy kyphosis has been reported to occur after decompression for spinal stenosis of the skeletally immature achondroplastic spine (254, 255). Ain et al. reported that

all 10 of their skeletally immature patients who had laminectomies and preservation of more than 50% of the facet joints developed progressive kyphotic deformities even though more than 50% of the facet joints were preserved. They recommended the addition of posterior instrumentation with pedicle screws and fusion when decompressive laminectomies are done in skeletally immature achondroplastic patients (255).

Pseudoachondroplasia. Kyphotic deformities also can occur in children with pseudoachondroplasia and are caused by wedging of multiple vertebral bodies in the thoracolumbar and thoracic spine. The kyphotic deformity in patients with pseudoachondroplasia differs from that in patients with achondroplasia. In patients with pseudoachondroplasia, the kyphosis involves multiple levels and is less acutely angular than the deformity in patients with achondroplasia, which involves only one or two levels. Bracing may prevent progression of this deformity, but surgery is indicated if progression occurs despite bracing. Spinal fusion with instrumentation can be performed safely in patients with pseudoachondroplasia because there is no associated stenosis of the spinal canal as in patients with achondroplasia (256, 257).

Spondyloepiphyseal Dysplasia Congenita. Thoracolumbar kyphotic deformities occur in approximately half of the patients with congenital spondyloepiphyseal dysplasia; these deformities usually respond to a modified TLSO (244). If surgery is needed for a progressive kyphosis, anterior and posterior fusions are recommended (257).

Diastrophic Dwarfism. Spinal deformity is a common finding in diastrophic dysplasia (258). These spinal deformities consist of cervical kyphosis, thoracic kyphoscoliosis, and lumbar hyperlordosis. Midcervical kyphosis occurs in 15% to 33% of patients with diastrophic dwarfism (244, 259, 260); however, Remes et al. (259, 260) and Herring (261) reported spontaneous improvement Progressive cervical kyphosis (more than 60 degrees) can be stabilized with a spinal fusion (228, 238, 239). If a posterior fusion is to be done, the increased incidence of cervical spina bifida in diastrophic dwarfism must be considered during dissection (259, 260, 262).

Mucopolysaccharidosis. Mucopolysaccharidoses are inherited lysosomal storage disorders caused by deficiency of the enzymes that are necessary for the degradation of glycosaminoglycans. There are at least 13 types of mucopolysaccharidoses. The more common names of this condition are Hurler, Hunter, Sanfilippo, Morquio, and Maroteux-Lamy syndromes. Bone marrow transplantation has increased the life expectancy of these patients. Before this treatment method became available, most children did not survive long enough to require intervention for spinal deformities. With increased survival, progressive kyphotic deformities of the spine with neurologic compromise have been reported (Fig. 19-37) (263–265). Despite bone marrow transplantation, the deposition of metabolites in bone is not reversed to the same extent as that in soft tissue (266).

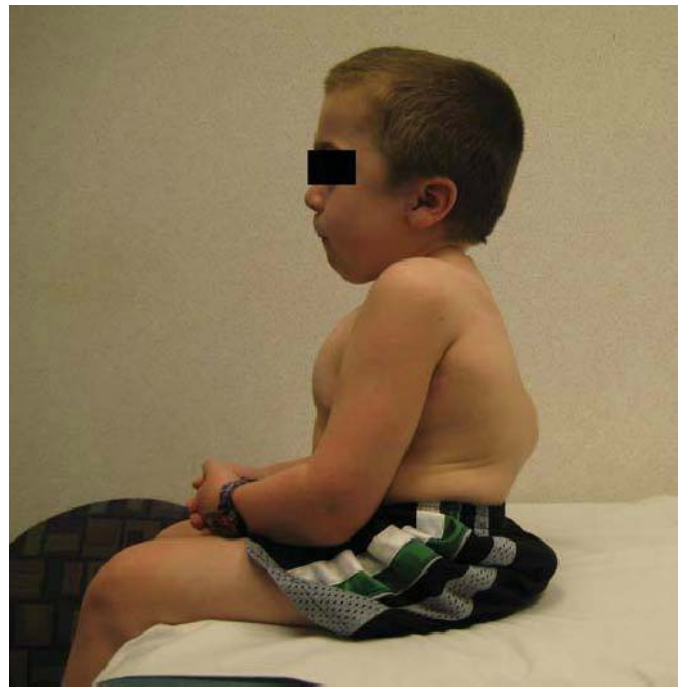


FIGURE 19-37. Progressive kyphotic deformity in a child with mucopolysaccharidosis.

Children with mucopolysaccharidosis develop thoracolumbar kyphosis, with anterior beaking and flattening of the vertebral bodies at the level of the kyphotic deformity. Swischuk (267) suggested a mechanical cause for the anterior beaking. He postulated that hypotonia results in thoracolumbar kyphosis, resulting in herniation of the nucleus pulposus into the anterior vertebral body, which causes the anterior beaking of the vertebral body. Field et al. (266), however, examined two specimens at postmortem and found that the end-plate formation was normal, but there was a failure of ossification in the anterosuperior part of the vertebral body.

Bracing can be used to prevent progression of the kyphotic deformity, but the effectiveness of this form of treatment has not been documented. Spinal fusion is recommended for progressive kyphosis in patients with mucopolysaccharidosis. Tandon et al. (264) reported good results with posterior spinal fusion, and Dalvie et al. (263) reported good results with anterior fusion and instrumentation. Further studies are needed to determine which approach is best, but the goal of surgery is to obtain a stable fusion of the involved area to prevent any further progression of the kyphosis (244, 257, 268).

Gaucher Disease. Gaucher disease is an uncommon hereditary glycolipid storage disorder characterized by the accumulation of glucocerebroside in the lysosomes of macrophages of the reticuloendothelial system. Splenomegaly with associated pancytopenia is the most common clinical manifestation. The skeletal manifestations are caused by infiltration of the bone marrow by Gaucher cells and include bone crisis, pathologic fracture, osteopenia, osteonecrosis, and osteomyelitis. Progressive kyphosis of the spine has been reported in these patients (269, 270). The proposed etiology of the kyphosis

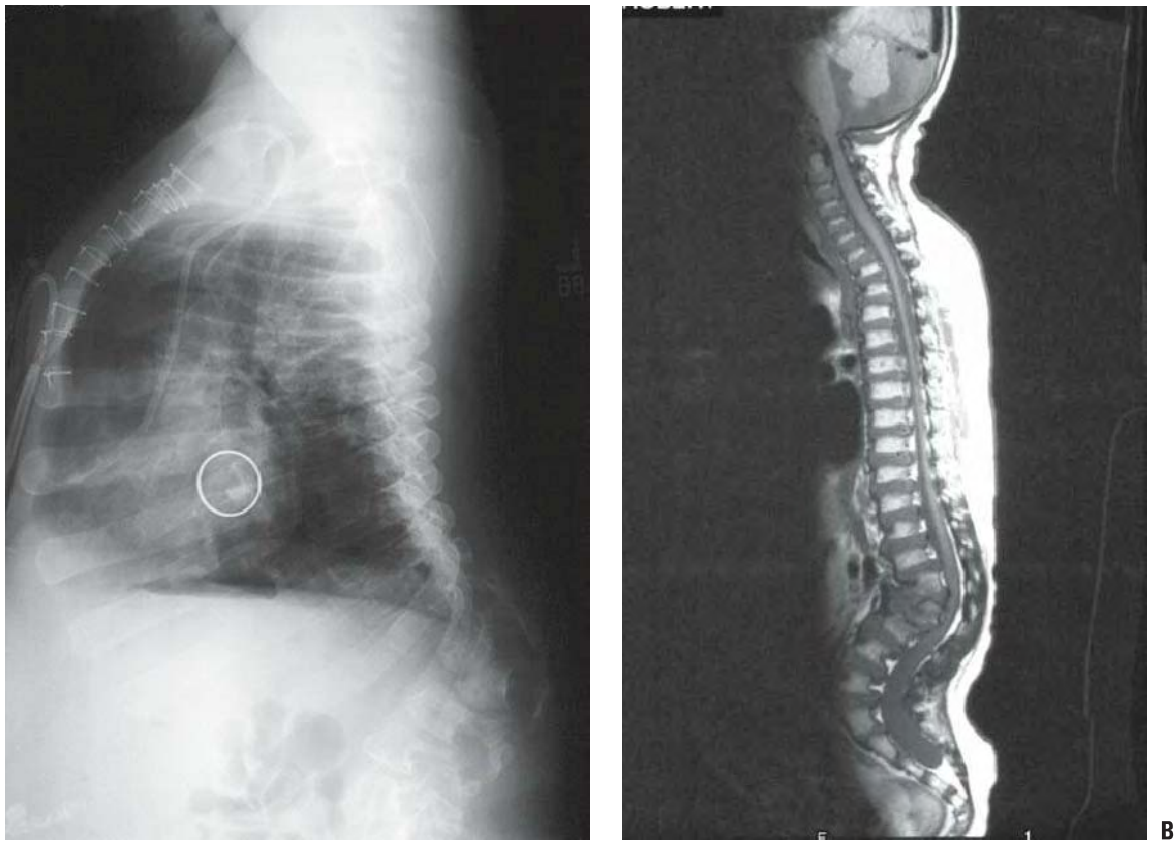


FIGURE 19-38. Lateral radiograph **(A)** and MRI **(B)** demonstrate progressive thoracolumbar kyphosis in a patient with Gaucher disease.

is infiltration of the bone marrow by Gaucher cells, resulting in bone crisis, osteopenia, and osteonecrosis that lead to vertebral-body collapse, often on multiple levels. Kyphosis can be progressive because of continued vertebral-body collapse or growth abnormalities secondary to vertebral-body collapse. If a progressive kyphosis develops, surgical intervention is recommended. If the spine is still flexible, posterior fusion and instrumentation are adequate, but if the deformity is rigid, anterior and posterior fusion and instrumentation are needed (Fig. 19-38) (269, 270).

Marfan Syndrome. Marfan syndrome is a generalized disorder of connective tissue that affects the supporting structures of the body, especially those in the musculoskeletal system. This syndrome is caused by mutations in coding of the genes for the glycoprotein fibrillin (271, 272). Spinal deformity is the most common skeletal abnormality in Marfan syndrome, and scoliosis is the most common of these spinal deformities (245, 273–278). Thoracic lordosis has been traditionally reported as the most common sagittal plane deformity (279, 280). In some patients, the thoracic lordosis becomes severe enough to compromise respiration. With the lordotic posture of the thoracic spine, an associated kyphosis or a relative kyphosis may develop in the lumbar spine. A third common spinal deformity associated with Marfan syndrome is thoracolumbar kyphosis, which affects approximately 10% of patients (Fig. 19-39). These spinal deformities usually occur during the juvenile growth period, before the adolescent growth spurt (280). Sponseller et al. (281) found that 41% of their patients with Marfan syndrome had a kyphotic deformity of more than 50 degrees, with a tendency toward longer kyphoses extending through the thoracolumbar junction.

Brace treatment has been recommended to try to halt the progression of spinal deformity but has been found to be ineffective (282, 283). Correction of kyphotic deformities requires anterior and posterior spinal fusion with segmental instrumentation (282). Thoracic lordosis is corrected by posterior segmental instrumentation to correct the lordotic deformity, followed by posterior fusion (284). Because dural ectasia erodes pedicles, a CT scan of the pedicles should be obtained to plan fixation. In addition, fusion should be extended to one level above and below the end vertebrae and include all curves. Complications are more frequent after surgical correction of spinal deformity in patients with Marfan syndrome than after spinal surgery in other patients and include infection (10%), dural tears (8%), instrumentation failure (21%), and pseudarthrosis (10%) (282).

Cervical spine abnormalities also are common in patients with Marfan syndrome, but clinical problems from these abnormalities are rare. Basilar impression and focal cervical kyphosis are the most frequently reported cervical spine abnormalities. Focal cervical kyphosis usually is associated with a lordotic thoracic spine (285).

Because of the increased incidence of cervical spine abnormalities, Hobbs et al. (285) recommended that patients with

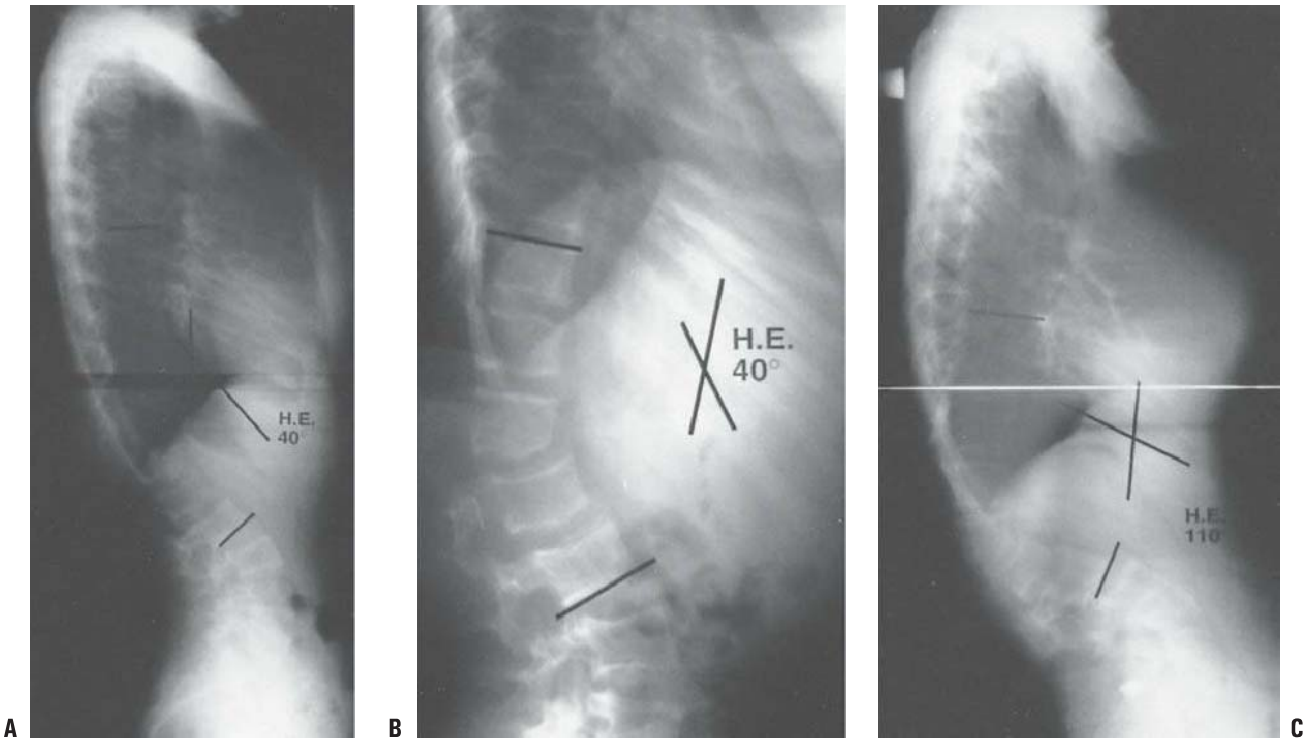


FIGURE 19-39. **A**,**B**: Lateral radiographs of a 17-year-old child with Marfan syndrome and a 40-degree progressive thoracolumbar kyphosis. **C**: Lateral radiograph of the same patient 3 years later shows that the thoracolumbar kyphosis has progressed to 110 degrees.

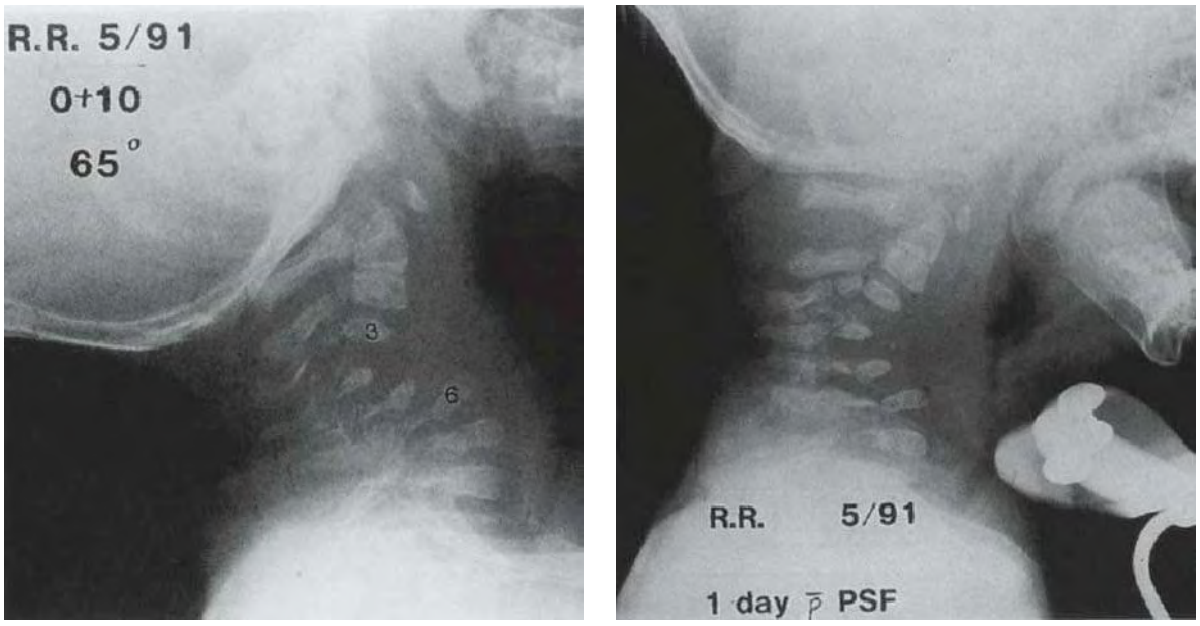
Marfan syndrome avoid sports that involve risks of high-impact loading of the cervical spine.

Larsen Syndrome. In 1950, Larsen et al. (286) described a congenital malformation syndrome (287) consisting of facial dysmorphism and hyperelasticity of the joints, with congenital dislocation of the knees and frequent dislocation of the hips and elbows (129, 288–291). Equinovarus or valgus foot deformities and ancillary calcaneal nuclei also are characteristic features of this syndrome. Abnormalities of the cervical spine, specifically cervical kyphosis, were not emphasized in the original description, and often this life-threatening finding is overlooked (198, 288, 292). Johnston et al. (287) reported cervical kyphosis and vertebral-body anomalies in five of nine patients with Larsen syndrome. The apex of the kyphosis usually occurs at the fourth or fifth cervical vertebra, with marked hypoplasia of one or two of the vertebral bodies (Fig. 19-40). Cervical kyphosis is present in infants with Larsen syndrome. Developmental delay may be attributed to hypotonia and dislocation of the knees or hips, but the underlying cause for developmental delay may be a chronic myelopathy from the cervical kyphosis. Cervical kyphosis and vertebral hypoplasia are easily demonstrated on lateral C-spine radiographs. Flexion and extension views usually are not needed and may be difficult to obtain safely in an infant. MRI scans will demonstrate spinal cord compression or compromise.

The recommended treatment recommendation for cervical kyphosis in Larsen syndrome is early posterior arthrodesis to stabilize the spine. An *in situ* posterior arthrodesis with autogenous iliac crest bone graft, followed by immobilization in either a halo or Minerva cast or custom orthosis, is recommended. Reduction of the kyphosis is obtained only in the postoperative halo or Minerva cast or orthosis stage of the treatment. Johnston et al. (287) found that, over a period of time following a solid posterior arthrodesis, a gradual correction of the kyphosis occurred because of continued anterior vertebral-body growth. Because the posterior arthrodesis is done at a young age, the patient must be followed for potential complications from continued anterior growth, which would result in lordosis. Johnston and Schoenecker (293) described a patient who developed neurologic symptoms from this growth-related lordosis.

Posttraumatic Deformities. Kyphosis can occur as a direct result of trauma to the spinal column or the spinal cord. Deformity can occur at a fracture site from a malunion, chronic instability leading to progressive deformity, paralysis after spinal cord injury, or from anterior growth arrest (294–298). Kyphosis at the fracture site is acute and spans a short segment of vertebrae. Paralytic kyphosis is a long, C-shaped deformity that spans many vertebral segments. Progressive kyphosis also may occur after development of a posttraumatic syrinx (299).

Kyphosis at a fracture site requires surgical intervention for correction. Anterior, posterior, and combined anterior and posterior procedures have been described for correction of posttraumatic kyphosis (300–304). Brace treatment has been ineffective for progressive paralytic kyphosis (294), and surgery is indicated for paralytic kyphosis of more than 60 degrees. If the kyphosis is flexible and can be reduced to <50 degrees, posterior fusion with segmental instrumentation can be done. If



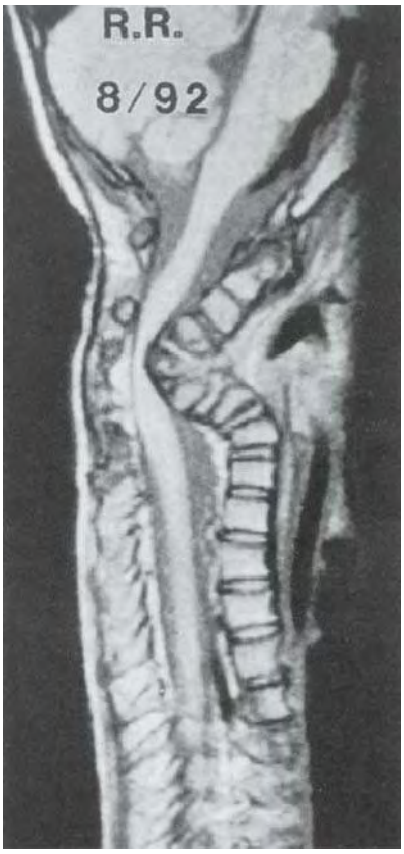


FIGURE 19-40. Larsen syndrome. **A**: Lateral radiograph of a 10-month-old patient showing kyphosis of 65 degrees, with correction to only 48 degrees in extension. **B**: Lateral radiograph immediately after posterior arthrodesis, showing the patient with orthosis and correction of kyphosis to 39 degrees. **C**: T2-weighted magnetic resonance image 15 months postoperatively shows severe impingement on spinal cord. Kyphosis had progressed to 110 degrees, and the patient was quadriplegic after a fall. (From Johnston CE, Birch JG, Daniels JL. Cervical kyphosis in patients who have Larsen syndrome. *J Bone Joint Surg Am* 1996;78:538.)

the kyphosis is rigid and cannot be reduced to <50 degrees on preoperative bending radiographs, anterior release and fusion should be (performed) followed by posterior fusion and segmental instrumentation (294, 298).

Neurofibromatosis. Kyphoscoliosis is common in patients with neurofibromatosis, although kyphosis may be the predominant deformity (305, 306). Funasaki et al. (307) found that 50% of their patients with neurofibromatosis and spinal deformity had an abnormal sagittal curve. The vertebral bodies are frequently deformed and attenuated at the apex of the kyphosis. Dystrophic vertebral-body changes may develop over time (308, 309). Crawford (308) and Durrani et al. (310) described this as modulation of the deformity, from a nondystrophic curve to a dystrophic curve. The kyphosis typically is sharp and angular over a relatively small number of vertebral segments. Severely angular kyphosis can cause neurologic compromise (311, 312). Lonstein et al. (313) found that cord compression due to spinal

curvature from neurofibromatosis was second only to congenital kyphosis as a cause of spinal cord compression. The kyphosis in patients with neurofibromatosis typically involves the thoracic spine or the upper thoracic spine. Involvement of the cervical and cervicothoracic vertebrae also has been reported (311, 314–318).

Kyphotic deformities with dystrophic changes tend to be progressive, and they more commonly lead to neurologic compromise.

Treatment of kyphoscoliosis in patients with neurofibromatosis begins with a thorough physical examination for neurologic abnormalities. MRI scans should be obtained to demonstrate any intraspinal lesions, such as pseudomeningocele, dural ectasia, or neurofibroma, which may cause impingement on the spinal cord (319). Any intraspinal lesions should be treated appropriately before spinal fusion and instrumentation are undertaken. Because posterior fusion alone has resulted in a high rate of pseudarthrosis (65%) (319), combined anterior and posterior spinal fusions combined with posterior instrumentation are recommended (320). Titanium instrumentation is preferred to allow for future MRI studies of the spine. Abundant autogenous bone grafts and prolonged immobilization may be required to obtain a solid fusion in these patients, and repeated bone grafting may be required 6 months after the initial surgery. Vascularized fibular or rib grafts also can be used for anterior fusion and structural support (109, 308, 314, 317, 321, 323).

Tuberculosis. The spine is involved in 50% of patients with skeletal tuberculosis (324). Spinal tuberculosis is the most dangerous form of skeletal tuberculosis because of its ability to cause bone destruction, deformity, and paraplegia. In childhood spinal tuberculosis, the extent and degree of abscess formation are greater than those in adult tuberculosis, but paraplegia is less common in children than in adults with spinal tuberculosis (325). The most frequent site of spinal tuberculosis in children is the thoracolumbar junction and its adjacent segments. Tuberculosis infection usually destroys the anterior elements of the spine and results in a significant angular kyphosis at the infected site. The involved anterior vertebral bodies usually fuse once the infection is adequately treated. In young children, continued growth of the intact posterior element can cause a late increase in kyphosis in an already kyphotic spine (325).

All forms of active spinal tuberculosis are treated with a complete course of chemotherapy. First-line drugs are streptomycin, isoniazid, and rifampin, and second-line drugs are ethambutol and pyrazinamide (326). Medical therapies for spinal tuberculosis will adequately treat the tuberculum infection in most cases (324, 327–330). Bracing or casting has been used along with medical therapy to try to prevent progression of kyphosis during therapy. Rajasekaran found an average increase in deformity of 15 degrees in all patients who were treated nonsurgically (331). The greatest increase in deformity occurred during the first 6 months of treatment.

Indications for surgery in spinal tuberculosis are spinal instability, neurologic involvement, prevention or correction of spinal deformity, drainage of significant abscesses, and diagnostic biopsy (327). Neurologic involvement and present or impending paraplegia are more obvious indications for surgical intervention than the other indications. Rajasekaran described four prognostic signs to predict spinal instability and late increase in deformity. When more than two signs are present, this is a reliable predictor of progressive deformity and spinal instability. These prognostic signs are (a) dislocation of the facets, (b) posterior retropulsion of the diseased fragments, (c) lateral translation of the vertebrae in the anteroposterior view, and (d) toppling of the superior vertebra (Figs. 19-41 and 19-42) (332). Other factors that lead to a significant increase in kyphosis in children who are not treated surgically are involvement of three or more vertebral bodies, initial kyphosis of more than 30 degrees, and age younger than 15 years (333–335).

Several different surgical approaches have been used in the treatment of spinal tuberculosis (336–340). Anterior debridement and strut grafting, with or without a posterior fusion and instrumentation, have the most consistent long-term results (326, 340–352). Good results have been reported with the use of allografts for structural support anteriorly (353–355). Anterior debridement and fusion with anterior instrumentation of the spine also have had positive results in the treatment of spinal tuberculosis (356, 357). Some correction of the kyphosis may be obtained at the time of surgery. Kyphosis also can be a problem in patients with healed spinal tuberculosis (358). The infected area of the anterior spine usually fuses, and continued growth posteriorly causes progressive kyphosis that can result in paraplegia. The presence of neurologic symptoms is an indication for anterior decompression and fusion, which can be followed by posterior fusion and instrumentation.

Juvenile Osteoporosis. Idiopathic juvenile osteoporosis is an acquired systemic condition that consists of generalized osteoporosis in otherwise normal prepubertal children (359). Although idiopathic juvenile osteoporosis is uncommon, associated kyphosis and back pain are common in patients with this condition.

Schippers (360) first described this condition in 1939 and, since that time, other authors have described its clinical findings and natural history (153, 360–367). The etiology of idiopathic juvenile osteoporosis is unknown. Laboratory values of serum calcium, phosphorus, alkaline phosphatase, parathyroid hormone, and osteocalcin are normal. The collagen type and ratios from skin biopsy samples also are normal. There have been some reports of a slight decrease in 1,25-dihydroxyvitamin D (365, 368, 369), but the significance of this finding is not known. Low serum calcitonin levels also have been reported, but treatment with calcitonin has not proven to be beneficial (342, 349). In contrast, Saggese et al. (366) noted normal serum calcitonin levels in their patients. Green (370) suggested that a mild deficiency of 1,25-dihydroxyvitamin D can explain most of the findings in idiopathic juvenile osteoporosis. During rapid growth phases, the deficiency is discovered because growth requirements cannot keep pace, causing a relative osteoporosis. When puberty occurs, the increase in sex hormone overcomes

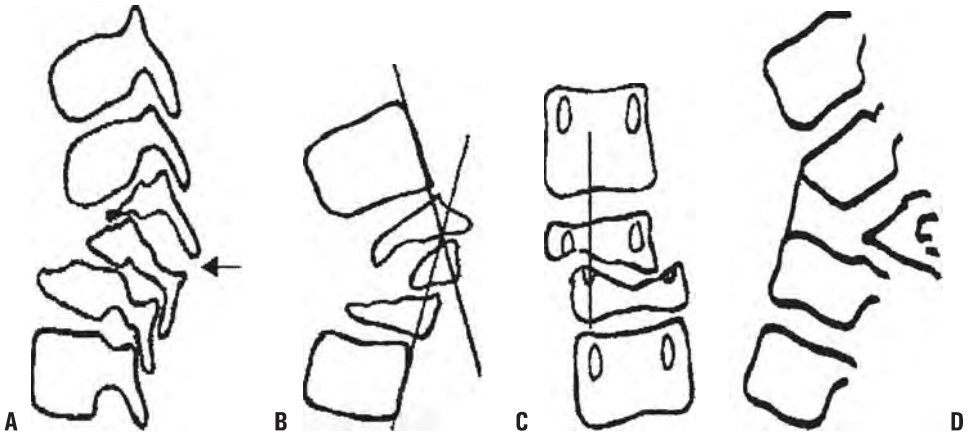


FIGURE 19-41. Radiologic signs for the spine at risk. **A**: *Separation of the facet joint.* The facet joint dislocates at the level of the apex of the curve, causing instability and loss of alignment. In severe cases, the separation can occur at two levels. **B**: *Posterior retropulsion.* This is identified by drawing two lines along the posterior surface of the first normal vertebrae above and below the curve. The diseased segments are found to be posterior to the intersection of the lines. **C**: *Lateral translation.* This is confirmed when a vertical line drawn through the middle of the pedicle of the first lower normal vertebra does not touch the pedicle of the first upper normal vertebra. **D**: *Toppling sign.* In the initial stages of collapse, a line drawn along the anterior surface of the first lower normal vertebra intersects the inferior surface of the first upper normal vertebra. "Tilt" or "toppling" occurs when the line intersects higher than the middle of the anterior surface of the first normal upper vertebra. (Reproduced with permission and copyright of the British Editorial Society of Bone and Joint Surgery. Rajasekaran S. The natural history of post-tubercular kyphosis in children. *J Bone Joint Surg* 1901;83B:954.)

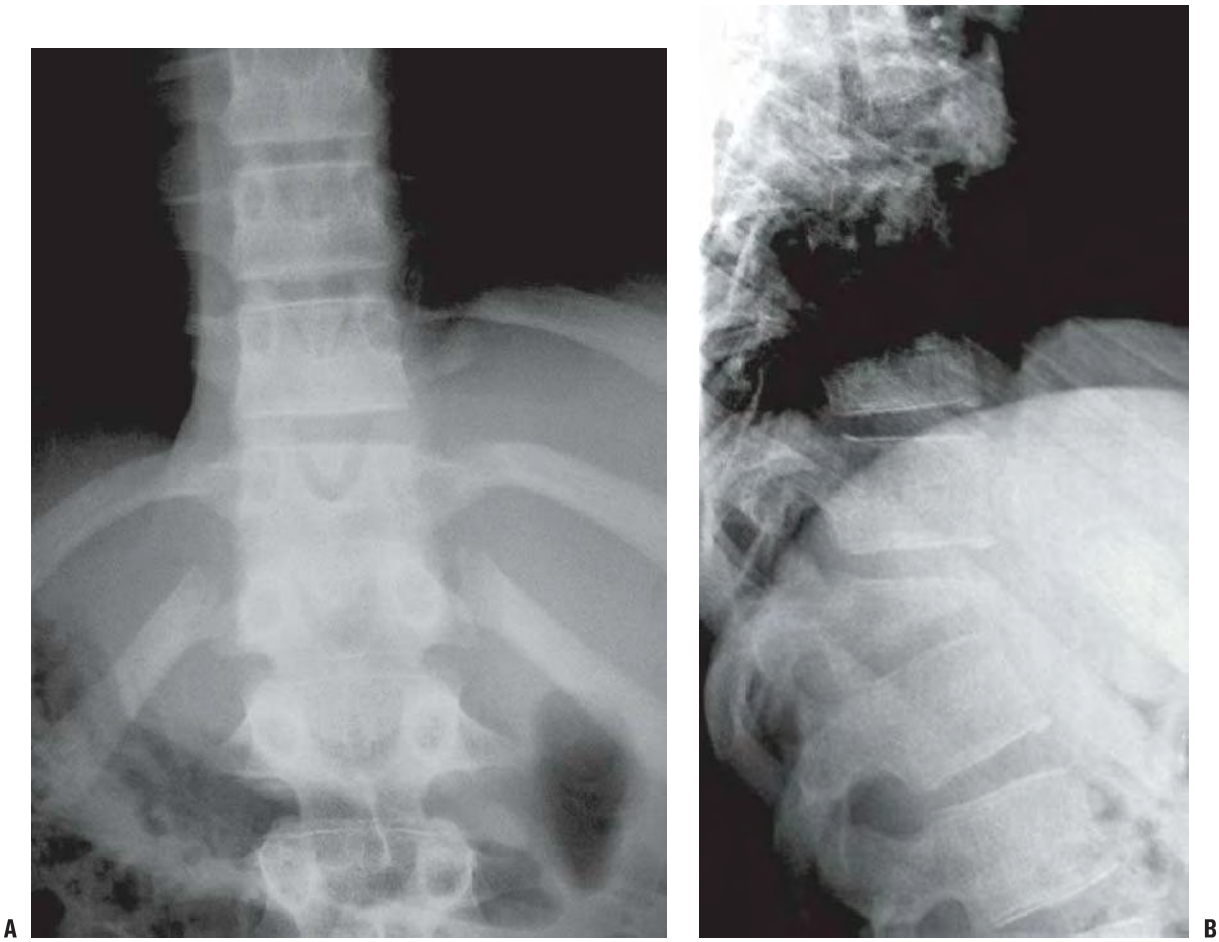
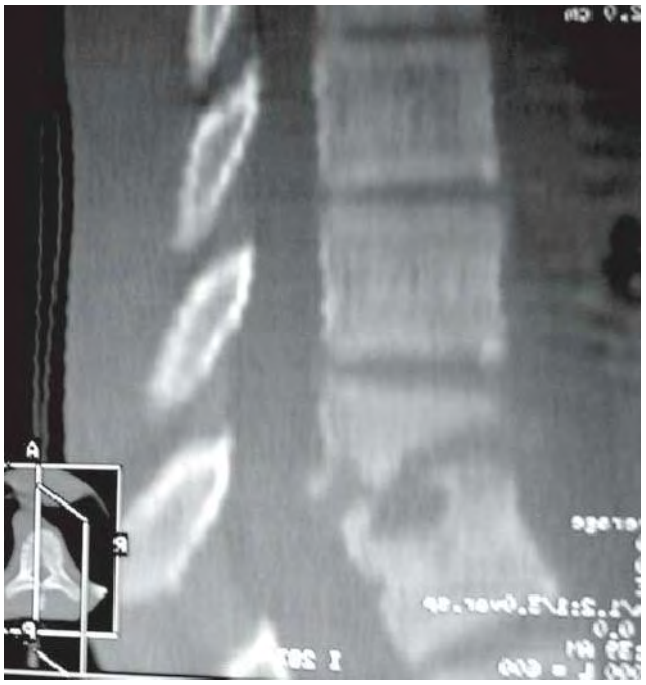


FIGURE 19-42. Anteroposterior and lateral radiographs **(A,B)** and a CT scan



C

FIGURE 19-42. (*continued*) **(C)** demonstrating vertebral body collapse secondary to tuberculosis.

the deficit in 1,25-dihydroxyvitamin D, and the relative osteoporosis improves. This theory has yet to be proved.

Clinically, these patients complain of insidious onset of back pain (371), lower extremity pain or fractures, and difficulty in walking (64, 271, 372, 373). Difficulty in walking may sometimes be the only finding. This condition occurs during the prepubertal period and is slightly more common in boys than in girls. Vertebral collapse or wedging, with resulting kyphosis, is common. Brenton and Dent (374) classified idiopathic juvenile osteoporosis as mild, moderate, and severe. Patients with the mild type have only back pain and vertebral fractures; those with the moderate type have back and lower extremity pain and fractures, with some limitation of activities but eventual return to normal function; and those with the severe form have back and lower extremity pain and fractures. Both metaphyseal and diaphyseal fractures can occur in the lower extremities. Patients with severe disease improve clinically but do not return to normal activity after puberty.

Plain radiographs show wedging or collapse of the vertebral bodies. A "codfish" appearance of the vertebral bodies can occur, with the superior and inferior borders of the vertebrae becoming biconcave (Fig. 19-43). Other studies that can be useful for following the progress of this disease are single-photon absorptiometry, dual-photon absorptiometry, and quantitative CT scanning (363, 365, 366). The problem with these tests is that normal ranges for adolescents and children are variable and have not been standardized.

Idiopathic juvenile osteoporosis is a diagnosis of exclusion. Other diseases that must be considered include metabolic bone diseases, leukemia, Cushing syndrome, lysinuric protein intolerance, type I homocystinuria, and osteogenesis imperfecta. The natural history of this condition is spontaneous improvement or remission at the onset of puberty. Associated kyphosis tends to improve after the onset of puberty.

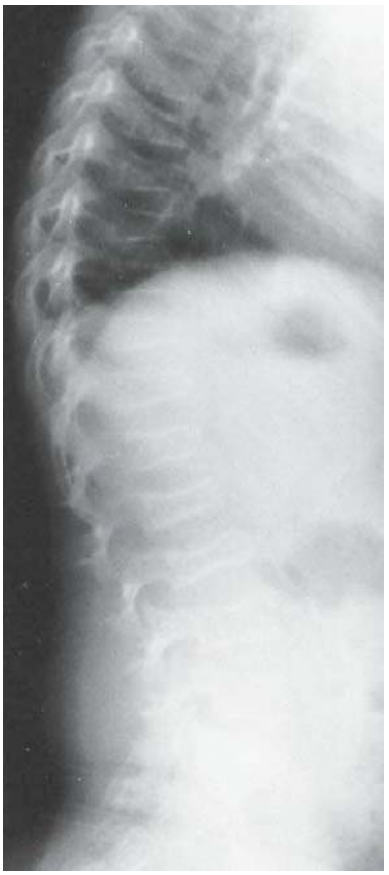


FIGURE 19-43. Lateral radiograph taken of a standing 10-year-old girl with idiopathic juvenile osteoporosis shows diffuse osteopenia, *multiple* "codfish" vertebrae in the thoracic and lumbar spine, and "coin" vertebrae in the upper thoracic spine secondary to extreme collapse. (From Green WB. *Idiopathic juvenile osteoporosis*. New York, NY: Raven, 1994.)

Treatment of idiopathic juvenile osteoporosis involves modification of activities, possible calcium and vitamin D supplementation, and supportive treatment of spinal deformities. It must be ensured that there is sufficient restriction of activities to prevent fractures, but not so much restriction as to cause an increase in osteoporosis. If a significant progressive kyphosis develops, the Milwaukee brace is the treatment of choice (364). The brace is to be worn until there is evidence of improvement of the osteoporosis. Operative therapy for this condition has been associated with a high complication rate because the poor bone quality makes instrumentation and fusion difficult (375).

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