Consultation with the Specialist: Adolescent Idiopathic Scoliosis

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CONSULTATION WITH THE SPECIALIST

Adolescent Idiopathic Scoliosis

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Objectives

After completing this article, readers should be able to:

1. Discuss the causes and natural history of adolescent idiopathic scoliosis.
2. List the indications for magnetic resonance imaging in scoliosis patients.
3. Describe how and when to examine for scoliosis.
4. Know how to determine the magnitude and pattern of a curve based on standing spine radiographs.
5. Recognize indications for referral to a specialist.

Author Disclosure

Drs Stewart and Skaggs did not disclose any financial relationships relevant to this article.

Case Presentation

A 16-year-old girl presents with a complaint of spine asymmetry and a history of occasional back pain under her right scapula. She denies bowel or bladder problems and has no neurologic complaints. A right rib hump is apparent on the Adams forward bending test, and she has anterior prominence in the left lower rib cage. No neurologic deficit is noted on examination. Her radiographs demonstrate a 55-degree right thoracic scoliosis from approximately T6 to T12 (Fig. 1). On bending films, the curve bends out to 27 degrees. The patient subsequently undergoes posterior spine fusion from T4 to L1 and has an uneventful postoperative course (Fig. 2). She requires no more pain medicine and returns to school within 3 weeks of the surgery. Six months after the surgery, the patient resumes playing soccer.

Definitions

Scoliosis is defined as curvature of the spine greater than 10 degrees observed on a standing posterior-anterior (PA) spine radiograph with associated vertebral rotation. The Cobb angle is the angle on the PA view between the superior endplate of the most tilted upper vertebrae and the inferior endplate of the most tilted lower vertebrae.

Scoliosis may occur in association with various conditions. Congenital scoliosis implies failure of segmentation or formation of some vertebral elements and may be associated with fused ribs and spinal cord anomalies. Congenital scoliosis may be seen at any age. Neuromuscular or paralytic scoliosis occurs in children who have muscular dystrophy, congenital myopathies, spina bifida, spinal cord injuries, and other congenital or traumatic neuromuscular conditions.

Idiopathic scoliosis, or spinal curvature without an identifiable cause in an otherwise normal child, is the most common type and represents approximately 80% of structural curves. Idiopathic scoliosis in children younger than age 3 years is referred to as infantile scoliosis; onset in children from age 3 to 10 years is referred to as juvenile scoliosis. Adolescent idiopathic scoliosis is defined as scoliosis whose onset is seen in children older than 10 years. This review focuses on adolescent idiopathic scoliosis, the most common form, touching briefly on other types of scoliosis. The most important roles

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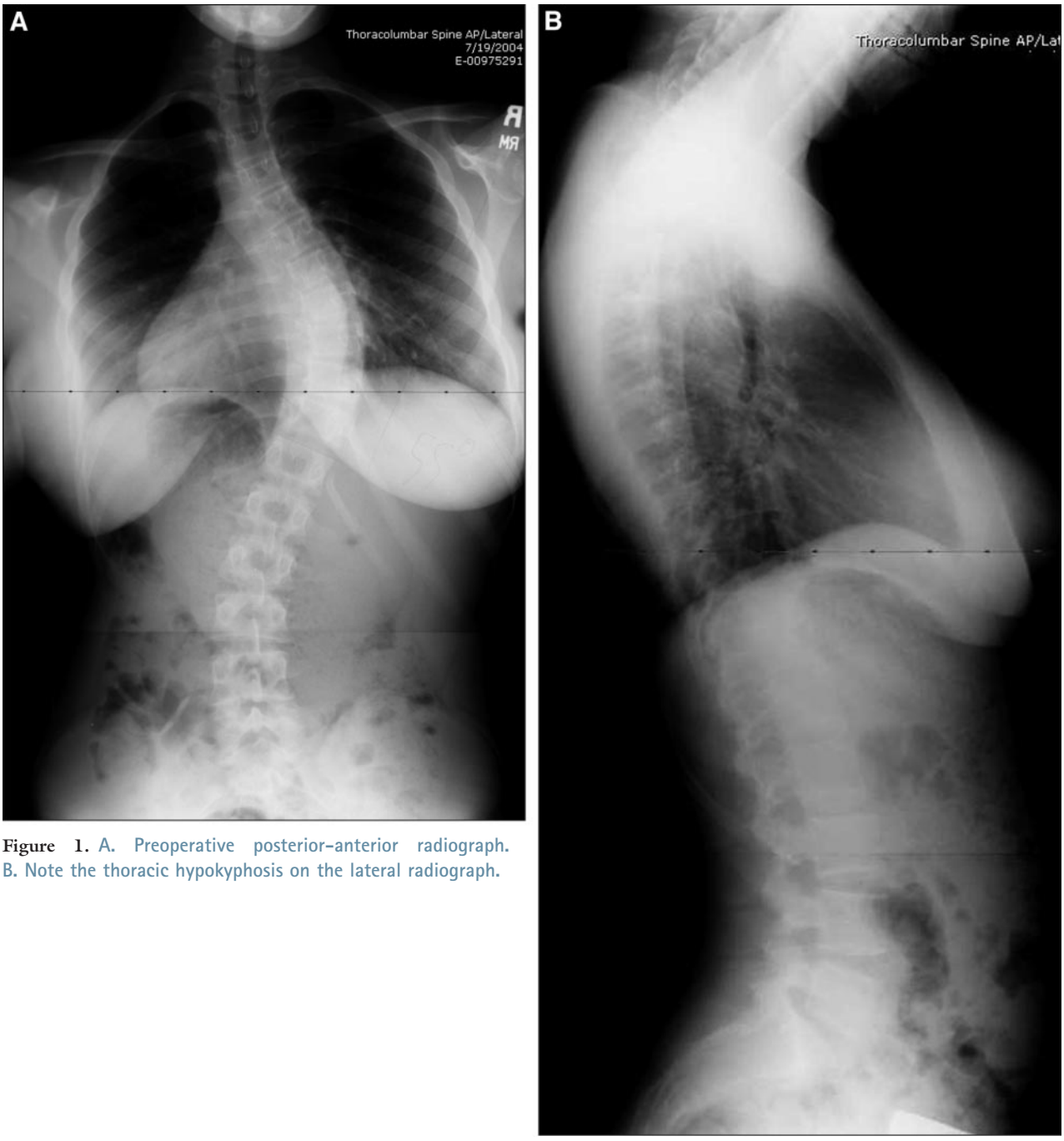

Figure 1. A. Preoperative posterior–anterior radiograph. B. Note the thoracic hypokyphosis on the lateral radiograph.

of the pediatrician in the evaluation of scoliosis are: 1) to ensure that scoliosis is truly idiopathic and that there are no other underlying treatable medical conditions associated with the patient's spinal curvature, and 2) to know when to refer a child to a specialist for additional evaluation.

Causes

Idiopathic scoliosis is more common in girls than in boys, and the female preponderance rises with increasing curve magnitude. Although many theories regarding the cause of adolescent idiopathic scoliosis have been studied, the underlying cause remains unknown. Studies have demonstrated a higher prevalence of scoliosis in families of patients,

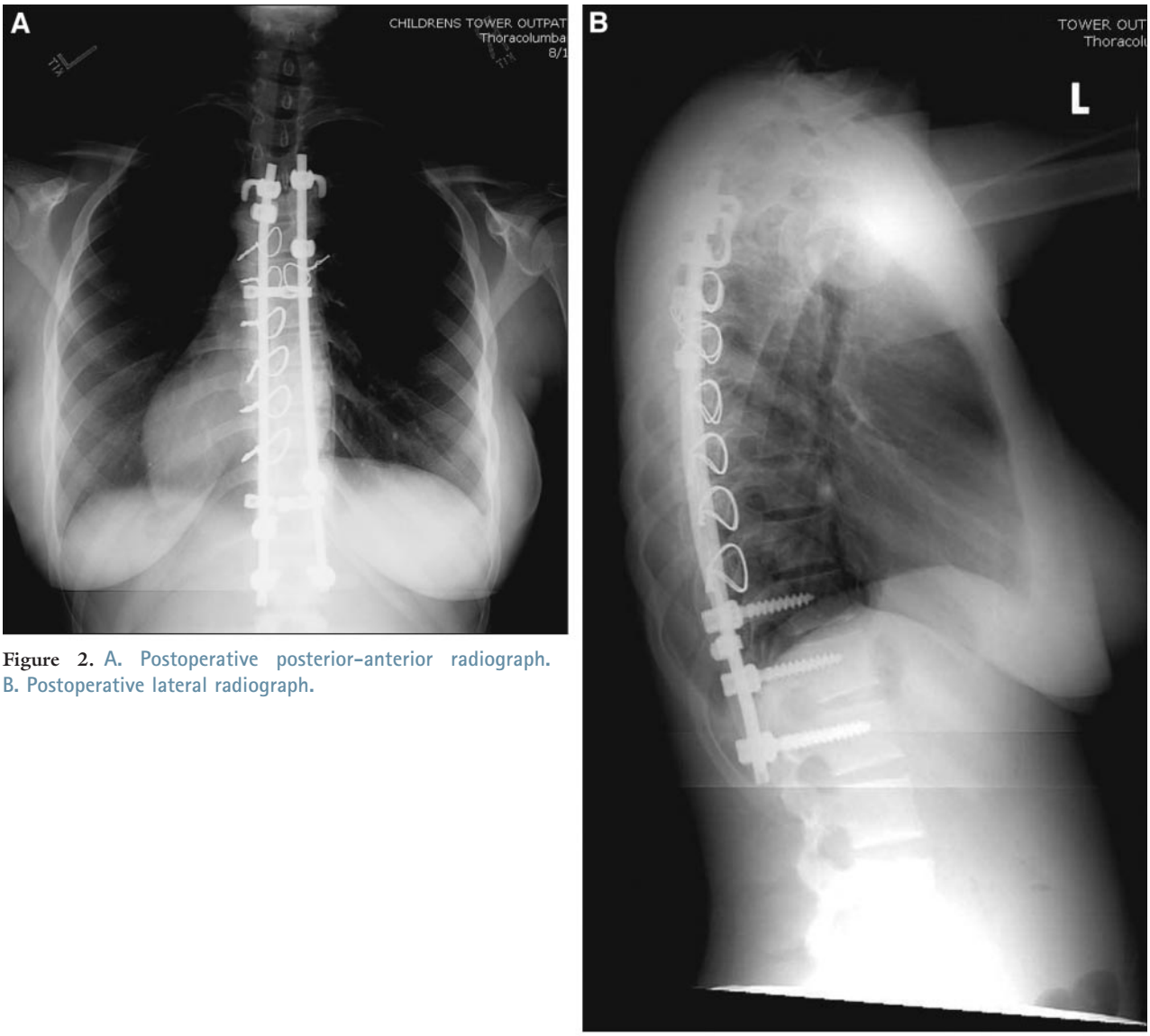

Figure 2. A. Postoperative posterior–anterior radiograph. B. Postoperative lateral radiograph.

suggesting some heritable factors. However, no single mode of inheritance has been linked conclusively to adolescent idiopathic scoliosis, and the causes of scoliosis generally are viewed as multifactorial. Patients should be informed that the risk of scoliosis in family members is slightly increased and that siblings should be screened when they approach adolescence.

Several other associated factors have been investigated. Some authors report lower melatonin levels in scoliosis patients compared with age-matched controls; other authors have been unable to reproduce these findings and report no difference. One investigation showed that idiopathic scoliosis patients have abnormalities of fibrillin, similar to the defects that have been seen in patients who have Marfan syndrome. Abnormalities of intervertebral disc collagen, platelets, and muscle and changes in vibratory sensation have been reported in scoliosis patients, but many authors believe these abnormalities to be results rather than causes of the scoliosis.

Natural History and Treatment Goals

Back pain, cosmetic issues, and pulmonary effects are the primary sequelae of untreated idiopathic scoliosis. According to American Thoracic Society criteria, children who have curves of more than 60

degrees have mild-to-moderate pulmonary dysfunction, which can become more severe with increasing curve size. Recent studies report that children who have curves as small as 20 degrees have decreased exercise tolerance due to pulmonary dysfunction. Lumbar and thoracolumbar curves have no proven impact on lung function. There is no evidence that patients affected with adolescent idiopathic scoliosis have any increase in mortality compared with unaffected individuals. In contrast, children who have infantile and juvenile scoliosis experience a large increase in mortality from cardiac and pulmonary causes that become apparent at age 40 to 50 years and sometimes before. The emphasis of treating early-onset scoliosis, therefore, differs from treatment for adolescent idiopathic scoliosis, focusing on maximizing the space available for the lungs by treating the chest wall deformity.

Congenital curves may worsen slowly, rapidly, or not at all. Neuromuscular curves often worsen relentlessly. Idiopathic curves can worsen at approximately 1 degree per month until the child achieves skeletal maturity during the adolescent growth spurt. The risk of idiopathic scoliosis progressing is related to the patient's age, menarchal status, skeletal maturity, and curve size and pattern, with young, premenarchal patients who have large curves experiencing the highest risk of progression. Progression of the scoliosis more rapidly than 1 degree a month is a warning sign that nonidiopathic processes may be involved and often warrants magnetic resonance imaging (MRI) of the spine.

Remaining growth is important in anticipating possible curve progression and length of bracing, but is highly variable. A variety of methods are available to estimate skeletal maturity, including estimating bone age by the method of Greulich and Pyle, Sexual Maturity Rating, onset of menses, Risser sign, and chronologic age. Growth continues for an additional 18 to 24 months following menarche, which can help the physician estimate a female's remaining time to grow. In premenarchal females, the history of beginning to form breast buds usually predates menarche by approximately 2 years and corresponds to a time of high growth velocity. The Risser sign is based on the ossification of the iliac apophysis across four quadrants from lateral to medial (1 to 4), with Risser 0 representing no ossification and Risser 5 representing full ossification with fusion of the apophysis to the ilium. Curve progression is most likely in younger patients who have considerable growth remaining and in those who have curves greater than 20 degrees. At the point of nearing skeletal maturity (Risser 3 and 4), one classic natural history study found a 1.6% probability of progression of at least 5 degrees for curves in the 5- to 19-degree range at presentation and a 23% chance of progression of curves that were already at 20 to 29 degrees. For patients in whom considerable growth remains (Risser 0 and 1), a 22% chance of progression was identified for curves between 5 and 19 degrees and a 68% chance for curves between 20 and 29 degrees.

School Screening

The role of school screening is controversial. Screening may lead to earlier intervention for children who have large curves, although most patients identified by school screening for scoliosis ultimately are found not to have a spinal deformity that requires treatment. The role of the physician is to determine if deformity exists and if follow-up or imaging studies are needed.

Clinical Evaluation

Screening for scoliosis should be performed annually as part of the health supervision visit. Screening should consist of inspecting the child's back from behind to assess for spinal, rib, or pelvic asymmetry and performing an Adams forward bending test to assess for rib rotation. These screening steps should not take more than 30 seconds. Younger children also should be evaluated for sitting balance and spine symmetry as a part of health supervision visits. Patients who have a positive family history should be screened early. Patients who have a positive screening result should undergo a more detailed history and examination.

History

The history begins by asking about any family history of scoliosis. One study noted that pain is a presenting complaint in 23% of patients who have adolescent idiopathic scoliosis; an additional 9% complain of pain at subsequent follow-ups. Although many adolescents and young adults experience some back pain, it generally is mild and activity-related. Severe pain, constant pain, night pain, and point tenderness are all warning signs to search for other causes. A history of unexplained or asymmetric weakness, sensory disturbance, bowel or bladder problems, or progressive impairment is concerning for neurologic involvement and should prompt focused examination and possible MRI of the spine. The time of menarche is noted for female patients.

Physical Examination

A vital goal of the evaluation of patients who have scoliosis is to rule out potentially treatable underlying dis-

orders. Especially in young children, the patient's height and weight should be evaluated relative to expected norms for the patient's age and sex because some skeletal dysplasias and syndromes are associated with scoliosis. The skin is inspected for a midline hairy patch or a dimple outside the gluteal cleft that could suggest an underlying spinal cord disorder. Café-au-lait macules suggest the possibility of neurofibromatosis. Marfan syndrome and other connective tissue disorders also are associated with an increased risk of scoliosis and should be kept in mind for patients who have excessive ligamentous laxity and marfanoid features on examination.

The patient is assessed for shoulder and waist symmetry while standing, and the spine is palpated for point tenderness. The Adam forward bending test is performed by asking the patient to bend forward at the hips, with the knees straight and arms hanging forward. The spine is inspected from behind for symmetry. A thoracic rib hump or lumbar asymmetry is present in patients who have significant idiopathic scoliosis.

Patients who have large, double-major curves can appear remarkably well balanced when viewed standing erect, but rib humps often become apparent when the patient bends forward. Because of an obligatory but variable coupling of rotation with spinal curvature, the rib rotation or asymmetry often suggests scoliosis. A scoliometer, a screening device used to measure rotation of the spine, is placed in the midline over the spot of maximum rotation during the Adams forward bending test to assess spinal rotation. A threshold of 5 to 7 degrees of rotation measured by the scoliometer often is used as the cutoff for orthopedic referral in school screening, which very roughly correlates to about a 20-degree Cobb angle.

A leg length discrepancy can result in spinal curvature compensating for pelvic obliquity, which is not idiopathic scoliosis. The examiner should ensure that the iliac crests are level when the patient stands. A limb length discrepancy may be quantified by placing blocks under the foot on the shorter side until the iliac crests are level. Radiographs taken with the leg length equalized by such a block show any true scoliosis independent of limb length discrepancy.

One of the most important parts of the physical examination is the neurologic assessment, which may uncover an underlying neurologic deficit that might be associated with the cause of the scoliosis (Table). Asking the patient to walk on heels, walk on toes, and single-leg hop on each foot usually can reveal any significant lower extremity motor weakness at the level of spinal nerve roots or above. Light touch sensation is assessed in both lower extremities to compare for symmetry. Range of motion of the hips, knees, and ankles is assessed for tightness or contractures. Popliteal angles are checked as the examiner tries to extend the knees with the hips flexed 90 degrees; tight popliteal angles lacking more than 20 to 30 degrees from full extension can be associated with spinal disease. Patellar, Achilles, and abdominal reflexes are assessed bilaterally, and the patient is evaluated for upper and lower extremity clonus and the Babinski sign. The superficial abdominal reflex is elicited by stroking the skin in the quadrants around the umbilicus. The reflex may be present, absent, or variable in unaffected patients, but abdominal reflexes that are consistently present on one side and absent on the other do not occur in healthy individuals and may warrant MRI of the spine. Foot deformity, especially when unilateral, also should raise suspicion of an intraspinal disorder.

Table. Sixty-second Neurologic Examination

- Have patient hop on each foot, one at a time
- Have patient walk on heels
- Check patient's reflexes
- Inspect patient's foot and dorsiflexion to assess muscle tone and clonus
- Test patient's sensation
- Check patient's popliteal angle

Adapted from Staying out of trouble. In: Skaggs DL, Flynn JM, eds. *Pediatric Orthopaedics.* Philadelphia, Pa: Lippincott; 2006. Used with permission.

The popliteal angle is measured by attempting to extend the knees in a supine patient whose hips are flexed to 90 degrees. A popliteal angle of 50 to 60 degrees or greater, measured by degrees lacking from full knee extension, demonstrates hamstring tightness, which can result from intraspinal disease and may warrant MRI of the spine.

Radiographic Evaluation

Patients who have scoliosis are evaluated via a PA spine radiograph to assess coronal curvature and a lateral radiograph to assess sagittal alignment. The curve is defined by the Cobb angle. The typical idiopathic thoracic curve has an apex to the right, and lumbar curves most commonly have a left apex. Left thoracic and right lumbar curves rarely occur in patients who have idiopathic scoliosis and should raise suspicion that other causes are involved. Lack of thoracic kyphosis is normal in idiopathic scoliosis patients, and the presence of excessive kyphosis suggests nonidiopathic causes. Radio-

graphs should be scrutinized for any vertebral anomalies that may suggest congenital scoliosis and widened pedicles that suggest diastematomyelia (abnormal midline bone or fibrous tissue in the spinal canal).

Except for patients who are unable to stand, initial radiographs of the spine should be obtained in a standing position because standing radiographs demonstrate a larger curve than do supine radiographs. If the iliac crest of one hemipelvis is significantly higher than the other on standing spine films, a leg length discrepancy probably is the cause of some or all of the spinal curvature. Hip dysplasia, subluxation, or dislocation also can create a leg length discrepancy and result in an oblique pelvis, which may not be recognized if the films do not include the hips. PA radiographs result in significantly less breast radiation than do anterior-posterior spine radiographs, and employing that technique represents the current standard of care.

MRI of the spine usually is not considered necessary in the routine evaluation of patients who have adolescent idiopathic scoliosis. When an MRI is ordered, it should include the cervical, thoracic, and lumbar spine, so abnormalities ranging from an Arnold-Chiari malformation to a tethered cord may be recognized. MRI is indicated for patients who have an abnormal history (abnormal pain, bowel or bladder incontinence), neurologic abnormalities on examination, atypical curve patterns or kyphosis, or other suggestions that the scoliosis is not idiopathic. For children who are ages 10 years or younger at the onset of scoliosis, MRI of the spine is indicated to assess for intraspinal disease. Approximately 20% of patients who have juvenile scoliosis (age 3 to 10 y) and 20% to 50% of patients who have congenital scoliosis have an intraspinal anomaly.

Treatment

The primary treatments for adolescent idiopathic scoliosis consist of observation, bracing, and surgery. We are not aware of any scientifically valid evidence that physical therapy, manipulation, electrical stimulation, diet, or other treatments correct scoliosis or reduce the risk of curve progression. Adolescent idiopathic scoliosis occurs at a time when most patients feel insecure about their bodies, and careful consideration should be given to the psychological aspects of the condition and treatments.

Bracing

Bracing often is considered for idiopathic curves in skeletally immature patients once the curve measures 20 to 30 degrees. The goal of bracing is to prevent or minimize curve progression. Correction of the scoliosis with bracing is uncommon. The effectiveness of bracing in the treatment of scoliosis is controversial, although most believe that a careful bracing program may have some effect on curve progression. One study reports that bracing is not effective in boys. Bracing does not appear to prevent or delay curve progression in idiopathic curves greater than 45 degrees or in neuromuscular curves greater than 20 degrees. Bracing generally is considered ineffective in the treatment of patients near or at skeletal maturity (Risser 4 and 5), although many factors such as height of the parents and family history of growth should be taken into account. Some studies have recommended against bracing in obese children and boys because efficacy could not be demonstrated. Different types of braces are available, depending on the curve pattern, with a custom-molded thoracolumbosacral orthosis being the most common. Studies show that most patients wear the brace significantly less than instructed and that they overreport brace compliance to the physician by approximately 150%.

Surgery

Long-term studies have shown that thoracic curves greater than 50 degrees and lumbar and thoracolumbar curves greater than 40 to 45 degrees can continue to worsen in adulthood at a rate of approximately 1 degree per year. Surgical intervention, therefore, may be recommended for such patients even after they achieve skeletal maturity. Skeletally mature patients who have smaller curves are not expected to worsen more than 10 to 15 degrees over an entire lifetime and typically do not require routine follow-up.

Surgery for adolescent idiopathic scoliosis generally consists of a spinal fusion and instrumentation (Fig. 2). A metal rod is attached to the vertebrae of the most curved portion of the spine, along with bone graft that eventually forms a solid construct analogous to that of concrete with reinforcing steel bars. Straight Harrington rods have been replaced by implants that allow for preservation of normal kyphosis and lordosis.

Surgery can be performed in the anterior spine, posterior spine, or both. Traditional anterior surgery through the chest is associated with decreased pulmonary function years later and, thus, has fallen out of favor. Although some centers promote thorascopic spinal instrumentation, this approach has not been adopted widely. Anterior surgery for thoracolumbar curves does not harm pulmonary function and may be able to leave more caudad vertebrae unfused, which may result in less stiffness and pain.

The most common surgical pro-

cedure is a posterior spinal fusion with instrumentation. For immature children who have significant growth remaining, an anterior and posterior surgery may be recommended to avoid the crankshaft phenomenon (continued anterior growth of the spine while the posterior growth is tethered by the fusion, leading to progressive rotational deformity). Postoperative regimens vary substantially. Our typical patient spends about 1 week in the hospital, returns to school in about 1 month, and slowly increases exercise until a full return to contact sports is allowed at 6 months.

Indications for Specialist Referral

Referral to an orthopedic surgeon for additional evaluation and management of scoliosis is indicated in otherwise healthy patients who have spinal curvature at or above 20 degrees because early treatment with bracing may slow or stop curve progression. In addition, patients who have atypical history, physical, or radiographic findings for idiopathic scoliosis should be referred. Some pediatricians prefer to refer patients based on clinical examination alone without prior radiographs. This procedure can minimize the number of pediatric visits needed and eliminate the frequent need to repeat referral radiographs in the orthopedist's office when outside films do not provide adequate spinal imaging. Early orthopedic referral also is recommended for patients who have any identified congenital spine anomalies and for patients who have juvenile, neuromuscular, or paralytic curves.

Summary

The most common form of scoliosis is adolescent idiopathic scoliosis or scoliosis with onset in children older than age 10 years without other underlying disorders. Rib rotation associated with spinal curvature can be appreciated on the physical examination. Curve magnitude may be determined from a standing PA spine radiograph. A spine MRI should be obtained for patients who have neurologic symptoms or other evidence of underlying pathology. Patients who have curves greater than 20 degrees, early-onset scoliosis, or any atypical findings should be referred to an orthopedic specialist. Small curves should be observed; large curves in a growing child may require bracing or surgery.

Suggested Reading

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PIR Quiz

Quiz also available online at www.pedsinreview.org.

10. Scoliosis is a back condition defined by the finding of a spinal curvature (with vertebral rotation) on a standing posterior-anterior radiograph of greater than:

    A. 2 degrees.
    B. 4 degrees.
    C. 6 degrees.
    D. 8 degrees.
    E. 10 degrees.

11. A variety of theories have been suggested for the cause of the most commonly seen form of scoliosis, which is idiopathic adolescent scoliosis. The *most* likely cause is:

    A. Autosomal dominant inheritance.
    B. Autosomal recessive inheritance.
    C. Multifactorial.
    D. Prenatal factors.
    E. Sex-linked inheritance.

12. The effects of scoliosis on pulmonary function are debatable, but increasing curves can cause pulmonary dysfunction. Accordingly, it is important in the follow-up of patients who have scoliosis to remember that mild-to-moderate pulmonary compromise can be anticipated with a curve greater than:

    A. 20 degrees.
    B. 30 degrees.
    C. 40 degrees.
    D. 50 degrees.
    E. 60 degrees.

13. A 14-year-old girl is brought to her pediatrician for the first time in 2 years. The mother has been taking her to a chiropractor for treatment of scoliosis. On examination, using the Adams forward bending test, a prominent posterior thoracic rib hump with obvious scoliosis is apparent. With much urging, a complete scoliosis spine series is obtained that reveals two curves involving the thoracolumbar areas. The Cobb angle is 80 degrees on the larger curve. The *most* appropriate course of action for this child at this point is:

    A. Electrical stimulation.
    B. Physical therapy and close follow-up.
    C. Prompt fitting of a custom-molded thoracolumbosacral orthosis.
    D. Repeat spine films in 3 months to assess for progression.
    E. Spinal fusion with instrumentation.

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