

- Describe the different forms of scoliosis
- Discuss the patient history and physical assessment of scoliosis
- Review the evaluation of scoliosis by radiographic and other imaging studies
- Explain the therapeutic options, including observation, bracing, and surgical intervention

Scoliosis: A straightforward approach to diagnosis and management

Scoliosis should be no more intimidating to manage than acute otitis media. This article reviews how to treat patients with the condition before and after referral to a specialist.

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Scoliosis management is a fundamental component of pediatric evaluation and care. All PAs who treat pediatric patients should feel comfortable approaching this condition, just as one would treating otitis media or strep throat. The essential steps are to properly identify children at risk, stabilize the driving forces, and enable a healthy, play-filled childhood. Scoliosis has been a treatable condition for more than 200 years; and although treatment is somewhat routine, significant advances in modalities have recently been made. In this way, scoliosis truly remains a classic problem with ever-evolving solutions.

Traditionally, scoliosis is described as an “S” curve; however, it is more than a two-dimensional condition. In someone with scoliosis, the spine actually becomes more of a helical shape because of dramatic changes in kyphosis and lordosis, creating a complex spinal deformity and treatment paradigm. This three-dimensional aspect of the scoliotic spine was not recognized until the 1990s.¹ The forces that twist the spine into these contorted positions are derived from imbalanced growth. Although the initiating causes are unknown, hereditary factors are likely to be involved.

Scoliosis is classified into three forms: congenital, neuromuscular, or idiopathic. This discussion will focus on idiopathic scoliosis (IS). This condition is labeled according to age of onset: Infantile (IIS), birth to 3 years; juvenile (JIS), 4 to 9 years; and adolescent (AIS), 10 years to adulthood. Most IS cases (approximately 85%) are AIS; of the remaining cases, 1% are IIS and up to 15% are JIS. Gender predominance changes with ascending age. The ratio moves from 3:2 male predominance in infants to 3:1 female predominance in children to an equal distribution in adoles-



cents.^{2,4} Although it appears that there may be a higher incidence of scoliosis in female adolescents, such discrepancy actually reflects a higher propensity for progression in girls.²

PHYSICAL EXAMINATION

A thorough patient history plays an important role in prompting more intense, frequent screenings and guiding treatment. Patients with visceral anomalies, limb deformities, or a family history of scoliosis must be monitored closely for onset. Recent growth surges, perimenarchal age, or prior thoracotomy may require heightened assessment of established curves.² Spinal pain is an unusual complaint for children. If pain is reported, a workup for neoplastic or neurologic causes may be indicated,⁵ especially if there are bowel and bladder complaints or the pain awakens the child from sleep during the night.

Infants Evaluation of the infant or toddler is particularly challenging because of obvious communication boundaries and cooperation issues. Most spinal curves will have formed prior to the onset of walking, which may make postural asymmetries less apparent. A thorough examination of the skin should be performed. Clues to other neurologic abnormalities include café au lait spots (more than six suggests neurofibromatosis type I), sacral dimples, or hairy patches (a potential harbinger of a tethered cord). The orientation of the shoulder girdles and their respective relationship to the pelvis can help reveal skeletal asymmetries. Examination of the head and face may reveal areas of flattening or plagiocephaly, which occurs in 80% to 100% of cases.³ A deformity of the head is often on the right side because most infantile curves are apex left.³ Neurologic examination is very difficult to obtain systematically; most information is derived from gross motor movements. Special attention should be paid to the abdominal reflex and ocular movements because they can indicate the presence of CNS abnormalities. Hip dislocations are 5 to 10 times more likely to occur in these patients; therefore, a complete hip examination, including Ortolani and Barlow maneuvers, must be performed.³

Children and adolescents Evaluation of these patients also begins with a detailed inspection of the skin. The shoulder and pelvic heights should be examined for obliq-



FIGURE 1. Adams forward-bending test

uities with the patient standing with shoes removed and heels approximated. From this position, have the patient bend forward with hands together in a “diving” form (see Figure 1). Prominent paraspinal musculature may be noted in the lumbar spine, or rib humps may be seen in the thoracic spine. An examiner’s eye may be fairly sensitive to abnormalities in spinal contour; however, a scoliometer, which can detect an abnormality of even a few degrees, should be used. A scoliometer reading of 7° to 10° is suggestive of a curve of approximately 20° on radiograph.⁶ Full motor, sensory, and reflexive tests also should be performed on the extremities. As in infants, abdominal and ocular responses can indicate the presence of cord and brainstem lesions. An absent gag reflex or a cavus foot⁵ may indicate CNS abnormalities as well. If an obliquity is noted in standing, the leg lengths should be checked to rule out a lower extremity problem.

RADIOGRAPHIC ASSESSMENT

Posteroanterior and lateral plain film views of the entire spine should be obtained on 14×36-inch films with the patient standing with shoes removed and any leg length dis-

KEY POINTS

- Traditionally, scoliosis is described as an “S” curve; however, the spine actually becomes more of a helical shape because of dramatic changes in kyphosis and lordosis.
- Patients with visceral anomalies, limb deformities, or a family history of scoliosis must be monitored closely for onset. Recent growth surges, perimenarchal age, or prior thoracotomy may require heightened assessment of established curves.
- The decision to treat a patient is conventionally based on the size of the curve, as determined by Cobb-angle measurements. Twenty degrees and 40° are considered indicators for bracing and surgery, respectively.
- Fusion is the ultimate and definitive treatment for progressive scoliosis; however, it is generally not performed until the patient is near skeletal maturity to ensure that maximum spinal height is achieved, lung development is optimized, and growing forces are quieted.

COMPETENCIES

- Medical knowledge
- Interpersonal & communication skills
- Patient care
- Professionalism
- Practice-based learning and improvement
- Systems-based practice

crepancy eliminated with blocks. Standard Cobb-angle measurements of both main and compensatory curves should be taken. The measurements are derived from the most angled vertebrae, using the vertebral endplates as reference points (see Figure 2). Also make note of the triradiate cartilages and iliac apophyses (Risser grade) to estimate remaining growth potential.

Rib-vertebral angle difference (RVAD) measurements should be taken in infant and juvenile patients to ascertain curve-progression risk. This relationship is referenced from the endplate of the apical vertebra, instead of the transitional segments that are used in the Cobb measurement. The angles created by the ribs at the apical spine segment on both sides are compared (see Figure 3). Greater differences in the angles have been shown to relate to higher rates of progression.^{3,7}

The Lenke system of classification for AIS was developed in 2001 to help identify curve types, assess risk for progression, and establish surgical guidelines.⁸ The system provides a consistent tool for clinicians to determine the overall curve profile. An in-depth review of the Lenke system is beyond the scope of this discussion. Essentially, the major, or largest,

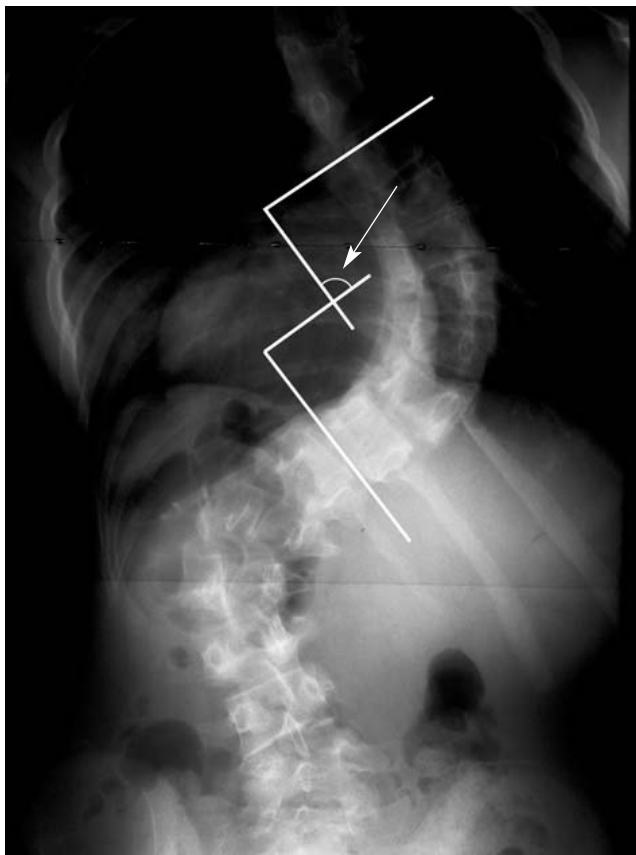


FIGURE 2. The Cobb angle (arrow) is determined by drawing lines from the most tilted vertebrae above and below the apex of the curve and then drawing intersecting lines perpendicular to the lines at the top and bottom vertebrae.

curve is identified, and the curve is designated as thoracic, thoracolumbar, or lumbar. In addition, the amount of kyphosis, the flexibility of each curve, and the severity of the lumbar curve are assessed to help determine the appropriate treatment.

EXTENDED EVALUATION

After completing the history and the physical and radiologic examinations, the clinician must determine whether more diagnostic testing is needed. Certain examination findings demand further investigation. For instance, scoliosis at a young age (IIS) is unusual and should automatically raise concern for neurologic anomalies.⁵ Approximately 20% of children younger than 10 years who present with scoliosis also have neural axis abnormalities.⁹ In addition, rapid progression, axial skeletal pain, left convex thoracic curves, or double major curves can be indicators of a neurologic abnormality.^{5,9} Any of these factors or an abnormal neurologic examination should prompt referral to a pediatric center for a noncontrast MRI of the entire spine; depending on the patient's age, the MRI may be obtained under sedation.⁵ Arnold-Chiari malformation, cervical or thoracic syrinx (syringomyelia), and tethered cord are diagnoses that must be ruled out.

TREATMENT HIERARCHY

The decision to treat a patient is conventionally based on the size of the curve, as determined by Cobb-angle measurements. Twenty degrees and 40° are considered indicators for bracing and surgery, respectively. Those triggers to treat are more appropriate for the adolescent population; newer evaluation and surgical options have changed the algorithm for the younger age-groups.

Observation Curvature of the spine is not considered scoliosis until the curve is more than 10°. Physiologic curves, as well as most scoliotic curves less than 25°, should be closely observed with serial radiographs. In 80% of infants with an RVAD less than 20°, the angle difference will spontaneously resolve and the patient should simply be observed.^{3,7} In patients with JIS, the RVAD should be less than 10°. Evaluation and plain radiographs should be performed every 4 to 6 months until the curve is determined to be stable. If it is stable, follow-up evaluation every 6 to 12 months is adequate. Growth spurts, onset of puberty, scoliometer measurement changes, or observable anatomic changes should prompt earlier follow-up.² If a curve surpasses 20°, consider referral to a specialist.

Bracing The primary mode of treatment for progressive scoliosis in the absence of more suitable alternatives is bracing. Molded thoracolumbar-sacral orthosis braces utilize the concept of three-point fixation to counteract the deforming forces on the spine, thereby limiting curve progression. The brace must be worn for 16 to 23 hours per day in order to be effective;¹⁰ but even with an optimal wear schedule, the benefit may be modest at best. Comparison studies of observation and bracing modalities showed no change in the prob-

ability of the need for surgical intervention.¹¹ In addition, low self-esteem, lifestyle changes, and poor adherence have been reported in braced patients.¹² If bracing or casting is determined to be the appropriate treatment, then it should be utilized in patients with IIS when the RVAD is more than 20°, which indicates an 80% risk of curve progression;^{3,7} in patients with JIS when the RVAD is more than 10°, a reported 100% risk of curve progression;⁴ and in patients with AIS when the Cobb angle is more than 20°.²

Surgery Many surgical options are available, with both anterior and posterior approaches. Most patients typically require a posterior spinal fusion with pedicle screws and rods (see Figure 4). The development of pedicle screws and contoured rods has revolutionized this procedure in the past 15 to 20 years; marked control over the anterior spinal column is achieved, which was not possible with previous techniques.^{1,13} The number of levels fused is patient dependent, but frequently the fusion spans the upper thoracic to middle lumbar levels. Surgical correction may include concurrent osteotomies to release bony tethers.

Spinal fusion is the treatment of choice for patients with large, progressive curvatures of more than 50°, and may be considered for children as young as 8 years.^{2,3} Without surgery, curves of more than 50° have a high likelihood of progressing 1° per year throughout adulthood.¹⁴ A recent study demonstrated that curve progression is linear and can be used to determine individual risk of progression across all curvatures.¹⁵ In the future, genetic testing may become the most accurate prognostic indicator.

NEW TREATMENTS

In the past 5 years, improvements in the devices used to correct curves have focused on the challenges of the growing spine. Fusion is the ultimate and definitive treatment for progressive scoliosis; however, it is generally not performed until the patient is near skeletal maturity to ensure that maximum spinal height is achieved, lung development is optimized, and growing forces are quieted. New minimally invasive techniques help to preserve the native spine in anticipation of eventual fusion and assist in guiding the growth of the maturing spine to theoretically provide some level of self-correction. The vertical expandable prosthetic titanium rib (VEPTR) device and vertebral staples are innovations that can help resolve poor interim stabilization.

VEPTR demonstrated early success and has evolved into a more versatile variation (the VEPTR II) in 2008. The device is an expandable, subcutaneous rod that provides various distraction forces to the trunk. VEPTR can be used to drive the pelvis down, elevate the shoulder, or expand the rib cage to correct the spinal deformity and preserve unimpeded growth. Multiple possible fixation points allow the VEPTR to span from rib to rib, rib to spine, or rib to pelvis¹⁶ (see Figure 5, page 44).

Insertion of the device requires a small incision over each of the two proposed attachment sites. A tract is developed in the adipomuscular tissue plane between the two incisions. If

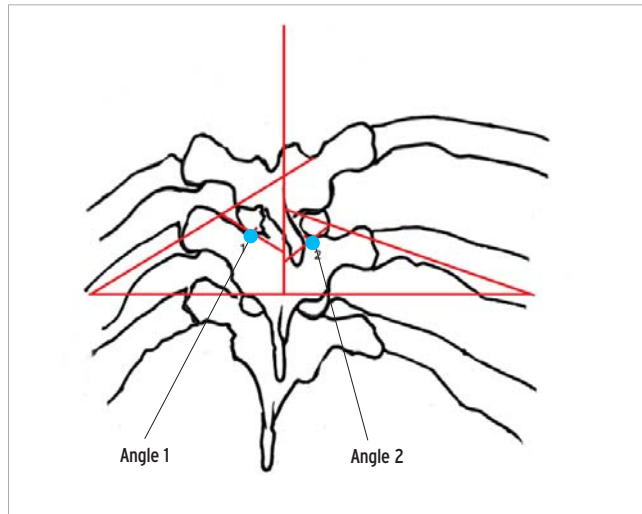


FIGURE 3. The rib-vertebral angle difference (RVAD) is determined by drawing a line along the inferior border of the apical vertebra and then drawing a second line perpendicular to the first. Connect the lines by going through the midpoints of the body of the ribs on either side of the apical vertebra, and extend them to meet the perpendicular line. The RVAD is angle 2 minus angle 1.

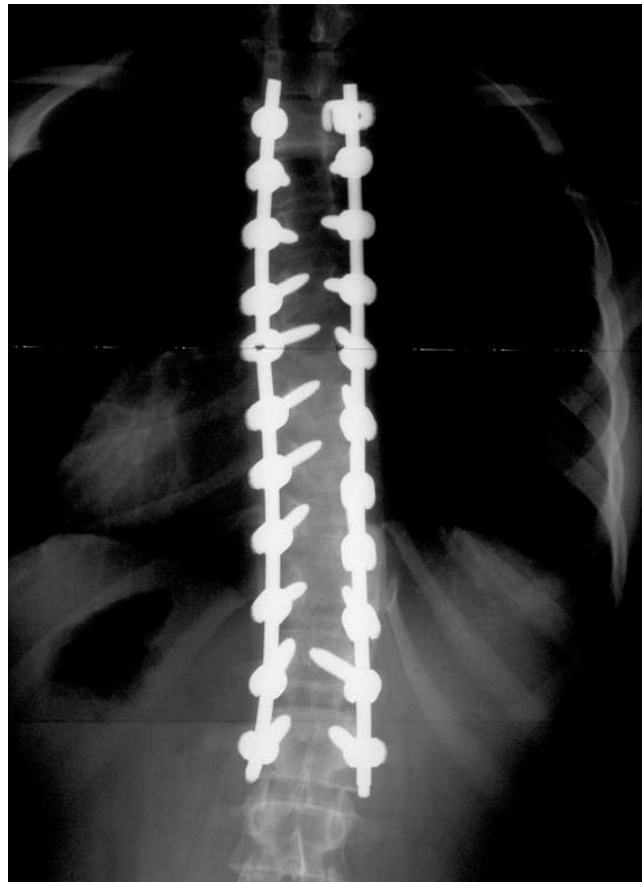


FIGURE 4. Spinal fusion with pedicle screws and contoured rod



FIGURE 5. Vertical expandable prosthetic titanium rib device spanning from rib to pelvis

necessary, an opening wedge thoracostomy is performed through the proximal incision site to provide chest wall excursion during correction. Once fixed, the VEPTR is ratcheted out to the appropriate length. Lengthening the device to keep in stride with changes in growth is a short surgical procedure performed every 4 to 6 months.¹⁶

Vertebral stapling to treat scoliosis was first investigated in the 1950s,¹⁷ but the results were dismal because of hardware and fixation failure. Newer metal materials now make this procedure possible. Nitinol, a melding of nickel and titanium, has a unique structural memory that allows it to be straight at room temperature and curve at body temperature; this enables the blades of the staple to clamp down on the bone once inserted, making the correction and fixation more stable.¹⁸

The surgical technique involves making multiple small thoracotomies over the apex of the thoracic curve, deflating the lung, and using thoracoscopy with fluoroscopy to identify the appropriate levels for stapling. The staple is introduced into opposing vertebral endplates to create a tethering effect between the two (see Figure 6). The goal is to limit growth

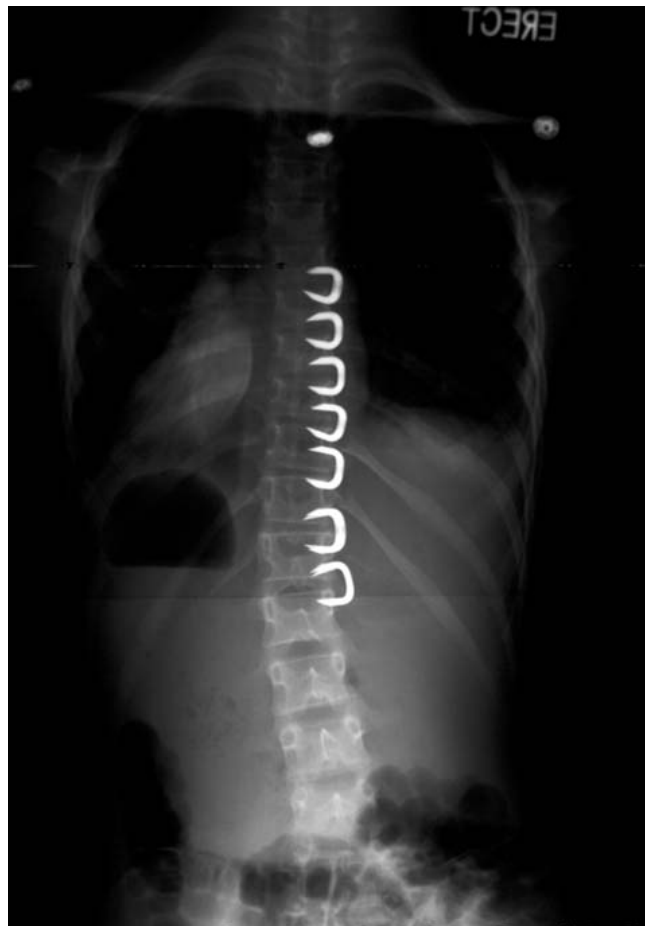


FIGURE 6. Vertebral staples inserted to limit growth on the convex side of a curve.

on the convex side and allow growth on the concave side, ultimately restricting progression of the curve and inciting correction. The lumbar curve is often corrected during the same procedure; the staples are introduced contralaterally through a small retroperitoneal approach.^{18,19}

Appropriate patients for stapling include those aged 4 to 14 years, those with curves from 20° to 45°, and those with curves less than 25° that have progressed 5° in 6 months.¹⁹ Stapling is contraindicated in patients whose vertebrae are too small, patients with an associated kyphosis more than 40°, or patients with curves that extend above T2 or below L4.¹⁹ Early data suggests that no progression is seen in stapled curves less than 30° and an 87% success rate is seen in curves less than 50° in patients older than 8 years.¹⁹

LIFE AFTER FUSION

Although surgery conclusively eliminates deformity, successful treatment of the scoliotic spine does not always end with fusion. These patients may need periodic surveillance and are likely to present frequently to the clinic, emergency department, or operating room.

Postoperative period Many patients have their fusion procedure performed by a specialist at a major medical center a long distance from home and, when they return, you will have to treat the postoperative conditions that can develop. One such condition, called *superior mesenteric artery (SMA) syndrome*, is the compression of the duodenum between the aorta and superior mesenteric artery where it branches off from the aorta. The anatomic alignment of the vessels forms a “nutcracker” configuration around the duodenum. Decreased fat stores around the bowel, particularly in thin patients, allow increased compression directly on the duodenum. Tension created from the surgical correction may contribute to triggering the sudden onset of an upper GI obstruction characterized by epigastric pain, distention, and bilious vomiting. SMA syndrome usually occurs within the first 5 to 7 postoperative days but may not develop until as late as a few weeks after surgery. Affected patients cannot take anything by mouth; treatment includes antiemetics and IV fluid repletion, placement of a nasogastric tube, and possible nasojejunum tube feeds for a few days to weeks.^{20,21}

Pregnancy A common concern for many patients after spinal fusion is pregnancy. Patients should know that there is little impact on the experience as a whole. Fused and unfused portions of a scoliotic spine have been shown to remain unchanged throughout pregnancy.²² Up to 40% of patients with a fusion may experience low back pain associated with the pregnancy but, for the vast majority, back pain is mild to moderate and has no functional impact.²² Cesarean deliveries may be performed slightly more often on patients with instrumented fusions, but no additional complications associated with labor and delivery have been found.²² However, administering epidural anesthesia is more difficult. Multiple attempts, dural punctures, subdural catheterization, and poor or failed anesthesia have been consistently reported.²³

Long-term prognosis Prior surgical techniques, such as Harrington rod fixation, successfully corrected the coronal plane, but the sagittal plane was neglected. Many patients developed *flat-back syndrome*. The spine was literally made rigidly flat, which caused problems with balancing the spine over the pelvis, and the body was forced to compensate. The extensor musculature was stressed and the lower lumbar levels hyperextended, leading to rapid onset of degenerative disk disease and subsequent low back pain. Current techniques are able to maintain natural anatomic relations and hopefully decrease the likelihood of aggressive spondylosis.¹

Scoliosis can have a significant impact on a patient’s quality of life and lung function. A balanced spine through instrumented fusion can biomechanically improve general activities of daily living by centralizing posture, normalizing gait, and improving overall endurance. Once the spine is brought back to midline, the ribs are elevated causing a relative expansion of the lungs and increase in pulmonary function.²⁴ For many, this is a worthy tradeoff for some loss of overall trunk flexibility. **JAAPA**

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REFERENCES

- de Jonge T, Dubouset JF, Illés T. Sagittal plane correction in idiopathic scoliosis. *Spine*. 2002;27(7):754-760.
- Lonstein JE. Scoliosis: surgical versus nonsurgical treatment. *Clin Orthop Relat Res*. 2006;443:248-259.
- Lincoln TL. Infantile idiopathic scoliosis. *Am J Orthop*. 2007;36(11):586-590.
- Mannherz RE, Betz RR, Clancy M, Steel HH. Juvenile idiopathic scoliosis followed to skeletal maturity. *Spine*. 1988;13(10):1087-1090.
- Schwend RM, Hennrijus W, Hall JE, Emans JB. Childhood scoliosis: clinical indications for magnetic resonance imaging. *J Bone Joint Surg Am*. 1995;77(1):46-53.
- Korovessis PG, Stamatakis MV. Prediction of scoliotic Cobb angle with the use of the scoliometer. *Spine*. 1996;21(14):1661-1666.
- Mehta MH. The rib-vertebra angle in the early diagnosis between resolving and progressive infantile scoliosis. *J Bone Joint Surg Br*. 1972;54(2):230-243.
- Lenke LG, Betz RR, Harms J, et al. Adolescent idiopathic scoliosis: a new classification to determine extent of spinal arthrodesis. *J Bone Joint Surg Am*. 2001;83-A(8):1169-1181.
- Gupta P, Lenke LG, Bridwell KH. Incidence of neural axis abnormalities in infantile and juvenile patients with spinal deformity. Is a magnetic resonance image screening necessary? *Spine*. 1998;23(2):206-210.
- Allington NJ, Bowen JR. Adolescent idiopathic scoliosis: treatment with the Wilmington brace. A comparison of full-time and part-time use. *J Bone Joint Surg Am*. 1996;78(7):1056-1062.
- Dolan LA, Weinstein SL. Surgical rates after observation and bracing for adolescent idiopathic scoliosis: an evidence-based review. *Spine*. 2007;32(19 Suppl):S91-S100.
- Andersen MØ, Andersen GR, Thomsen K, Christensen SB. Early weaning might reduce the psychological strain of Boston bracing: a study of 136 patients with adolescent idiopathic scoliosis at 3.5 years after termination of brace treatment. *J Pediatr Orthop B*. 2002;11(2):96-99.
- Zeiller SC, Lee J, Lim M, Vaccaro AR. Posterior thoracic segmental pedicle screw instrumentation: evolving methods of safe and effective placement. *Neurol India*. 2005;53(4):458-465.
- Weinstein SL, Ponseti IV. Curve progression in idiopathic scoliosis. *J Bone Joint Surg Am*. 1983;65(4):447-455.
- Marty-Poumarat C, Scattin L, Marpeau M, et al. Natural history of progressive adult scoliosis. *Spine*. 2007;32(11):1227-1234.
- Campbell RM Jr, Smith MD, Hell-Vocke AK. Expansion thoracoplasty: the surgical technique of opening-wedge thoracostomy: surgical technique. *J Bone Joint Surg Am*. 2004;86-A(suppl 1):51-64.
- Smith AD, von Läckum WH, Wylie R. An operation for stapling vertebral bodies in congenital scoliosis. *J Bone Joint Surg Am*. 1954;36(A:2):342-348.
- Betz RR, Kim J, D’Andrea LP, et al. An innovative technique of vertebral body stapling for the treatment of patients with adolescent idiopathic scoliosis: a feasibility, safety, and utility study. *Spine*. 2003;28(20):S255-S265.
- Betz RR, D’Andrea LP, Mulcahey MJ, Chafetz RS. Vertebral body stapling procedure for the treatment of scoliosis in the growing child. *Clin Orthop Relat Res*. 2005;(434):55-60.
- Hod-Feins R, Copeliovitch L, Abu-Kishk I, et al. Superior mesenteric artery syndrome after scoliosis repair surgery: a case study and reassessment of the syndrome’s pathogenesis. *J Pediatr Orthop B*. 2007;16(5):345-349.
- Tsirikos AI, Anakwe RE, Baker AD. Late presentation of superior mesenteric artery syndrome following scoliosis surgery: a case report. *J Med Case Reports*. 2008;2:9.
- Orvoma E, Hillesmaa V, Poussa M, et al. Pregnancy and delivery in patients operated by the Harrington method for idiopathic scoliosis. *Eur Spine J*. 1997;6(5):304-307.
- Ho AM, Ngan Kee WD, Chung DC. Should laboring parturients with Harrington rods receive lumbar epidural analgesia? *Int J Gynaecol Obstet*. 1999;67(1):41-43.
- Kim YJ, Lenke LG, Bridwell KH, et al. Prospective pulmonary function comparison following posterior segmental spinal instrumentation and fusion of adolescent idiopathic scoliosis: is there a relationship between major thoracic curve correction and pulmonary function test improvement? *Spine*. 2007;32(24):2685-2693.



ON THE WEB

• Additional views of spinal fusion, VEPT, and vertebral staples, including radiographs taken before the surgical interventions.

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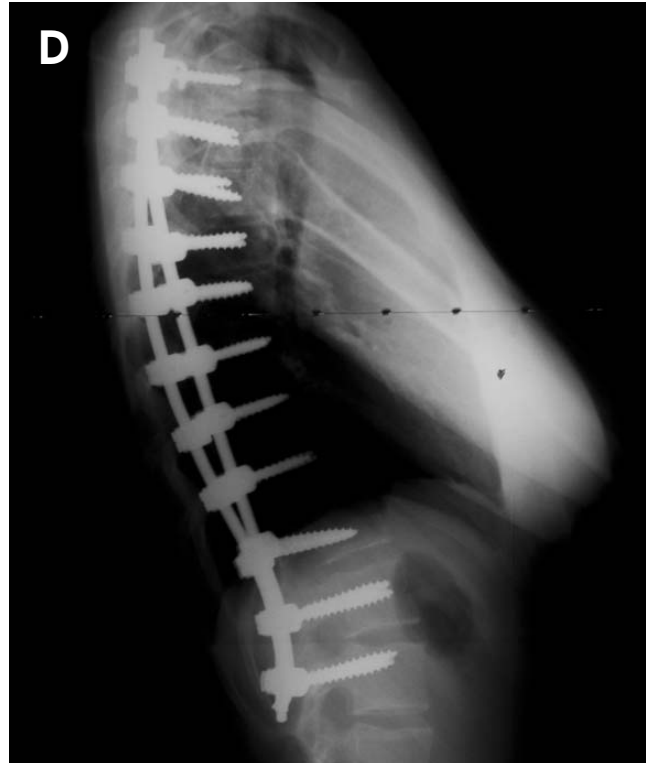
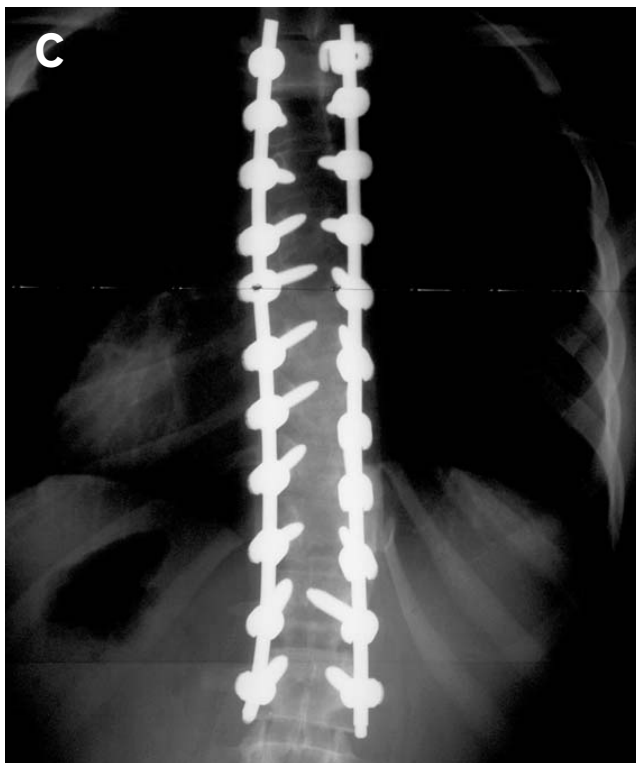
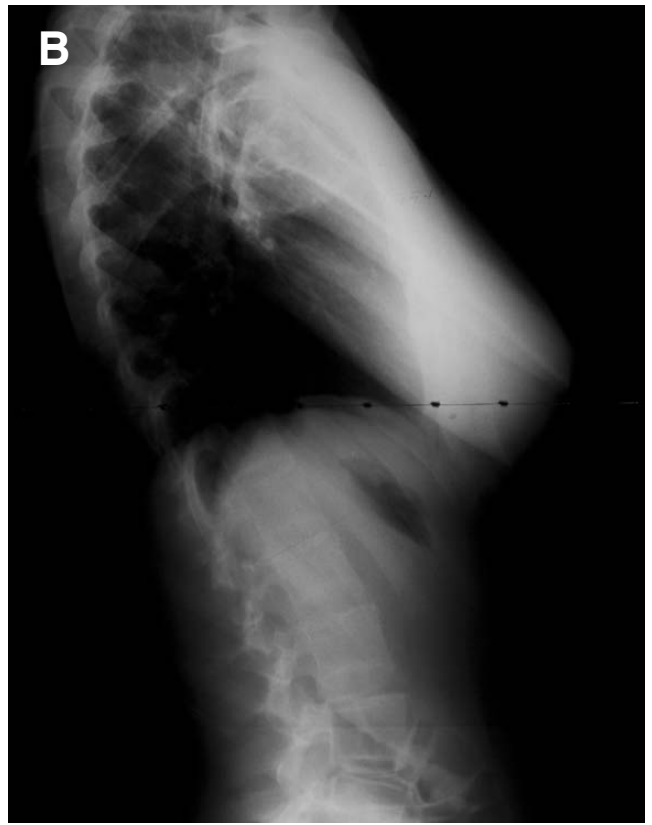


FIGURE 4. Spinal fusion with pedicle screws and contoured rod
Preoperative radiographs of posteroanterior (A) and lateral (B) views; postoperative radiographs of posteroanterior (C) and lateral (D) views.

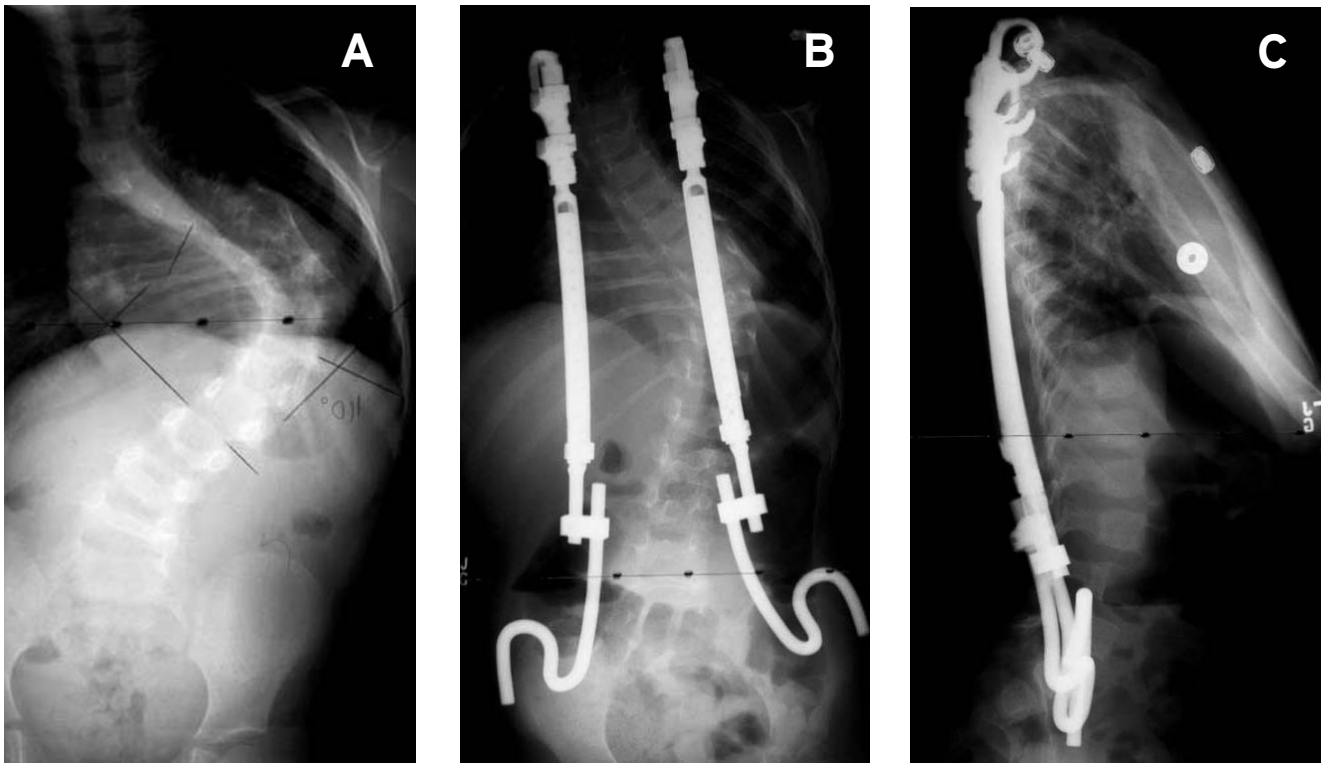


FIGURE 5. Vertical expandable prosthetic titanium rib device spanning from rib to pelvis
Preoperative radiograph, posteroanterior view (A); postoperative radiographs of posteroanterior (B) and lateral (C) views

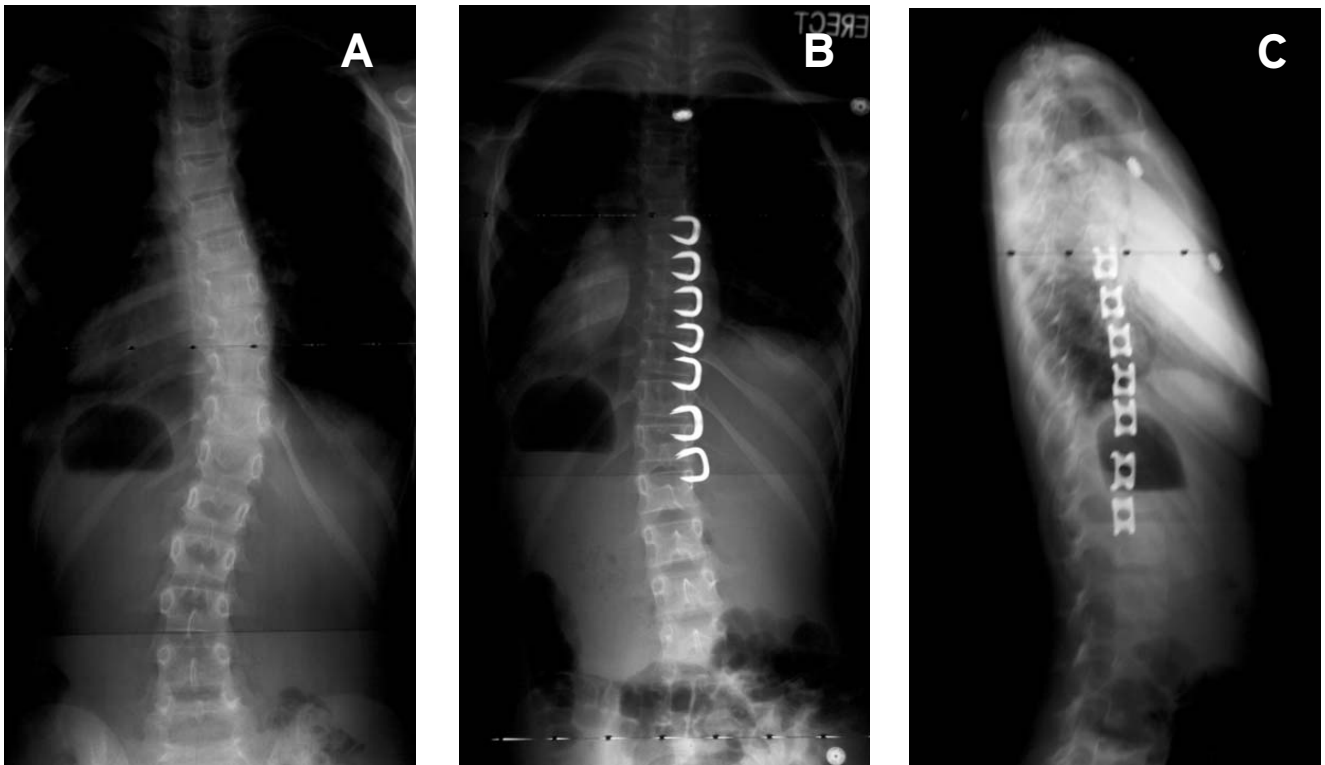


FIGURE 6. Vertebral staples inserted to limit growth on the convex side of a curve. Preoperative radiograph, posteroanterior view (A); postoperative radiographs of posteroanterior (B) and lateral (C) views