CHAPTER 11

Scoliosis

Definition

The term *scoliosis* (first used by Galen, A.D. 131–201) is derived from the Greek word meaning crooked.[301] One of the most common deformities of the spine, scoliosis has been recognized since ancient times, with descriptions of normal and abnormal spinal curves found in the *Corpus Hippocraticum.* In 1741, André devised the crooked spine as his symbol for orthopaedics.[11]

Currently, scoliosis is defined as a lateral deviation of the normal vertical line of the spine which, when measured on a radiograph, is greater than 10 degrees (Fig. 11–1). Because the lateral curvature of the spine is associated with rotation of the vertebrae within the curve, a three-dimensional deformity occurs. This complex deformity represents abnormal movement in three planes: (1) intervertebral extension in the sagittal plane, leading to lordosis of the scoliotic segment, (2) lateral intervertebral tilting in the frontal plane, and (3) a rotatory component in the axial plane.[349] This results in torsion of the spine, with the most significant abnormality located at the apical region. As the deformity worsens, structural changes develop in the vertebrae and rib cage. Relationships between intrathoracic and abdominal organs may become distorted as the deformity becomes severe, but rarely are the organs' functions compromised.

Classification of Scoliotic Curves

A variety of terms are used to describe different types of scoliotic curves. Table 11–1 provides definitions for the most common classifications. In general, scoliosis is a deformity that may have its genesis in fetal life, infancy, childhood, or adolescence, although the residual deformity persists throughout adult life.

Idiopathic Scoliosis

Idiopathic scoliosis, in which the cause of the deformity has not been established, is the most common type of scoliosis, accounting for nearly 80 percent of patients with structural scoliosis. The diagnosis of idiopathic scoliosis, however, can only be made after a thorough physical examination has ruled out neurologic causes or other syndromes (e.g., neurofibromatosis) and radiographic analysis has excluded congenital anomalies. Idiopathic scoliosis may have its onset at any age during growth, but usually it has three fairly well-defined peak periods: (1) in the first year of life, (2) at age 5 to 6 years, and (3) after age 11 years to the end of skeletal growth.

Thus, idiopathic scoliosis is frequently divided into three groups based on the age at onset. *Infantile idiopathic scoliosis* refers to a scoliosis recognized in patients less than 3 years old. In patients between ages 3 and 10 years, the condition is referred to as *juvenile idiopathic scoliosis.* The term *adolescent idiopathic scoliosis* is used when the deformity is recognized between age 10 years and skeletal maturity, although typically it is noted before the onset of puberty. Adolescent idiopathic scoliosis is the most common of the three groups. Scoliosis recognized after skeletal maturity is defined as *adult scoliosis.*

Adolescent Idiopathic Scoliosis

PREVALENCE

In general, two types of prevalence studies have been used to determine the percentage of people afflicted with scoliosis. The first type, which is based on reviews of chest radiographs made for tuberculosis, has several limitations: (1) there was minimal, if any, imaging of the lumbar spine, (2) many of the radiographs were underpenetrated, and (3) the film size was small.[402,429] The second type of study has been based on school screening programs. Numerous studies of this kind have been reported, and they have provided a more accurate picture of the prevalence of scoliosis.* The prevalence of radiographic curves measuring at least 10 degrees ranges between 1.5 and 3.0 percent, that of curves exceeding 20 degrees between 0.3 and 0.5 percent, and that of curves exceeding 30 degrees between 0.2 and 0.3 percent.

*See references 18, 57, 93, 102, 151, 258, 304, 306, 362, 385, 419, 469, 500.

There is a definite relationship between idiopathic scoliosis and sex, particularly as the curve magnitude increases. The ratio of affected females to males is 1:1 for curves between 6 and 10 degrees, 1.4:1 for curves between 11 and 20 degrees, 5.4:1 for curves exceeding 21 degrees but not needing treatment, and 7.2:1 for curves requiring orthopaedic intervention.[385] This gender prevalence for idiopathic scoliosis—that is, an equal prevalence between the sexes for small curves (less than 10 degrees), with increasing female prevalence for the larger and progressive curves—has been reported by other authors.[18,93,258,385] The clinical significance of these observations is that curve progression is more common in girls.

NATURAL HISTORY

Understanding the natural history of scoliosis is essential to determining when treatment is necessary and, if it is utilized, whether or not the treatment is effective. Unfortunately, there are few current natural history studies that examine curve progression in the untreated, skeletally immature scoliosis population.[59,259,316] Today, when a child is evaluated by the orthopaedist for scoliosis and found to have a moderate-sized spinal curvature, orthotic management usually is started in an effort to prevent curve progression.

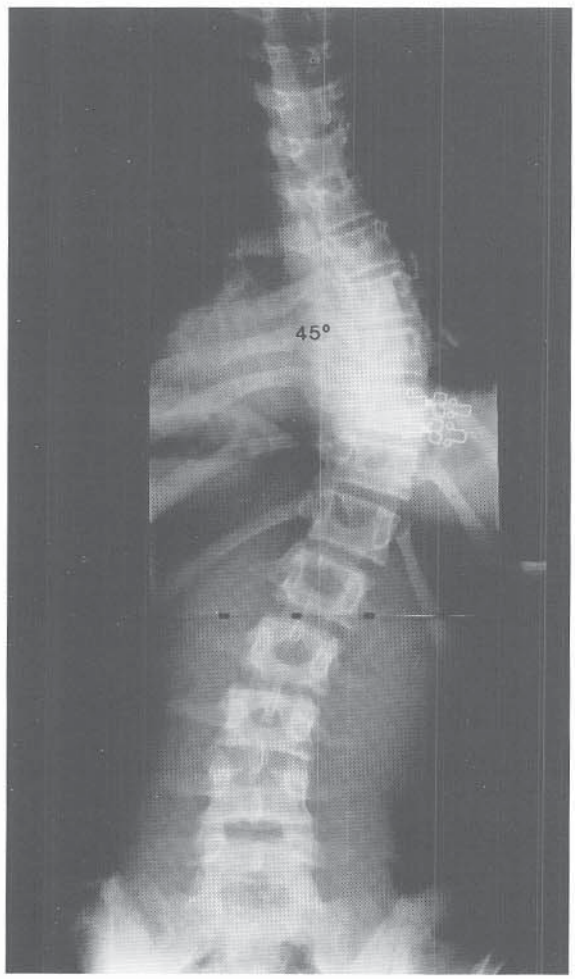


FIGURE 11–1 PA radiograph of the thoracolumbar spine of a 13-year-old girl showing a 45-degree right thoracic scoliosis.

TABLE 11–1 **Glossary of Terms Describing Different Types of Scoliotic Curves**

Structural curve: Represents a segment of the spine that has a fixed lateral curvature.
Nonstructural (functional) curve: A curvature that does not have a fixed deformity and may be compensatory in nature. The curve may be a result of leg length discrepancy (and so disappears when the patient is supine), poor posture, muscle spasm, or other cause.
Primary curve: The first or earliest curve present.
Compensatory curve: A secondary curve located above or below the structural component that develops in order to maintain normal body alignment.
Lordoscoliosis: Structural scoliosis associated with increased swayback or loss of normal kyphosis within the measured curve; nearly always present in idiopathic scoliosis.
Kyphoscoliosis: Noted as an increased round-back on the lateral radiograph, the condition may represent a true kyphotic deformity (as seen in some pathologic conditions) or it may represent such excessive rotation of the spine that the lateral radiograph is actually reflecting the scoliotic deformity. (In idiopathic scoliosis, true kyphotic deformity does not occur.)
Cervicothoracic curve: Any spinal curvature in which the apex is at C7 or T1.
Thoracic curve: A spinal curvature in which the apex is between T2 and T11.
Thoracolumbar curve: A spinal curvature in which the apex is at T12, L1, or the T12–L1 interspace.
Lumbar curve: A spinal curvature in which the apex is between L1 and L4.
Lumbosacral curve: A spinal curvature in which the apex is at L5 or below.
Double curve: Scoliosis in which there are two lateral curves in the same spine.
Double major curve: Scoliosis in which there are two structural curves that usually are of similar size and rotation.
Double thoracic curve: Scoliosis with a structural upper thoracic curve, a larger, more deforming lower thoracic curve, and a relatively nonstructural lumbar curve.
Idiopathic scoliosis: A structural curve for which the cause has not been definitely established.
Congenital scoliosis: Scoliosis due to bony abnormalities of the spine that are present at birth. These anomalies are classified as failure of vertebral formation and/or failure of segmentation.
Neuromuscular scoliosis: Scoliosis due to a neurologic disorder of the CNS or muscle.
Hysterical scoliosis: A nonstructural deformity of the spine that develops as a manifestation of a psychological disorder.
Adult scoliosis: Spinal curvature present after skeletal maturity. It may be due to any cause.

There is no consensus in the literature regarding the definition of curve progression. Measurable increases in curve sizes of 5, 6, and 10 degrees have all been reported as representative of progression.* Most current studies use increases exceeding 5 or 6 degrees as indicating definite progression. As more is learned about adolescent idiopathic scoliosis, information from older retrospective studies regarding the likelihood of curve progression becomes less useful because these studies tended to focus on large curves and included patients with scoliosis of different etiologies.

Natural History Before Skeletal Maturity. Most patients with mild idiopathic scoliosis will not experience problems during their lives because of their deformity. Reports in the literature indicate that individuals with untreated curves less than 20 degrees are at low risk for progression, particularly as

*See references 6, 59, 120, 158, 209, 255, 259, 261, 316, 322, 361, 387.

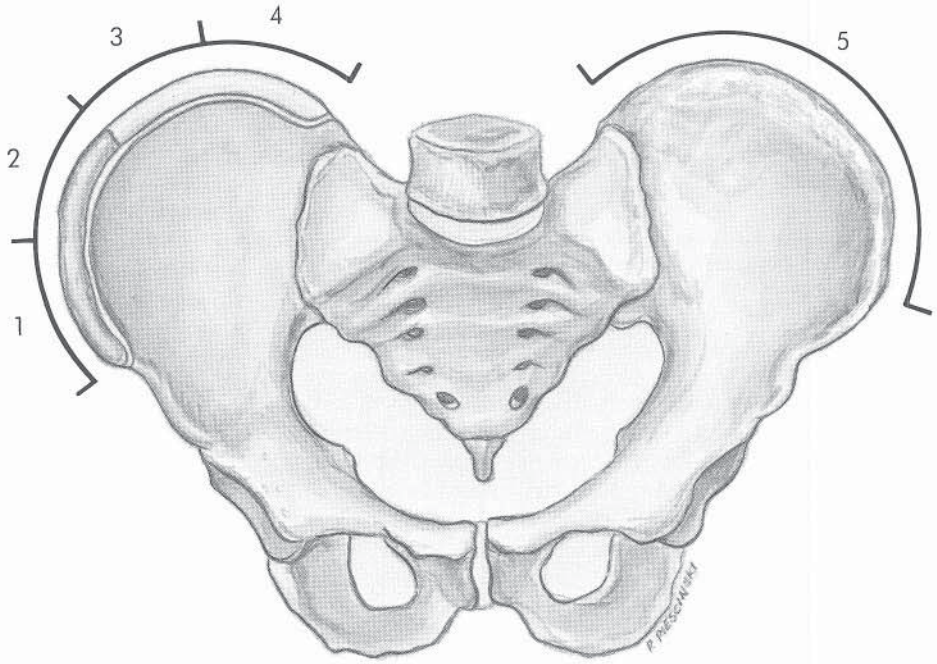


FIGURE 11–2 The Risser sign proceeds from grade 0 (no ossification) to grade 4 (all four quadrants show ossification of the iliac apophysis). When the ossified apophysis has fused completely to the ilium (Risser grade 5), the patient is skeletally mature.

they approach skeletal maturity.[259] Some patients, however, have curves that progress over the years and may ultimately lead to health problems. Thus, it is important to recognize factors associated with curve progression. Several factors that may help predict progression are patient sex, remaining growth, curve magnitude, and curve pattern.[59,259,316] Factors of no predictive value for curve progression before skeletal maturity include a family history of scoliosis, patient height-weight ratios, lumbosacral transitional anomalies, thoracic kyphosis, lumbar lordosis, and spinal balance.[59]

SEX. Females clearly compose the majority of patients whose curves progress and ultimately require treatment.[18,93,258,385] Although the exact reason for this phenomenon is not well known, hormonal influences have been proposed.[2,164,416]

REMAINING GROWTH. The young patient's remaining growth usually is assessed by two maturity indices: (1) the Risser sign (a skeletal marker) and (2) in females, menarchal status (a physiologic marker). A third index, peak height velocity, is currently gaining greater recognition as a means of assessing remaining skeletal growth.

The Risser sign is a radiographic measurement based on the ossification of the iliac apophysis.[380] Interpreted from a routine scoliosis radiograph (posterior-to-anterior projection of the spine and pelvis on a 36-inch cassette), the ossification begins on the lateral aspect of the iliac apophysis and progresses medially (Fig. 11–2). Divided into four quadrants, the Risser sign proceeds from 0 (no ossification) to Risser 4, in which all four quadrants show ossification ("capping") of the apophysis. When the ossified apophysis has fused completely to the ilium (Risser 5), the patient is fully skeletally mature. Patients with Risser sign 0 or 1 (and, to a lesser extent, Risser sign 2) are at greatest risk for curve progression because a significant amount of spinal growth remains.

Menarchal status is a clinical measurement applicable only to females. The premenarchal girl is still in the active growth period. After menarche, the girl enters the deceleration phase of growth, and the likelihood of curve progression lessens. The Tanner index of maturity,[436] which is based on an assessment of breast and genital development, is another clinical index that has been used to determine a child's remaining growth and thus indirectly the risk of curve progression.

Peak height velocity (PHV) is a measurement of the maximum skeletal growth that occurs during the adolescent growth spurt (Fig. 11–3). The value for PHV, calculated from changes in a patient's height measurements over time, is fairly consistent throughout the published literature and is reported to be about 8.0 cm per year for girls and 9.5 cm per year for boys.[127,252,436] The reported average age of PHV in North American girls is approximately 11.5 years. Triradiate cartilage closure, a radiographic index of maturity, occurs after PHV and before Risser 1 and menarche. For PHV to be clinically useful, serial height measurements must be obtained. Six-month intervals are generally used. Shorter

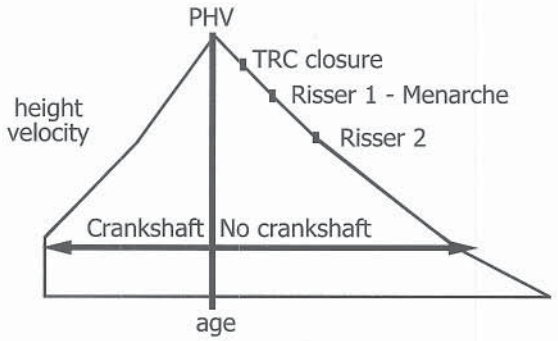


FIGURE 11–3 A schematic drawing of height velocity. Triradiate cartilage (TRC) closure occurs after the period of peak height velocity and before Risser 1 and menarche are attained. (Modified from Sanders JO, Little DG, Richards BS: Prediction of the crankshaft phenomenon by peak height velocity. Spine 1997;22[12]:1352–1356.)

TABLE 11–2 **Incidence of Curve Progression Based on Curve Magnitude and Risser Sign**

Risser Sign	Percentage of Curves That Progressed	
	Curves 5–19 Degrees	*Curves 20–29 Degrees*
Grade 0 and 1	22%	68%
Grade 2, 3, or 4	1.6%	23%

Modified from Lonstein JE, Carlson JM: The prediction of curve progression in untreated idiopathic scoliosis during growth. J Bone Joint Surg 1984;66-A:1061.

time intervals may result in significant measurement error. If height data are not available from the patient's records, the information can often be obtained from the family, school, or pediatrician. Although PHV requires analysis of serial height measurements collected over time, it is the earliest and best index available to demonstrate that growth is slowing and the risk of curve progression is diminishing.

CURVE MAGNITUDE. The size of the existing curve, when the scoliosis is recognized, also is helpful in predicting curve progression. The combination of this factor with assessment of remaining growth is quite helpful in predicting the natural history of scoliosis in young patients. Immature patients (premenarchal, Risser 0) with curves exceeding 20 degrees are at substantial risk for progression of spinal deformity (Table 11–2).* For immature patients with curves exceeding 25 to 30 degrees, the risk of curve progression is believed to be significant enough that orthotic management at the time of initial evaluation has been recommended.[261,316]

CURVE PATTERN. The curve pattern is useful in predicting curve progression. Double curves and thoracic curves are most likely to progress, followed by thoracolumbar curves. Lumbar scoliosis is least likely to worsen.[59,259]

Natural History After Skeletal Maturity. In general, the rate of progression of scoliosis in adulthood is much slower than in adolescence and is very much dependent on the size of the curve once skeletal maturity has been reached. Regardless of the curve pattern, curves less than 30 degrees in the mature individual are unlikely to progress. Conversely, approximately two-thirds of curves that exceed 50 degrees worsen, with thoracic curves progressing nearly 1 degree per year.[462] Lumbar curves also tend to progress in adulthood, and will do so at a smaller magnitude than 50 degrees if they are accompanied by a transitory shift between the lower vertebrae.

Looking at the long-term status of adults with untreated scoliosis, several Swedish studies reported an overall mortality rate greater than that predicted by the national statistics for mortality.[315,321,343] However, these studies included patients with nonidiopathic scoliosis and patients with infantile deformities. When examined selectively, the mortality rate for patients with adolescent idiopathic scoliosis appeared to be the same as expected for the general population.[343] A significant increase in the mortality rate was identified for those patients with infantile and juvenile idiopathic scoliosis. Respiratory failure and cardiovascular disease accounted for most early deaths. Respiratory failure developed in adults with severe scoliosis (exceeding 110 degrees) as normal aging further reduced their ventilatory capacity.[342] However, patients operated on for scoliosis tended not to develop respiratory failure, which suggested a preventive effect of corrective surgery.[344]

Chronic back pain, although not related to size or location of the curvature, is more common in adults with scoliosis.[315,460] The pain usually does not interfere with the patient's ability to work or perform daily activities. Lumbar osteoarthritis may also be seen in up to 83 percent of adults with scoliosis, but it is not necessarily associated with the duration or intensity of back pain. Despite outwardly apparent deformities due to long-standing untreated scoliosis, the majority of individuals have no significant psychological difficulties when compared with persons without scoliosis (the sole exception being a slight dissatisfaction with body image).[461]

In summary, thoracic scoliosis exceeding 50 to 60 degrees in adulthood may progressively worsen and potentially reduce the person's pulmonary function. Lumbar curves, especially those greater than 50 degrees, also are likely to progress in adult life, leading to osteoarthritis. Therefore, even when cosmetic factors are not taken into account, aggressive treatment of the child with a significant spinal deformity is justified.

SCOLIOSIS SCREENING

School Screening Programs. A number of medical organizations have supported the general screening of children for scoliosis. The Scoliosis Research Society has recommended screening of children between 10 and 14 years of age.[398] The American Academy of Orthopaedic Surgeons has advocated screening of girls at ages 11 to 13 years and of boys at 13 to 14 years.[8] The American Academy of Pediatrics has recommended screening at routine health supervision visits at ages 10, 12, 14, and 16 years.[9] In some states, scoliosis screening is mandated by law. In a 1989 survey, 15 states reported statutes and five states had administrative regulations requiring screening.[17] Thirty-one states had voluntary screening programs. The range of school grades screened is from 3 through 12, although most screening is performed in grades 5 through 9.

The clinical logic behind school screening for idiopathic scoliosis assumes that (1) screening is an accurate and reliable method for detecting curvatures, (2) early detection results in improved health outcomes, and (3) brace therapy is effective in altering the natural history of the deformity.[4,308,400,468] The implications in these assumptions are that (1) small curvatures detected through screening are likely to progress to curvatures of clinical significance, (2) scoliosis causes significant health problems, and (3) the benefits of early detection outweigh the potential adverse effects of screening and treatment. The many proponents of school screening believe that these assumptions are successfully addressed by school screening programs.* Earlier detection and the institution of brace treatment have reportedly reduced the need for operative intervention.†

*See references 59, 152, 255, 259, 316, 385.

*See references 51, 60, 61, 155, 203, 253, 257, 258, 260, 267, 304, 368, 419, 458, 478, 480.

†See references 60, 131, 203, 253, 258, 261, 304, 316, 444.

The idea of general scoliosis screening, however, is not universally accepted. Some authors have argued that school screening programs (1) have not been helpful in reducing the prevalence or incidence of scoliosis requiring treatment, (2) are not cost-effective, and (3) result in unnecessary referrals to orthopaedic surgeons or radiologists of children with no scoliosis or only a mild degree of curvature that does not require treatment.‡ The British Orthopaedic Association and the British Scoliosis Society issued a statement in 1983 advising against a national policy of screening for scoliosis in the United Kingdom.[56] More recently, the United States Preventive Services Task Force stated that there is insufficient evidence to recommend for or against the routine screening of asymptomatic adolescents for idiopathic scoliosis.[35,399,400,448] In addition, the task force reported that prior literature did not support routine clinical visits specifically for scoliosis screening or for examinations at specific ages during adolescence. Further clinical research to demonstrate the effectiveness of school screening was recommended.

Screening Methods. Several clinical signs are indicative of possible scoliosis and are frequently used in screening programs. These include shoulder asymmetry, unequal scapular prominence, appearance of an elevated or prominent hip, greater space between the arm and body on one side (with the arms hanging loosely at the side), head not centered over the pelvis, and a positive Adams forward-bending test.

The Adams test is performed by examining the patient from the rear, having the child bend forward until the spine is horizontal, and noting whether one side of the back appears higher than the other (Fig. 11–4). This test is the best noninvasive clinical method for evaluating scoliosis.[1,60,83]

One of the constant features of a structural scoliosis is axial rotation of the vertebrae affected by the curve. The spinous processes always rotate toward the concavity of the curvature. Rotation of the thoracic vertebrae is also impaired by rotation and deformity of the attached rib cage, with elevation on the side of the convexity and depression on the side of the concavity. This asymmetry is significantly accentuated when the patient bends forward.

Examining the patient in the forward-bent position is the standard method used to detect a mild degree of curvature in mass screening programs. However, the mere presence of asymmetry observed during this maneuver does not necessarily indicate a scoliotic deformity severe enough to warrant referral to an orthopaedic surgeon. This is a point of contention for those who believe that screening leads to excessive and unnecessary referrals.

In an effort to quantitatively assess the asymmetry and, by doing so, establish an appropriate degree of deformity necessary to justify referral for medical evaluation, Bunnell introduced the scoliometer (Fig. 11–5) in 1984.[60] This specially designed inclinometer (similar to a level used in a woodshop) measures the angle of vertebral rotation. When using the scoliometer, it is important that the screener stand behind the patient to view the back (as in the Adams forward-bending test). The screener's eyes should be on a horizontal plane with the maximum deformity of the back. If the patient bends forward approximately 45 degrees, the outline of the trunk at the level of the thoracic spine is seen.

*See references 60, 131, 151, 203, 253, 258, 261, 304, 306, 316, 362, 444.

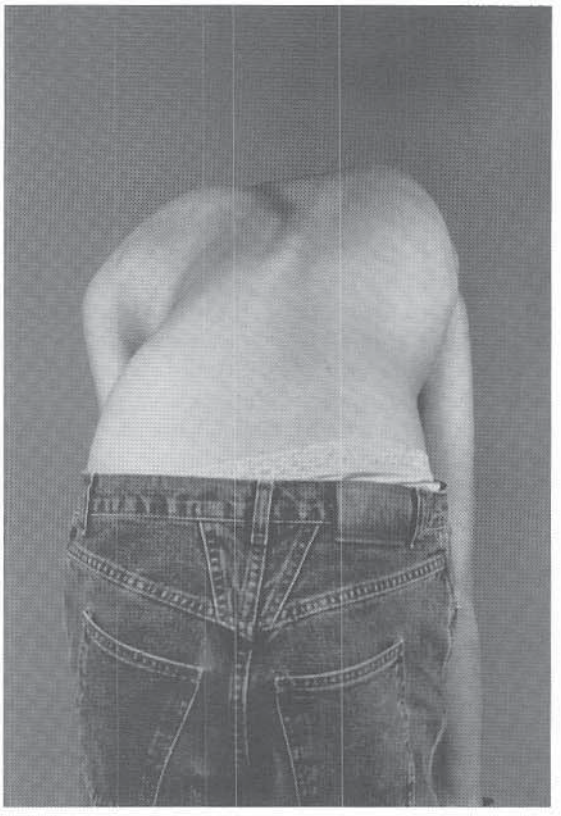


FIGURE 11–4 The Adams forward bending test. The patient is viewed from behind and is asked to bend forward until the spine is horizontal. When scoliosis is present, one side of the back appears higher than the other.

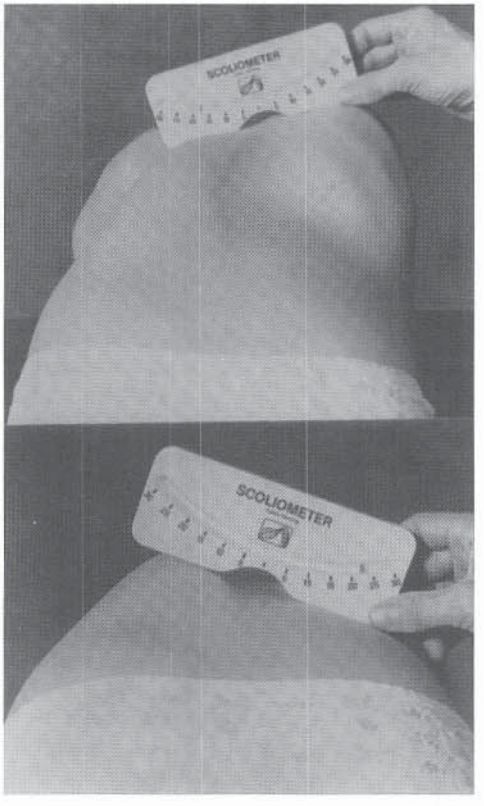


FIGURE 11–5 The scoliometer is a specially designed inclinometer used clinically to measure the angle of vertebral rotation.

With further bending, the outline of the trunk at the level of the thoracolumbar spine is seen, followed finally by the outline at the level of the lumbar spine.

If a rotational deformity of the back is noted at any level, the scoliometer is placed gently on the back at the apex of the deformity, perpendicular to the long axis of the body, and the angle of inclination is read directly from the scale. Originally, the recommendation for orthopaedic referral was a 5-degree angle of trunk rotation at any level of the spine.[60] By doing so, the chances of missing a curve exceeding 20 degrees were small. However, because this recommendation continued the likelihood of excessive referrals, it has been modified to a 7-degree angle of trunk rotation.[61] With this criterion, the chances of missing a curve greater than 30 degrees (the curve magnitude at which bracing would usually be initiated) are small. With this approach, the referral rate would be approximately 3 percent of those persons screened, with a 95 percent detection rate of those curves requiring brace treatment. By using this method, over-referral can be avoided and the cost-effectiveness of spinal screening programs can be maintained.

ETIOLOGY

The exact cause of idiopathic scoliosis remains unknown despite considerable investigation into its etiology. Although growth has a significant influence on the deformity, it is not considered a causative factor. Over the past 10 years, much of the research into the etiology of scoliosis has focused on central neurologic dysfunction, connective tissue abnormalities, and genetic factors. These influences have supplanted previous theories that idiopathic scoliosis was caused by a biochemical or nutritional deficiency,[170,196,496] structural defects,[64] or endocrine abnormality.[416] Most likely the true cause is multifactorial and involves several of the aforementioned factors.

Neurologic Dysfunction. The bulk of the current literature continues to support an underlying neurologic abnormality as the primary etiologic factor in idiopathic scoliosis. Dysfunction of the vestibular, ocular, and proprioceptive systems causes an interruption of equilibrium that is indicative of abnormalities involving the posterior column of the proximal portion of the spinal cord, brain stem, and cerebral cortex.* Responses to vibratory stimuli have been reported to be significantly reduced and to be asymmetric between left and right sides in scoliotic patients when compared with controls.[24,497] These findings support the concept that an aberration in the function of the posterior column pathway of the spinal cord may have a role in the etiology. Other investigators, however, have not been able to corroborate this opinion.[287] Altered balance affecting foot posture and gait, particularly pes cavus, has been reported.[68,146] In addition to abnormalities in sensory pathways, motor dysfunction has been reported, suggesting that the organization of the entire brain is asymmetric in individuals with scoliosis.[150]

Another proposed neurologically based theory for idiopathic scoliosis involves the role of melatonin in regulating normal spine growth. Secreted by the pineal gland, this neurohormone controls the circadian rhythm. Experiments on pinealectomized chickens revealed that melatonin deficiency contributed to the development of scoliosis in this model, probably by interfering with the normal symmetric growth of the proprioceptive system involving the paraspinal muscles and the spine.[269–271,273] Significantly lower melatonin levels have been reported in patients with scoliosis when compared with controls.[270] Other investigators, however, have refuted this finding.[179]

Connective Tissue Abnormalities. Another continued focus of research is alterations in connective tissue involving the spine, paraspinal muscles, and platelets in patients with scoliosis.* Differences in collagen have been found between normal individuals and those with adolescent idiopathic scoliosis. However, this finding is not universal.[454] These changes may be secondary to the mechanical effects of the spinal deformity rather than reflecting mutations in the collagen itself. This theory has been substantiated by segregation analysis of genetic markers linked to the structural genes encoding types I and II collagen.[69]

Other components of connective tissue may also be abnormal. In histologic studies of the ligamentum flavum in scoliotic patients, the elastic fiber system has been shown to have disarranged fibers, a marked decrease in fiber density, and a nonuniform distribution of fibers throughout the ligament.[163] These findings suggest that the elastic fiber system (which is predominantly fibrillin) may play a significant role in the pathogenesis of idiopathic scoliosis in some individuals. Bone mineral density has also been shown to be lower in young adolescents with scoliosis.[77] It is uncertain, however, whether this finding is related to the primary etiology of the disease or whether it is secondary to the asymmetric mechanical forces associated with the back deformities.

The paravertebral musculature in scoliosis patients may exhibit abnormalities in the muscle spindle,[264] in individual muscle fiber morphology,[190,390,506] in histochemistry,[422] and on electromyography.[5,369] Some of these changes are more pronounced in severe curves, but they are believed to be secondary to muscle adaptation to the curve and not a primary cause of the deformity.[190]

Abnormal platelet structure and function have been reported in patients with scoliosis.† Calmodulin, a calcium-binding receptor protein found in platelets and skeletal muscle, regulates the contractile protein system (actin and myosin). If there is an underlying systemic contractile disorder, platelets and skeletal muscle would both be affected. Thus, measurable abnormalities of calmodulin in platelets are indicative of skeletal muscle abnormalities. Platelet calmodulin levels in adolescents with progressive scoliosis are significantly higher than the levels seen in normal individuals or in patients with stable curves.[219] Although this finding cannot be implicated as the direct cause of scoliosis, it may be a useful predictor of curve progression.

Genetic Factors. Because idiopathic scoliosis can be seen in multiple members of the same family, researchers have attempted to determine the cause of the condition based on the genetic factors involved. Several extensive clinical studies of affected families were conducted approximately 30 years ago.[86,379,500] Because of a high prevalence of familial scoliosis

*See references 24, 55, 68, 146, 150, 211, 245, 325, 392, 495, 497, 503.

*See references 69, 77, 163, 190, 219, 246, 298.

†See references 137, 219, 246, 312, 345, 389.

(6.9 to 11.1 percent of first-degree relatives), a dominant inheritance pattern or a multifactorial mode of inheritance was suggested as the cause of adolescent idiopathic scoliosis (Fig. 11–6). Not all studies suggested a genetic basis; some instead implicated higher maternal age at the time of childbirth.[95] However, evidence of a strong genetic tendency in adolescent idiopathic scoliosis was recently reaffirmed with a meta-analysis report on scoliosis in twins.[214] Monozygous twins had a significantly higher rate of concordance than dizygous twins, and the curves in monozygous twins developed and progressed together.

Studies are underway to identify the specific gene or genes that cause scoliosis and its progression.[298] This new investigative frontier uses genetic linkage to analyze genomic DNA from families with apparent autosomal dominant inheritance of adolescent scoliosis. Analysis of families through a candidate gene approach would potentially enable investigators to link a gene or a genetic locus to the disease, track the disease within a pedigree, and predict the genotype of future individuals. In this way, specific markers for abnormal structural genes may be identified, thereby providing more insight into the etiology of idiopathic scoliosis.

PATHOPHYSIOLOGY

The extent of structural changes varies with the degree of scoliosis.[121,381] These changes are greatest at the apex of the curve and diminish toward either end. In structural scoliosis, the rotation of the vertebral body is to the convex side of the lateral curvature, so that the spinous processes of the vertebrae are rotated toward the concavity of the curve.

Forces in compression and distraction act on the growing spine and produce changes in the vertebrae, which become wedge-shaped, higher on the convex side and lower on the concave side (Fig. 11–7). The vertebral body becomes condensed on the concave side as a result of the greater pressure, and is expanded and thinned on the convex side. In addition to the changes in the frontal and axial planes, the scoliotic portion of the spine is lordotic in the sagittal plane.[100,101] This three-dimensional deformity is appropri-

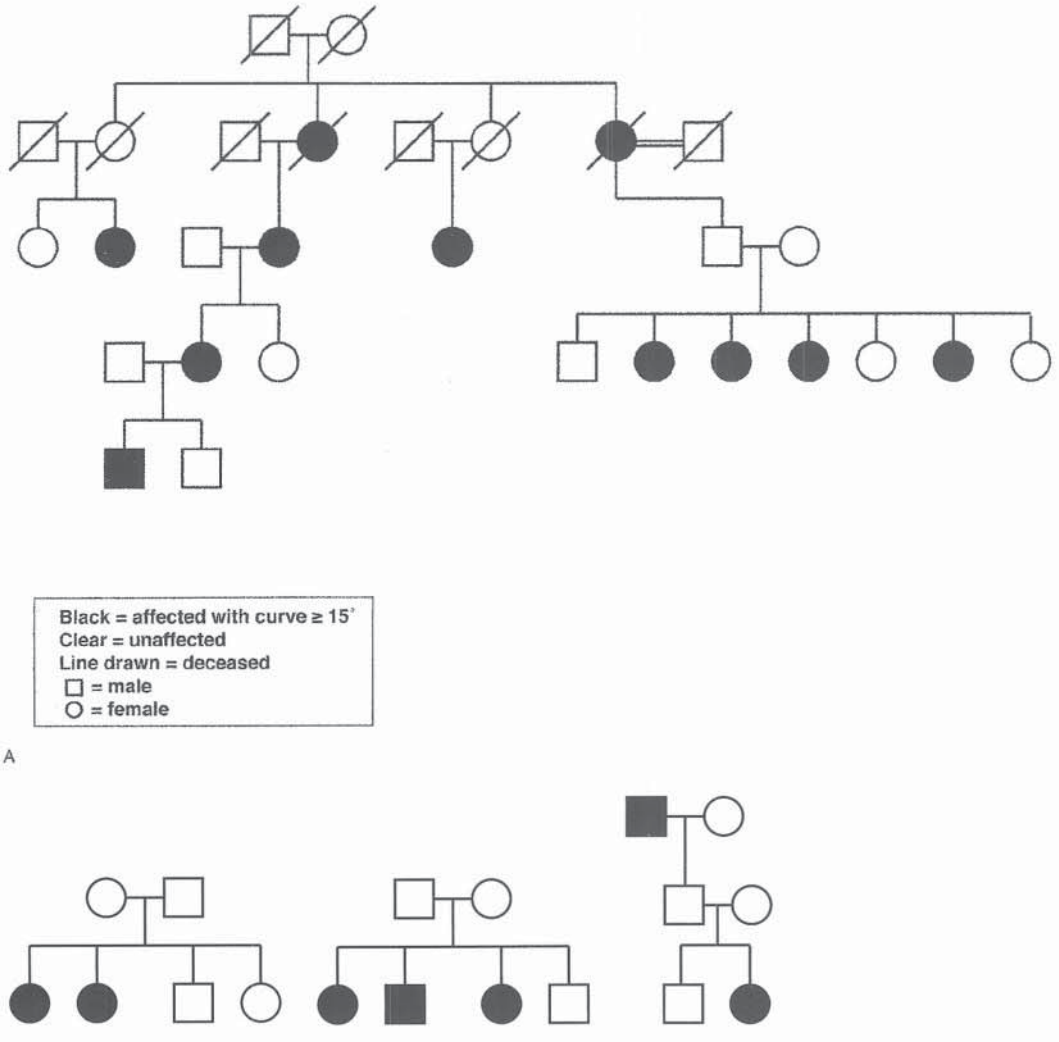


FIGURE 11–6 **A,** A family tree of five generations demonstrates an apparent dominant pattern of inheritance. **B,** Three other small family trees reflect probable multifactorial modes of inheritance.

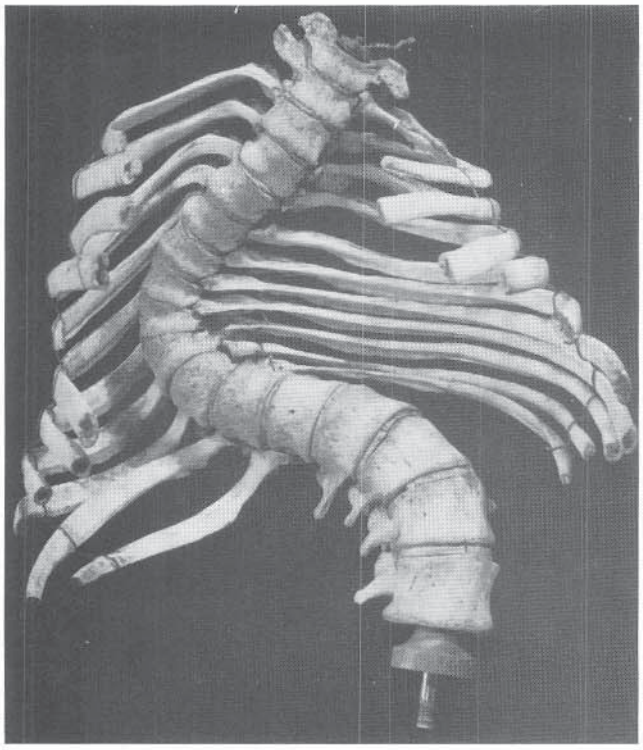


FIGURE 11–7 Gross anatomic specimen of a spine showing changes that developed with an extremely severe right thoracic scoliosis. The vertebral bodies became trapezoidal, with the narrower side on the concavity. The rotation of the spine is so severe in this specimen that the anterior aspect of the apical region is facing 90 degrees to the right. (From James JIP: Scoliosis, p 13. Baltimore, Williams & Wilkins Co, 1967.)

ately termed *torsion of the spine* and is greatest at the apical region.[100,101,109,347–349]

In the scoliotic spine, there are associated changes in the neural canal and the posterior arch. In more severe deformities, the laminae on the convex side are broad and widely separated, while those on the concave side are narrow and close together (Fig. 11–8). The pedicles are shorter and stubbier on the concave side. The transverse processes more closely approach the sagittal plane on the convex side and are more in the frontal plane on the concave side. The intraspinal canal becomes distorted because of the misshapen pedicles and articular processes (Fig. 11–9).

As a result of pressure over time, the intervertebral disks on the concave side narrow and show degenerative changes. The adjoining portion of the vertebra becomes sclerotic, with marginal lipping.

The thoracic cage is also affected by the deformity. Because of rotation of the thoracic vertebrae, the ribs on the convex side are directed posteriorly, producing a rib prominence that, in severe cases, may be referred to as a "razorback." On the concave side, the ribs are rotated forward, potentially producing prominence of the anterior chest wall. The sternum may be asymmetric and laterally displaced from the midline. The breasts often are mildly asymmetric owing to the chest wall deformity. Breast asymmetry often is a major concern of patients.

Because of the spinal deformity, the thoracic cavity is no longer symmetric. Its capacity is diminished on the convex side and increased on the concave side. In severe cases in which there is marked angulation of the ribs posteriorly, lung function may be altered.[342,449]

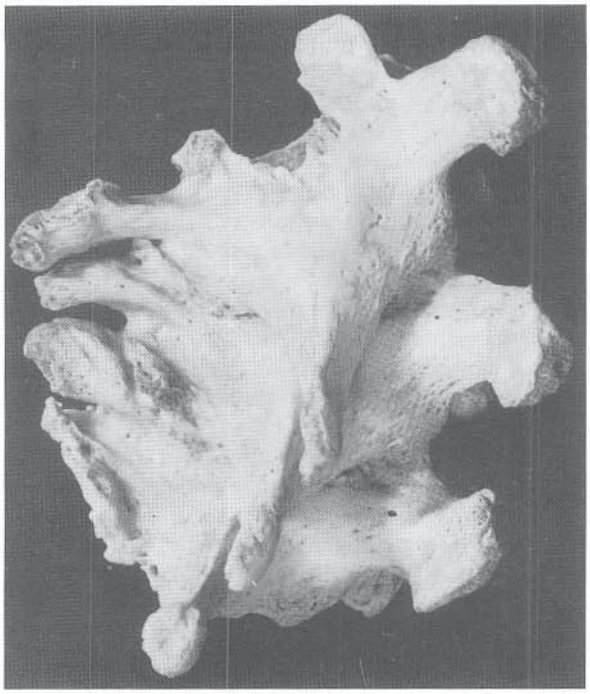


FIGURE 11–8 The posterior elements of the spine at the apical region of this severe scoliosis show notable deformity, with the laminae on the concave side being narrow and close together. (From James JIP: Scoliosis, p 15. Baltimore, Williams & Wilkins Co, 1967.)

In severe cases of scoliosis in which the shape of the intraspinal canal is distorted, the spinal cord may be stretched over the concave side, but rarely is there any neurologic deficit. Cord compression with neurologic deficit usually occurs only in extreme deformities that are accentuated by marked thoracic kyphosis.

FIGURE 11–9 The intraspinal canal is slightly distorted, owing to short, misshapen pedicles. The transverse processes are asymmetric. (From James JIP: Scoliosis, p 15. Baltimore, Williams & Wilkins Co, 1967.)

PATIENT EVALUATION

Presenting Complaint. Adolescents with scoliosis usually do not seek medical evaluation because of back discomfort but rather because of some physical aspect of their deformity. These abnormalities include a high shoulder, one-sided prominence of a scapula or breast, elevated or protuberant iliac crest, and asymmetry in flank creases and the trunk. Except for being noticed personally by the adolescent, these findings often are first appreciated by someone else during school screening programs for scoliosis or during back-to-school examinations by the family physician.

Although uncommon, back pain is present in individuals with idiopathic scoliosis more often than was previously thought. Nearly 32 percent of adolescents with idiopathic scoliosis complain of back discomfort at some point in time (23 percent upon presentation and 9 percent during the period of observation).[366] There is a significant association between back pain and patient age greater than 15 years, skeletal maturity greater than or equal to Risser sign 2, postmenarchal status, and a history of injury. Back pain does not seem to be related to patient sex, a family history of scoliosis, limb length discrepancies, magnitude or type of curve, or spinal alignment. In patients who do present with back pain, the source of discomfort can be identified only 10 percent of the time, despite the use of appropriate imaging studies. The most common causes of discomfort are associated spondylolysis, spondylolisthesis, or Scheuermann's kyphosis. Less likely causes include spinal cord syrinx, disk herniation, tethered spinal cord, and tumor. A painful left thoracic curve or an abnormal neurologic finding is most predictive of an underlying pathologic condition of the spinal cord.

When an adolescent with presumed idiopathic scoliosis has back pain, a careful history should be obtained, a thorough physical examination performed, and plain radiographs ordered. If findings on this initial evaluation are normal, a diagnosis of idiopathic scoliosis can be made, the scoliosis can be treated appropriately, and nonsurgical treatment can be initiated for the back discomfort. It is not necessary to perform extensive diagnostic studies to evaluate every adolescent with scoliosis and back pain. If the patient's symptoms persist and significantly restrict normal activities and if the neurologic examination is normal, a technetium bone scan may be useful. If the neurologic examination is abnormal, magnetic resonance imaging (MRI) of the spinal cord is indicated. Unlike backache in adults with lumbar scoliosis, backache in adolescents usually is not due to degenerative arthritis in the posterior articulations or to nerve root irritations.

Respiratory symptoms are uncommon in patients with adolescent idiopathic scoliosis. Studies have shown that cardiopulmonary compromise usually does not occur until curve magnitude approaches 100 degrees, vital capacity becomes less than 45 percent, or thoracic lordosis significantly narrows the anterior-posterior dimensions of the chest.[342,344] Most curves are operatively treated well before spinal deformity becomes this severe.

Neurologic deficits also are rare in adolescent idiopathic scoliosis. Should the adolescent describe any suspicious symptoms (e.g., persistent neck pain, frequent headaches, ataxia, weakness), meticulous attention must be given to the neurologic portion of the physical examination. If any neurologic deficits are found or if the convexity of the thoracic curve is to the left, appropriate imaging of the neural axis is undertaken.[73,126,323,366,397,443] Normally, the convexity of thoracic curves in adolescent idiopathic scoliosis is directed to the right. Abnormal left thoracic curves are more common in the presence of an underlying syrinx.[366,397]

Physical Examination. The physical examination of the adolescent with idiopathic scoliosis should be performed with the patient properly draped. It is convenient if the patient wears a swimsuit. Alternatively, the patient may be dressed in underpants and an examination gown open at the back. The patient's entire back, including the shoulders and the iliac crests, needs to be visible.

The skin is inspected closely for abnormalities such as midline hemangiomas, hair tufts, and dimpling in the lumbosacral region. Any of these surface findings may indicate the presence of an underlying spinal cord abnormality such as tethered cord or diastematomyelia. The spinous processes are palpated from the cervical region to the sacrum for any deficiencies or areas of discomfort. Occasionally absence of a spinous process is noted, which usually corresponds to a spina bifida occulta seen on the spine radiograph (Fig. 11–10).

With the patient standing, the examiner should determine whether the iliac crests are level. If they are not, a lower limb length discrepancy is likely to be present, which can be quantified by placing measured blocks under the short extremity until the iliac crests are level. Lower limb

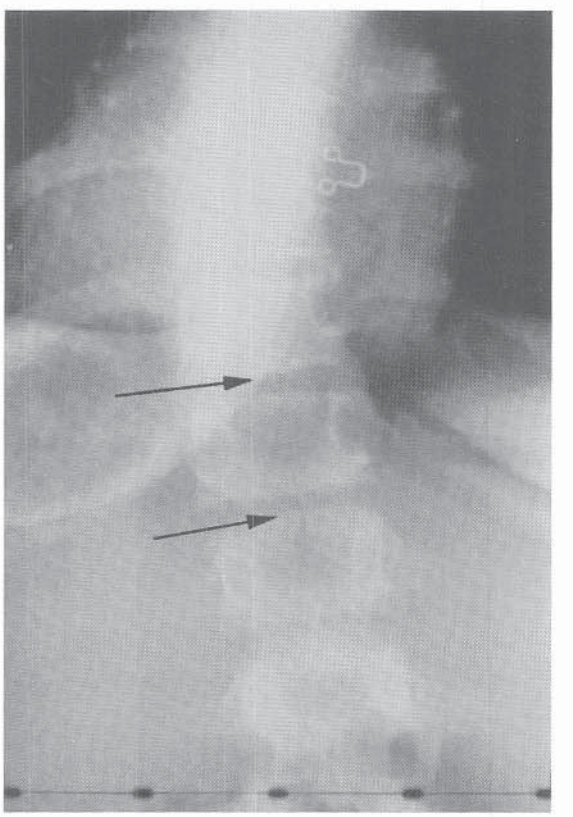


FIGURE 11–10 PA radiograph of the spine of a 12-year-old girl. The spinous processes of T11 and T12 were not detectable on palpation. The radiograph shows spina bifida occulta at the same levels (*arrows*).

length discrepancy can be responsible for the appearance of scoliosis, and the condition must not be overlooked. The back is then examined for asymmetry of the shoulders and flank creases, unequal scapular prominence, a prominent iliac crest, and increased space between the arm and body on one side compared with the other side (with the arms hanging loosely at the side) (Fig. 11–11).

Although these findings are consistent with scoliosis, the best noninvasive clinical test for evaluating spinal curvature is the Adams forward-bending test (see Fig. 11–4).[1,60,83] With this test, the degree and direction of associated rotation of the vertebrae are demonstrated most clearly. The examiner observes the adolescent from behind as the patient bends forward at the waist until the spine is horizontal. The patient's knees should be straight, the feet together, the arms dependent, and the palms in opposition. Vertebral rotation will cause one side of the back to appear higher. This is noted as a rib prominence in the thoracic region or as a paraspinal fullness in the lumbar region. This asymmetry can be quantified with the scoliometer, which, if used over time, can provide measurements documenting change (see Fig. 11–5).[60]

Often, if the patient is inspected from the front, an asymmetry of the pectoral regions, breasts, or rib cage may be evident. Although these asymmetries are probably related to the spinal curvature, they may also be seen in individuals without scoliosis. Occasionally the breast asymmetry is the primary concern of the patient and parents. Families should be informed that correcting the scoliosis may have little, if any, influence on this asymmetry.

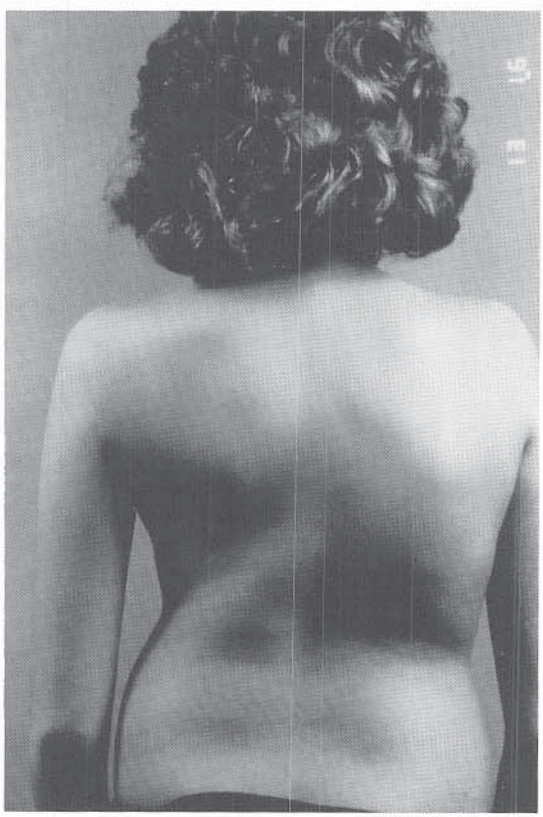


FIGURE 11–11 Clinical appearance of a 13-year-old girl with a right thoracic and left lumbar scoliosis. The right scapula is prominent, and the space between the left arm and body is increased. The shoulders are level.

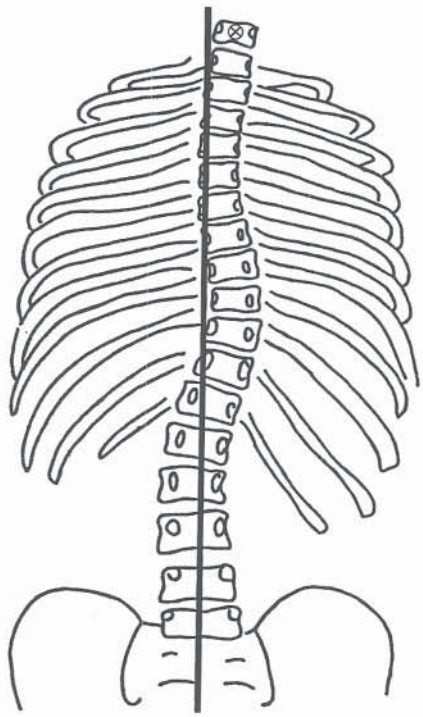


FIGURE 11–12 A plumb line held at the spinous process of C7 (*x*) should not deviate from the center of the gluteal fold (center sacral line) by more than 1 to 2 cm. (From Richards BS, Birch JG, Herring JA, et al: Frontal plane and sagittal plane balance following Cotrel-Dubousset instrumentation for idiopathic scoliosis. Spine 1989;14[7]:733–737.)

Spinal balance is assessed by two different methods. The first way is to determine the alignment of the head over the pelvis. The head is almost always positioned directly above the gluteal crease in patients with idiopathic scoliosis. To assess this balance, a plumb line is held from the base of the skull or from the spinous process of C7. Normally the plumb line should not deviate from the center of the gluteal crease by more than 1 to 2 cm (Fig. 11–12). If it does, this finding should be considered atypical, and a meticulous neurologic examination is necessary to rule out coexisting neurologic pathology. The second method of evaluating spinal balance is to assess the position of the trunk over the pelvis.[26,138,372] Unlike the position of the head over the pelvis, there may be significant imbalance of the trunk over the pelvis in patients with idiopathic scoliosis (Fig. 11–13). This is particularly true in single thoracic curve patterns.

Next, the examiner inspects the patient from the side and observes the sagittal contours of the spine. Normally, in idiopathic scoliosis, the sagittal plane is mildly hypokyphotic in the scoliotic segment. In more severe cases the sagittal plane may actually be lordotic, leading to a very narrow anteroposterior (AP) diameter of the thoracic cage. In the very rare severe cases, there may be as much as 90 degrees of rotation in the apical vertebrae within the curve. In this instance the examiner is actually assessing the AP aspect of the spinal curvature when viewing the spine from the side. The resultant clinical appearance is that of an apparently increased kyphosis in the sagittal plane because, in reality, the scoliotic deformity is being viewed from the side (see Fig. 11–7).

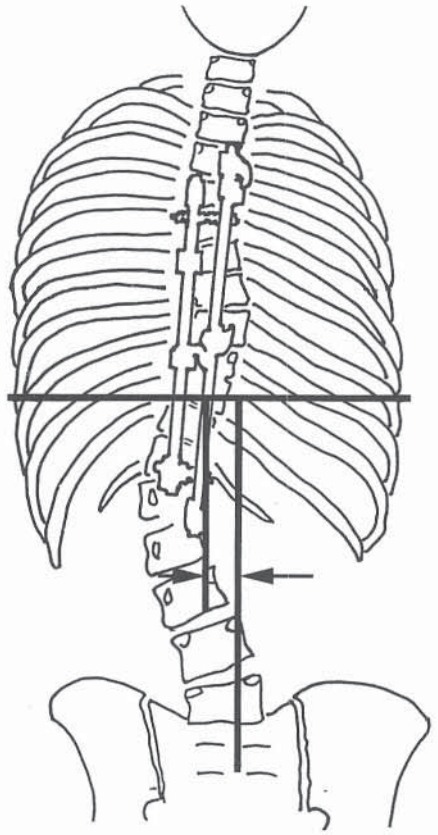


FIGURE 11–13 To measure trunk balance, two vertical lines are drawn on a radiograph. The first vertical line is the center sacral line. The second vertical line bisects a horizontal line drawn from the peripheral edges of the ribs of the apical vertebra. The distance between the two vertical lines quantifies the amount of trunk imbalance. (From Richards BS: Lumbar curve response in type II idiopathic scoliosis after posterior instrumentation of the thoracic curve. Spine 1992;17[8 Suppl]:S282–S286.)

Neurologic Examination. Because idiopathic scoliosis is basically a diagnosis of exclusion, a thorough evaluation is necessary to rule out a neurologic cause for the deformity. The neurologic examination begins by assessing the patient's reflexes. Examination of the superficial abdominal reflexes is useful for determining which patients should undergo MRI to rule out syringomyelia (Fig. 11–14).[504]

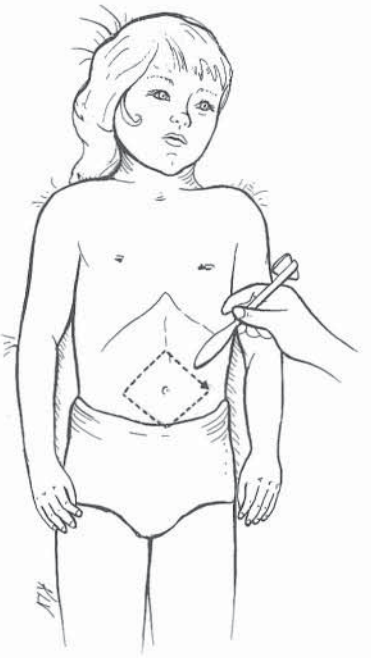


FIGURE 11–15 The abdominal reflex examination is performed with the patient supine. A bluntly pointed handle of a reflex hammer is used to lightly stroke the skin in each quadrant over a distance of 10 cm. Asymmetry of the reflex between sides is abnormal.

The abdominal reflex examination is performed with the patient supine on an examination table with the arms relaxed along the side of the body. An area approximately 10 cm above and below the umbilicus and to each anterior axillary line is exposed. With the patient relaxed, the bluntly pointed handle of a reflex hammer is used to lightly stroke the skin in each quadrant over a distance of 10 cm (Fig. 11–15). The stroke starts lateral to the umbilicus near the anterior axillary line and is directed diagonally toward the umbilicus in each quadrant. The umbilicus is observed for deviation toward the side of the test. If these reflexes are consistently present on one side and absent on the other side, further workup is warranted, since this finding does not occur in normal patients with scoliosis. However, other variations might occur, such as absent reflexes in all quadrants.

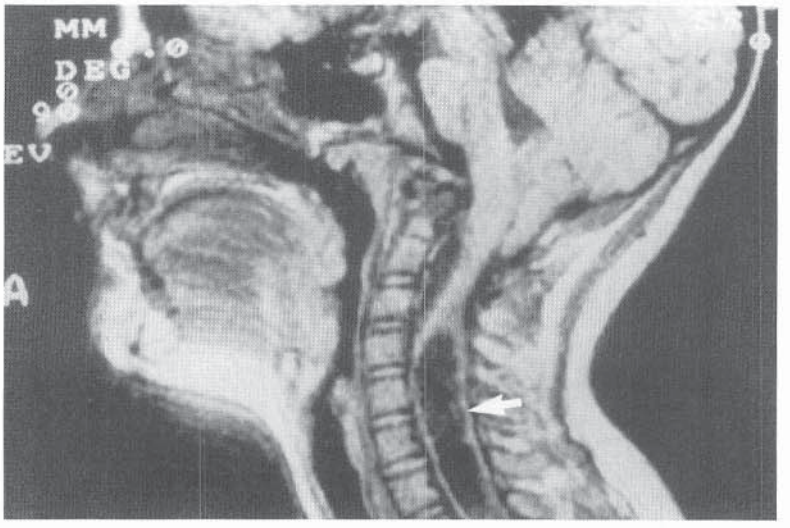


FIGURE 11–14 Diagnostic imaging of the spinal cord and canal is necessary in children with abnormal neurologic findings. MRI is the optimal study for assessing the neural axis. A large cervical syringomyelia (*arrow*) is evident on this MR image of the head and neck. (From Richards BS: Back pain in childhood and adolescence: the clinical assessment. J Musculoskeletal Med 1998;15:39.)

The patellar and Achilles tendon reflexes should also be tested, with the expectation that they will be symmetric.[504] Muscle testing and examination of the range of motion in all four extremities should always be conducted but need not take long to perform. The hands and feet should be examined for abnormal posture and for evidence of abnormal sensation (excessive callus formation or nail bed irregularities). Abnormal findings may be the only clinical evidence of underlying pathology of the neural axis, such as syringomyelia or tethered cord.

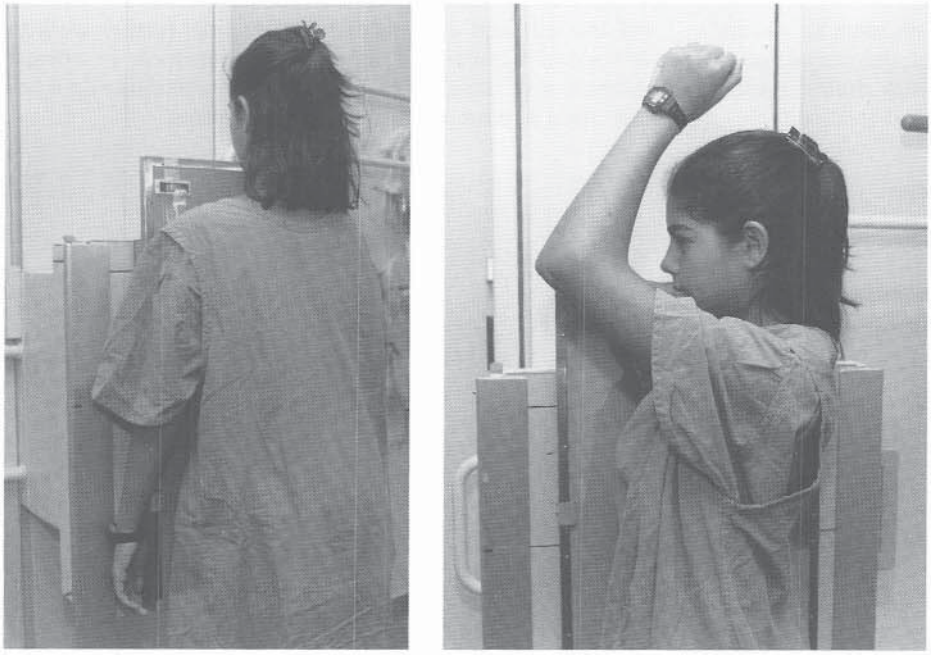
Patient Maturity. Sexual maturity can be assessed during the physical examination according to the Tanner system.[435] This system assesses breast and pubic hair development in girls and genital and pubic hair development in boys. However, while the Tanner system may provide an indication of the patient's physical maturity, more practical clinical emphasis is placed on the patient's menarchal status and increase in height over time, and on an assessment of skeletal indicators for maturity (e.g., Risser sign, open or closed triradiate cartilages).

IMAGING STUDIES

Plain Radiography. The initial examination of the spine should include posteroanterior (PA) and lateral radiographs on 36 × 14-inch film cassettes. By using these long cassettes, nearly all of the important radiographic features can be assessed on a single film. On the PA projection, these features include the curve pattern in its entirety, the type of scoliosis (congenital or idiopathic), the overall balance of the spine and trunk, skeletal maturity (as determined by the Risser sign, triradiate cartilages, or capital femoral physis), and the presence of a lower limb length discrepancy (pelvic tilt). The lateral projection is useful initially to evaluate the global sagittal contour of the thoracic and lumbar spine, determine the presence and severity of thoracic hypokyphosis, and screen for spondylolysis and spondylolisthesis. In very young children, 17 × 14-inch film cassettes may be large enough to provide all of this information. However, these shorter cassettes are too small to be used for adolescent patients. With female patients, the examiner should always inquire as to when their last menstrual period took place, so that if pregnancy is suspected, the radiographic evaluation should be postponed.

Because studies have shown that the risk of breast and thyroid cancer may be slightly increased in scoliosis patients undergoing multiple radiographic examinations, methods to reduce the amount of x-ray exposure have evolved over the years.[7,97,114,181,243] The PA projection has replaced the AP projection for frontal plane assessment because the former results in significantly less radiation exposure to breast and thyroid tissue.[128,141,157,243] Although the quality of bone detail may be somewhat less on the PA projection than on the AP projection, the difference is negligible when compared with the significant reduction in radiation exposure. Other methods of reducing the amount of radiation exposure include the use of specially designed leaded acrylic filters, high-speed screen-film systems, beam collimation, specially designed cassette holder and grid, and radiopaque breast and gonadal shields.[157] Occasionally, however, the need to assess bone detail may preclude the use of shields. Newer techniques, such as digital radiography, may result in even less radiation exposure.

After the initial radiographic evaluation has been accomplished, an effort is made to limit the number of follow-up films, thereby reducing the amount of radiation exposure. During the course of routine follow-up examinations, only the PA projection is needed. There is *no* set time interval from one radiographic examination to the next for all patients. The period between evaluations depends on the maturity of the patient and the size of the spinal curvature. For



A B

FIGURE 11–16 During the radiographic evaluation, the patient stands erect with the knees straight and the feet together. The PA projection (**A**) reduces exposure to breast tissue. During the lateral view (**B**), the arms are held forward to allow clear visualization of the spine.

example, a premenarchal, Risser 0, 11-year-old girl with a 25-degree thoracic curve should return for radiographic reevaluation after a 4-month interval. On the other hand, a 2-year postmenarchal, Risser 4, 14-year-old girl with a 30-degree curve need not return for reevaluation before 1 year. In most cases the interval between radiographic evaluations ranges from 4 to 6 months.

During the radiographic evaluation, the patient should stand as erect as possible with the knees straight and the feet together (Fig. 11–16). If possible, the patient should be barefoot, so that if lower limb length inequality is suspected, the appropriate lift can be placed under the short limb. Unsupported sitting views are taken if the patient is unable to stand. There should be no twisting of the trunk. To ensure sufficient cephalad visualization, the upper limit of the cassette should extend to the external auditory meatus. In the upright lateral projection, the patient's shoulders are flexed forward 90 degrees with the arms resting on a stand. Having the patient in this position avoids imposing confusing shadows of the upper extremities over the spine. Although AP radiographs of the left hand and wrist are used by some orthopaedists to determine skeletal age, maturity is more commonly assessed from the Risser sign on the iliac crest.

Bending radiographs obtained with the patient supine are usually reserved for preoperative evaluations of spinal flexibility. The information gained from these AP radiographs can be helpful in determining appropriate fusion levels.

MEASUREMENT OF CURVE MAGNITUDE. The Cobb method is considered the standard for measuring curve size.[19] The measurement is started by determining the end vertebrae (top and bottom of the curve). The cephalad end vertebra's superior surface and the caudad end vertebra's inferior surface will have the greatest amount of tilt into the curve (Fig. 11–17). The intervertebral space on the concave side of the curve usually is wider above the cephalad (top) vertebra and narrower below it. The opposite applies to the inferior surface of the caudad (bottom) vertebra. Using a transparent plastic goniometer, the examiner draws lines perpendicular to the top vertebra's superior surface and the bottom vertebra's inferior surface (Fig. 11–18). The angle formed by the intersection of these lines is the Cobb angle. If a second curve is present below the primary curve, the original curve's bottom vertebra becomes the cephalad end vertebra for the second curve and the same line along its inferior surface is used.

Although the Cobb method is considered the standard measurement technique, there is always some variation between different observers' measurements. This variability averages 7.2 degrees if the end vertebrae are not preselected but improves to 6.3 degrees when they are preselected.[309] Another reported aspect of the accuracy of the Cobb method is that there would have to be a measurement difference of 10 degrees between radiographs taken at different times to achieve a 95 percent statistical confident that a true change in curve size had occurred.[67] This finding is of particular interest because many studies that report on the effectiveness of brace management for scoliosis use a criterion of a 5- to 6-degree change in curve size to determine the success or failure of brace treatment. This information reinforces the importance of meticulous line drawings and precise measurements.

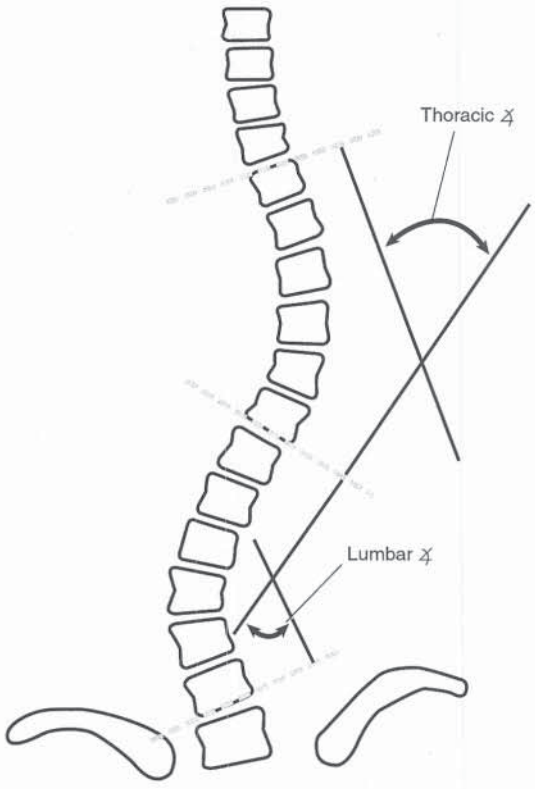


FIGURE 11–17 The Cobb measurement. The vertebrae with the greatest amount of tilt are selected as the end vertebrae. Lines are drawn perpendicular to the end-plates of the vertebrae. The angle formed at the intersection of these lines is the Cobb angle. If a second curve is present below the primary curve, the original curve's lower vertebra becomes the top vertebra for measuring the second curve, and the same line along its surface is used.

MEASUREMENT OF VERTEBRAL ROTATION. The Perdriolle method and the Nash-Moe method are the two most common means of assessing vertebral rotation on a plain frontal radiograph. The Perdriolle method uses a transparent "torsionmeter" that is overlaid on the radiograph (Fig. 11–19).[346] The edges of the curve's apical vertebra and its rotated pedicle constitute the landmarks. This method has been found accurate for measuring rotations that are less than 30 degrees.[336] However, once the scoliotic spine has undergone instrumentation, the landmarks of the apical vertebra may become obstructed by the shadows of the rods or hooks, making accurate measurements nearly impossible.[373]

In the Nash-Moe method, the relationship of the pedicle to the center of the vertebral body is observed on the AP radiographs, and the rotation is divided into five grades: zero (0) when both pedicles are symmetric, grade I when the convex pedicle has moved away from the side of the vertebral body, grade III when the convex pedicle is in the center of the vertebral body, grade II when rotation is between grades I and III, and grade IV when the convex pedicle has moved past the midline (Fig. 11–20).[317]

Computed tomography (CT) can also be used to assess vertebral rotation.[91,188,494] Although CT is more expensive, its

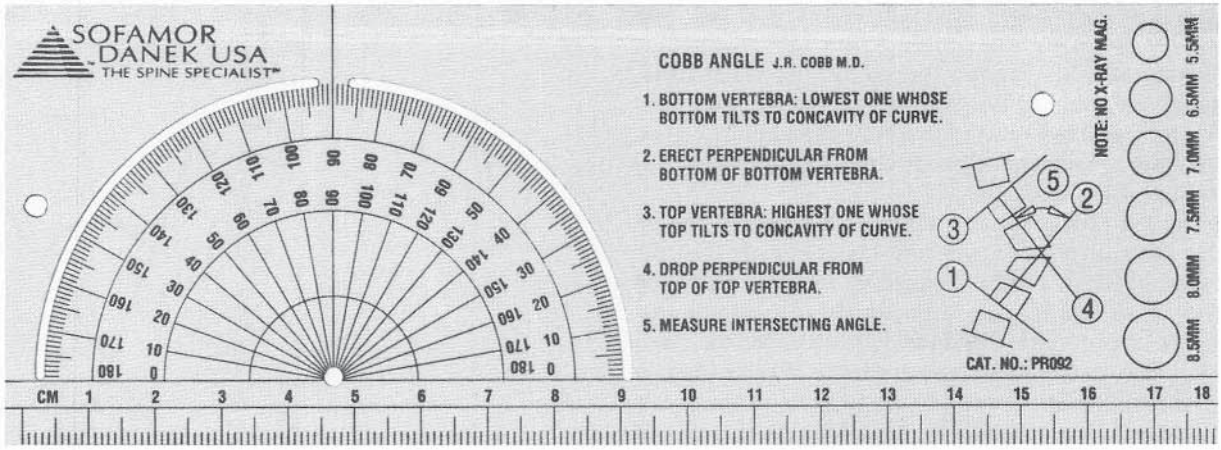


FIGURE 11–18 A transparent plastic goniometer is used to draw the lines and measure the angles. (Courtesy of Medtronic Sofamor Danek, Memphis, TN.)

accuracy is better than the Nash-Moe method. For example, vertebrae of Nash-Moe grade 0 have been found to have up to 11 degrees of rotation when measured using CT.[180]

MEASUREMENT OF KYPHOSIS AND LORDOSIS ON LATERAL RADIOGRAPHS. The end vertebrae are the last vertebrae that are maximally tilted into the concavity of the curve. In the thoracic area, the upper end vertebra usually is T3 or T4 and the lower end vertebra is T12. Perpendicular lines are drawn to the inferior and superior end-plates. The angle formed between the two perpendicular lines represents the degree of thoracic kyphosis. Normal thoracic kyphosis ranges from 20 to 45 degrees. There is no kyphosis or lordosis at the thoracolumbar junction (between T11 and L1–2).[37] Lumbar lordosis usually begins at L1–2 and gradually increases caudally to the sacrum. To measure lumbar lordosis, the lower end vertebra for the thoracic curve becomes the upper end vertebra. The lower end vertebra for the measurement of lumbar lordosis usually is L5 or S1. Although recent attempts have been made to determine normal lumbar lordosis,[78,446] currently there is no consensus. Reported normal values range from 50 to 65 degrees.[78,446] The thoracic and lumbar regional alignment is very similar between adolescents and adults.[453]

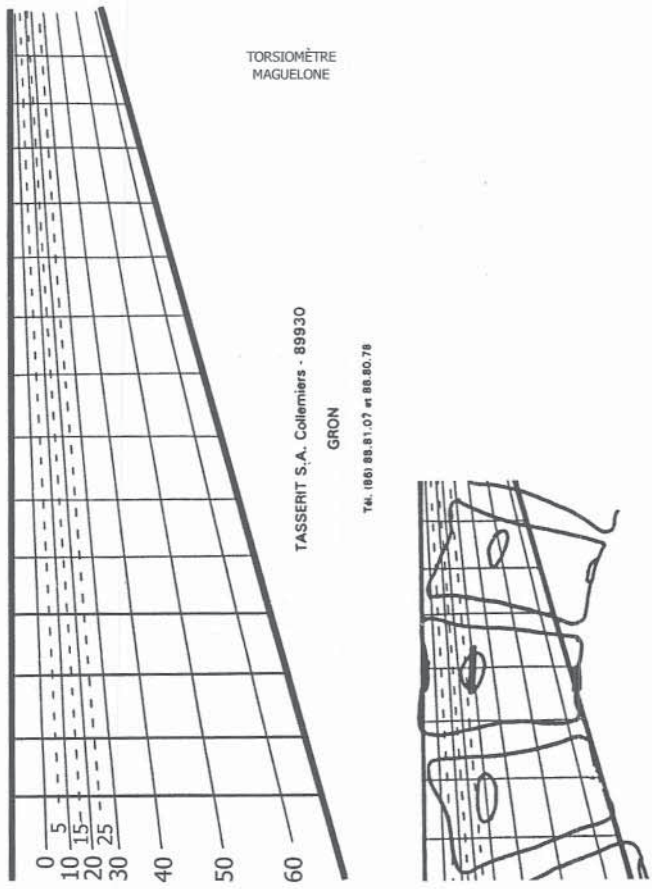


FIGURE 11–19 *Left:* The Perdriolle torsionmeter, a clear template. *Right:* The torsionmeter's outer margins are aligned over the vertebra's lateral borders. The line intersecting the center of the pedicle shadow (convex side) estimates the amount of spinal rotation. (From Richards BS: Measurement error in assessment of vertebral rotation using the Perdriolle torsionmeter. Spine 1992;17[5]:513–517.)

Surface Imaging. In an effort to decrease the amount of radiation exposure during the course of scoliosis management, techniques have been developed to assess body surface changes in scoliosis patients. The goals of surface imaging are to develop methods that appropriately identify scoliosis, monitor curve progression, and provide information that can be used for treatment decisions. However, natural history data and, in most cases, treatment decisions are based on Cobb angle radiographic measurements made from upright radiographs.

For surface imaging systems to be useful, they must demonstrate some consistency with Cobb angle measurements. Moire topography, raster-stereophotography, and the Integrated Shape Imaging System (ISIS) are three of the sophisticated techniques that have evolved, with all of them using computer analysis of digitized topographic information (Fig. 11–21). The presence, level, and side of scoliosis curvature have been documented nicely by these techniques in patients with standard rotation; however, it has not been possible to determine the magnitude of the scoliosis with sufficient accuracy from topography for it to be used for most clinical purposes.[92,391,427,445,491] These techniques continue to be investigated in an effort to determine their appropriate role in the management of scoliosis.

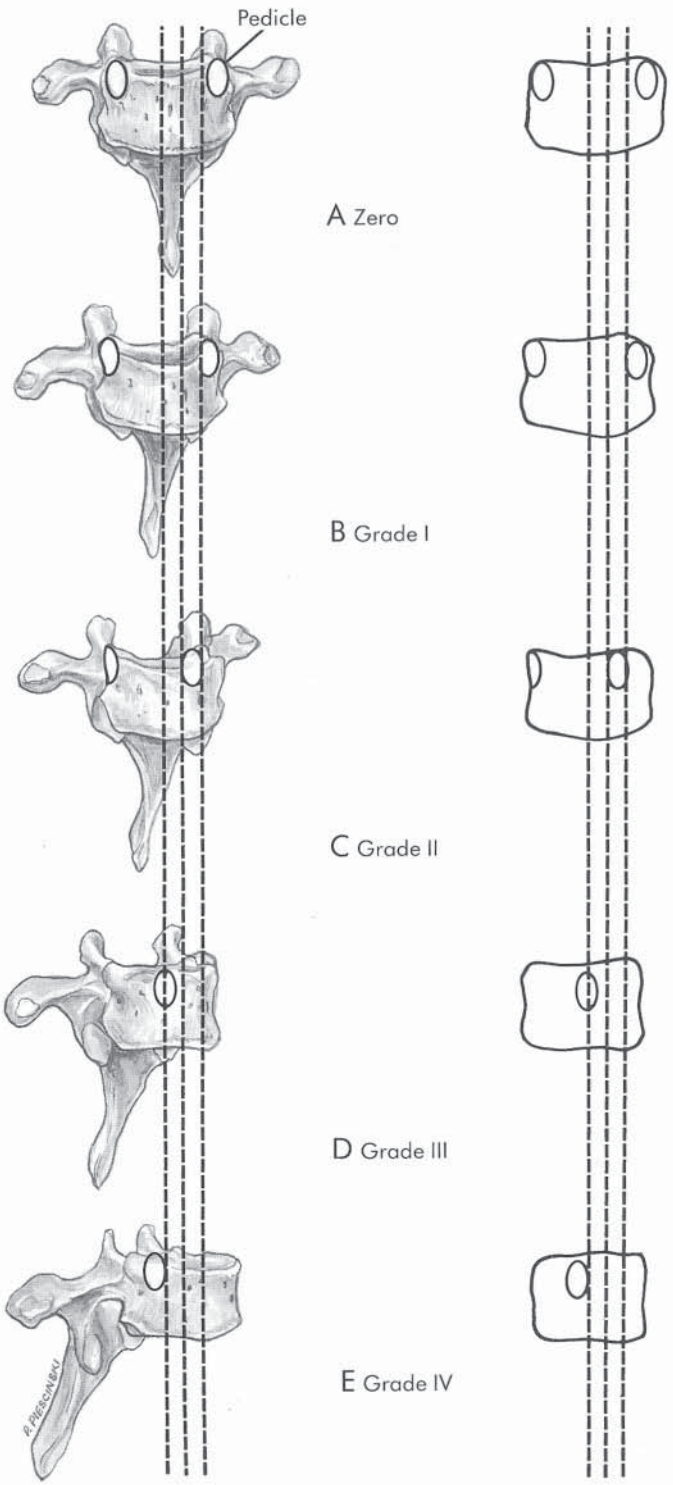


FIGURE 11–20 The Nash–Moe method of assessing vertebral rotation. Grade 0: The pedicles are symmetric and equidistant from the sides of the vertebral body. Grade I: The pedicles in the convexity of the curve have moved from the lateral border of the vertebral body. Grade II: The pedicle on the convex side is intermediate between grades I and III. Grade III: The pedicle on the convex side is in the center of the vertebral body. Grade IV: The pedicle on the convex side has rotated past the midline. (Redrawn from Nash C, Moe J: A study of vertebral rotation. J Bone Joint Surg 1963;51-A:223.)

Magnetic Resonance Imaging. By providing a clear anatomic picture of abnormalities that can occur within the spinal canal, MRI can be an extremely valuable tool in the assessment of scoliosis. Syringomyelia, Arnold-Chiari malformations, abnormalities in the brain stem, hydromyelia, spinal cord tumors, spinal cord tethering, and diastematomyelia have all been identified in individuals previously thought to have idiopathic scoliosis.[144,244,397,405,484] However, because these abnormalities are rare, the use of MRI as part of routine screening programs is impractical and cost-prohibitive. MRI usually is reserved for patients whose idiopathic scoliosis presentation appears atypical.

First, though, the typical patient must be defined. This is a girl who presents during adolescence, is asymptomatic with no neurologic deficits, and has a right thoracic curve that follows one of several defined curve patterns.[4] Although atypical idiopathic scoliosis has never been specifically defined, it generally includes those patients with neck pain and headache (particularly with exertion) and abnormal neurologic findings such as ataxia, weakness, and progressive foot deformities; those patients with unusually rapid curve progression; or those patients requiring surgery who have left thoracic curves or asymmetric abdominal reflexes. Curves exceeding 70 degrees do not increase the likelihood of finding a spinal cord anomaly.[326] Routine preoperative MRI is probably not indicated in typical adolescent idiopathic scoliosis if the neurologic examination is normal.[326,405,484]

Computed Tomography. Although CT may clearly demonstrate congenital abnormalities in the spine, it is rarely,

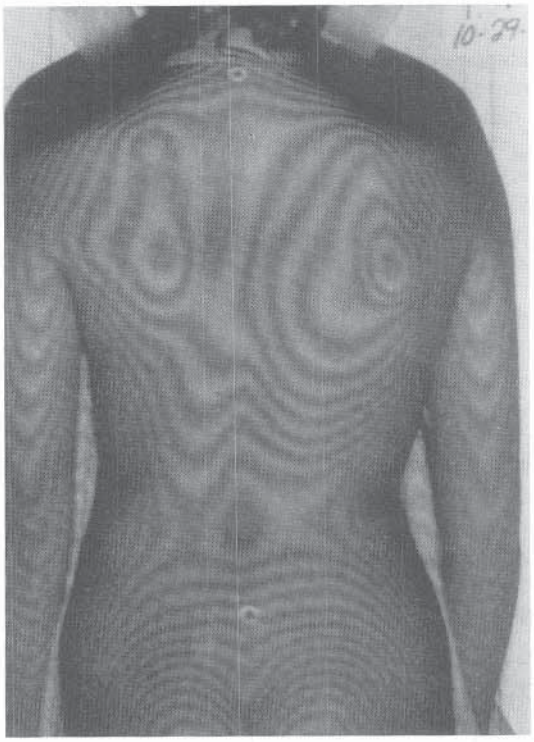


FIGURE 11–21 Moire topographic photograph. This surface imaging system produces an image that can be read in the same way as contour lines on a map. (From Stokes IA, Moreland MS: Concordance of back surface asymmetry and spine shape in idiopathic scoliosis. Spine 1989;14[7]:73.)

if ever, needed in the routine assessment of idiopathic scoliosis. It remains a useful tool postoperatively (particularly with three-dimensional reconstruction) in assessing bone fusion mass if pseudarthrosis is suspected. Pedicle screw placement can be verified and changes in spinal rotation can be documented.[91,167,238,247,493,494] In addition, CT-myelography affords improved evaluation of the spinal cord when retained metal implants limit the effectiveness of MRI.

GENERAL TREATMENT CONCEPTS

Most adolescents with idiopathic scoliosis will not require treatment, because of the low probability that their curves will progress.[4,259] Treatment therefore is warranted only for those patients whose scoliotic curves are at substantial risk of worsening over time or for patients with severe curves at presentation. A clear understanding of the risk factors discussed earlier in the Natural History section is useful in determining which patient is in need of treatment, regardless of whether the individual is skeletally immature or mature.

In selecting treatment, the physician must consider the adolescent's remaining growth potential, the severity of the curve at the time of detection, and the pattern and location of the scoliosis. The cosmetic appearance of the individual and any social factors that may have an impact on treatment also enter into the decision-making process. The treatment choices available are observation, nonsurgical intervention, and surgical intervention. Table 11–3 outlines general treatment guidelines.

OBSERVATION

In general, no treatment is needed for curves that are less than 25 degrees, regardless of the patient's maturity. Follow-up examinations are necessary, with the interval between visits depending on the patient's maturity and the size of the curve. For example, a premenarchal Risser 0 adolescent with an initial curve measuring 24 degrees should undergo follow-up examinations every 3 to 4 months. A brace may be needed if the curve progresses further. For more skeletally mature patients (Risser 3 or greater), longer intervals between visits (i.e., 6 months) are appropriate because curve progression usually occurs at a slower rate, if at all. Clearly, predetermined guidelines do not apply to all cases, and follow-up must be individualized.

The magnitude of the patient's curve at the time of initial presentation also helps to determine the frequency of follow-up visits. In general, for growing children with small curves (less than 20 degrees), the next follow-up evaluation should be approximately 6 months later. If the curve is between 20 and 30 degrees, radiographs should be obtained 3 to 4 months later because treatment may be necessary if curve progression of 5 degrees or more occurs. For those patients whose curves do not progress, observation continues and the interval between visits gradually lengthens as maturity approaches.

There remains some debate as to what truly constitutes curve progression. Traditionally, an increase in curve size greater than 5 to 6 degrees has been taken as representing progression. However, a 7- to 10-degree change in measurement is more accurate if a 95 percent confidence level is used to determine true progression.[67,309] This should be taken into consideration when deciding whether the measured change in the patient's scoliosis warrants either nonsurgical or surgical intervention. Nevertheless, the reader should be aware that throughout the literature, a 5- to 6-degree measured change is considered indicative of curve progression. Not all progressive curves exceeding 30 degrees require treatment; the decision depends on the adolescent's maturity and the size of the curve.

It is imperative that physicians treating patients with scoliosis know which individual warrants treatment. Table 11–3 provides general treatment guidelines. Actively growing adolescents (Risser $\leq$ 2) with curves between 30 and 45 degrees should be started on brace therapy at the time of their initial visit.[387] In the very immature patient (Risser 0 and premenarchal if female) with a curve exceeding 25 degrees, bracing should be started immediately.[261,387] In most cases, growing adolescents with curves exceeding 45 to 50 degrees require operative stabilization, as other forms of treatment are ineffective in controlling or correcting the scoliosis. Skeletally mature individuals with curves exceeding 50 to 55 degrees also are at risk for continued curve progression and therefore should be considered for surgical treatment.[462] Possible exceptions include patients with well-balanced double curves less than 60 degrees whose clinical appearances are acceptable to them. Continued observation would be necessary to establish further progression of their scoliosis, which would necessitate surgery.

NONSURGICAL TREATMENT

To be considered effective, nonsurgical treatment must prevent curve progression in those who are most at risk (curves of 25 to 45 degrees in Risser 0 or 1 patients), be of benefit with all curve patterns, result in an acceptable cosmetic appearance at the end of treatment, and reduce the need for surgery. In other words, nonsurgical treatment must improve the patient's outcome when compared with the expected natural history. Over the years a great deal of experience has been gained with various forms of nonsurgical treatment, some of which have proved effective (e.g., bracing) and others of which have not demonstrated any

TABLE 11–3 **Guidelines for Treating Patients with Idiopathic Scoliosis**

Curve Magnitude	Risser Sign Grade 0/Premenarchal	Risser Sign Grades 1 or 2	Risser Sign Grades 3, 4, or 5
<25 degrees	Observation	Observation	Observation
30–45 degrees	Brace therapy (begin when curve exceeds 25 degrees)	Brace therapy	Observation
>45 degrees	Surgery	Surgery	Surgery (when curve exceeds 50 degrees)

beneficial effect (e.g., electrical stimulation, exercises, biofeedback).

Orthotic (Brace) Treatment. Historically, Ambrose Pare is credited with being the first to use metal braces, in the form of armor, to treat patients with scoliosis. Since then, various types of braces and casts have been advocated, such as the suspensory plaster cast of Sayre and the hinge or turnbuckle cast of Hibbs and Risser. In 1946 the Milwaukee brace was developed to replace the use of postoperative plaster immobilization. Later the brace was used as a nonoperative method of treatment where passive, active, and distraction forces were thought to be necessary to prevent curve progression. Subsequent studies have shown that the corrective forces of a brace are passive in nature and that the predominant corrective component is the transverse loading of the spine through the use of corrective pads.[12,74,341,498] In the 1960s, thermoplastics were introduced into orthotic manufacturing, leading to the thoracolumbosacral orthoses (TLSOs) of today.

INDICATIONS FOR BRACE TREATMENT. Brace treatment is restricted to immature children in an attempt to prevent curve progression during further skeletal growth. Bracing is indicated in growing adolescents (Risser 0, 1, or 2) who, on presentation, have curves in the range of 30 to 45 degrees or who have had documented progression exceeding 5 degrees in curves that initially measured 20 to 30 degrees. These patients should have deformities that are considered cosmetically acceptable and the patients should be realistically willing to wear the brace the prescribed amount of time. Low-profile braces (TSLOs) are the most commonly used orthoses today, but their use is restricted to patients whose curve apex is at T7 or below. Fortunately, this is the case in most curve patterns in adolescents with idiopathic scoliosis.

CONTRAINDICATIONS TO BRACE TREATMENT. There are several contraindications to brace treatment. First, most studies concur that larger curves (exceeding 45 degrees) in the growing adolescent cannot be effectively controlled by a brace and that these patients need surgical treatment. Even if progression could be controlled by a brace, the cosmetic appearance associated with these larger curves is often felt to be unacceptable because of excessive trunk shift and rib prominence. There is an exception, though, to this general rule of avoiding bracing in patients with larger curves. Very immature adolescents who have not yet reached their peak height velocity and who have large curves (approximately 50 degrees) may benefit from a brace in an effort to delay progression until further maturity is reached. Bracing in these patients may avoid the need for additional anterior spinal fusion to prevent the crankshaft phenomenon.

The second contraindication to bracing is for patients who find the wearing of an orthosis to be emotionally intolerable. Appropriate psychological counseling, though, may result in acceptance of a brace by the adolescent. The third contraindication is extreme thoracic hypokyphosis. In these cases, normal positioning of the pads within the brace could exacerbate the rib deformity. In cases where hypokyphosis is less than or equal to 20 degrees, corrective pads should be lateralized to eliminate any anteriorly directed derotation forces. Fourth, bracing will not benefit the skeletally mature adolescent (Risser 4 or 5 and, if female, 2 years postmenarchal). Finally, a relative contraindication to bracing is a high thoracic or cervicothoracic curve, because this deformity ordinarily does not respond to orthotic treatment.

COMPARISON OF ORTHOSES. Numerous reports in the literature attest to the effectiveness of brace treatment.* In most of these studies, bracing was considered effective if the curve being treated remained within 5 to 6 degrees of its original magnitude on completion of treatment. Some of these studies included low-risk patients (Risser 3 to 5, curves less than 20 degrees), patients still undergoing treatment, patients who had had previous treatment, and children less than 10 years old. In some studies, patients may have been eliminated from the patient population because of noncompliance. All of these factors make comparisons between studies difficult, particularly when one is trying to assess the effectiveness of bracing in patients most at risk (Risser 0 or 1, premenarchal girls, 25- to 45-degree curves). Some of the more recent literature, however, has been more consistent in focusing on this population at greatest risk.[6,209,261,316,360] These studies and a recent meta-analysis of the bracing literature[387] strongly reinforce the idea that bracing remains effective in controlling curve progression.

Numerous orthoses are available today. Most are named after their place of origin; some examples include the Milwaukee brace,[261] the Boston brace,[120,166] the Wilmington brace,[6,28] and the Charleston brace.[360] All of these braces have been reported to be effective in preventing curve progression. Before deciding which brace to use, the orthopaedist should be familiar with the advantages and disadvantages of the various orthoses.

The Milwaukee brace, introduced by Blount, Schmidt, and Bidwell in 1946, was the original modern design. The device consisted of three main components: a pelvic girdle, a suprastructure, and lateral pads (Fig. 11–22). Over time, there have been some design modifications to this brace. In the current model the pelvic girdle is made of thermoplastic material and is created from a positive mold of the patient's pelvis. The suprastructure consists of one anterior and two widely separated posterior uprights, plus a cervical ring with a throat mold and occipital piece. In many cases a low-profile, over-the-shoulder structure may be used in place of the more standard neck-ring design. The lateral pads apply pressure to the apical vertebrae.

The Milwaukee brace has been shown to be effective in controlling curve progression in idiopathic scoliosis, with the largest recent series reported by Lonstein and Winter.[261] The authors found that patients with curves between 20 and 39 degrees who used the Milwaukee brace were less likely to have curve progression exceeding 5 degrees when compared with a similar patient population that received no treatment. However, as with all braces, the success achieved with the Milwaukee brace has not been universal.[322] Today, when there is a strong emphasis on self-image, use of the Milwaukee brace has decreased greatly and has largely been replaced by the use of equally effective lower-profile braces. Low-profile TLSOs, such as the Boston brace, Wilmington brace, and Miami brace,[286] can often be hidden under loose shirts

*See references 6, 12, 26, 27, 74, 119, 120, 131, 133, 158, 209, 212, 231, 261, 303, 316, 322, 334, 360, 361, 387, 449, 467, 476, 483.

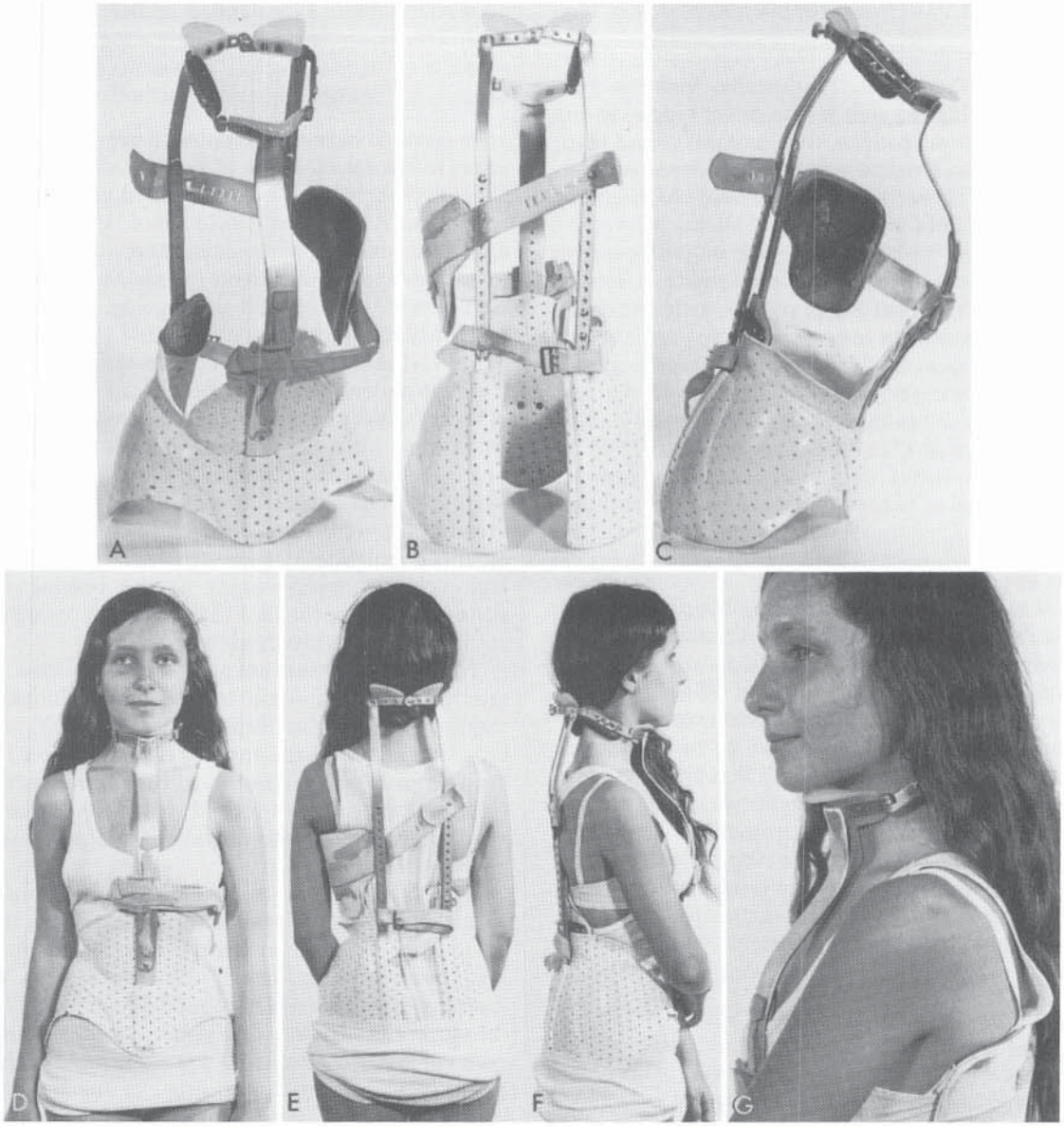


FIGURE 11–22 Modern design of the Milwaukee brace. **A** to **C,** Anterior, posterior, and lateral views of the orthosis (see text for discussion). **D** to **G,** An adolescent girl wearing the Milwaukee brace. Note the following: (1) The *pelvic girdle* is made of a thermoplastic material called Orthoplast. It is self-hinging, perforated for ventilation, lightweight, and waterproof. It can be remolded by using a heat gun to relieve areas of skin pressure and to accommodate the increasing size of the pelvis. Deterioration of the Orthoplast pelvic girdle is prevented by daily washing with soap and water. (2) The throat mold is snugly approximated, and there is complete lack of pressure against the mandible. (3) The occipital pad fits the lower occiput accurately, following its contour. (4) The uprights fit closely to the torso, allowing room for deep breathing, lateral shift, pelvic tilt, and abdominal exercises.

or sweaters, providing the adolescent with a more acceptable alternative.

The Boston brace was introduced in 1971 by Hall and Miller.[166] Its design consists of a prefabricated, symmetric thoracolumbar-pelvic module with built-in lumbar flexion and areas of relief opposite areas of pressure (Fig. 11–23). Braces are individually constructed by the orthotist based on a blueprint created from the patient's full-length radiograph. This brace is the most commonly used TLSO today and, like the Milwaukee brace, has been shown to be effective in controlling curve progression.[120,209,303,334,467] It has been reported that the Boston brace exerts derotational forces on the scoliosis; however, a recent study found no lasting improvement in derotation of the spine.[467] The brace is effective in treating either single- or double-curve patterns in which the apex of the most cephalad curve is located at T7 or below. To control a thoracic curve whose apex is higher requires that a suprastructure be fitted to the brace. Rarely is this actually done.

The Wilmington brace was described in 1980.[62] It is custom-made from a positive mold of the patient's torso in which the scoliosis is maximally corrected in a Risser- or Cotrel-type cast. The indications for the Wilmington brace are the same as those for the Boston brace. It also has the same limitations (i.e., it is not effective in curves with a apex above T7–8). This brace has not enjoyed the popularity

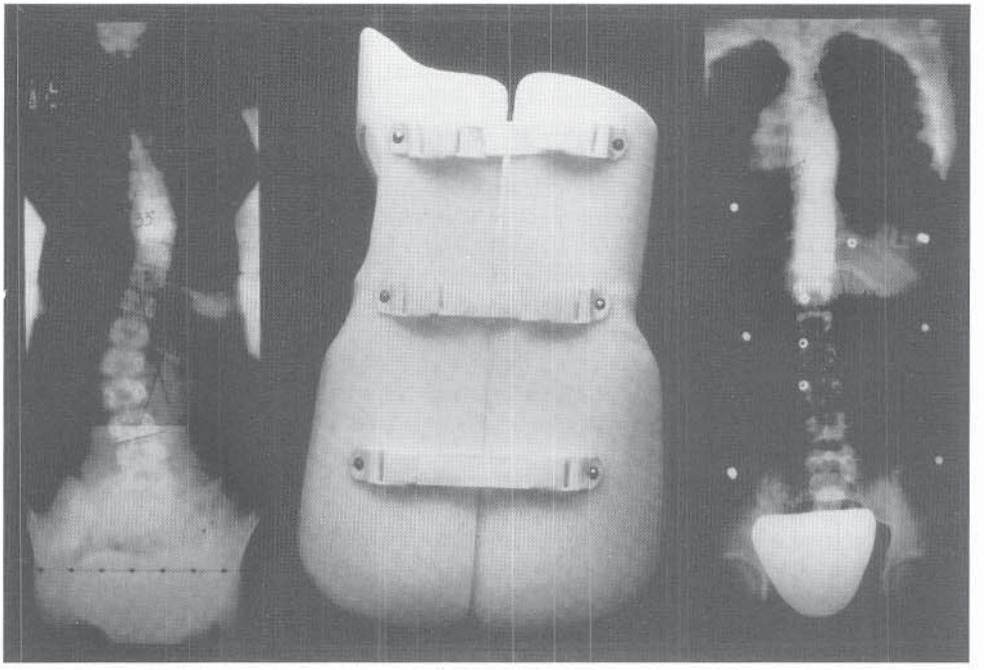


FIGURE 11–23 The Boston brace. This orthosis is a prefabricated, symmetric thoracolumbar-pelvic module with built-in lumbar flexion and areas of relief opposite areas of pressure. Each brace is individually modified based on the patient's full-length spine radiograph. This brace is the most common thoracolumbosacral orthosis used today.

of the Boston brace, although it continues to be used by several institutions.[6]

The number of hours each day that the brace needs to be worn remains uncertain. Originally, 20 to 22 hours per day was advocated for the Milwaukee brace in immature adolescents with progressive curves. This same recommendation was made for the lower-profile TLSOs. Faced with this situation, adolescents understandably experienced some emotional distress, and poor compliance with brace wear was not uncommon.[103] As a result, the idea of part-time use of braces evolved, with the goal for daily use being approximately 16 hours. With this program, most adolescents choose not to wear the orthosis during school hours. Over the past 13 years, there have been several studies reporting that part-time use appears to be as effective as full-time wear in controlling curve progression.[6,120,158]

The Charleston brace was developed based on the concept that part-time use may be effective.[129,360,361] This brace holds the patient in maximum side-bending correction (Fig. 11–24) and is worn at night only, for 8 to 10 hours. The side-bending force that the brace exerts does not allow its use in the upright position, thus making wear feasible only when the patient is recumbent. The main appeal of this brace is the limited number of hours of daily use, all of which are accomplished during sleep.

Despite preliminary studies which reported that the Charleston brace was as effective as the Milwaukee and Boston braces,[129,361] there remain some skeptics who doubt that such a limited amount of time spent in a brace can successfully control curve progression. One long-term report suggests that the Charleston brace exerts a favorable effect on the natural history of adolescent idiopathic scoliosis and that its continued use is justified.[360] However, in a comparison of the Boston brace with the Charleston brace, Katz and associates found that in patients with curves between 36 and 45 degrees, 83 percent treated with a Charleston brace experienced curve progression of more than 5 degrees, compared with 43 percent of patients treated with a Boston brace.[209] Overall, the Boston brace was found to be more effective than the Charleston brace in preventing curve progression and in avoiding the need for surgery. When choosing between the two braces, the authors concluded that the Charleston brace should be reserved for single lumbar or thoracolumbar curves less than 35 degrees.

BRACE TREATMENT PROTOCOLS. In an effort to form a consensus from the literature on the effectiveness of bracing (including

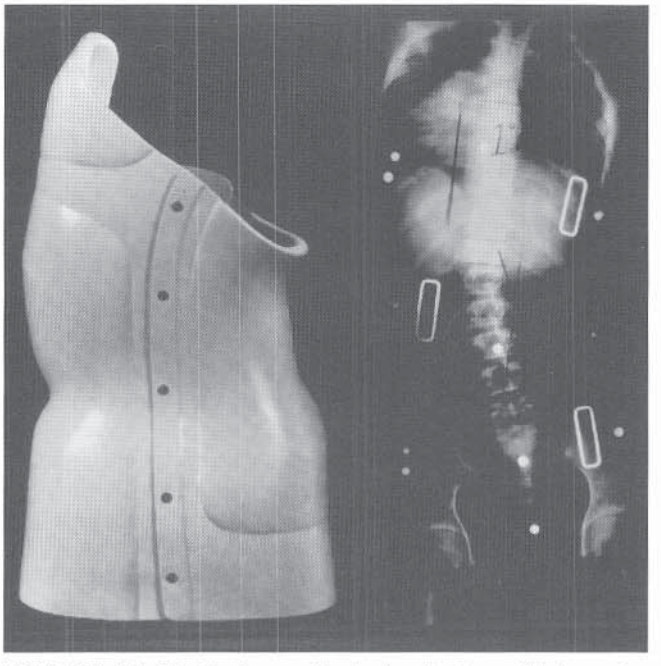


FIGURE 11–24 The Charleston nighttime bending brace. This brace tries to maintain maximum side-bending correction. It is worn at night only for 8 to 10 hours.

whether part-time bracing controls curve progression as effectively as full-time bracing), the Prevalence and Natural History Committee of the Scoliosis Research Society conducted a meta-analysis on more than 1,900 patients from 20 studies.[387] They concluded that bracing (with TLSOs or the Milwaukee orthosis) is indeed effective in controlling curve progression in idiopathic scoliosis and that full-time bracing (23 hours per day) is more effective than part-time bracing (8 to 16 hours per day). The latter finding is further supported by a recent study in which patients' use of the Boston brace was divided into three groups: noncompliant (less than 12 hours per day), part-time (12 to 18 hours per day), and full-time (18 to 23 hours per day) wear.[300] At the conclusion of treatment, the greatest success was achieved by the full-time group and the poorest results occurred in the noncompliant group.

The issue of compliance with brace wear remains very subjective, being dependent on the child's or family's report. Better objective means of documenting compliance would be useful in determining the effectiveness of "full-time" bracing in comparison to "part-time" bracing. From the information currently available, it appears that the highest success rate from a bracing program can be expected when a patient uses the orthosis 20 hours or more per day. However, it also appears that part-time bracing still exerts a favorable influence on the natural history of adolescent idiopathic scoliosis.

When brace treatment has been chosen for the patient, certain general guidelines should be followed. Once the brace has been constructed and fitted to the patient by the orthotist, the patient should work up to the prescribed number of hours each day. After 2 to 4 weeks, the adolescent should return to the orthopaedist's office for the initial brace evaluation. At that time, any problems with the wearing of the brace (e.g., intolerable pressure points) will have been identified by the patient and can be addressed by the orthotist. Equally important, an in-brace radiograph should be obtained to verify the amount of curve correction that is being achieved. With the Boston brace, a minimum of 40 to 50 percent of curve correction should be obtained in the brace.[209] With the Charleston brace, the amount of in-brace correction should approach 90 percent for flexible curves and 70 percent for rigid curves for it to be effective. Regardless of the type of brace used, insufficient in-brace curve correction leads to an unsatisfactory outcome, one that differs little from the expected natural history.[322] If proper correction cannot be obtained with brace use, orthotic treatment should be discontinued.

During brace management, follow-up visits are scheduled at 4-month intervals for the rapidly growing adolescent with a large curve. The interval may be extended to 6 months for those patients nearing maturity whose curves have shown no recent changes. During these visits, a single standing PA thoracolumbar radiograph is obtained with the patient out of the brace. Curve progression, if it has occurred, is readily identifiable, and appropriate adjustments to the treatment program are made. Some physicians obtain the radiographs with the patient wearing the brace in order to show the brace's effect on both the curve and spinal balance.[254] However, recognition of curve progression may be missed if the patient is imaged wearing the brace.

With female patients, if the brace has been successful in controlling curve progression, plans can be made to discontinue treatment when the girl is approximately 18 to 24 months postmenarchal and Risser 4, and when no further increase in her height has occurred (Fig. 11–25). Rather than tapering use of the brace, we discontinue its use completely at that time. In male patients, curves exceeding 25 degrees have a tendency to progress even when Risser 4 maturity has been reached.[208] Therefore, in boys, bracing may need to be continued until Risser sign 5 is achieved. Frequently this will not occur before the later teenage years, which makes brace-wear compliance truly a challenge to all involved.

Electrical Stimulation. Electrical stimulation was used as an alternative to bracing in the early 1980s. Surface muscle stimulators were placed over the muscles on the convex side of the scoliotic curve and were activated for approximately 8 to 10 hours each night. In Canada, electrode stimulators were actually implanted in the paraspinal muscles. Although some preliminary success has been reported with transcutaneous stimulation,[10] most studies found that this form of treatment did nothing to favorably alter the natural history of scoliosis.[38,112,316,327] Today, electrical stimulation is no longer considered a useful method in the management of idiopathic scoliosis.

Physical Therapy/Biofeedback. The purposes of spinal exercise are to improve posture, to increase the strength of the trunk muscles, and to maintain spinal flexibility. Although muscle conditioning is beneficial to a patient's overall well-being, there is no evidence to support the concept that exercises or physical therapy programs are helpful in controlling or improving scoliosis. Likewise, spinal manipulations and biofeedback have not been shown to alter the natural history of scoliosis.

SURGICAL PROTOCOLS

The primary goals of surgical intervention in the treatment of scoliosis are to reduce the magnitude of the deformity, to obtain fusion in order to prevent future curve progression, and to do so safely. Operative treatment should result in a well-balanced spine in which the patient's head, shoulders, and trunk are centered over the pelvis. Ideally, when this is accomplished, a significant amount of curve correction will be achieved.

Current instrumentation systems are able to exert stronger corrective forces on scoliotic spines than was possible 15 to 20 years ago with the use of Harrington instrumentation. They are also more complicated to use and require a significant amount of training. These newer systems, which include the Cotrel-Dubousset (CD) instrumentation, Texas Scottish Rite Hospital (TSRH) instrumentation, and Isola instrumentation, became popular in the mid-1980s and remain so today. Each instrumentation system allows the scoliosis surgeon to achieve increased curve correction, improved sagittal contouring, brace-free postoperative mobilization, and MRI compatibility (with the availability of titanium components).

The expansion of technology in the 1990s resulted in the availability of numerous other systems that provide similar advantages (e.g., AO Universal Spine system, Moss Miami

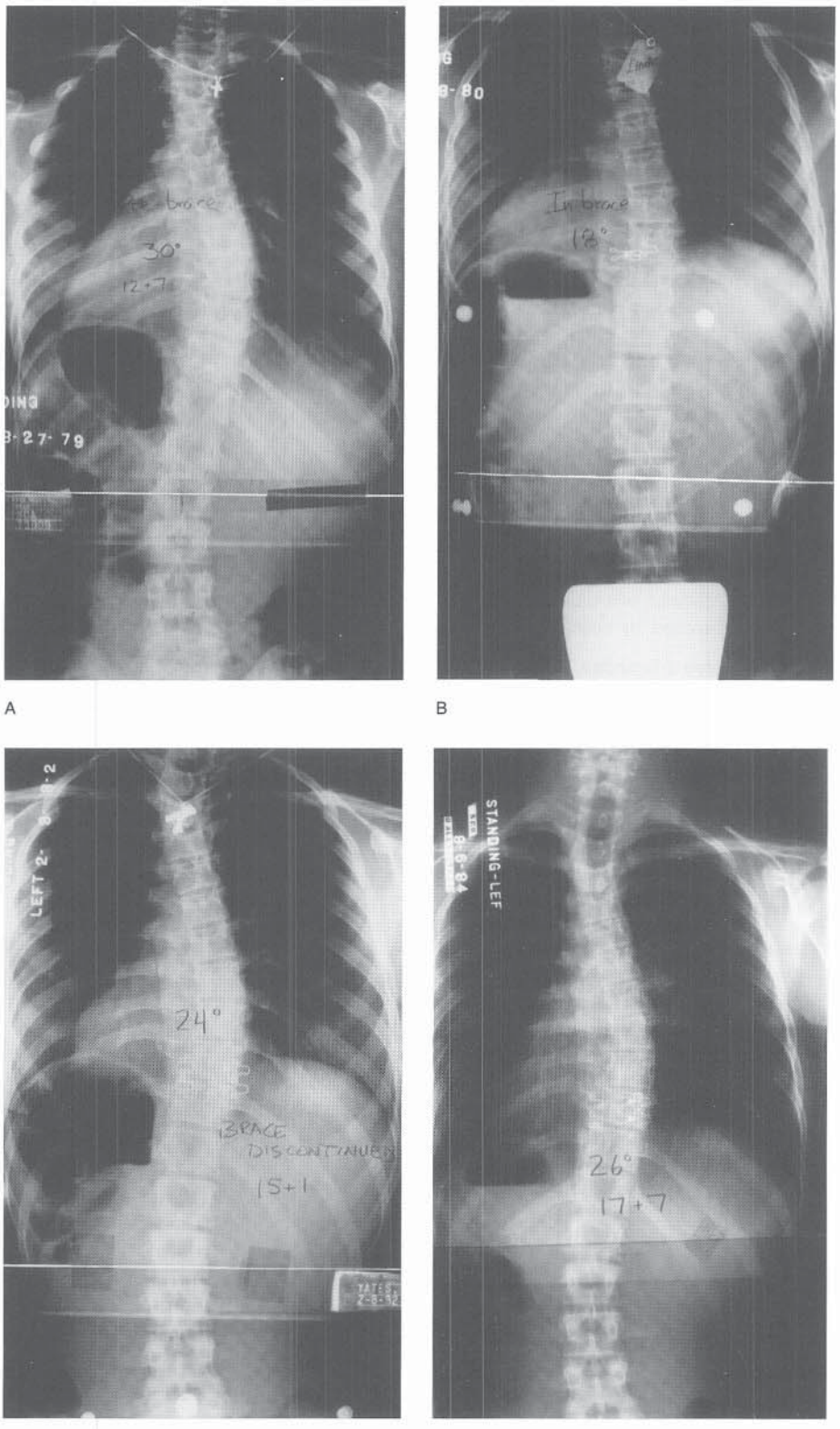


FIGURE 11–25 Radiographic findings with brace wear in a premenarchal girl age 12 years 7 months. **A,** Initially she had a 30-degree thoracic curve. Her Risser sign was 0. **B,** Treatment in a Boston brace was begun, with in-brace correction to 18 degrees. **C,** Brace wear was continued until the patient was 2 years postmenarchal and had a Risser sign 4. **D,** Two and one-half years later, the curve remained stable at 26 degrees.

instrumentation, Synergy spine system, CD Horizon, Kaneda scoliosis system). Familiarity with one or more of these systems, including their limitations, is helpful when planning surgical treatment for the various curve patterns seen with idiopathic scoliosis.

Indications for Surgery. Although various factors enter into the surgical decision making, the magnitude of the scoliotic curve remains the primary factor. Curves less than 30 degrees at skeletal maturity are unlikely to progress, regardless of curve pattern, and do not require surgery. Tho-

racic curves and double major curves that exceed 50 degrees at skeletal maturity have a significant probability of worsening over time and nearly always warrant operative intervention.[462] Thoracolumbar and lumbar curves of lesser magnitude, when associated with marked apical rotation or translatory shift, also have a propensity to worsen over time in mature patients. In these cases, surgery should be considered when the curves exceed 40 to 45 degrees.

In addition to curve magnitude, the patient's appearance (as perceived by the child, the family, and the surgeon) factors into surgical decision making. Patients and their families usually are concerned most about this aspect of the deformity. The patient's spinal balance may be decompensated, with the thorax shifted noticeably away from the midline; the rib prominence may be severe due to excessive rotation; and the shoulders and hips may appear uneven.

It is uncommon that back pain by itself serves as an indication for scoliosis surgery. Nearly 30 percent of patients with scoliosis describe associated back pain, but in less than 10 percent of these symptomatic patients will a definite cause of the discomfort be found.[366] The treating orthopaedist, therefore, should not assume that the patient's discomfort will be remedied by spinal fusion.

Preoperative Planning. Preoperative planning must take into consideration the patient's curve pattern and spinal balance, preoperative curve flexibility, neurologic status, rib deformities, physical maturity and future growth potential, and other surgery-related needs (transfusion requirements, bone grafting, spinal cord monitoring, and postoperative pain management). The surgeon's selection of instrumentation depends on personal experience, the availability of the various systems, and the choice of anterior or posterior instrumentation.

CURVE PATTERNS. Knowledge of the various curve patterns seen in idiopathic scoliosis is needed for proper preoperative planning, as this will influence the selection of instrumentation, the length of required spinal fusion, and the decision to pursue an anterior or posterior approach. The King-Moe classification system, introduced in 1983, described five different curve patterns (Fig. 11–26).[221] This classification system continues to be used regularly; however, its reliability and reproducibility have recently been questioned.[90,236]

The King type curve patterns are described in Table 11–4. King type II and III curves appear to be most common and constitute the majority of curves treated by surgery. Other curve patterns that are not described by the King-Moe classification system include single thoracolumbar, single lumbar, and double major S-shaped curves. In double major curves the thoracic and lumbar components are similar in size, are structural, and have limited flexibility (Fig. 11–27). It is important to differentiate double major curves from King type II curves.

The "stable vertebra" refers to the inferior vertebra in the thoracic curve that is most closely bisected by the center

TABLE 11–4 **King-Moe Classification System of Curve Patterns**

King type I and II curves: S-shaped curves in which both the thoracic curve and the lumbar curve cross the midline.
King type I curve: Lumbar curve is larger than the thoracic curve by 3 degrees or more. On supine bending radiographs, the thoracic curve is more flexible than the lumbar curve.
King type II curve: Thoracic curve is equal to or larger than the lumbar curve. On side-bending radiographs, the lumbar curve is more flexible than the thoracic curve.
King type III curve: A thoracic curve in which the lower level does not cross the midline.
King type IV curve: A long thoracic curve in which L5 is centered over the sacrum but L4 tilts into the long thoracic curve.
King type V curve: A double thoracic curve in which T1 is tilted into the convexity of the upper curve (the upper curve is structural on side bending).

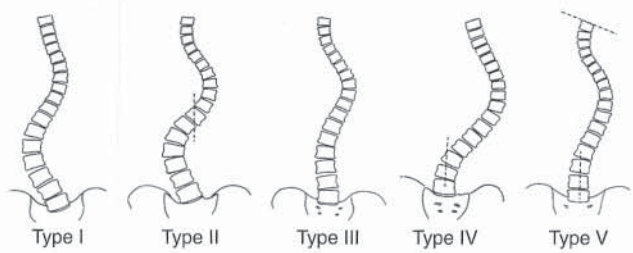


FIGURE 11–26 Diagrammatic representation of the King-Moe classification of idiopathic scoliosis. Five distinct curve patterns were described. This classification continues to be used regularly, although its reliability and reproducibility have been questioned. (From King HA, Moe JH, Bradford DS, et al: The selection of fusion levels in thoracic idiopathic scoliosis. J Bone Joint Surg 1983;65-A:1302.)

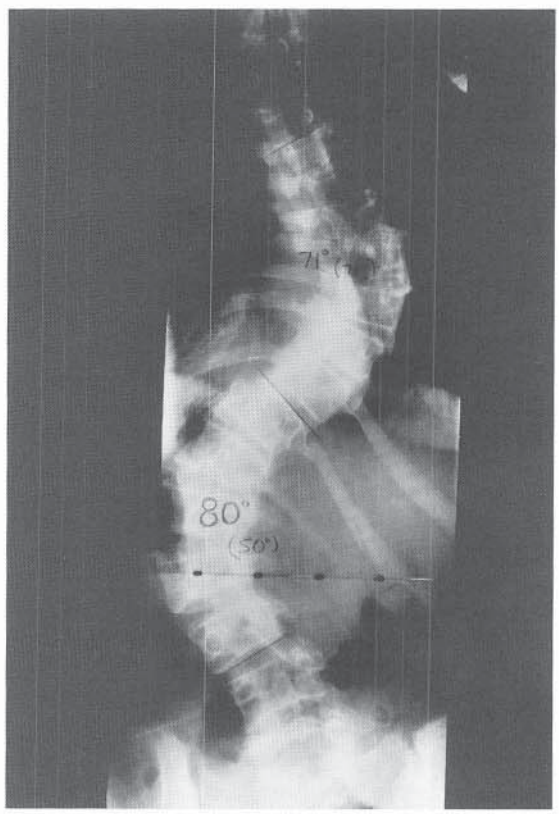


FIGURE 11–27 Double major curve pattern. The thoracic and lumbar components are similar in size, are structural, and have limited flexibility. It is important to differentiate double major curves from King type II curves.

sacral line (CSL) (Fig. 11–28). The CSL is a vertical line drawn upward from the center of the sacrum. King found the concept of the stable vertebra useful in determining appropriate fusion levels for all curve types. He recommended that the lower level of Harrington instrumentation stop at the stable vertebra. In this way, modest curve correction could be achieved and spinal balance maintained. For type II curves, King recommended a selective thoracic fusion, leaving the lumbar curve free of instrumentation. This approach resulted in satisfactory spinal balance and preserved valuable lumbar motion after Harrington instrumentation.

RECOMMENDATIONS FOR SELECTING FUSION LEVELS BASED ON CURVE PATTERNS

King Type I. The surgical options depend on the magnitude and flexibility of the thoracic curve. Commonly, the thoracic component is large, structural, has notable rotation, and clearly crosses the midline, and posterior spinal instrumentation is necessary to achieve a well-balanced spine. On occasion the thoracic component is relatively small (less than or equal to 30 degrees), barely crosses the midline, and has minimal rotation. In these cases, anterior spinal instrumentation of the lumbar curve only may result in maximum frontal plane and rotational correction of the lumbar curve, plus maintenance of spinal balance. A small residual thoracic curve will remain, but it usually is not noticeable.

King Type II. Selective fusion of the thoracic curve with instrumentation to the stable vertebra has led to excellent results when Harrington instrumentation is used. Curve correction approaching 40 percent and maintenance of spinal balance have been consistently reported.[285] With the advent of the newer "derotation" instrumentation systems in the latter 1980s, however, selective thoracic fusions often led to spinal imbalance. This is manifested as a shift in the patient's trunk or head (or both) to the left of midline.*

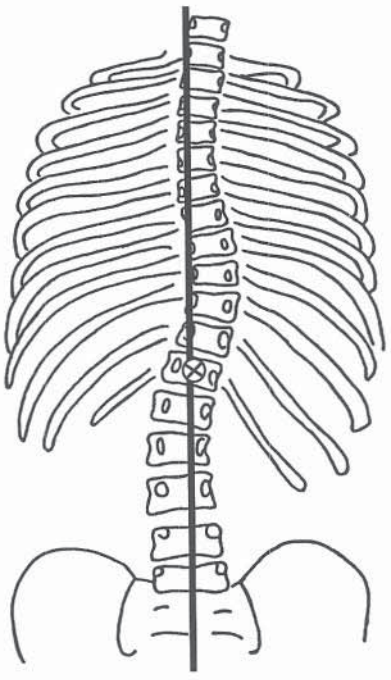


FIGURE 11–28 The stable vertebra (*x*) is the inferior vertebra in the thoracic curve that is most closely bisected by the center sacral line. This reference point has been used in determining appropriate fusion levels during preoperative planning. (From Richards BS, Birch JG, Herring JA, et al: Frontal plane and sagittal plane balance following Cotrel-Dubousset instrumentation for idiopathic scoliosis. Spine 1989;14[7]:733–737.)

Many theoretical explanations have been offered for this imbalance following selective fusions with the newer generation implants. Such explanations include improper choice of fusion levels, overcorrection of the thoracic curve, incorrect identification of curve patterns, lumbar curve stiffness and progression, and hook patterns. Certainly the newer implant systems imparted a stronger corrective force to the spine than the Harrington instrumentation. Instead of two points of fixation, as was used with Harrington distraction instrumentation, multiple hooks were utilized that rigidly attached the rod to the spine. Once the rod was seated in the hooks and the scoliosis was corrected by the 90-degree rod rotational maneuver, the forces imparted to the spine could not be sufficiently balanced by the uninstrumented lumbar component. Fortunately, in most cases where there was some postoperative imbalance, improvement over time occurred as the uninstrumented lumbar curve adapted (Fig. 11–29).[280,372]

When using the newer generation of instrumentation for selective fusions in King type II curves, the surgeon should keep in mind two principles that will help minimize the chances of postoperative imbalance. First, the instrumentation should not extend beyond the stable vertebra. Fusing one or two levels lower than this tends to shift the patient's trunk to the left. Second, the rotation maneuver of the primary rod should be less than a complete 90 degrees. Limiting the rotation maneuver will avoid overcorrection or excessive straightening of the thoracic curve. The uninstrumented lumbar curve can then compensate satisfactorily.[34,285,305,439] The preoperative planning for selective thoracic instrumentation and fusion of a King type II curve is shown in Figure 11–30.

Failure to properly distinguish King type II curves from true double major curves was also responsible for some of the imbalance following selective thoracic fusions. Useful guidelines have subsequently been developed to help differentiate King type II and double major curve patterns.[240] Relative ratios between the thoracic and lumbar curves with regard to their size, rotation, and deviation from the midline can be assessed preoperatively on a standing radiograph (i.e., thoracic curve parameter ÷ lumbar curve parameter). If the ratios are less than 1.0, both curves require fusion. If the ratios are greater than 1.2 for curve size and deviation and greater than 1.0 for rotation, selective thoracic fusion can be safely performed. With true double major curves, both curves must be included in the posterior fusion to achieve a balanced spine using CD, TSRH, Isola, and other new systems. The preoperative planning for instrumentation of double major curves is shown in Figure 11–31.

Recently, selective anterior fusion on the convexity of the thoracic curve in King type II patterns (using screws and either a threaded or a smooth rod) has been advocated.[39] Reported advantages over posterior instrumentation include improved balance, correction of a hypokyphotic thoracic

*See references 29, 34, 54, 240, 280, 284, 305, 372, 374, 439.

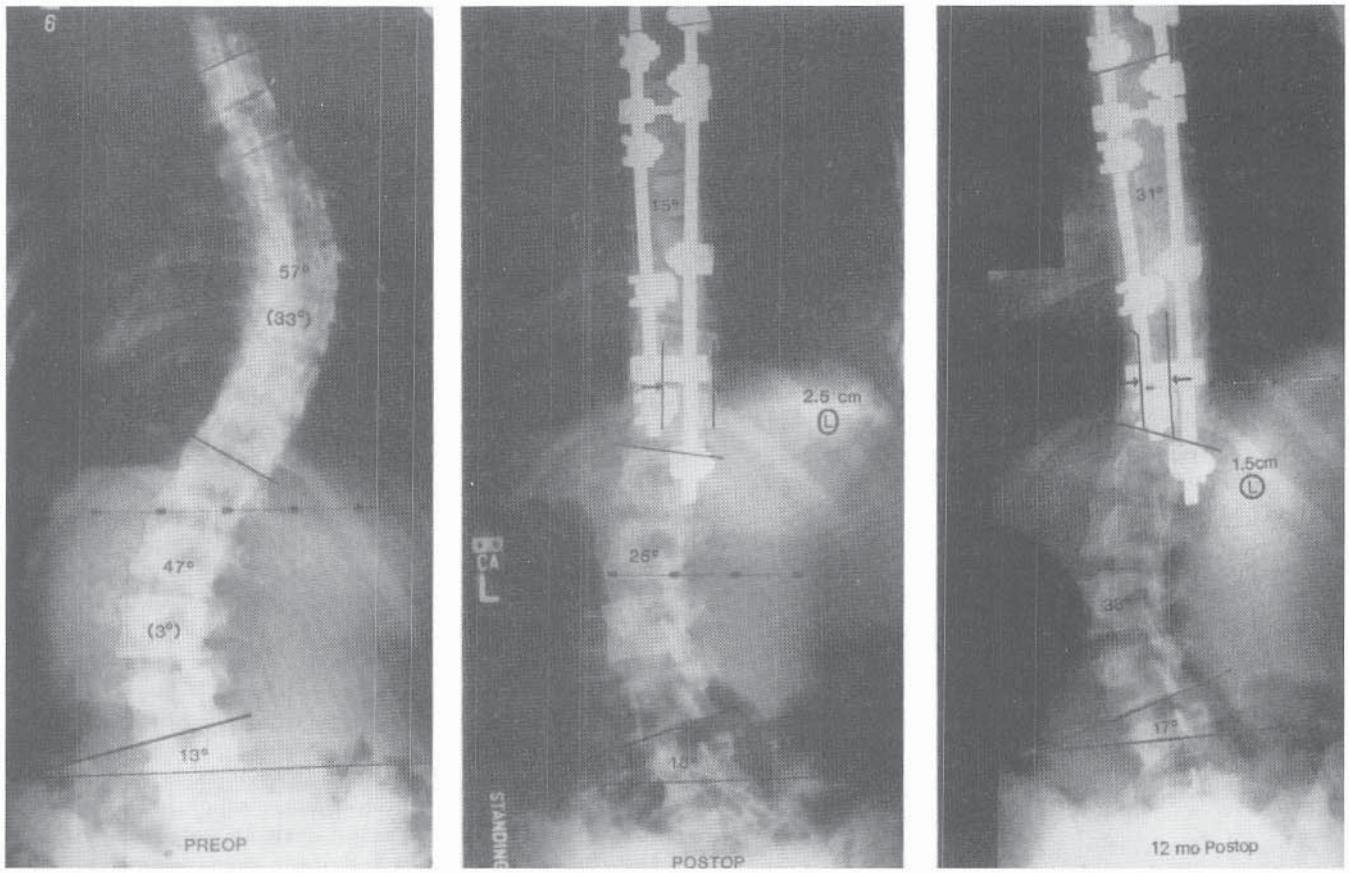


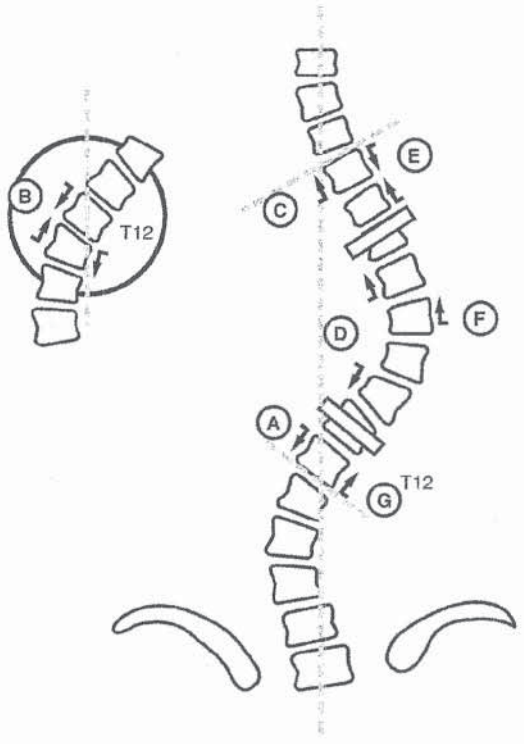
FIGURE 11–29 Preoperative, postoperative, and follow-up standing AP radiographs of a 17-year-old girl. *Left:* Preoperatively the lumbar curve was very flexible and the L4-pelvic obliquity measured 13 degrees. *Middle:* Postoperatively the lumbar curve remained larger than the thoracic curve, the L4-pelvic obliquity remained, and the trunk shifted 2.5 cm to the left. *Right:* At 12-month follow-up the trunk shift had improved, the lumbar curve remained larger than the thoracic curve, and the L4-pelvic obliquity persisted. (From Richards BS: Lumbar curve response in type II idiopathic scoliosis after posterior instrumentation of the thoracic curve. Spine 1992;17[8 Suppl]:S282–S286.)

spine, and preservation of more inferior motion segments. Preliminary studies, however, have reported breakage of the threaded rod in 31 percent of patients.[39] This complication may be remedied in the future by the use of a larger rod. Thoracoscopically placed anterior instrumentation is now being used in some centers familiar with thoracoscopic techniques.

King Types III and IV. Patients with King type III and IV patterns have single thoracic curves. Clinically, patients with these curve patterns have a more pronounced trunk imbalance to the right than is seen in patients with double curve patterns. Posterior instrumentation (using CD, TSRH, or Isola systems) predictably results in a significant amount of curve correction and, perhaps more important, a marked improvement in the patient's spinal balance. In type III curves, excellent correction can be achieved and spinal balance maintained when the inferior level of instrumentation stops one level cephalad to the stable vertebra.[374] The preoperative planning for instrumentation of a King type III curve is shown in Figure 11–32. In some type IV curves, instrumentation may be stopped two levels cephalad to the stable vertebra. Recent experience with anterior instrumentation has shown that even more inferior motion segments may be preserved when this technique is used.[39,206]

King Type V. King type V curves are double structural thoracic curves in which the first thoracic vertebra is tilted into the upper curve. The patient's shoulder on the side of the convexity of the upper curve often is elevated. The upper curve and the shoulder elevation may worsen if only the lower thoracic component is instrumented. Therefore, most King type V patterns require posterior instrumentation of both thoracic curves. The instrumentation should be extended up to the second thoracic vertebra if (1) T1 is tilted into the upper curve and the shoulder is elevated on the convex side of the upper curve, (2) the upper curve is greater than 30 degrees, with limited flexibility, or (3) the transitional vertebra between the curves is located at T6 or below.[234,242,475,479]

Single Thoracolumbar and Lumbar Curves. Optimal correction of single thoracolumbar curves, lumbar curves, or King type I curves with minimal thoracic components is achieved by anterior instrumentation. Curve correction greater than 75 percent and nearly complete rotational correction have been reported. In the early 1990s, constructs using one or two solid rods were developed that resulted in a decreased incidence of pseudarthrosis, better maintenance of restored sagittal lordosis, and elimination of postoperative brace immobilization.[187,199,205,207,447] The preoperative planning for in-



King Type II

FIGURE 11–30 Planning of selective thoracic fusion for a King type II curve.

1. The surgeon must correctly identify the curve pattern as one amenable to selective fusion. This can be done by assessing the relative ratios between the thoracic and lumbar curves with regard to size, rotation, and deviation from the midline on the standing preoperative radiograph. If the ratios are less than 1, both curves require fusion. If the ratios are greater than 1 (rotation), selective thoracic fusion can result in satisfactory correction and balance.

2. Hook sites are first planned for the concave side of the curve. The surgeon identifies the stable vertebra by determining which vertebra is bisected most closely by the center sacral line (drawn vertically on the radiograph). The inferior end vertebra of the thoracic curve is often also the stable vertebra. If close inspection of the lateral standing radiograph reveals no obvious junctional kyphosis at this level, then the inferior, downwardly directed sublaminar hook is placed here (*A*). Some authors have recommended extending beyond this level into the upper lumbar curve and reversing the hooks (*B*). This approach should be used cautiously because of reports of increased spinal imbalance.

3. An upwardly directed pedicle hook is placed at the upper end vertebra (*C*).

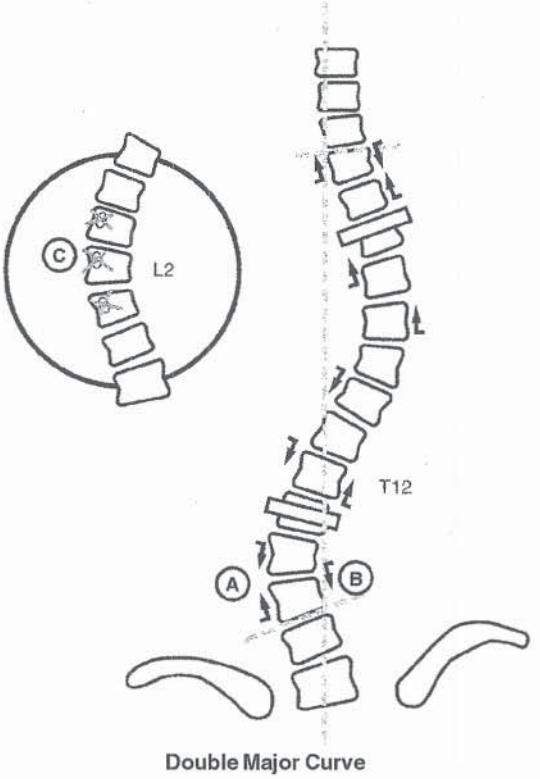
4. The two concave-side intermediate apical hooks are placed three interspaces apart (*D*). If they are placed closer together, difficulty may be encountered in securing the rod to the hooks. If they are placed farther apart, insufficient control is gained at the apical region of the curve. A pedicle hook is placed pointing up and a sublaminar hook is placed pointing down.

5. Placement of the second rod is now planned for the convex side. Placement of this rod does not result in further correction of the deformity but increases the torsional strength of the construct. A claw is placed at the upper two levels on the convexity (*E*). The most cephalad hook is placed pointing down on the transverse process of the upper end vertebra. A pedicle hook is placed pointing up one level caudal to the transverse hook. A two-level claw is recommended for ease of rod insertion, improved strength of the claw, and ability to perform facet arthrodesis in between. This claw construct allows compression along the convex rod between distant hook sites. The transverse process by itself is not strong enough to allow sufficient compression.

6. An upwardly directed pedicle hook is placed at the apex on the convexity (*F*). This is often the most prominent hook site because of the spinal rotation. Occasionally it is not used because its prominence may be excessive in the thin adolescent.

7. The inferior convex-side hook site is directed upward at the same level selected for the caudal hook site for the concave-side rod (*G*). If the hooks have been reversed as described in B, then the convex-side inferior hook is reversed in its orientation. Compression of the distal claw on the concave rod will trap this downgoing hook on the convex side, making a very stable distal attachment.

8. The planned hook sites must allow room for cross-links. Cross-links are placed as the final step, following partial rotation, distraction, and compression of the two rods. They should be placed as far superior and inferior as possible to maximize strength for torsional control of the construct.



Double Major Curve

FIGURE 11–31 Planning fusion for double major curve patterns.

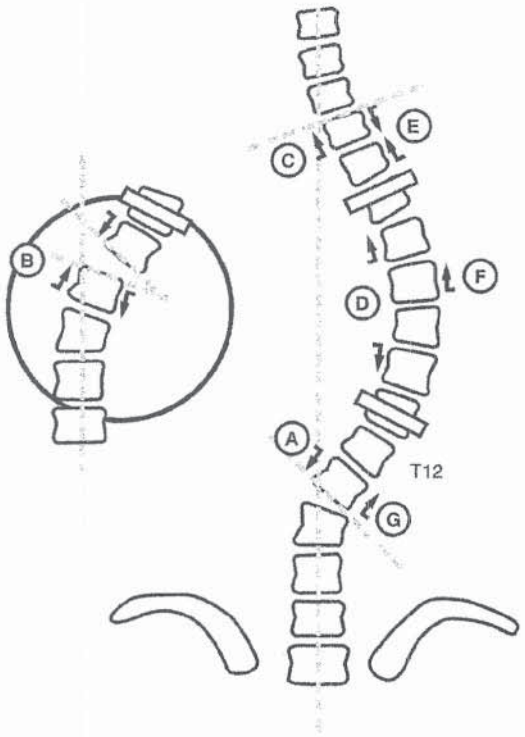
1. The hook sites used for the thoracic component of the deformity are almost identical to those used for selective thoracic fusion (see Fig. 11–30).

2. For the lumbar component, the instrumentation usually extends to L3. Fusion to L4 should be considered only if the L3 vertebral body deviates significantly away from the midline and if the L3–4 disk space remains wedged open on the side of the convexity. Fusion to L5 for idiopathic scoliosis is almost never indicated.

3. The convexity of the lumbar curve is approached first. An upwardly directed sublaminar hook is placed at L3, and either an upwardly directed or a downwardly directed sublaminar hook is placed at L2. The author prefers the downward direction to achieve a "claw" inferiority (*A*). This provides firm fixation and will allow compression across the entire convexity of the lumbar curve.

4. On the concavity of the lumbar curve, a downwardly directed sublaminar hook is placed at L3 (*B*). Compression of the L2–3 claw on the convex rod in the lumbar curve will trap the downwardly directed L3 hook on the concavity, making a very stable distal attachment.

5. Pedicle screws may be used as an alternative to hooks on the convexity of the lumbar curve and would be placed at L1, L2, and L3 (*C*). These screws allow increased correction to be achieved in the lumbar spine when compared to hooks. If this technique is used, a pedicle screw should also be placed in the L3 pedicle on the right (in place of a hook). Familiarity with pedicle screw placement is a prerequisite for this technique.



King Type III

FIGURE 11–32 Planning of fusion for single thoracic curves (King type III).

1. Hook sites are first planned for the concave side of the curve. The operator identifies the stable vertebra by determining which vertebra is bisected most closely by the center sacral line (drawn vertically on the radiograph). The inferior end vertebra of the thoracic curve is often one or two levels superior to the stable vertebra. The instrumentation can safely end one level cephalad to the stable vertebra with the expectation of excellent spinal balance. In this illustration, the lower level of instrumentation is L1. A downwardly directed sublaminar hook is placed on L1 (*A*). However, if the curve has started to reverse itself (i.e., the T12–L1 disk space opens toward the left), then the hooks are reversed. A downwardly directed sublaminar hook is placed at T12 and an upwardly directed sublaminar hook is placed at L1 (*B*). Placement of hooks in this orientation allows increased distal fixation, compression across an "open" T12–L1 disk space, and initiation of lumbar lordosis. This is the author's preferred hook configuration.

2. An upwardly directed pedicle hook is placed at the upper end vertebra (*C*).

3. The two concave-side intermediate apical hooks are placed three interspaces apart (*D*). If they are placed closer together, difficulty may be encountered in securing the rod to the hooks. If they are placed farther apart, sufficient control is not gained at the apical region of the curve. A pedicle hook is placed oriented upward and a sublaminar hook is placed oriented downward.

4. Placement of the second rod is now planned for the convex side. Placement of this rod does not result in further correction of the deformity but increases the torsional strength of the construct. A "claw" is placed at the upper two levels on the convexity (*E*). The most cephalad hook is placed oriented downward on the transverse process of the upper end vertebra. A pedicle hook is placed oriented upward one level caudal to the transverse hook.

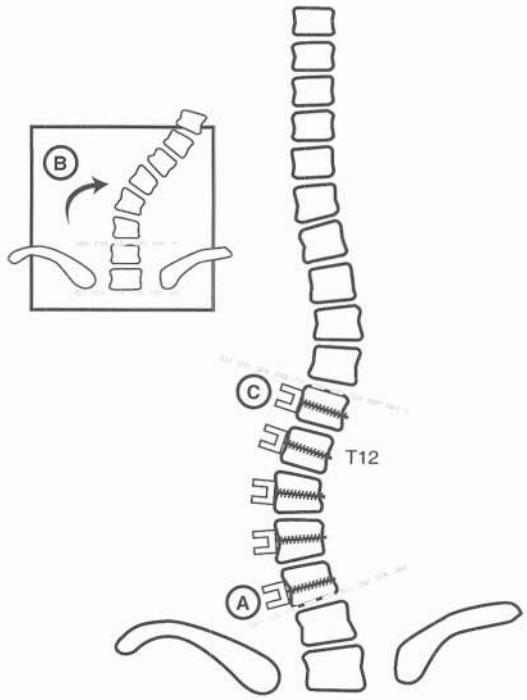
5. An upwardly directed pedicle hook is placed at the apex on the convexity (*F*). This is often the most prominent hook site because of the spinal rotation. On occasion it is not used because its prominence may be excessive in the thin adolescent.

6. The inferior convex-side hook site is directed upward at the same level selected for the caudal hook site for the concave-side rod (*G*). If the hooks have been reversed, as described in B, then the convex-side inferior hook is reversed in its orientation.

7. The planned hook sites must allow room for cross-links. Cross-links are placed as the final step, following rotation, distraction, and compression of the two rods. They should be placed as far superior and inferior as possible.

strumentation of a thoracolumbar curve is shown in Figure 11–33.

PREOPERATIVE CURVE FLEXIBILITY. Preoperative curve flexibility can best be assessed from side-bending radiographs. We use supine radiographs because they realistically reflect the amount of curve correction that can be achieved posteriorly with the newer generation instrumentation systems. Some surgeons use standing side-bending radiographs in an effort to predict residual balance and derotation of the lumbar spine (personal communication, Jean Dubousset). A great deal of caution must be exercised, though, if consideration is given to using side-bending radiographs for determining inferior levels of posterior instrumentation. When instrumentation is not taken far enough inferiorly, spinal decom-



Thoracolumbar Curve

FIGURE 11–33 Planning for anterior instrumentation of a thoracolumbar curve.

1. This technique is best for single thoracolumbar or lumbar curves. If this approach is considered for King type I curves, the accompanying thoracic component should be less than 30 degrees and should have minimal rotation.

2. Thoracolumbar curves treated with anterior instrumentation most commonly have five segments instrumented, with the inferior level ending at L3. The lower end vertebra of the measured curve is usually the best selection for the inferior instrumentation level and is determined from the standing PA view (*A*). This vertebra usually has little or no rotation. If instrumentation is discontinued one level cephalad to this, the disk space (between the last instrumented vertebra and the lower end vertebra) often remains notably oblique and does not regain normal balance. The long-term effects of this are not well understood. Sufficient flexibility of the spine below the planned instrumentation levels should also be confirmed on supine bending radiographs to ensure satisfactory postoperative balance. Unless a congenital obliquity is present at the lumbosacral region, the lower end vertebra nearly always becomes sufficiently horizontal (*B*).

3. The proximal level of instrumentation extends to, but not above, the superior end vertebra (*C*).

4. The surgical approach should be through the rib bed of the vertebra located one level cephalad to the most proximal vertebra to be instrumented.

pensation and "adding-on" to the curve may occur.[452] An important rule to remember is that both the proximal and distal levels of instrumentation are best determined by close scrutiny of the standing PA and lateral radiographs, not the bending radiographs. If anterior instrumentation is considered in thoracolumbar or lumbar curves, information from the bending radiographs may help confirm selection of the proper inferior vertebral level. This level must demonstrate the ability to become nearly horizontal with the sacrum on bending. Unless it does so, the trunk may remain out of balance over the pelvis. Preoperative traction films may also be helpful in assessing curve flexibility, particularly in curves exceeding 50 degrees.[357]

NEUROLOGIC STATUS. If a subtle neurologic abnormality is detected in an otherwise normal individual (e.g., asymmetric abdominal reflexes), then MRI of the entire spinal canal should be considered to rule out syrinx, cord tethering, or diastematomyelia.[504] Preoperative MRI should also be performed in patients with left thoracic curves because of the association with intracanal abnormalities.[366,397] The MRI study can be ordered when surgery is scheduled.

RIB DEFORMITIES. Excessive rotation of the thoracic spine may lead to extreme rib prominences, the most severe being described as "razorback." Fortunately, the razorback deformity is rare in idiopathic adolescent scoliosis (it is more commonly seen in nonidiopathic conditions such as neurofibromatosis). Because there are no definite guidelines that identify which patients should undergo thoracoplasty to reduce the rib deformity, how often the procedure is performed is often based on the surgeon's personal perspective. Suggested guidelines for patients who may benefit from thoracoplasty include a preoperative rib prominence exceeding 10 degrees (measured from a tangential radiograph with the patient bent forward 90 degrees), preoperative curves greater than 60 degrees, and flexibility less than 20 percent.[173]

In addition to improving the patient's cosmetic appearance, partial resection of three to five apical ribs provides bone graft in amounts sufficient to obviate an iliac crest graft.[25,145,173] Internal thoracoplasty has been safely performed when an anterior procedure has accompanied posterior surgery.[411] A prerequisite for thoracoplasty during posterior surgery is normal lung function. Thoracoplasty is contraindicated in a patient with compromised preoperative pulmonary or cardiac status.

FUTURE GROWTH POTENTIAL. The patient's maturity is assessed physiologically (by noting of rapid growth spurts and menarchal status) and skeletally (with the Risser sign). Most individuals with adolescent idiopathic scoliosis are quite mature by the time surgical intervention is needed (i.e., postmenarchal and past their peak growth period). As a result, scoliosis correction achieved by posterior spinal instrumentation and fusion usually is maintained over time and is not adversely affected by residual remaining growth in the anterior portion of the spine. In the immature child, however, a posterior fusion by itself may be inadequate because continued anterior spinal growth will produce progression of the scoliotic deformity over time. This event is known as the *crankshaft phenomenon* and is often associated with worsening of the curve, increased rib prominence, and deterioration in spinal balance.* Dubousset coined the term crankshaft phenomenon because he observed that the entire spine and trunk gradually rotated and deformed as the anterior portion of the spine continued to grow and twist around the axis of the fusion mass (in a manner similar to an automobile crankshaft) (Fig. 11–34).[110]

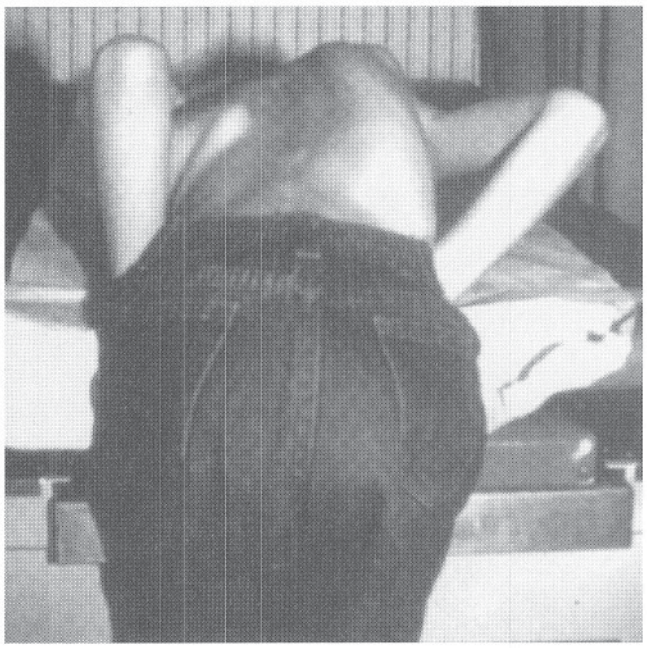


FIGURE 11–34 Clinical photograph demonstrating the crankshaft phenomenon in a young male patient several years after posterior spinal fusion. (From Richards BS: The effects of growth on the scoliotic spine following posterior spinal fusion. In Buckwalter JA et al (eds): Skeletal and Growth Development: Clinical Issues and Basic Science Advances, p 581. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1998.)

To better understand why the crankshaft phenomenon occurs, knowledge of spinal growth is important. Growth occurs in three areas of each vertebra: the vertebral end-plates, the cartilage overlying the articular processes, and the neurocentral synchondroses (Fig. 11–35). The two end-plates (physeal regions) in each vertebra primarily provide for longitudinal growth in the spine, at a rate of 0.7 mm per year per segment in the thoracic spine and 1.0 to 1.2 mm per year per segment in the lumbar spine. Posterior fusion arrests growth only of the posterior articular processes and does not affect any of the growth that remains anteriorly in the end-plates or in the neurocentral synchondroses. In younger patients, this results in continued growth of the anterior portion of the spine, even in the presence of a thick posterior fusion mass. Progression is proportional to the number of unfused growth centers and the number of years of growth remaining. Infantile and juvenile idiopathic scoliosis patients are most at risk; however, this phenomenon also occurs in the adolescent who is very immature at the time of posterior fusion.

Although difficult to quantify in severity, the crankshaft phenomenon can best be appreciated by examining serial clinical photographs that demonstrate progressive changes in the rib deformities, narrowing of the chest, and imbalance in the thoracic and lumbar spine. Radiographs can also

*See references 106, 110, 154, 168, 233, 235, 371, 393, 394, 410.

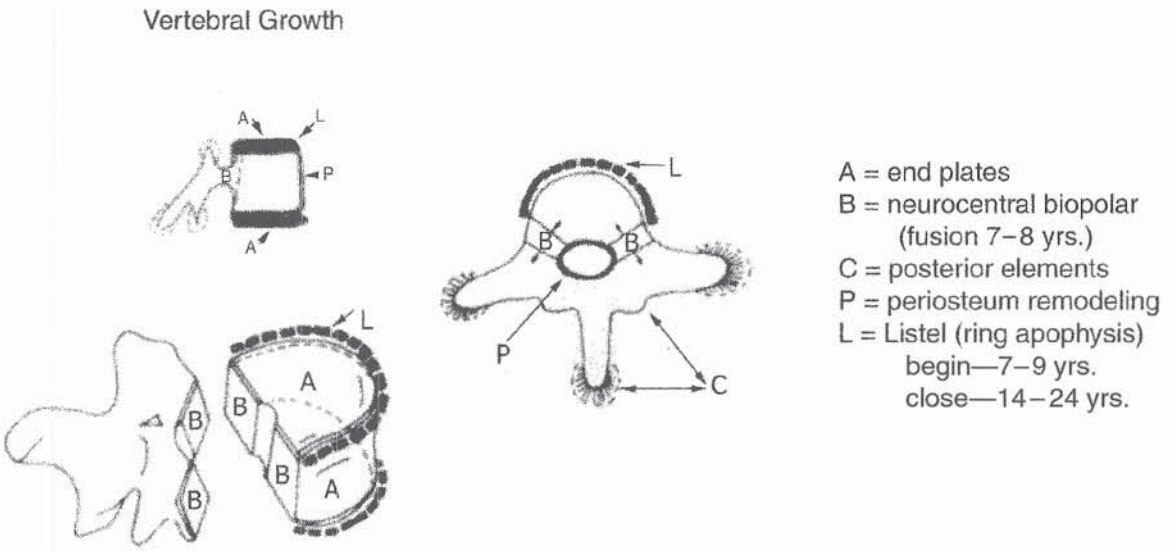


FIGURE 11–35 Three areas of vertebral growth—the vertebral end-plates, the cartilage overlying the articular processes, and the neurocentral synchondroses. (From Dubousset J, Herring JA, Shufflebarger H: The crankshaft phenomenon. J Pediatr Orthop 1989;9[5]:541–555.)

demonstrate progressive changes over time, such as (1) changes in the curve size, rotation, and rib vertebral angle differences, (2) translation of the apical vertebra toward the chest wall on the convexity, and (3) changes in the vertical inclination of the instrumentation. Radiographic changes greater than 10 degrees in curve size, apical vertebral rotation, and rib vertebral angle difference are all thought to reflect progression of the crankshaft phenomenon.[168,233,393] However, during the first 6 to 12 months following surgery, it is important not to automatically assume that changes in radiographic measurements are a result of the crankshaft phenomenon, as these changes are often due to stress relaxation of the spine, gradual maturation of the fusion mass, and realignment of the curve.

For female adolescents in need of surgery who have not yet reached their peak height velocity, who are premenarchal, and whose triradiate cartilages remain open, strong consideration should be given to combining anterior and posterior fusions to prevent the crankshaft phenomenon.[106,233,410] For the anterior spinal fusion, a conventional open thoracotomy approach is now being compared with the newer, less invasive video-assisted thoracoscopic surgery (VATS).[182,294,320,457] The advantages of VATS include muscle sparing, improved cosmetic results (less scar), and, perhaps, greater access to the entire length of the thoracic spine. Instruments are used through multiple intercostal portals to resect disk material, perform anterior release, and insert bone graft. However, extensive training is required before VATS is performed. Further research on the usefulness of this newer technique is needed.

TRANSFUSION REQUIREMENTS. Several procedures are available to reduce the need for homologous blood transfusions in patients undergoing posterior spinal instrumentation for scoliosis. These include controlled hypotensive anesthesia, autologous blood predonation of one or two units, acute normovolemic hemodilution, intraoperative and postoperative salvage of shed blood, and transfusion decisions based on clinical judgment rather than on a predetermined hemoglobin value.

Various combinations of these methods have been shown to significantly reduce exposure to homologous blood products in scoliosis surgery.* The combination of predonated autologous blood, hypotensive anesthesia, and intraoperative salvage of shed blood is probably the one most frequently used at the present time for healthy individuals with idiopathic scoliosis.[307,314] Intraoperative salvage of shed blood, the most expensive of the techniques available, is most effective when blood loss is expected to exceed 500 to 1,000 mL. Acute normovolemic hemodilution appears to be a satisfactory alternative to the use of predonated autologous blood.[82,335]

BONE GRAFTING. The primary goal of scoliosis surgery is achieving a solid fusion, which is enhanced by meticulous cleaning of soft tissue from the spine, facetectomies, decortication, and adequate bone grafting. Autogenous bone grafting continues to be the standard. Sources of autograft (which are generally determined by the surgical approach) include the posterior iliac crest, the spinous processes, and the ribs. A vertical incision over the posterior iliac crest may be the best harvesting method, as this approach decreases the risk of cutaneous nerve injury.

Over the past decade, there have been numerous reports on successful fusions in idiopathic scoliosis using allograft, frozen bank-stored bone as a substitute for autogenous bone.[22,45,105,123,428] Pseudarthrosis rates did not increase; however, follow-up was relatively short in some of the studies.[105,123] Reported advantages of the use of bank-stored bone include less blood loss, decreased operative time, and avoidance of the morbidity associated with iliac crest harvesting. To minimize the danger of transmission of HIV, hepatitis virus, and any other potential viral pathogen, the donor blood and tissue are tested at the site of recovery, with testing usually continued throughout the harvesting process.

*See references 82, 130, 143, 193, 227, 307, 314, 335, 354, 412, 413.

The freeze-dried cancellous bone is usually exposed to low-dose gamma radiation to sterilize all nonsystemic bacterial and fungal contaminants. Current research with bone morphogenic protein (BMP) indicates that it will have a significant role in spine fusions in the near future, perhaps obviating bone grafting.[376]

Spinal Cord Monitoring. Spinal cord monitoring, using somatosensory-evoked potentials (SSEPs), has become the standard of care during scoliosis surgery. This test records the sensory function of the spinal cord and provides continuous monitoring throughout the procedure.[13,75,76,176,311,324] It may, however, be adversely affected by changes in anesthetic level and perfusion. More recently, the development of monitoring by motor-evoked potentials has allowed monitoring of the spinal cord motor tracts.[36,149,338,396,426,432] When used in conjunction with SSEPs, the chance of unrecognized injury to the spinal cord is minimized. Unlike SSEPs, motor-evoked potentials have been shown to be reliable under isoflurane or desflurane anesthesia.[36]

The wake-up test, an evaluation of motor function, is no longer regularly used if spinal cord monitoring is available and found to be normal throughout surgery. The wake-up test is performed if changes in SSEPs are noted during correction of the spine. For this test, the anesthesiologist allows the patient to regain partial consciousness and motor function during the surgical procedure.[165] A recent study reported that the ankle clonus test was more accurate than the wake-up test and SSEP monitoring in predicting neurologic compromise.[188] Clonus should be present for a brief period as anesthesia is lightened. Its absence is abnormal.

Postoperative Pain Management. Patient-controlled analgesia (PCA) and epidural analgesia are the two methods used regularly in the management of postoperative pain. PCA provides safe and effective analgesia in children as young as 5 years. It allows the patient to self-administer small preprogrammed doses of opioids via a pump connected to the patient's intravenous tubing.[275] This enables the patient to titrate an opioid blood level in direct response to the changing intensity of pain. The built-in safety mechanism of PCA systems prevents oversedation. In addition, PCA devices can deliver a continuous infusion so that therapeutic levels of analgesia are maintained during sleep.

The use of epidural analgesia in scoliosis surgery has become increasingly popular, as it provides excellent pain relief.[14,215,275,404] At the end of the surgical procedure but prior to closure, the surgeon inserts an epidural catheter. The catheter is tunneled lateral to the incision and is usually left in place for 48 to 72 hours. Low-dose opioids are infused to provide effective analgesia, usually under the direction of pain management teams experienced with this technique. Close monitoring of the patient's respiratory status and the use of pulse oximetry are necessary until 24 hours after the infusion has been discontinued. Postoperative pulmonary toileting is optimized with this technique.

Ketorolac, an injectable nonsteroidal anti-inflammatory drug (NSAID), is effective for short-term management of moderate to severe postoperative pain. It is often used in conjunction with opioids because the combination provides more effective analgesia than either drug alone. However, recent research suggests that NSAIDs in doses typically used for postoperative pain significantly inhibit spinal fusion, and they perhaps should be avoided during the early postoperative period.[151]

POSTERIOR SPINAL INSTRUMENTATION

Harrington Instrumentation. Harrington developed his technique in the late 1950s and first reported it in 1962.[171,172] In this system, hooks are attached to posterior elements of the spine—facets, laminae, and transverse processes (Fig. 11–36). These hooks apply distraction forces to the concave side of the spinal curve using a ratchet mechanism. Compression forces are applied to the convex side of the thoracic curve at the base of the transverse processes, with the amount of force adjusted by tightening nuts on a threaded rod. Because distraction was the major corrective force, the distraction instrumentation was often used alone without the compression system.

For 25 years, Harrington instrumentation was the standard by which other systems of spinal instrumentation were assessed. Long-term follow-up studies have reported that approximately 30 to 40 percent of curve correction is maintained through the years with Harrington instrumentation.[99,192,263,297,466] However, with the development of the newer generation of spinal instrumentation systems in the 1980s, the limitations of the Harrington system became evident. Minimal, if any, three-dimensional correction of the spine can be achieved using simple distraction along the concavity; as a result, the thoracic rib hump is not corrected.[16,277,395,466]

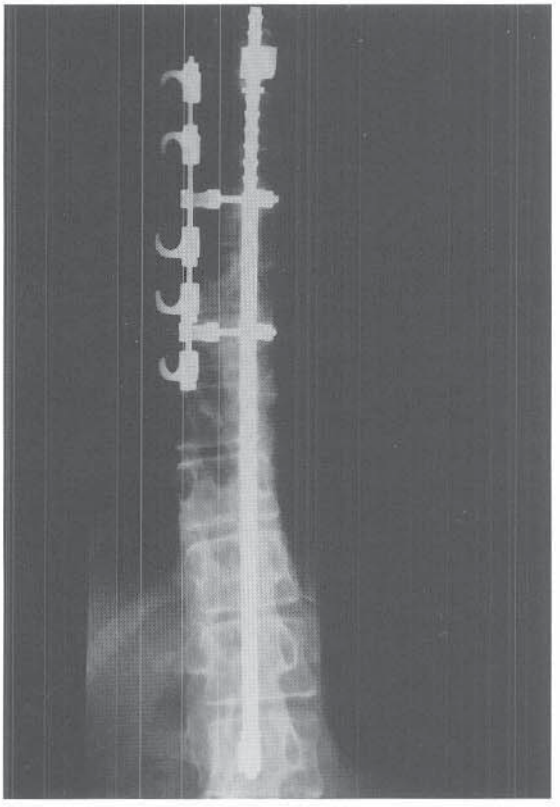


FIGURE 11–36 Harrington instrumentation. Distraction is applied along the rod between two hooks on the concavity of the curve. Compression forces are then applied on the convex side of the curve at the base of the transverse processes.

Second, distraction forces tend to flatten the spine and diminish or obliterate the normal lumbar lordosis.[70,192] Finally, the Harrington instrumentation construct does not provide sufficient stability to allow for brace-free postoperative mobilization. Thus, a cast or brace is required.

Over the long term, implant breakage occurs in as many as 40 percent of those instrumented with Harrington rods.[99] Today, Harrington instrumentation is rarely used in the surgical treatment of adolescent idiopathic scoliosis. The operative technique of Harrington instrumentation is described and illustrated in detail in the second edition of Tachdjian's *Pediatric Orthopedics.*[433]

Luque Double L-Rod Segmented Instrumentation. In this system, two contoured $\frac{3}{16}$- or $\frac{1}{4}$-inch stainless steel rods are wired to the spine at every vertebral level using sublaminar wires (Fig. 11–37). Each rod is bent to a right angle at one end, contoured to maintain thoracic kyphosis and lumbar lordosis, and wired transversely to the other. Unlike the Harrington system, the Luque construct provides sufficient stability to allow brace-free postoperative mobilization. This was of tremendous benefit to patients with paralytic deformity and insensitive skin. Unfortunately, the risk of neurologic damage during the passage of the sublaminar wires has precluded the regular use of Luque instrumentation for idiopathic scoliosis. Often this trauma is relatively minor, such as sensory dysesthesias that resolve within 2 or 3 weeks. However, major complications in the form of partial or total paralysis can occur.

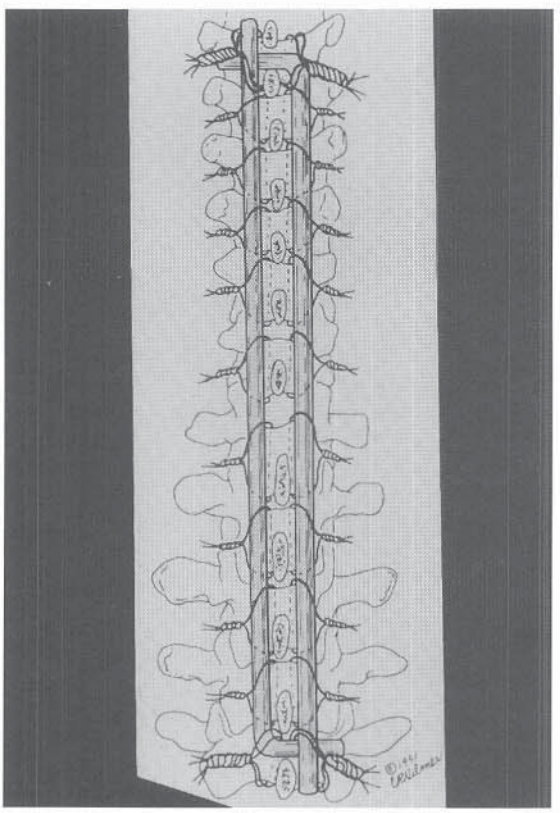


FIGURE 11–37 Luque double L-rod segmented instrumentation. Each rod is bent to a right angle at one end, contoured to maintain thoracic kyphosis and lumbar lordosis, and then wired to the spine at every vertebral level using sublaminar wires.

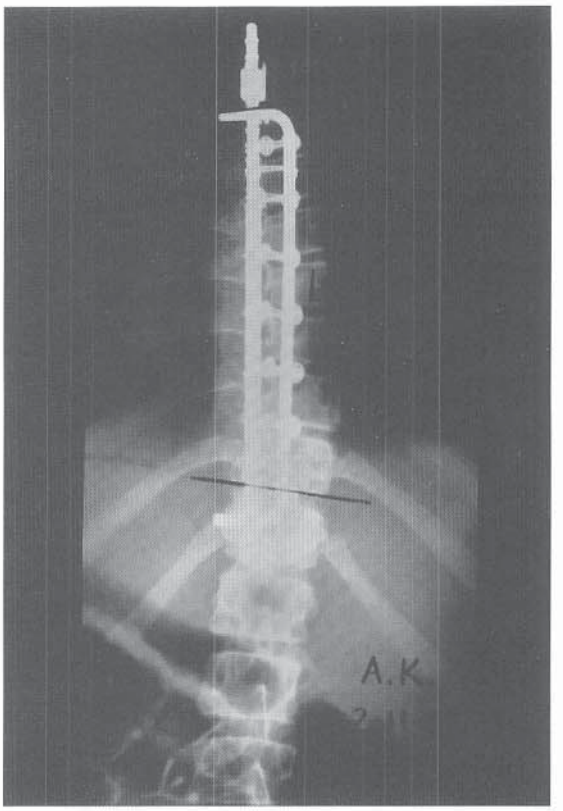


FIGURE 11–38 Wisconsin segmental spinal instrumentation (WSSI). Double button wires are passed through a hole at the base of each spinous process in the segment of the spine undergoing fusion. Because the wires are not sublaminar, the risk of inadvertent spinal cord injury during passage is lessened. The wires are then fixed to a Harrington distraction rod on the concavity of the curve and to a Luque rod on the convexity of the curve.

Wisconsin Segmental Spinal Instrumentation. In this segmental spinal instrumentation technique, purchase is achieved at each level of the spine by passing double button wires through a hole at the base of the spinous process (Fig. 11–38).[107,177,319] The wires are placed deep in the laminae but they do not enter the spinal canal. The wires are then fixed to a Harrington distraction rod on the concavity of the curve and to a Luque rod on the convexity. The rods are contoured into the postural sagittal curves, thereby avoiding flattening of the spine. Tightening the wires to both rods corrects the scoliosis. Postoperative immobilization is not required, as the segmental fixation is stable (Plate 11–1).

The distinct advantage of the Wisconsin segmental spinal instrumentation (WSSI) system over the Luque system is that the WSSI does not violate the neural canal with wires, yet it still provides adequate stability to allow brace-free postoperative mobilization. Technically the procedure is relatively simple and not time-consuming. Although it does not translate (rotate) the spine, as the newer generation multiple hook systems do, the WSSI system has provided satisfactory correction of scoliosis and maintenance of the sagittal contour, and it is relatively inexpensive.[177,178,198,319]

Cotrel-Dubousset Instrumentation. The Cotrel-Dubousset (CD) instrumentation system is a multiple hook system that was developed by Cotrel and Dubousset in France in the early 1980s and introduced into the United States in

the mid-1980s (Fig. 11–39).[84] The system revolutionized posterior instrumentation for idiopathic scoliosis by enhancing the surgeon's ability to improve the three-dimensional orientation of the spine. This was accomplished through the "derotation maneuver" popularized by Dubousset.[23,85] When the contoured rod is secured to the spine with various hooks, it is rotated 90 degrees. This maneuver restores near-normal sagittal contour, achieves significant curve correction, and improves the rotation or translation of the spine. CD instrumentation utilizes numerous hooks but no sublaminar wires, thereby avoiding encroachment of the neural canal.

With the CD technique, nearly all of the deformity correction is achieved with the rotation maneuver of the first rod. Placement of the second rod increases the construct's strength. When these two rods are rigidly united together with a rod-connecting device, sufficient torsional stability is achieved to allow brace-free postoperative mobilization.

Numerous reports have documented significant improvements in the correction of idiopathic scoliosis using CD instrumentation. Rib deformities are reduced, curve correction in the range of 48 to 69 percent is achieved and maintained, and near-normal sagittal alignment is restored.* The ability to preserve lumbar lordosis in curves requiring long fusion to L3 or L4 avoids the long-term "flat back" problems that were seen with Harrington distraction instrumentation.

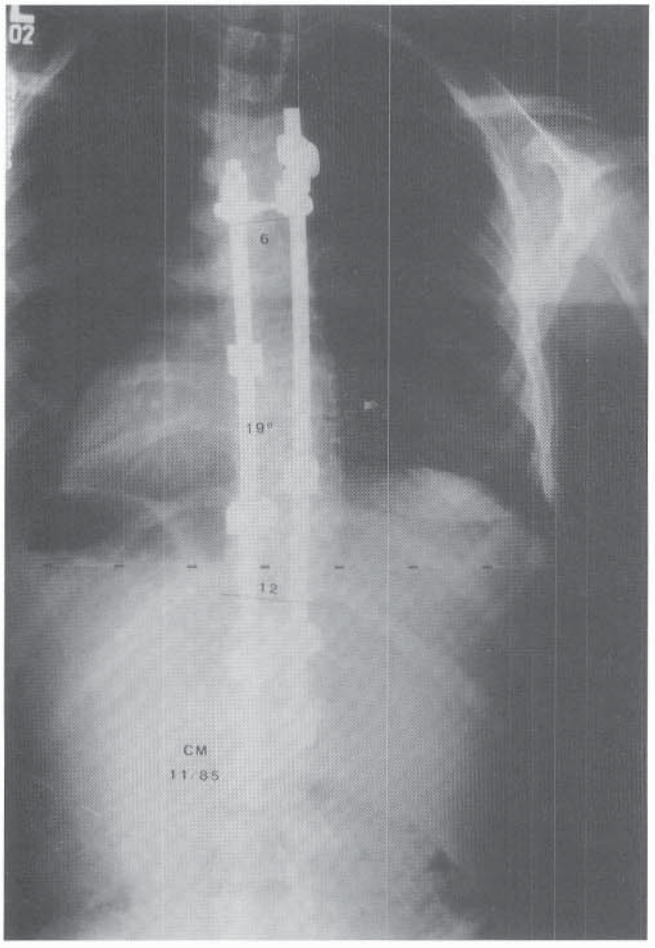


FIGURE 11–39 Cotrel-Dubousset (CD) instrumentation. The first contoured rod is secured to the concave side of the spine with multiple hooks and is rotated 90 degrees. This maneuver improves the sagittal contour and achieves significant curve correction. Placement of the second rod increases the construct's strength. CD instrumentation utilizes numerous hooks but no sublaminar wires.

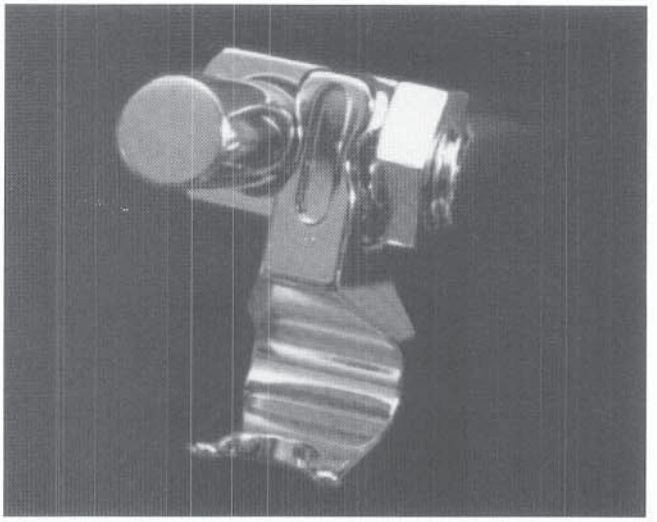


FIGURE 11–40 Texas Scottish Rite Hospital (TSRH) instrumentation. This system, like Cotrel-Dubousset instrumentation, uses multiple hooks to attach smooth, precontoured rods to the spine. The hooks attach to the rod through a three-point clamping mechanism provided by an eyebolt-nut system. (From Richards BS, Herring JA, Johnston CE II, et al: Treatment of adolescent idiopathic scoliosis using Texas Scottish Rite Hospital instrumentation. Spine 1994;19[14]:1598–1605.)

Texas Scottish Rite Hospital Instrumentation. The Texas Scottish Rite Hospital (TSRH) Instrumentation system was introduced in 1988 and, like the CD system, uses multiple hooks (and occasionally screws) to attach smooth, precontoured rods to the spine.[21, 200] All of the hooks and screws attach to the rod through a three-point clamping mechanism provided by an eyebolt-nut system (Fig. 11–40). The hook design is open, so that a rod can be inserted from above for ease of assembly. A small recess in the hook allows the rod to be captured within the hook while the eyebolt nut is still only partially tight. Once the system is assembled, selective compression, distraction, and rotation maneuvers can be performed to correct the spinal deformity. These maneuvers follow the principles introduced by Cotrel and Dubousset.[85] The technique of this multihook system is illustrated in Plate 11–2.

Studies evaluating TSRH instrumentation in the surgical treatment of idiopathic scoliosis have reported findings similar to those seen with CD instrumentation.[43,375,421]

Isola Instrumentation. The Isola instrumentation system was introduced in the late 1980s (Fig. 11–41). Its name was derived from a butterfly species because of the similarity seen in the early implant component.[19,20] The Isola system is based on the principles and designs conceived by Harrington and refined by Asher. These principles (which are applicable to all of the newer generation instrumentation systems)

Text continued on page 258

*See references 42, 52, 91, 135, 160, 192, 229, 230, 237, 239, 241, 356, 364, 374, 388, 409, 451, 463, 490, 494.

Wisconsin Segmental Spinal Instrumentation

OPERATIVE TECHNIQUE

In this system, fixation is achieved at each level of the spine by passing double button wires through a hole at the base of the spinous process. The system results in segmental fixation similar to that achieved with the Luque system but is safer in that it does not violate the neural canal with wires. It provides adequate stability to allow brace-free postoperative mobilization.

The spine is exposed in the same fashion for any posterior instrumentation technique. A Harrington rod is used on the concave side of the curve. The upper and lower distraction hook sites are prepared for the Harrington rod. Once these hook sites are prepared, facetectomies are performed at all of the remaining levels that will be included in the fusion area.

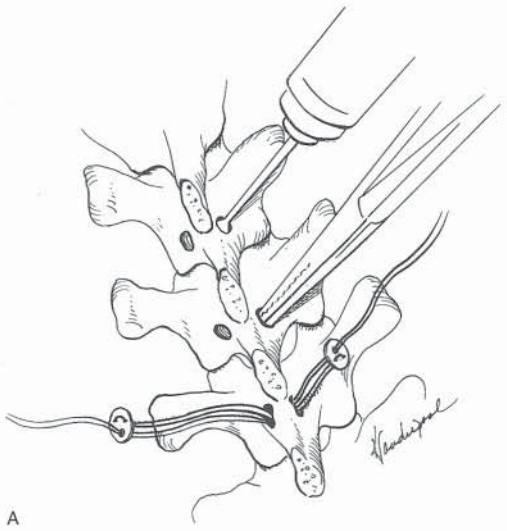
A, Next, the base of the spinous process at each level (of the interval to be instrumented) is prepared. Either an awl or a powered bur can be used to create the hole at the base of the spinous process. It is important while using the curved awl that the hand of the surgeon be held low to the spine, to avoid inadvertent penetration into the spinal canal. The tip of each button wire is introduced through the hole on each side of the base of the spinous process. The implants (two at each level) interlock when the beaded wire from one implant passes through the hole of the button on the opposite implant.

B, The buttons are then pulled snugly against the base of the spinous process. Before the Harrington distraction rod is placed on the concave side of the curve, the transverse processes are decorticated.

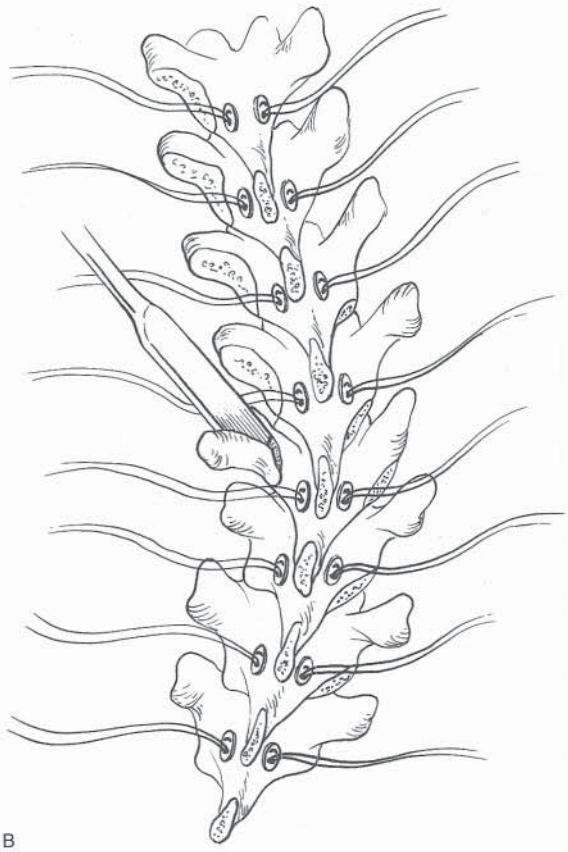
C, The Harrington rod is then placed in its hooks at the upper and lower levels. Distraction is gradually applied to provide some straightening of the spine. The rod is contoured so that physiologic sagittal curve alignment might be achieved. The wires are then tightened around the Harrington distraction rod to pull the concavity of the curve toward the Harrington rod.

D, After the distraction maneuver has been completed and the wires have been tightened, the Luque rod is contoured for the convex side curvature. The L-rod can be contoured slightly straighter so that further correction of the scoliosis might be achieved once the wires are in place. The ends of the L-rod are bent to right angles to the long axis of the rod. Sagittal plane contouring of this rod is done in the same way as for a Harrington rod. The L-rod is then tightened to the spine with the segmental wires. The transverse limbs at the ends of the rods should be long enough to span the spinal canal and prevent rotation toward it. The wires are tightened first at the apex and then alternately at each proximal and distal level, working toward the upper and lower ends of the instrumented segment of the spine. It is important that the correction be achieved by the assistant pushing the rod to the spine and not by twisting the wires alone. Further torsional stability of the construct can be obtained by locking the two rods together using the TSRH cross-link system.

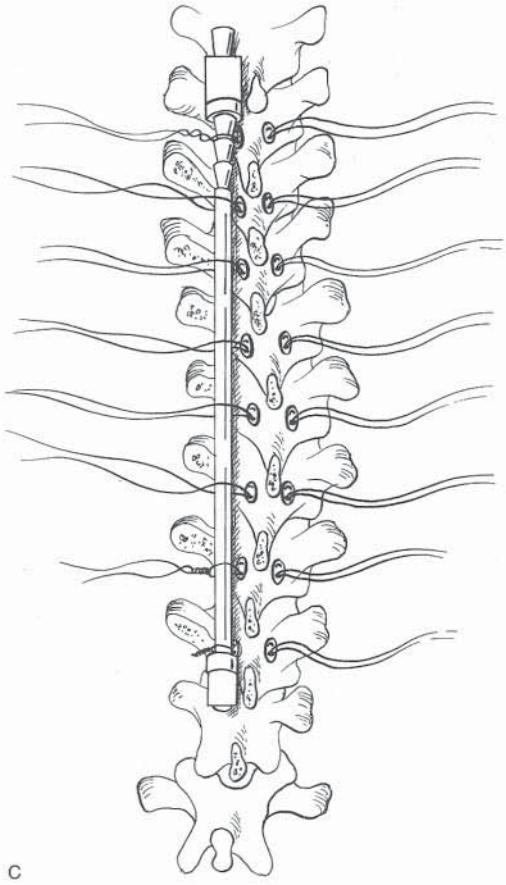
PLATE 11–1 Wisconsin Segmental Spinal Instrumentation



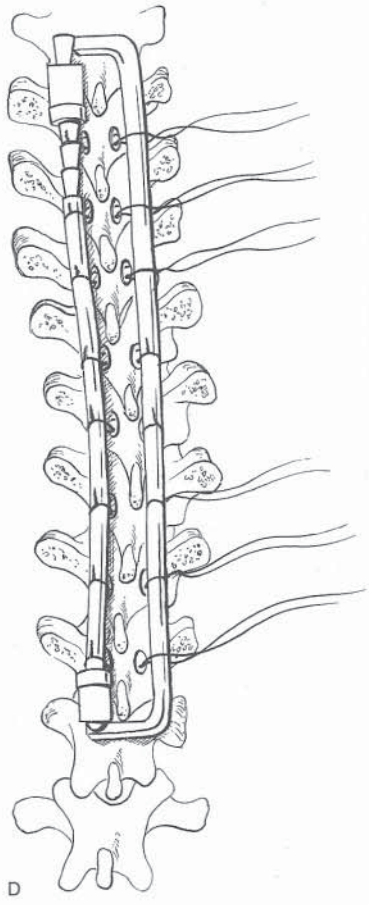
A



B



C



D

Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation

TSRH IMPLANT COMPONENTS (A–E)

A, The TSRH implants utilize a three-point shear clamp mechanism. The eyebolt attaches to the rod and, with tightening of the nut, secures the rod to the uprights of all of the hooks or screws. This adjustability of the nut allows selective tightness and reversal of the tightening mechanism. This will allow for rod rotation while maintaining distraction and compression, all without the need for nonimplanted extra devices. Wrenches (open or closed) are used to achieve final tightening of the eyebolt nut to the hook.

B, *1:* The pedicle hook sizes include pediatric (*left*), low-profile (*center*), and standard (*right*). The tines of the hook are designed to grasp the pedicle, while at the same time the buttress slant of the hook shoe fits precisely against the resected edge of the lamina. This makes the hook rotationally stable and unlikely to displace laterally.

2: Thoracic laminar hooks, used in the thoracic spine, are oriented in a caudal direction. These hooks are designed to achieve a "press fit" against the laminar edge and the underside of the lamina in order to minimize protrusion into the canal and prevent further intrusion in the canal during the rod assembly and correctional maneuvers.

3: Round laminar hooks are used most frequently in the lumbar spine. They can be directed upward underneath the laminar edge, directed downward under the laminar edge, or placed around a transverse process. The pediatric or various adult hook sizes accommodate patient anatomic variations.

4: Cranial-angled laminar hooks are placed in an upwardly directed position under the hyperlordotic lumbar lamina so that the center post is not oblique to the rod. This will provide more anatomic fitting at the inferior level of instrumentation for the upwardly directed hook.

C, Two types of screws are available for fixation in the lumbar or lower thoracic spine. The center post screw (*1*) has the upright identical to those seen in the hooks. As the screw has firm purchase within the pedicle and vertebral body, there is no "toggle" for the screw uprights, and therefore the rod fit must be contoured exactly. If this contouring is not possible, the variable-angle screw (*2*) allows connection of the rod to the screw at any degree of angle or rotation to the rod. When screws are used posteriorly in the spine, the rod is always attached in the medial position to the screws.

D, Two smooth rods are available ($\frac{3}{16}$ inch and $\frac{1}{4}$ inch). All rods have a shot-peened surface for fatigue resistance, have hexagonal ends for rotation (with a wrench), are available in titanium, and come in different degrees of stiffness, which allows proper matching to the spine requiring correction.

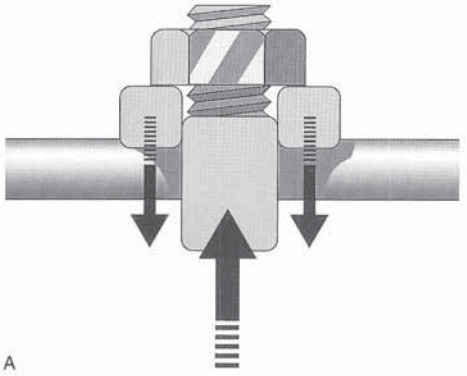
E, The TSRH cross-link provides rigid locking of the rods together, which will result in increased torsional stiffness of the two-rod construct (*top*). Generally, two cross-links are used for every construct. They are placed in the most cephalad and caudad position allowable. As shown in Figures 11–30 through 11–32, placement of the eyebolt should be preplanned on the drawings to ensure that satisfactory space is available for the cross-link. A low-profile cross-link is also available that does not require preplacement of the eyebolts (*bottom*).

OPERATIVE TECHNIQUE

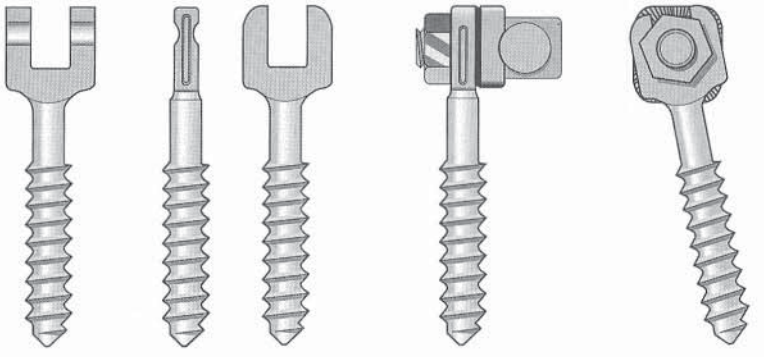
General anesthesia is administered via endotracheal intubation. Intravenous access is obtained, followed by placement of a radial arterial line. Perioperative antibiotics, usually first-generation cephalosporins, are given.

Autologous blood is made available by one of two methods. Preoperative donation of one or two units can be done. This blood is then available at the time of surgery in the form of packed red blood cells. The second method is to perform hemodilution once the patient has undergone anesthesia but before the skin is incised. This method allows two units of whole blood to be obtained over a period of 20 minutes. The operation is carried out while the patient is in a hemodiluted state. During closure of the incision, the blood is transfused back into the patient.

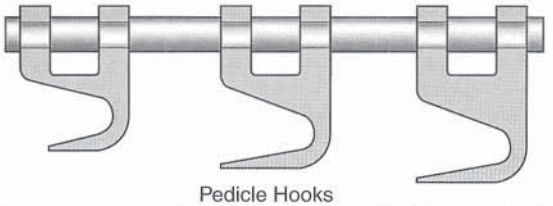
PLATE 11–2 Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation



A



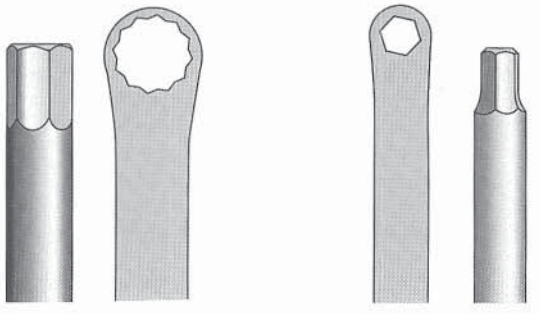
C, 1 Center Post C, 2 Variable Angle



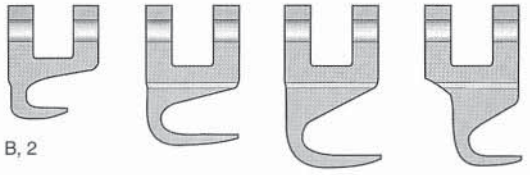
Pedicle Hooks

PEDIATRIC	LOW PROFILE	STANDARD

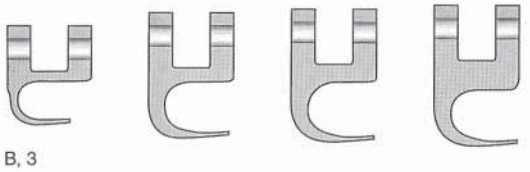
B, 1



D

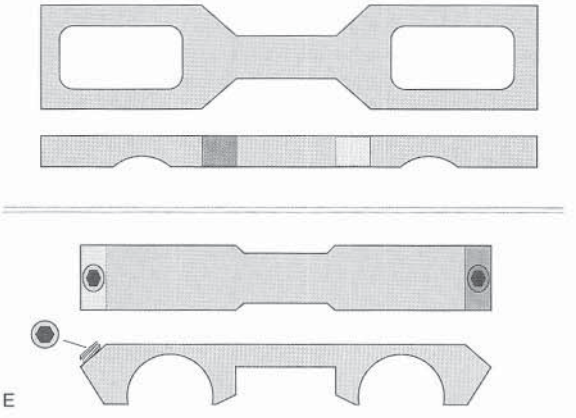


B, 2



B, 3

B, 4



E

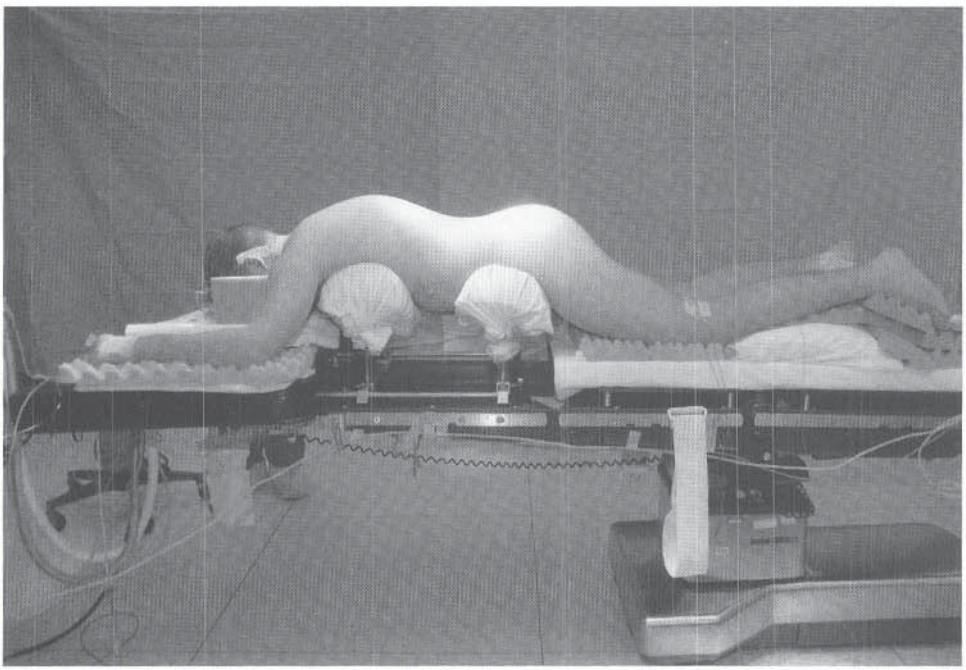
Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation *Continued*

F, *Positioning the Patient.* Under the supervision of the surgeon, the patient is placed prone on a Relton-Hall frame. Gel pads are placed over the four support pads of the Relton-Hall frame to further cushion the chest and inguinal region. The abdomen is free of any contact to minimize blood loss. The upper pads rest on the upper chest just lateral to the nipple region. The shoulders are abducted and the elbows are flexed. The axillae should be free of pressure. There should not be any stretch occurring across the brachial plexus or pressure over the ulnar nerve (at the elbow). The lower pads make contact at the ilioinguinal region. Pressure over the lateral femoral cutaneous nerve, unless satisfactorily padded, can lead to a temporary dysesthesia postoperatively in the anterior thigh.

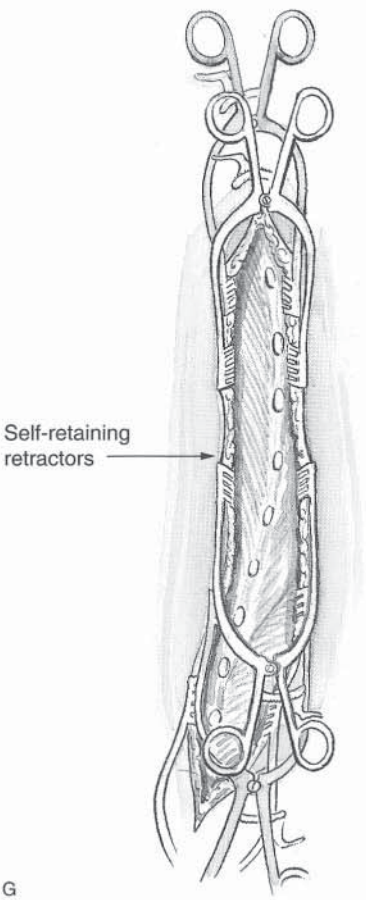
The entire back is prepared using Betadine, with application beginning at the base of the hairline and continuing to the gluteal cleft. Both iliac crests are included in the surgical field. After preparation and draping, a Betadine-impregnated sticky drape is applied. An adequate area must be exposed so that the incision never extends to the edge of the drapes.

G, *Incision.* The length of the skin incision is determined by the number of levels requiring fusion. The scalpel takes the incision down to the dermis. To minimize bleeding, electrocautery is used to continue the incision down through the dermis into the subcutaneous tissues. An alternative to this technique is to infiltrate the intradermal tissue with epinephrine and then to incise sharply down to the subcutaneous tissues. Self-retaining retractors are then placed into the wound to keep the skin edges under tension and provide exposure of the spinous processes.

PLATE 11–2 Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation



F



G

Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation *Continued*

H, Once the spinous processes have been exposed, the median raphe is incised sharply down to bone. A Kelly clamp may provide proper orientation for the incision. Dissection in this avascular plane minimizes blood loss.

I, Cobb elevators are used to subperiosteally expose the posterior elements. The exposure extends laterally to the tips of the transverse processes of all of the levels included in the fusion. In the thoracic spine, the dissection is easiest if it is begun distally and proceeds proximally. This facilitates the subperiosteal elevation of the oblique attachments of the short rotator muscles and ligaments from the lamina. Meticulous dissection should be performed to prepare the posterior elements for the upcoming fusion.

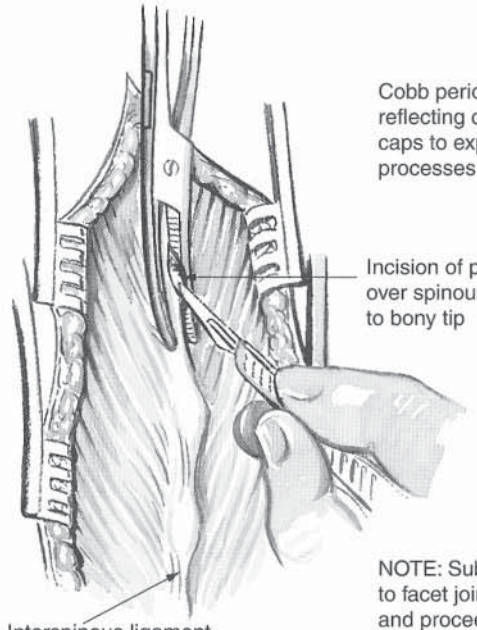
J, As the subperiosteal dissection continues, each level is packed firmly with gauze to minimize the bleeding.

K, Once the dissection is completed, packing is removed and self-retaining retractors are placed at the proximal, distal, and intermediate areas. Further cleaning of the surgical field is then performed using rongeurs, curets, and electrocautery.

To confirm the proper levels for fusion, an intraoperative localizing radiograph is obtained. A towel clip is placed over the spinous process of the most inferior vertebra exposed. Usually this vertebra will be located in the upper lumbar region. An AP radiograph is then obtained. Regardless of the surgeon's expertise, a radiograph should always be performed to avoid the inadvertent selection of the wrong vertebral level.

After exposure of the surgical field, hook (or screw) sites are prepared. Preoperative planning for proper hook (or screw) placement should be noted on the radiograph and should be familiar to all of those assisting in the operation (see Figs. 11–30 through 11–32 for techniques of selecting appropriate hook sites).

PLATE 11–2 Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation

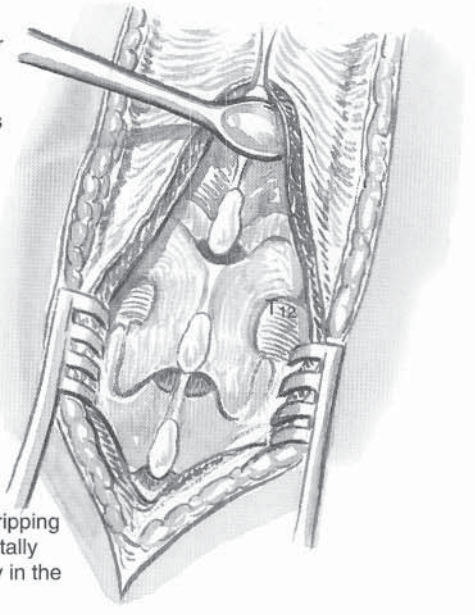


Interspinous ligament

H

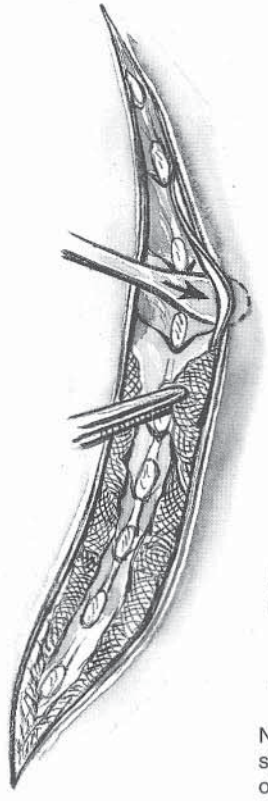
Cobb periosteal elevator reflecting cartilaginous caps to expose spinous processes on both sides

Incision of periosteum over spinous processes to bony tip

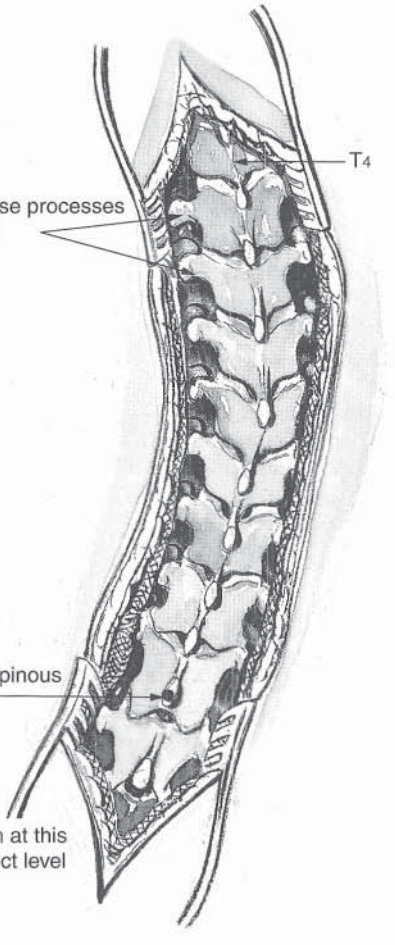


NOTE: Subperiosteal stripping to facet joints begins distally and proceeds proximally in the thoracic spine.

I



J



Transverse processes exposed

Metal markers on spinous process of T12

NOTE: X-ray is taken at this step to confirm correct level of exposure

K

Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation *Continued*

L, Pedicle hooks must be placed accurately to achieve maximum stability and grasp during the rotation or translation maneuvers that are used to correct deformity. Proper placement involves careful cutting of a hook site in the thoracic facet's inferior process. The insertion includes identification of the inferior edge of the thoracic pedicle (*A*). The proposed facet resection should remove just enough of the inferior process (*B*) so that the tines of the pedicle hook engage the pedicle and grasp it, while simultaneously achieving a "press fit" against the inferior edge of the resected lamina (*C*).

M, A properly placed pedicle hook.

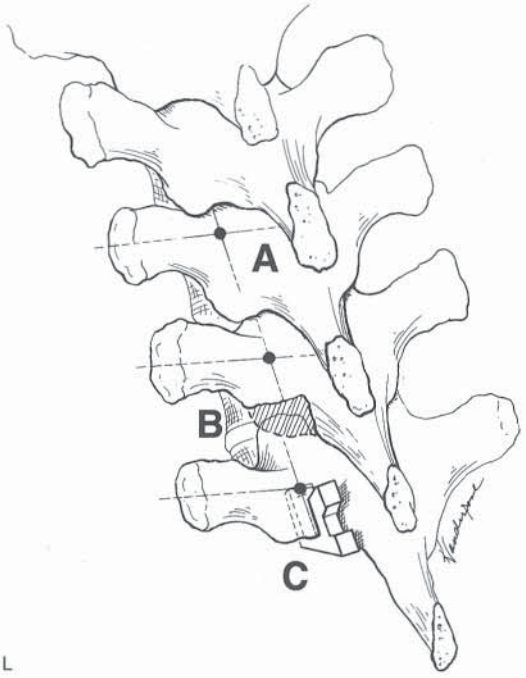
N, Laminar hooks directed cephalad are placed under the inferior edge of the lamina by dissecting with a sharp lamina elevator. A space is created for the shoe of the hook between the undersurface of the lamina and the ligamentum flavum.

O, In this diagram of a right thoracic curve, there are four hooks placed on the concavity. At the most cephalad site is an upwardly directed pedicle hook. At the most caudad site is a downwardly directed lumbar laminar hook. The intermediate hooks include an upwardly directed pedicle hook and a downwardly directed thoracic laminar hook.

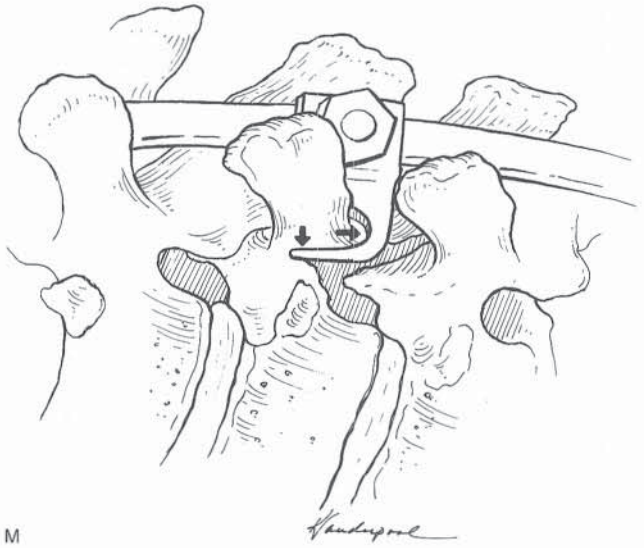
Before the first rod is attached to the hooks, final preparation of the spine is done. The hook sites on the convex side of the curvature are prepared, and facetectomies are then performed at all of the levels included in the fusion. This will increase spinal mobility so that the maximum amount of correction is achieved during the rotational maneuver. Bone graft should be placed over the decorticated laminae before seating the rod.

P, The first smooth $\frac{1}{4}$-inch TSRH rod is contoured. The proper contour is obtained by examining the lateral radiograph, determining the proper thoracic kyphosis and lumbar lordosis over the instrumentation levels, and bending the rod to match this. Placement of the hex-ended side of the rod toward the head facilitates the rotational correction maneuver. Often the rod is attached first to the proximal and distal hook sites. The intermediate two hook sites are then brought to the rod using an accessory "corkscrew" tool. The eyebolts for the cross-links should be placed on the rod before engagement of the rod by the hooks.

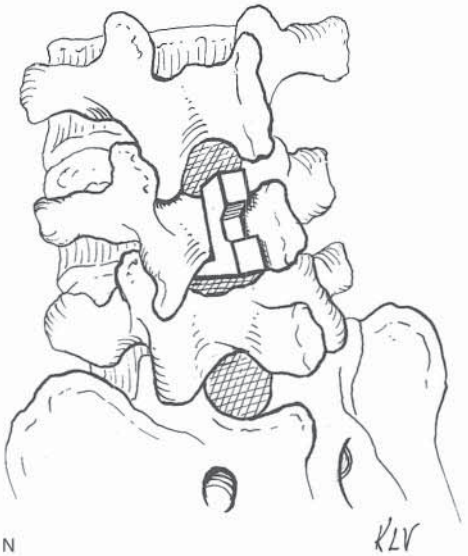
PLATE 11–2 Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation



L



M



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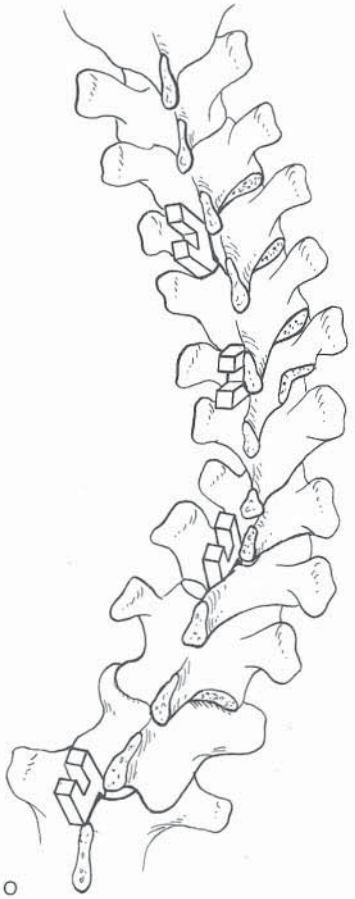

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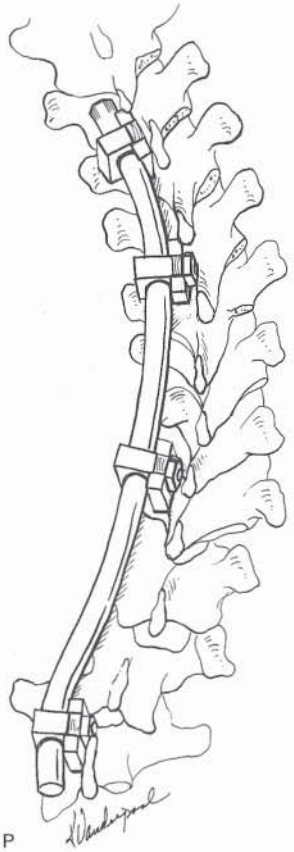


P

Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation *Continued*

Q, An intermediate thoracic laminar hook has been placed into the canal via a laminotomy. A corkscrew device is used to push the rod and preplaced eyebolt down into the uprights of the hook.

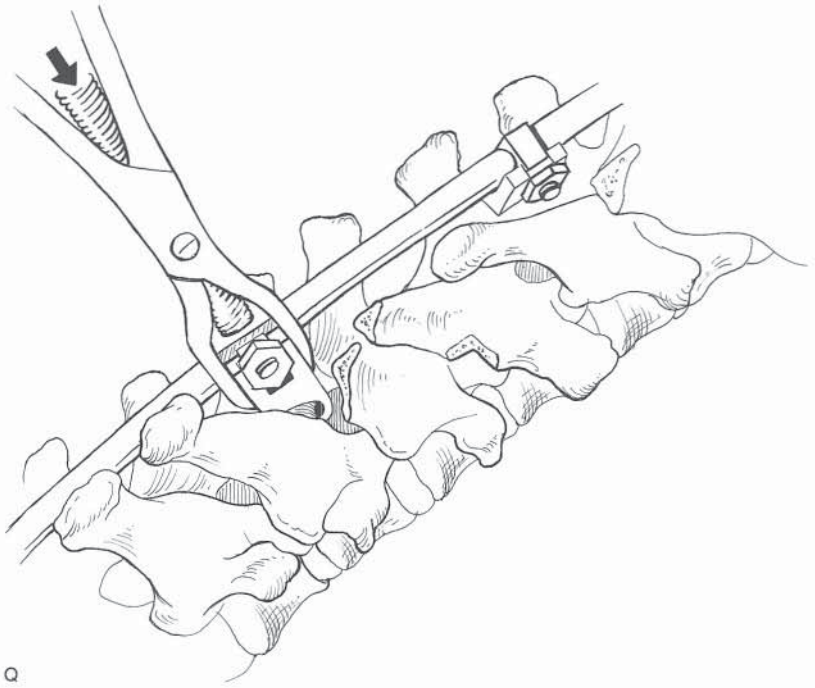
R, Once the rod is provisionally secured to the hooks, the rotational maneuver is performed. The spine will lengthen slightly with this maneuver. Distraction should be maintained between the intermediate hook sites, ensuring that they stay engaged during the rotation maneuver.

S, The spinal deformity is corrected by rotating the rod 90 degrees. Rotation is greatly facilitated by using the small wrench over the hexagonal end of the rod. On occasion, a vise grip applied to the distal end of the rod will assist in this rotation manuever. Rotation should be performed slowly to avoid possible intermediate-level hook disengagement. The most cephalad pedicle hook and most caudad laminar hook usually maintain excellent purchase during the correction maneuver.

The amount of curve correction is greatly determined by the flexibility of the spine. All of the correction that is achieved, is achieved with the first rod. After rod rotation, slight distraction is placed at each of the hook sites to ensure firm purchase. The nut on each eyebolt is tightened completely.

T, Further correction of the spinal deformity should not be expected with the second rod. However, this rod does significantly increase the torsional strength of the construct. The proximal end of the rod is secured to the spine with a "claw." This claw is created by placing a downwardly directed laminar hook over the transverse process at the most cephalad vertebral site. An upwardly directed pedicle hook is placed one level lower. These two hooks are then compressed along the rod to provide firm proximal fixation. Once this "claw" is firmly secured on the uppermost segments of the convex rod, compression can be applied along the rod at the remaining hook sites.

PLATE 11–2 Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation



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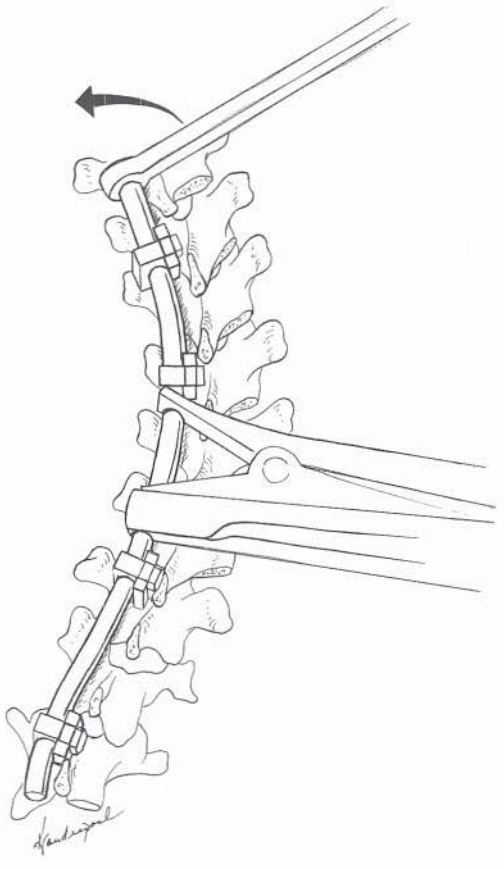

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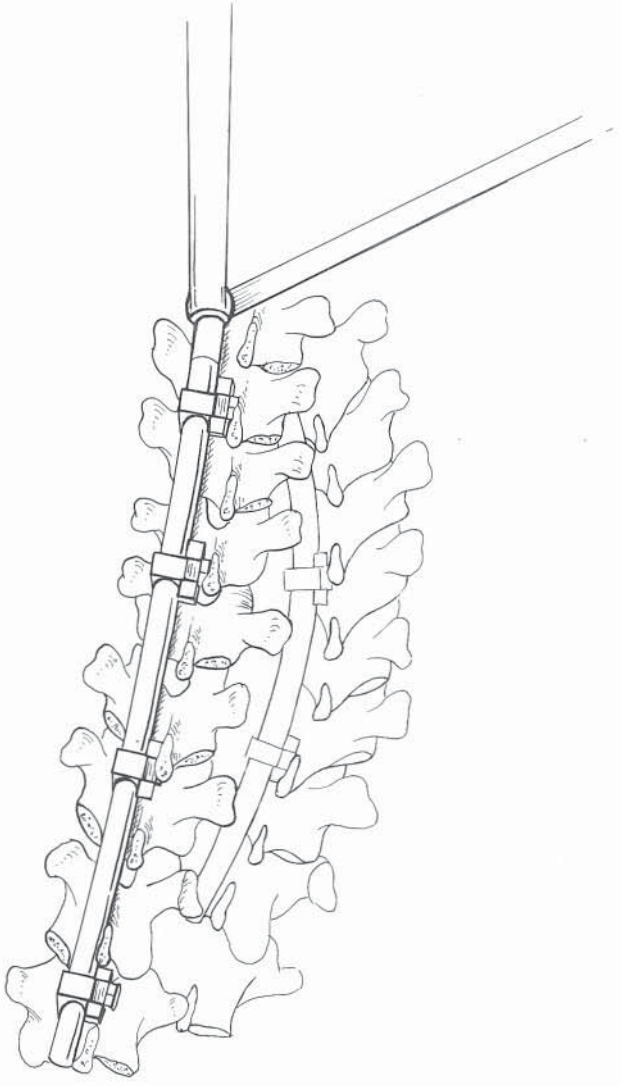

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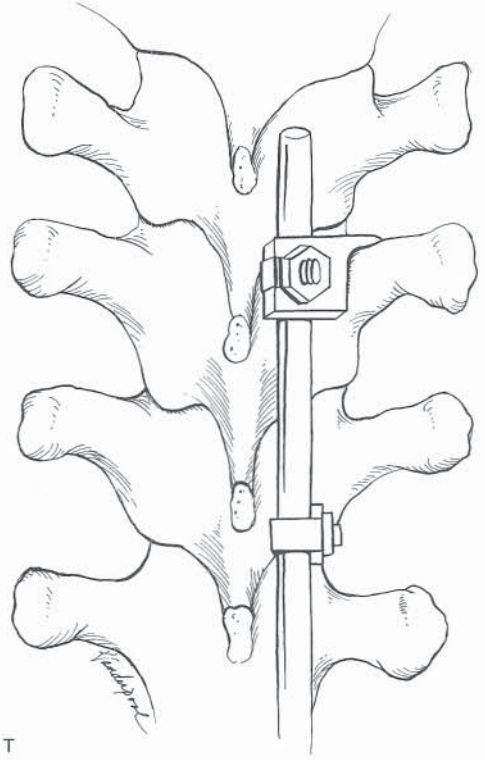


T

Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation *Continued*

U, An alternative method of fixation of the proximal site of the convex rod is to use a downwardly directed sublaminar hook. This configuration is used if the transverse process site does not provide satisfactory fixation for the claw configuration as described in T. At the intermediate hook site on the convex rod is an upwardly directed pedicle hook, which is usually the most prominent hook in this two-rod construct. If this hook is too prominent, it may be excluded.

At the inferior hook site on the convex rod is an upwardly directed sublaminar hook.

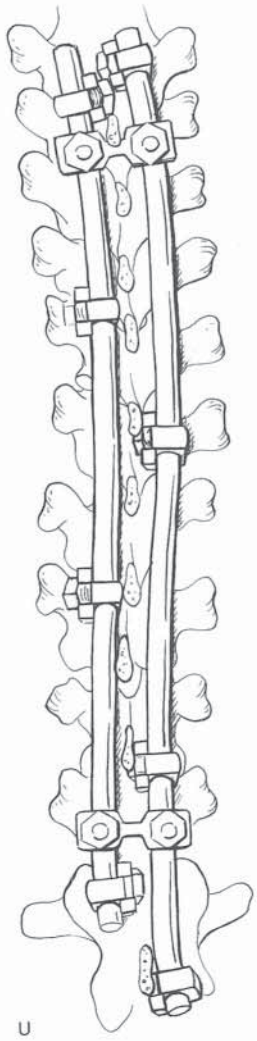
The eyebolts for the cross-link should be placed on the rod before engaging the rod with the hooks.

V, The rods must fit precisely in the grooves of all of the hook uprights (*right*). The corkscrew device is helpful in facilitating proper fit when the rod does not appear to be fully seated. For final tightening of the nut, a minimum of 150 inch-pounds of torque is applied. If the proper fit is not achieved (*left*), no amount of tightening of the nut will produce a stable three-point clamp between the hook and the rod. As a result, the rod could disengage from the hooks over time.

W, The cross-link is then placed at the proximal and distal ends of the rod to create a rectangular construct, thus ensuring maximum torsional stability. Intraoperative assembly of the cross-link is facilitated by using the nut starter, which allows two or three threads to be engaged on the eyebolt stem. The nut is then fully tightened to 150 inch-pounds of torque.

X, Once the construct has been fully assembled, the surrounding bone is decorticated. Autogenous iliac crest bone graft is then placed along the transverse processes, facet joints, and lamina.

PLATE 11–2 Posterior Spinal Instrumentation and Fusion Using TSRH Instrumentation



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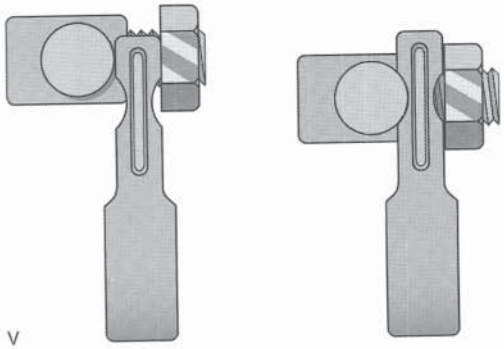

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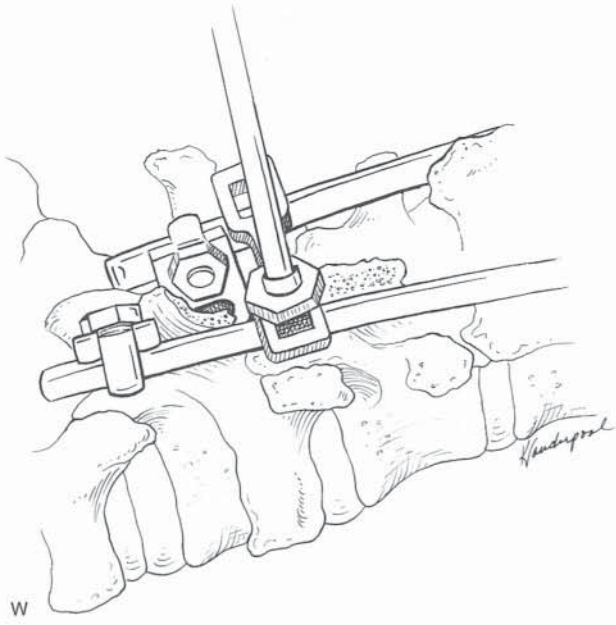

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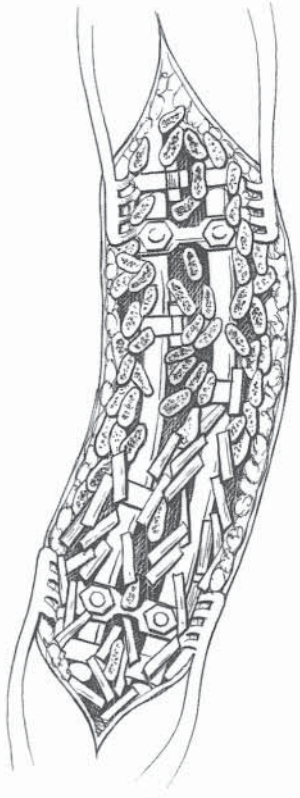


X

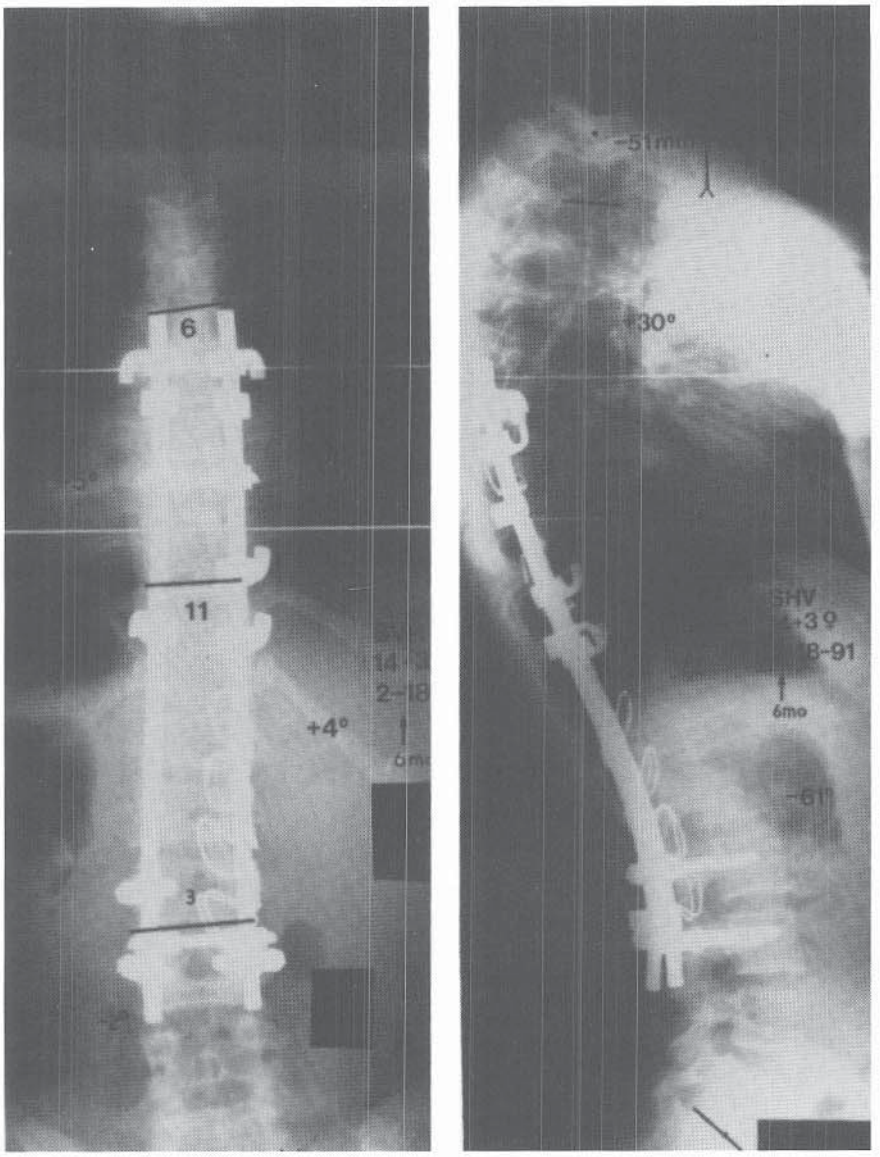


FIGURE 11–41 Isola instrumentation. To secure the rods to the spine and to achieve curve correction, numerous hooks, lumbar pedicle screws, and sublaminar wires are utilized with this system. (From An H: Spinal Instrumentation, p 343. Baltimore, Williams & Wilkins Co, 1992.)

include (1) precise description of the spinal deformity, with special emphasis on spinal balance, (2) anchors to laminae, transverse processes, and pedicles, and (3) segmental fixation. Unlike the CD or TSRH instrumentation, the Isola technique includes lumbar pedicle screw fixation and sublaminar wires as additional methods of obtaining purchase to the spine. The effectiveness of this additional fixation in correcting scoliosis has led to the occasional use of lumbar pedicle screws with CD and TSRH instrumentation. As with CD and TSRH instrumentation, translational and angular correction are nicely achieved with the Isola system.

Complications Associated with Newer Generation Posterior Instrumentation. Although the newer instrumentation systems offer increased versatility in the treatment of spinal deformities, they also involve the implantation of a substantial amount of hardware, including numerous hooks, two rods, and two rod-connecting devices. A 1 to 10 percent incidence of delayed wound infection has been reported, presumably related either to the increased amount of hardware or to the multiple hook-rod connections.[111,370,456,470] Some of these episodes of delayed drainage have been aseptic and were attributed to micromotion at the hook-rod interface.[111,470] Micromotion causes metal debris, which leads to a foreign body reaction forming a false membrane and fluid that finally results in loosening of the implant. Rather than representing an aseptic process, it is more likely that these delayed infections resulted from low-virulence organisms that were seeded at the time of surgery and remained quiescent over an extended period of time.[370,456]

Other potential complications include paralysis and implant failure. Very low rates of neurologic deficits have been reported, and problems have occurred mainly when an anterior diskectomy was performed at the same surgical setting as the posterior procedure.[53] The etiology is primarily thought to be related to vascular insult secondary to division of segmental vessels.

Implant failure is uncommon with double rod systems. On occasion, inferior hooks may dislodge. If this occurs and curve correction is lost, revision may be necessary. Use of a single rod posteriorly with multiple hooks was shown to

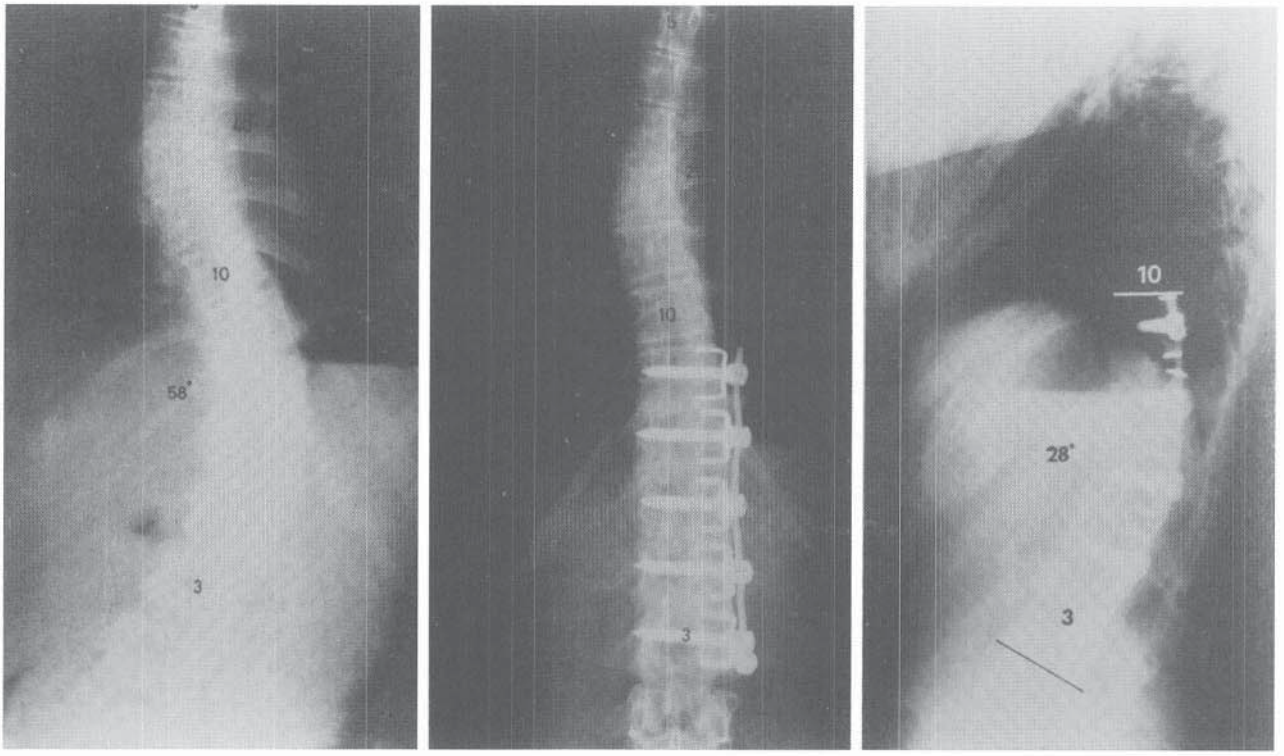


FIGURE 11–42 Dwyer anterior instrumentation. A flexible titanium cable is attached to vertebral body screws on the convexity of the curve. By tensioning the cable between screws, curve correction could be achieved. Unfortunately, excessive instrument-related kyphosis within the operate lumbar segments and an unacceptable rate of pseudarthrosis have been reported with this system. (From Hsu LC, Zucherman J, Tang SC, et al: Dwyer instrumentation in the treatment of adolescent scoliosis. J Bone Joint Surg 1982;64-B:538.)

lead to an unacceptable rate of rod breakage[459] and is no longer recommended.

Additional Newer Generation Posterior Instrumentation. As the technology for spinal instrumentation evolves, the number of available multiple-hook posterior implants for the correction of scoliosis continues to expand (e.g., AO Universal Spine system, Moss Miami instrumentation, Synergy spinal system, and CD Horizon). However, the principles established over the past 10 years with the CD, TSRH, and Isola systems continue to be followed. To maximize efficiency in the treatment of scoliosis, formal training with one or more of these newer generation instrumentation systems is necessary.

In 1995, Suk and associates described a more aggressive posterior approach in which segmental fixation is achieved with thoracic and lumbar pedicle screw fixation.[431] Slightly better correction in the frontal plane deformity and improvement in rotation are reported with this technique. However, only experts familiar with its use should attempt the technique because of the risks associated with multiple level screw placement.[58]

ANTERIOR SPINAL INSTRUMENTATION

Dwyer Instrumentation. Dwyer introduced anterior spinal instrumentation for scoliosis in 1965.[115–117] By using a flexible titanium cable attached to screws in vertebral bodies, shortening the long convex side of the scoliotic curve effectively corrected thoracolumbar and lumbar curves (Fig. 11–42). Although the concept was sound, the instrumentation was not stable enough to provide satisfactory long-term results. Its limitations included the inability to adjust the instrumentation following crimping of the screw-cable connection, a lack of rotational stability, which resulted in an unacceptable rate of pseudarthrosis,[224] and instrument-related kyphosis within the operated lumbar segment. With the evolution of newer anterior spinal instrumentations, the Dwyer system is rarely, if ever, used today. The operative technique of Dwyer instrumentation is described and illustrated in detail in the second edition of Tachdjian's *Pediatric Orthopedics.*[433]

Zielke Ventral Derotation Spondylodesis Instrumentation. In 1973, Zielke advanced Dwyer's concept by introducing derotation of the instrumented segment. Instead of using a flexible cable, as was done with Dwyer instrumentation, a derotation-lordosation outrigger device manipulated a threaded $\frac{1}{8}$-inch rod on the convexity of the curve. Combining this maneuver with the blocking open of disk spaces with bone graft before compression could, in theory, prevent the unintentional development of kyphosis. The threaded rod provided segmental adjustability, as the compression nuts could be repeatedly tightened (Fig. 11–43).

Thoracolumbar and lumbar curve correction in the range of 70 to 85 percent and rotational correction in the range of 42 to 60 percent have been reported with Zielke instrumentation.* As with Dwyer instrumentation, postoperative

*See references 147, 169, 204, 205, 302, 310, 333, 365, 430.

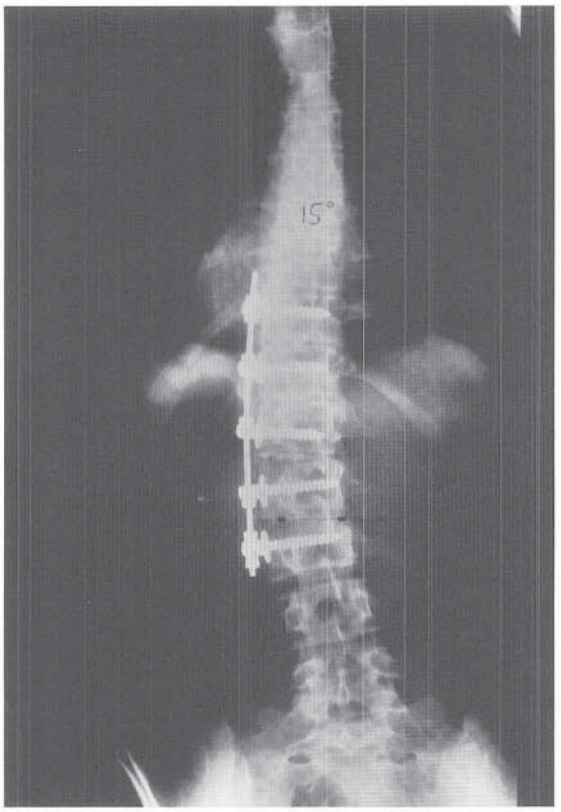
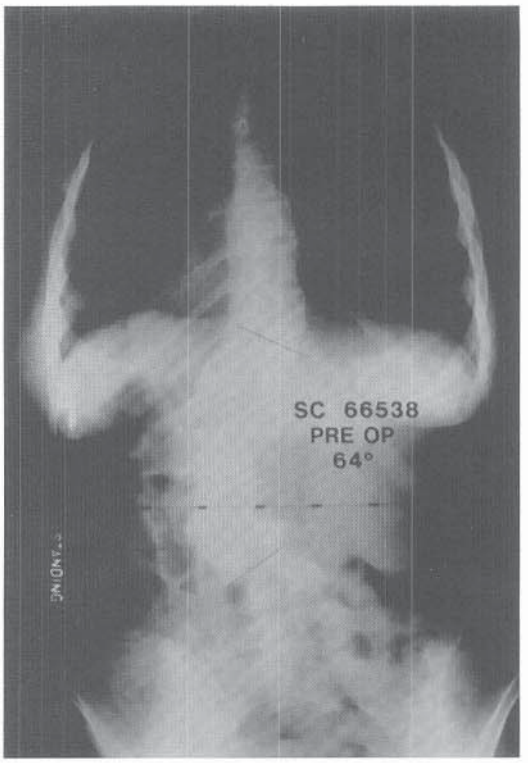


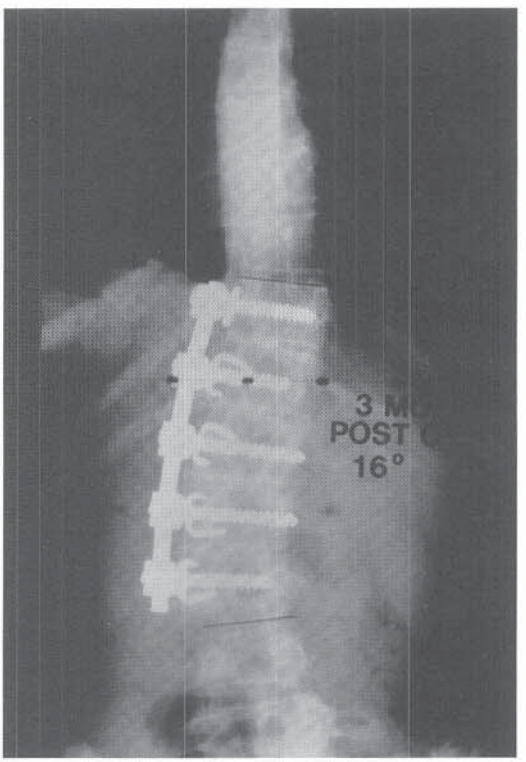
FIGURE 11–43 Zielke ventral derotation spondylodesis (VDS) anterior instrumentation. A threaded $\frac{1}{8}$-inch rod is attached to vertebral body screws on the convexity. Use of a derotation-lordosation outrigger device and blocking open of disk spaces with bone graft prevent, in theory, the unintentional development of kyphosis. Excellent frontal plane curve correction is achieved.

brace immobilization is required, as the threaded rod is not sufficiently strong enough to allow brace-free ambulation. Limitations of Zielke's technique include pseudarthrosis in 5 to 20 percent of instrumented segments.[169,189,430] In addition, a 2- to 8-degree increase in kyphosis within the instrumented segment, despite the use of the lordosation device, has been reported.[265,310,328] The operative technique of Zielke instrumentation is described in the second edition of Tachdjian's *Pediatric Orthopedics*.[433]

Newer Generation Solid Rod Anterior Instrumentation for Thoracolumbar and Lumbar Deformity. Toward the end of the 1980s, solid rod constructs were introduced for anterior instrumentation. TSRH instrumentation extended the concepts of Zielke by using a stiffer, smooth, solid rod as the longitudinal connection between vertebral screws (Fig. 11–44).[199,200,447] The resulting stiffer, fatigue-resistant construct enhances the maintenance of correction and the likelihood of arthrodesis without postoperative external immobilization in most cases. Deformity is corrected by rotation of a 6.4-mm rod, precontoured for lordosis (similar to CD instrumentation principles for thoracic curves posteriorly, only in reverse). Thus, lordosis is created and maintained by the stiffness of the construct and by structural anterior bone grafting. Although compression can be applied segmentally after rod rotation, the current recommended technique actually uses minimal interbody compression, as any



A



B

FIGURE 11–44 **A** and **B,** TSRH anterior instrumentation. Use of a stiffer, smooth, solid rod as the longitudinal connection between vertebral body screws helps maintain curve correction and increases the likelihood of arthrodesis without postoperative external immobilization. Lordosis is created and maintained by the stiffness of the construct and structural anterior bone grafting.

compression anteriorly in the lumbar spine can produce undesirable segmental kyphosis. Because deformity correction is achieved by gradual rod rotation, corrective forces are evenly distributed all along the construct simultaneously, rather than applied acutely or gradually at a single segment. We have experienced good results without immobilization and currently use no postoperative orthoses. The technique of the anterior solid rod instrumentation for thoracolumbar or lumbar scoliosis is detailed in Plate 11–3.

Another anterior technique, popularized by Kaneda, utilizing a two-rod construct has been reported to be successful, with 90 percent frontal plane correction and excellent sagittal plane reorientation (Fig. 11–45).[186,187,207] This emphasizes the importance of construct stiffness and the stability provided by the second rod.

Anterior Instrumentation for Thoracic Deformity. Dwyer proposed this technique in the 1960s, but found correction unsatisfactory with the cable systems.[116] More recently the use of anterior instrumentation for thoracic deformity has reemerged. In 1988, Harms began repopularizing the idea after conjecturing that anterior correction without a posterior derotation maneuver of the thoracic curve in King type II deformities would prevent the lumbar curve from decompensating, as had been described following selective posterior instrumentation (Fig. 11–46).[39] In addition, better correction of thoracic kyphosis could be obtained by removing the disks. A 3.2-mm threaded rod was used in the early cases but was found to fail in nearly one-third of the patients. More recently, a 4-mm threaded rod has been used, without any breakage reported to date. Experience has shown that a shorter thoracic fusion can be more successfully accomplished with this system than with posterior instrumentation. Further success has been reported with anterior thoracic instrumentation using two thin rods on the convexity of the curve.[206]

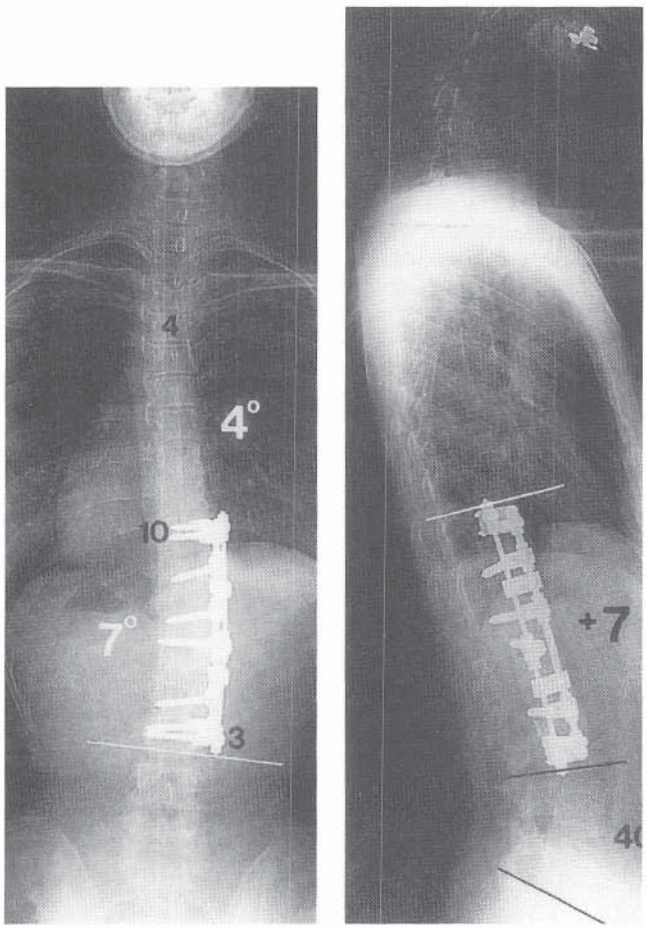


FIGURE 11–45 Kaneda anterior instrumentation. This two-rod system increases the stiffness of the construct, enhancing the likelihood of successful arthrodesis. (From Kaneda K, Shono Y, Satoh S, et al: New anterior instrumentation for the management of thoracolumbar and lumbar scoliosis: application of the Kaneda two-rod system. Spine 1996;21[10]:1250–1261.)

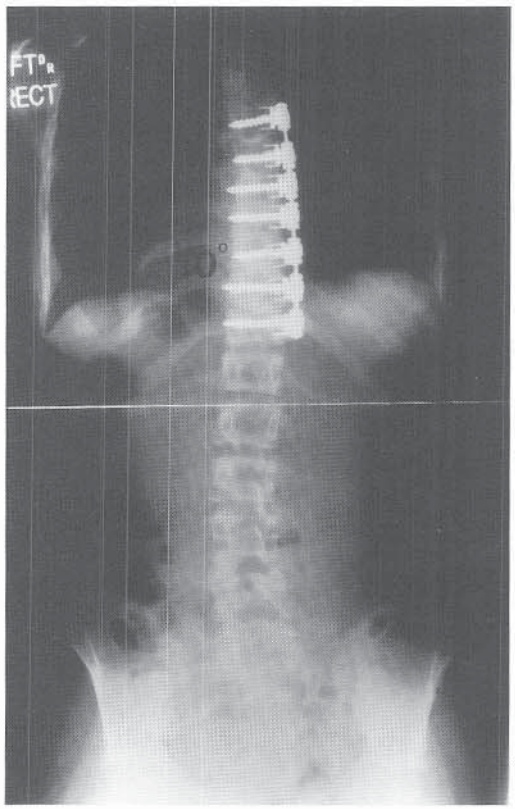


FIGURE 11–46 Anterior thoracic instrumentation. A threaded rod is attached to vertebral body screws on the convexity of the thoracic curve. A shorter segment of thoracic fusion can be accomplished with this system than with posterior instrumentation.

Infantile and Juvenile Idiopathic Scoliosis

Despite many advances in management, the etiology of infantile and juvenile idiopathic scoliosis, like that of adolescent idiopathic scoliosis, is unknown. There is no evident neuromuscular disorder clinically, and radiologically the vertebrae appear normal without congenital abnormality. The differentiation between these two entities is based on the age of the patient. Infantile idiopathic scoliosis is diag-

Text continued on page 276

Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis

OPERATIVE TECHNIQUE

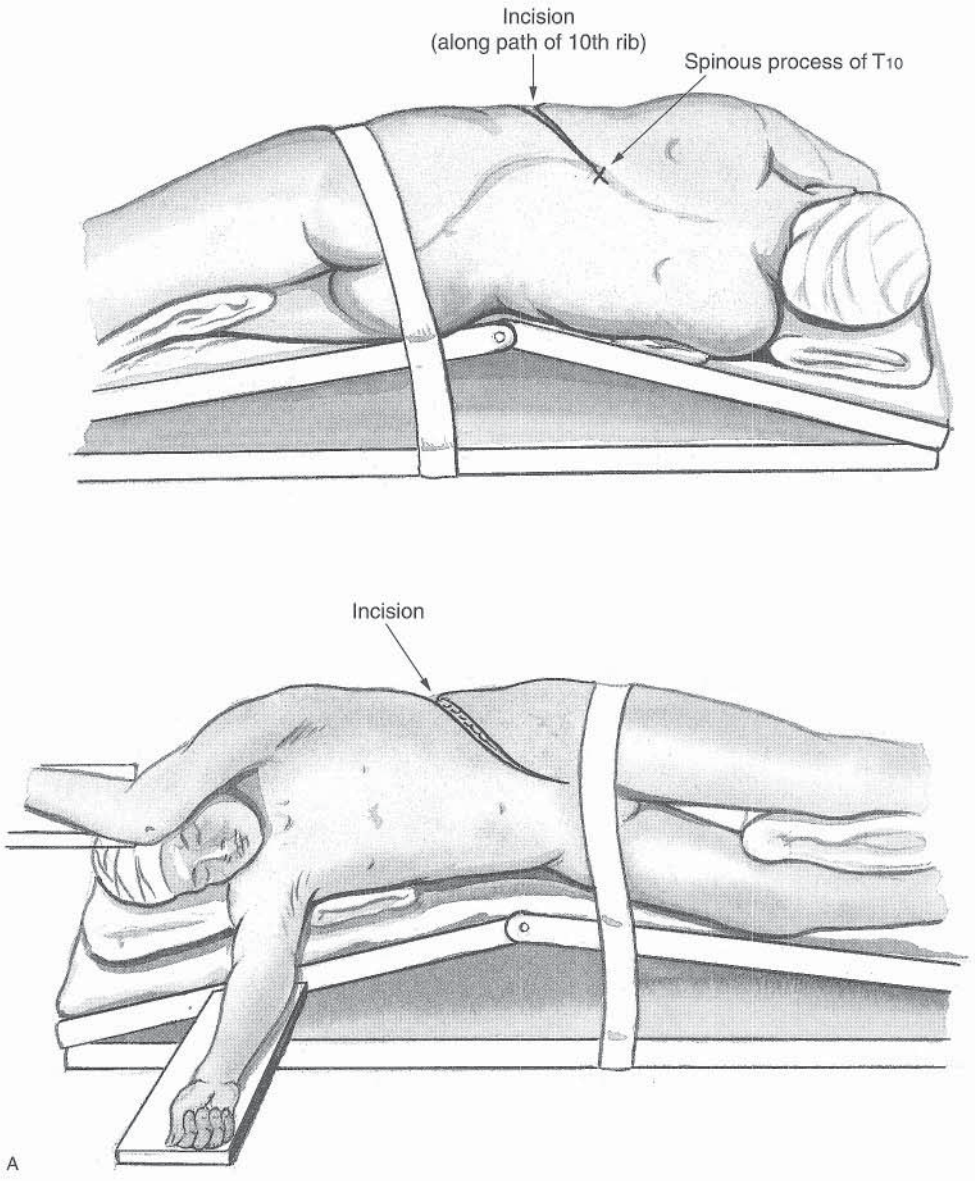
In this plate, the thoracoabdominal approach for exposure of the lower thoracic and lumbar spine is described.

A, *Positioning.* Under the direction of the surgeon, the patient is placed in the lateral decubitus position (convexity of the curve is upward). A roll is placed under the axilla of the dependent arm. The body is supported with a deflatable bean bag. The upper arm is flexed forward and slightly abducted. The operating table may be temporarily flexed (at the apex of the scoliosis) to facilitate excision of the intervertebral disks.

Approach. It will be necessary to remove a rib for exposure of the spine. Ideally, the rib that is removed is the one immediately cephalad to the uppermost vertebral body requiring instrumentation. For instrumentation between T11 and L3, removal of the tenth rib will allow adequate exposure.

Skin Incision. The incision begins lateral to the spinous process of T10 (or T9) and extends along the course of the tenth rib to the costocartilaginous junction, then across the upper abdomen to the lateral edge of the rectus abdominis. Here it turns distally toward the symphysis pubis, stopping at the level of the umbilicus.

PLATE 11–3 Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis



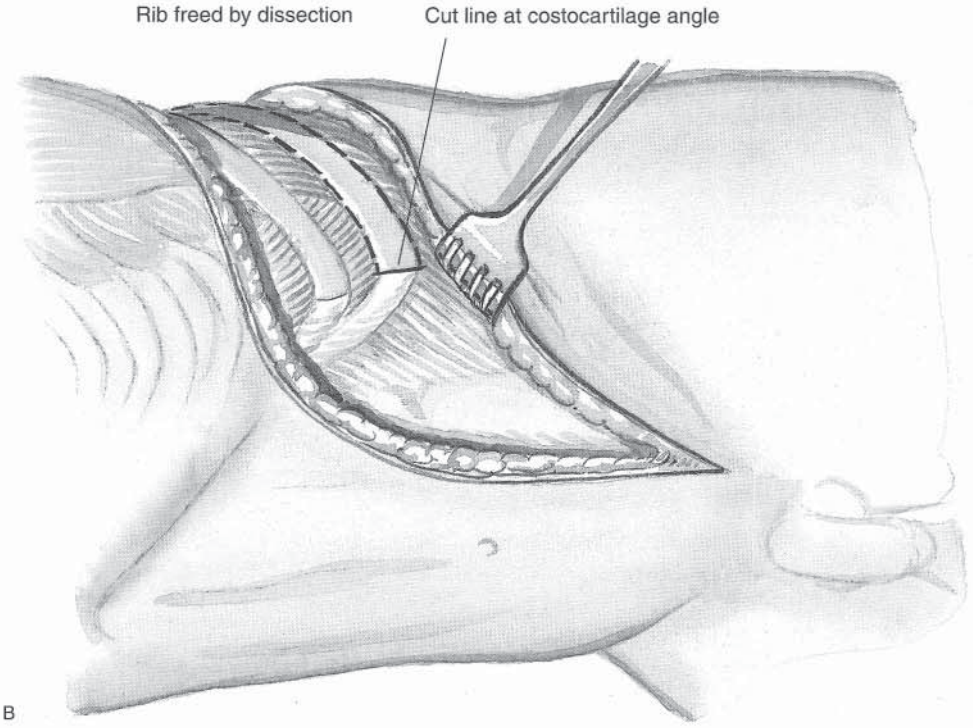
A

Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis *Continued*

B, The tenth rib is freed subperiosteally, divided at its costocartilaginous junction, and removed. This creates a larger working aperture and provides a source of autogenous bone graft.

C, Once the costal cartilage of the tenth rib is split, the retroperitoneal space is identified and entered.

PLATE 11–3 Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis



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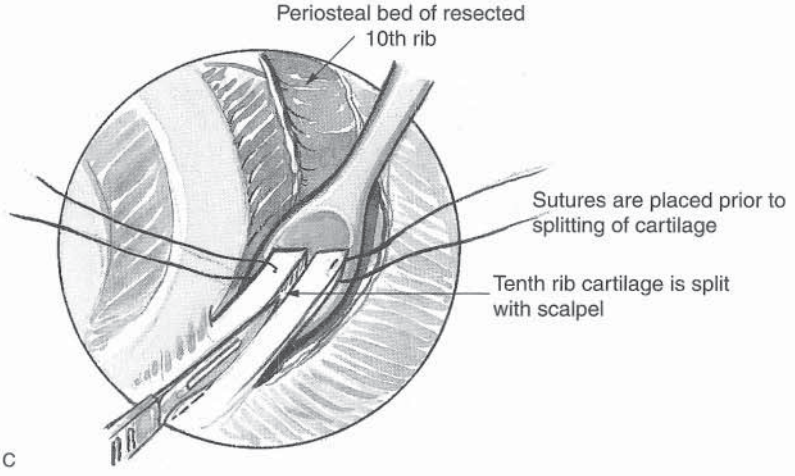


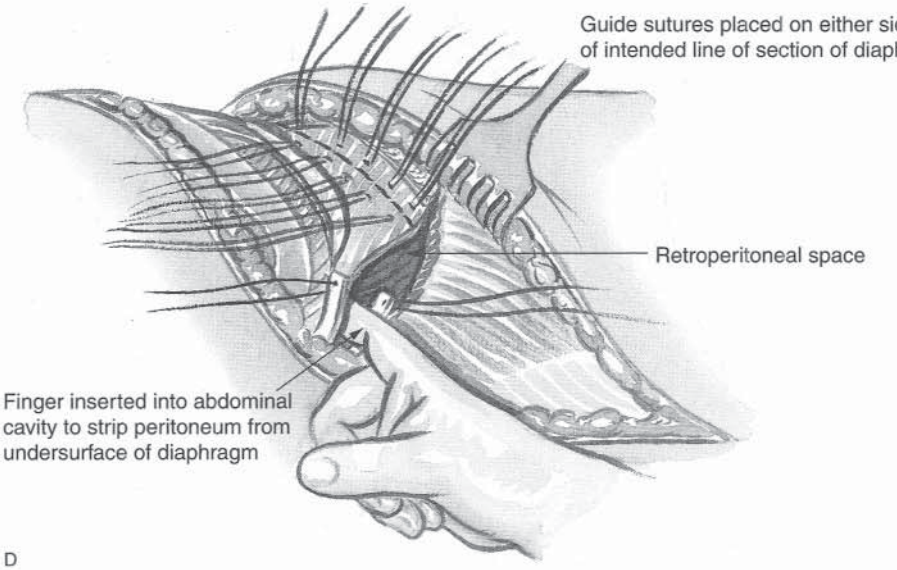
C

Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis *Continued*

D, Using blunt finger dissection, the operator separates the peritoneum from the inferior aspect of the diaphragm. Once freed, the viscera lie safely away from the vertebral bodies. Identification sutures are placed on either side of the intended line of division of the diaphragm, which is $\frac{1}{2}$ to $\frac{3}{4}$ inch from its periphery. Placement of several of these sutures will facilitate proper closure of the diaphragm later on.

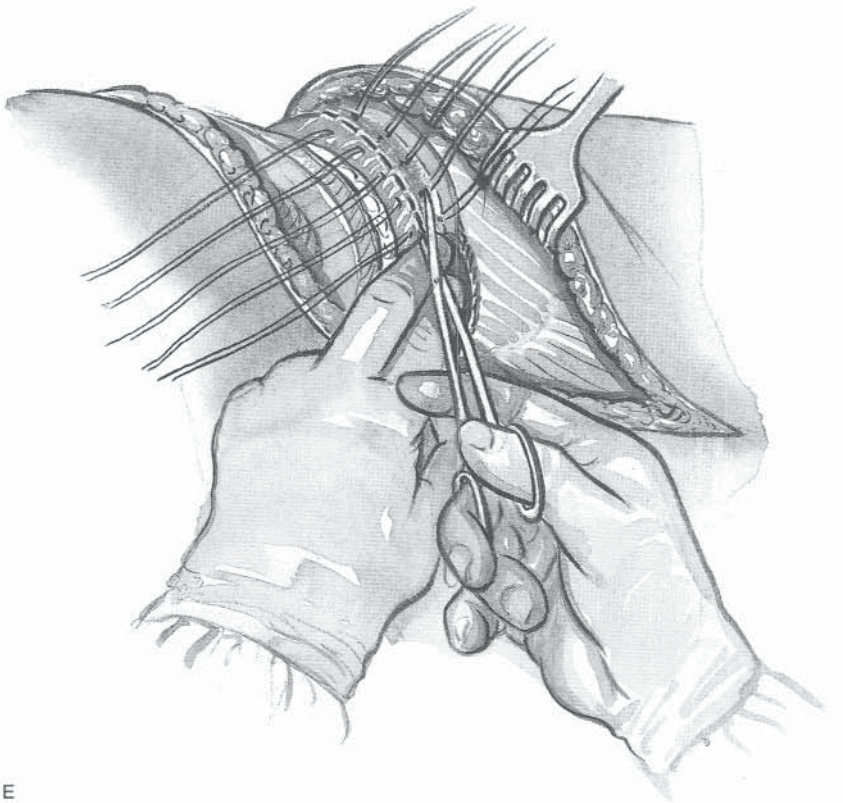
E, The diaphragm is sectioned from its costal attachments.

PLATE 11–3 Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis



D

Diaphragm (free from peritoneum on undersurface) is divided circumferentially 1.5 cm. from its costal attachment



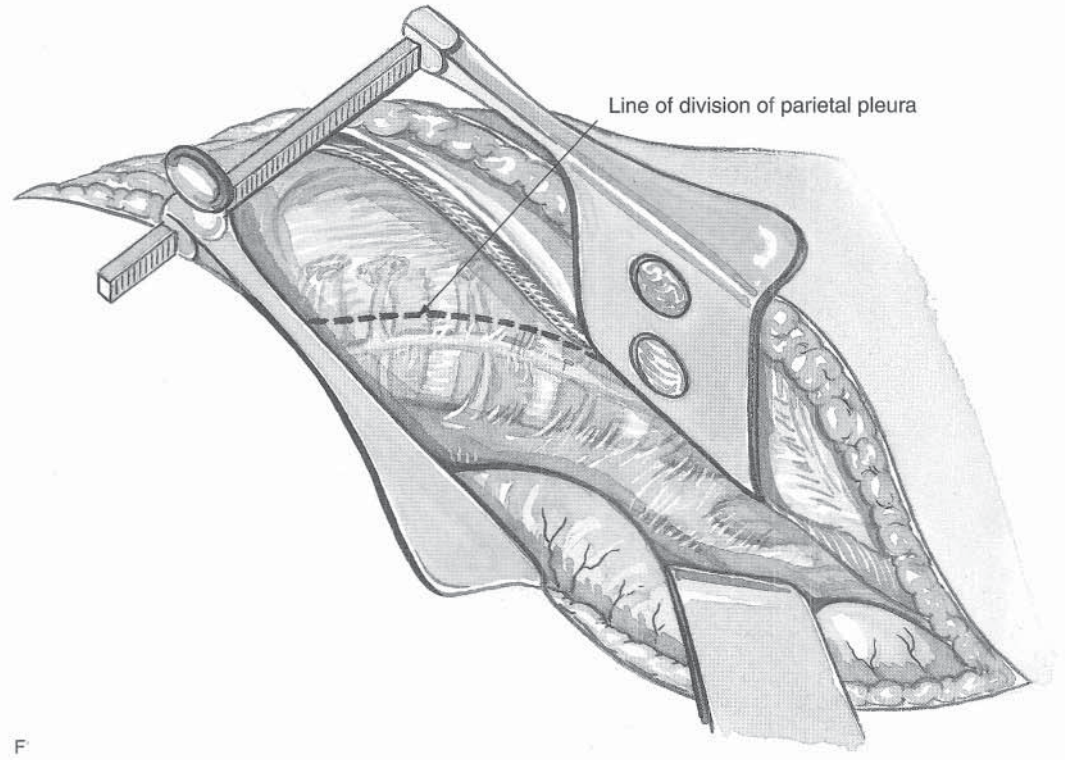
E

Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis *Continued*

F, Next, the parietal pleura is incised along the thoracic vertebral bodies that are to be included in the fusion.

G, In the lumbar region, the psoas muscle is gently elevated off the vertebral bodies and intervertebral disks and retracted posteriorly. The segmental vessels are ligated in the middle of each vertebral body included in the fusion. The aorta and vena cava are protected with retractors, and the anterior longitudinal ligament is partially excised with a sharp scalpel. Each disk within the levels selected for fusion is removed with various rongeurs and curets.

PLATE 11–3 Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis



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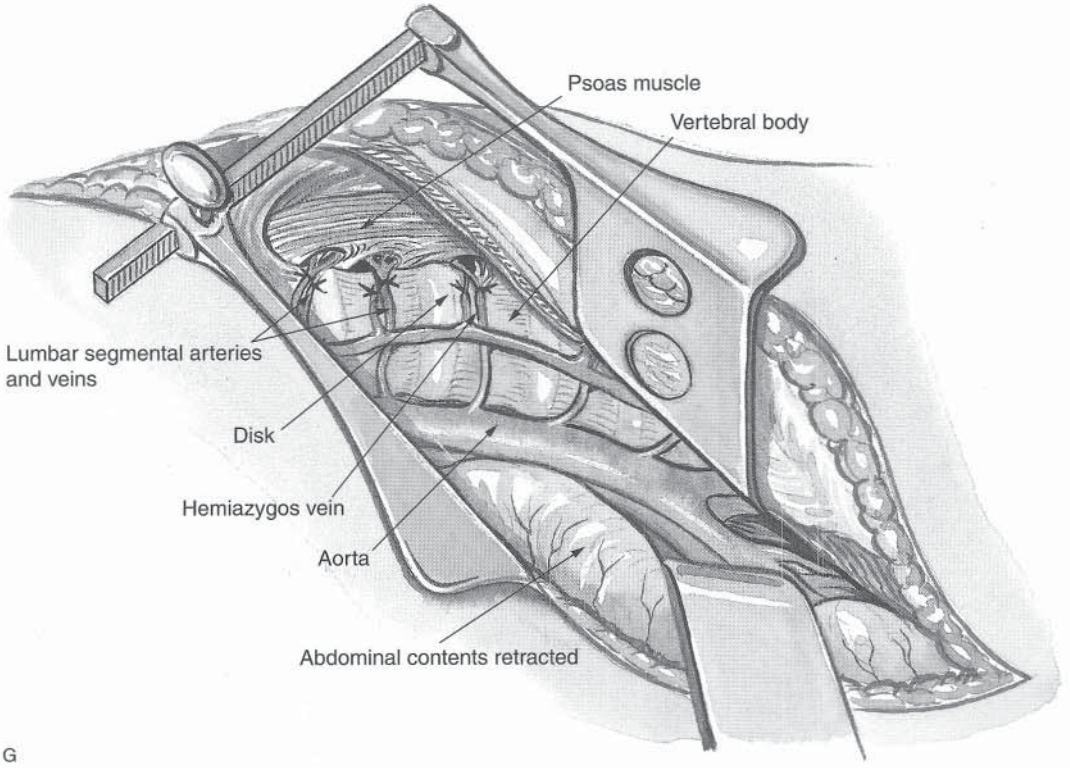
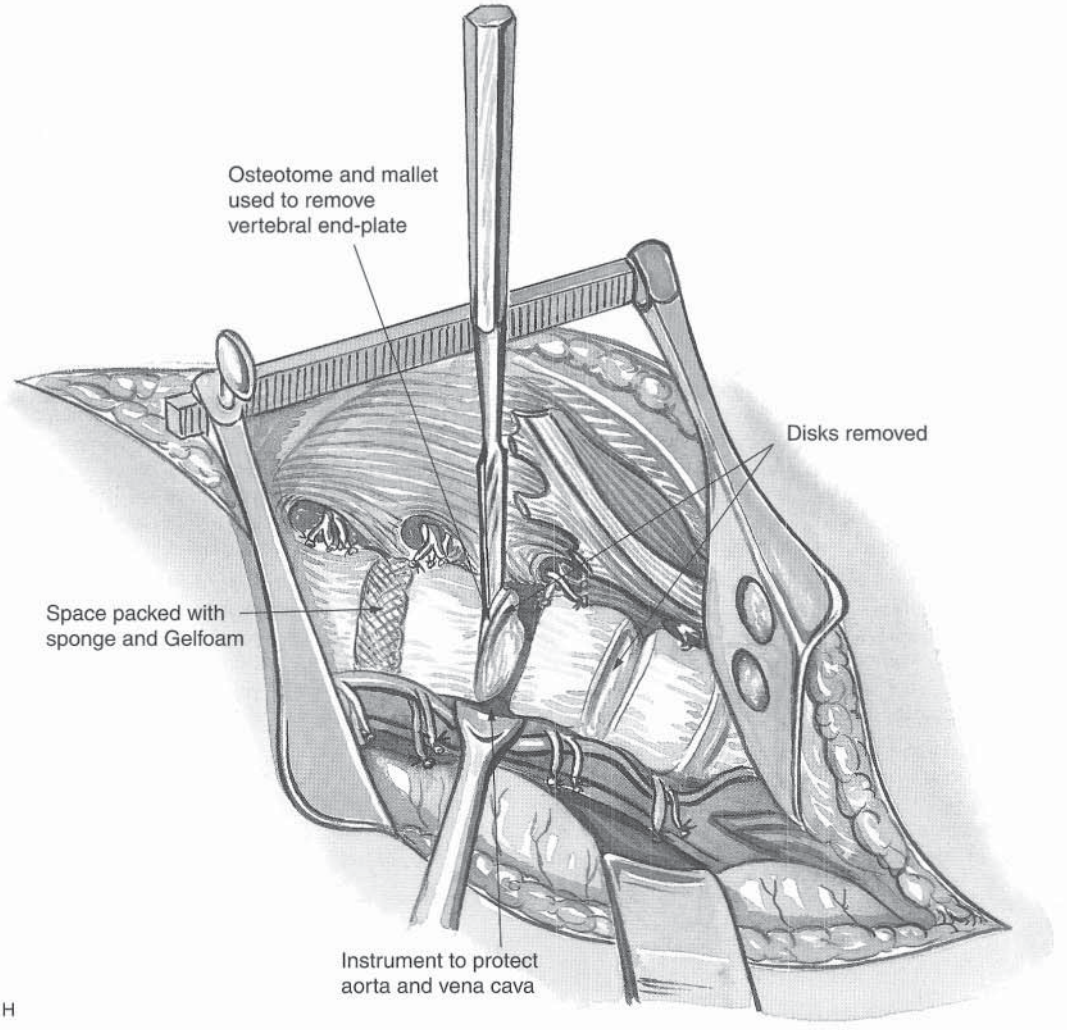


G

Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis *Continued*

H, With a curet or sharp osteotome and mallet, the operator removes the vertebral cartilaginous end-plates and retained pieces of disk. In correction of kyphosis, most of the annular ligamentous tissue down to the posterior longitudinal ligament is removed. In scoliosis, however, the outer annular fibers need not be fully removed. The disk spaces are then temporarily packed with Gelfoam to minimize bleeding. If the operating table was flexed to facilitate excision of intervertebral disks, it should be flattened at this time.

PLATE 11–3 Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis



H

Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis *Continued*

I, For the anterior instrumentation, screws should be supplemented with staples whenever possible. Biomechanical testing has documented a 50 percent increase in pull-out strength in axial load (compression or distraction) when a staple is used, compared to a disk washer with the screw or a screw by itself. A 6.5-mm screw should be used whenever possible to maximize stability.

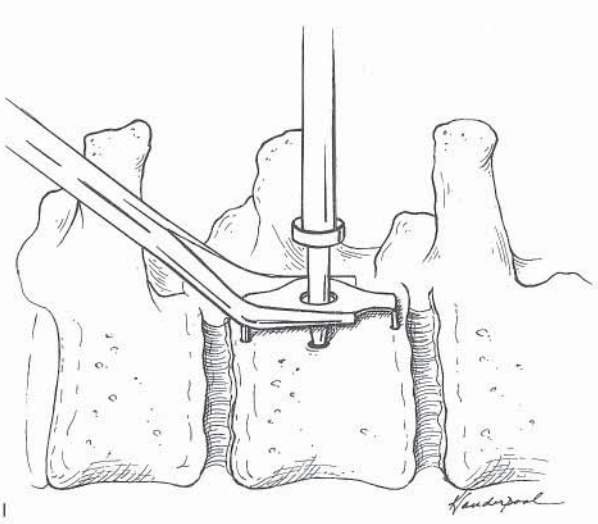
Screws should be placed in the posterior one-third of the vertebral bodies, and the appropriate-sized staple should be used as a template to start the hole for the vertebral body screw. Prongs of the staple should go into the bony end-plate. It is sometimes helpful to use a small osteotome to cut the cortex of the vertebral body at the end-plates to get the prongs of the staple started. The awl can then be used to start the hole for the vertebral screw. The staple should be impacted into place using the awl. To make the staple flush with the lateral vertebral surface, it is sometimes helpful to "shave" the cephalic and caudal bony prominences next to the disk spaces with a rongeur. It is not necessary to drill or tap a vertebral body because the screws are self-tapping.

J, Each screw should be long enough for its tip to penetrate the far cortex. The correct length can be estimated using a depth gauge placed through the adjacent disk space to the opposite vertebral body cortex.

It is helpful to palpate the tip of the screw exiting the opposite vertebral cortex when the final threads are tightened. This requires a relatively circumferential exposure of the vertebral body. If the disk space is adequately visualized and the direction of the screw confirmed by comparing its path to an imaginary path through the disk space, there should be little concern about the safety of the tip protruding on the opposite side.

K, The screws should be placed in a relatively straight line cephalad to caudad. There is no need to offset the screws for better correction, as the corrective forces imparted to the spine come from the contouring of the solid rod. The rod should be contoured to obtain the desired lordosis in the lumbar spine. In the typical case, maximum lordosis will be bent into the rod between L1 and L3, with a relatively straight segment from T11 to L1 (the thoracolumbar junction should be essentially straight).

PLATE 11–3 Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis



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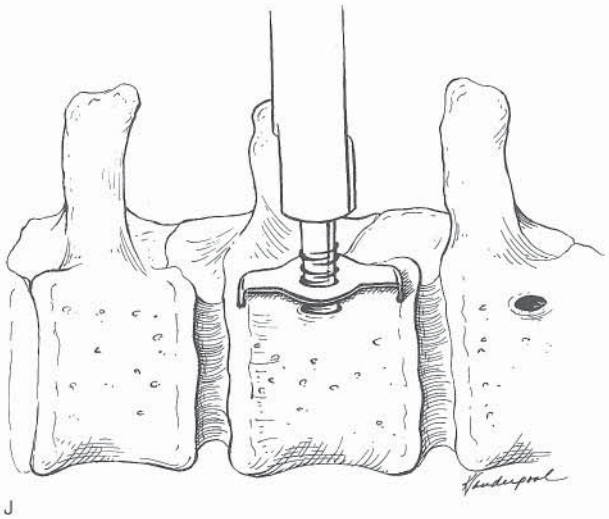

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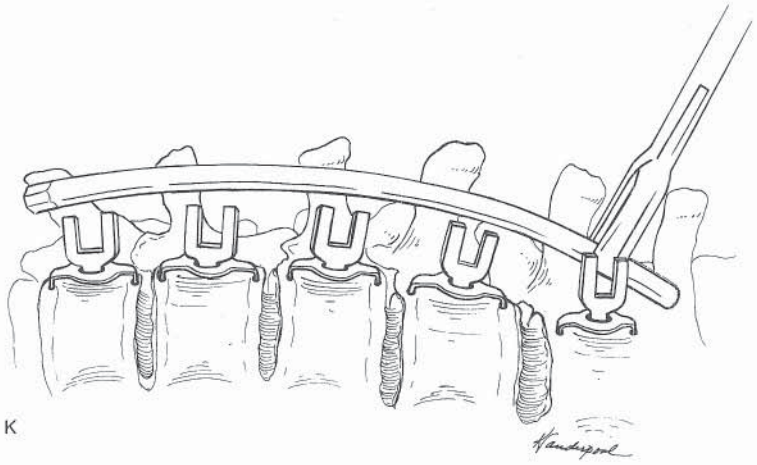


K

Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis *Continued*

L, Eyebolts are then placed on the rod and the rod is seated in each screw successively. The rod must be cut very accurately, to avoid excessive length protruding distal to the most caudal screw or proximally into the chest. The hex-end side of the rod (needed to facilitate rotation) is directed toward the patient's head. The rod is usually seated at the most caudal screw first and then seated successively in each more proximal screws. As each screw-eyebolt attachment is completed, the nut should be tightened slightly so that the rod will not displace from the screw head goal posts during the remainder of the assembly maneuvers.

The rod should be placed posterior to the screw heads, with the nuts facing toward the abdominal contents. This allows the nuts to be tightened quickly using the T-handled torque wrench.

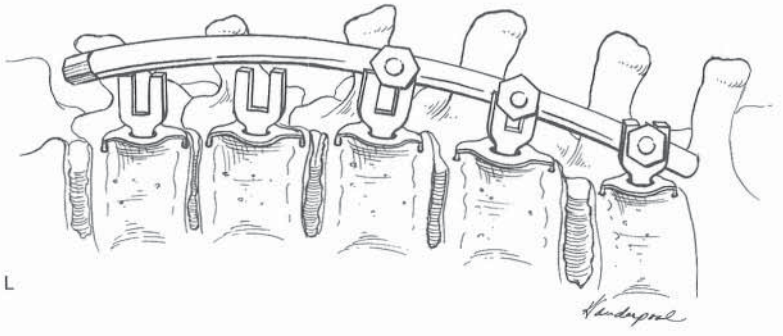
M, After all eyebolts have been seated in the screw heads, rotational correction of the scoliosis is accomplished by rotating the rod 90 degrees into lordosis. This maneuver is most easily accomplished using the hexagonal-end wrench. Depending on the amount of contouring of the rod and the diameter of rod used, this maneuver usually completely corrects a moderately sized lumbar curve.

The 6.4-mm "flexible" rod is used for the routine idiopathic lumbar curve. This rod has proved to have satisfactory stiffness such that it will impose a lordotic correction on the typical scoliosis curve.

N, After the rotation maneuver, the disk spaces are noticeably opened anteriorly as a result of the increased lordosis. At this point, all of the evacuated disk spaces should be packed with the autogenous rib bone. Several pieces of rib should be cut into 5- to 10-mm segments. One of these segments is placed inside the anterior edge of each disk space (to form a strut). This is done to ensure that the lordosis of each disk space is maintained following mild compression between screws. An alternative method to maintain lumbar lordosis is to place a metal mesh cage into the inferior-most disk space. Additional bone graft (allograft or iliac crest graft) may be needed to fill the disk spaces. After the bone graft insertion, a very small amount of compression should be applied segmentally to lock the bone graft into place, and the nuts tightened using the torque wrench. We usually gently compress the end of the construct toward the middle screw. Excessive compression should be avoided to minimize the chance of creating kyphosis in the instrumented segment.

O, Prior to final tightening of any nut, the operator confirms that the rod is properly placed in the groove of the screw head. If the rod is not properly seated (*left*), no amount of nut tightening will ensure a stable connection. If the rod has popped up slightly during the rotation maneuver, particularly at the end vertebra, it should be reseated after rotation but prior to final tightening, using a screw holder and corkscrew device. Only when the rod is ascertained to be precisely in the grooves of the screw head (*right*) should the nut be finally tightened to 150 inch-pounds of torque.

PLATE 11–3 Anterior Instrumentation of the Spine for Thoracolumbar or Lumbar Scoliosis



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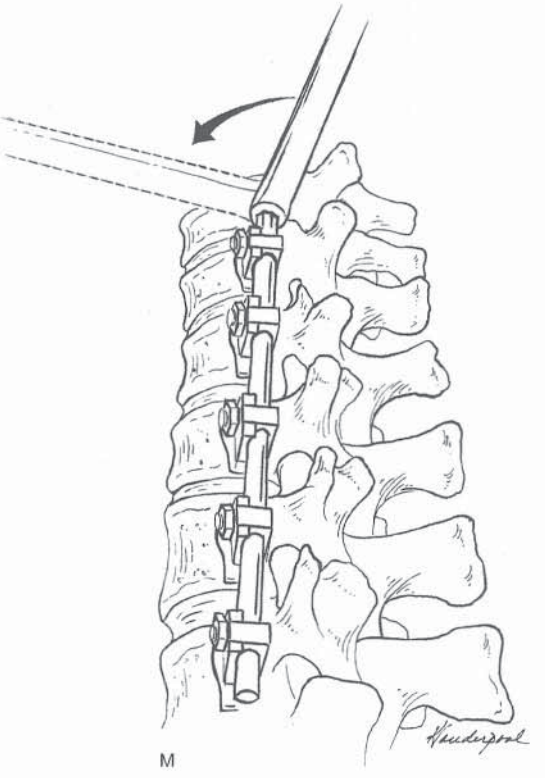

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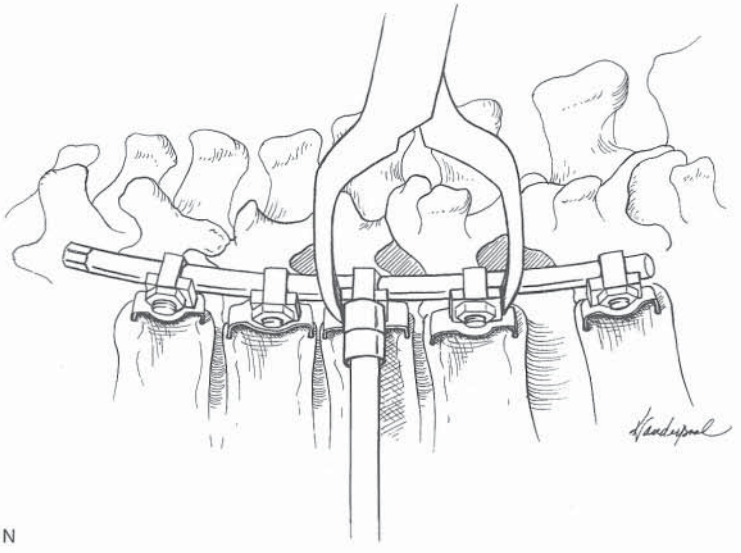

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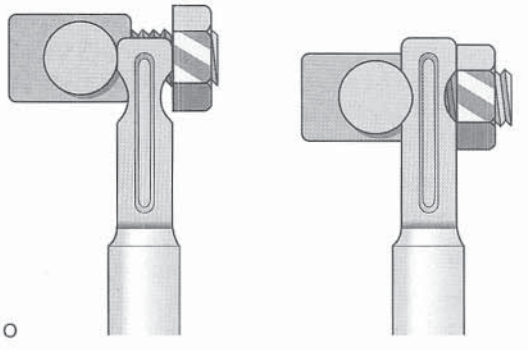

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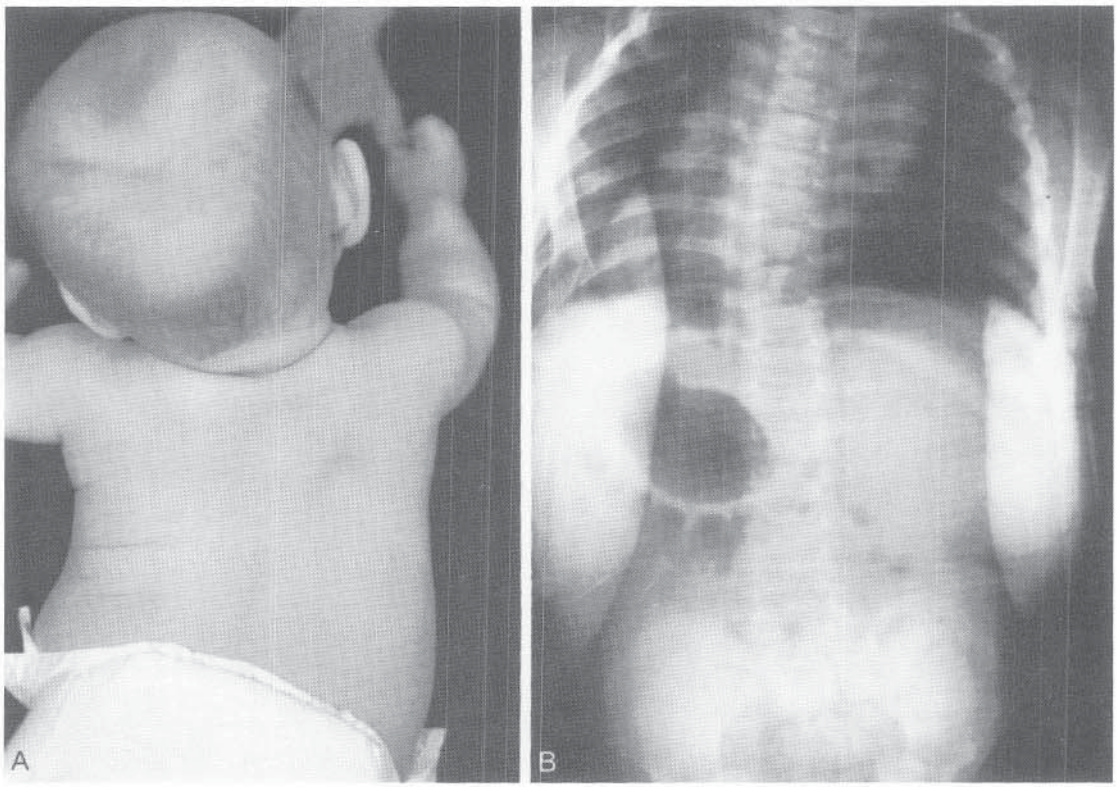


FIGURE 11–47 Infantile scoliosis in a 6-month-old infant. **A,** Plagiocephaly of the head. **B,** AP radiograph of spine showing scoliosis.

nosed in those less than 3 years old. Patients ages 3 to 10 years are classified as having juvenile idiopathic scoliosis.

INFANTILE IDIOPATHIC SCOLIOSIS

Infantile idiopathic scoliosis is more common in boys than in girls and is more common in Europe than in North America. In a 1973 report from Massachusetts, infantile curves accounted for only 0.5 percent of idiopathic curves.[379] During a similar period in Great Britain, infantile curves accounted for nearly 41 percent of reported idiopathic scoliosis. However, with the passage of time, the relative frequency of infantile scoliosis in Europe has declined and now represents only 4 percent of those with idiopathic scoliosis.[290] The authors attributed this decrease to a change in patient position (from supine to prone) during sleep.

Clinical Features. The majority of children with infantile idiopathic scoliosis are diagnosed within the first 6 months of life. Most have left-sided thoracic curve patterns (Fig. 11–47).[501] Plagiocephaly is a common association and is attributed either to the position assumed during sleep or to intrauterine molding.[225,248,433]

Natural History. Many curves resolve over time without treatment, but there is controversy about the frequency of spontaneous resolution.[195,197,248] In one study, 92 percent of curves spontaneously resolved,[248] while another source reported spontaneous resolution in only 20 percent of cases.[195]

Methods for Predicting Progression. In an effort to distinguish between curves that were likely to worsen and those that were likely to resolve spontaneously, Mehta in 1972 developed a useful radiographic measurement known as the rib-vertebral angle difference (RVAD).[295] This measurement, shown to be reproducible and valid, described a relationship between the apical vertebra (of the thoracic curve) and its ribs (Fig. 11–48).[226,283] The angle is formed by a line drawn perpendicular to the end-plate of the apical vertebra and a line drawn along the center of the rib. The RVAD is calculated by subtracting the angle value of the convex side from the concave side. The difference was found to be useful in predicting curve behavior. In Mehta's report, 83 percent of the curves that resolved had an initial RVAD measuring less than 20 degrees, whereas 84 percent of the curves that

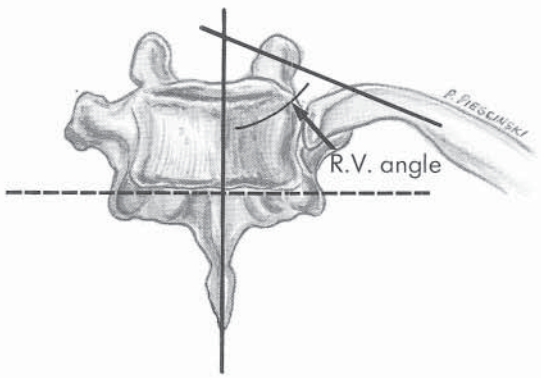


FIGURE 11–48 Method of measuring Mehta's rib-vertebral angle (RVA). A perpendicular line is drawn to the end-plate of the apical vertebra. Next, lines bisecting the head and neck of the ribs on each side of the apical vertebra are drawn. The angle formed by the intersection of these two lines is the RVA.

progressively worsened had an RVAD exceeding 20 degrees. Today, this point differentiation of 20 degrees continues to be a useful parameter.

Mehta also described a phase 1 and a phase 2 radiographic appearance, the phase depending on the position of the rib head (Fig. 11–49). In phase 1, the rib head on each side of the apical vertebra does not overlap the vertebral body. In phase 2, the rib head overlaps the convex side of the vertebral body. In phase 2, progression of the infantile curve is certain and measurement of the RVAD is unnecessary.

Mehta further subdivided progressive infantile idiopathic scoliosis into benign and malignant forms.[295] Each form demonstrated rapid worsening in the first 5 years of life, a more gradual progression between 5 and 10 years of age, and then marked deterioration during the adolescent growth spurt. The malignant form was distinguished by more severe progression early on, resulting in greater difficulty in management.

Factors Influencing Curve Progression. Factors that influence curve progression in infantile scoliosis include the patient's age at onset, the magnitude of the curve at initial assessment, and an association with developmental problems.[433] Curves that develop during the first year of life have a greater likelihood of resolving spontaneously, whereas curves developing after 1 year of age have a poorer prognosis.[248,290] Larger curves will most likely progress, but even curves less than 20 degrees must be carefully followed. Developmental problems, especially mental deficiencies, are more prevalent in children with progressive curves.[80,81] Other abnormalities found in association with infantile scoliosis include developmental dislocation of the hip, congenital heart disease, congenital ventral hernias, and prematurity.[185,415,501,502]

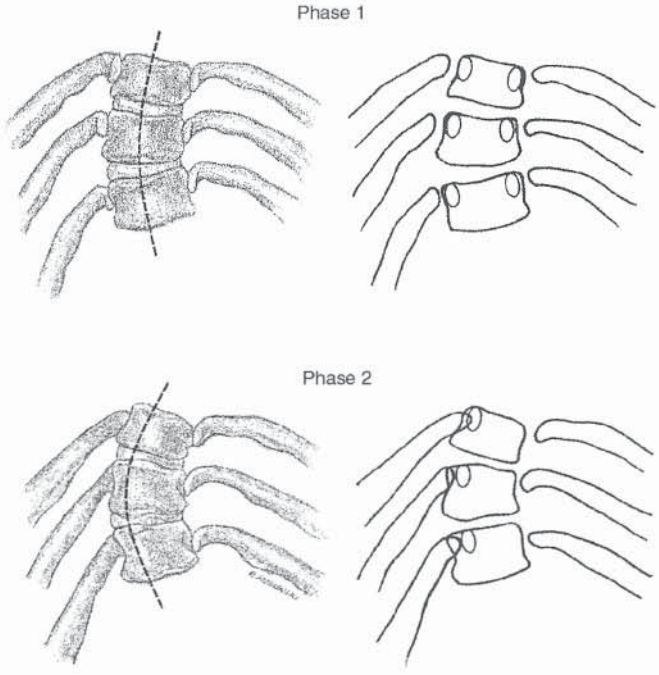


FIGURE 11–49 The two phases in progression of infantile scoliosis as seen on the PA radiograph. In phase 1, the rib head on the convex side does not overlap the vertebral body. In phase 2, the rib head on the convex side overlaps the vertebral body.

Neural Axis Abnormalities. Recent reports have shown an increased incidence of neural axis abnormalities in patients with infantile and juvenile scoliosis.[162,244] Although the numbers reported are very small, the incidence of neural axis abnormality (on MRI) in those with infantile curves may approach 50 percent. If the curve is progressive, MRI of the entire spine is recommended. Most, if not all, of these patients will require sedation for MRI.

Management of Curves That Resolve. If at the time of initial evaluation the curve is less than 25 degrees and the RVAD is less than 20 degrees, active treatment is not required and radiographic follow-up should be accomplished every 4 to 6 months.[225] Having the infant sleep in the prone position rather than supine may be of benefit. Most infantile curves that resolve do so by age 1 to 2 years.[71,248,290,440] Occasionally, several years may pass before the curve has resolved.[455] Follow-up should continue even after resolution, though, because scoliosis may recur in adolescence.

Nonoperative Treatment for Progressive Curves. Nonoperative treatment should be undertaken without delay in those with infantile scoliosis and demonstrated progression of the curve. Left untreated, these curves can easily exceed 70 degrees by age 10 years. Further worsening can occur during the periods of rapid adolescent growth. The goal of brace treatment is to control curve progression until patient size and skeletal growth have been achieved, to allow a one-time spinal stabilization procedure to be performed. Parents should have a clear understanding early on that operative intervention is almost always inevitable. A curve that resolves in a brace would probably have resolved without treatment.

For the young child with a flexible curvature, the physician should use either a Boston TLSO or a modified Milwaukee brace, preferably worn full-time. Successful management requires strong parental support, along with frequent adjustments by the orthotist.

Occasionally a young child with infantile scoliosis is not seen by a physician until the scoliotic deformity is large and inflexible. In this situation a corrective cast is needed prior to bracing. Casts are usually applied under general anesthesia, with the use of longitudinal traction and lateral pressure placed over the apex of the curve. Two or three serial casts may be necessary (changed every 2 to 4 weeks) to gain sufficient correction to allow a thermoplastic orthosis to then be used. This treatment approach may have to be repeated over the course of several years. On rare occasions, consideration must be given to the use of halo traction (progressively increasing the amount of distractive forces) to allow the initially inflexible spine to correct sufficiently that a corrective cast can be applied, followed by the program described.

Operative Treatment. Surgical treatment is indicated for curves that cannot be controlled by serial casting or orthotic treatment. Fortunately, an aggressive nonoperative approach will usually allow delay of operative considerations until the child has reached the age of 7 or 8 years. By that time, definitive operative stabilization can be considered.[371] Treatment then usually consists of an anterior diskectomy and fusion, followed by posterior instrumentation with fusion. With this method, significant curve correction is achieved and the potential problem of the crankshaft phenomenon

is avoided.[175] This is the author's preferred treatment method (Fig. 11–50). Occasionally, preoperative halo traction over a period of 6 to 10 weeks may be necessary before surgery in the young child with a rigid, severe curve.

Other surgical alternatives can be considered. Anterior and posterior hemiepiphysiodesis on the convex side of the curve has been performed in an attempt to allow the unfused concave portion of the spine to gradually correct the deformity. This technique has been reported to be successful in patients with congenital short-segment scoliosis caused by hemivertebra; however, the approach is much less successful in the young patient with idiopathic scoliosis. A recent study on convex spinal epiphysiodesis for managing progressive infantile idiopathic scoliosis (mean age at surgery was 6

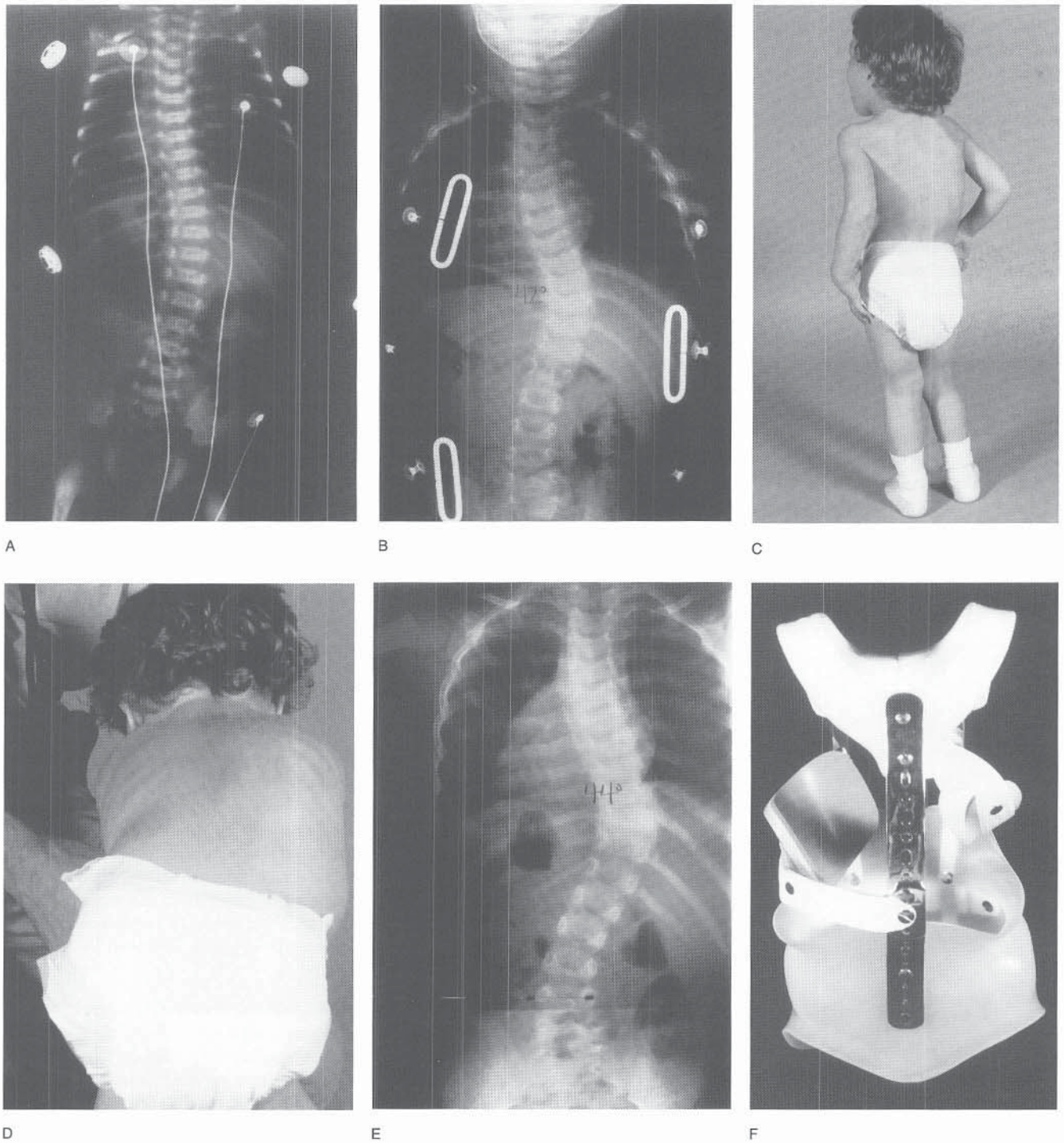


FIGURE 11–50 Treatment of infantile scoliosis. Radiograph of the spine of a newborn infant demonstrated a mild right lower thoracic idiopathic scoliosis (**A**). On presentation at age 13 months, the rib-vertebral angle measured 27 degrees (phase 1). At age 16 months, the curve measured 42 degrees with the child in a brace (**B**). At age 3 years 8 months, scoliosis was clearly evident clinically (**C, D**) and measured 44 degrees radiographically (**E**).

years) found that patients who underwent epiphysiodesis without instrumentation exprienced progressive worsening of the curvature.[278] When hemiepiphysiodesis is combined with instrumentation that is repeatedly lengthened, the results have been somewhat better.

Another option, subcutaneous instrumentation without fusion, has been used in young children in an effort to control progressive spinal deformity while temporizing the eventual need for posterior arthrodesis. Proponents of this approach report success despite lengthy treatment courses,

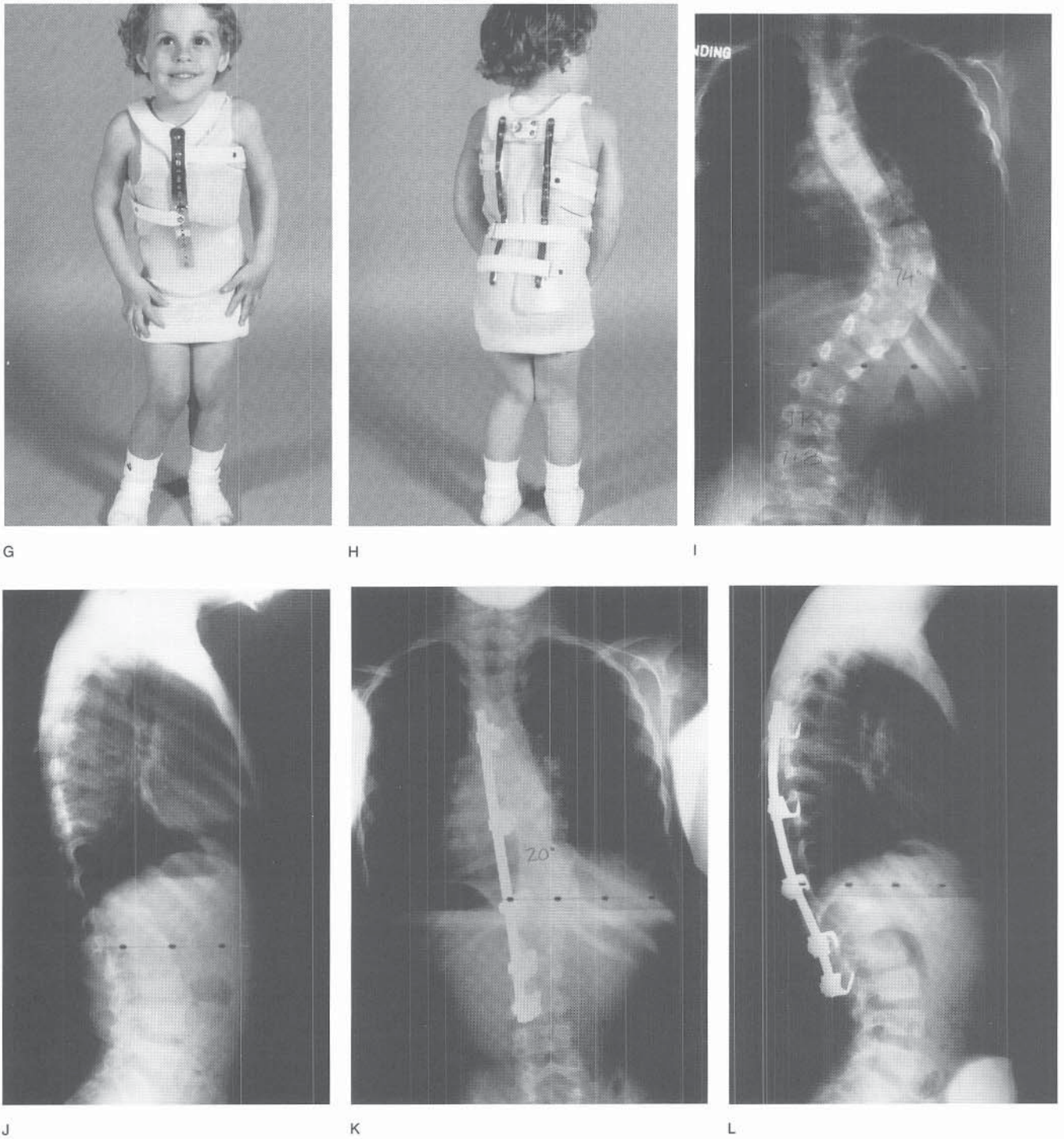


FIGURE 11–50 *Continued.* Full-time use of a modified Milwaukee brace (**F** to **H**) during that time appeared to be beneficial. Unfortunately, despite compliance with the brace program, the curve increased to 74 degrees by age 7 years 10 months (**I, J**). Preoperative MRI was normal. Anterior diskectomy and fusion (T7–L1) followed by posterior instrumentation and fusion (T5–L2) was then carried out during the same anesthesia episode. One year later, the curve correction was maintained at 20 degrees and sagittal balance was normal (**K, L**). Aggressive nonoperative treatment usually allows surgery to be delayed until the child is 7 or 8 years old.

repeated operations for distraction along the spine, and frequent instrumentation-related difficulties.[223,263] In a recent study, 67 children underwent initial instrumentation at an average age of 7.8 years.[223] Over a period of 3.2 years, an average of 4.4 repeat distractions or rod replacements were performed. As a result, the average longitudinal spinal growth within the instrumented area was 3.1 cm (i.e., 1.2 cm per year). Apical anterior growth arrest, in conjunction with posterior instrumentation without fusion, has led to better reported outcomes.[340] Unintentional spontaneous fusion along the posterior spine can occur following instrumentation without arthrodesis.[134] The reported benefits of these treatment methods must be carefully scrutinized today, particularly when compared with the results of combined anterior fusion and posterior instrumentation with fusion. The combined fusion technique allows for significant curve correction, immediate gain in spinal height, prevention of progressive deformity, and minimal complications.

JUVENILE IDIOPATHIC SCOLIOSIS

Juvenile idiopathic scoliosis is diagnosed in children ages 3 to 10 years. For the population affected by idiopathic scoliosis (infantile, juvenile, and adolescent), the prevalence of the juvenile form is 8 to 12 percent in Europe and 13 to 16 percent in the United States.[195,384] The deformity usually is recognized clinically by age 6 to 7 years.[276] The female-male ratio ranges from 1.6:1 to 4.4:1, with the difference increasing with age.[132,384] Convex-right thoracic curve patterns are most common in juvenile scoliosis. Relatively few patients have thoracolumbar or lumbar curves.

Natural History. Juvenile scoliosis usually progresses slowly during the period of steady spinal growth (ages 5 to 10 years). After 10 years, progression is more rapid. A recent study reported curve progression in 95 percent of children diagnosed with juvenile idiopathic scoliosis.[384] Of those patients followed to maturity, 86 percent of them required spinal fusion.

Predicting Curve Progression. Unlike in infantile scoliosis, use of the RVAD has not been found to predict progression of curves in juvenile scoliosis.[442] Patients with progressive curves have a steady increase in the RVAD, while those whose curves will resolve usually show a decrease in the RVAD. If the RVAD does not improve following bracing of a progressive curve, then it is likely that spinal fusion will be required as definitive treatment. The level of the most rotated vertebra at the apex of the primary curve appears to be the most useful factor in determining the prognosis of patients with juvenile idiopathic scoliosis. Those with the apex of the curve at T8, T9, or T10 have an 80 percent chance of requiring spinal arthrodesis by age 15 years.[384] The predictive value of two other factors once thought to be associated with a poor prognosis—thoracic kyphosis of less than 20 degrees, and left-sided curves in boys—has recently been questioned.

Neural Axis Abnormalities. Recent MRI studies have provided more insight into juvenile idiopathic scoliosis.[122,162,244,397,423] The incidence of neural axis abnormalities in these patients is 18 to 26 percent. Most of these children are asymptomatic and have no physical signs (other than scoliosis) of an underlying neural axis abnormality. The MRI abnormalities include Chiari type I malformations with cervical syrinx, thoracic syrinx, brain stem tumor, dural ectasia, and a low-lying conus. Many of these abnormalities may benefit from neurosurgical treatment. As a result, some authors have recommended MRI during the initial evaluation of patients presumed to have juvenile idiopathic scoliosis. If scoliosis surgery is planned, it is imperative that MRI evaluation be undertaken preoperatively. Neurologic deficit following spinal surgery has been reported in patients with a neural axis abnormality that was not recognized preoperatively.[323]

Treatment. It may be difficult to differentiate infantile from early juvenile scoliosis in the 4- to 5-year-old child who presents with idiopathic scoliosis. This is particularly true for boys with single left thoracic curves. Many of these curves may be large at the time of initial evaluation; if so, active treatment should be started immediately.

Generally, curves less than 25 degrees require only observation, with a return visit scheduled in 4 to 6 months. If there is 10 degrees or more of progression in curves initially less than 20 degrees or if there is 5 degrees of progression in curves intially between 20 and 25 degrees, active treatment should be started. Larger curves should be treated immediately.

Nonoperative management is similar to that described for children with infantile idiopathic scoliosis. Most curves will require brace treatment. Some of the less flexible larger curves may require cast correction at the beginning.[296]

Operative management would follow recommendations similar to those delineated for the child with infantile idiopathic scoliosis who has a progressive severe deformity (Fig. 11–51).

Congenital Spine Deformities

Congenital deformities of the spine are caused by anomalies in the growing vertebrae. These anomalies may be subtle and found incidentally on radiographs obtained for some other reason, or they may be complex and lead to severe spinal deformity with accompanying neurologic deficits. Congenital scoliosis, congenital kyphosis, or a combination of the two are the deformities encountered. They are generally much less common than idiopathic scoliosis.

ETIOLOGY

The cause of congenital vertebral anomalies remains unknown. During embryologic development, these abnormalities develop in the spine between the fifth and eighth weeks of gestation, but it is very uncommon to identify any traumatic or teratologic type of maternal insult during this stage of pregnancy.

Recent research has found that carbon monoxide exposure with its resultant hypoxia has led to reproducible congenital spinal deformities in mice offspring.[250] These deformities include wedged, hemi-, fused, and missing vertebrae, as well as fused ribs. The severity of the deformities was found to be related both to the dose of carbon monoxide and to the gestational time when exposure occurred. Correlating with this basic science study, the same institution reported

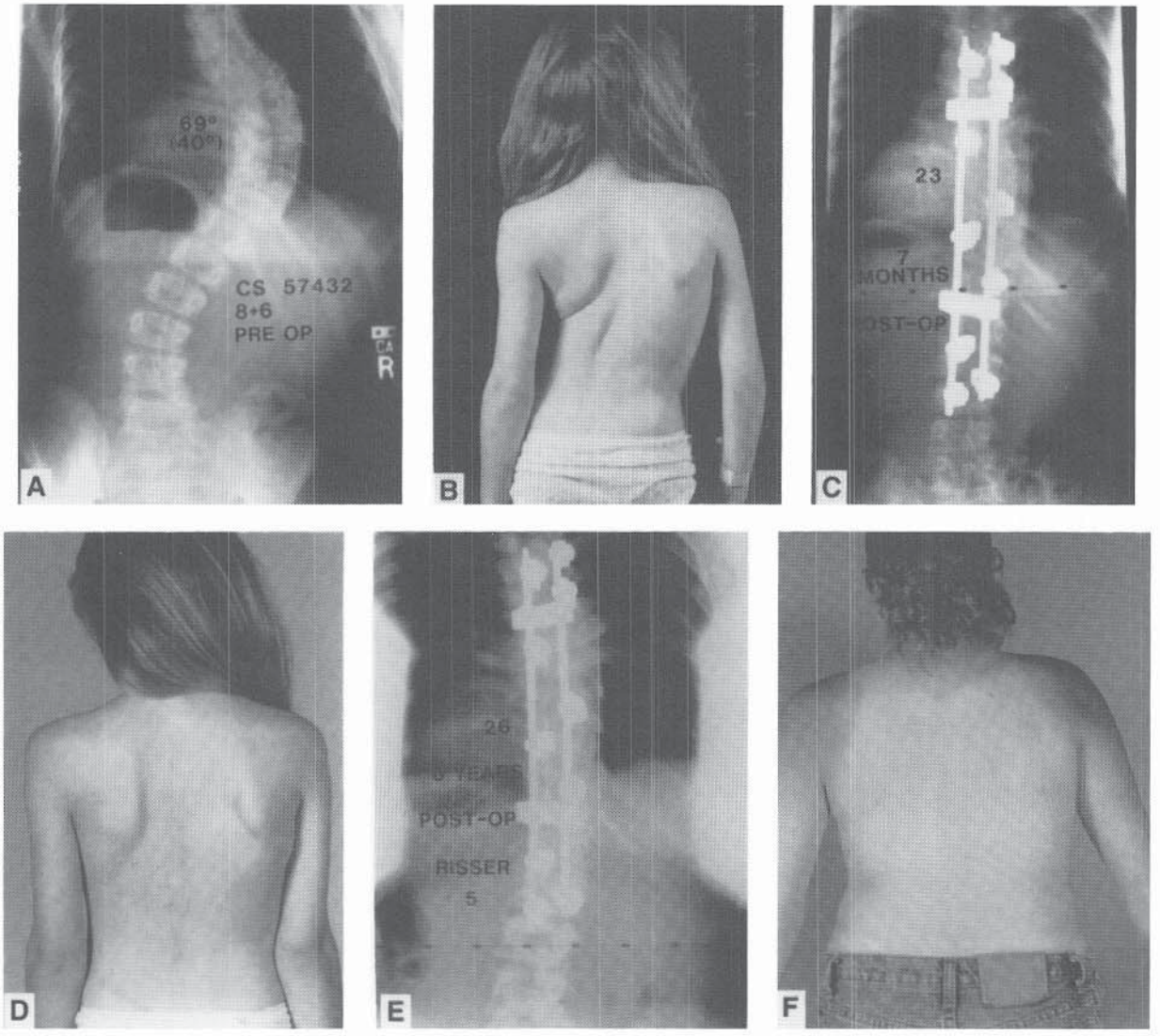


FIGURE 11–51 Treatment of juvenile scoliosis. A skeletally immature patient underwent anterior and posterior spinal fusion to prevent progressive changes caused by the crankshaft phenomenon. **A** and **B,** Preoperative radiographic and clinical appearance at age 8 years 6 months. **C** and **D,** Appearance 7 months following surgery. **E** and **F,** Appearance 6 years following surgery. Although the trunk was slightly shorter than normal, it was without deformity. (From Richards BS: The effects of growth on the scoliotic spine following posterior spinal fusion. In Buckwalter JA et al (eds): Skeletal and Growth Development: Clinical Issues and Basic Science Advances, p 585. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1998.)

clinical data that indicated a potential increase in exposure to fumes (chemical fumes and carbon monoxide) in mothers of children with congenital spine deformities.[249]

Investigation into genetic causes has provided little insight. A positive family history can be found in approximately 1 percent of patients with congenital spinal deformities.[473] An isolated anomaly, such as a hemivertebra, usually occurs as a sporadic event and carries no risk of a similar abnormality in further offspring.[499] Studies on identical twins of whom one twin was affected have shown no genetic pattern.[174,352,358] Only recently has a report of siblings with similar lumbar congenital deformities suggested the possibility of genetic causes.[136] Scientists have identified the human gene HuP48, a member of the *PAX* family of developmental control genes, as having a role in establishing the segmented pattern of the vertebral column.[417] As yet, no mutations in this gene have been found in those with vertebral segmentation defects. A chromosomal aberration, a deletion of 17p11.2, has been reported in congenital scoliosis but needs further verification.[194] As yet, no definitive cause for anomalous vertebral development has been established.

ASSOCIATED ABNORMALITIES

The neural axis, vertebral column, and other organ systems develop at a similar stage in utero. Goldberg and associates suggested that congenital vertebral anomalies arise from a nonspecific insult during this embryonic period that destabilizes the developmental control systems and may result in congenital malformations of any organ undergoing concurrent epigenesis.[153] The most common associated finding is spinal dysraphism, a general category that includes numer-

ous abnormalities such as diastematomyelia, syringomyelia, diplomyelia, Arnold-Chiari malformations, and intraspinal tumors.[50,289,291,359] The prevalence of one of these associated neural axis abnormalities is approximately 40 percent. Diastematomyelia is the most frequently recognized lesion. All of the abnormalities are best identified with MRI.

Once an intraspinal abnormality (such as a diastematomyelia spur) has been identified, it should be neurosurgically addressed if a progressive neurologic deficit has developed or if surgical correction of the scoliotic deformity is needed (Fig. 11–52).[291,323] To some physicians the mere presence of a potentially tethering intraspinal lesion may be sufficient reason for prophylactic surgical treatment. The rationale for this early aggressive approach is to address the lesion prior to the development of any neural dysfunction.[98] Any of these neural axis lesions may or may not be associated with a more visible clinical abnormality such as a hairy patch, a nevus, or a distinct neurologic deficit. Subtle deficits can be present, and therefore a careful neurologic examination is imperative for any patient with a congenital spinal abnormality.

In addition to the neural axis abnormalities, approximately 60 percent of patients will have associated abnormalities affecting any of seven other systems.[31] Approximately 20

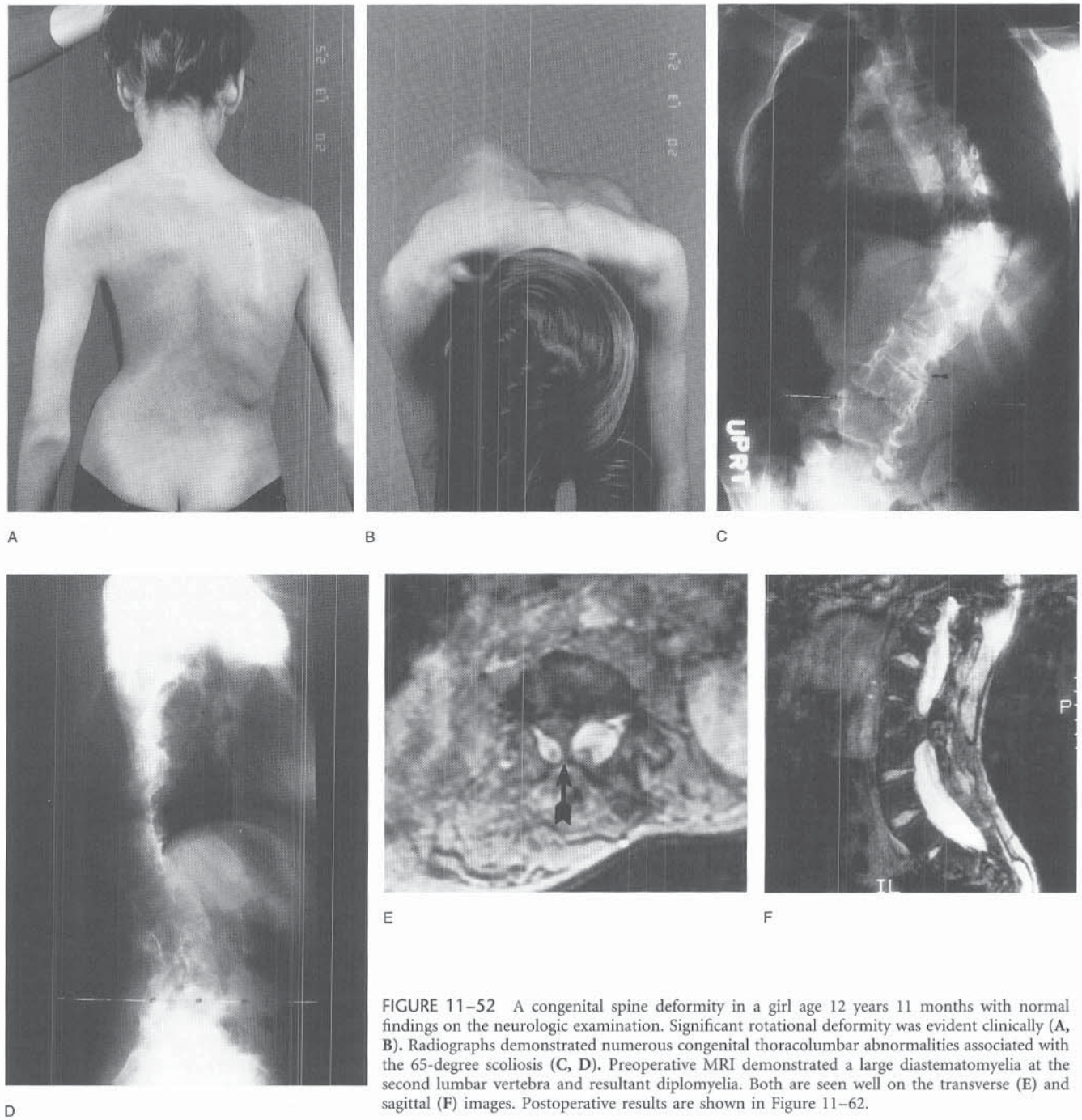


FIGURE 11–52 A congenital spine deformity in a girl age 12 years 11 months with normal findings on the neurologic examination. Significant rotational deformity was evident clinically (**A, B**). Radiographs demonstrated numerous congenital thoracolumbar abnormalities associated with the 65-degree scoliosis (**C, D**). Preoperative MRI demonstrated a large diastematomyelia at the second lumbar vertebra and resultant diplomyelia. Both are seen well on the transverse (**E**) and sagittal (**F**) images. Postoperative results are shown in Figure 11–62.

percent of patients will have an anomaly of the genitourinary tract system.[87,118,159,268] Cardiac anomalies are seen in approximately 12 percent of patients.[31,44] Other abnormalities include cranial nerve palsy, radial hypoplasia, clubfeet, dislocated hip, Sprengel's deformity, imperforate anus, and hemifacial microsomia.

CONGENITAL SCOLIOSIS

Congenital scoliosis may not become evident until later childhood even though the vertebral anomalies are always present at birth. In the child less than 3 years old, differentiation between infantile idiopathic scoliosis and congenital scoliosis can be difficult. Close examination of the radiographs will usually reveal the vertebral abnormalities present in congenital scoliosis.

The variety of vertebral anomalies that exist in congenital scoliosis lead to an unpredictable natural history. The deformity may remain mild or it may progress dramatically over time, ultimately resulting in severe spinal deformity and pulmonary compromise.[94] Understanding which vertebral anomalies put the scoliotic spine most at risk for progressive deformity allows the treating physician to intervene at the appropriate time.

Classification. Two basic types of abnormalities lead to congenital scoliosis: defects of vertebral formation and defects of vertebral segmentation (Fig. 11–53). Hemivertebrae and wedged vertebrae are examples of defects of formation. Defects of segmentation include block vertebrae, unilateral bars, and the worrisome unilateral bars accompanied by hemivertebrae. Approximately 80 percent of the vertebral anomalies associated with congenital scoliosis can be easily classified into one of the two above types. The remaining 20 percent cannot be precisely classified. Many patients may have a combination of deformities in which one type predominates.

Failure of Formation. Defects of formation may be partial or complete. Partial unilateral failure of formation produces a wedged or trapezoidal-shaped vertebra that contains two pedicles, although one of them may be hypoplastic. The associated scoliosis slowly worsens and may not require treatment.

True hemivertebrae are caused by complete failure of formation on one side and result in laterally based wedges consisting of half the vertebral body, a single pedicle, and a hemilamina. When present in the thoracic spine, hemivertebrae are usually accompanied by an extra rib. Hemivertebrae may be fully segmented (most common), semisegmented, nonsegmented, or incarcerated (least common) (Fig. 11–54). Distinguishing between these various types is important because the associated differences in growth potential have a profound effect on the eventual severity of the spinal deformity. A *fully segmented hemivertebra* has the highest likelihood of progressive deformity because it is separated from the adjacent vertebrae by intact end-plates and intervertebral disks. The hemivertebra is nearly always located at the apex of the scoliosis. Lower thoracic and thoracolumbar curves tend to worsen more rapidly than curves at other levels. Should there be two or more hemivertebrae on the same side of the spine, the deformity progresses

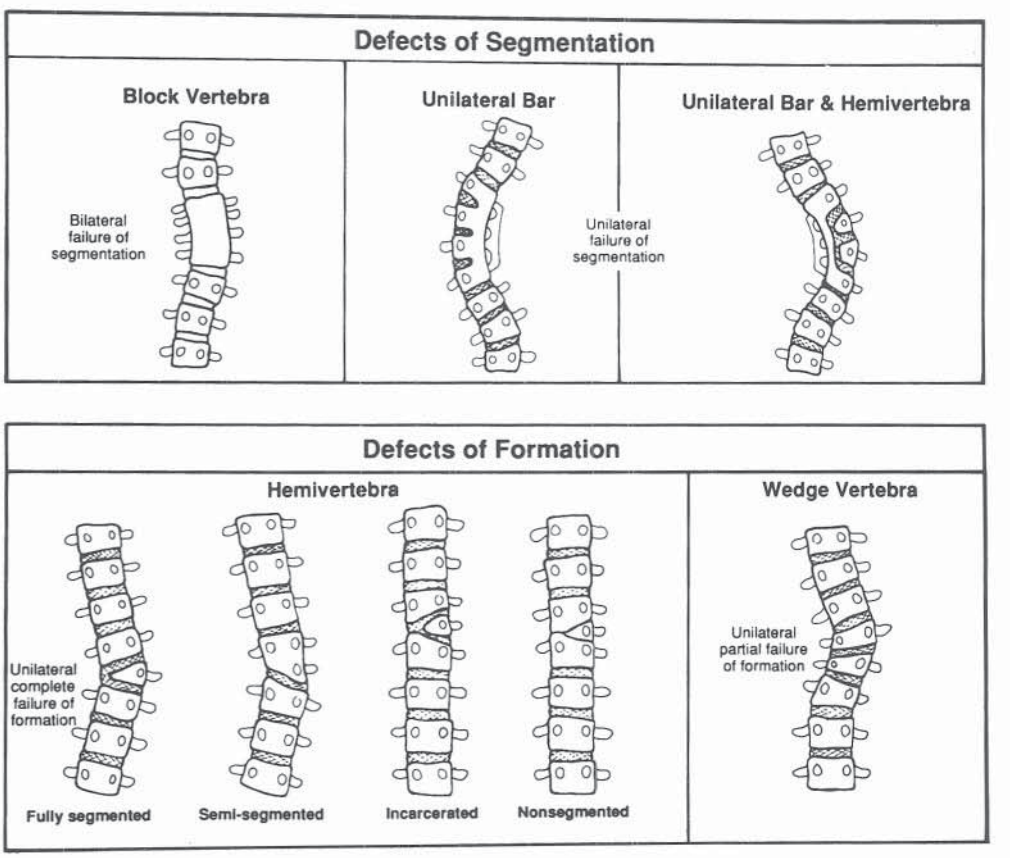


FIGURE 11–53 Congenital scoliosis: defects of formation and defects of segmentation. (From McMaster MJ: Congenital scoliosis. In Weinstein SL (ed): The Pediatric Spine: Principles and Practice, p 229. New York, Raven Press, 1994.)

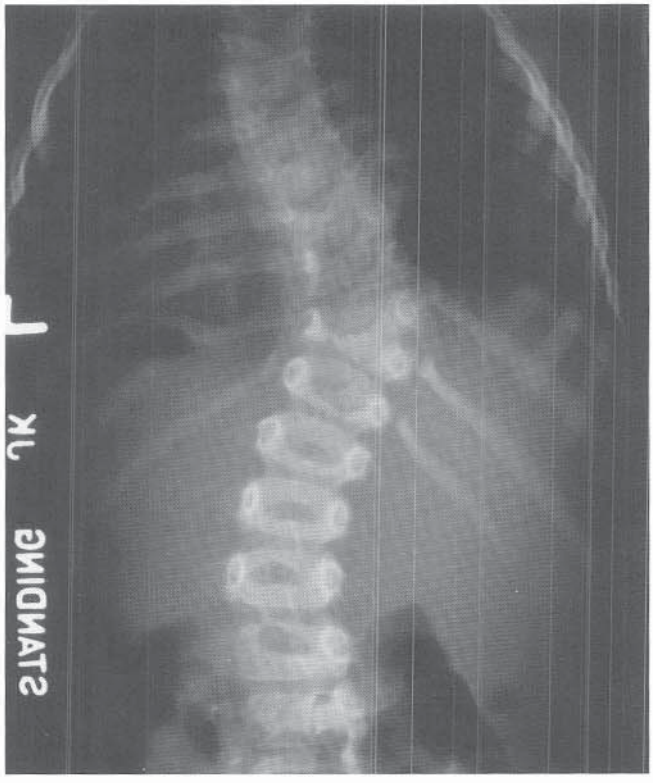


FIGURE 11–54 Spine radiograph of a 2-year-old girl with a 53-degree congenital scoliosis and semisegmented hemivertebrae at levels T8 and T10. A rib accompanies each of these hemivertebrae. This deformity is partially balanced by a left-sided hemivertebra at the T4 level. The deformity slowly progressed to 61 degrees by 9 years of age, at which time she underwent fusion.

at a faster rate. Conversely, the spinal deformity may be balanced and nonprogressive if two hemivertebrae are situated opposite one another.

A fully segmented hemivertebra at the lumbosacral junction creates significant obliquity between the spine and pelvis and is usually accompanied by a long compensatory scoliosis in the lumbar or thoracolumbar region. This readily apparent deformity is best treated surgically (usually by hemivertebrectomy) at an early age, before the compensatory curve becomes fixed (Fig. 11–55).

A *semisegmented hemivertebra* is separated from one adjacent vertebra (superior or inferior) by a normal vertebral growth plate and disk, but is fused to the other adjacent vertebra. Although the growth of the spine should remain balanced, the hemivertebra can induce a slowly progressive scoliosis. Treatment is necessary only if the deformity is progressive (see Fig. 11–54).

A *nonsegmented hemivertebra* is fused to both adjacent vertebrae (above and below) and therefore has no vertebral end-plates or adjacent disks. In the absence of any asymmetric growth, the nonsegmented hemivertebra does not cause progressive spinal deformity. An *incarcerated hemivertebra* is more ovoid in shape and smaller than a fully segmented (nonincarcerated) hemivertebra. The vertebrae above and below compensate for this hemivertebra and, as a result, there is minimal, if any, scoliosis.

Failure of Segmentation. Defects of segmentation result in a bony bar or bridge between two or more vertebrae, either

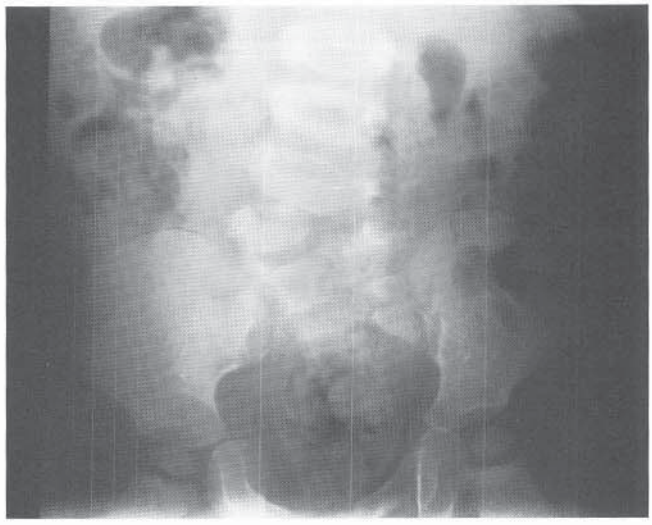


FIGURE 11–55 Standing radiograph of an 18-month-old boy showing significant obliquity of the pelvis due to a hemivertebra at the L5 level. Postoperative findings are shown in Figure 11–63.

unilaterally or involving the entire segment. Circumferential, symmetric failure of segmentation leads to a *block vertebra* (Fig. 11–56). This does not cause any angular or rotational spinal deformity but does lead to some loss of longitudinal growth. Klippel-Feil syndrome in the cervical spine represents a severe form of this failure of segmentation.

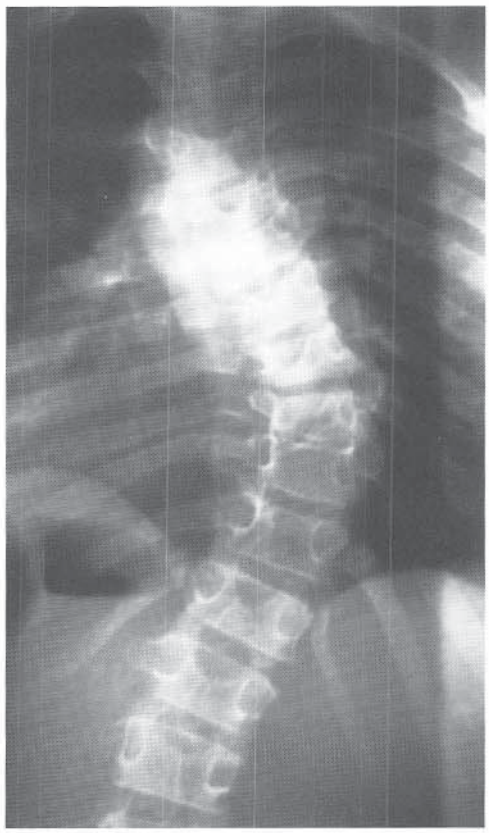


FIGURE 11–56 Radiographic appearance of a 10-year-old girl with a 52-degree scoliosis and block vertebrae at T5–6 and T9–10. The deformity cannot be attributed to the mere presence of these abnormalities. MRI of the spinal canal was normal.

A *unilateral failure of segmentation* of two or more vertebrae (unilateral bar) is the most common cause of congenital scoliosis. Usually a bar of bone fuses the disk spaces, pedicles, and facet joints on one side of the spine, precluding growth on the concavity (Fig. 11–57). Growth usually proceeds on the convexity and leads to worsening of the deformity. Rib fusions or other rib abnormalities on the concavity of the scoliosis are often seen adjacent to the bony bar bridging the vertebrae.

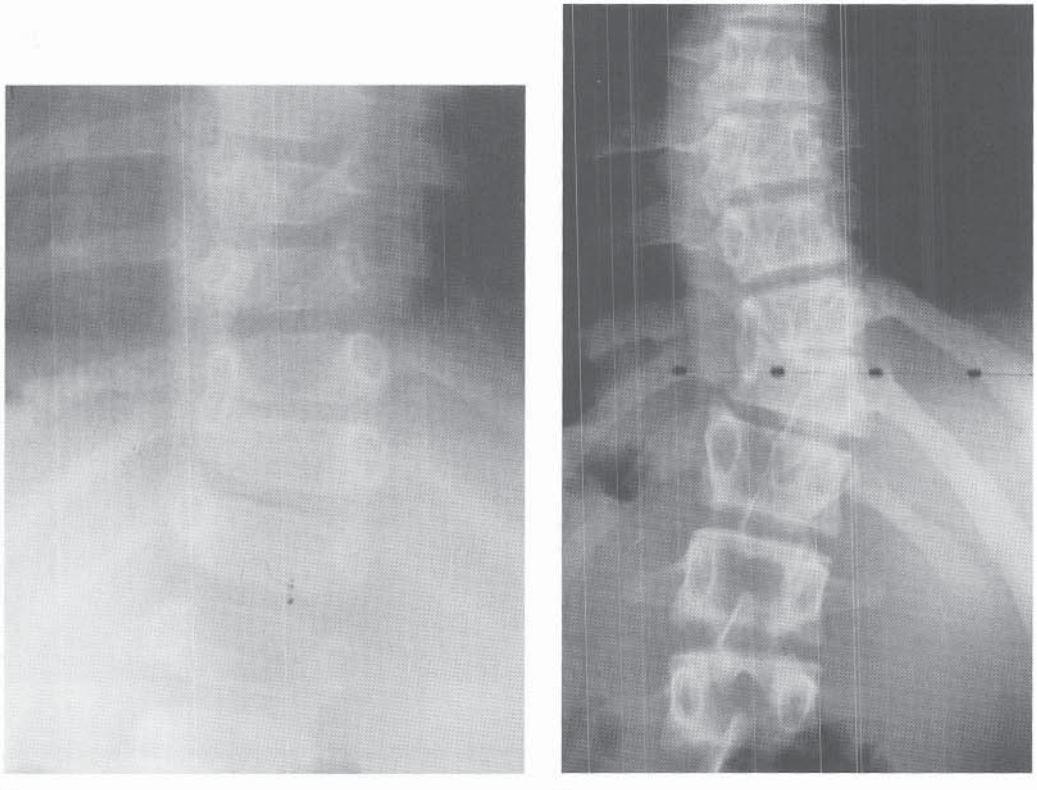
A subgroup of patients with unilateral failure of segmentation will have one or more hemivertebrae located on the opposite convex side of the curve. For an individual with congenital scoliosis, this combination carries the worst prognosis in that it produces the most severe and rapidly progressive deformity. Curves of this kind located in the thoracolumbar spine can be expected to exceed 50 degrees by age 2 years. Without treatment, patients with thoracolumbar, midthoracic, or lumbar curves become severely deformed at an early age owing to a combination of shoulder imbalance, severe distortion of the rib cage, decompensation of the trunk, and/or pelvic obliquity that produces an apparent leg length discrepancy.

In addition to the deformities involving the thoracic and lumbar spine, congenital scoliosis involving the cervical and cervicothoracic spine can lead to significant deformities of the neck and position of the head (Fig. 11–58).[418] The deformities in the neck can result in persistent tilt of the head (apparent torticollis) because the relatively few normal vertebrae above the area of the segmentation defects cannot provide sufficient compensation for balance. Nearly 50 percent of those with congenital cervical or cervicothoracic scoliosis have associated Klippel-Feil abnormalities.[418,438,441]

Natural History. The rate of curve progression and the final severity of the congenital scoliosis are related to two factors: the type of vertebral anomalies present and the patient's remaining growth at the time of diagnosis. The two periods of accelerated growth during which congenital scoliosis worsens most rapidly are the first 2 years of life and the adolescent growth spurt.

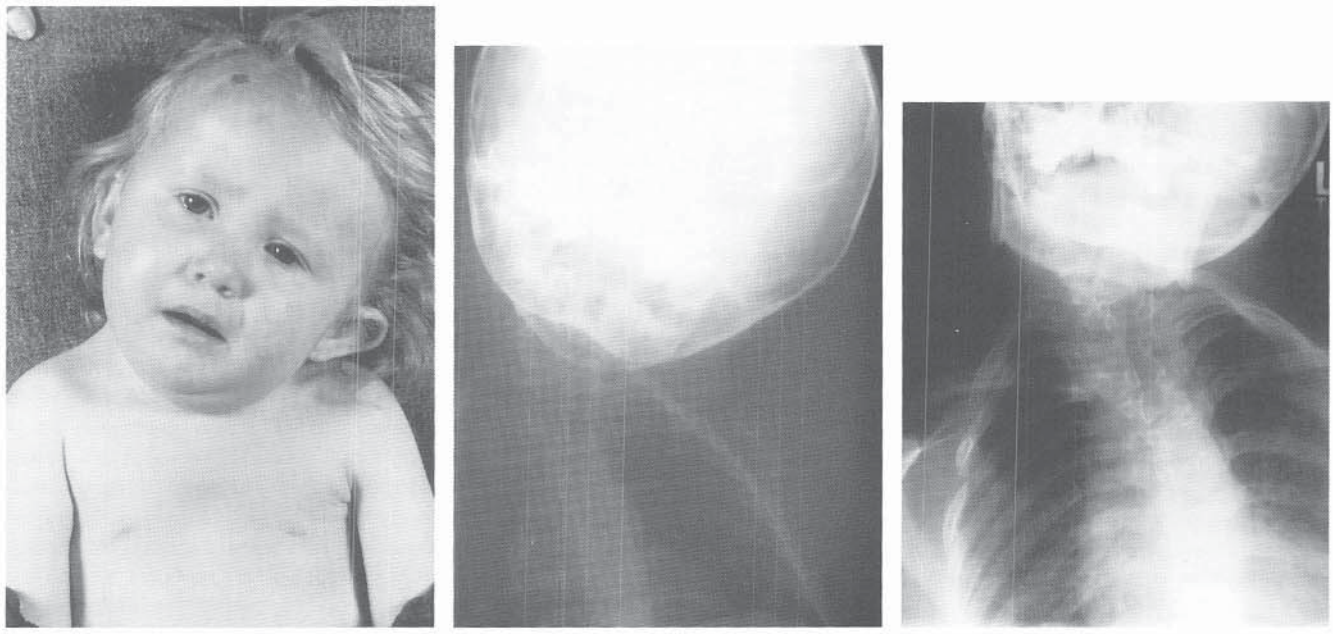
The type of congenital scoliosis in which progression is a certainty is the unilateral unsegmented bar with contralateral hemivertebra (one or more).[46,288,289,471,474] Thoracolumbar curves of this type have the worst prognosis and deteriorate 7 degrees per year before age 10 years, increasing to 14 degrees per year during the adolescent growth spurt. Severe spinal deformity will predictably occur unless surgical intervention is undertaken.

The next most severe deformities are caused by isolated unilateral unsegmented bars, followed, in order of decreasing severity, by multiple fully segmented hemivertebrae, a single fully segmented hemivertebra, and a block vertebra. In an excellent study of the natural history of congenital scoliosis, McMaster and Ohtsuka reviewed 202 patients followed past age 10 years without treatment. They found only 11 percent of the curves to be nonprogressive, 14 percent to be slightly progressive, and 75 percent to be significantly progressive.[292] At follow-up, 36 percent of the patients had



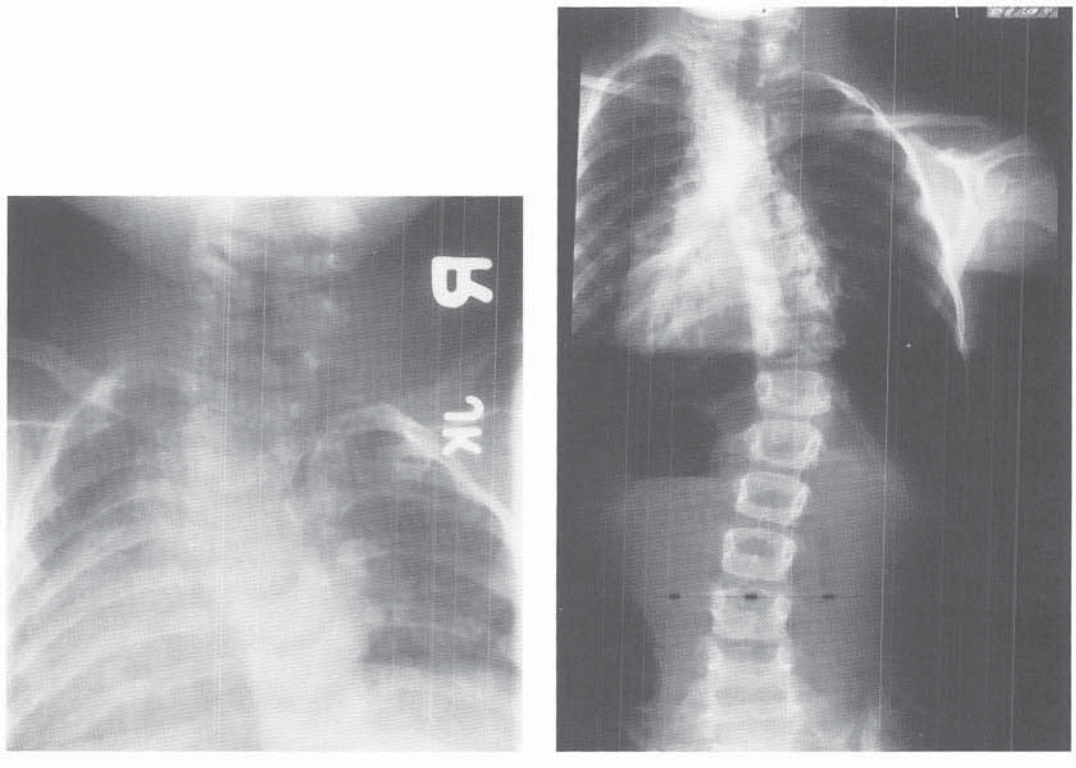
A B

FIGURE 11–57 An 11-month-old boy had a unilateral failure of segmentation noted on a radiograph obtained because of mild back asymmetry (**A**). On a radiograph obtained at age 16 years, no significant change had occurred in the spinal alignment, even though no intervention had been undertaken (**B**).



A B C

FIGURE 11–58 **A** and **B,** Clinical and radiographic appearance of a 2-year-old girl with Klippel–Feil syndrome and congenital cervicothoracic scoliosis due to numerous vertebral abnormalities. **C,** Eleven years later, without operative intervention, the curve was of similar size, and the head tilt was mild in its clinical appearance. Neurologic findings were normal.



A B

FIGURE 11–59 Radiographic appearance of a girl with numerous congenital abnormalities in the upper thoracic spine, including hemivertebrae on the convexity and fused ribs on the concavity. These abnormalities are well demonstrated on a radiograph obtained at age 7 months **(A).** Because of the high risk of progression, anterior and posterior in situ fusion between T2 and T8 was performed at age 16 months. A compensatory curve developed below which measured 40 degrees at age 7 years **(B).** This compensatory curve was subsequently managed with a brace.

curves between 40 and 60 degrees and 28 percent had curves exceeding 61 degrees.

The future behavior of congenital scoliosis due to a combination of the previously described abnormalities becomes extremely difficult to predict. The need for treatment will be determined over the course of numerous visits as the nature of the curve becomes evident.

Compensatory curves in the otherwise normal spine develop more commonly in patients with congenital scoliosis and a curve apex at T5, T6, or T7 (Fig. 11–59). As the congenital curve deteriorates, this secondary curve may worsen, become inflexible, and require treatment.[288] Patients with severe lumbar or thoracolumbar congenital curves may be unable to develop compensatory curves large enough to maintain a balanced trunk. In this instance, notable pelvic obliquity and apparent lower limb length inequality are unsightly compensatory mechanisms utilized to keep the trunk vertical.

Radiographic Evaluation. The radiographic detail of the vertebral abnormalities is best seen on films obtained prior to the development of significant deformity. Often, this is during infancy, when a radiograph is taken while the child is supine. As the child grows and the congenital scoliosis progressively worsens, the bony detail becomes less clear. On first presentation, coned-down radiographs of the affected area will provide the most information about the vertebral anomalies. Associated abnormalities that may also be noted on plain radiographs include diastematomyelia (midline bone spur), spina bifida occulta, and congenital rib fusions on the concavity of the curve.

Although the early supine radiographs best reveal bone detail, they cannot be used in the assessment of curve progression. The initial upright radiograph must serve as the baseline study against which further curve progression will be measured. The variability in measuring angles in congenital scoliosis is larger than that in idiopathic scoliosis because of skeletal immaturity, incomplete ossification, and anomalous development of the end vertebrae.[251] As a result, interpretation of actual progression becomes more difficult. Concerted efforts should be made to measure the curves with similar end points, detecting subtle yet steady progression of the curvature, and assessing secondary or compensatory curves. The most recent radiographs should be carefully compared with one of the earliest upright radiographs to ascertain whether slow yet steady progression has occurred. It is not uncommon for radiographs taken 4 to 6 months previously to reveal only slight progression when compared with current radiographs. If comparisons are made with radiographs obtained several years earlier, the steady changes that develop become more evident. Consistent measurement of the secondary curve may also reveal its progression and point to the need for treatment.

In severe congenital scoliosis, plain radiographs may not provide sufficient detail of the vertebral abnormalities. Should surgical intervention be necessary, CT with three-dimensional reconstruction may be helpful in the preoperative planning.

MRI of the spine should be performed in all patients with congenital scoliosis who are undergoing surgical intervention. Approximately 40 percent of patients will have an intraspinal abnormality, which in all cases should be evident on MRI (Fig. 11–60).[289] Today, reconstructed MRI images can provide a clear picture of the canal contents despite the severe three-dimensional deformity associated with some cases of congenital scoliosis.

Nonoperative Treatment. Bracing is much less successful in the treatment of congenital scoliotic deformities than it is in idiopathic scoliosis. Use of a brace to control curve progression secondary to unsegmented bars or hemivertebrae is universally unsuccessful and should not be attempted. However, bracing can be considered as a means of temporarily controlling a long flexible compensatory curve below the congenital component, thereby allowing further spinal growth prior to operative intervention. If the congenital or compensatory component worsens during the period of brace wear, operative intervention should be undertaken without delay. As a generalization, few patients with congenital scoliosis will benefit from the use of an orthosis.

Operative Treatment. The primary goal of surgery is to stop further progression of the congenital spinal deformity. If a partial correction can be safely obtained, that is an added benefit. Even for relatively small curves (less than 40 degrees), once progression has been confirmed, surgical intervention should be undertaken. This concept needs to be emphasized: surgery should be performed *before* a major deformity develops (Fig. 11–61). Young patients who have undergone spine fusion still need to be followed to maturity, as a progressive deformity can develop above or below the fused sites. Later surgery may yet be required in these individuals.

Various operative approaches can be used, the choice depending on the maturity of the patient, the location of the deformity, and the type of congenital deformity. These approaches include anterior and posterior convex hemiepiphysiodesis, anterior and posterior spinal fusion, posterior fusion with or without instrumentation, hemivertebra excision, and spine osteotomies.

ANTERIOR AND POSTERIOR CONVEX HEMIEPIPHYSIODESIS. This approach is ideal for patients with a small but progressive, scoliosis in which there is growth potential in the concavity of the curve.[216,279,482] It is best suited for patients less than 5 years old with curves less than 40 degrees that are caused by fully segmented hemivertebrae. Theoretically, the fusion on the convexity should allow the open disks on the concavity to continue growing and should lead to some progressive correction. This method is of no value if a unilateral unsegmented bar is present on the concavity. No instrumentation is used in these young patients. In the thoracic spine, the anterior hemiepiphysiodesis could be performed thoracoscopically, if preferred. The open technique would be required for thoracolumbar or lumbar hemiepiphysiodesis. A transpedicular approach for the anterior hemiepiphysiodesis has been described.[213,220] Only a portion of the intervertebral disk and end-plate is removed. Posteriorly, only the lamina on the convexity is approached; it is then decorticated, facetectomies are performed, and bone graft is applied. Postoperative management includes a period of cast wear for 4 to 6 months to allow the convex growth arrest to consolidate.

ANTERIOR AND POSTERIOR FUSION. This approach is used in immature individuals in whom continued anterior growth on the convexity would lead to development of the crankshaft

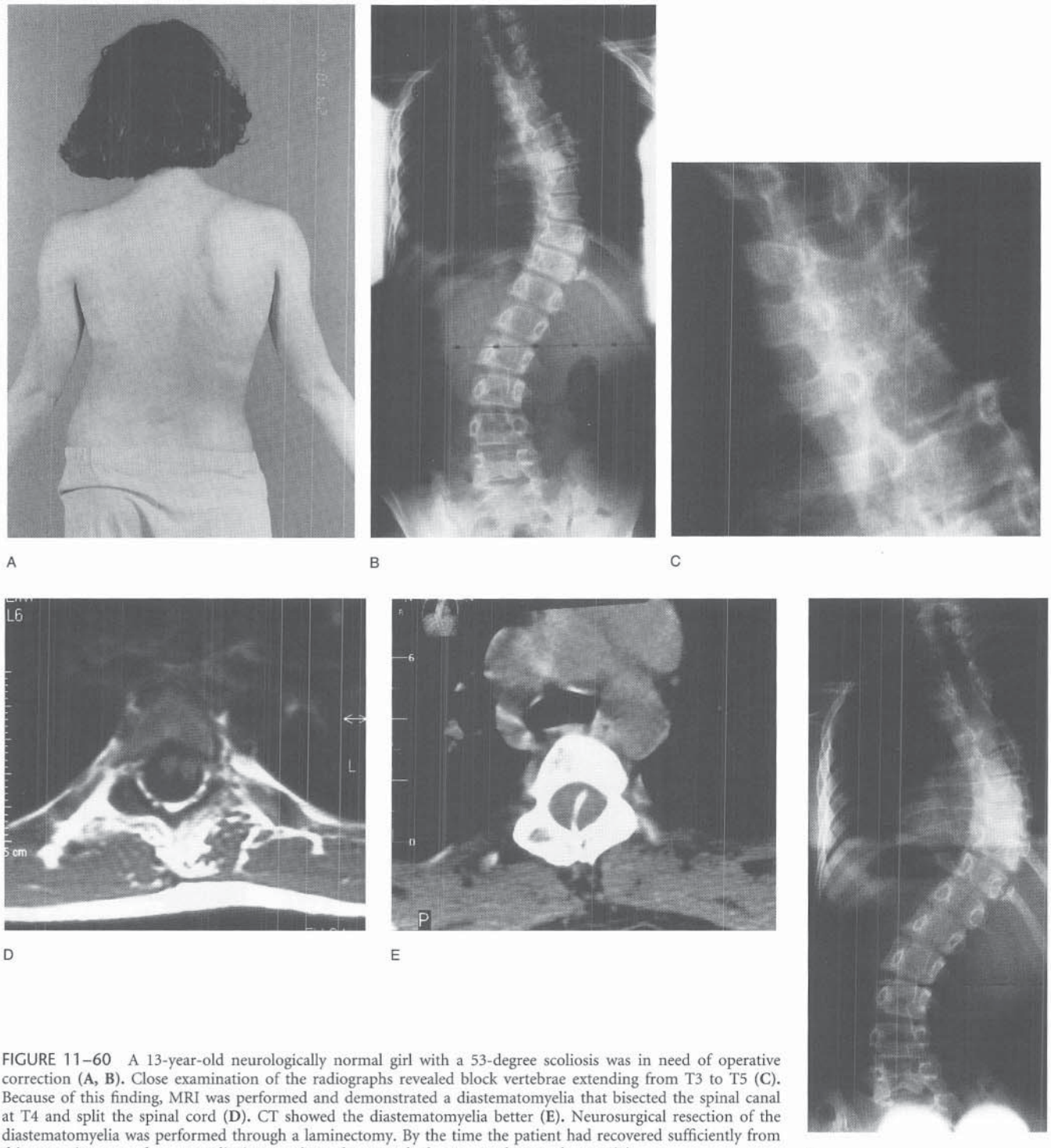
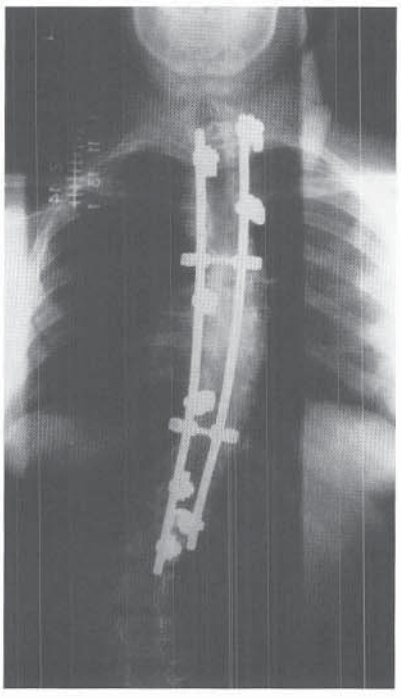


FIGURE 11–60 A 13-year-old neurologically normal girl with a 53-degree scoliosis was in need of operative correction **(A, B)**. Close examination of the radiographs revealed block vertebrae extending from T3 to T5 **(C)**. Because of this finding, MRI was performed and demonstrated a diastematomyelia that bisected the spinal canal at T4 and split the spinal cord **(D)**. CT showed the diastematomyelia better **(E)**. Neurosurgical resection of the diastematomyelia was performed through a laminectomy. By the time the patient had recovered sufficiently from this operation to undergo a scoliosis procedure, the curve had progressed to 62 degrees **(F)**.

phenomenon.[110,175,437] Children most in need of this approach have unilateral unsegmented bars with (or sometimes without) contralateral hemivertebrae (see Fig. 11–61). In the very young child, fusion should extend to one level above and one level below the anomalous vertebrae. Doing so may prevent "adding on" of the curve in subsequent years. Postoperatively, a cast is needed for 4 to 6 months, until healing has been achieved. Placement of posterior subcutaneous rods on the concavity of the curvature, without performing posterior fusion, has been advocated by some as a means of achieving partial correction of the curve beyond the anomalous vertebrae.[156] Conceptually, this approach would allow postponement of a definitive fusion procedure until the child is older (more than 10 years old). At the author's institution definitive fusion is performed at age 7 to 8 years because our experience with the subcutaneous rod and repeated lengthenings has shown little or no benefit.



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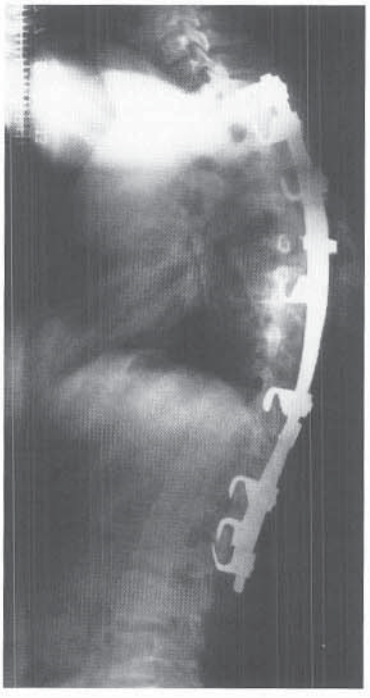
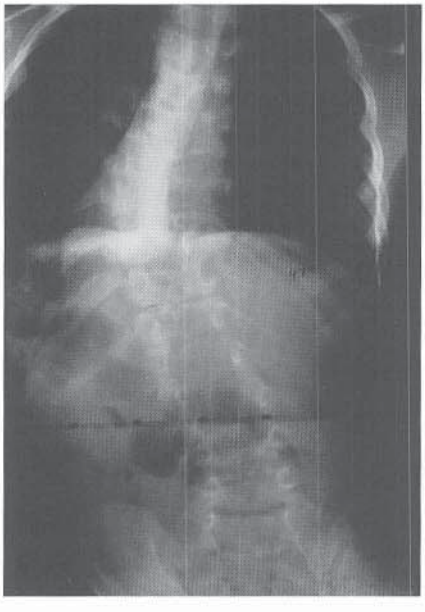
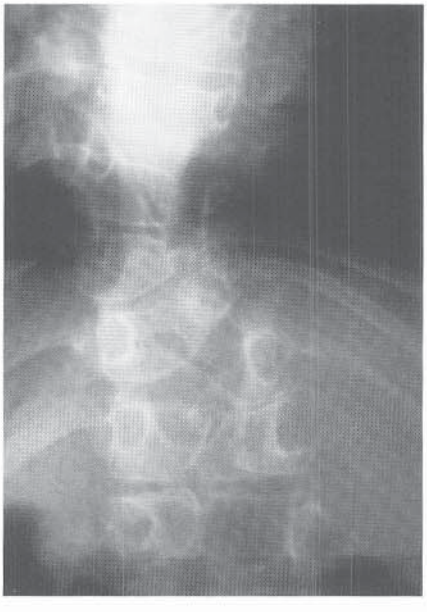


H

FIGURE 11–60 *Continued.* Two years after posterior spinal instrumentation and fusion, the spine was well balanced and she remained neurologically normal (**G, H**).



A



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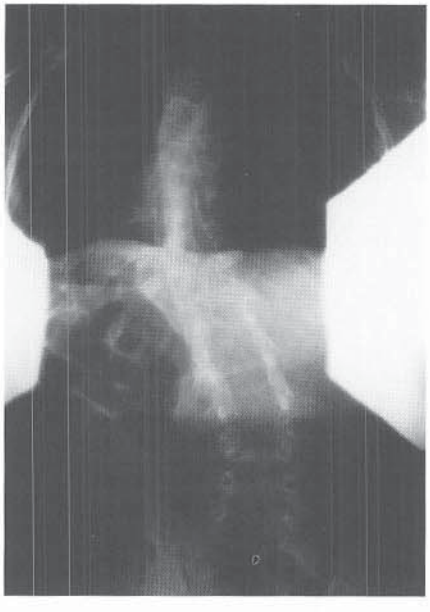


C

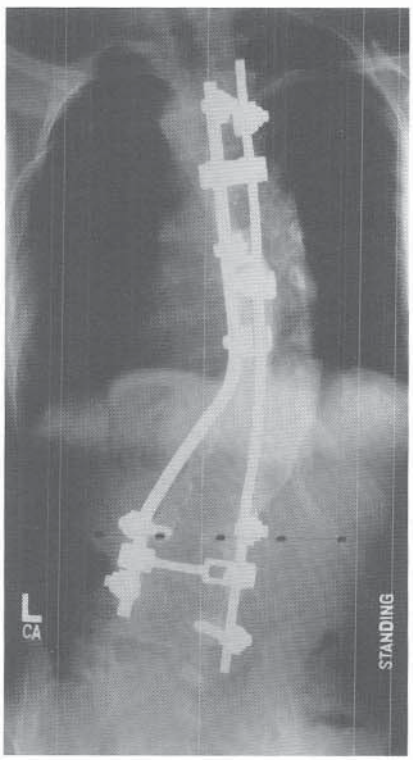
FIGURE 11–61 A neurologically normal 6-year-old girl had a left congenital thoracic curve measuring 47 degrees (**A**). It had increased a small amount (6 degrees) over 4 months. The decision was made to operate before a major deformity developed. The preoperative bending radiograph clearly demonstrated a bar formation in the concavity at T8–10 (**B**). (Five years previously the patient had undergone resection of a diastematomyelia at the thoracolumbar junction.) Anterior and posterior fusion without instrumentation was performed. After surgery the patient was immobilized for 4 months in a Risser cast. Four years later the curve measured 25 degrees, she had good spinal balance, and she remained neurologically normal (**C**).

POSTERIOR SPINAL FUSION. Posterior spinal fusion is indicated in the older child with progressive congenital scoliosis in whom the crankshaft phenomenon is unlikely to develop or in younger children who do not have normal anterior growth potential. Unlike children with idiopathic scoliosis, many young children with congenital scoliosis do not develop crankshaft progression because the anterior growth plates are abnormal.[262,487] These patients are not always easily identified preoperatively, and consequently the decision to include anterior fusion is difficult.

If any correction of the deformity with instrumentation is anticipated, preoperative MRI of the neural canal is essential to rule out diastematomyelia, syrinx, tumor, and other abnormalities. If identified, these lesions need to be addressed neurosurgically before the spinal fusion is performed. If spinal instrumentation is used, spinal cord monitoring is imperative, and use of the wake-up test should be considered.[481] Any deformity correction that is achieved is obtained through the flexible, normal portion of the curve, not in the rigid, congenitally anomalous region (Fig. 11–62). Of all the forms of scoliosis, congenital scoliosis carries the highest risk of neurologic complications following intraoperative correction.

Slow, gradual correction of severe deformities can be achieved in some individuals by means of preoperative halo traction used for 6 to 12 weeks. At our institution, patients use it while sleeping, walking, or in a wheelchair. A home traction program is possible but requires very close monitoring for any neurologic change (numbness, tingling, weakness). When correction has been achieved or has reached a plateau, the spine is stabilized by instrumentation and fusion.

HEMIVERTEBRA EXCISION. Very few patients with congenital scoliosis secondary to a hemivertebra need to have the hemivertebra excised. Most can be managed with safer procedures already described. Hemivertebra excision carries a significant risk of temporary, and occasionally permanent, neurologic injury to a nerve root.[184] This procedure is indicated for patients with a fixed decompensation in whom adequate alignment cannot be achieved through other procedures—usually patients with a hemivertebra at the fourth or fifth lumbar level. Excision of the hemivertebra at this level is safer than in the upper lumbar or thoracic region as the cauda equina is more tolerant of manipulation than the area surrounding the spinal cord. The major advantage of resection of the hemivertebra is that it allows maximum correction of the deformity and realignment of the spine.[48,65,184,222] This surgical approach requires two exposures, the first anterior and the second posterior. The anterior approach allows removal of the body of the hemivertebra and its adjacent disks back to the spinal canal, with removal of the anterior half of the pedicle. The patient is then repositioned and the posterior elements are excised through a secondary midline approach. Correction is then achieved internally with posterior compression instrumentation on the convexity or externally with a cast during the postoperative period. Immobilization with a cast is needed for 4 to 6 months until fusion has been achieved (Fig. 11–63). The main complication of this procedure is nerve root compression on the convexity caused by the pedicle on the proximal segment pressing against the nerve root in the area of resection. Excision of a lumbosacral hemivertebra is contraindicated if a second hemivertebra on the opposite side exists in the lumbar region.



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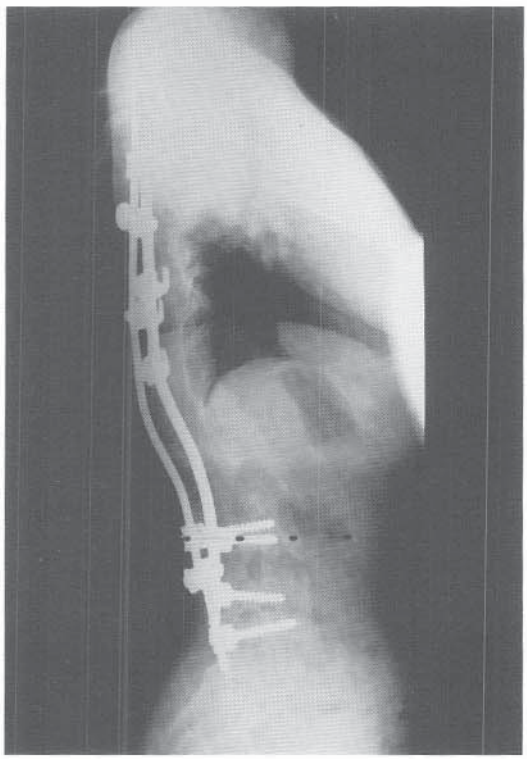
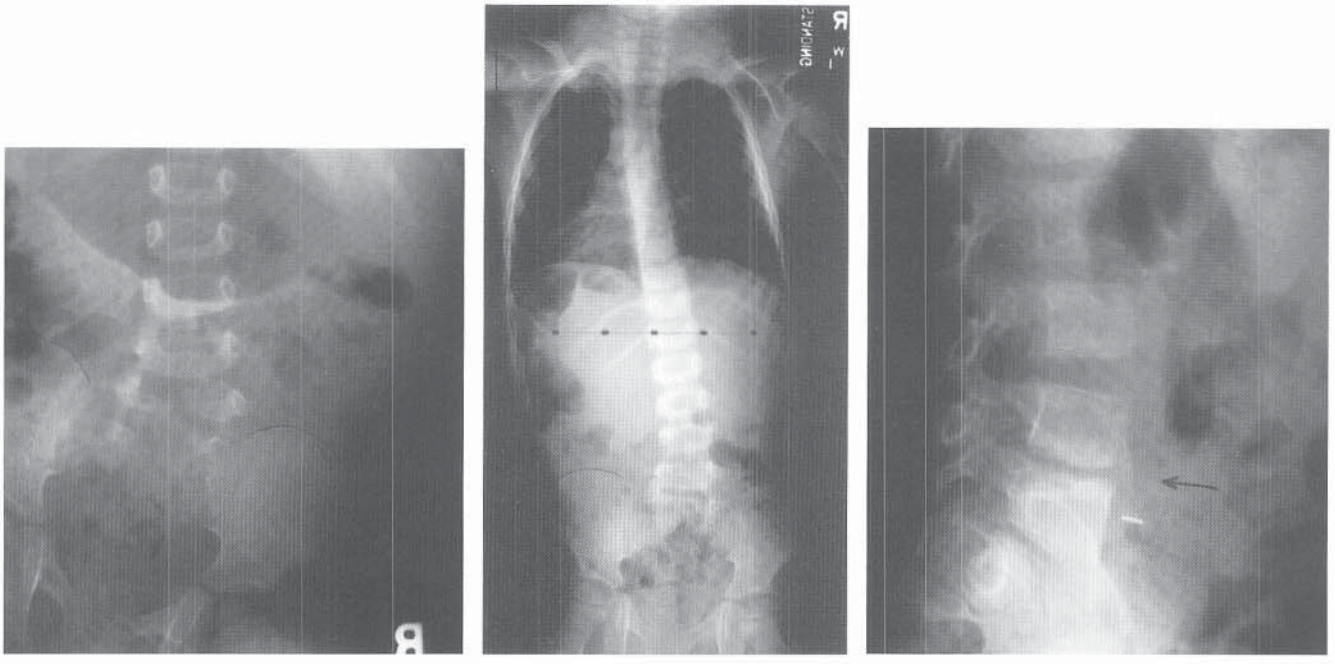


B

FIGURE 11–62 The patient whose preoperative imaging findings are shown in Figure 11–52 underwent posterior spinal instrumentation and fusion 4 weeks after resection of the diastematomyelia. Eighteen months later (at age 14 years 6 months), her spine remained balanced, with a residual 49-degree curve (**A, B**).



A B C

FIGURE 11–63 The patient with a hemivertebra shown in Figure 11–55 had significant pelvic obliquity due to the L5 hemivertebra (**A**). At age 3 years he underwent resection of the L5 hemivertebra through both anterior and posterior approaches. Correction was maintained with a cast rather than with internal fixation. Twenty-seven months later, the obliquity was much improved (**B, C**). Although there was a stable, fibrous nonunion between L4 and the sacrum, the patient remained asymptomatic.

OSTEOTOMY OF THE SPINE. This approach is reserved for the older child who has a rigid, severe, angular scoliosis and significant spinal decompensation. Anterior and posterior wedge resection osteotomies are performed and the spine is then instrumented for correction (Fig. 11–64).[138,184] If this procedure is undertaken in the thoracic spine, resection of ribs may be required. This approach should be performed only by the very experienced spine surgeon, as the risk of neurologic complications is high.

CONGENITAL KYPHOSIS

Congenital kyphosis represents an abrupt posterior angulation of the spine due to a localized congenital malformation of one or more vertebrae.[108,161,471,481] Fortunately, this condition is less common than congenital scoliosis, for the potential consequences of paraplegia are far greater in those with congenital kyphosis.

Classification. Congenital kyphosis is caused by defects of vertebral body formation (type I), defects of vertebral body segmentation (type II), or a combination of the two (type III) (Fig. 11–65). In contrast to congenital scoliosis, failure of formation in congenital kyphosis is the most common type and will tend to produce more severe deformities than are seen with kyphosis due to failure of segmentation. These vertebral abnormalities may also lead to frontal plane deformity, resulting in kyphoscoliosis.

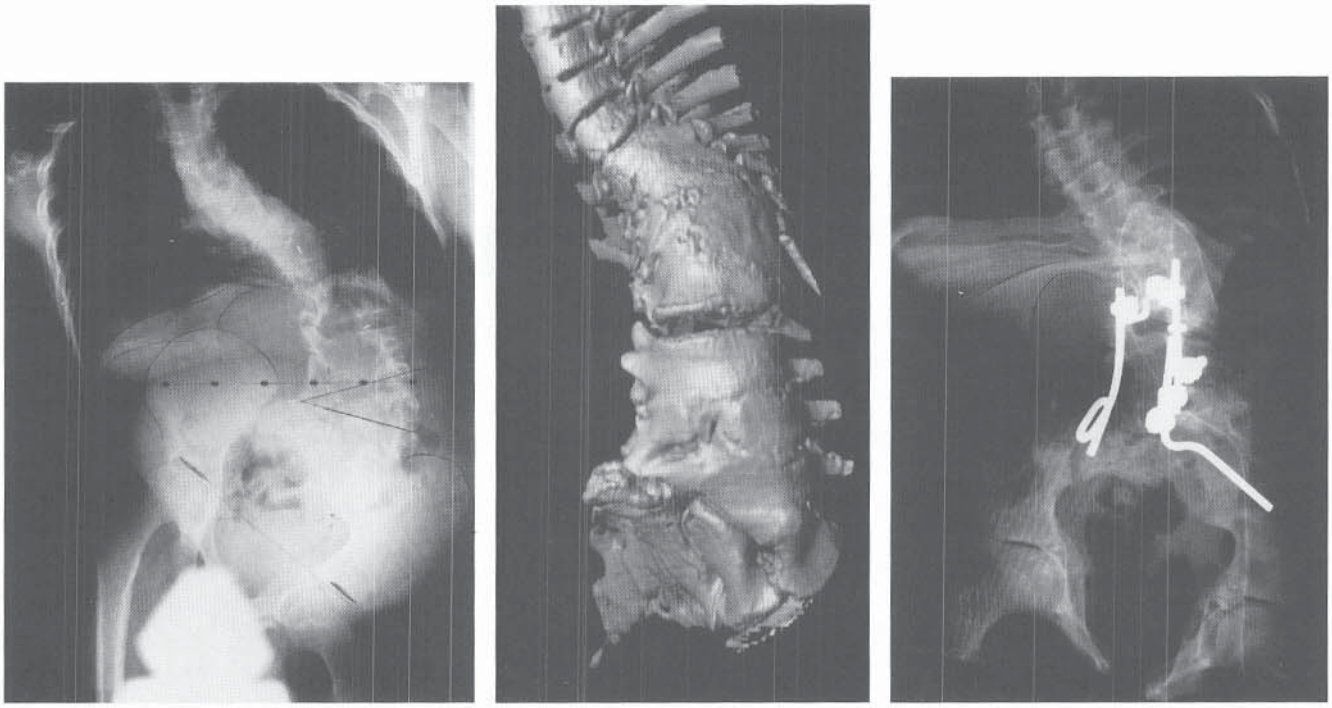
Failure of Formation (Type I). In kyphosis due to defects in vertebral body formation, part or all of the vertebral body is deficient (Fig. 11–66). Several contiguous levels may be affected, which produces greater deformity. In general, the posterior elements (spinous process, pedicles, transverse processes) remain present and accompany the deficient vertebral body. Growth continues normally in the posterior portion of the spine but not anteriorly. As a result, relentless progression of deformity usually occurs.

Defects in formation place the patient at a much greater risk for developing paraplegia than defects in segmentation. The kyphotic junction may be quite unstable, particularly when the apex is between T4 and T9. Paraplegia can occur at any age but is most common during the adolescent growth spurt. Reports of acute-onset paraplegia following minimal trauma in young children testify to the potentially fragile neurologic status.

Failure of Segmentation (Type II). In kyphosis due to failure of segmentation, the anterior portion of two or more adjacent vertebral bodies are fused together. This deformity tends to be less progressive, produces less deformity, and is associated with a much lower risk of paraplegia than kyphosis due to defects in formation.[282] The area most commonly affected is the lower thoracic or thoracolumbar spine (Fig. 11–67).

Natural History. The apical area of the kyphosis can occur at any level but most frequently occurs between the tenth thoracic and the first lumbar level.[293] There appears to be no relationship between the severity of the kyphosis and its location in the spine. Progression of these deformities is most rapid during the adolescent growth spurt.

Congenital kyphosis due to either failure of formation (type I) or mixed anomalies (type III) tends to be relentlessly



A B C

FIGURE 11–64 A 12-year-old neurologically normal boy had severe fixed pelvic obliquity (**A**). There was only one motion segment (L1–2 disk) between the T7 and the pelvis because of the numerous congenital abnormalities and previous spinal fusions. Three-dimensional reconstruction of CT images allowed viewing of the spine throughout a 360-degree rotation (**B**). A 32-degree wedge osteotomy performed through a posterior exposure improved but did not fully correct the pelvic obliquity (**C**).

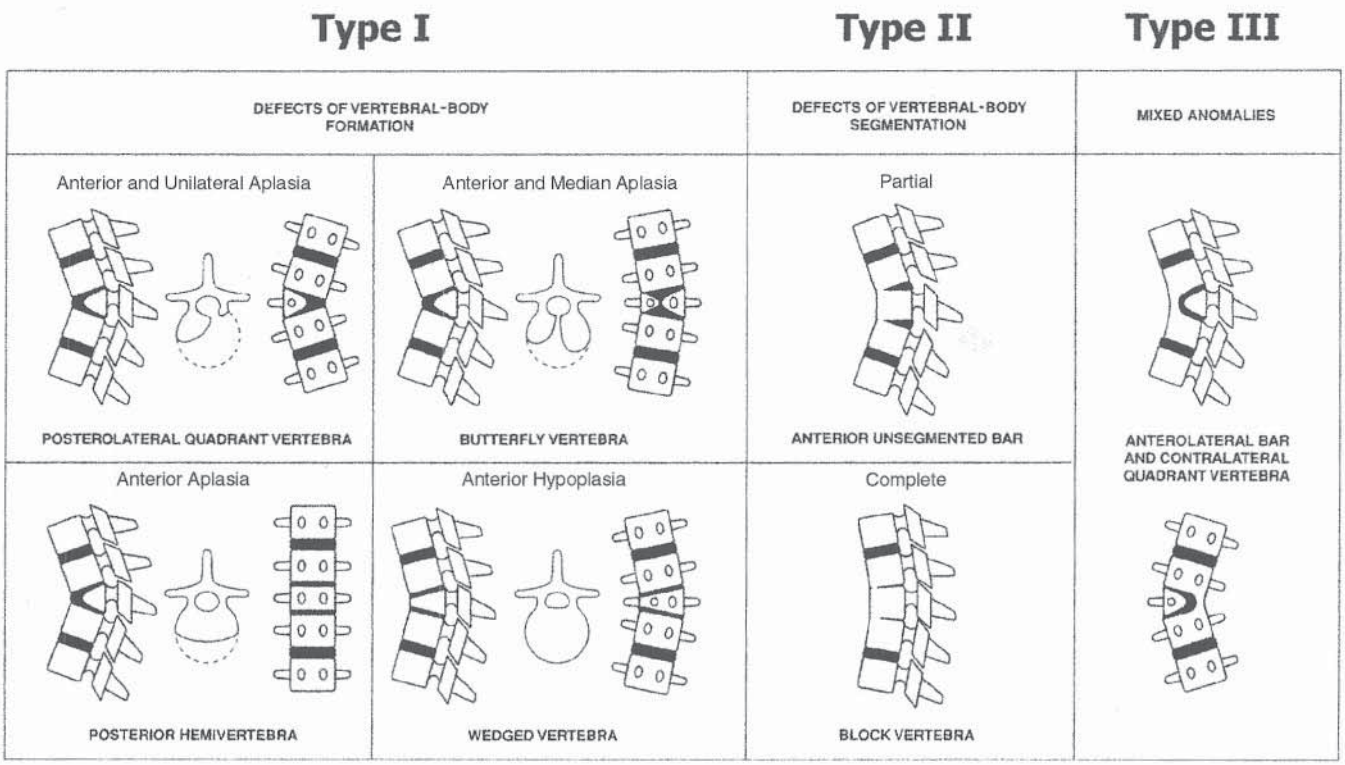
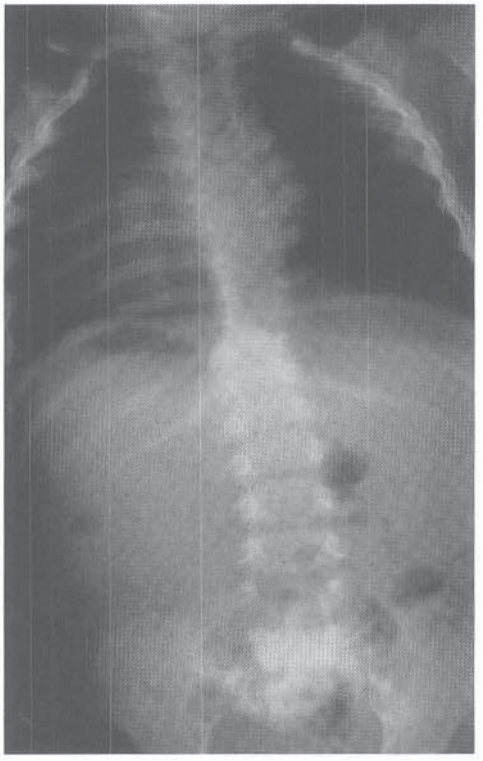


FIGURE 11–65 Congenital kyphosis type I results from defects in vertebral body formation, type II results from defects of segmentation, and type III results from a combination of the two. (From McMaster MJ, Singh H: Natural history of congenital kyphosis and kyphoscoliosis. J Bone Joint Surg 1999;81-A:1369.)



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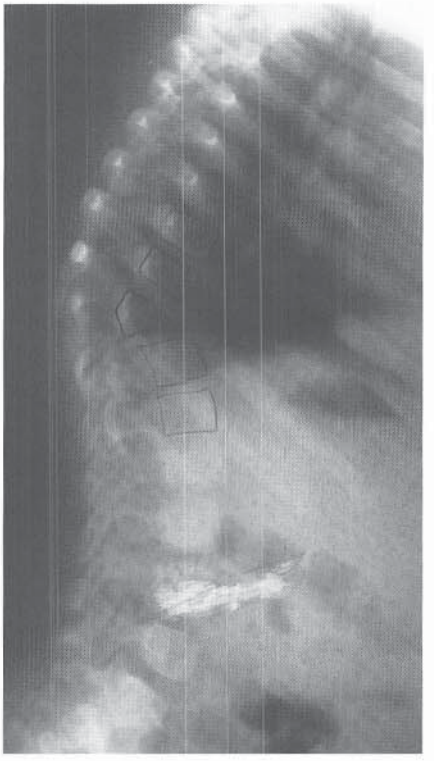

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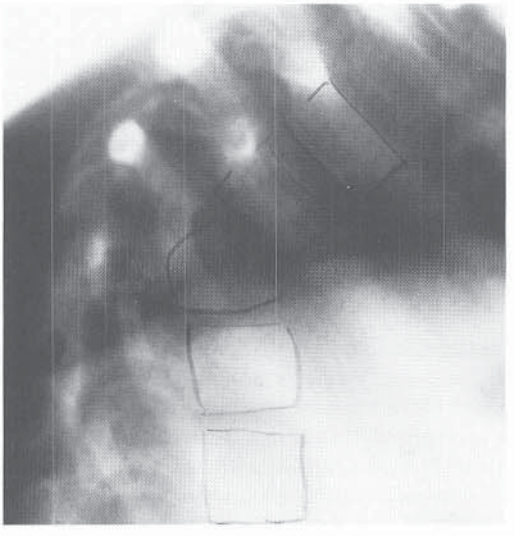

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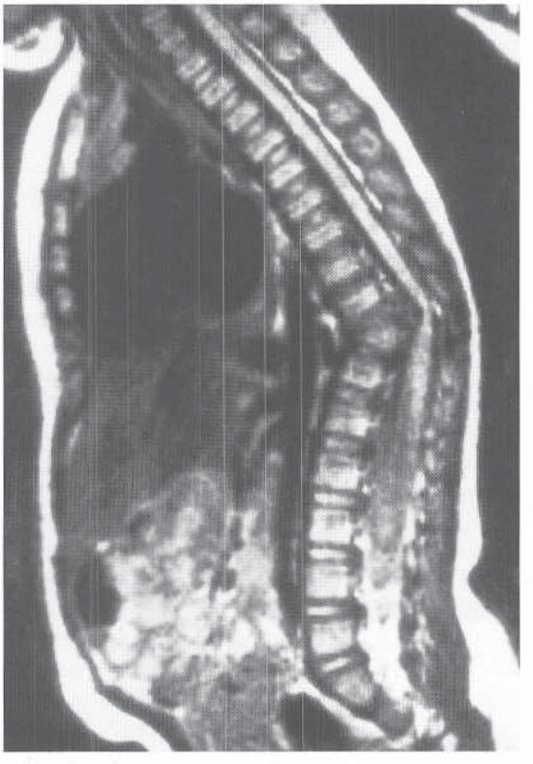
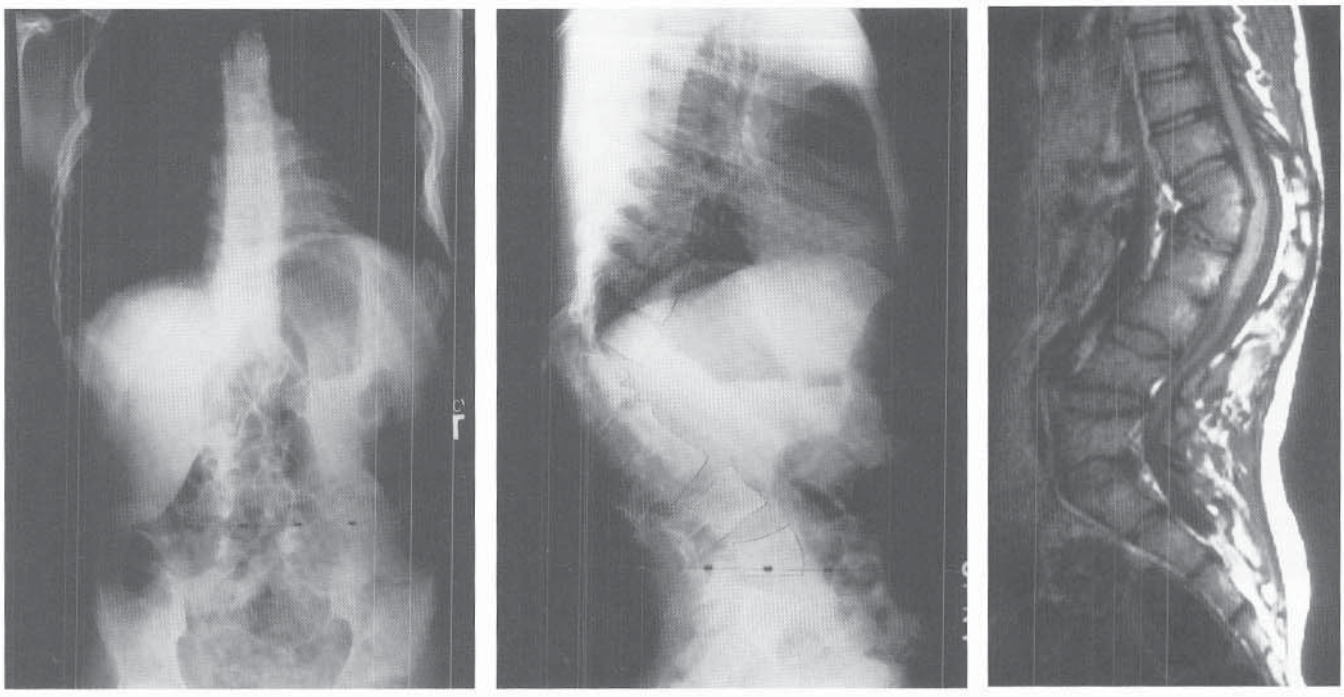


D

FIGURE 11–66 Chest radiographs obtained in a 14-month-old girl to evaluate an upper respiratory tract infection showed an abnormality at T11. On further radiographic evaluation, the abnormality was determined to be kyphosis due to failure of vertebral body formation (**A–C**). A 51-degree kyphosis was measured. MRI demonstrated abrupt angulation of the spinal cord at this level (**D**). The child was neurologically normal.



A B C

FIGURE 11–67 This neurologically normal 14-year-old boy had back pain and progressive worsening of his back appearance. Radiographs demonstrated failure of segmentation at T12–L1 and L2–3 with a resultant 82-degree localized kyphosis (**A, B**). MRI showed a normal spinal cord (**C**).

progressive.[293,353,488] Deformities due to two adjacent type I vertebral anomalies will progress more rapidly and with more severity than deformities due to a similar single anomaly.

Kyphosis due to failure of segmentation (type II) is much less progressive, produces less severe deformity, and has a very low likelihood of resulting in paraplegia.

Clinical Features. Although congenital kyphosis has been diagnosed prenatally, it may not be clinically evident in the newborn or infant.[420] Suspicion may first be raised following examination of a chest radiograph obtained for evaluation of an unrelated event, such as a respiratory infection. As the child begins standing and walking, a localized prominence may become noticeable or palpable. The child is usually asymptomatic and has no spinal tenderness. In the adolescent, the predominant clinical complaint tends to be lower back discomfort caused by secondary lumbar hyperlordosis. A mild scoliosis may accompany the kyphosis.

On occasion, the child with a congenital kyphosis may develop a myelopathy or paraplegia secondary to spinal cord compression. Reports of mild trauma producing the sudden onset of paraplegia in children who have unrecognized acute type I kyphosis make one appreciate the delicate underlying neurologic status in this condition. When congenital kyphosis due to a defect in vertebral formation is diagnosed, a meticulous neurologic examination should be performed to identify any subtle abnormalities. Plans for surgical intervention should begin immediately.

Radiographic Evaluation. Congenital kyphosis is best visualized on a lateral radiograph of the spine. It may not be evident on the frontal plane view. Once identified, a coned-down lateral view of the specific area provides greater bone detail.

MRI provides the clearest picture of the spinal cord and vertebral bodies in very young children. It should be ordered immediately for those whose kyphosis is due to failure of formation (see Fig. 11–66). Spinal cord compression may be evident on MRI before any clinical neurologic deficits become apparent. Three-dimensional CT imaging of the spine with reconstructed images is very useful in the evaluation of the vertebral anomalies, especially in the older child. Both of these tests (MRI and CT) should be performed before any operative intervention is undertaken.

Treatment. Nonoperative treatment has no beneficial effect on congenital kyphosis, and the use of an orthosis is inappropriate. Once kyphosis due to type I or type III abnormalities is recognized, plans for surgical intervention should be made. For adolescents with mild type II kyphosis (failure of segmentation), close monitoring for progression is reasonable. If the deformity is recognized at a younger age, operative intervention should be considered.

FAILURE OF FORMATION (TYPE I). Defects of formation are more common than defects of segmentation, can lead to more severe deformity, and have a greater potential for producing paraplegia. For these reasons, once this form of congenital kyphosis is diagnosed, surgical intervention is indicated, even in the infant. The main goal is to prevent paraplegia. All other goals, such as improved spinal alignment and cosmetic appearance, are secondary.

If the kyphosis is recognized in a child less than 5 years old and is less than 45 degrees, simple posterior fusion without instrumentation may be considered. A hyperexten-

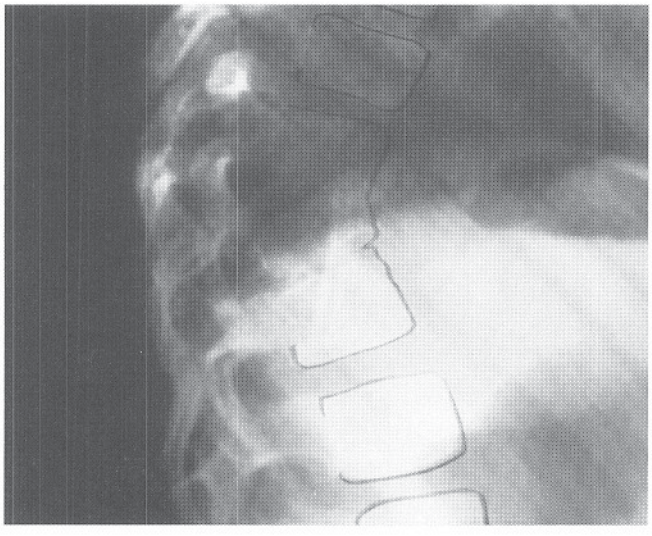
sion cast is used postoperatively for 4 to 6 months, followed by a TLSO for another 6 months. Successful outcomes with posterior fusion have been reported.[293,485] This approach allows some growth to occur anteriorly in the abnormal region of the spine, which over time may result in progressive improvement in the localized kyphosis. Reexploration and augmentation of the graft at 6 months has been advocated.

An alternative approach for this same young child is to combine an anterior fusion using a rib strut with posterior fusion during the same surgical intervention (Fig. 11–68). This approach produces some improvement in the sagittal plane alignment immediately and increases the likelihood of a solid fusion. It eliminates any further correction that might occur from anterior growth. In the older child or the adult, the combination of anterior and posterior arthrodesis is mandatory.[489] The anterior arthrodesis is performed first. Following excision of the gristle-like soft tissue anteriorly, some distraction is attempted. Any distraction that is achieved can then be maintained with rib-strut grafts. Vascularized rib-struts heal more rapidly, should always be used in those who have previously unsuccessful attempts at anterior fusion, and may be considered for the initial fusion procedure (Fig. 11–69).[47,49,401] Spinal cord monitoring is essential. In the older child, instrumentation may be considered during the posterior arthrodesis if it is not too prominent.

As with any congenital vertebral anomaly, associated dysraphic spinal lesions should be ruled out prior to surgical intervention. Preoperative MRI accomplishes this.

If a neurologic deficit is present at the time the congenital kyphosis is recognized, treatment should be undertaken immediately. If this deficit is minimal (increased reflexes, positive Babinski's sign, but no loss of motor, bowel, or bladder function), a formal anterior decompression of the spinal cord is not necessary. Following a solid anterior and posterior arthrodesis, the subtle neurologic deficits have the potential to resolve. On occasion, patients present with mild paraparesis of recent onset. In these individuals the apical flexibility of the kyphotic deformity should be assessed with a hyperextension radiograph. If the apex is flexible, some improvement in the paraparesis may be achieved by placing the recently compromised spinal cord at rest with, for example, a halo vest, cast, or, on occasion, minimal halo traction.[108] *Halo traction should not be considered in those with a rigid, inflexible kyphotic apex because of the risk of progressive neurologic deterioration.* Very close monitoring is needed. If recovery occurs, then spinal fusion anteriorly and posteriorly can be performed without the need for decompression. If the deficits do not resolve, then anterior and posterior arthrodesis must be combined with an anterior decompression of the spinal cord. Unless the child is very small at the time of surgery, these procedures can be accomplished during the same operative episode. The decompression must be performed anterior to the compressed cord by removing the posterior aspect of the vertebral body. Posterior laminectomy will not relieve the spinal cord compression.

FAILURE OF SEGMENTATION (TYPE II). Defects of segmentation are best treated at a young age, before significant deformity has developed. Posterior spinal fusion, followed by cast immobilization, is all that is needed. The fusion should span the unsegmented levels plus one level further both cephalad and caudad. Correction of the kyphosis should not be expected, although mild improvement from the cephalad and caudal extensions is possible.[217] In young children, their size



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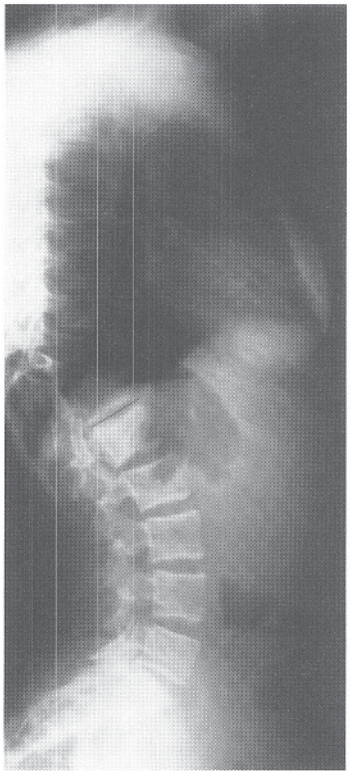


B

FIGURE 11–68 The patient whose imaging findings are shown in Figure 11–66 was treated with an anterior rib strut graft between T10 and T12, followed by posterior in situ fusion. A small amount of correction of the kyphosis was achieved. Two years postoperatively the kyphosis measured 35 degrees and the rib strut had been incorporated but was still visible radiographically **(A)**. Ten years postoperatively the kyphosis remained unchanged **(B)**. The patient is neurologically normal.

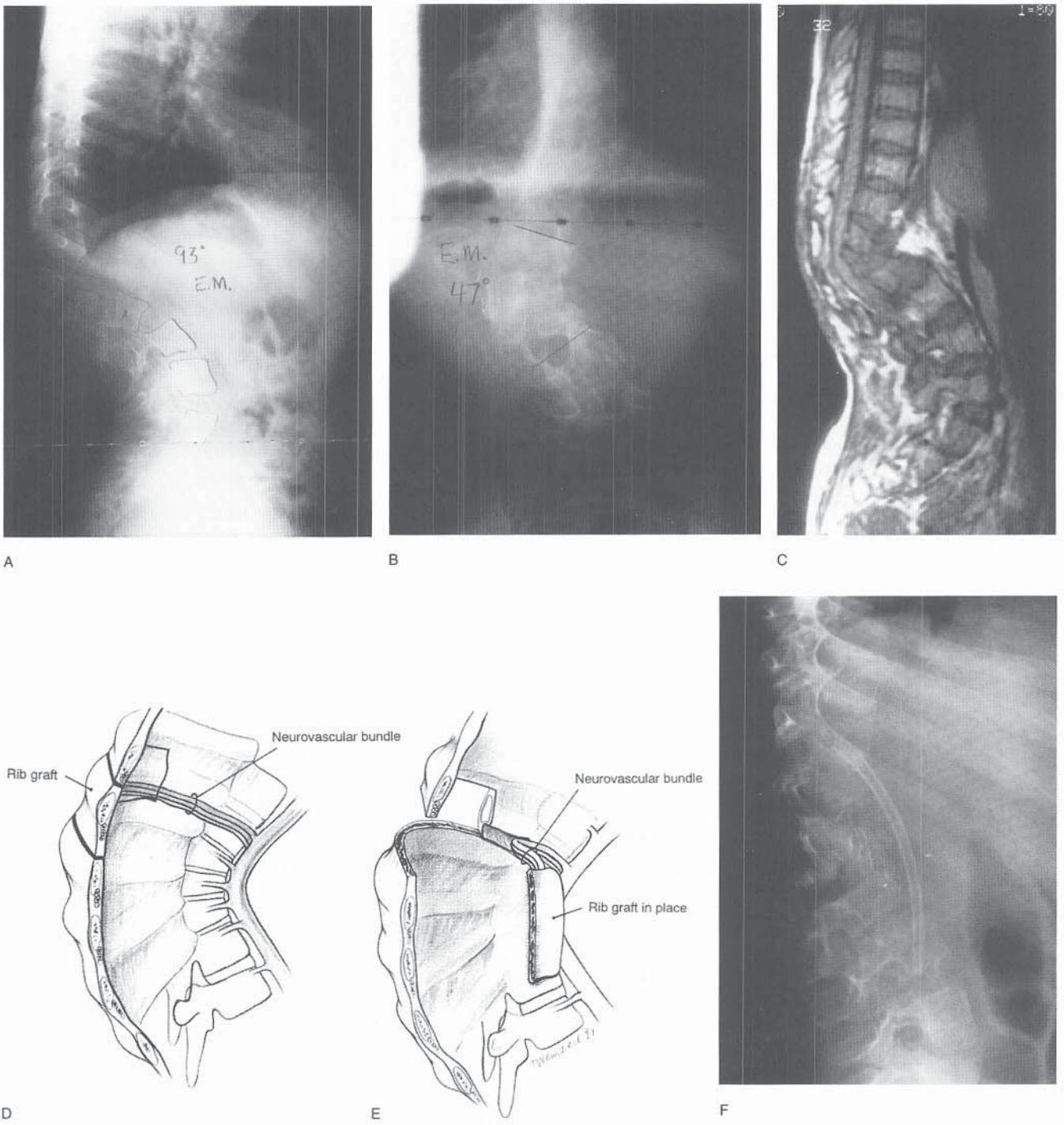
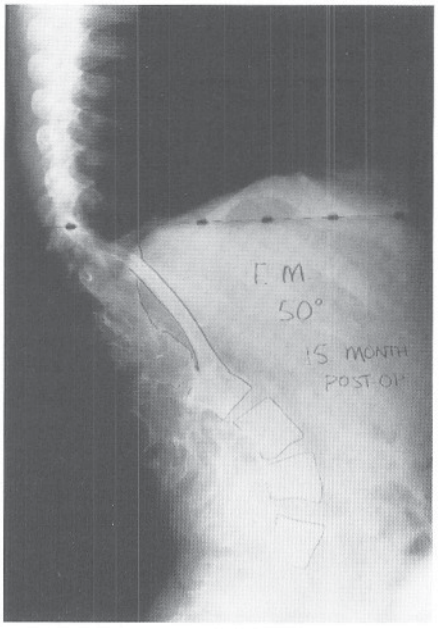


FIGURE 11–69 **A** and **B,** Radiographic appearance in a 10-year-old boy with spondyloepiphyseal dysplasia and an abrupt 93-degree kyphosis at L1 (and 47-degree scoliosis) due to a small posterolateral quadrant vertebra. He had previously undergone an attempt at anterior and posterior fusion with posterior instrumentation; that intervention was complicated by infection. The hardware was removed and the deformity worsened. He remained neurologically normal. **C,** MRI demonstrated the abrupt kyphosis and its effect on the spinal cord. **D** and **E,** A vascularized rib strut graft was used during the repeat anterior and posterior fusion. The neurovascular bundle was isolated from the rib near its origin to allow the rib to be cut. The rib selected for a graft usually corresponds to the upper vertebral level requiring fusion (D). When the rib is seated into the vertebral bodies, the vascular bundle must be free of tension. The ends of the ribs should be exposed subperiosteally for a length of 1 cm to allow secure fixation into the vertebral bodies (E). (D and E from Herring JA: Anterior spinal surgery. In Weinstein SL (ed): The Pediatric Spine: Principles and Practice, p 1419. New York, Raven Press, 1994. Copyright © 1994 by Lippincott, Williams & Wilkins.) **F,** The vascularized rib was fixed in the T10 and L3.



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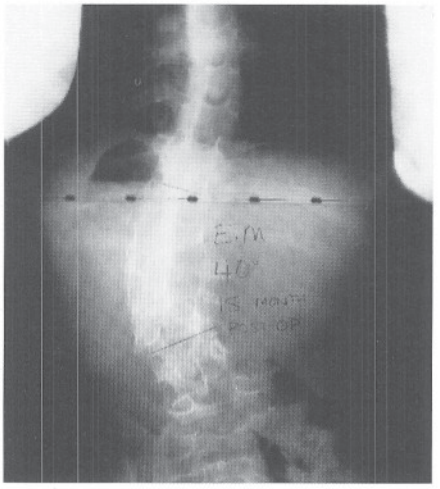


H

FIGURE 11–69 *Continued.* **G** and **H,** Fifteen months postoperatively, the kyphosis was stable at 50 degrees and the scoliosis was at 40 degrees. Five years later the radiographic appearance was unchanged and the patient remained neurologically normal.

precludes the use of instrumentation. In the older child or adolescent, posterior compression instrumentation may lessen the need for external immobilization.

In the older child with severe kyphosis, some correction of the deformity may be achieved through osteotomy of the unsegmented anterior region. When combined with posterior compression instrumentation, this approach may result in some improvement in the sagittal plane.

MIXED ANOMALIES (TYPE III). Mixed anomalies are least common but, when present, usually produce a kyphoscoliotic deformity. Because of their association with failure of segmentation, type III anomalies usually require posterior arthrodesis only.

SEGMENTAL SPINAL DYSGENESIS, CONGENITAL DISLOCATION OF THE SPINE, AND CONGENITAL VERTEBRAL DISPLACEMENT

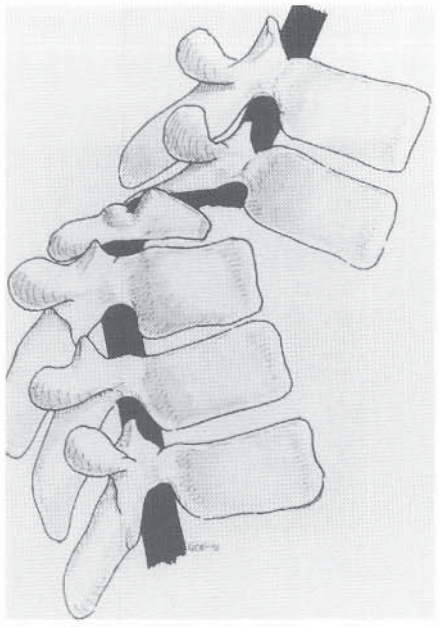
Segmental spinal dysgenesis, congenital dislocation of the spine, and congenital vertebral displacement of the spine are conditions that create the most severe localized kyphosis of the spine and lead to a neurologic deficit in 50 to 60 percent of patients.[124,139,191,403,505] These conditions can be difficult to differentiate from one another.

Segmental spinal dysgenesis is characterized by a focal spinal deformity, usually located at the thoracolumbar junction or the upper lumbar spine.[124,139,140,191] The deformity frequently includes severe kyphosis; either anterior, posterior, or lateral subluxation of the spine; scoliosis in association with the severely stenotic spinal canal; and absent nerve roots. All of these patients have a localized stenosis of the spinal canal at the level of the involvement and the osseous canal has an hourglass shape. No pedicles, spinous processes, or transverse processes are seen at the level of involvement. Commonly, there is an offset in the sagittal plane between the cephalad and caudad segments of the spine at the level of the dysgenesis. Decompression of the stenotic canal results in some improvement in neurologic function in 20 percent of patients. Early anterior and posterior arthrodesis in patients with segmental spinal dysgenesis is indicated, as progressive kyphosis inevitably develops, often resulting in the neurologic deficits.

Congenital kyphosis due to failure of formation (type I) can be similar to, and may be confused with, segmental spinal dysgenesis. Whereas type I congenital kyphosis represents failure of formation of the vertebral body, the pedicles and posterior elements are present. The associated severe spinal stenosis of segmental spinal dysgenesis is not present in congenital kyphosis. Although many patients with segmental spinal dysgenesis have a fixed neurologic deficit, neurologic function in patients with congenital kyphosis is generally good at birth, with paraplegia subsequently developing as a result of untreated instability and worsening kyphotic deformity.

Congenital vertebral displacement occurs when the spinal column is displaced at a single vertebral level, resulting in an abrupt displacement of the neural canal (Fig. 11–70).[403] This displacement can occur in the presence of a posteriorly located hemivertebra in which the pedicles, transverse processes, and spinous processes can be present. As in segmental spinal dysgenesis, the potential for severe neurologic deficits is quite high. For this disorder, combined anterior and posterior arthrodesis of the spine is needed in an effort to prevent the development of neurologic deficits. For those with neurologic deficit of recent onset or progressive neurologic deficit, decompression of the spinal cord is indicated.

The *congenital dislocated spine* was first described in 1973 by Dubousset.[505] Like the other disorders described above, it is associated with spinal kyphosis and a high likelihood of neurologic deterioration. The posterior elements are abnormal in all patients with congenital dislocated spine. The various stages of posterior dysraphism range from agenesis of the lamina with pathologic changes of the articular facets



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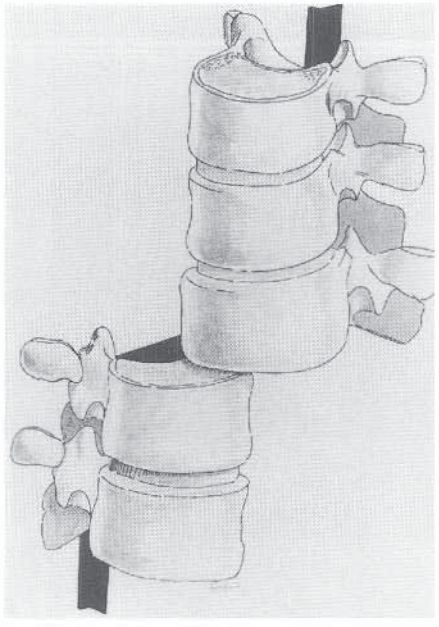

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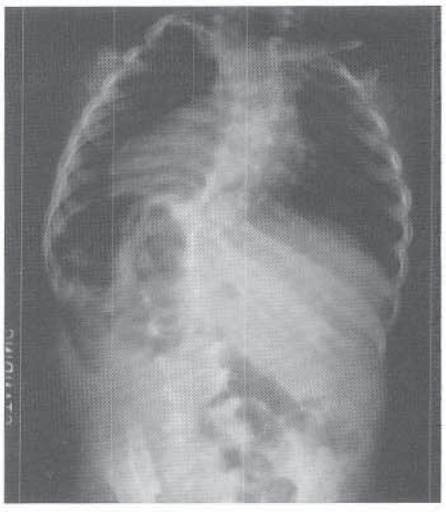

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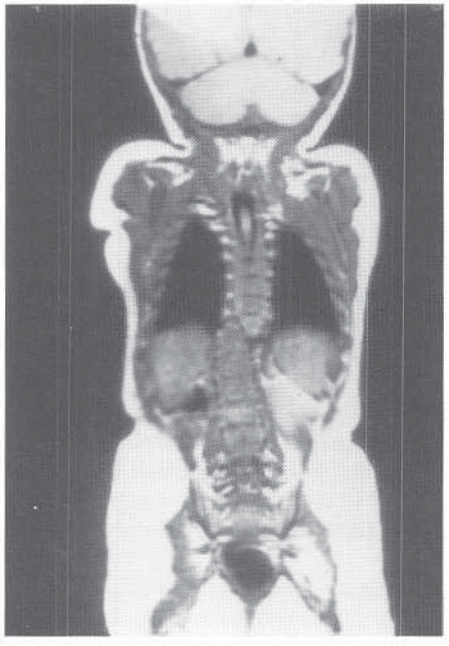


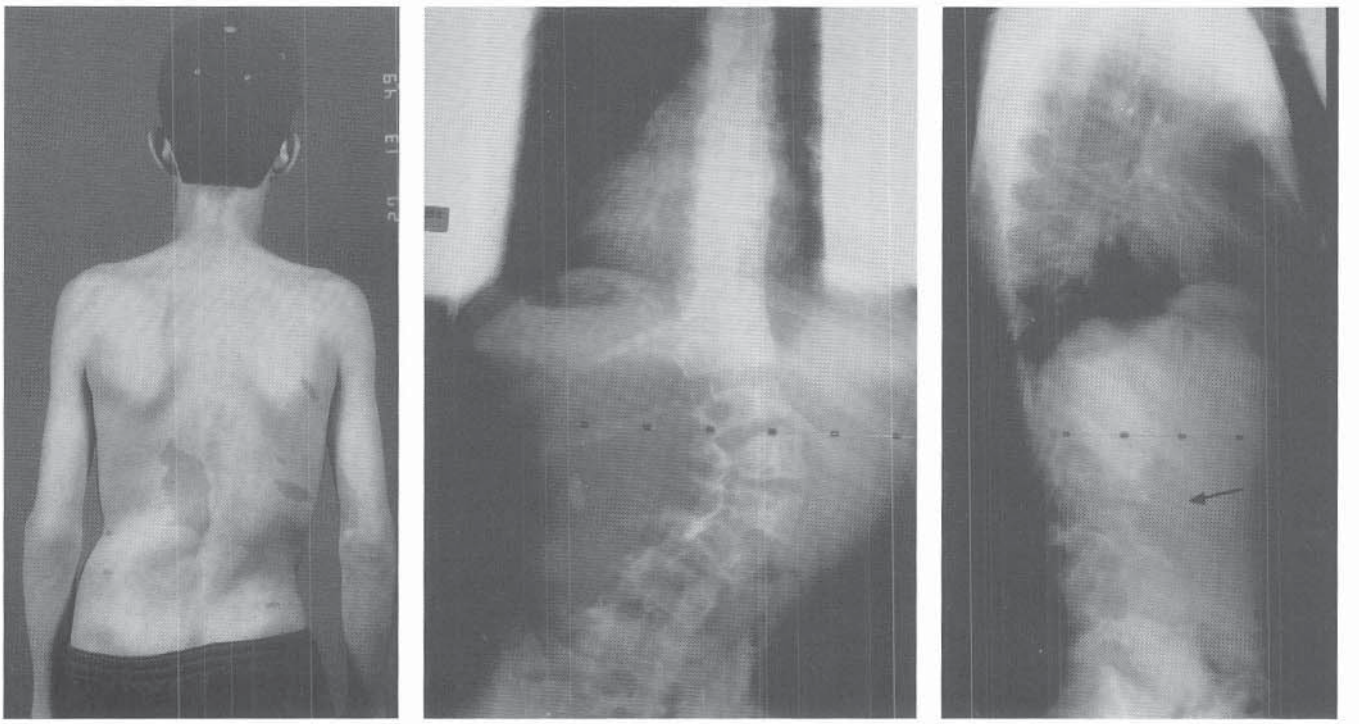
D

E

FIGURE 11–70 There are two types of congenital vertebral displacement. In type A, there is posterior displacement of the caudal vertebrae with anterior angulation of the spinal canal. A hemivertebra is noted in the area of the deformity **(A)**. In type B, there is rotatory subluxation with resultant rotatory translation of the neural canal **(B)**. (A and B from Shapiro J, Herring J: Congenital vertebral displacement. J Bone Joint Surg 1993;75-A:656.) The type B deformity is illustrated by a girl age 2 years 8 months with an abrupt congenital vertebral displacement at the T10 level **(C)**. This was further demonstrated on MRI **(D)** and CT **(E)**.

to total absence of posterior elements with the spinal cord under an otherwise normal skin. This description is similar to that of segmental spinal dysgenesis. Anterior and posterior spinal fusion is indicated, as posterior spinal fusion alone is insufficient to achieve solid fusion in this type of congenital instability. Exploration and augmentation of the posterior fusion mass should be considered because of a high rate of occurrence of pseudarthrosis in this abnormality, as with the others. No sudden extemporary correction should be attempted in older patients with severe angular kyphosis and progressive neurologic deficit. Function must be favored over cosmetic appearance. Neurosurgical decompression should be used only for a proven recent and progressive neurologic deficit.

These three entities all represent very severe forms of localized kyphosis in the spine. They share similar characteristics, and differentiation between them may be difficult. Early recognition of their severity is imperative and the appropriate operative intervention should be undertaken.



A B C

FIGURE 11–71 **A,** Clinical appearance of a 16-year-old male patient with neurofibromatosis and a thoracolumbar scoliosis with the convexity to the right. **B** and **C,** PA and lateral radiographs of the spine demonstrating the dystrophic nature of the curve, as evidenced by erosions of the vertebral bodies.

Other Causes of Scoliosis

NEUROFIBROMATOSIS

Scoliosis is the most common skeletal manifestation of neurofibromatosis (Fig. 11–71).[88,89,218] Typically it is located in the thoracic spine, has a short, sharply angled curve, and involves four to six vertebra. The reported incidence is between 10 and 60 percent.[3,72,88,104,183,414] The higher incidence rates reported may have been biased in that they were derived from populations of neurofibromatosis patients followed by musculoskeletal specialists. The most recent report (10 percent incidence of scoliosis) appears to reflect more accurately the entire neurofibromatosis population.[3]

The cause of spinal deformity in neurofibromatosis is unknown. Proposed explanations include primary mesodermal dysplasia, osteomalacia, erosion or infiltration of the bone by localized neurofibromatosis tumors, and endocrine disturbances.[88]

Neurofibromatosis scoliosis can be either dystrophic or nondystrophic, depending on accompanying abnormalities specific to this disorder.* Differentiation between the two is important because the prognosis and management differ significantly. *Dystrophic scoliosis* is more common, has a greater tendency to progress, and includes a subgroup (those with severe kyphoscoliosis) at risk for developing neurologic deficits.[414] *Nondystrophic scoliosis* more closely resembles idiopathic scoliosis in both its curve patterns and its behavior. Recently it has become clear that nondystrophic curves in younger children can modulate into the more worrisome dystrophic type over the course of several years.[88]

Nondystrophic Scoliosis. This group has the more favorable outlook among neurofibromatosis patients affected by scoliosis. The clinical appearance, radiographic findings, and behavior of the curve tend to be similar to those found in idiopathic scoliosis. However, nondystrophic deformities usually become apparent at an earlier age than idiopathic curves and have a slightly higher likelihood of progressive deformity. The management of nondystrophic curves is similar to that for idiopathic scoliosis. Curves less than 25 degrees can be observed closely without active intervention. Brace treatment appears to be effective for skeletally immature individuals with curves between 25 and 40 degrees.[218] However, once nondystrophic curves of neurofibromatosis exceed 40 degrees, posterior spinal fusion with instrumentation is recommended (Fig. 11–72). Close follow-up after surgery is needed, for two reasons. First, there is a higher likelihood of pseudarthrosis in this population. Second, over time, some of the nondystrophic curves evolve to show characteristics of dystrophic scoliosis.

Dystrophic Scoliosis. In dystrophic scoliosis, short, sharply angled curves develop at an early age, often as young as 3 years. Radiographic features that help to differentiate dystro-

*See references 66, 88, 142, 218, 232, 331, 414, 465.

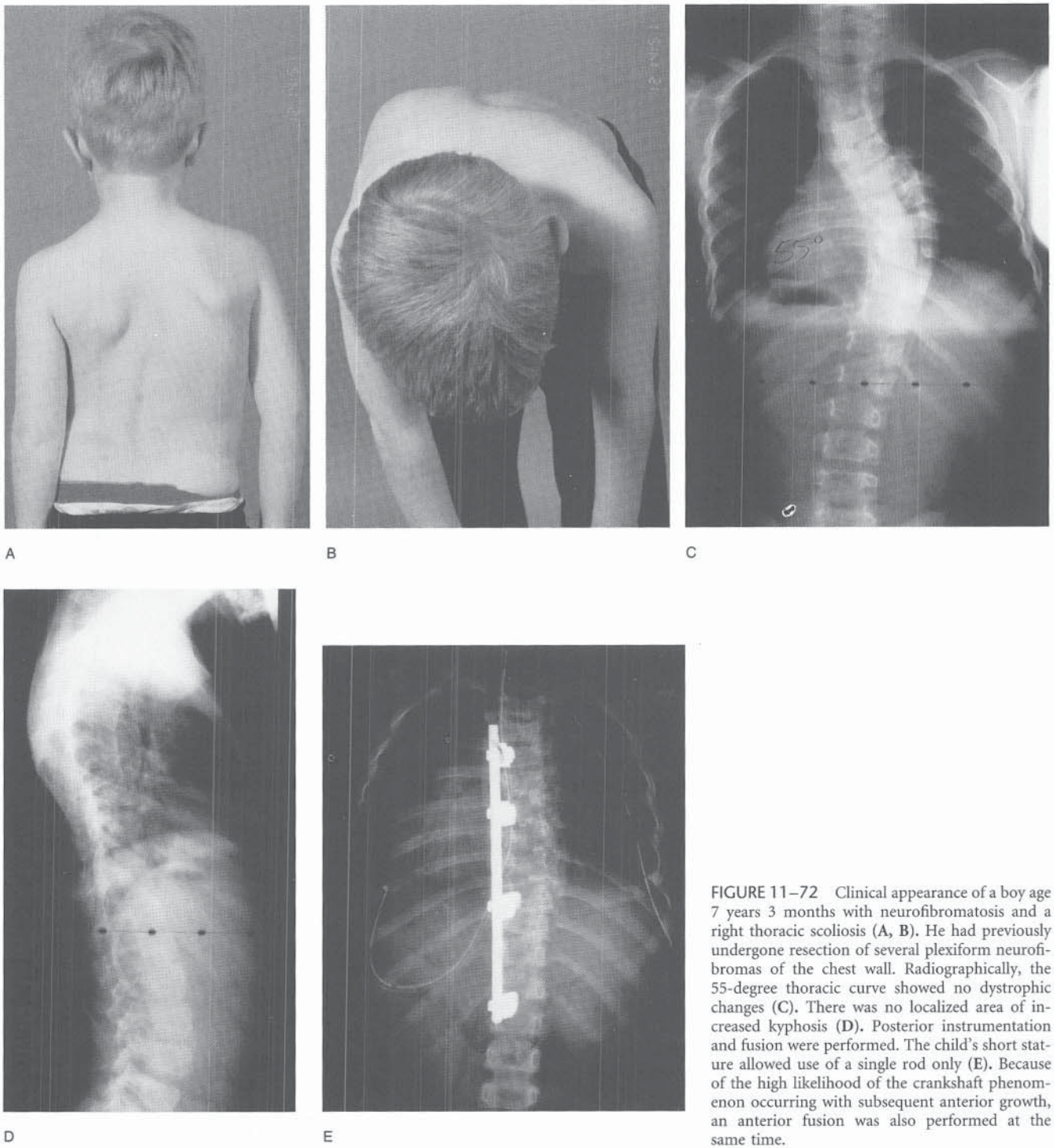


FIGURE 11–72 Clinical appearance of a boy age 7 years 3 months with neurofibromatosis and a right thoracic scoliosis (**A, B**). He had previously undergone resection of several plexiform neurofibromas of the chest wall. Radiographically, the 55-degree thoracic curve showed no dystrophic changes (**C**). There was no localized area of increased kyphosis (**D**). Posterior instrumentation and fusion were performed. The child's short stature allowed use of a single rod only (**E**). Because of the high likelihood of the crankshaft phenomenon occurring with subsequent anterior growth, an anterior fusion was also performed at the same time.

phic curves from nondystrophic curves include vertebral scalloping, spindled transverse processes, severe apical vertebral wedging and rotation, foraminal enlargement, defective pedicles, penciling (narrowing of the proximal portion) of the ribs, the presence of paravertebral soft tissue lesions, and, rarely, subluxation between vertebral bodies. Some of these findings seen in dystrophic scoliosis may result from direct erosion of the bone by intraspinal neurofibromas, paraspinal neurofibromas, or dural ectasia. Dural ectasia is an expansion in the width of the thecal sac thought to be due to an increase in hydrostatic pressure.

Fortunately, most dystrophic curves are not accompanied by an excessive amount of kyphosis.[414] If this combination is present, there is a significant potential for developing neurologic deficits. Kyphosis may occur in one of two ways. An abrupt angular kyphosis may be present from the very early stages of the deformity, or there may be a more gradual kyphosing scoliosis that results from progression and rota-

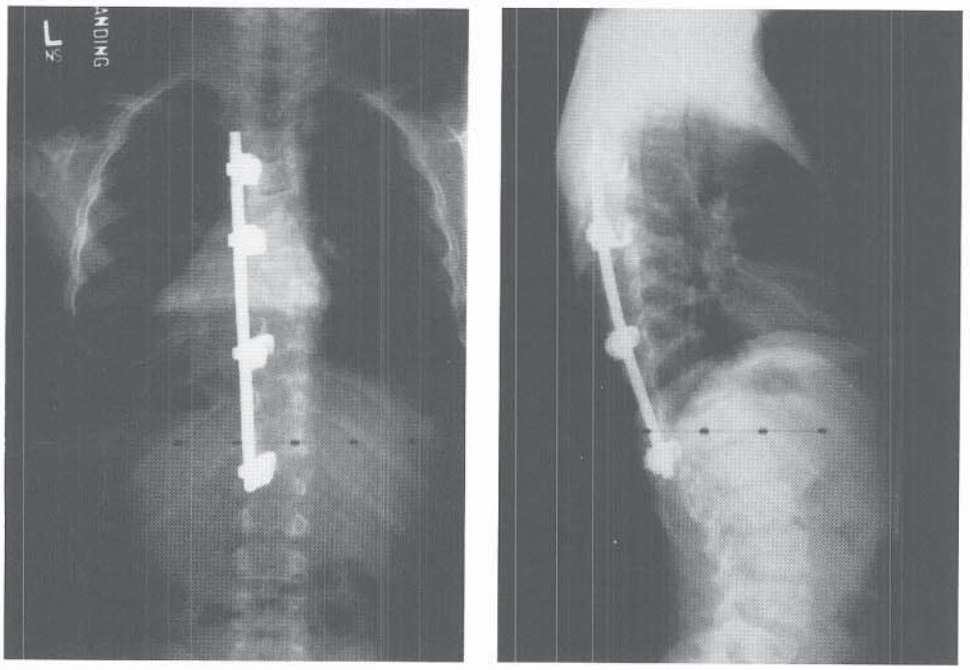


FIGURE 11–72 *Continued.* Two years later the curve correction was maintained and solid fusion was evident anteriorly (**F**). The sagittal plane alignment remained satisfactory (**G**).

F G

tion of the scoliosis (Fig. 11–73).[142] Recognition of either of these is important because, once established, they always require prompt combined anterior and posterior spinal fusions.

Treatment for Dystrophic Scoliosis. Nonoperative management of dystrophic scoliosis is almost always unsuccessful. These curve patterns need early and aggressive surgical intervention, even in the younger child. Delay only leads to progressive deformity, which may be as rapid as 8 degrees per year in the frontal plane and 11 degrees per year in the sagittal plane.[66] Most patients will have marked progression before the age of 10 years. One need not wait until the adolescent growth spurt to witness severe deformity. Characteristics of dystrophic scoliosis that correlate with an excessive risk of progression include an early age at onset, a high Cobb angle at the time of initial evaluation, the presence of vertebral scalloping, penciling of multiple ribs, and apical vertebral rotation exceeding 11 degrees (Perdriolle measurements[346]).[142]

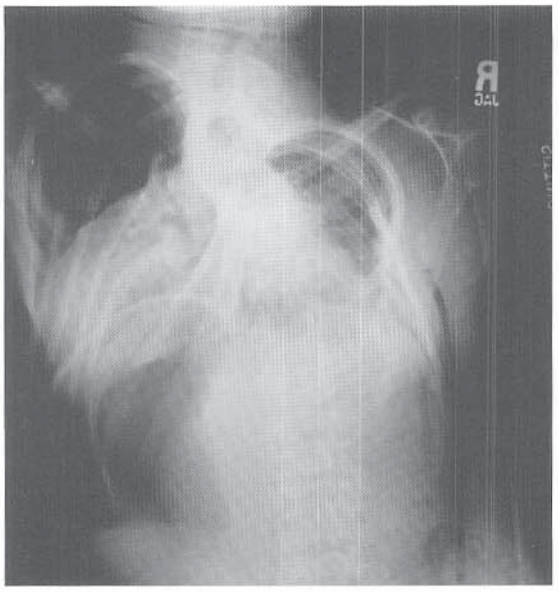
Prior to surgery, a thorough neurologic examination is essential to identify any subtle abnormalities. MRI and CT should always be performed. MRI will demonstrate neurofibromatosis lesions in the thorax, paravertebral region, neural foramina, or spinal canal. Dural ectasia, pseudomeningoceles, and spinal cord compression (due to localized kyphosis, rib impingement, or mass effect from neurofibromas) can also be detected using MRI. CT demonstrates scalloping of the vertebral bodies anteriorly, erosion of the posterior portion of the vertebral body and/or lamina from dural ectasia, and the presence of ribs within the spinal canal (Fig. 11–74). Three-dimensional CT reconstruction is invaluable in clarifying the anatomy of severe deformities and is helpful in preoperative planning.

Posterior spinal fusion with instrumentation alone can be used for certain patients with dystrophic curves.* These patients have curves between 20 and 50 degrees with a kyphosis of less than 50 degrees (no sharp angulation). Because the risk of developing pseudarthrosis remains higher than in the idiopathic population, consideration should be given to performing imaging studies (tomography) 6 months after surgery. If the fusion mass appears inadequate, repeat bone graft augmentation may be necessary.

Anterior fusion in addition to posterior fusion is needed for the majority of patients with dystrophic curves. The combination of anterior and posterior fusion increases the likelihood of successful fusion. Longer fusions are generally indicated, even in young patients. Curve progression in patients with neurofibromatosis can occur even in the presence of a solid arthrodesis.[465]

Severe kyphoscoliosis absolutely requires anterior fusion in addition to posterior fusion. Thorough anterior diskectomy, bone grafting, and rib (or tibia) strut placement are needed (Fig. 11–75). In some patients with exaggerated kyphosis, the apical rotation may be so severe that the vertebral body faces posterolaterally. With this deformity, placement of the strut graft can be extremely difficult, and the anterior approach to the spine may need to be from the concave side. Vertebral body erosion secondary to intrathoracic neurofibroma or dural ectasia can also significantly interfere with anterior exposure and fusion. Dysplastic posterior elements limit the ability to achieve strong posterior internal fixation. Every effort should be made to stabilize the spine, as stabilization improves the likelihood of a successful outcome. Postoperative immobilization in a cast or orthosis is clearly indicated when the vertebrae are weak, the severity and location of the kyphosis cause an excessive strain at certain hook sites, or the quality of bone does not allow

*See references 40, 88, 218, 331, 408, 465.



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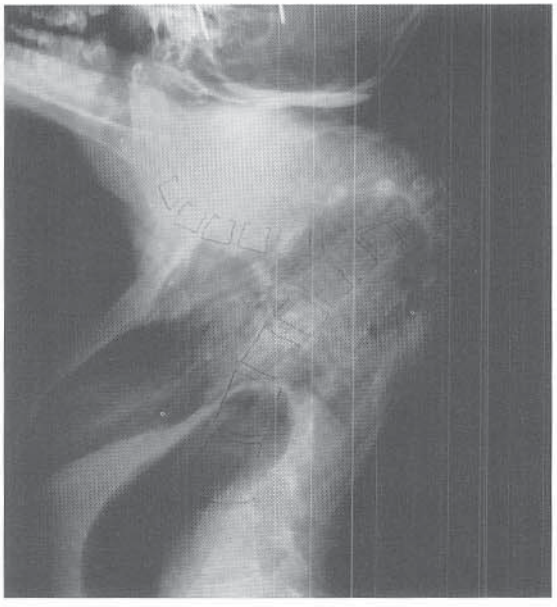

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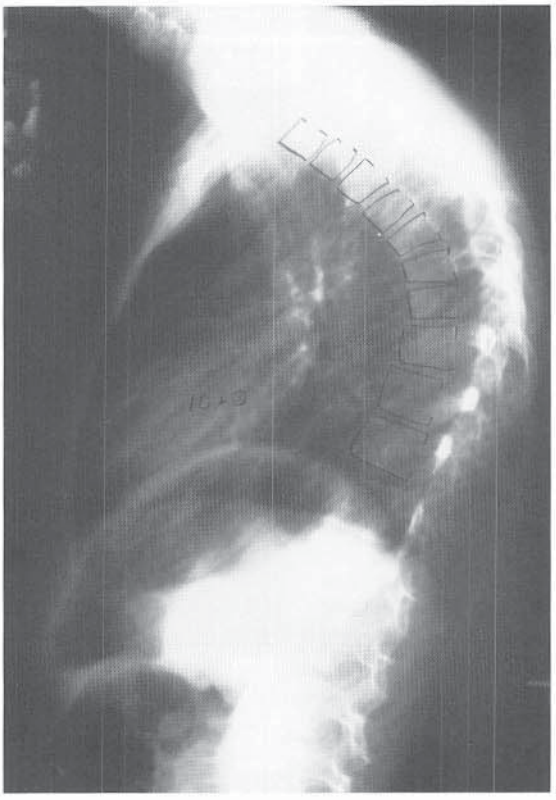

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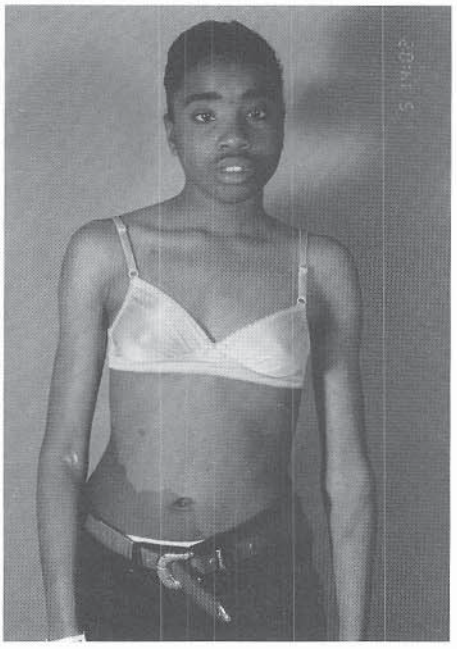
FIGURE 11–73 **A** and **B,** Severe kyphosis in a 15-year-old girl with neurofibromatosis. She remains ambulatory but complains of progressive weakness. **C,** A radiograph obtained elsewhere at age 10 years showed a scoliosis of approximately 60 degrees and a moderate thoracic kyphosis, but no intervention was undertaken.

fixation points for instrumentation. Despite meticulous attempts at anterior and posterior fusion, pseudarthrosis remains a significant concern.[88,414,486]

Excessive kyphosis is the most frequent cause of neurologic deficits in patients with neurofibromatosis and spinal deformities. Should a neurologic deficit be present, partial vertebrectomy is needed to decompress the spinal cord. Laminectomy for spinal cord decompression or prophylactic laminectomy for kyphoscoliosis should be avoided as it destabilizes the spine, increases the kyphosis, removes bone stock needed for successful posterior fusion, and, most important, does not relieve the anterior compression on the spinal cord. Neurologic deficits can also result from impingement on the spinal cord by neurofibroma lesions within the spinal canal.[355] Differentiating impingement by neurofibroma lesions from kyphotic impingement is needed in order to correctly address the problem surgically. MRI should help clarify the difference.

MARFAN'S SYNDROME

Marfan's syndrome, one of the more common connective tissue disorders, has a 0.01 percent prevalence in the general population.[424] Scoliosis is the most common spinal deformity in this condition, with a prevalence approaching 50 to 55 percent.[382,383,424,434] In addition, 6 percent of the Marfan popu-



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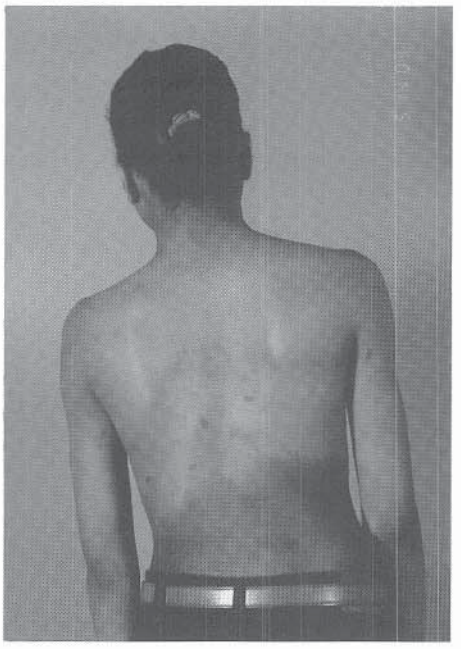

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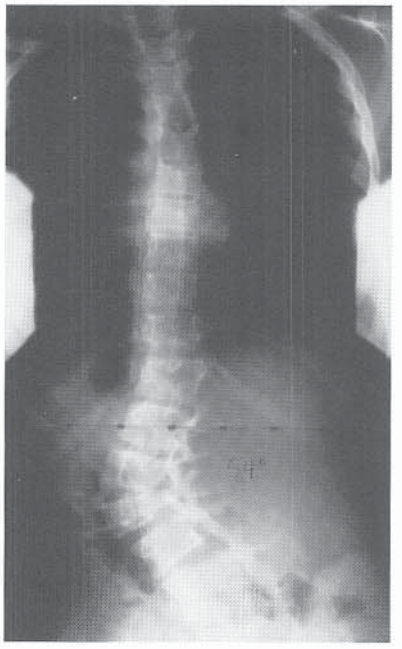

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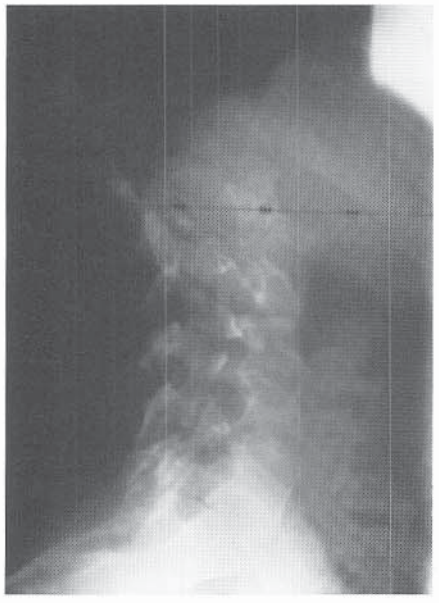

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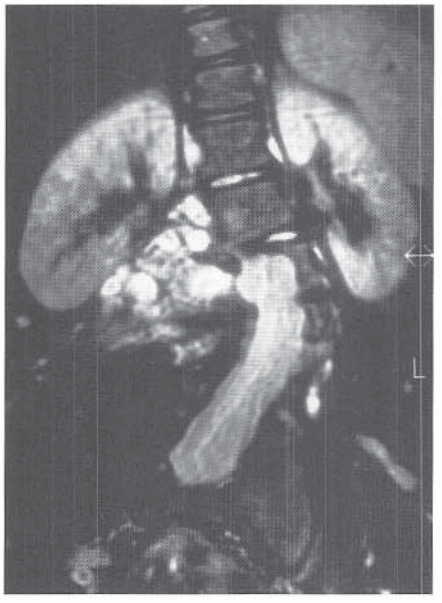

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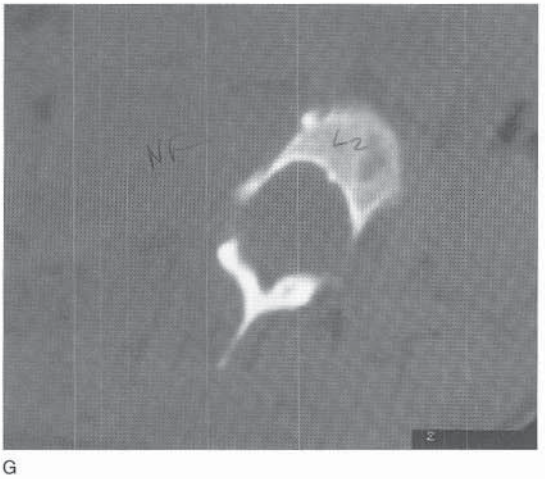

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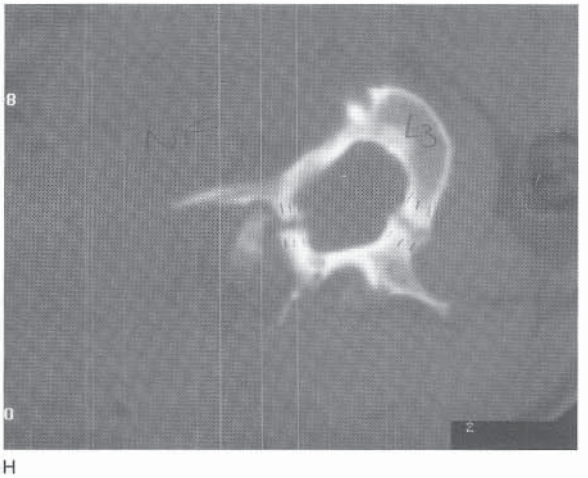

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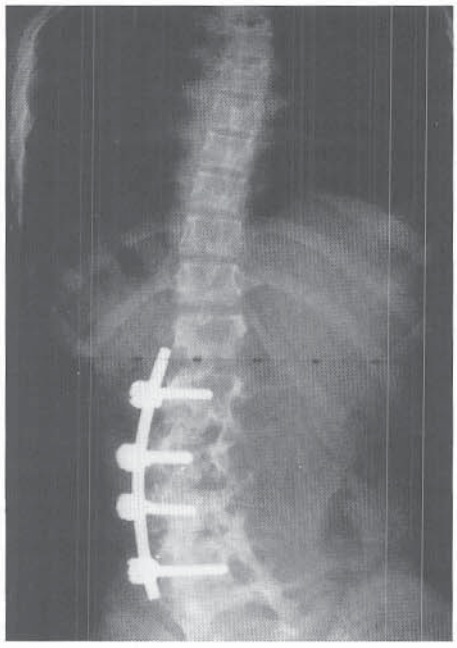
FIGURE 11–74 *See legend on following page*



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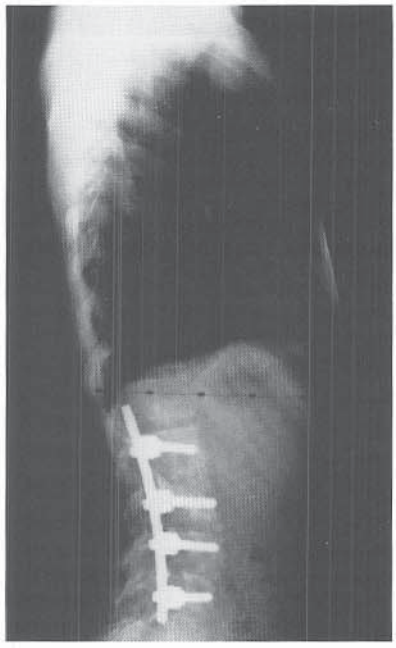

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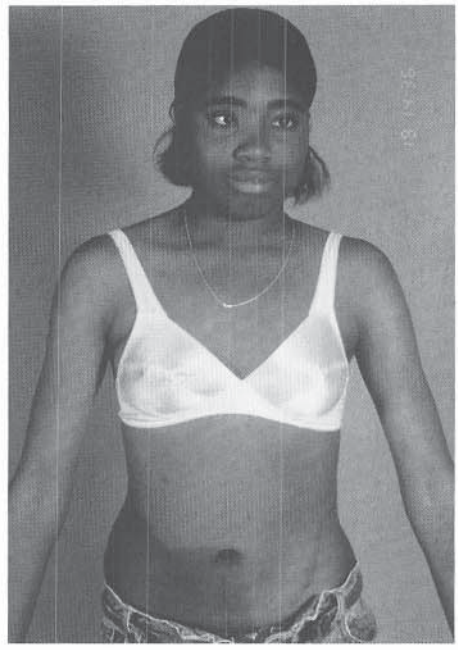

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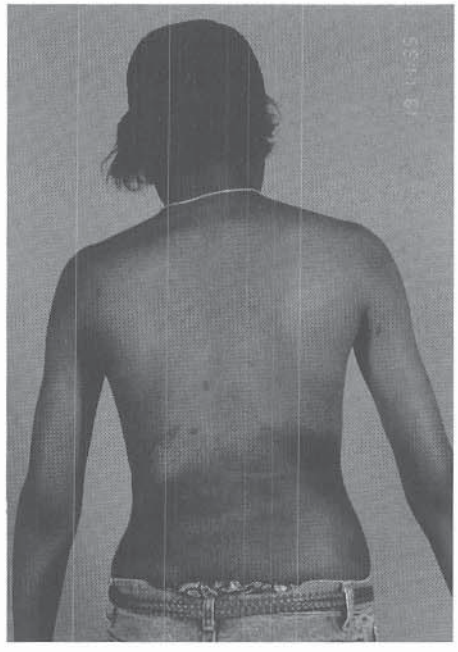

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FIGURE 11–74 *Continued.* **A** and **B,** Clinical appearance of a girl age 14 years 4 months with an extremely large café-au-lait discoloration on the trunk. **C** and **D,** Radiographs showed 54-degree lumbar scoliosis with flattening of the L3 body and posterior scalloping of numerous lumbar vertebrae. **E** and **F,** Preoperative MRI showed a large neurofibroma in the concavity of the lumbar spine. **G** and **H,** CT demonstrated significant enlargement of the spinal canal, narrow depth of the vertebral bodies, thin posterior elements, and spondylolysis of the pars and pedicle of L3. **I–L,** Anterior instrumentation and fusion between L1 and L4 improved the patient's frontal and sagittal plane balance.

lation has spondylolisthesis. Although Marfan's syndrome is an autosomal dominant disorder, no familial pattern of scoliosis has been identified.

The curve patterns in scoliosis seen in Marfan's syndrome are similar to those seen in idiopathic scoliosis, although there is a slightly higher rate of triple curves and thoracolumbar curves (Fig. 11–76). Scoliosis is equally distributed between males and females, in contrast to the female predominance in idiopathic scoliosis.

The sagittal profile of the spine in Marfan's syndrome varies widely and requires close examination when planning treatment. Increased lordosis in the thoracic spine has been thought common,[472,477] but a recent report found that increased thoracic kyphosis (greater than 50 degrees) may be found in as many as 45 percent of patients with Marfan's syndrome and scoliosis.[424] When the transitional point between thoracic kyphosis and lumbar lordosis is below the second lumbar vertebra, a long, broad thoracic kyphosis is evident. If a localized kyphosis exists at the thoracolumbar junction, the thoracic spine above generally is hypokyphotic.

Back pain is more frequent in patients with Marfan's

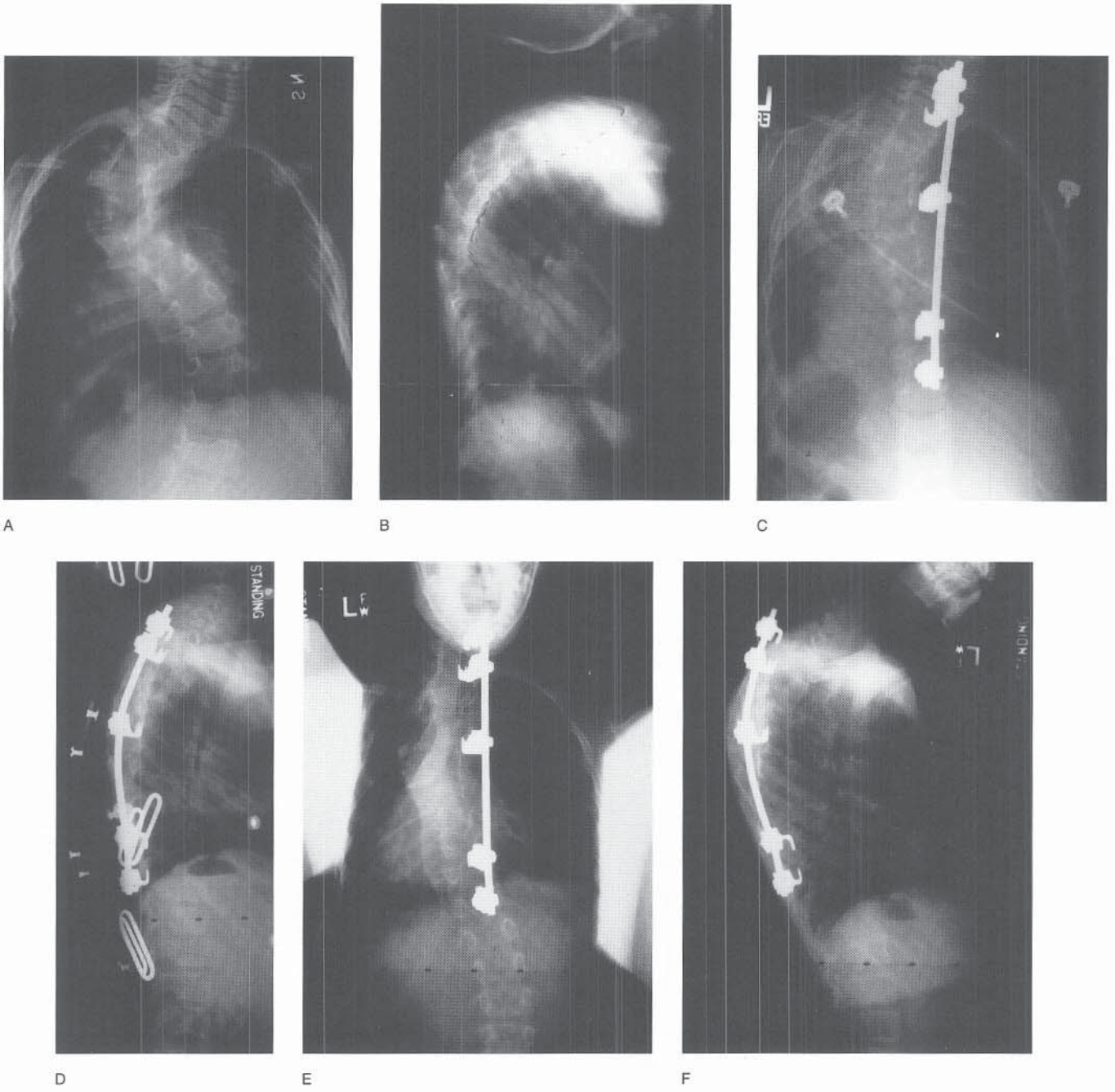


FIGURE 11–75 **A** and **B,** Radiographic appearance of a boy age 3 years 2 months with neurofibromatosis and severe upper thoracic kyphoscoliosis. **C** and **D,** A rib strut was placed anteriorly and a single rod was used posteriorly during fusion. **E** and **F,** Nearly 5 years later, frontal plane alignment remained satisfactory but the kyphosis was increasing above the instrumentation.

syndrome than in the general population. However, there is no significant difference in back pain between patients with scoliosis and those without.

No well-defined natural history studies of scoliosis in patients with Marfan's syndrome exist, although certain trends are evident. Curves identified in infancy progress dramatically.[425] These curves do not resemble the curves of infantile idiopathic scoliosis in that they are not expected to resolve spontaneously and most are right thoracic in configuration. In older but still skeletally immature patients, all curves more than 30 degrees will likely progress at least 10 degrees and will reach at least 40 degrees by maturity.

Unfortunately, brace treatment has not been shown to be effective in controlling scoliosis in Marfan's syndrome.[41,228,424,425,472] In infants, curves almost always progress to the point of needing operative intervention. Nevertheless, bracing curves less than 40 degrees in infants may be temporarily useful for postponing surgery. This is important be-

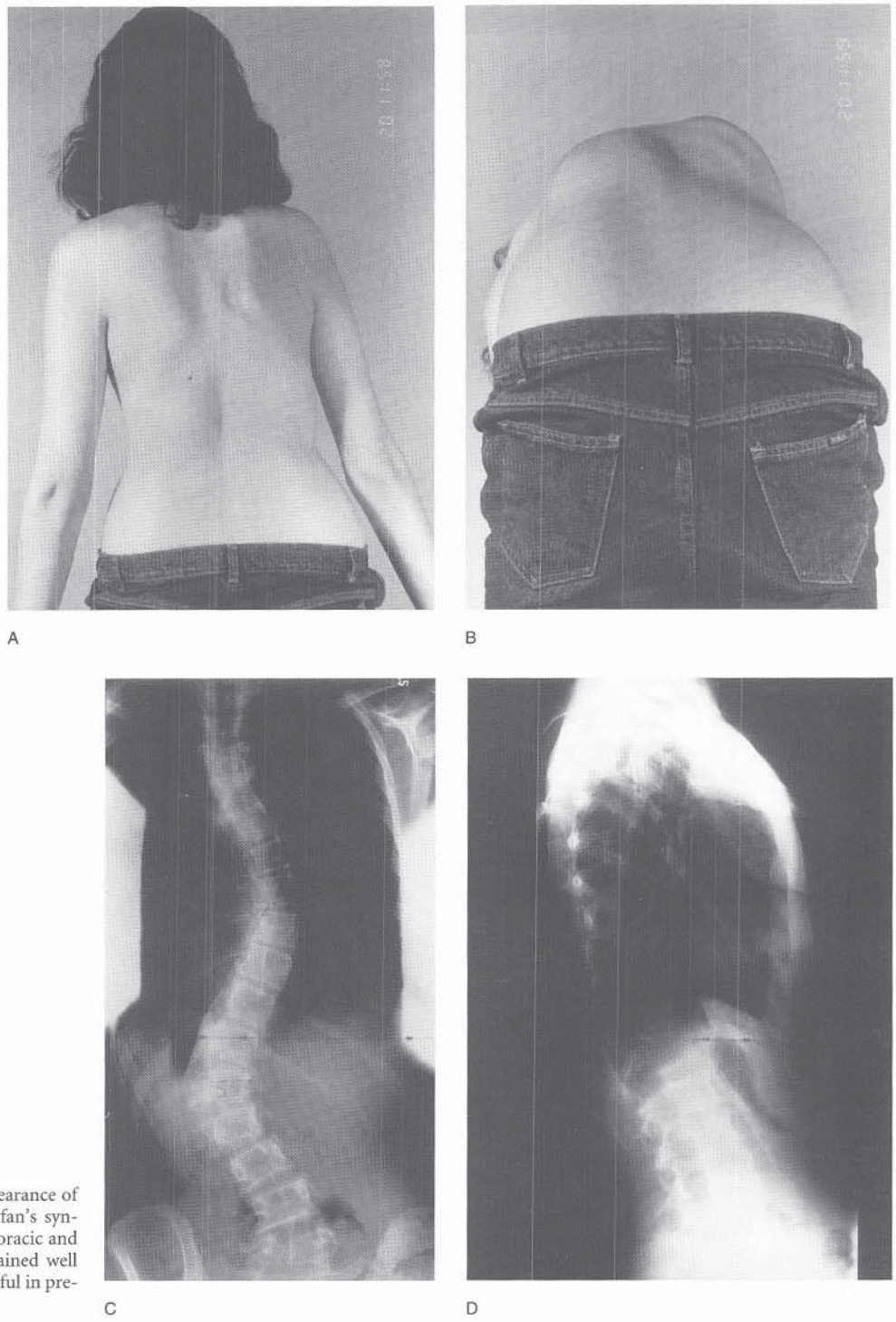


FIGURE 11–76 **A** and **B,** Clinical appearance of a girl age 12 years 2 months with Marfan's syndrome. **C** and **D,** Despite progressive thoracic and thoracolumbar scoliosis, the spine remained well balanced. Brace treatment was unsuccessful in preventing curve progression.

cause, in children with Marfan's syndrome and scoliosis, surgical intervention before age 4 years is associated with significant cardiac morbidity. Bracing in older children may also be of temporary benefit by allowing sufficient maturity to be gained, thereby necessitating only posterior surgery.

Surgery is indicated when scoliosis exceeds 45 degrees in adolescents or 50 degrees in adults. The procedure of choice remains posterior spinal fusion with segmental instrumentation.[202,228,472] Fusion levels are selected in the same way as for idiopathic scoliosis (Fig. 11–77). Careful attention to sagittal plane alignment is critical, since there can be significant variation from normal. Thoracolumbar or lumbar kyphosis may require anterior release prior to posterior instrumentation in order to achieve sufficient sagittal plane flexibility. Patients tend to have a higher incidence of pseudarthrosis, although its true incidence remains unknown.[228]

Instrumentation and fusion in patients who are identified during infancy usually produces modest correction, in the

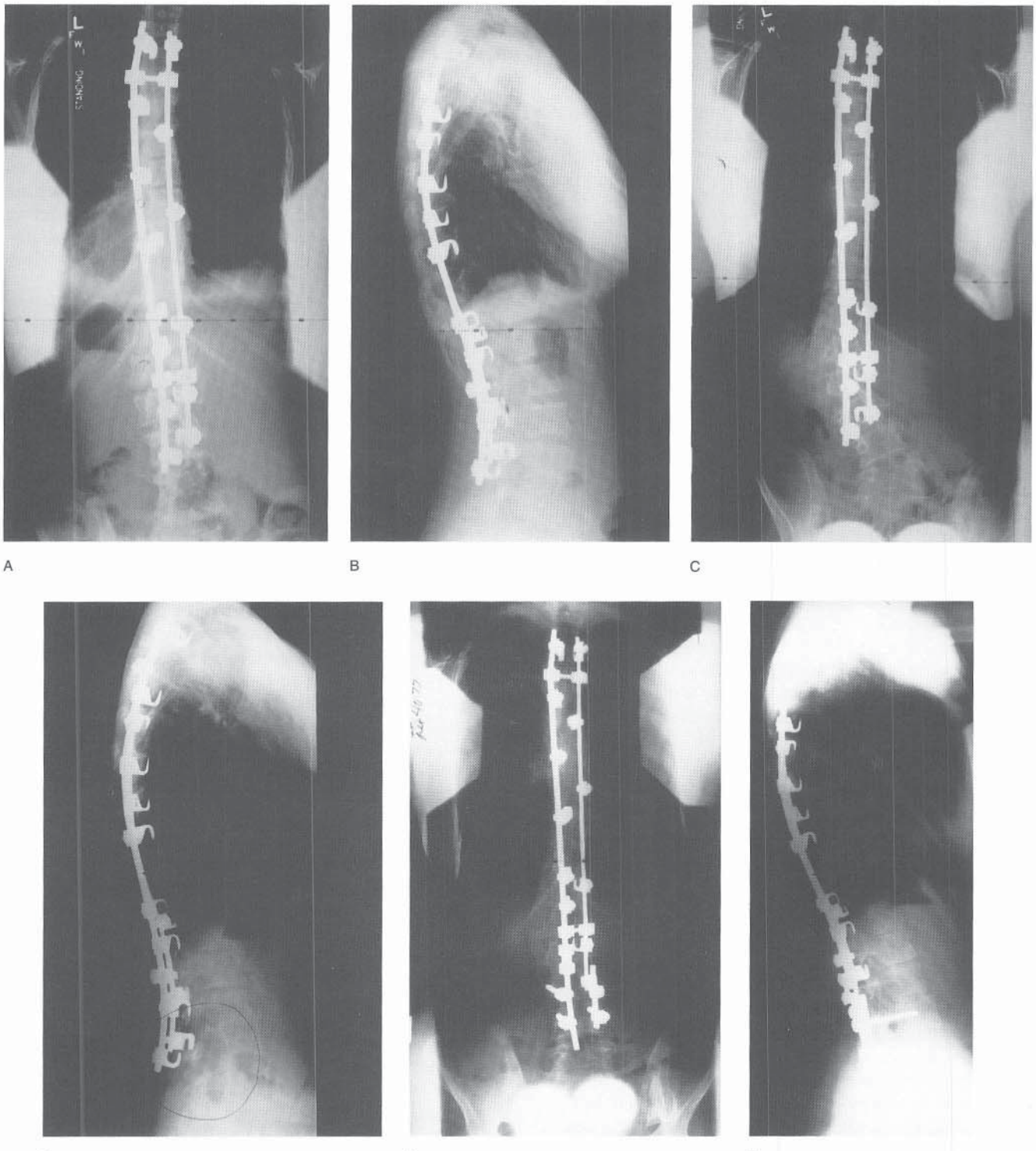


FIGURE 11–77 The patient whose clinical and radiographic findings are shown in Figure 11–76 underwent posterior instrumentation and fusion. Excellent correction was achieved **(A, B)**. Five months later, she noticed worsening balance in the lower spine. Radiographs showed that the inferior portion of the instrumentation had dislodged **(C, D)**. The patient was decompensated to the left and had lost correction of the lumbar curve. The lower segment was revised using pedicle screws, rod extensions, and rod cross-links. Three years later her balance remained improved and she was asymptomatic **(E, F)**.

range of 20 percent.[425] Surgery should be delayed until the child is older than 5 years of age. It should not be performed earlier because many patients with large curves before this age will succumb spontaneously to cardiac complications. If possible, anterior fusion should be added to prevent the development of the crankshaft phenomenon or to address excessive kyphosis.

Relative contraindications to performing corrective surgery for spinal deformity include cardiac insufficiency and a dissecting aortic aneurysm. If present, these conditions should be addressed before orthopaedic intervention is undertaken.

CONGENITAL HEART DISEASE

The association of scoliosis with congenital heart disease is well established.* Owing to advances in heart surgery for this condition, children are now living longer than in the past. Many with severe scoliosis are candidates for operative correction.

The incidence of scoliosis associated with congenital heart disease has been reported to be approximately 4 percent.[407] For those with congenital heart disease who have undergone cardiac surgery, the incidence of scoliosis is higher (11 percent).[210] This observation has led some authors to conclude that there may be an association between thoracotomies for congenital heart disease and the development of scoliosis.[210,450] Others refute this conclusion.[363] Although surgical intervention for heart disease in children may be associated with the development of scoliosis, it would appear that these two events have a multifactorial relationship.

Two types of scoliosis are seen in conjunction with congenital heart disease: congenital scoliosis and developmental scoliosis. For congenital scoliosis, the curve patterns and subsequent natural history appear to be unaffected by the coexisting congenital heart disease. Curve progression requires limited spinal fusion performed according to the standard guidelines described previously in the section entitled Congenital Scoliosis.

Children with developmental curves present at an average age of 11 years.[125,210] Left and right convexities occur with equal frequency. Usually, however, convex left thoracic curves are found in the upper thoracic spine and convex right curves are seen in the lower thoracic region.[210] There is no significant relationship between the age of the child at the time of cardiac surgery, age at the onset of scoliosis, and the severity of the scoliosis (Fig. 11–78).

Those with mild developmental curves (less than 30 degrees) require only observation. In children with developmental curves exceeding 30 degrees, the curves may progress as much as 9 degrees per year, tend not to respond to bracing, and are likely to require posterior spinal fusion. Before spinal surgery is initiated, repair of cardiac anomalies or temporary cardiac shunting procedures should be completed. Intraoperative management by experienced cardiopulmonary anesthesiologists and postoperative intensive care support are requisite for orthopaedic surgical intervention.

*See references 30, 33, 125, 201, 210, 266, 329, 351, 367, 386, 407.

THORACOGENIC

Many patients who develop scoliosis following thoracotomy have congenital heart disease.[450] Other conditions requiring thoracotomy (e.g., repair of tracheoesophageal fistula) can also lead to scoliosis.[464,492] Most, but not all, curves have the convexity toward the operated side. On occasion, two ribs fuse together at the thoracotomy site and function as a tether. In this instance, the concavity of the scoliosis is toward the operated side.

Younger patients who have a larger number of ribs resected or who have undergone multiple thoracotomies are at a higher risk for developing scoliosis.[96,113] Usually resection of the posterior portion of the ribs leads to the deformity. Anterior resection of the ribs does not tend to produce significant scoliosis.

Brace treatment for larger curves is usually ineffective, possibly because of the inability to apply corrective forces to the abnormal chest wall. Operative intervention with posterior spinal instrumentation usually results in a successful outcome.

POSTLAMINECTOMY

Spinal deformity following a one-level laminectomy is uncommon.[351] Usually the deformities encountered in children result from multilevel laminectomies performed for intraspinal tumors or trauma.[63,337,339] The age of the patient and the anatomic level of the laminectomy are important factors. Laminectomies performed in the cervical or thoracic spine commonly lead to progressive kyphosis. When laminectomies are performed in the lumbar spine, excessive lordosis may result. In addition to these sagittal plane deformities, scoliosis and rotatory deformities may also occur. Many of the conditions for which laminectomies are performed (e.g., trauma, neurofibromatosis, syringomyelia) can, by themselves, also produce a spinal deformity.

Over 50 percent of children undergoing multilevel laminectomies in the cervical region will develop spinal deformities.[15,32] Most of these deformities are kyphotic and usually span short segments. Some of the deformities are more gradual and involve more vertebrae (Fig. 11–79). "Swan-neck" lordotic deformities also can occur and are thought to represent compensatory mechanisms to maintain alignment of the head over the thorax.

In the thoracic spine, kyphosis may occur as short, sharply angled deformities or may have a more gradual angle and extend over several vertebrae (Fig. 11–80). These sagittal plane abnormalities result from the destabilizing effect of laminectomies, during which the spinous processes, interspinous and supraspinous ligaments, laminae, and ligamentum flavum are all removed. Preserving facet joints is important, as these joints contribute significantly to the stability of the spine.[313,351]

Several other factors may contribute to postlaminectomy kyphotic spinal deformities. Vertebral body defects caused by trauma or tumor (e.g., eosinophilic granuloma) lessen resistance to compressive flexion forces. Radiation therapy is often required in patients who have undergone laminectomies for spinal cord tumors. Radiotherapy damages the growth plates, adding to the deformity.

Scoliosis occurs less frequently than sagittal plane defor-

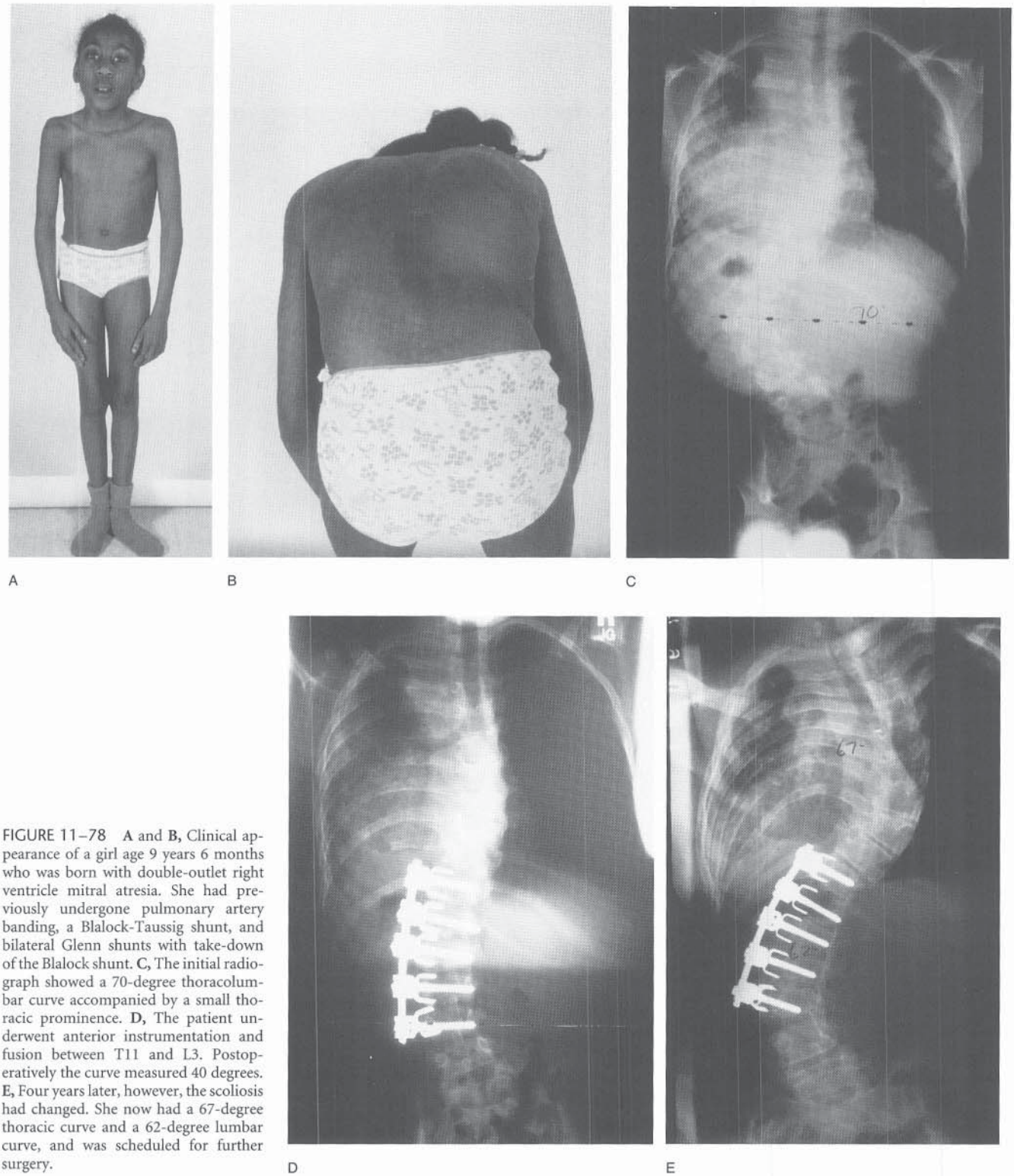
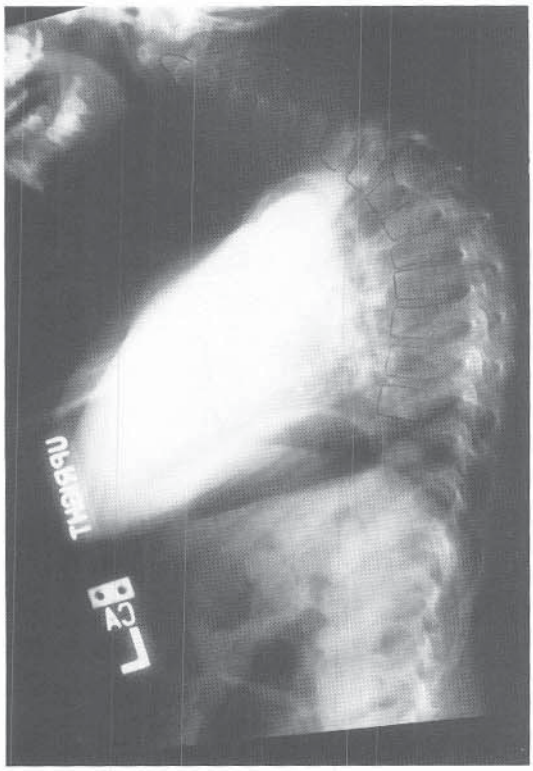


FIGURE 11–78 **A** and **B,** Clinical appearance of a girl age 9 years 6 months who was born with double-outlet right ventricle mitral atresia. She had previously undergone pulmonary artery banding, a Blalock-Taussig shunt, and bilateral Glenn shunts with take-down of the Blalock shunt. **C,** The initial radiograph showed a 70-degree thoracolumbar curve accompanied by a small thoracic prominence. **D,** The patient underwent anterior instrumentation and fusion between T11 and L3. Postoperatively the curve measured 40 degrees. **E,** Four years later, however, the scoliosis had changed. She now had a 67-degree thoracic curve and a 62-degree lumbar curve, and was scheduled for further surgery.

mity. It, too, usually involves the area of the laminectomies. Less commonly, collapsing scoliosis develops below this area. In such an instance, it is usually related to a neuromuscular deficit that resulted from a spinal cord tumor or its treatment.

In the lumbar spine, an extensive laminectomy may result in lumbar hyperlordosis.[256] Rhizotomy, which has recently become popular in the treatment of cerebral palsy, requires lumbar laminectomies to expose the nerve rootlets. Although rhizotomy has been reported not to result in increased changes in sagittal plane deformities,[350] at the author's institution several examples of relentlessly progressive postoperative hyperlordotic deformities have been encountered.



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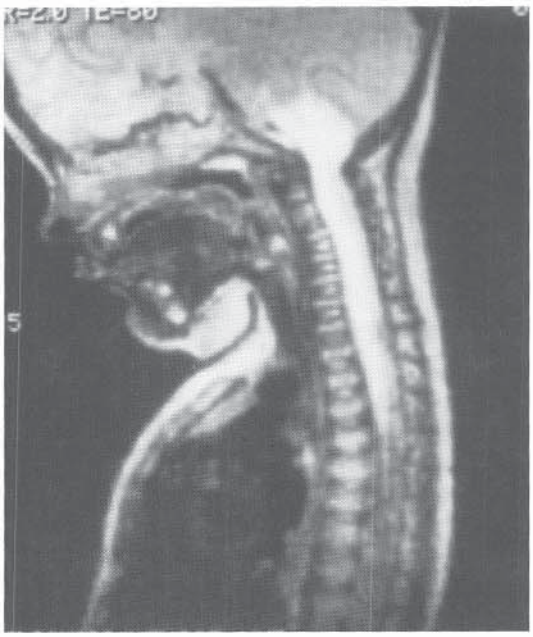

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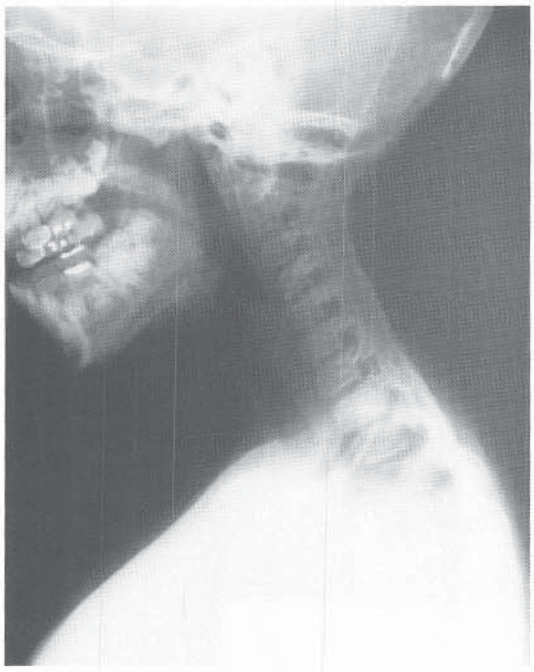

C

FIGURE 11–79 Radiograph appearance of a girl age 5 years 10 months who presented with excessive cervicothoracic kyphosis secondary to multiple laminectomies **(A).** At age 10 months a small cell neural ectodermal tumor of the cervical spinal cord had been diagnosed **(B).** Craniotomy and laminectomies down to the T5 were performed, followed by chemotherapy and radiation therapy. At the time of presentation (age 5 years 8 months), she underwent posterolateral fusion performed using autogenous iliac crest bone as the graft material, followed by immobilization in a halo vest. Three years later the fusion remained solid, with excellent sagittal plane alignment **(C).**

Treatment for an established postlaminectomy spinal deformity generally requires further stabilization surgery. Therefore, all efforts should be directed toward preventing this deformity. Laminoplasties in the thoracolumbar spine at the time of initial surgery have been helpful in preventing the occurrence of kyphosis.[299,406] Facetectomies should be avoided if possible. Preoperative discussions between neurosurgeons and orthopaedic surgeons prior to tumor resections should be undertaken to determine whether immediate operative fusion would be of benefit.

Preventive bracing of the spine following laminectomies has not proved effective in stopping subsequent deformities. Therefore, if a brace is utilized, it should be with the understanding that it may only temporarily be of benefit. Once

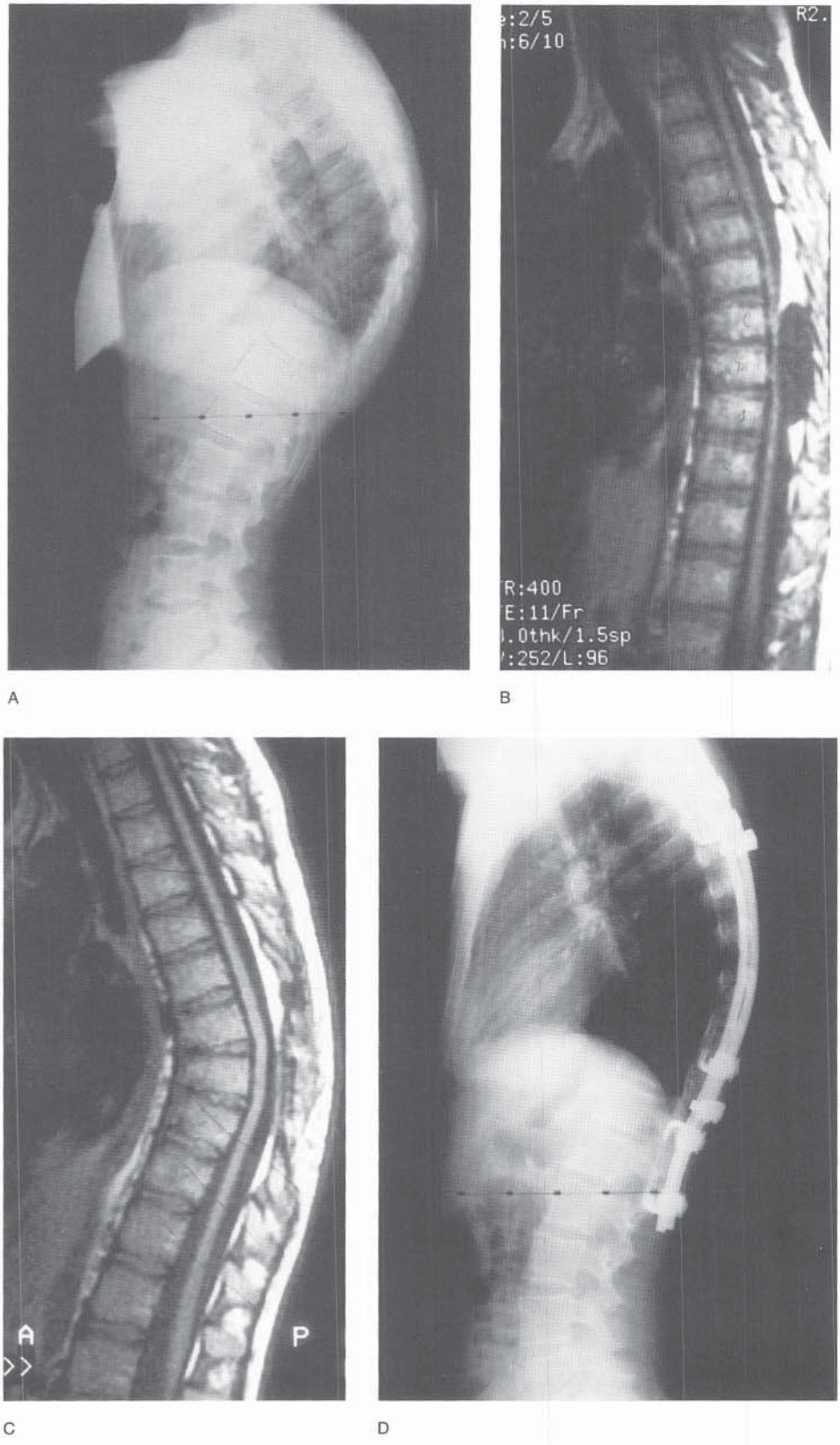

FIGURE 11–80 A 13-year-old boy presented with an increasing kyphosis in the thoracic spine. It measured 62 degrees between T5 and T10 (**A**). One year previously he had undergone resection of an arachnoid cyst (**B**) through laminectomies over three levels. At the time of presentation, a recent MRI showed no presence of the cyst but did demonstrate progressive localized kyphosis (**C**). Anterior and posterior fusion was performed using posterior titanium instrumentation. The improvement seen in the sagittal plane at 6 months (**D**) has been maintained over time.

notable kyphosis has been identified in the cervical or thoracic spine, surgical fusion is needed to correct and stabilize the deformity. In the thoracic spine, if the deformity is mild, posterior fusion with instrumentation by itself may be sufficient. If the deformity is more severe, or if there is short, sharply angled kyphotic deformity, then anterior fusion (with a rib-strut graft, as needed) should precede the posterior spinal instrumentation and fusion.[337] In the cervical spine, anterior fusion is often performed in combination with posterior fusion.[15,32]

IRRADIATION

For some spinal cord tumors, radiation therapy may be the primary form of treatment. In the growing child, doses to the vertebral column exceeding 1,000 rads may have an inhibitory effect on the physeal regions.[318] As a result, asymmetric growth may develop, leading to a scoliotic or kyphotic deformity. Spinal deformity may not always be caused by a distortion of the vertebral growth but may also result from soft tissue fibrosis and contractures.

Very young children who receive radiation (often for Wilms' tumor or neuroblastoma) are at greatest risk for developing spinal deformities.[274,281,377,378] Long-term follow-up of these individuals is necessary, as deformities can worsen notably during the adolescent growth spurts. Every effort should be made to exclude the spine or pelvis from the radiation field in young children.

For treatment, bracing has not proved effective in arresting progression of irradiation-induced spinal deformity. However, it continues to be used in patients with scoliosis exceeding 20 degrees in an effort to delay progression of the deformity. Should scoliosis exceed 40 to 45 degrees, operative intervention should be undertaken. Healing may be prolonged, and consideration should be given to repeat bone grafting 6 months postoperatively. The risk of postoperative complications, such as pseudarthrosis and infection, is increased.

For postirradiation kyphosis, anterior fusion is needed along with posterior fusion in an effort to avoid the likelihood of pseudarthrosis.[332] For sharply angled kyphotic deformities, a vascularized rib-strut graft is recommended. Postoperative bracing for 6 to 12 months should be considered in these patients.

HYSTERICAL SCOLIOSIS

Hysterical scoliosis is a diagnosis of exclusion.[332,435] The curvature generally has a long C-shaped appearance, with trunk imbalance, lack of abnormal neurologic or other physical findings, and no radiographic evidence of vertebral rotation. There may be a change in the pattern or the severity of the scoliosis from day to day. The curve generally resolves when the individual is supine.

A thorough neurologic evaluation is necessary to rule out rare unusual causes, such as spinal cord tumor. Bone scan and MRI may be necessary to confirm this. Laboratory studies (complete blood cell count with differential and sedimentation rate) will rule out infection. Once organic causes have been ruled out, the treatment for hysterical scoliosis requires psychological (or psychiatric) therapy. Orthotic management should not be undertaken, as it may reinforce the underlying personality disorder.

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