Scoliosis and Kyphosis

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Sometimes with secret pride I sigh,
To think how tolerant am I;
Then wonder which is really mine:
Tolerance, or a rubber spine?

― Ogden Nash

Learning Objectives:
1. Understand the difference between normal and abnormal curvatures of the spine
2. Appreciate the role of the patient’s history in diagnosing idiopathic scoliosis
3. Perform the Adams test and understand its role in evaluating for scoliosis
4. Understand the results of spinal radiographs in the context of prognosis, referrals, and treatment
5. Differentiate between common types of adolescent kyphosis

Primary References:

CASE ONE:

A 12-year-old patient, Bendice Pine, presents to your office after a concern for scoliosis was noted on routine school screening. The child looks as if she has some questions that she wants to ask, but she is quickly overshadowed by her mom, Nerva Smother. Before you can even start your standard scoliosis discussion, she demands that you “repeat the test from school because I’ve never noticed anything wrong with my daughter.” She tells you that she does not want Bendice to have to wear a brace. You assure Ms. Smother that you will repeat the exam and answer all of her questions, and proceed with a brief background explanation before taking a history.

1. What is scoliosis and what are the different types?

It is normal to have a certain degree of spinal curvature in both the coronal and sagittal planes. Scoliosis is defined as a lateral curvature of the spine in the coronal plane; when that curvature is greater than 10 degrees according to the Cobb classification (see below) it is considered abnormal. There are two main categories of scoliosis, congenital and idiopathic, and three subtypes of idiopathic scoliosis: infantile (most rare), juvenile (ages 3-10) and adolescent. Idiopathic scoliosis accounts for 80% of abnormal spinal curves. Idiopathic adolescent scoliosis, the most common form, is more common in girls, who are also 5-8 times more likely than boys to have their curves increase in size and to need treatment.

2. What are the key pieces of the history that you want to keep in mind when assessing a child for scoliosis?

Age of onset, presence of back pain, and other neurological complaints should be considered. If a neurologic or anatomic process underlies the scoliosis, it is no longer considered idiopathic. The age of onset can provide information about the rate of progression, potential for more progression, and associated pathology. Abnormal developmental milestones in young children may be suggestive of a
neuromuscular cause (i.e., cerebral palsy, muscular dystrophy) or birth defects related to abnormal spinal development. The vertebral defects of VACTERL association can result in scoliosis. Juvenile onset of scoliosis is associated with occult intraspinous abnormalities in about 20% of patients, and should prompt consideration of an MRI from the craniocervical junction to the sacrum.

While 23% of over 2000 adolescents referred for evaluation of scoliosis presented with back pain in a retrospective study, there was no correlation between presence of pain and the degree of spinal curvature. Pain other than mild musculoskeletal pain (i.e., severe constant pain, pain occurring mainly at night, and/or point tenderness), should be a red flag for an underlying condition such as a tumor of the spinal column, osteoid osteoma, infections, a syrinx, or tethered spinal cord, and may warrant a more comprehensive workup.

Neurological complaints such as loss of bowel or bladder control, numbness, or trouble with proprioception can signal one of a number of neuromuscular diseases associated with abnormal curvature of the spine, including muscular dystrophy, spinal cord injury, congenital myopathies, and spina bifida.

Studies have demonstrated a higher prevalence of scoliosis in families of patients, but no specific mode of inheritance is yet identified. Certain inherited syndromes, such as Marfan syndrome, are associated with abnormal spinal curvature.

The age of menarche is used as a marker of skeletal maturity. Continued growth is expected for 18 to 24 months following menarche. In children with idiopathic adolescent scoliosis, curve progression is most likely in those who have considerable growth remaining (see the discussion of Risser sign below).

3. **What is the utility of school screening?**

There is debate over the utility of school screening for scoliosis as well as screening for scoliosis in the annual physical exam. In a 2015 joint statement, the American Academy of Orthopaedic Surgeons, the Scoliosis Research Society, the Pediatric Orthopaedic Society of North America, and the American Academy of Pediatrics reinforced their prior 2007 recommendation for routine screening of girls at ages 10 and 12, and boys at age 13 or 14. An expert panel concluded that there can be substantial benefits from early treatment of idiopathic scoliosis, and that in light of incomplete and conflicting data, screening is justified. Note that this recommendation includes two screenings for females, since females are afflicted with scoliosis that requires treatment three to four times more frequently than males. By screening at both ages 10 and 12, one can capture variation in maturity. In addition, the International Scientific Society on Scoliosis Orthopaedic and Rehabilitation Treatment (SOSORT) recommends school screening programs for the early diagnosis of idiopathic scoliosis.

In a 2018 statement, the U.S. Preventive Services Task Force (USPSTF) arrives at a different conclusion, stating that “the current evidence is insufficient to assess the balance of benefits and harms of screening for adolescent idiopathic scoliosis in children and adolescents aged 10 to 18 years.” This statement is in contrast to their 2004 recommendation against screening. The American Academy of Family Physicians echoes the updated USPSTF position.

In order to understand the recommendations, one must understand the questions to ask when considering a screening program.

- There must be evidence that the screening test is sensitive; for scoliosis, can a physical examination of the back detect curves?
- What are the adverse effects of screening? For scoliosis, adverse effects of screening include exposure to radiation, parental worry, and excess referral to orthopedic surgeons.
- Will screening (early detection) help? Do people with a positive screen who do have scoliosis have better outcomes (in terms of back complaints, disability, or psychosocial complaints) than those who were diagnosed as a result of complaints like back pain or parental realization of back asymmetry?
- Do treatments designed to minimize curvature of the spine actually work?
- Do these treatments cause adverse effects?
• Is the screening test specific? Do curves diagnosed by screening exam actually progress to curves of clinical significance? Specificity of a screening test becomes more important as the implications of false positive results increase.
• Do people whose curves progress have more complications, or might there be some other cause of symptomatology?

The USPSTF summarizes their analysis of these questions as follows:

The USPSTF found no direct evidence regarding the effect of screening for adolescent idiopathic scoliosis on patient-centered health outcomes. The USPSTF found inadequate evidence on the treatment of idiopathic scoliosis (Cobb angle <50° at diagnosis) in adolescents with exercise (2 small studies) or surgery (no studies) or its effects on health outcomes or the degree of spinal curvature in childhood or adulthood. The USPSTF found adequate evidence (5 studies) that treatment with bracing may decrease curvature progression in adolescents with mild or moderate curvature severity (an intermediate outcome). However, it found inadequate evidence on the association between reduction in spinal curvature in adolescence and long-term health outcomes in adulthood.

In contrast, the pro-screening SORORT recommendations factor in studies demonstrating that curves greater than 25° before skeletal maturity will continue to progress and curves greater than 50° will progress 1-2° per year after skeletal maturity. However, they acknowledge that “heterogeneity of the study protocols limits generalizability of the recommendations.”

Providers must balance the incomplete data against expert recommendations, while factoring in state-specific regulations, when making individual practice decisions regarding routine screening for idiopathic scoliosis.

4. Now that a school nurse has raised the issue, why would you repeat the exam and how would you do it?

The job of the primary care provider is to perform a more thorough examination to assess for spinal deformity and associated conditions. The patient is first asked to stand with upper body minimally clothed so that the spine is as visible as possible. The feet should be toed up to a line and separated so that the knees do not interfere with each other. The back is inspected for significant (>1cm) shoulder or scapular asymmetry, and for a difference in the distance between the arm and waist on either side. In addition, leg length discrepancy can result in spinal curvature, so it is important to note whether the iliac crests are level when the patient stands. The Adams forward bending test is then performed by asking the patient to bend forward at the hips with the knees straight and the arms hanging forward. As the patient bends forward, the spine is inspected from behind for an elevation of either side of the paraspinal area or rib cage. Of note, this is a good opportunity to assess for abnormalities that may be indicators of other causes of abnormal spinal curvature (i.e., spinal tufts or café-au-lait spots). A comprehensive neurologic exam should be administered, including tests for deep tendon reflexes, abdominal reflexes, clonus, spasticity, and Babinski.

Due to association of rib rotation with spinal curvature, the scoliometer is often used to measure the degree of rotation. The scoliometer, which works like a carpenter’s level (with a moveable ball inside to indicate the angle), is a measuring device which is placed in the midline over the spot of maximal spine rotation during the Adams test. The measurement should be repeated twice to improve accuracy.

CASE TWO:

Bendice’s back exam is normal; it is symmetrical on standing and forward bending. Mrs. Smother and Bendice are relieved. Interestingly, your next patient, Curva Sure, is a 12-year-old female whose mother has noticed a difference in her shoulder heights and is coming to you for evaluation. The history is otherwise unremarkable. On exam, you note asymmetry in the shoulder height and arm-to-waist distance, as well as spinal curvature on the Adams forward bending test.
5. How would you counsel Curva and her mom? What is your plan for further investigations?

It is important for the pediatric provider to exclude any underlying medical conditions that could cause or contribute to the patient’s spinal curvature. Based on the unremarkable history and the age of onset, the most likely diagnosis is idiopathic adolescent scoliosis. The family should be told that it is likely to see progression of her curve during the adolescent growth spurt of up to 1 degree per month.

Recent studies have indicated that pulmonary dysfunction leading to decreased exercise tolerance can occur in children with curves as small as 20 degrees, and commonly in those with curves of more than 60 degrees. You might screen Curva’s younger siblings as they approach adolescence due to the slightly increased risk in family members of those with idiopathic scoliosis.

One next step is a radiographic evaluation of the patient’s spine via standing PA and lateral spinal radiographs. These films will provide information as to the exact degree of curvature using a measurement system known as the Cobb angle, which is the angle of intersection of two perpendicular lines: one drawn from the cranial border of the most angled upper vertebrae and the other from the caudal border of the most angled lower vertebrae. A rotation of 5 to 7 degrees measured by the scoliometer is often used as a cut-off for orthopedic referral as it roughly correlates to a Cobb angle of 20 degrees. In some communities, spine surgeons prefer that primary care providers refer patients prior to obtaining radiographs.

Predictions of prognosis are informed by the Risser sign, a scale from 0 to 5 which correlates with the degree of ossification of the iliac crest as noted on radiograph. A higher score indicates a greater level of skeletal maturity and is associated with a lower probability of progression of the curve. This skeletal maturity index can be roughly correlated to the stages of puberty: the average female reaches a Tanner stage I at 11 years of age, the beginning of the growth spurt at 11.5 years of age, a Risser grade 1 at 12 years of age, and has an onset of menarche between 12 and 13 years of age.

If a patient’s curve is greater than 30 degrees, and she is skeletally immature (Risser 0-1) or has not yet reached menarche, there is a very high likelihood that she will have significant progression of her curve and is a candidate for bracing.

More than 90% of patients with idiopathic scoliosis have a curvature that is right thoracic, left lumbar, (like a backwards S). Any direction or location that differs from this should be further evaluated with an MRI total spine (posterior fossa to conus) for intraspinal anomalies causing the scoliosis (i.e., intraspinal anomalies). Other reasons to evaluate with MRI include rapid progression, excessive kyphosis, structural abnormalities, neurologic symptoms or pain, foot deformities or asymmetric abdominal reflexes.

CASE continued:

Curva and her mom return for a follow-up appointment in one week. You (with the help of a kind radiologist) find that the x-ray is significant for a right thoracic convexity of 25 degrees. You’ve decided to refer them to a pediatric orthopedist for further evaluation and management, but Ms. Sure wants some information about treatment options before leaving your office.

6. What are the main treatment options that you will discuss with them?
Although referral to an orthopedist is usually made on the basis of an abnormal Adams test, spinal rotation of 5-7 degrees as measured by a scoliometer (in those practices that use them) or Cobb angles approaching 20 degrees are often used as cut-offs for orthopedic referral, as well. There are three main options for treatment: observation, bracing, and surgery. As the primary care provider, it may be helpful to discuss these three options with patients and their families prior to referral. Observation is usually indicated for more skeletally mature patients with smaller curves, due to the decreased risk of lifetime progression, until at least 2 years past menarche. Options include serial physical exams and/or serial spinal radiographs at 4-6 month intervals until skeletal maturity is reached. Since curve progression is greater during the adolescent growth spurt, more frequent radiographs may be obtained during this time.

Physical therapy is popular in Europe to strengthen the trunk musculature. However, there is poor quality of evidence confirming the efficacy of physical therapy in reducing the risk of progression of scoliosis.

Bracing is usually considered for patients with less mature skeletons (Risser <3) who have curves greater than 20-30 degrees or demonstrate progression. Bracing has been shown to prevent or minimize progression of the curve. A 2013 multi-center randomized trial to clarify the role of bracing ended early owing to the efficacy of the intervention. Bracing significantly reduced the progression of high risk curves, and the benefit increased with longer hours of brace wear. However, critics of the study point out that 48% of patients in the observation group had treatment success (did not require surgical correction), as did 41% in the intervention group who spent little time wearing the brace. Poor adherence to bracing is common, particularly in the adolescent population, due to the social and emotional impact of the intervention. Bracing may hold promise for a subset of patients though it is not yet clear which patients would benefit compared to observation alone.

Surgical treatment is typically reserved for patients with curves greater than 40-50 degrees. This is because curves greater than 50 degrees will progress approximately 1 degree per year even in sexually mature patients. The most common surgery is posterior spinal fusion with instrumentation, in which a metal rod is attached to the vertebrae of the most curved portion of the spine along with a bone graft that eventually forms a solid frame. Skeletal maturity and degree of related pulmonary dysfunction are other important factors that are taken into consideration. Recent studies have questioned the utility of surgery even in patients with curves of 50-75 degrees. While these patients are more likely to have pulmonary symptoms and back pain, left untreated there is no increase in mortality, and there is insufficient evidence that surgery is effective in treating back pain associated with scoliosis. Furthermore, patients with moderate curves who are untreated may have no disadvantageous effects on quality of life compared to a healthy population.

CASE THREE:

A week later, Ms. Smother is back in your office - this time, she has brought her 15-year-old son, Kinkeese Pine. She tells you that for the past year she has noticed that he always has his shoulders hunched. She thinks it makes him look sloppy and, given the recent experience with his sister, is concerned that “if he doesn’t stand up straight, he’ll stay that way forever!” Kinkeese doesn’t feel that there’s anything wrong and is tired of his mother being on his case.

7. What are the two most common types of pediatric kyphosis and how would you distinguish between them on physical exam?

Kyphosis is the curvature of the thoracic spine in the sagittal plane (with a posterior apex); normal kyphosis is typically a curve of 20-45 degrees; the term is more commonly used to refer only to excessive curvature.

Postural roundback (also known as flexible kyphosis) is not permanent and can be voluntarily corrected by hyperextending the back. Scheuermann’s kyphosis is a rigid structural deformity, most often mid-thoracic, which cannot be voluntarily corrected. When observed from the side during an Adam’s forward bending test, a patient with Scheuermann’s kyphosis may have an angulated “A-frame” spine.
as opposed to a smoothly rounded back in postural roundback. Scheuermann’s kyphosis is radiographically defined by an angle of kyphosis exceeding 45 degrees due to the anterior “wedging” (by 5 or more degrees) of at least 3 adjacent vertebrae at the apex of the curve. Onset typically occurs in early adolescence. There is an increased prevalence among males, with taller males being at higher risk for developing severe disease, and an autosomal dominant inheritance pattern. Surgical correction is reserved for patients with large deformities. Refer to figure 669-4 in Nelson’s Textbook of Pediatrics, 17th edition, page 2285, for an illustrative example of a radiograph in Scheuermann’s kyphosis and to the Resources section for an illustration.

CASE continued:

You ask him to stand up straight in the office and his thoracic curvature normalizes.

8. How would you counsel Kinkeese and his mom?

The key to counseling these families is that most patients with postural roundback will outgrow the deformity. Serial physical exams and extension exercises may be part of the treatment plan as well. If Kinkeese had not been able to straighten his spine with hyperextension, or if he had had an A-frame deformity on forward bending, you would have referred him to an orthopedist for further management of likely Scheuermann’s kyphosis. The typical treatment is with extension and strengthening exercises and follow-up with serial lateral radiographs. If pain persists, or if the angle of kyphosis exceeds 60 degrees, bracing or other orthopedic intervention may be warranted. Bracing is indicated in a skeletally immature patient who has moderate deformity that spans several levels and is flexible. Severe kyphosis >70 degrees can progress into adult life and cause back pain, respiratory complications, and neurological issues.

Additional References:

Resources:

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