

Congenital Scoliosis

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INTRODUCTION

Congenital scoliosis is defined as a lateral deviation of the spine associated with one of a broad range of congenital vertebral malformations (CVMs) that can form during *in utero* development. It is distinct from other spinal deviations in which malformations do not occur and can present as an isolated spine anomaly or be associated with a large number of visceral organ and syndromic abnormalities. In Smith's "Recognizable Patterns of Human Malformation," over 40 syndromes have CVMs listed as one of the presenting features (1).

The malformations are always present at birth, but the development of the scoliosis may occur over time. They can occur in any part of the spinal column and are believed to be a result of the disruption of the process of somatogenesis that occurs between the 5th and 8th weeks of gestation (2–8). These malformations are not unique to humans as several other species develop such abnormalities in response to teratogenic exposures or from genetic predispositions (9–14).

Of great importance is the high incidence of associated abnormalities in other organ systems that can lead to significant adverse impacts upon the health and well-being of affected individuals. In addition to this is the realization that management of the developing deformity is often extremely challenging and, due to the variation of presentation, highly individualized. This chapter focuses upon children with congenital vertebral anomalies. We do not discuss infantile scoliosis or other conditions where there may be deformity without malformations.

INCIDENCE/PREVALENCE

The true incidence and prevalence of congenial vertebral malformations is unknown. Many individuals without visible deformity are undoubtedly not found, and many series intermix patients with syndromic or other neuromuscular conditions. Large population studies utilizing screening chest x-rays for tuberculosis suggested a thoracic spine incidence of 0.5 to 1:1,000 live births (15), with more recent ultrasound screening studies finding an incidence of 0.1 to 0.3:1,000 live births with many of these children having other complex birth defects (16, 17). In general, the incidence should be considered to be rare, with kyphotic CVMs even rarer.

MECHANISM OF SPINE FORMATION

It is useful to review the evolving knowledge and understanding of spine formation and development as it may relate to the formation of CVMs. Much of this knowledge has been gained from the observation of human embryonic development (18) and study of murine models of spine formation. During gastrulation, four identified *Hox* gene clusters containing 39 genes (*Hox A, B, C, and D*) are believed to determine positional information along the rostrocaudal axis of developing vertebrates (11, 18, 19). They are expressed in cells of the developing mesoderm and ectoderm that later form the somites, which in turn form the vertebrae, ribs, and muscles. Vertebrae are derived from the paraxial mesoderm that forms from the superficial epiblast cells growing into the primitive streak during gastrulation forming the paraxial mesoderm. These develop into the segmental units of the somite, which then subdivide into ventral sclerotome (vertebral precursor) and dorsal dermatomyotome (muscle, skin, rib precursors) units (3, 18, 20). Induction of the sclerotome is signaled by

the notochord and the floor plate of the neural tube. This involves the Sonic Hedgehog protein (21). Subsequent fusion of the ventral and dorsal sclerotomes then forms vertebrae (3, 18, 20). It is believed that a molecular segmentation clock that is mediated through the Notch signaling pathways

controls vertebrate segmentation (3, 10, 12, 22) with formation of somites in a rostral to caudal sequence. This involves periodic expression of 50 to 100 cyclic genes. Genes that are believed to impact this pathway are lunatic fringe (*LFNG*), *DLL3*, and *MESP2* (23, 24) (Fig. 18-1).

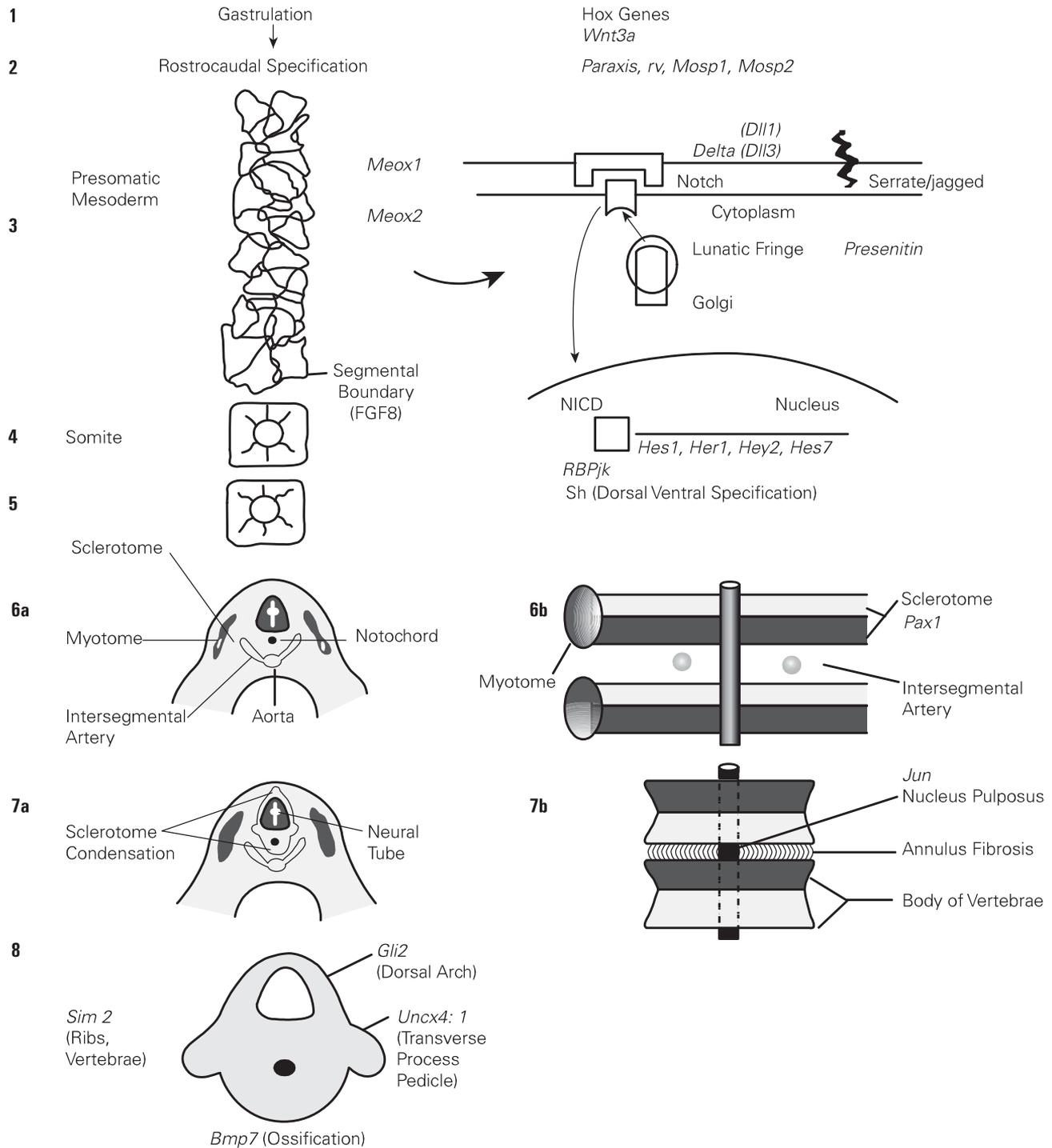


FIGURE 18-1. Schematic diagram showing pathway of murine embryologic development of somites with potential gene interactions highlighted. Segmentation is operated through a molecular clock operating through the Notch signaling pathway. Segment boundary formation is mediated by fibroblast growth factor 8. Sclerotome formation and subdivision into dorsal and ventral halves is mediated by *Pax 1* and *Meox1*.

ETIOLOGY

The true etiology of the development of CVMs is unknown. The spectrum of the disorder is very large, ranging from syndromic cases such as Jarcho-Levin syndrome to isolated hemivertebrae, and a number of environmental and genetic associations have been identified, suggesting that both genetic factors and teratogenic effects from the environment play a role in the disturbances of normal spine formation.

Anoxia created by carbon monoxide exposure to mouse and chick models at a critical time and specific dose has been demonstrated to cause CVMs very similar to those found in humans (9, 25–27). Similar outcomes in humans due to exposure to carbon monoxide have been postulated, but not clearly established (28). Other environmental factors have been clinically associated with the development of CVM. Alcohol exposure with fetal alcohol syndrome (29), anticonvulsant medications Valproic acid and Dilantin (30–33), retinoic acid (31), hyperthermia (34), maternal insulin-dependent diabetes (35, 36), and folate deficiency (37) have all been implicated in abnormalities of the spine in humans.

Model organisms such as the chick and mouse strongly suggest that there is a genetic contribution to the development of CVM (3, 11, 12, 38). In humans, the heterogeneous clinical manifestations, variety of morphologic features, and phenotypic presentations described as failures of formation or failures of segmentation had led to the belief that the majority of cases were sporadic events (39). Recent evidence suggests that there is a strong role for genetic factors in the development of human CVMs. The genetic transmission of some cases of CVM has been shown to have a familial recurrence rate of 3% (20, 40), and an increasing number of vertebral malformation syndromes have identifiable genetic etiologies. Spondylocostal dysostosis patients have a loss of normal vertebral morphology throughout the entire spine and have been shown to have mutations in *delta-like 1* (*DLLS*), *mesoderm posterior 2* (*MESP2*), and *LFNG*. Mutations in these affect the Notch signaling pathway and highlight the importance of Notch in vertebral column formation in humans (3, 41, 42). Spondylothoracic dysostosis (commonly referred to as Jarcho-Levin syndrome) is a confusing array of CVMs, all of which have rib anomalies with fusion of all ribs. A homozygous recessive nonsense mutation in *MESP2* has been identified. Alagille syndrome is an autosomal dominant condition with bile duct paucity, cardiac, eye, kidney, pancreas, and vertebral anomalies in 22% to 87% of affected individuals. Mutations in *JAG1* and *NOTCH2* have been found in these patients (2, 3). Klippel-Feil syndrome was first reported in 1894 (43) and described in 1912 (44). Most cases have been found to be sporadic in families, but autosomal dominant, recessive, and X-linked forms have been reported. There has been recent evidence that *PAX1* mutation and notch pathway mutations can be found in patients with Klippel-Feil syndrome (8, 45–47). As isolated CVM are often sporadic occurrences with a given family, candidate gene analysis has been used. Using mouse–human synteny analysis, 27 eligible loci have been identified, of which 21 cause

vertebral malformations in the mouse. Six of these—*PAX1*, *WNT3A*, *DLL3*, *SLC35A3*, *T(Brachyury)*, and *TBX6*—have been studied in some detail and show promise as potential loci for the formation of CVMs in humans (3, 13, 14, 46–48).

ASSOCIATED CONDITIONS

CVMs do not occur in isolation. As they develop during a critical stage of organogenesis, as many as 61% have other associated malformations (49), and the development of progressive deformity may lead to secondary organ involvement. The malformations seen are in general neurologic and visceral, with most tied to the level of the CVM.

Neurologic abnormalities have been described in 18% to 38% of patients with CVM (50–54). A prospective evaluation of consecutive patients with CVM found that 38% had an intraspinal anomaly, with higher concentrations among patients with cervical and thoracic CVM, mixed patterns of segmentation and formation, and congenital kyphosis (50). Half of these patients had physical findings to suggest that they had an intraspinal lesion. Diastematomyelia was the most common finding followed by intraspinal lipoma, tethered cord, Chiari malformation with or without associated syringomyelia, dermoid cyst, and epidermoid cyst (Figs. 18-2 to 18-4). For patients with an isolated hemivertebrae, there may also be a high incidence of intraspinal anomalies. Belmont found that of their 76 patients with C3VM, 29 had an isolated hemivertebrae, and of these, 8 (28%) had an MRI abnormality (55). Thirteen had abnormal clinical findings. A review of these patients and published series of similar patients suggests that an abnormal finding on the history of physical examination had an accuracy of 65%, sensitivity of 59%, specificity of 87%, a negative predictive value of 72%, and a positive predictive value of 74% (45, 50, 51, 56, 57).

For patients with known congenital cardiac abnormalities, the incidence of scoliosis has been found to be as high as 10% (58), but most of these are normally segmented (26, 58). Farley found that only 11/48 children (23%) with congenital heart disease (CHD) who had scoliosis had congenital scoliosis. The converse relationship of a higher incidence of CHD (baseline incidence 0.5% in the general population) in patients primarily found to have a congenital scoliosis has not been demonstrated, but it is believed that there is a higher incidence. Basu et al. (50) found cardiac abnormalities in 26% of patients with CVM. Two-thirds of these had knowledge of the cardiac abnormality before they presented for evaluation of their spine. Of those who did not have prior knowledge, of the cardiac defect, 2/10 (20%) did proceed to active management.

The incidence of renal anomalies in CVM has been reported as 26% to 37% (49, 50, 54, 59–61). The most common anomalies have been unilateral renal agenesis, urinary duplication, ectopic kidney, reflux, and a horseshoe kidney. The incidence of renal anomalies may be higher if there is an associated rib anomaly in addition to the CVM (52). In most of the series, the renal anomalies were unsuspected, leading to general recommendations to screen children for these problems.



FIGURE 18-2. **A:** Example of diastematomyelia—plain radiographs may reveal a central canal bony process as the child matures. **B:** Plain radiographs. **C:** MRI findings.

Morbidity associated with large progressive increases in deformity has largely been related to the development of restrictive lung disease and is believed to be a function of both altered alveolar multiplication and altered dynamics of breathing due to constriction of the thorax (62–72). As compared to idiopathic scoliosis, the loss in vital capacity for a given Cobb angle is believed to be 15% greater for individuals with CVM (73). Several series have shown a progressive loss in vital capacity for progressive curves but have shown that if there is no progression and no associated severe rib anomalies, the vital capacity average for individuals with CVM is 87% of predicted (67, 69,

73–75). Similarly, diffusion capacity for nonprogressive CVM has been found to be normal, suggesting that congenital scoliosis without fused ribs may not be associated with pulmonary hypoplasia or thoracic insufficiency syndrome (TIS) (72). Overall, however, the greater the deformity and the more proximal the deformity, the greater the apparent long-term impact upon spirometry (62, 63, 66, 69). For severe progressive curves, body mass index (BMI) less than the 5th percentile can be common, and this may create a higher risk situation for the operative management of these children. One of the primary clinical correlates to measurable declines in functional vital capacity



FIGURE 18-3. Example of syrinx with Chiari I malformation.

(FVC) in CVM is an altered BMI (74). Another is increased asymmetry of ventilation/perfusion (V/Q), which occurs in about 50% of children with TIS and congenital scoliosis (75).



FIGURE 18-4. Example of a tethered spinal cord with an intraspinal lipoma.

Coupled with this is the observation that diffusion capacity does decline with worsening deformity presumably due to a reduction in the total alveolar surface area (72). Cobb angle has not been correlated to either FVC changes or to altered function as measured by V/Q scans (68, 71). Surgical interventions aimed at stabilizing the chest/spine in patients with TIS have been shown to preserve growth-related changes in vital capacity over time but do not appear to be able to catch up for lost function (68, 70).

CLASSIFICATION/NATURAL HISTORY

Early reports of CVM suggested a benign natural history (76), but subsequent reports by Winter and Moe (77) and MacEwen et al. (78) identified severe deformities from high-risk patterns of deformity. Subsequent classifications for CVM have been several (77, 79–83). Most have focused upon trying to assign a risk of progression by study of the natural history of deformities presenting to surgeons. Despite new knowledge gained by three-dimensional (3D) imaging using computed tomographic (CT) scans (79) and information about genetic mechanisms by which CVMs may form, the classification schemes proposed by Winter and Moe (77) and Nasca et al. (82) and modified by McMaster and Ohtsuka (80) remains the most useful and predictive (Fig. 18-5). The system defines malformations as either failures of formation, failures of segmentation, or combinations of these. The malformations can occur in frontal, sagittal, or both planes. The likelihood of progression is predicted by the anticipated potential for either unbalanced growth created by uneven growth potential due to failures of formation (hemivertebrae, wedge vertebrae) or growth retardation due to tethering created by failures of segmentation (bar). Approximately 10% of patients have patterns of CVM that are not definable by this classification scheme. The highest risk of progression was shown by McMaster to be in situations where there is unbalanced growth opposite an asymmetric tether as in a unilateral unsegmented bar with a contralateral hemivertebrae. The average annual rate of progression was found to be from 5 degrees to >10 degrees per year. The next highest risk of progression was in those with a unilateral unsegmented bar followed by double hemivertebrae (80) (Fig. 18-6). Reports of untreated patients followed in surgical clinics document a risk of progression to curves >40 degrees at maturity from 37% to 84% (76, 77, 80). For any given deformity type, there is variability in progression that may be due to the fact that it is difficult to account for variation in anterior and posterior CVMs at any given level. The most rapid times of progression were in the first 5 years of life and from ages 10 to 14 years. If there were clinically evident deformities in the first year of life, a worse overall prognosis was found (84, 85). The risk of progression has been predicated upon the belief that there is no growth potential of involved segments with CVM, leading to deformity and a shortened trunk (85). Campbell and Hell-Vocke (86) have questioned this with evidence of growth in CVM segments used forced growth techniques.

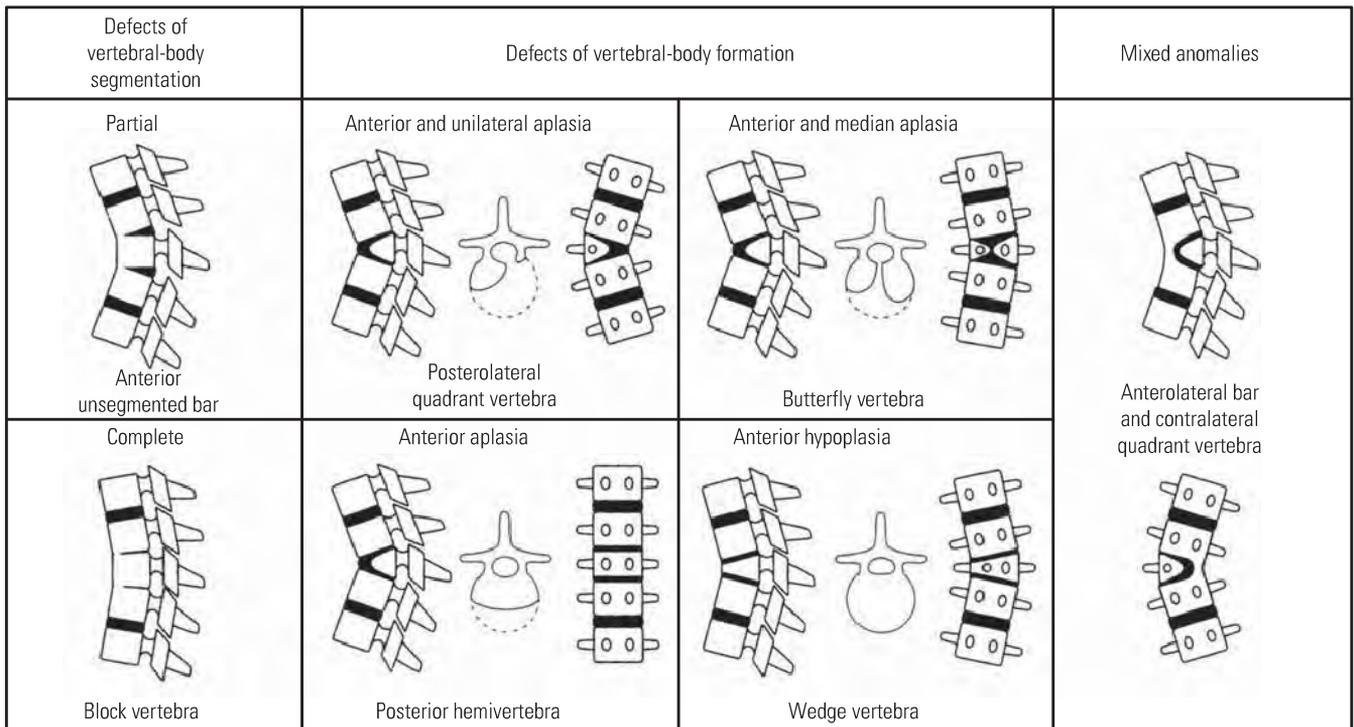
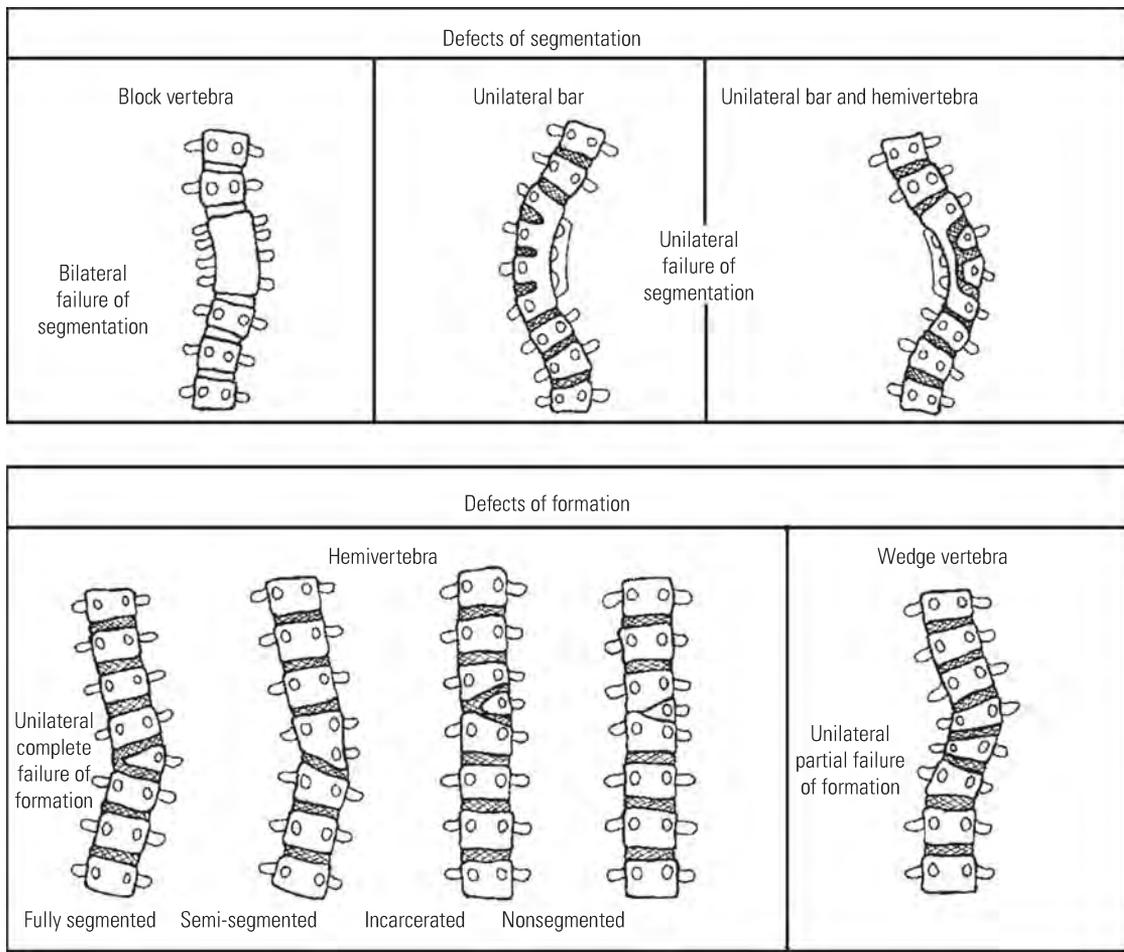


FIGURE 18-5. **A:** Classification of congenital scoliosis showing failures of formation and segmentation. Incarcerated hemivertebrae have compensatory deformities proximal and distal to it. The highest risk for progression lies with a hemivertebrae opposite a unilateral bar. **B:** Classification of congenital kyphosis. A high percentage of true congenital kyphosis deformities will progress and can lead to paraplegia especially if in the thoracic spine.

Site of curvature	Type of congenital anomaly					
	Block vertebra	Wedge vertebra	Hemivertebra		Unilateral unsegmented bar	Unilateral unsegmented bar and contralateral hemivertebrae
			Single	Double		
Upper thoracic	<1°–1°	★–2°	1°–2°	2°–2.5°	2°–4°	5°–6°
Lower thoracic	<1°–1°	2°–3°	2°–2.5°	2°–3°	5°–6.5°	6°–7°
Thoracolumbar	<1°–1°	1.5°–2°	2°–3.5°	5°–★	6°–9°	>10°–★
Lumbar	<1°–★	<1°–★	<1°–1°	★	>5°–★	★
Lumbosacral	★	★	<1°–1.5°	★	★	★

No treatment required
 May require spinal surgery
 Require spinal fusion
★ Too few or no curves

Ranges represent the degree of derotation before and after 10 years of age

FIGURE 18-6. Risk of progression of types of CVM as compiled by McMaster. Highest rates of progression were in unilateral unsegmented bar and contralateral hemivertebrae.

The clinical impact of progressive deformities associated with CVM has focused historically on respiratory consequences and more recently upon quality-of-life measures. There is no clear association that has been established between classifications of CVM and mortality or morbidity. Patients with severe progressive deformities have been demonstrated to have severe restrictive lung disease and significant morbidity with early death (62–64, 85, 86). They have also been found to have a significantly diminished quality of life as measured by the Child Health Questionnaire in the domains of physical limitations and caregiver burdens, but not in psychosocial domains (87). Surgical intervention for these deformities has not yet been shown to alter these natural histories. It appears that pulmonary function decline can be stabilized with surgical treatment in some cases, but improvement in depressed vital capacity has not been observed (68, 70). Similarly, diminished quality of life does not appear to be improved after intervention (88).

EVALUATION OF PATIENTS WITH CVM

The wide range of presentations and the high incidence of visceral anomalies that can be present in patients with CVMs mandate that all evaluations include a careful history and physical examination. Presentations will range from severely involved children with obvious visceral and structural anomalies to those with nonprogressive and asymptomatic malformations.

Neurologic. Many relatively uninvolved children will be referred due to incidental findings on imaging studies obtained for other reasons. Growth charts should be reviewed as children with CVMs may have altered growth velocities and can present with disproportionate growth retardation. Delays in developmental milestones such as walking and running and potty training can be important signs of an underlying spinal dysraphism as can symptoms of back pain in very young children. Physical findings of posterior midline trunk hairy patches, large nevi, or hemangiomas, atrophy of extremities,

a cavus foot (Figs. 18-7 and 18-8), or any neurologic abnormalities (weakness, balance difficulties, sensory, hyperreflexia, asymmetric abdominal reflexes) can indicate an underlying spinal dysraphism in 50% to 80% of cases (19, 50, 51). It is widely recommended that an MRI of the neural axis be obtained prior to surgical intervention for CVM (45, 50–53, 56, 78, 89–91). Less clear is whether there is benefit to the routine ordering of spine MRI scans for asymptomatic children with CVM who are not to undergo surgery. The positive and negative predictive value of physical examination and clinical history is 74% and 72% respectively for determination of MRI abnormalities (45, 50). As many younger children will require an anesthetic for a spine MRI, in asymptomatic nonprogressive deformities in children, observation to an older age is recommended. Intrauterine MRI has been tested for defining neural axis lesions in CVM. It has been found to be better than intrauterine ultrasound, but its overall accuracy and role in managing CVM are as yet unknown (92–95). For neonates with a suspected abnormality of the neural axis, ultrasound has been shown to be an effective screening tool and can be used



FIGURE 18-7. Cavus foot. High incidence of neurologic abnormalities associated with this finding.

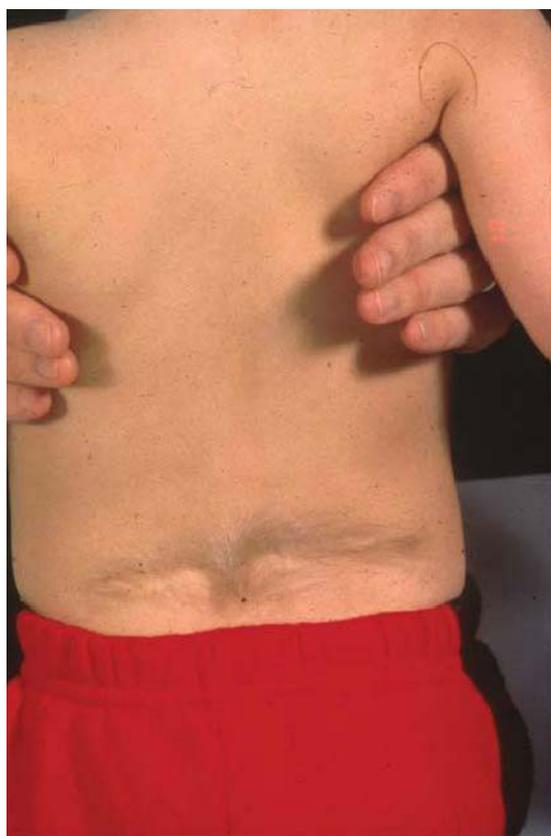


FIGURE 18-8. Midline hairy patch consistent with an underlying spinal dysraphism.

in a cost-effective manner depending upon the expertise of the individual performing it (8, 47, 54).

Renal. As up to one-third of patients may have an underlying renal anomaly and many will be asymptomatic, evaluation of the urinary system is generally advised (29, 49, 50, 52, 59, 69). Renal ultrasound has replaced contrast intravenous pyelogram as the imaging study of choice (60, 89). For patients who are undergoing MRI for evaluation of the neural axis, renal anomalies can be well-visualized when a modified spine MRI is ordered (96).

Cardiac. As many as two-thirds of patients with cardiac abnormalities and CVM will have had diagnosis made by their primary care provider or a cardiologist prior to presentation for evaluation of their CVM (50, 58). For those who have not had prior evaluation and who are asymptomatic, the majority of the cardiac anomalies that may be present will not require active treatment. Cardiac screening is recommended for patients who are to undergo surgical treatment (89).

Pulmonary. Children with progressive and severe deformities are at risk for developing significant restrictive lung disease (62, 69, 73, 75, 82). The linkage between the spine, thorax, and lung growth and function has been brought to light by Campbell et al. (64) through their definition of TIS as the

inability of the thorax to support normal lung growth and function. It is recommended that children who are to undergo surgical intervention for progressive disease have a pulmonary evaluation prior to surgery. There are currently no established guidelines related to respiratory function that guide decisions for surgical intervention.

The assessment of lung function in children is made difficult by their inability to cooperate with pulmonary function testing (97). For very young children, CT lung volumes have been offered as a surrogate (98–100), but correlation between measured changes in volume and change in respiratory function as measured by physiologic testing has not been established for these populations of normal children. Right lung CT volumes have been found to highly correlate with preoperative right lung function as measured by ventilation perfusion scans, but to have poor correlation after surgery for patients with TIS (101). This implies that alterations in volume alone do not predict impact upon respiratory function. Current concerns related to the risk of radiation exposure to children from CT scans should bring to light risk–benefit questions when ordering these tests (102–104). Recently, dynamic chest and diaphragm motion has been evaluated using ultrafast dynamic MRI in an effort to define how chest wall perturbations may affect respiratory efforts through their impact upon chest wall compliance and diaphragm motion. Correlation with spirometry data has been done in healthy, normal subjects, with the technique showing great promise for the assessment of individuals with pathologic conditions (105, 106).

Children with TIS will manifest with hypercarbia and hypoxemia in advanced stages of their disorder (62–65, 67, 74). Serologic evaluation with capillary blood gases looking for elevation of serum bicarbonate indicating CO_2 retention and of serum hemoglobin levels that would indicate hypoxemia is recommended at the time of initial evaluation (64, 65).

Radiographic. Biplanar radiographs have been the main mechanism for evaluation and guidance of decisions related to the surgical intervention for patients with CVM. For complex CVM, inter- and intraobserver reliability of the Cobb angle appears to be 10 degrees (107–109). Campbell et al. has developed the techniques of space available for the lung (SAL) and the interpedicular line ratio as alternatives for measuring deformity (64, 110). The reliability of these measures has not yet been established and correlation to respiratory changes has not been made. As pointed out earlier, the risk of curve progression is not always predicted by classification schemes utilizing biplanar radiographs. This coupled with the increasing use of pedicle screw fixation has prompted 3D evaluation of CVM prior to surgery using CT (111–113) and interest in developing classification systems based upon CT data (79) (Fig. 18-9). No consistent threshold value has been established at which intervention should be performed, but it has generally been recommended that documentable progression at a prepubertal age should be strongly considered for surgery

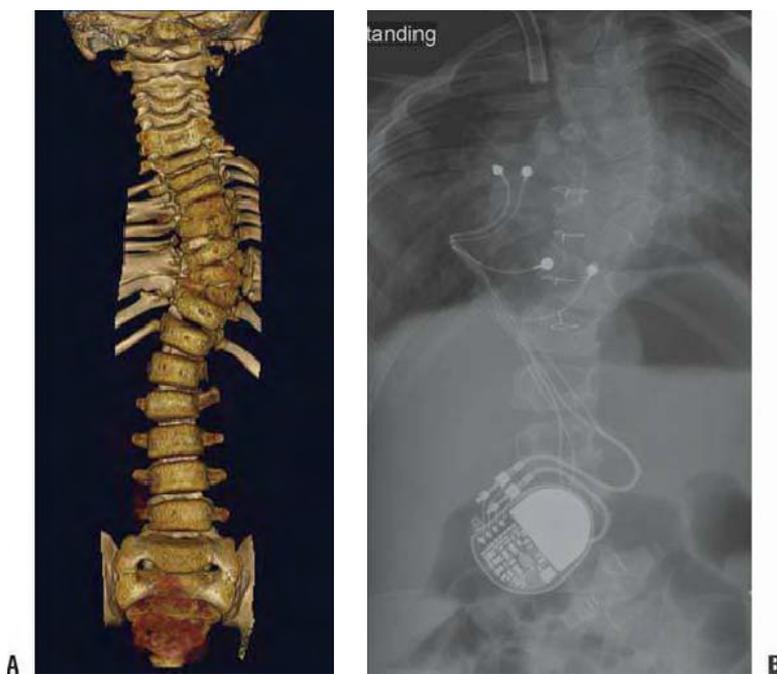


FIGURE 18-9. Three-dimensional CT scan (**A**) of a complex CVM (**B**). Note the mix of posterior and anterior defects.

(38, 77, 90, 114). As patients with CVM often have deformity at multiple levels, it is recommended that all patients have radiographic evaluation of their entire spine upon presentation, with special attention given to cervical spine abnormalities prior to surgery (89).

SUMMARY

Patients who present with CVM have identifiable defects in segmentation during early fetal development. They will present with a wide range of phenotypic expressions and have varying degrees of risk for progression and morbidity. Careful evaluation of these patients for associated visceral and neural axis abnormalities is advised upon initial presentation and before surgical intervention. Consistent criteria for surgical intervention have not been identified, but in general rapid progression of deformity at a prepubertal age carries a poor prognosis and surgical intervention is recommended. Three-dimensional imaging is rapidly becoming a standard in preoperative evaluation and planning. The remainder of this chapter focuses on the evolving techniques for intervention.

TREATMENT

Nonoperative Treatment

Observation. The child with a congenital vertebral anomaly requires close observation during growth until maturity. The anomaly, although present at birth, may not manifest itself as a clinically visible deformity until many years later,

after sufficient asymmetric growth has resulted in scoliosis. Hence, the classification of the deformity and the predicted natural history is crucial to determining the prescribed treatment. The patient can be monitored with radiographs at 6- to 12-month intervals depending on the type of malformation, location of the anomaly(ies), prior curve behavior, and the age of the child relative to spinal growth. Changes in the congenital deformity can be rapid in infancy and again in the preadolescent growth spurt, but relatively quiescent in middle childhood (5 to 10 years of age) (115). The location of the CVM is also important. Progressive cervical CVMs produce significant shoulder asymmetry and neck deformities (Fig. 18-10), and progressive limbosacral CVMs can produce a truncal shift or pelvic obliquity and coronal imbalance (Fig. 18-11). Some thought should be given to avoidance of radiographs at frequent, routine intervals just as a reflex reaction due to the cumulative nature of radiation to a young person over her lifetime (116). It can be very helpful to obtain the initial radiographs of a child, for example, a chest x-ray in the newborn nursery, in order to identify hemivertebrae, congenital bars, or fused ribs and then compare to films that are more contemporary in order to gain an understanding of the sometimes-complex pathology (Fig. 18-12). In a child <2 years, supine films are adequate, but after standing age, erect, full-length radiographs should be obtained in the posterior–anterior projection and lateral, when necessary. Clinicians should be aware that positional changes when x-rays are obtained during this development phase (sitting to standing) may drastically change the measured Cobb angle.

This archive of films should be carefully evaluated for changes not only in the congenital area and its associated curve but also in the compensatory (noncongenital) areas,

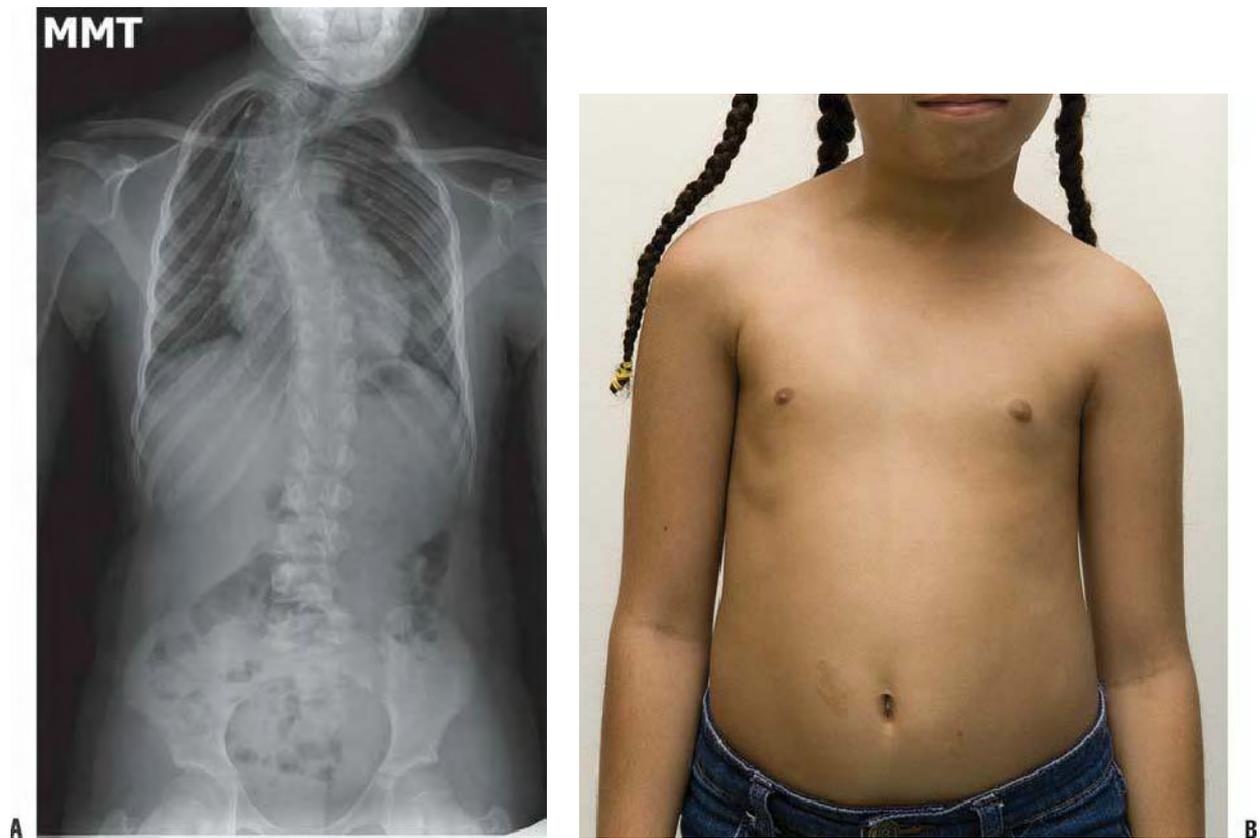


FIGURE 18-10. Progressive congenital kyphoscoliosis in an 8-year-old female. **A:** Erect radiograph with multiple vertebral anomalies centered at the cervicothoracic area including hemivertebra, wedge vertebrae, and a vertebral bony bar. **B:** Clinical photo illustrating the child's head tilt, neck and shoulder asymmetry.

truncal shift, or shoulder asymmetry as proxies for progression in the primary congenital curve. Small, subtle changes can be missed if one only examines the most recent studies; errors compounded and the window for optimal intervention can be missed. Due to the malformations, identification of the landmarks used to measure the Cobb angle(s) can be troublesome and inaccurate among and between observers (117). Consistency of the end points used must be maintained when measuring the radiographs over time; carefully remeasure the prior studies when necessary to determine if progression has occurred (107). The vertebral endplates are most commonly used as endpoints to calculate the Cobb angle, but pedicles may be used if endplate anatomy is unclear.

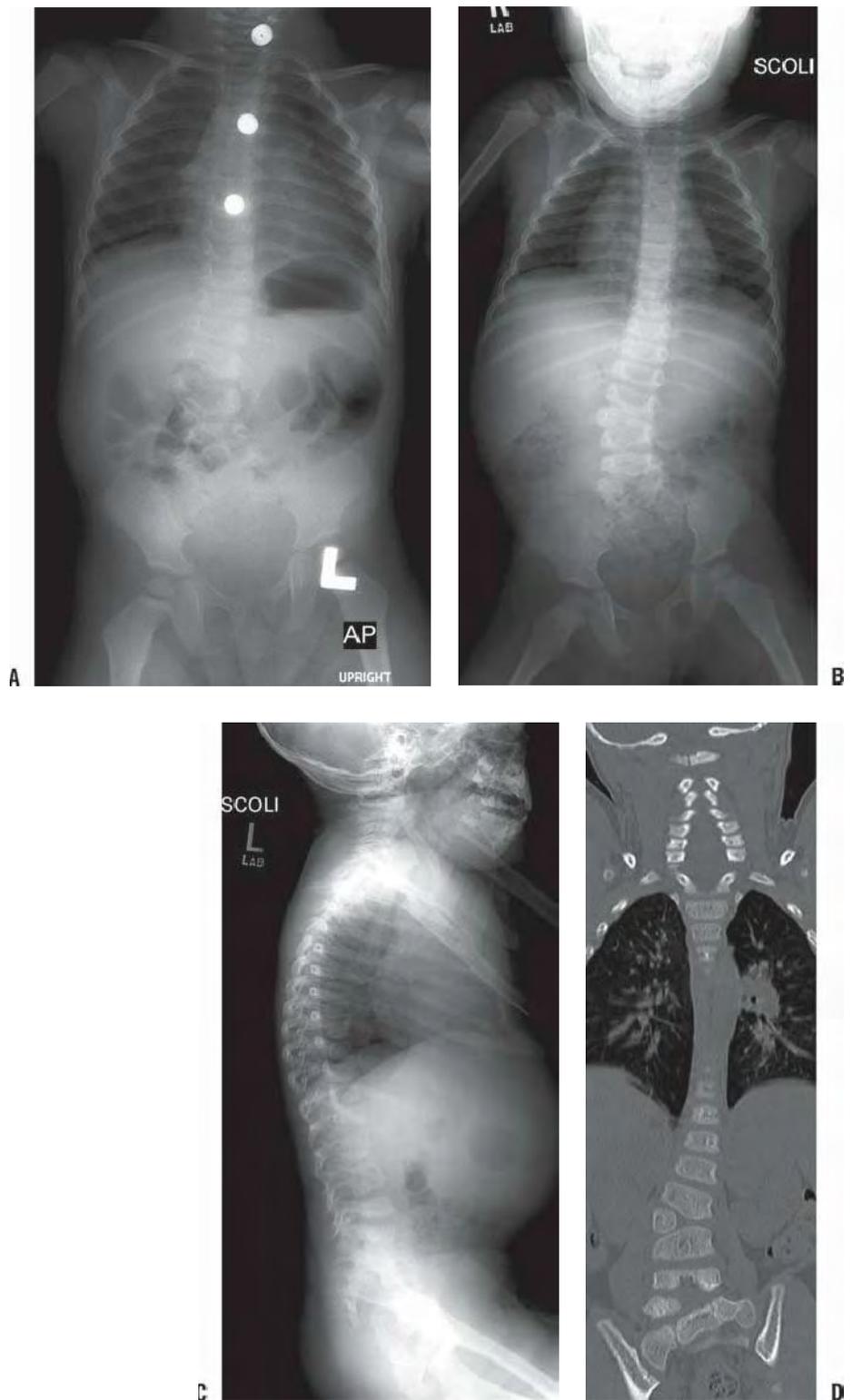
If there is progression of the compensatory, noncongenital scoliosis, without a change in the congenital primary curve, the possibility of spinal cord dysraphism needs to be investigated with an MRI scan of the entire spine. Spinal cord tethering, thickened filum terminale, syrinx, or lipoma may be present. However, as mentioned previously, given the fairly high rate of intraspinal anomalies noted on MRI in patients with congenital scoliosis, even isolated hemivertebra, routine use for all cases of congenital scoliosis may be justified (118).

Nonsurgical Treatment. There is little evidence that bracing is effective for congenital scoliosis; the congenital portion

of the curve is usually short and inflexible. However, Winter et al. (119) reported that an orthosis may be useful in certain congenital circumstances: compensatory curves above or below the congenital segment, long sweeping congenital curves with a flexible component, and for maintaining truncal and spinal balance following localized fusion during growth. A brace can be used in a young child as a supplement to surgery to curtail activities and protect instrumentation in soft bone. Bracing is not appropriate for congenital kyphosis, lordosis, or scoliosis with a known poor prognosis, such as a unilateral bar with a contralateral hemivertebra.

Halo Traction. Previously viewed as only of historical interest, halo-gravity traction is a viable adjunct in the modern treatment of severe scoliosis and is safe for congenital patients, in general. The exception is a patient with a short, sharp angular deformity associated with kyphosis—proceed with caution or avoid traction altogether. Used preoperatively before posterior correction and fusion or between staged anterior release procedures, halo-gravity traction is effective at maximizing correction of stiff deformities and well tolerated by patients, since they are allowed out of bed to a wheelchair or walker in traction. This treatment also has medical benefits; in the week(s) preceding surgery, patients can perk up nutritionally, gain weight, and improve pulmonary function and bone density.

FIGURE 18-11. Progressive lumbosacral congenital scoliosis in a 2-year-old male. **A:** Erect posterior–anterior (PA) radiograph of the child at 18 months showing ipsilateral hemivertebrae at L2 and L5/S1. **B:** Erect PA radiograph at 2 years of age showing progression of the lumbosacral scoliosis, truncal shift, coronal imbalance and pelvic obliquity. **C:** Lateral radiograph with mildly decreased lumbar lordosis. **D:** Coronal reconstructed computed tomography (CT) scan illustrating lumbar hemivertebra and lumbosacral hemivertebra with oblique sacrum.



Initially, traction should start with light weight and increase slowly while the patient is monitored for complaints or neurologic changes, and the maintenance of traction is variable from 2 weeks to many months. Clinical studies are hard to analyze since a small group of congenital patients are reported within a group of other etiologies of severe scoliosis, but most authors agree that there is improvement of the Cobb angle (28% to

35%), truncal height and balance, SAL, frontal and sagittal alignment, and avoidance of neurologic injury (120–123).

Surgical Treatment

Management Themes. After a thorough analysis of the deformity, the natural history or documentation of progression may be an indication for surgery. A multimodality evaluation

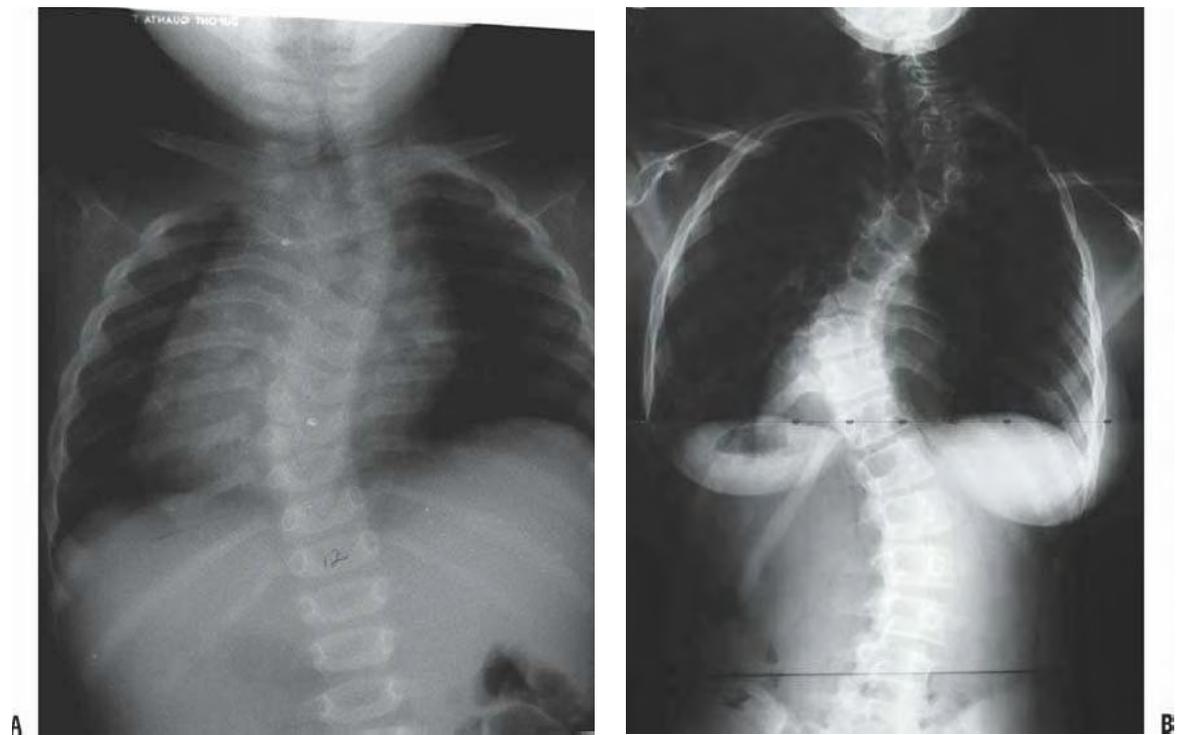


FIGURE 18-12. Progressive congenital scoliosis with development of compensatory thoracolumbar curve. **A:** Chest radiograph in the newborn nursery of an infant female showing contralateral thoracic hemivertebrae (hemimetameric shift). **B:** PA erect radiograph 14 years later showing the natural history: progression of both congenital curves and development of a compensatory lumbar curve.

of the patient is necessary to assess and manage any associated anomalies, pulmonary function, and nutritional status. The surgical treatment of congenital scoliosis can generally be divided into procedures that will prevent or control further deformity, gradually correct deformity, or ones that acutely correct the deformity. Fusion *in situ* is the classic example of a procedure for prevention or control; convex anterior hemiepiphyodesis and posterior hemiarthrodesis may gradually correct congenital scoliosis when done at the appropriate time, with the remaining growth and hemivertebrae resection or spinal osteotomy having the potential to acutely correct even severe deformities.

Historically, experts have asserted that it is easier to prevent a deformity than to correct one, and this reiterates the principles of vigilance in observation and knowledge of the natural history of the type of anomaly (124, 125). However, the ability to predict the natural history of many mixed, complex anomalies seen in congenital scoliosis is variable and sometimes an irrelevant issue if the patient presents when the deformity is severe (Figs. 18-13 and 18-14) or previous procedures have proved to be unsuccessful. The armamentarium of surgical options, therefore, must include a spectrum of techniques from the simple to the complex procedure for salvage when there is no other option. The goals of management of patients with congenital scoliosis are that at skeletal maturity, the patient should have a balanced spine, maximized pulmonary function, optimized spinal height, and no deterioration of neurologic function.



FIGURE 18-13. Progressive congenital scoliosis and associated anomalies in a 9-year-old female. The mixed anomalies include hemivertebrae, block vertebrae, wedge vertebrae, a vertebral bony bar, and fused ribs. She also has congenital short femur and a tethered cord.

In Situ Fusion. If a curve is predicted to worsen or progression has been documented, an *in situ* fusion can halt progression and is ideal for stabilizing a small curve, provided it is done at the proper time in a young child. The classic anomaly appropriate for this treatment is a unilateral bar or a unilateral bar with a contralateral hemivertebra that is diagnosed early with no associated deformity, since there is thought to be little potential growth on the concavity of

the deformity. *In situ* fusions are safe from a complication standpoint and technically simple to perform even in small children, but may be unreliable to control progression due to crankshaft and add-on phenomenon. The growth potential of the anterior column of the spine must be assessed as minimal by MRI or CT before a posterior tether (fusion) is applied in a young child, or progression of the curvature may result even in the case of a solid arthrodesis (126–128).



FIGURE 18-14. Hemivertebra in a 4-year-old female. **A:** PA radiograph illustrating progressive thoracic scoliosis due to a hemivertebra at T7. The risk of severe compensatory curves warrants surgical intervention. **B:** Lateral radiograph showing increased thoracic kyphosis. **C:** Coronal reconstructed computed tomography (CT) scan illustrating thoracic hemivertebra. **D-G:** Three-dimensional reconstructed CT scan in multiple planes of rotation. This type of preoperative modeling is useful for planning resection procedures.



E



F



G

FIGURE 18-14. (Continued)

When the potential for anterior growth exists, the patient should have an anterior growth cessation procedure (epiphysiodesis or fusion) performed, open or thoracoscopically, in the thoracic spine. Occasionally, an *in situ* fusion may offer correction over time—if there is growth potential on the concavity of a scoliosis or anteriorly for kyphosis—but more often, it is meant to prevent progression of a relatively short congenital deformity in an otherwise balanced spine.

Most commonly performed posteriorly for a kyphosing scoliosis, the procedure can be performed anteriorly as well, as a lordosing deformity would dictate. The fusion levels selected must include all of the vertebrae in the measured Cobb angle and typically extend at least to a level above and below the anomalous segment(s) (129) or the entire length on the contralateral side of a bar (130). Classically described for small children, postoperative immobilization is provided by a cast, but newer, downsized instrumentation may be implanted safely to provide more stable fixation (131), reduce the chance of pseudarthroses, and limit the immobilization time (132). The concern over an extensive fusion of the spine in a young child is that it stops growth. Inhibition of spinal growth can lead to a disproportionate posture, lead to a shortened trunk, hinder lung development, and lead to TIS (133–135).

Convex Hemiepiphysiodesis/Hemiarthrodesis. This procedure offers partial growth arrest on the convex side of the deformity with remaining growth, and thus, little or no concave growth should be expected. Most commonly applied to isolated hemivertebra in young children, unilateral failures of formation with modest curves are the deformities that are most appropriate for this technique. Some correction can be achieved over time, depending on the age and growth potential of the child at the time of the procedure; some correction is also initially obtained in the postoperative cast. Long-term results show that <15 degrees of total correction can be achieved, with some patients obtaining no correction of the curve or truncanal imbalance (136–138). The procedure is generally conducted with an anterior approach (open or thoracoscopic), extirpation of the disks and cartilaginous growth plates, anterior spinal fusion and a posterior spinal hemiarthrodesis on the convexity over the same segments (the concave side is not exposed since it could lead to spontaneous fusion), and postoperative immobilization in a cast or a brace in a somewhat corrected position.

Results are variable, but the best outcomes seem to be in patients younger than 5 years with an isolated lumbar hemivertebra with a progressive curve <40 to 50 degrees (139, 138). One of the complications is failure to achieve correction. Roaf (124) reported that 60% of his patients had correction of at least 10 degrees, whereas Keller et al. (140), with a transpedicular approach showed that 37% of curves improved, 42% remained the same, 16% progressed 10 to 15 degrees, and 5% progressed more than 15 degrees. Thompson et al. (138), in their series of 30 patients showed that 76% had an improvement of the Cobb angle; the best corrections were obtained in the lumbar spine rather than in the high thoracic or thoracic spine. This procedure was not recommended in cases with associated kyphosis,

since anterior fusion could contribute to the kyphotic component (141), but more recent refinement of the technique by Cil et al. (142), with extension of the anterior hemiarthrodesis all the way to the posterior longitudinal ligament may avoid sagittal deterioration. Convex hemiepiphysiodesis and *in situ* fusion are low-risk procedures, but due to the limited correction and unpredictability of future correction with growth, other methods are more reproducible for treatment of congenital scoliosis.

HEMIVERTEBRA EXCISION

Excision of the hemivertebrae offers immediate correction of the congenital deformity and restoration of truncal balance over a short segment. The ideal indication is a young patient (<5 years of age), with an isolated hemivertebrae in the thoracolumbar, lumbar, or lumbosacral area with progression and/or associated coronal imbalance. Cervicothoracic hemivertebrae can cause marked shoulder and neck imbalance and are amenable to excision but obviously carry with them an increased risk of neurologic deficit due to the manipulation of the column around the spinal cord needed to perform the procedure and associated vertebral artery anomalies (143). The inflexible thorax around the CVM may pose limits to the correction. The spectrum of the technique of hemivertebrae excision ranges from staged anterior and posterior resection procedures, simultaneous anterior and posterior resection, and posterior-only excision. Small stature instrumentation, multimodality neurophysiologic monitoring, advances in surgical techniques, and proper experience have lowered the complication rate and improved outcomes (Figs. 18-15 to 18-21).

The excision of hemivertebrae, first described by Bradford and Boachie-Adjei (144) and then subsequently by Klemme et al. (145), was outlined as a sequential anterior, then posterior excision, which offered a safe, circumferential exposure of the spine and allowed removal of the disks above and below the hemivertebra, in addition to the bone itself. Then, the corresponding lamina and pedicle can be removed, the convexity compressed posteriorly, and a small area fused with instrumentation, and the bone that is removed can be used for graft. In an effort to save the operative time needed to reposition the patient and drape again between the anterior and posterior procedures, both can be performed simultaneously. This allows proper control of the sagittal plane and the ability to titrate the excision for the proper correction, better visualization, reduced risk of crankshaft, and pseudarthrosis since the disks are completely removed (146, 147). In a series reported by Hedequist et al. (147a), 18 patients underwent simultaneous exposures with excision and instrumentation at an average age of 3 years, and the authors achieved 70% correction without any neurologic complications or pseudarthroses.

Hemivertebra excision performed as a posterior-only procedure is growing in popularity since the report of Ruf and Harms (148) in 2002. They described excellent results in patients younger than 6 years who underwent posterior-only hemivertebrae excision and segmental transpedicular screw instrumentation for curves with an average Cobb angle of 45 degrees. Curve correction to 14 degrees (75%) was

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Hemivertebra Excision (Figs. 18-15 to 18-21)

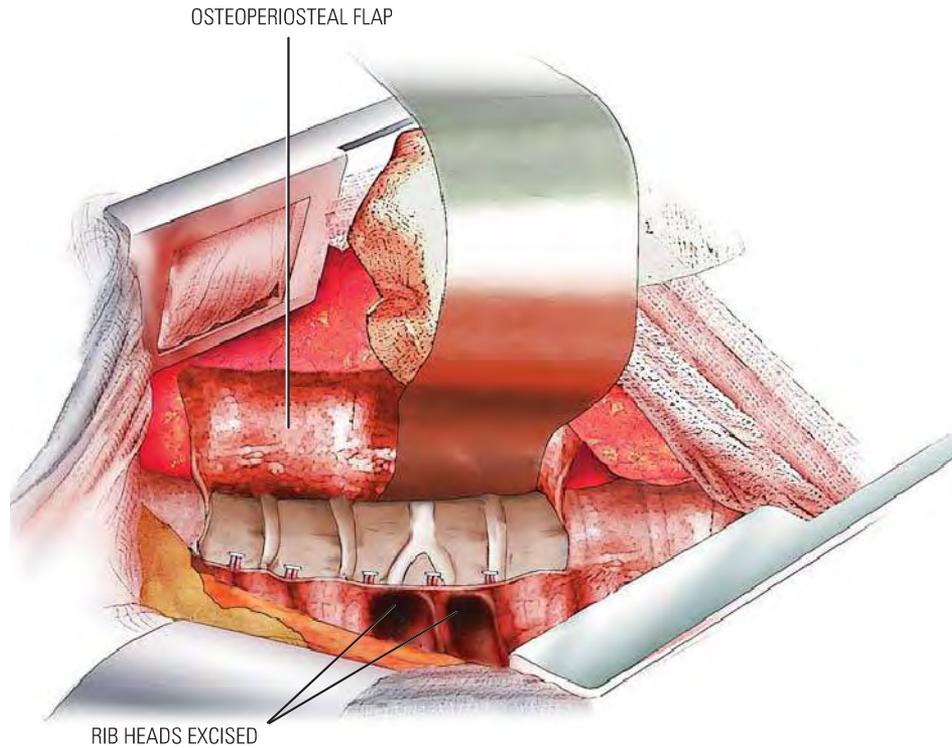


FIGURE 18-15. Hemivertebra Excision. The patient is taken to the operating room and placed on a standard operating table in the lateral decubitus position with the hemivertebra side facing up. Standard lateral decubitus positioning is used with an axillary roll and padding under the peroneal nerve, between the legs, under the down arm, and between the arms. In general, we prefer to flex the operating table to allow maximal visualization and opening up of the disk spaces. The patient's hemithorax and hemiabdomen are prepped from just beyond the midline anteriorly to just beyond the midline posteriorly.

Spinal cord monitoring should be done throughout the procedure and can be supplemented by a wake-up test.

If a thoracic hemivertebra is to be excised, a standard thoracotomy incision is made, removing the rib one or two levels above the hemivertebra. If the hemivertebra is at the thoracolumbar junction, a standard thoracoabdominal approach through the 11th or 10th rib may be used. This allows exposure and excision of a hemivertebra down to the L2 or L3 level. For lumbosacral junction hemivertebrae, a standard retroperitoneal approach is used. It is good practice to identify the hemivertebra to be excised with radiographic markers, such as a Keith needle, before exposure, to ensure that the correct level is approached.

In exposing the hemivertebra, after segmental vessel ligation, either an extraperiosteal or subperiosteal exposure can be used. We prefer to use a subperiosteal exposure to allow a flap with osteogenic potential to overlay the resection site. The spine must be subperiosteally exposed from the inferior aspect of the vertebral body above to the superior aspect of the vertebral body below. The osteoperiosteal flap must be elevated all the way around to the opposite side as shown. The flap allows protection of the great vessels on the concavity of the curvature.

In the thoracic spine, the rib head at the level of the hemivertebra should be removed to allow exposure of the transverse process and the disk space. The rib head at the next inferior vertebra in the thoracic spine may also need to be removed to allow for adequate exposure of the disk space. Bleeding can be controlled with bone wax.

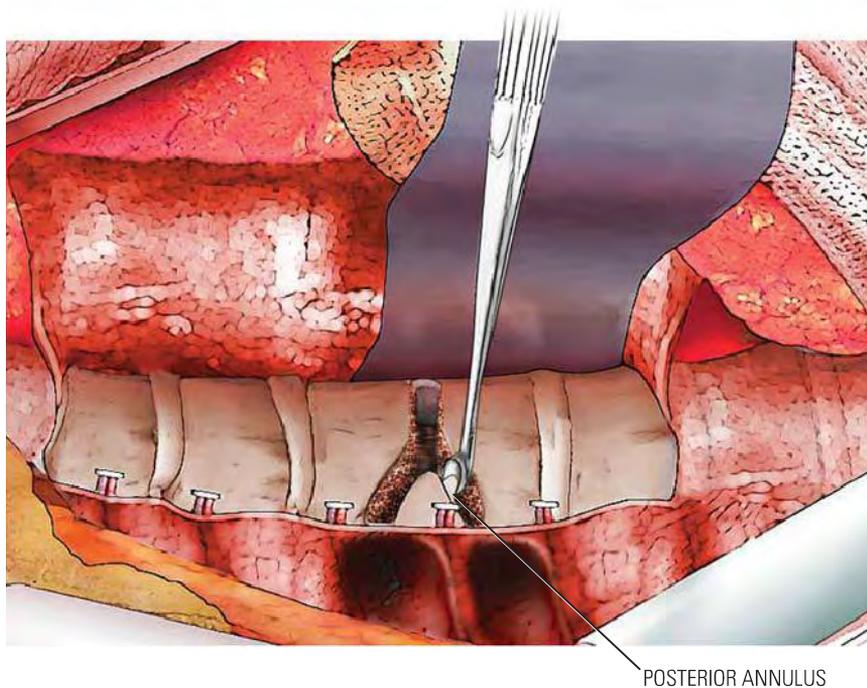


FIGURE 18-16. All disk materials above and below the hemivertebra are excised back to the posterior annulus using knife, curette, and rongeurs. We tend to use the rongeurs at the more superficial levels and then curettes as one approaches the posterior annulus.

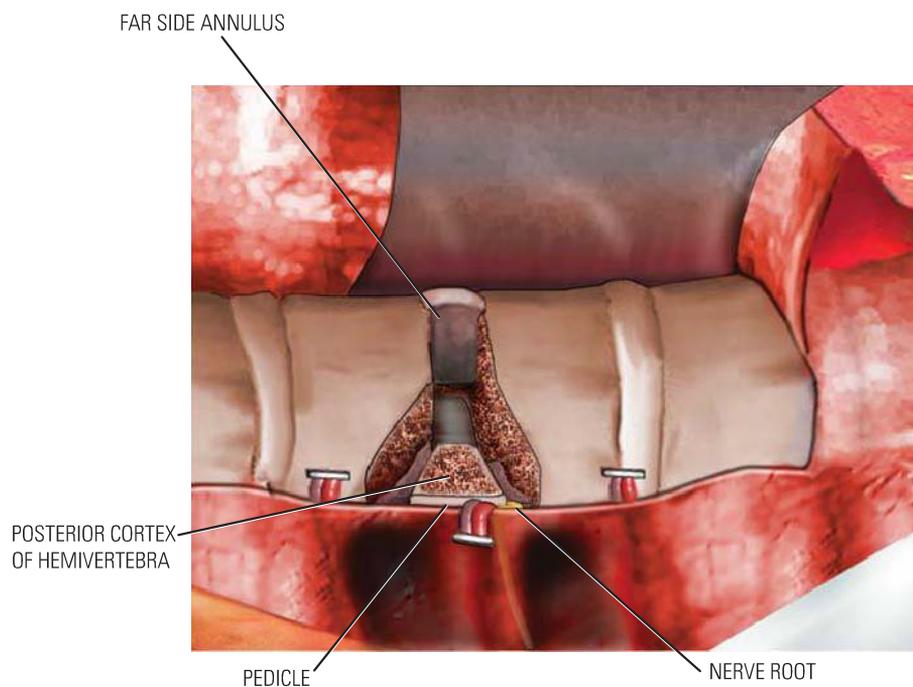


FIGURE 18-17. The inferior end plate of the vertebra above and the superior end plate of the vertebra below must also be excised. This can be done with curettes or Cobb elevators.

It is important to leave a portion of the most lateral annulus intact to act as a stabilizing tether during the posterior hemivertebra removal and closure of the gap. It is important to excise all of the soft tissues opposite to the hemivertebra. This may appear as disk material or fibrocartilage. Failure to excise all of this tissue except for the most lateral annulus will result in ineffective closure of the gap.

After the disk material has been removed, the hemivertebra excision is begun. This can be accomplished with a burr, rongeur, or curette. We prefer to use rongeurs and curettes. The bone removed is saved to be used as bone graft in filling some of the gap anteriorly and also posteriorly. We use rongeurs to remove the bulk of the hemivertebrae and then curettes, curetting from the concave to the convex side toward the surgeon. All the cancellous bone is removed back to the posterior cortex. In the thoracic spine, the transverse process is subperiosteally dissected anteriorly and rongereed off at its base.

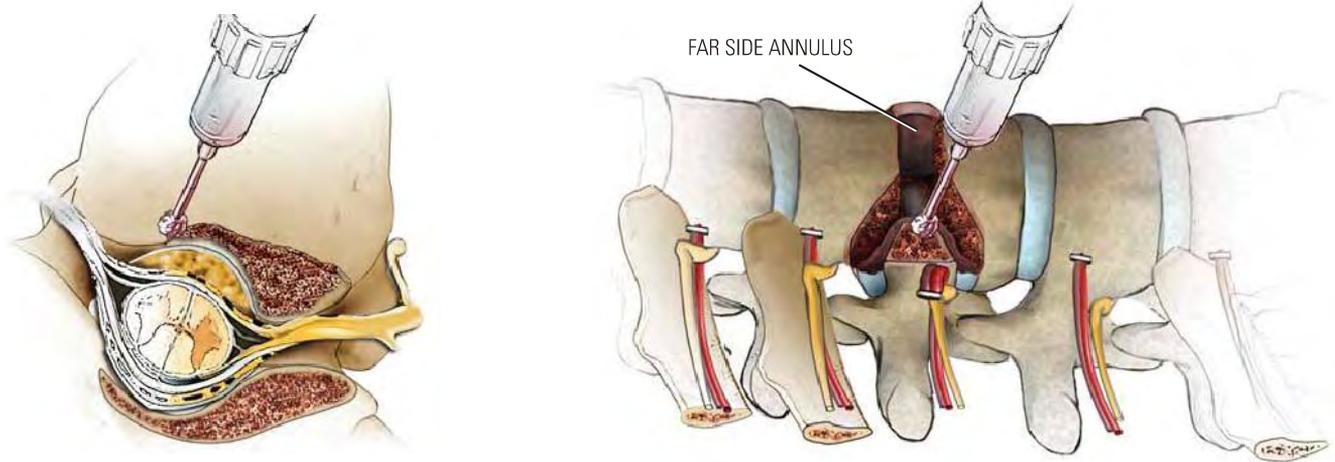


FIGURE 18-18. Final removal of the posterior vertebral cortex can be done in one of several ways. We prefer to use a diamond-tipped dental burr to enter the spinal canal in the midline. After a small hole is made, a small Harper-Kerrison rongeur may be used to remove the entire posterior cortex of the hemivertebra.

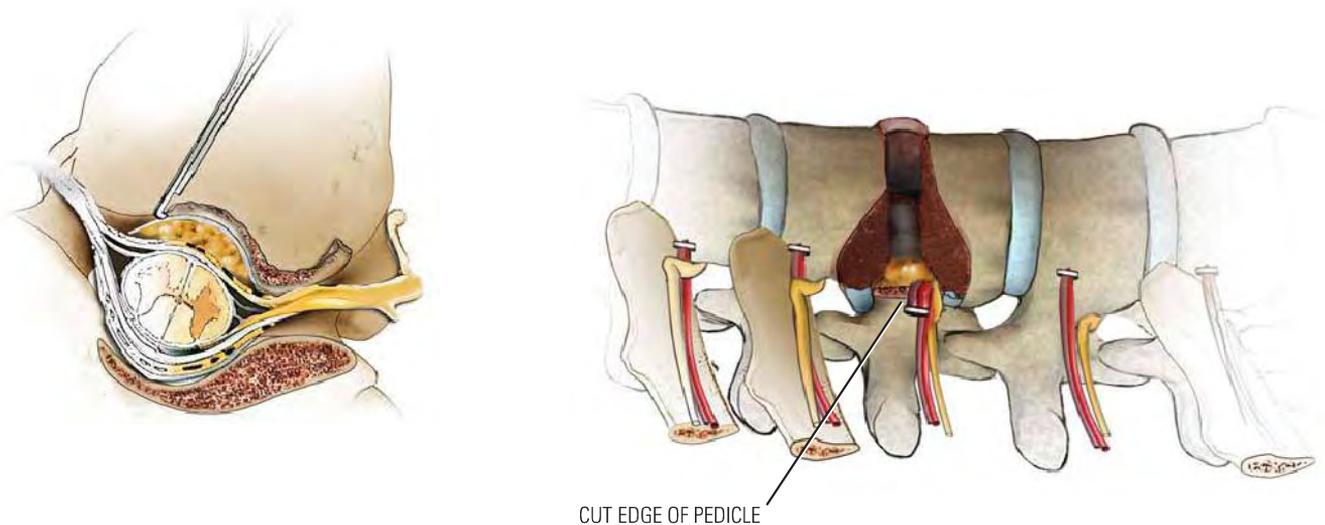


FIGURE 18-19. After the cortical bone is removed from the apex of the hemivertebra all the way back to the pedicle, the annular tissues superiorly and inferiorly can also be removed. The neural foramina both above and below the hemivertebra must be opened completely and the nerve roots visualized. Any bleeding that is encountered should be addressed with bipolar cautery or packing with thrombin-soaked Gelfoam.

The final structure removed anteriorly is the pedicle of the hemivertebra. The pedicle can be removed in several ways. It can be gradually nibbled away with narrow-nosed rongeurs or, as we prefer, the central portion drilled until decancellated followed by the outer walls being removed with either rongeurs or curettage toward the pedicle center.

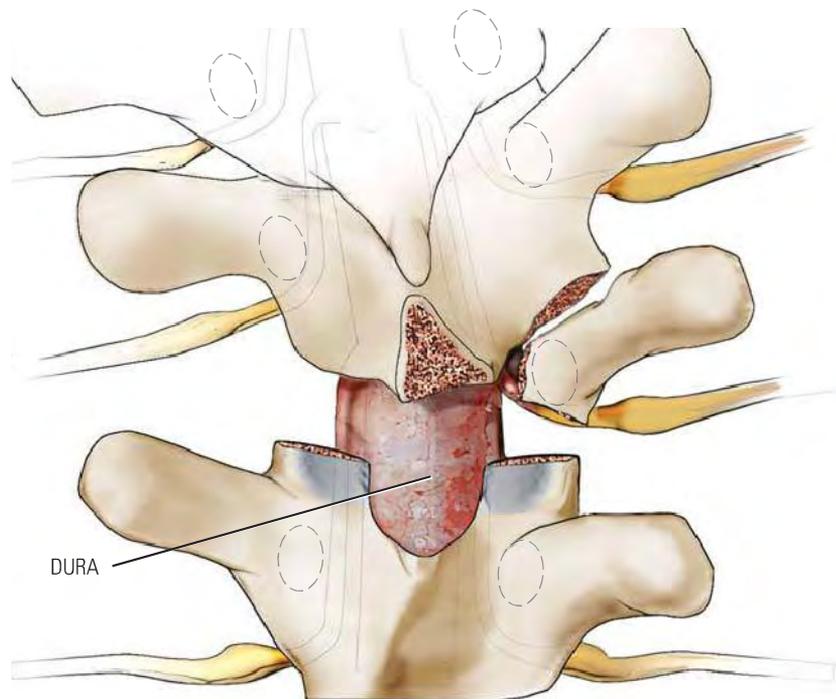
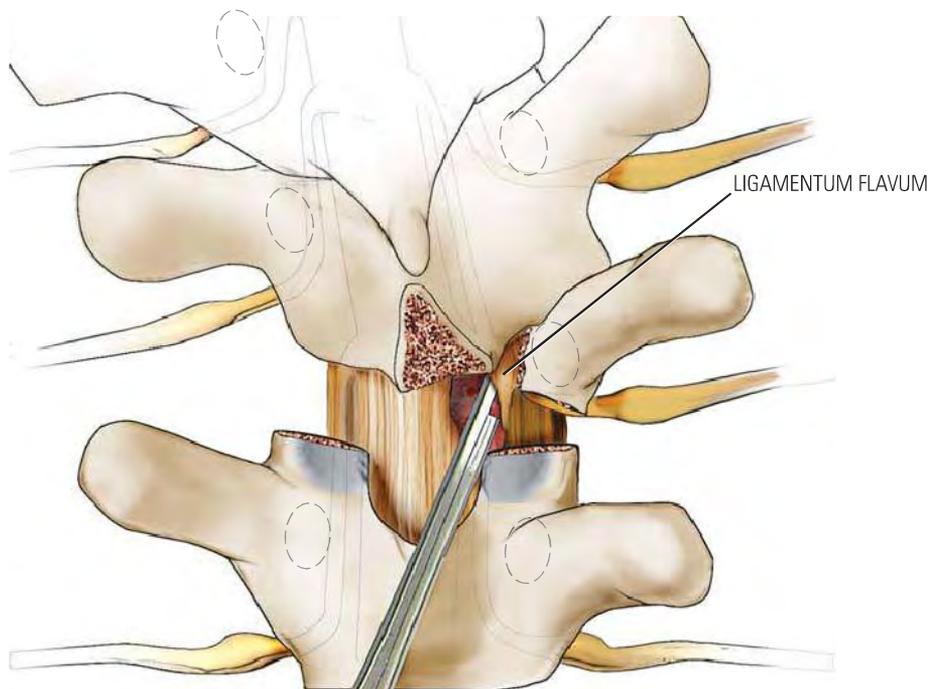
The pedicle bone should be nibbled away as far posteriorly as possible.

The dura is covered with a Gelfoam or Oxycel pad or fat graft, and then bone chips from the excised hemivertebra are loosely placed into the space. The osteoperiosteal flap is then sutured back down loosely, and in the chest the pleura is repaired. The wound is then closed in the standard fashion.

The second portion of the procedure can be done in the lateral decubitus position.

However, we prefer to roll the patient prone and transfer the patient to a spine frame (e.g., Jackson table, Relton-Hall frame). In some children, we prefer to use two laminectomy rolls (made of rolled bath blankets or foam), one under the chest and one under the iliac crests, leaving the abdomen free.

FIGURE 18-20. In the posterior approach, the spine is exposed from a standard midline exposure overlying the area of the hemivertebra. A radiograph is taken to determine the appropriate level for excision. The spine is then exposed subperiosteally. The ligamentum flavum above and below the hemilamina is carefully incised and removed with the Kerrison rongeur. Any bleeding is controlled by bipolar cautery or packing with thrombin-soaked Gelfoam. After the ligamentum flavum has been removed, the hemilamina is gradually removed from the midline to its lateral aspect. We generally do this with standard narrow-nosed rongeurs or Harper-Kerrison rongeurs. It is much easier to remove the posterior arch from the posterior approach when more of the pedicle has been removed by the anterior procedure.



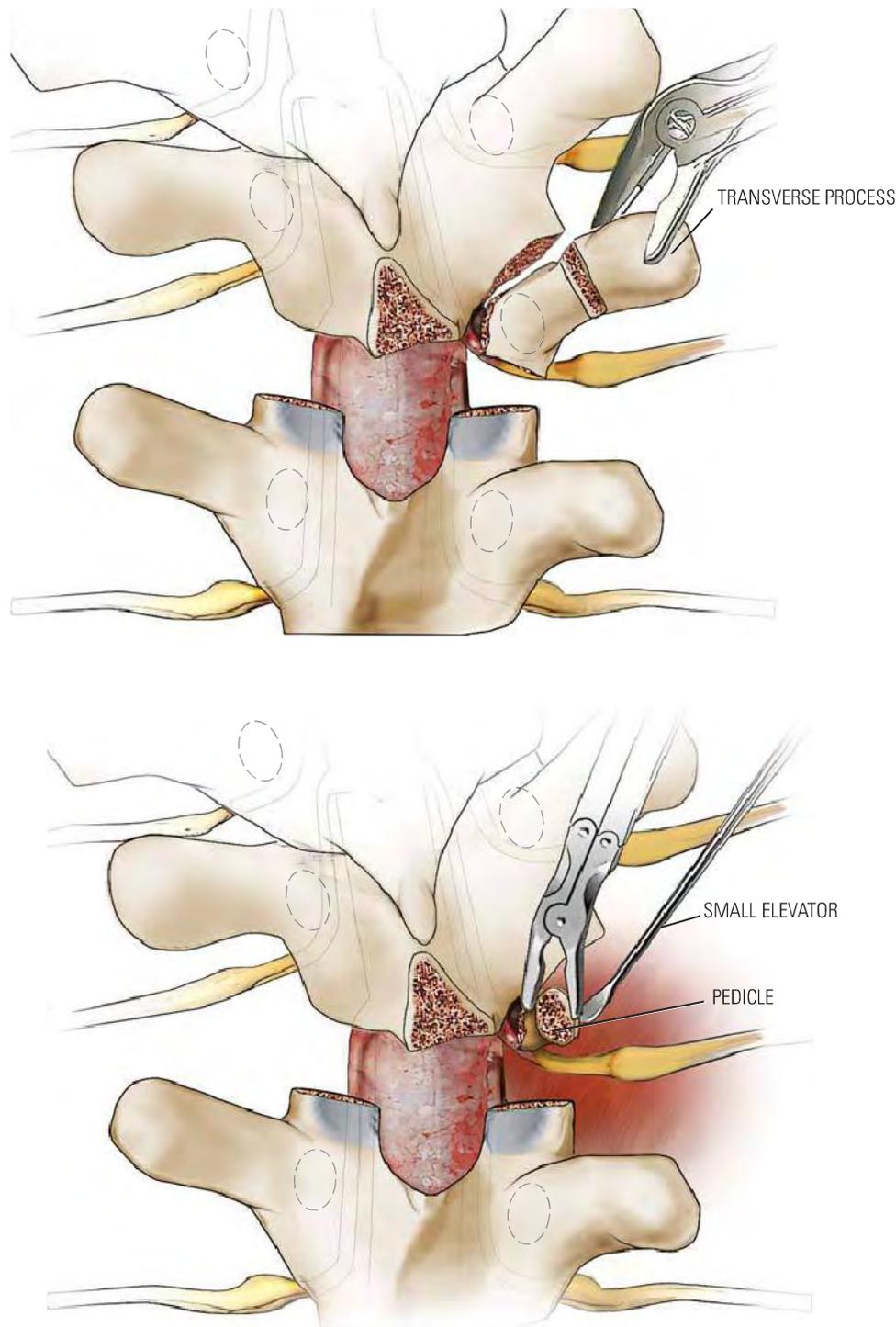


FIGURE 18-21. In the lumbar spine, the transverse process is transected at its base with a narrow-nose or Kerrison rongeur. In the thoracic spine, if the transverse process was left intact in the anterior approach, it must be removed at this point. After the lamina, facets, and transverse processes have been completely removed, the pedicle can be grasped with a rongeur or hemostat; with one prong inside the center of the pedicle and one on the cortical margin, the pedicle is slowly removed. If resistance is encountered, it is most important to use a small elevator to free the margin of the pedicle and remove it either *in toto* or piecemeal with narrow-nose rongeurs or curettes.

After all the bone is removed, the surgeon can easily visualize the dural sac with the nerve roots above and below the hemivertebra. The surgeon must take great care when watching the closure of the apical wedge osteotomy to make certain that the downgoing pedicle does not impinge on the two nerve roots going through the single neural foramen. If there is any concern about pedicle impingement, the superior pedicle must be thinned or even excised to prevent nerve root impingement. The laminae above and below can then be decorticated and the portions of the rib removed from the anterior portion of the procedure; the morselized hemivertebrae can be used for bone graft.

FIGURE 18-21. (*Continued*) When the hemivertebra to be excised is at the lumbosacral junction, a separate incision may be necessary to expose the iliac crest and obtain cortical and cancellous bone.

In very young patients, internal fixation is generally not possible, and the defect is closed and maintained in that position by casting. In toddlers and older children, small compression devices may be applied using the rod and hook configuration. Another alternative in older patients is the use of pedicle screws in the pedicle above and below the hemivertebra, with a small rod connecting the two screw heads. If no internal fixation is used, after wound closure, the patient must be turned carefully onto a pediatric spica table and a body spica cast that includes both legs down to the ankles applied. The cast should extend proximally up to the clavicle line. The patient's torso should be bent into the convexity of the curvature to maintain correction. Radiographs should be obtained in the operating room to ensure that the wedge is closed and that adequate correction has been obtained. It is also important to make certain that pressure is applied to the apex of the deformity posteriorly while the plaster is drying, to prevent the spine from drifting into kyphosis. If adequate correction has not been obtained, the cast can be wedged while the patient is still under anesthesia. If internal fixation is used, the surgeon will decide whether casting is necessary. If the surgeon believes that the internal fixation was very stable at surgery, the patient may be placed either in a cast or a thoracolumbar spinal orthosis after surgery, with a single-leg extension, depending on the surgeon's preference. In our opinion, external immobilization is important regardless of the stability of the internal fixation.

maintained at an average 3.5 years follow-up with no neurologic complications (Fig. 18-22). The purported advantages were the avoidance of an anterior surgery with similar correction of local deformities. The use of transpedicular instrumentation at this age may raise eyebrows due to concern over the risk of closure of the neurocentral synchondrosis (NCS) and iatrogenic spinal stenosis. The NCS closes at 3 to 6 years of age,

but the spinal canal diameter at birth is already roughly two-thirds of adult size (149–151). In subsequent reports of the technique by Ruf and Harms (152, 153), no canal stenosis was found in young children upon follow-up imaging years later. However, somewhat contradictory to these findings, transpedicular instrumentation in compression mode across the NCS

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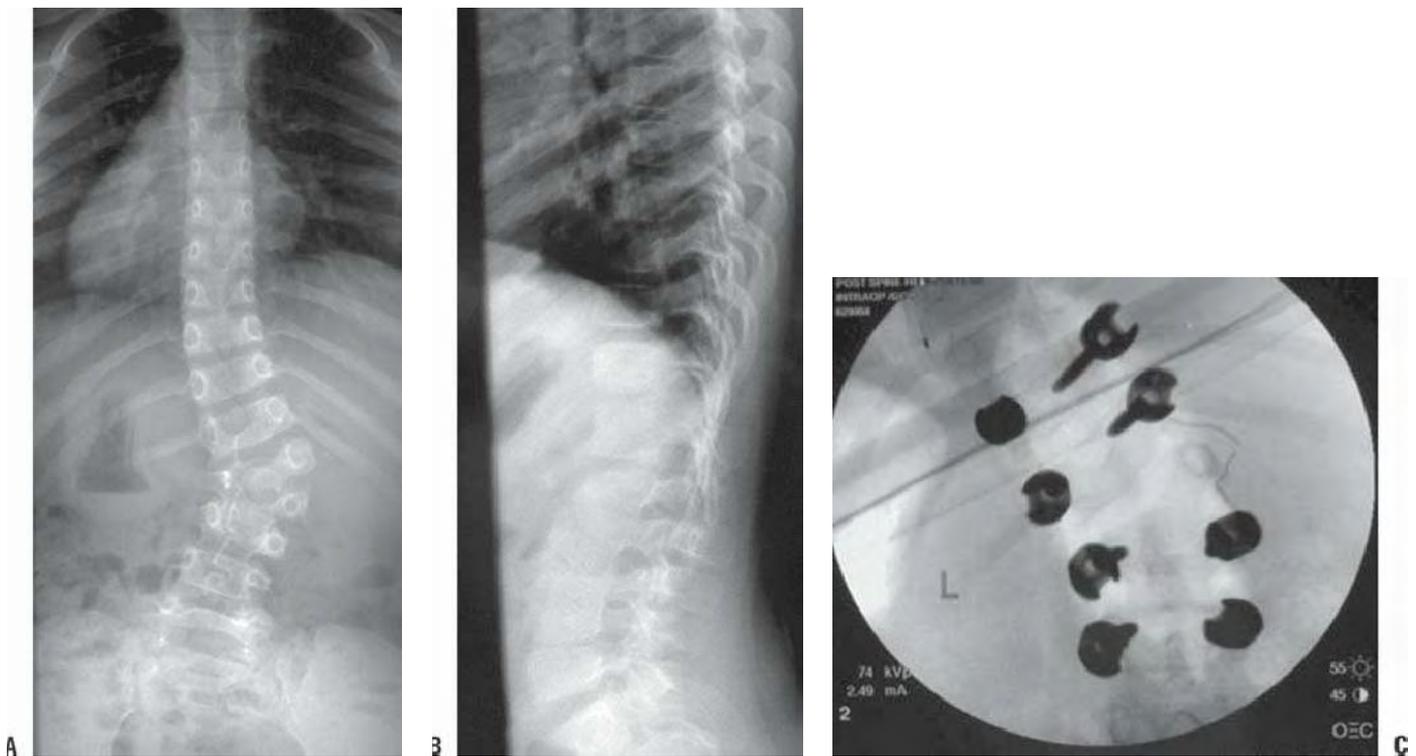


FIGURE 18-22. Posterior-only hemivertebra resection in a 5-year-old female. **A:** PA erect radiograph illustrating L1 hemivertebra. **B:** Lateral erect radiograph showing mild thoracolumbar kyphosis. **C:** Intraoperative fluoroscopy after posterior exposure to localize CVM and place pedicle screws.

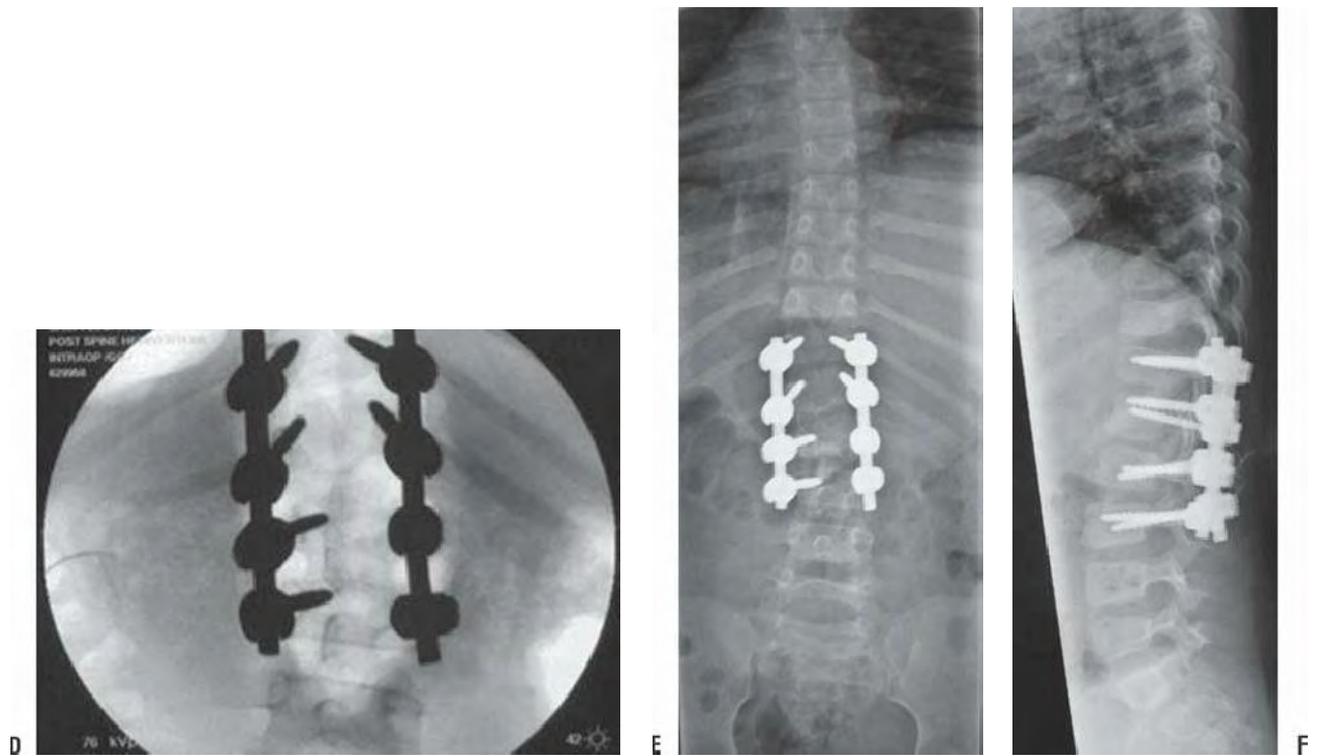


FIGURE 18-22. (Continued) **D:** Intraoperative fluoroscopy after resection of hemivertebra and compression of pedicle screw and rod instrumentation across the convex defect and correction of scoliosis. **E:** Postoperative PA radiograph showing maintenance of correction and excellent coronal balance. **F:** Lateral erect view showing restoration of thoracolumbar lordosis.

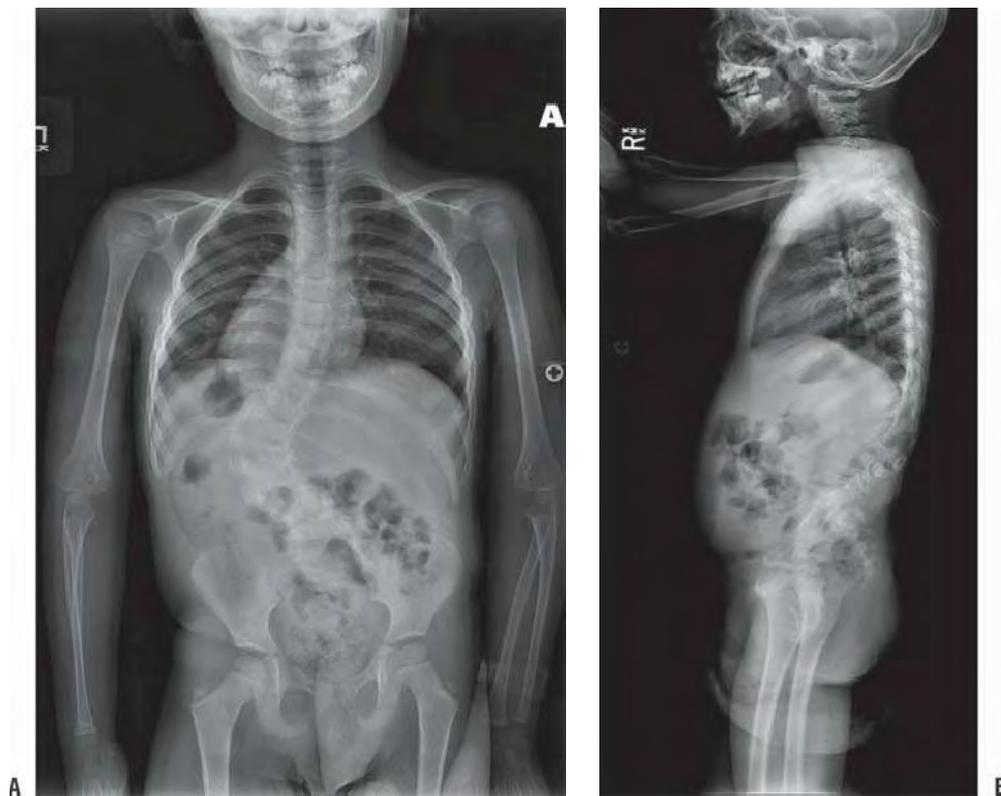


FIGURE 18-23. Posterior-only hemivertebra excision, asymmetric pedicle subtraction osteotomy and multilevel fusion in a 4.5-year-old boy with progressive congenital kyphoscoliosis. **A:** PA erect radiograph illustrating thoracolumbar congenital scoliosis. **B:** Lateral radiograph showing thoracolumbar kyphosis near the apex of the scoliosis. **C:** Sagittal MRI scan.

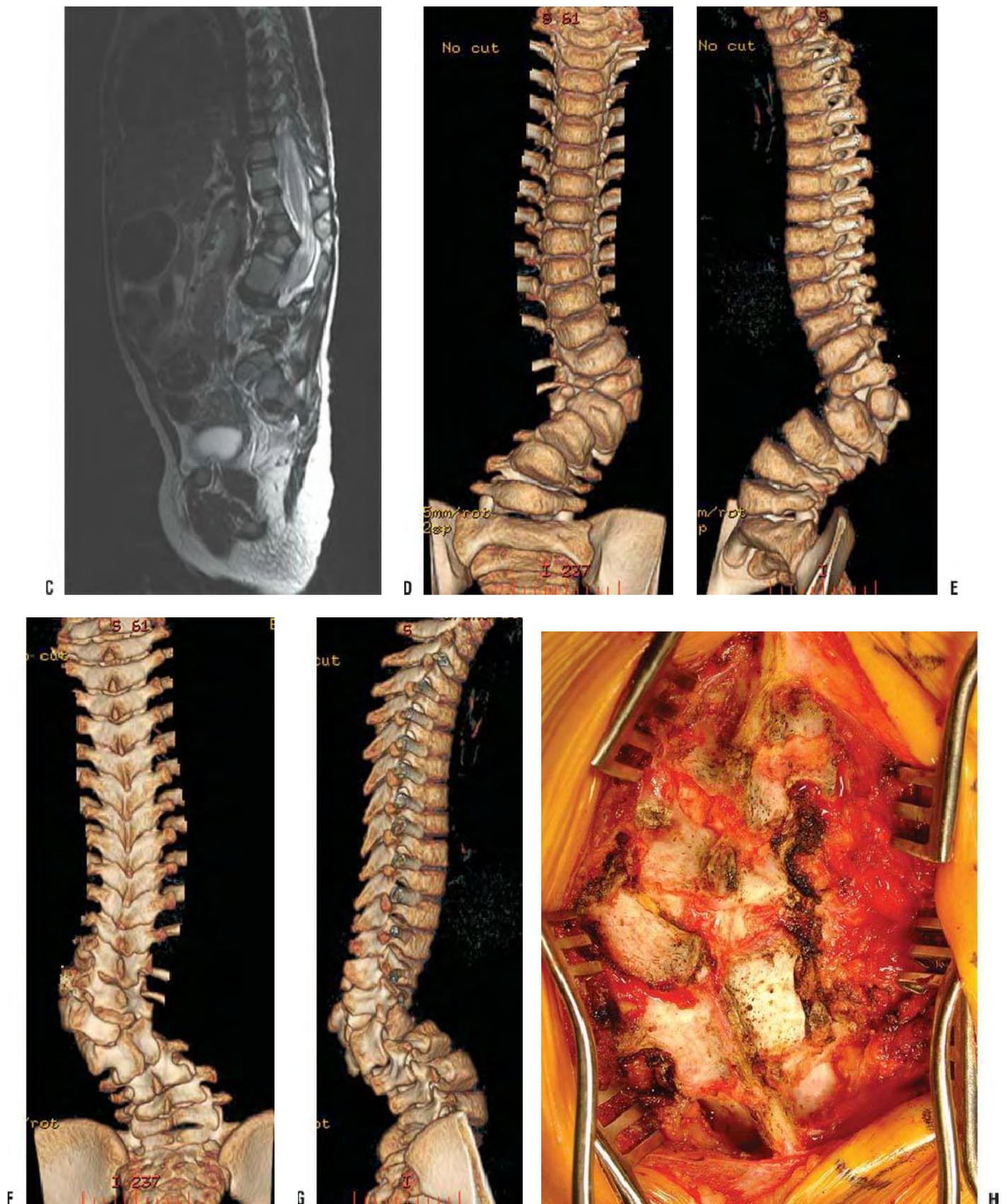


FIGURE 18-23. (Continued) **D-G:** Three-dimensional reconstructed CT scan in multiple planes of rotation. This type of preoperative modeling is useful for planning resection procedures in complex deformities since it identifies anatomy, confirms relationships between and anterior and posterior elements and visualizes the sagittal plane with great clarity. **H:** Intraoperative photograph after subperiosteal exposure of the levels to be treated. The posterior elements and landmarks can be correlated with the CT scan images to confirm the level of resection.

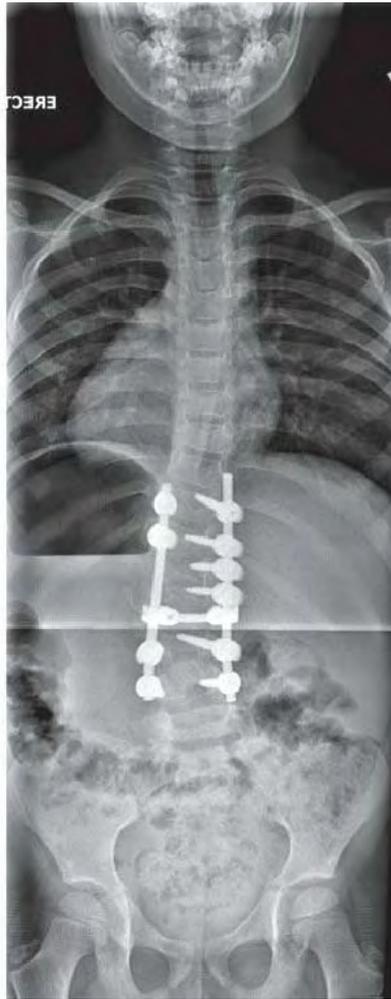
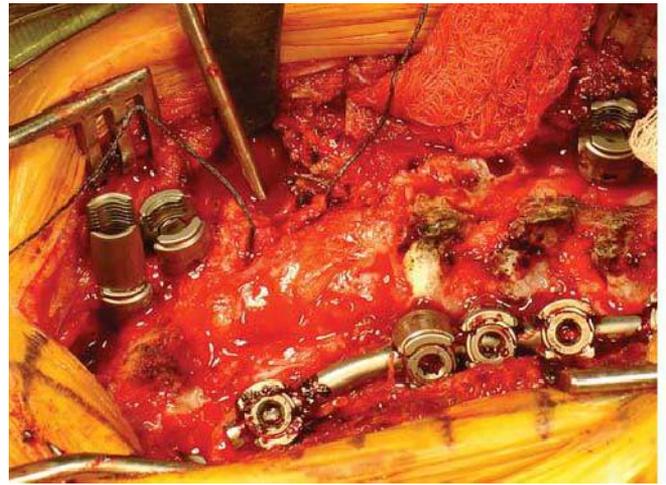


FIGURE 18-23. (Continued) **I:** Intraoperative fluoroscopic image to confirm levels and pedicle location with needles or Kwires. **J:** Intraoperative photograph after pedicle screw insertion, during hemivertebra resection and stabilization with temporary rod. **K,L:** PA and lateral radiographs 2 years postoperatively showing maintenance of correction and balance in both planes.

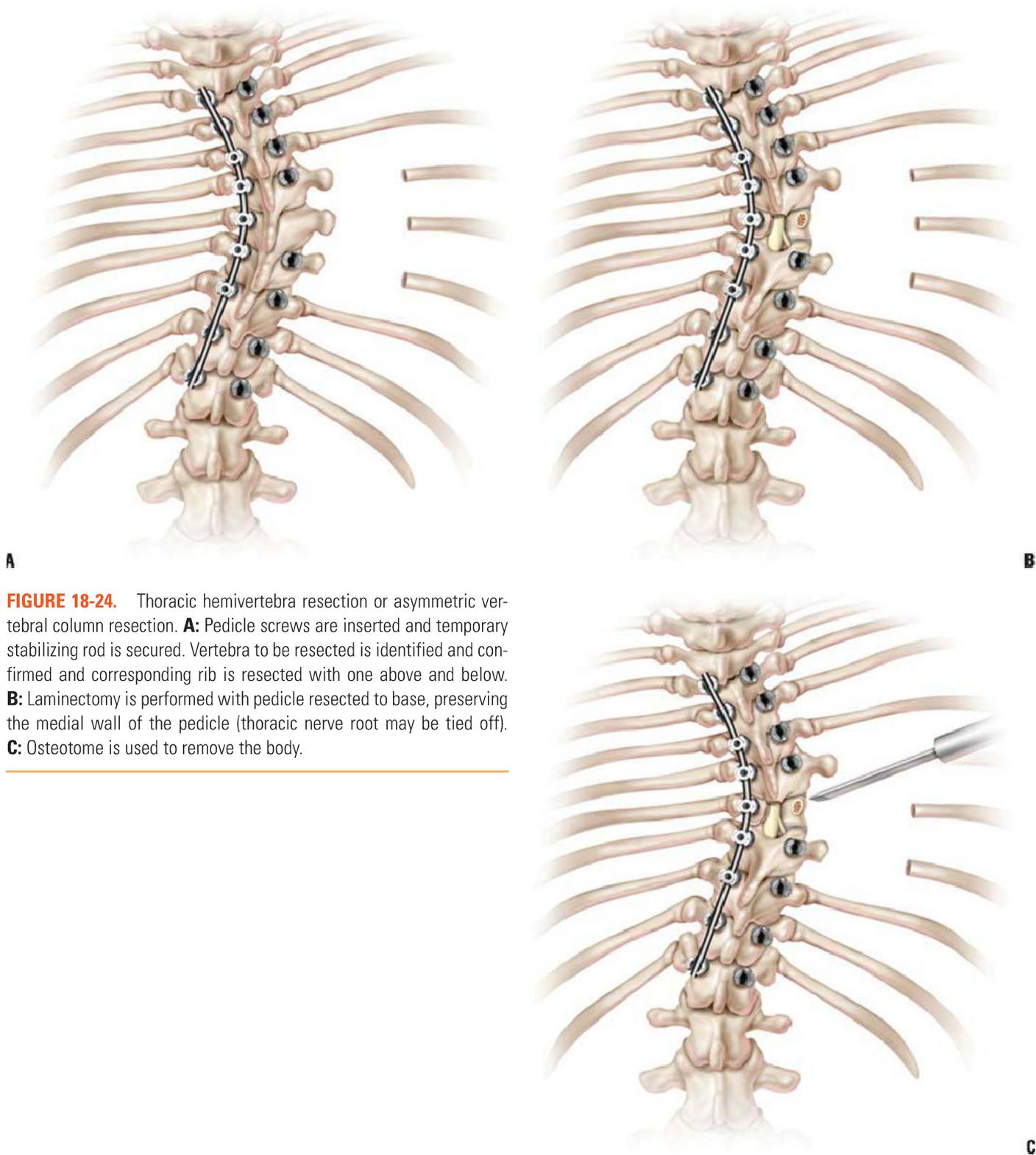


FIGURE 18-24. Thoracic hemivertebra resection or asymmetric vertebral column resection. **A:** Pedicle screws are inserted and temporary stabilizing rod is secured. Vertebra to be resected is identified and confirmed and corresponding rib is resected with one above and below. **B:** Laminectomy is performed with pedicle resected to base, preserving the medial wall of the pedicle (thoracic nerve root may be tied off). **C:** Osteotome is used to remove the body.

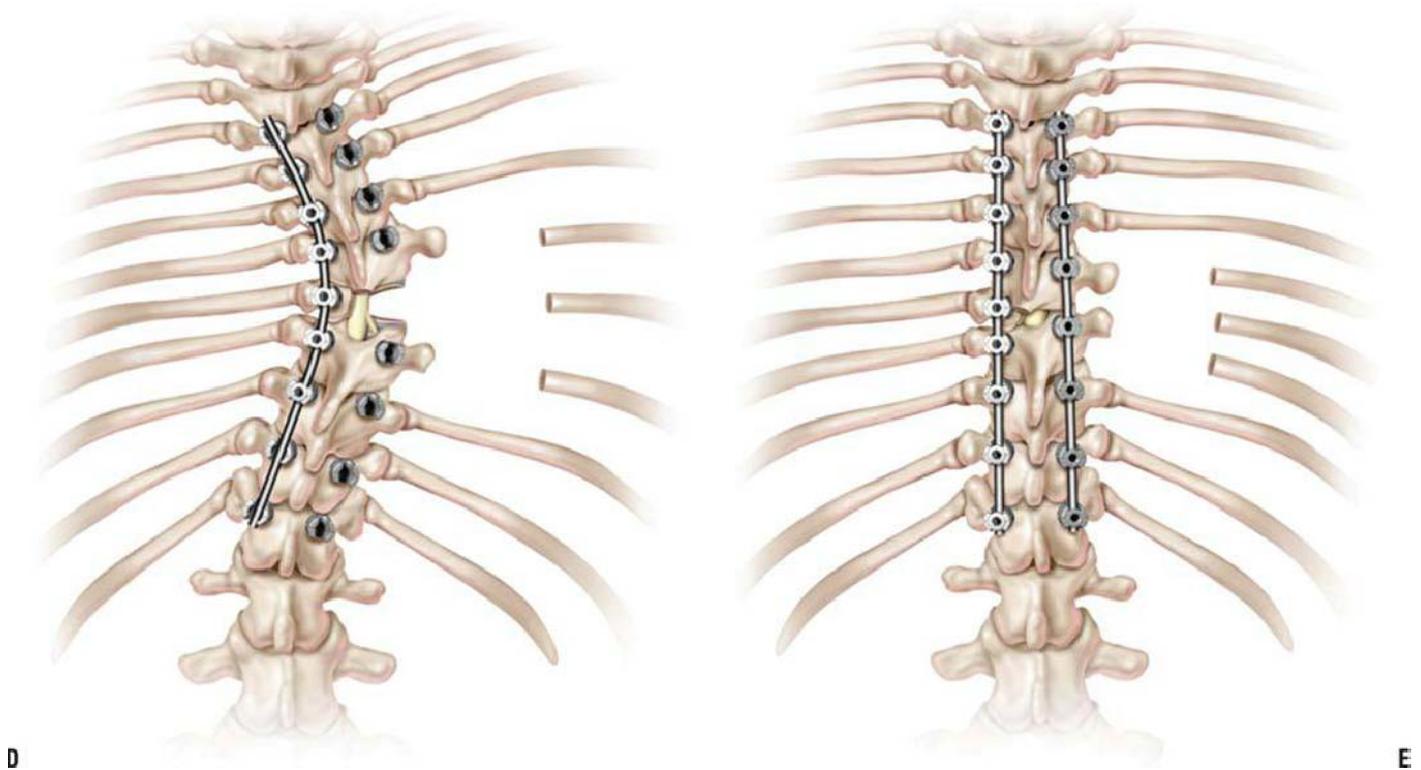


FIGURE 18-24. (Continued) **D:** Pituitary rongeurs and curettes are used to remove the disks above and below. The resection must reach the concave side in order to allow the convexity to close. **E:** Temporary working rods are exchanged for the final construct, the convexity is compressed and the resection is closed (a structural cage may be necessary for anterior column support). Care is taken to avoid subluxation of the spinal column, excessive buckling of the dura or laminar impingement. (Images courtesy of James Millerick.)

(and subsequent closure) did produce canal narrowing on the ipsilateral side in a porcine study (154).

Suk et al. (155) prefers the posterior-only procedure as well, due to its decreased operative time, ability to address the deformity at the apex, and correction by controlled shortening across the resection gap. Shono et al. (156) reported obtaining 64% correction with posterior-only techniques in 12 adolescent patients and no complications. In a multicenter comparison of three techniques (hemiepiphyodesis or *in situ* fusion, instrumented correction and fusion or instrumented hemivertebra resection) in 76 patients, Yaszay et al. (157) and the Harms Study Group showed that the resection group had better correction, shorter fusions, less blood loss, and shorter operative times, but a slightly higher complication rate.

Posterior-only resection is technically easier in a kyphotic deformity than a lordotic situation, since the apex of the deformity in kyphoscoliosis presents itself posteriorly (Fig. 18-23). Hemivertebra resection should include a costotransversectomy when performed in the thoracic spine for added exposure and visualization of the disks above and below the hemivertebra; furthermore, rib head excision creates flexibility in a stiff thoracic deformity that may become important in the correction (Fig. 18-24). Sacrifice of a thoracic nerve root is sometimes necessary for adequate exposure for resection or insertion of a structural cage.

Adequate resection of the hemivertebra is of paramount importance since excising the anomalous vertebra just to the midline, where the bony portion may end, without extension to the concave side with adequate release and division of a bar, for instance, will not result in sufficient correction of the curve. A “wedge” resection to the concave edge is usually necessary to achieve full correction of a single congenital deformity, which will theoretically prevent progression of secondary, longer curves in the noncongenital portion of the spine (158).

The ideal time for resection is dependent on many factors, but early resection is favorable before secondary structural curves develop and a longer fusion is necessary. Resection before age 2 poses challenges for stabilization with instrumentation, but casting is an option for external immobilization in the youngest patients. Early resection seems to make neurologic deficit less likely, as numerous reports advocate (148, 159). Neurologic complications are typically due to direct manipulation or contusion of the spinal cord or nerve root, or stretching (distraction) on the concave side with correction.

Contraindications to hemivertebra excision include spinal dysraphism at the same segment, inability to stabilize the resection with instrumentation or rigid external support, the presence of rigid deformities above or below the planned resection, which may not correct and cause spinal imbalance, and vascular anomalies associated with the CVM.

Fusion with Correction and Instrumentation. This procedure is commonly performed when correction of multiple segments in the congenital deformity and or secondary curves is desired and the principle of balancing the spine is most important. It should be noted that correction of the congenital part of the curve carries with it significant difficulty compared to idiopathic scoliosis due to the stiffness of the deformity and risk of neurologic deficit, but with meticulous technique and proper monitoring, safe correction is possible. Standing x-rays will define the stable zones, truncanal decompensation, congenital curves, and compensatory ones. With the use of bending and/or traction x-rays, we can define the flexibility of these respective deformities and then begin to formulate a plan to rebalance the spine. Choosing levels for instrumentation can be difficult, especially in scenarios when there are multiple noncontiguous anomalies (160). The entire curve must be included, but areas above or below the congenital curve may not behave in the same manner as idiopathic scoliosis. Rigid unbalanced curves may cause shoulder imbalance or truncanal decompensation and an unhappy patient. Traction x-rays may be helpful in determining how much correction will still keep reasonable balance.

The use of instrumentation allows correction and stabilization until the arthrodesis heals, and the use of titanium instrumentation allows further imaging of the spinal cord, if needed. Proper preoperative planning of the levels to be included, anatomical challenges, and instrumentation needs are paramount. The surgeon should choose a versatile system with a robust inventory of proper size implants to fit the patient and take into account the biomechanics of different rod diameters and materials when planning the correction, especially in small children.

In segmented congenital deformities, facetectomy, ligamentum flavum resection, posterior osteotomies (or resections), and anterior discectomies are useful strategies to make a long, severe, rigid curve more flexible. Patients with significant growth remaining may be candidates for anterior discectomies and fusion to obtain better rotational correction and decrease the risk of crankshaft.

The correction of severe deformities is a challenging endeavor fraught with increased neurologic risk. Osteotomies of congenital fusions or bars will aid in correction, but inherent in this arena are complications of bleeding, cord or nerve root manipulation, and frequently inadequate pedicles to obtain segmental anchorage. Whenever possible, the osteotomy should shorten the spine and rely on compression correction rather than lengthening with distraction. In the most rigid, deformed, fused, or unsegmented areas, when there is no other option, vertebral column resection (VCR) is a technique to obtain profound correction over a short segment (see below).

Osteotomy of Prior Fusion or Revision Surgery. Occasionally, patients may present with a history of previous treatment such as an *in situ* fusion, and with subsequent growth and/or progression of the deformity, they have severe, rigid curves with pain, truncanal decompensation, pelvic obliquity,

or neurologic deficit. Reconstructive surgery is indicated for these patients and may be achieved by anterior or posterior osteotomies of the previous fusion including VCR to rebalance the spine and instrumentation to restore spinal imbalance and provide the best chance for stable arthrodesis (Fig. 18-25). The basic principles are similar to the treatment of large deformities mentioned above, in which resection is effective for focal correction, anterior release and/or posterior-based osteotomies are used to correct the spine or induce flexibility over longer segments, and rebalancing the spine and fusion is performed with segmental instrumentation.

Leatherman (160a) is credited with introducing a two-stage corrective procedure for severe congenital deformities in 1969. He recognized that a posterior fusion alone in a rapidly progressing deformity may not prevent further progression, particularly in cases with unilateral bars. Furthermore, he taught the principle that in order to avoid the danger of traction paraplegia (which had resulted from Harrington distraction instrumentation around that time), the spine must be shortened as well as straightened. As such, a two-stage resection of the vertebral column was described by Leatherman and Dickson (129) in 60 patients with congenital spinal deformities for whom a two-stage corrective procedure provided excellent results. The first stage was anterior resection of the vertebral body, and the second stage was posterior resection, fusion of the curve, and instrumentation. In patients with congenital scoliosis and a mean age of 11 years (2+3 years to 16+8 years), an average correction of 47% was achieved, with no significant complications including paresis. These results are especially impressive considering that nonsegmental compression and distraction instrumentation was used and surgeries were performed without contemporary monitoring.

For the rigid, focal deformities, in either a primary or revision situation, where there is no other option, VCR can obtain dramatic correction over a short segment, but even the experienced surgeon should be prepared for excessive bleeding and assume the substantial risk of neurologic deficit. Multimodality intraoperative neurologic monitoring is mandatory since more than a quarter will have intraoperative neurologic events, and transcranial motor-evoked potentials are the most sensitive for prediction of postoperative neurologic deficit and allow prompt action intraoperatively to reduce the risk of permanent injury (161). Adequate laminectomy for visualization, stabilization with temporary working rods, undercutting the ends of the resection, and anterior structural grafting with a cage (to avoid shortening) may avert spinal subluxation and cord impingement. In a prospective, multicenter study of 147 patients who underwent VCRs for pediatric spinal deformity, the overall postoperative neurologic deficit rate was 13% (0.7% permanent), most of them at the spinal cord level, and common risk factors included kyphosis, congenital abnormalities, and revision surgery (162). The overall radiographic correction rate was excellent for kyphoscoliosis (51%) and for congenital deformity (46%), and Scoliosis Research Society (SRS) scores were significantly improved in the self-image, satisfaction domains, and total scores (163) (Fig. 18-26).



FIGURE 18-25. Osteotomy of prior fusion and revision surgery in a 10-year-old male with lumbosacral congenital scoliosis. **A,B:** PA and lateral radiographs of the boy who presented with a history of prior *in situ* fusion at age 4 with progression of the deformity, coronal and sagittal imbalance. **C,D:** CT scans showing the congenital elements and presumed pseudarthrosis.

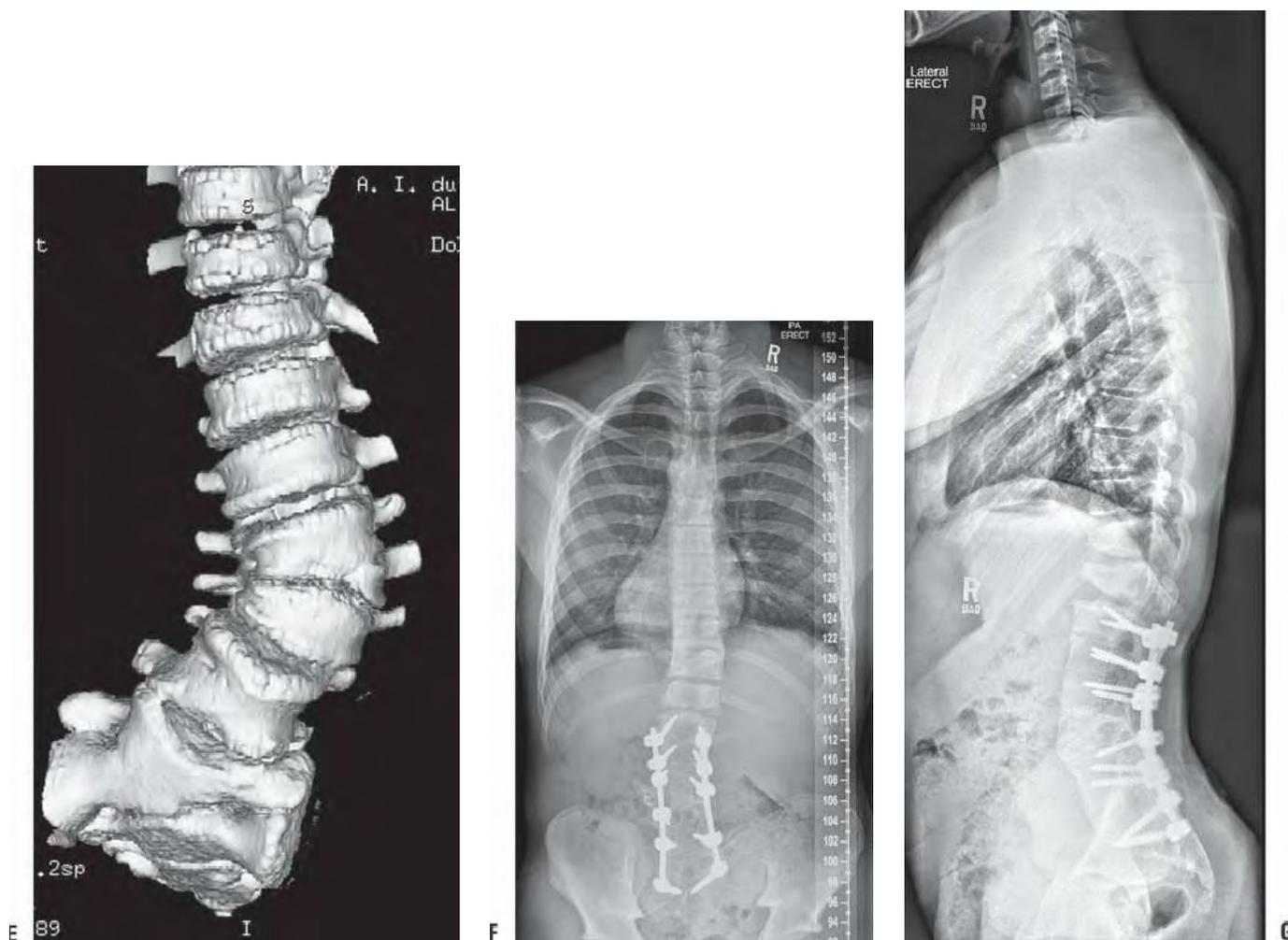


FIGURE 18-25. (Continued) **E:** CT scans showing the congenital elements and presumed pseudarthrosis. **F,G:** Postoperative radiographs 5 years after surgery showing excellent maintenance of correction and reconstruction of lumbar lordosis and sagittal balance.

Current segmental instrumentation has allowed earlier mobilization of the patient and improved correction, but has not decreased the technical complexity of the resection operation or the possible complications. In an effort to reduce the operative time and morbidity of staged or simultaneous anterior/posterior procedures, single posterior resections were devised. Suk et al.'s (164) series of posterior VCRs in an adult group of 38 congenital scoliosis patients described 63% correction of the scoliosis, but described major complications including paralysis, root injuries, fixation failures, infections, and hemopneumothoraces. Lenke's report of posterior VCR procedures for severe spinal deformities in 43 patients (heterogeneous group of adults and children) described excellent results with no permanent neurologic deficits in this challenging group of patients. He reiterated the need for spinal cord monitoring to prevent neurologic deficits since 18% patients lost intraoperative motor-evoked responses and promptly returned to baseline with surgical intervention (165).

The ability to treat severe deformities through an all-posterior vertebral resection has obviated the need for a circumferential approach in both primary and revision surgery except in special situations of lordotic deformities. Lenke et al. (166) reported on 35 consecutive pediatric patients who underwent posterior VCR of one to three levels; the average OR time was 460 minutes and average estimated blood loss (EBL) was 691 mL. Twelve patients had congenital scoliosis and had an average correction of 24 degrees (60%).

Considerable experience and skill are necessary to achieve results as described above and a stepwise progression with Ponte or Smith Peterson osteotomies, pedicle subtraction osteotomies, hemivertebra resection, and subsequently VCR is a reasonable approach. Careful preoperative planning, localization, and identification of complex anatomy are paramount to proper decision making for osteotomy and execution of the planned procedure. The authors have found 3D CT scans and even scaled models of the spine

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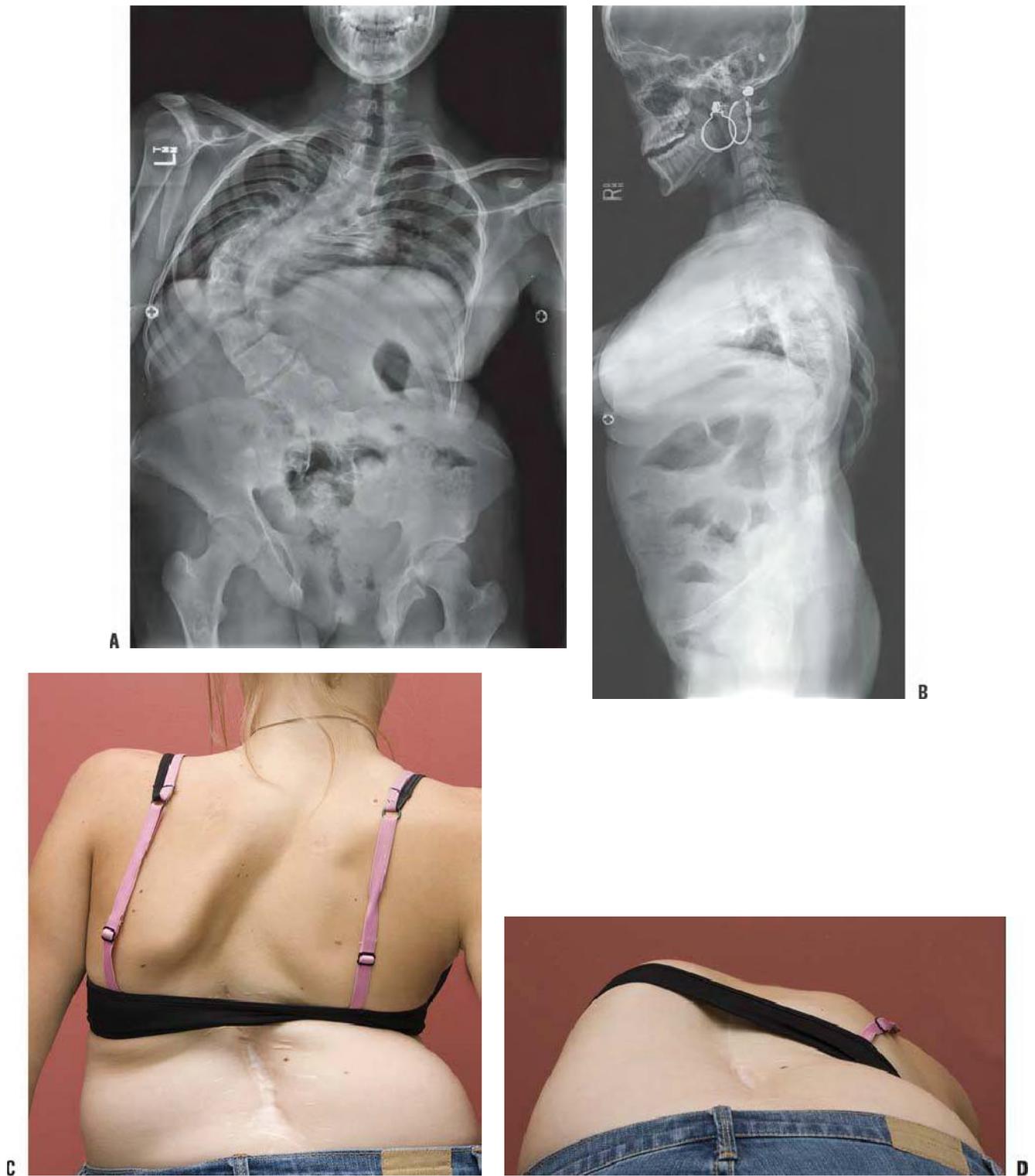


FIGURE 18-26. Multiple vertebral column resections in a 16-year-old female with congenital scoliosis and cloacal exstrophy. **A,B:** Preoperative PA and lateral radiographs showing a scoliosis of 105 degrees 11 years after a previous *in situ* fusion of the lumbo sacral area. **C,D:** Clinical photos of the patient showing severe truncal decompensation and chest wall deformity with forward bend.

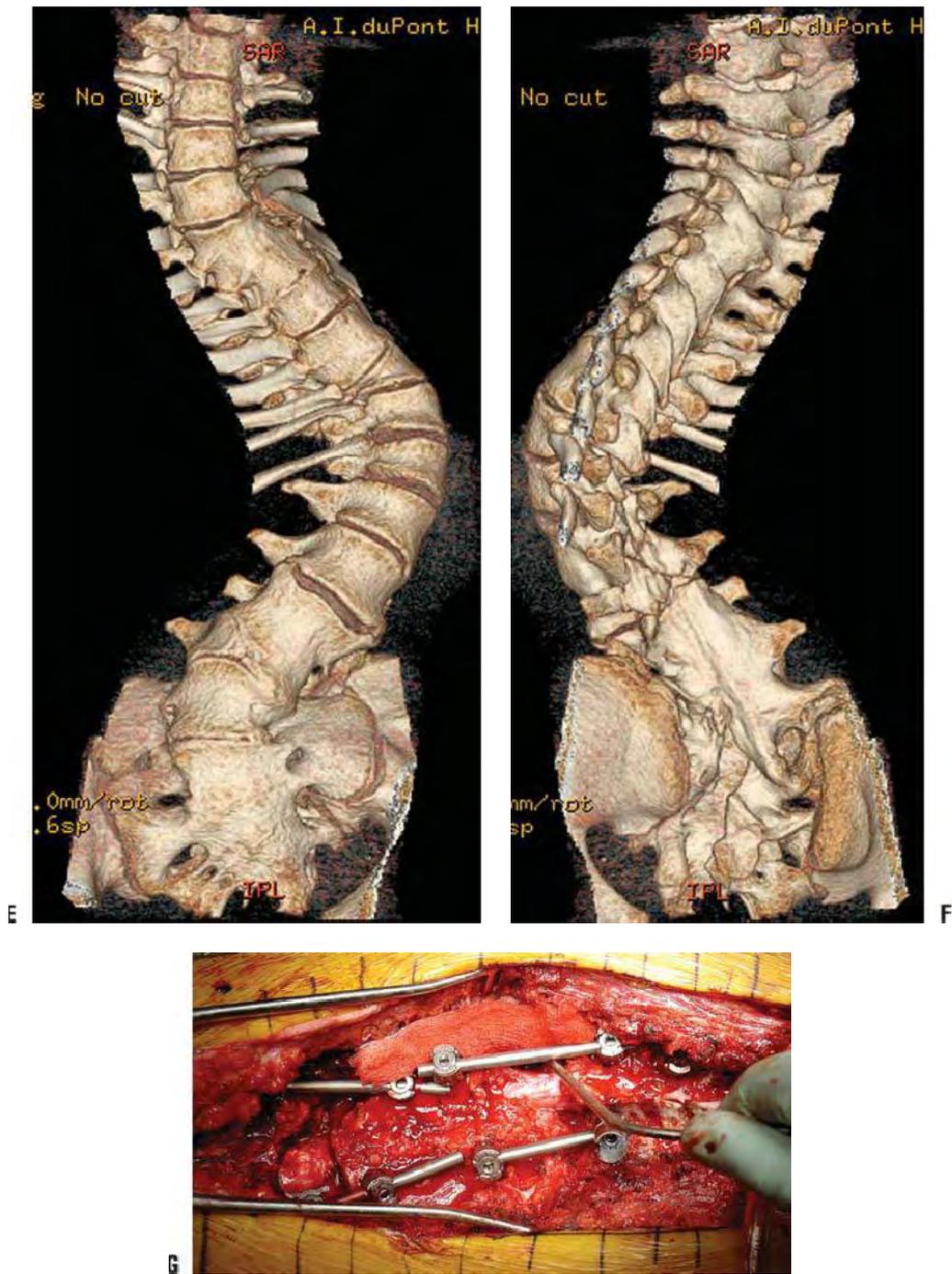


FIGURE 18-26. (Continued) **E,F:** Two perspectives of the spinal deformity illustrated by reformatted 3D CT scans. **G:** Intraoperative photo of the procedure showing two level VCR (L5 and T11) with temporary stabilizing rods prior to final correction.

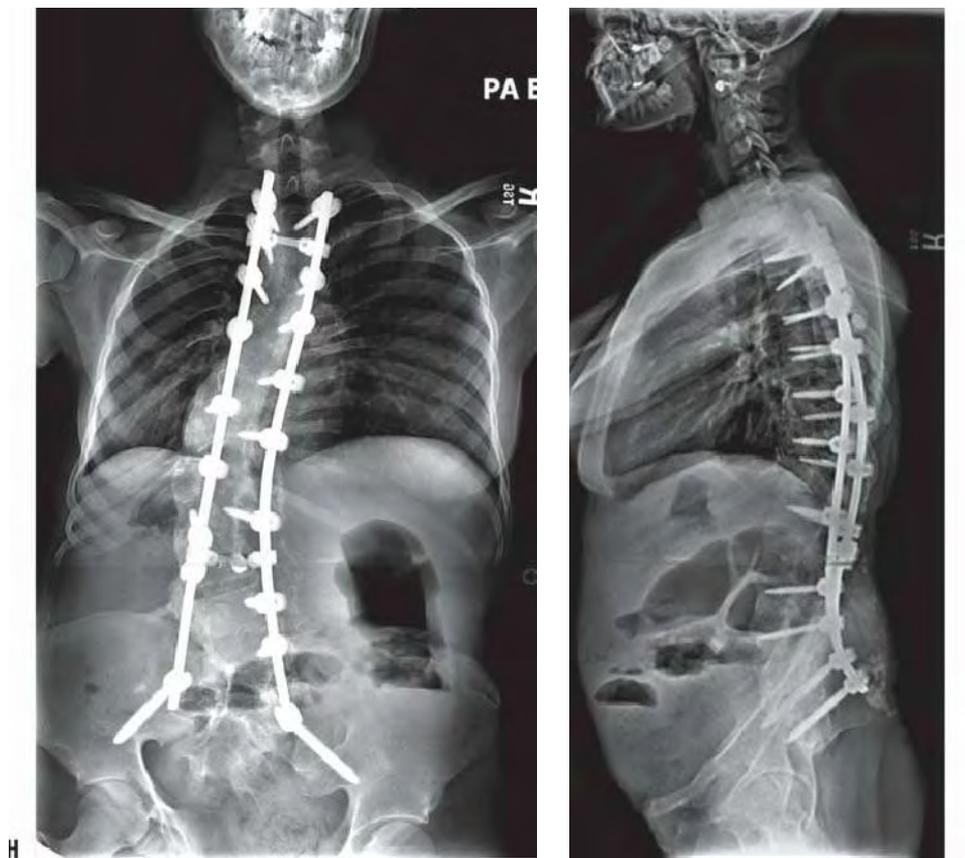


FIGURE 18-26. (Continued) **H,I:** PA and lateral radiographs 2 years after surgery.



FIGURE 18-27. Growth rods in the management of congenital scoliosis in a 6-year-old female with vertebral-anus-cardiovascular-trachea-esophagus-renal-limb-bud (VACTERL) syndrome. **A:** Erect PA radiograph showing a progressive scoliosis of 82 degrees and significant chest wall deformity. **B:** Lateral erect radiograph; note proximal thoracic kyphosis.

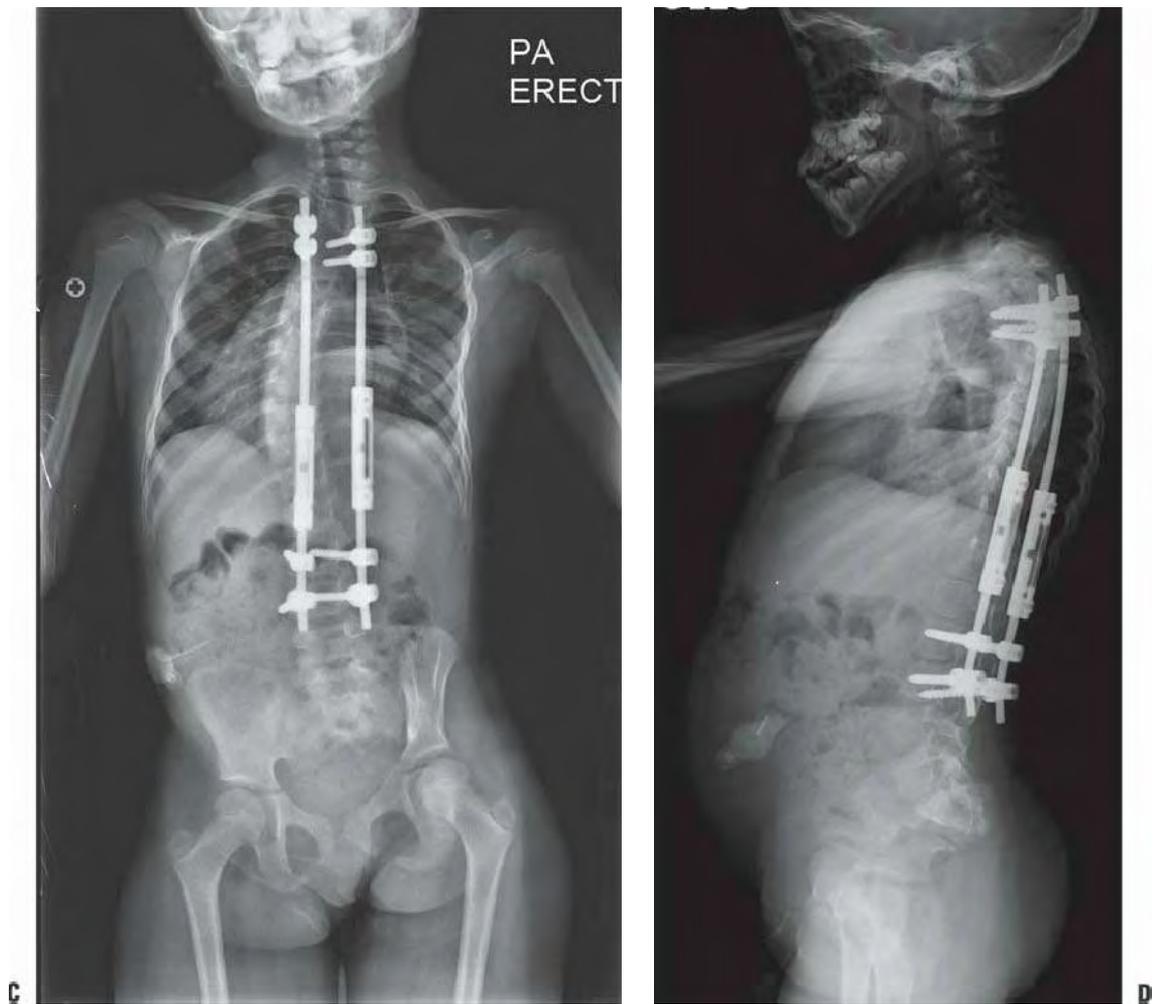


FIGURE 18-27. (Continued) **C,D:** Erect PA and lateral radiographs 18 months after dual growing rod implantation with fusion only at the proximal and distal anchor levels. The intervening tandem connectors at the thoracolumbar junction are lengthened every 6 months to maintain longitudinal spinal growth. **E:** Postoperative clinical photo of the patient.



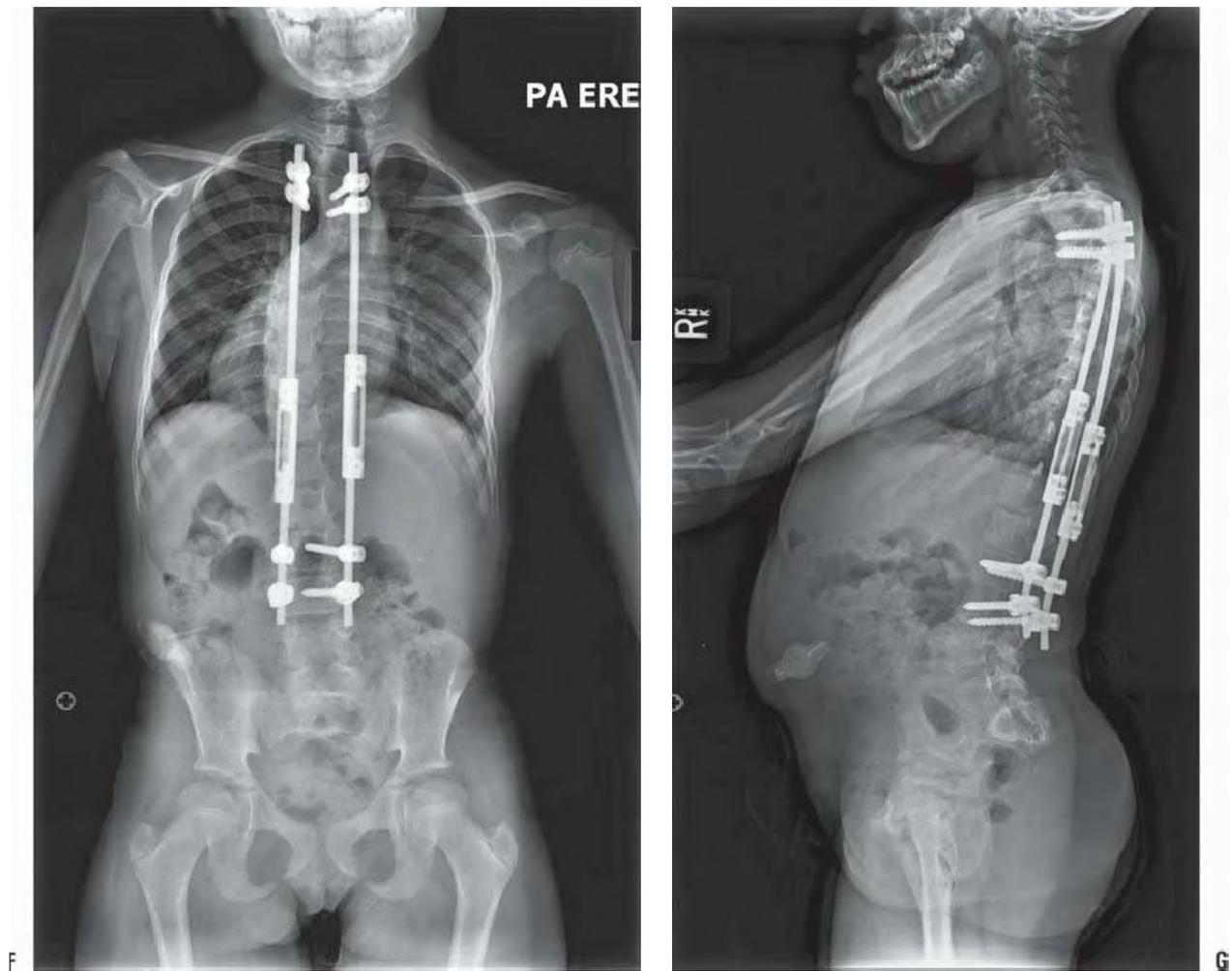


FIGURE 18-27. (Continued) **F,G:** Erect radiographs 4 years after initial implantation: six lengthenings and over 8.5 cm increase in T1-S1 height.

constructed from these scans to be very useful in resection and osteotomy procedures. Not infrequently, important information can be gleaned from this step in planning. The risk of substantial bleeding in any osteotomy is to be expected, and the risk of neurologic deficit (at the spinal cord level or nerve root compromise) is considerable. The surgeon's most trusted team should be present, and communication should be open and clear to prevent an intraoperative complication.

Growth Modulation and Future Directions. An alternative to fusion techniques is the use of growing rod techniques that correct the deformity but allow for spinal growth (Fig. 18-27). Preserving growth with control of the spinal deformity in a young child is an attractive concept that may be an option in some patients, especially when the deformity is recognized and referred early in its evolution. In very young children, long

sections of the spine involved with CVMs or compensatory curves should not be fused whenever possible, especially in the thoracic spine, to avoid a significant reduction in pulmonary function (135, 167). Congenital scoliosis is associated with short stature and decreased truncal height; combining this with a multilevel spinal arthrodesis can have a further detrimental effect on spinal growth, truncal height, and pulmonary function, leading to TIS (134). The growth of the spine is the greatest in the first 5 years of life, and sitting height is about 60% of that of an adult by age 5 years (115), so growth should be encouraged in this period without sacrificing control of the curve. The use of interval lengthening of posterior spinal instrumentation without fusion, the “growing-rod” technique, has a role in the management of early-onset scoliosis, including congenital scoliosis (145) without rib fusions. The growing rod technique is effective and reliable for treating congenital deformities in growing children if the CVM

segments are somewhat flexible, the anomalous segment is too large for resection, or other definitive treatment or is accompanied by a long, structural compensatory curve. The most commonly used dual rod systems have been described by Akbarnia and Thompson with moderate success; modest gains in spinal length of 3 to 7 cm (1.2 cm per year); improvement in SAL; and an acceptable complication rate of superficial infections, dislodged implants, and broken rods (168, 138). The technique involves anchorage at the proximal and distal stable zones with hooks or screws after subperiosteal dissection; fusion is desirable in these small areas away from the apex for anchor stability. Rods are then tunneled submuscularly joined by tandem connectors placed at the thoracolumbar junction, where the interval distraction is performed for lengthening the spine. An apical fusion, excision, or osteotomy for the congenital portion of the curve can be performed and then growing rods implanted to lengthen the spine and accommodate growth in the compensatory curve(s) (169). Apical fusions in patients with single-rod growing systems do create challenges such as increased force to lengthen and smaller gains (170, 171) as well as crankshaft, but with dual rods these difficulties may be obviated. The vertical expandable prosthetic titanium rib (VEPTR) device is indicated for use in patients with congenital scoliosis and fused ribs to address thoracic hypoplasia and prevent TIS when a large amount of growth remains (172) (see below). Unfortunately, both of these approaches require a scheduled operative lengthening recommended at 6-month intervals and ultimately are treated with a spinal fusion in most cases when no further lengthening can be achieved or the accelerative phase of growth is over. An alternative is a growth guidance system (Shilla) that corrects the scoliotic apex of the curve with a limited fusion and allows for continued guided growth at the proximal and distal portions of the spine, avoiding the need for routine operative lengthenings. Clinical data are limited, but McCarthy et al. (173) caprine animal model showed an average of 48 mm of growth at the ends of the rods near the sliding screws over 6 months. There were no implant failures, but minor wear debris was observed.

VEPTR (VERTICAL EXPANDABLE PROSTHETIC TITANIUM RIB)

The VEPTR was developed by Campbell (174) for the management of TIS. This heterogeneous group of conditions is defined as those in which the function of the thorax is insufficient to support normal growth and development. These conditions may be naturally occurring or can be iatrogenic. All forms of early onset scoliosis, many skeletal dysplasias, and congenital or acquired chest wall deformities including early spinal fusions are included. VEPTR devices currently are used under a humanitarian device exemption status by the FDA after undergoing an IDE trial in the early 2000s. This status requires institutions using the device to have an active institutional review board approval and restricts its use to neuro-

muscular scoliosis conditions, congenital scoliosis, constricted hemithorax conditions such as fused ribs or costal dysplasias, and acquired severe chest wall deficiencies.

Historically, it was believed that growth of vertebral segments within a region of congenital malformations did not occur. As such, treatments focused on short-segment fusions and convex hemiepiphysiodesis that would include segments outside of the malformed segment in an effort to gain compensatory correction. The notion that guided or forced growth management of congenital scoliosis deformities could not be done in congenital abnormalities was challenged by Campbell et al. (175) with their use of rib-based systems using the VEPTR (Fig. 18-28). In an uncontrolled and nonselective group of patients with congenital scoliosis with and without rib fusions, application of rib-to-rib- or rib-to-spine-based distraction devices with an expansion thoracoplasty (174) led to elongation of the concave side of the deformity by an average of 8 mm/year and of the convex side by 8.3 mm/year over mean follow-up of 4 years. Stabilization of the spinal deformity and secondary overall increase in lung volumes has been reported (175), leading many to believe that the combination of congenital spinal malformations and rib fusions is the ideal situation under which to use VEPTR devices (176–182).

The technique of VEPTR for congenital scoliosis and fused ribs has been described in detail by Campbell et al. (174, 175). The key components are an opening wedge thoracostomy through the fused rib segments, a lateral rib-to-rib-based strut, and a medial rib-to-rib- or rib-to-spine-based hybrid construct (Fig. 18-28A–D). In the absence of fused ribs, the role of thoracostomy has increasingly come into question due to concerns about altered chest wall compliance with the scarring created by the procedure. Rib anchors are recommended to be just lateral to the rib heads and avoidance of anchors above the 2nd rib is recommended due to the risk of brachial plexus injuries by proximal migration of the anchors (176, 183). Subperiosteal dissection of the ribs is done for placement of the rib cradles, and with the introduction of VEPTR II, multiple anchor points and ribs can now be used. Distal spine anchors are designed to be via laminar hooks. The off-label use of pedicle screw anchors has been introduced with recommendations to have at least two distal anchors to decrease the incidence of screw migration through the pedicles. We have found that use of a ventriculoperitoneal shunt passer and chest tube facilitates submuscular passage of the hybrid rod for cases that will have a distal spine anchor. No standard exists for the frequency of expansions. Borrowing from the experience of “growing rod” systems, recommendations have been for intervals of 6 months to optimize spinal growth (178, 184).

The incidence of complications related to the use of VEPTR implants has paralleled that of other growth preservation systems. Anchor migration, implant failure, wound infections, and loss of control of the deformity have all been reported (185–187).

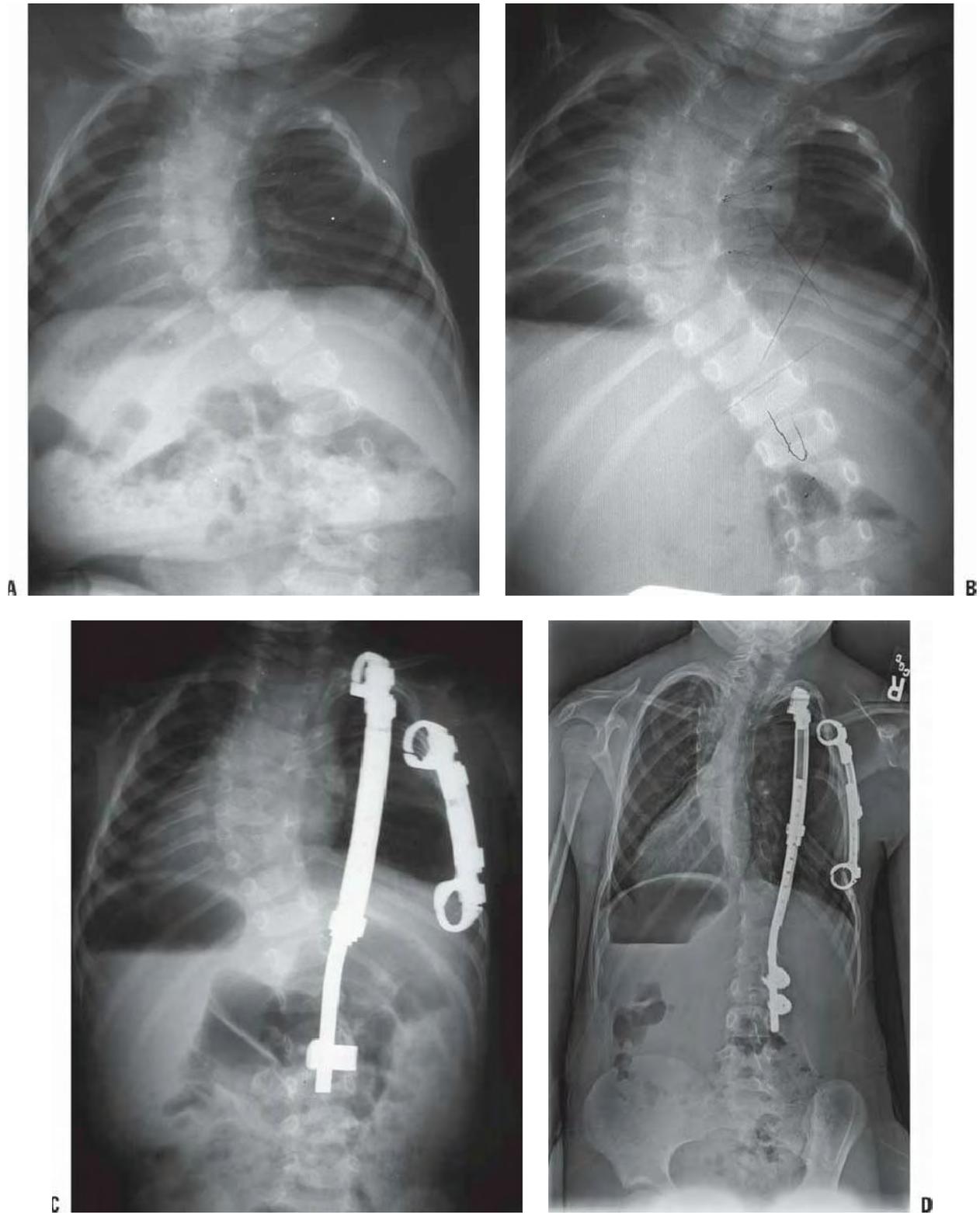
VEPTR (Vertical Expandable Prosthetic Titanium Rib) (Fig. 18-28)

FIGURE 18-28. Congenital scoliosis with multiple fused ribs **A:** 6-month-old female. **B:** Age 20 months. **C:** VEPTR implantation age; 23 months **D:** Age 11 years.

CONCLUSIONS

The treatment of congenital scoliosis is among the most interesting to the surgeon treating spinal deformities and focuses on early diagnosis and surgical treatment before large curves develop. The authors cannot emphasize enough the importance of the observational period of the treatment of patients with congenital scoliosis, namely giving thought to the evolution of the deformity with growth and acting at the appropriate time if surgery is necessary. Vertebral anomalies with a progressive natural history need to be managed aggressively. Outcomes are generally better with simple procedures with a favorable complication profile. *In situ* fusion or hemiepiphysiodesis are viable options if there is little to no deformity present, but the potential for progression is high. For moderate deformities, correction with instrumentation and arthrodesis will provide satisfactory results. Young children with an isolated hemivertebra are excellent candidates for resection if progression is noted; this keeps the area of arthrodesis small and avoids the development of compensatory curves. Extensive spinal arthrodesis should be avoided, especially before age 5, and growing rods can be used in children with long segmented areas of the spine to modulate growth and control the curve. TIS and congenital scoliosis with fused ribs seems to be best managed by the VEPTR device and expansion thoracostomy. For severe, rigid deformities in older children, osteotomies or resections to rebalance the spine with attention to the more structural compensatory curves may be needed. All too often, in a referral practice, we have seen missed opportunities in which a simpler procedure could have been performed, saving the patient and family from a much larger, complicated procedure and protracted recovery.

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