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SCOLIOSIS

Developed by Sahaj Puri and Dr. Selvakumar for PedsCases.com.
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Introduction:

Hi everyone! Welcome to this PedsCases podcast where we will be reviewing scoliosis in the pediatric population. My name is Sahaj Puri and I'm a third year medical student at McMaster University. This episode was created in collaboration with Dr. Selvakumar, a general pediatrician at McMaster University.

Learning Objectives:

By the end of this podcast, the listener should be able to:

1. Define scoliosis and briefly describe the major categories of this condition.
2. Describe an approach to evaluating a child who may be presenting with scoliosis, including key components of the history and physical exam.
3. Review the radiographic findings of scoliosis and how they may guide indications for management.
4. List options for conservative management as well as active interventions for scoliosis.

Clinical Case:

Let's begin with a clinical case. You are working with your preceptor in a general outpatient pediatrics clinic. Your preceptor asks you to see Ellie, an 11 year old girl, who is a new patient to the clinic. You enter the room and introduce yourself to Ellie and her mom, Sandra. You learn that Sandra has noticed that her daughter's shoulders appear uneven. Additionally, Ellie has noticed that her back is a bit sore after she has been standing for a long time in band practice. Upon further history, Ellie's mother confirms that Ellie was born at term via spontaneous vaginal delivery. She did not require a NICU stay and has a generally unremarkable past medical history. Sandra remembers that her aunt had scoliosis growing up and wonders whether this may be something Ellie is also presenting with.

On exam, you observe that Ellie's right shoulder appears slightly elevated compared to her left. When you ask Ellie to stand straight, it also appears as though her right arm is slightly shorter than the left one. A thorough neurological examination reveals no significant abnormalities. You return to your preceptor to share your findings and discuss relevant next-steps for Ellie's care.

Keep this case in mind as we go through the rest of the podcast and we'll return to Ellie's concern at the end of our discussion.

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Defining Scoliosis and its Categories:
Scoliosis is defined as an abnormal lateral curvature of the spine over 10 degrees with vertebral rotation.[1] If a curve falls under 10 degrees, it is thought to represent spinal asymmetry, rather than scoliosis.[2] The degree of the curvature is identified through radiographs, which we'll discuss in greater depth later in this episode. Scoliosis can be categorized based on its etiology and is often classified as idiopathic, congenital or neuromuscular in nature.[2]

Idiopathic Scoliosis
Idiopathic scoliosis is the most common type of scoliosis, representing about 85% of cases in children and adolescents.[2] It can be further subdivided based on age group. Infantile idiopathic scoliosis is inclusive of children up to 3 years of age.[2] Juvenile idiopathic scoliosis is seen in children 4 to 9 years of age and adolescent idiopathic scoliosis is seen in those 10 years or older.[2]

Of these subcategories, adolescent idiopathic scoliosis is the most common type and is often seen around the time a patient is entering a growth spurt.[2] Adolescent idiopathic scoliosis impacts about 2-4% of adolescents.[1] Although the general incidence of scoliosis is quite similar between males and females, it has been found that females have almost a 10 times greater risk for progression of their curve.[1]

As evident from the name, the pathophysiology of idiopathic scoliosis is not well understood at this time. There is evidence that a genetic aspect may underlie adolescent idiopathic scoliosis, although no clear inheritance pattern has been identified.[2] It has been found that if both parents have idiopathic scoliosis, their child is approximately 50 times more likely to require treatment for scoliosis relative to the general population.[1] Mutations in the CHD7 and MATN1 genes are found to be involved in scoliosis development, although many other unidentified genes may also play a role.[2]

Congenital Scoliosis
Congenital scoliosis is a less common type of scoliosis that is thought to impact about 1 in 10 000 newborns.[4] The vertebrae form between 8 to 12 weeks of gestation, and malformation of the vertebrae in utero during this period can lead to scoliosis.[4] When evaluating a newborn with scoliosis, we may find hemivertebrae, which are vertebrae that are only partially formed.[4] Sometimes, we may find partially fused vertebrae.[4] In other cases, babies may present with a combination of these concerns underlying their presentation.[4]

Neuromuscular Scoliosis
Neuromuscular scoliosis is another, more rare cause of scoliosis. It is primarily seen in children with underlying disorders of their muscles, brain or spinal cord.[5] This can lead to an inability to effectively align the spine. It is commonly associated with conditions such as cerebral palsy, myelodysplasia and spinal muscular atrophy.[5]

Given that idiopathic adolescent scoliosis is the most common type of scoliosis, we will primarily focus the rest of this podcast around a discussion of this presentation.

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Initial Evaluation of A Child Suspicious for Scoliosis:

When evaluating a child for suspected scoliosis, a thorough history and physical exam are vital components to consider.

Key Components of the History

Children initially presenting with scoliosis may approach a physician with concerns about one shoulder being higher than the other.[3] Other chief complaints may include a concern about clothes not hanging straight on their body.[3]

It is important to ask the child and their caregiver about any family history of scoliosis.[2] To obtain more information about possible etiologies, the child should be asked about neurological concerns. This includes any numbness, paresthesias or weakness in their limbs.[2] Children should also be evaluated for any red flags of back pain, such as bowel or bladder incontinence.[2]

Of note, scoliosis is not typically painful for most patients.[1] However, children may sometimes complain of back pain or fatigue, particularly with prolonged sitting or standing.[3] If a child is presenting with red flags of back pain, including pain that awakens them from sleep, we may worry about a more concerning underlying etiology, rather than an idiopathic scoliosis.[2]

Key Components of the Physical Examination

In addition to obtaining a history, it's important to perform a thorough neurological exam.[2] The child should be observed walking normally, on their toes, heels as well as on the medial and lateral aspects of their feet.[2] A high arch on the child's foot may indicate an underlying neurological disorder that is contributing to the child's presentation.[2] A dermatological exam should also be done, with attention to features such as cafe au lait spots, which may indicate conditions such as neurofibromatosis.[2]

In addition to these exams, more targeted exam maneuvers for scoliosis should be performed as well. For example, the clinician may palpate the iliac crests to observe for symmetry of the pelvis.[2] Of note, neuromuscular scoliosis can often present with an oblique pelvis, where one side is tilted higher than the other.[6] Additionally, newborns with congenital scoliosis may present with a tuft of hair or sacral dimple.[5] The clinician should also look for a leg-length discrepancy, asymmetrical gap between the waist and patient's arms when they are hanging by their sides, as well as asymmetry of the scapula or ribs.[2]

A detailed examination of the back includes inspection from various angles. For example, inspection from a lateral view may reveal increased or decreased kyphosis, which is the outward curvature of the spine.[2] The Adam's forward bend test is another commonly employed physical exam maneuver that aims to look for rotational asymmetry.[2] For this test, the child keeps their knees straight and bends forward - almost as if they were trying to touch their toes![2] The clinician should inspect the patient's spine by standing behind the child to look for any asymmetry.[2]

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Sometimes, an inclinometer, or scoliometer, can be used to assess the level of asymmetry on an Adam's forward bend test.[2] The scoliometer is placed on the child's back during the exam and allows us to measure the angle of trunk rotation.[2] A scoliometer measurement of under 5 degrees is unlikely to require follow up, while a degree greater than 10 should prompt further investigation.[1]

Further Imaging for Investigation of Scoliosis:

Upon suspicion of scoliosis, clinicians should refer children for radiographic imaging of their spine. Ideally, we should have AP views of the child while they are standing to ensure view of the thoracic and lumbar spine.[2]

The magnitude of a child's spinal curvature is defined using the Cobb angle, which is measured using x-rays.[2] Since the Cobb angle can tell us about the severity of the scoliosis, it can be used to guide subsequent treatment. To determine the Cobb angle, we must first identify the top and bottom vertebrae involved in the curve.[2] These would be the vertebrae that are tilted the most off of the baseline horizontal plane that we would expect a vertebra to lie upon.[2] A line is then drawn along the superior aspect of the top vertebra.[2] Another line is drawn along the inferior aspect of the bottom vertebra.[2] The angle at which these two lines intersect is the Cobb angle.

Children presenting with a Cobb angle under 20 degrees are considered to have mild scoliosis.[2] Curves between 20 and 40 degrees are thought to be moderate, while those above 40 degrees are severe.[3] Large curves, such as those over 70 degrees, may cause complications such as restrictive lung disease and severe back pain.[2]

Scoliosis curves are described by the location of the apical vertebra within the curve, such as whether it is thoracic or lumbar, as well as the direction that the curve is facing.[2] When evaluating adolescent idiopathic scoliosis, it has been noted that almost all thoracic curves have an apex facing to the right.[2] On the other hand, almost all lumbar curves have an apex facing the left.[2] The reason for this is not well understood at this time. Apart from x-rays, no other imaging is routinely used for scoliosis management. However, if a child presents with a left thoracic curve, rather than the typical right thoracic curve, an evaluation with MRI may be warranted to investigate for underlying etiologies.[2]

In other more rare cases, such as congenital scoliosis, children may also undergo further testing with echocardiography and ultrasound to investigate for possible abnormalities within their organs.[4] This is because spinal formation is concurrent with the growth of other organs in the body, including those within the genitourinary, cardiac and nervous systems.[4]

Curve Progression

Before we move on to a more detailed discussion of the key principles of scoliosis management, it is important to recognize factors that may be involved in disease progression. As mentioned, periodic x-rays can be used as a tool to track curve progression over time. Determining the child's risk of progression is important as it can guide the interventions that a physician may need to consider for their patient.

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Risk factors for increased progression of a curve include sex assigned at birth, as females tend to have a greater risk.[1] Additionally, the child's age and relative skeletal maturity can influence their risk of curve progression.[2] Children who are skeletally immature are at a higher risk of their curve worsening.[2] We can estimate the child's remaining growth using the Risser stage, which grades the relative ossification of the iliac apophysis via radiograph images.[2] The Risser stage ranges from 0 to 5. A grade of 0 indicates no ossification, which is associated with a very high risk of progression.[2] A grade of 5 indicates full closure of the iliac apophysis, indicating that the patient is skeletally mature.[2] It is thought that a female has reached skeletal maturity around 2 years post menarche.[2] Males are thought to reach this benchmark when they could be shaving daily, which is thought to be around 16 years of age.[2] The degree of the patient's curve upon presentation is also important, as those with a Cobb angle over 25 degrees are thought to have an increased risk of progression.[1]

Although parents may be concerned that their child's heavy backpack or slouching has caused their scoliosis, we can assure them that these are not underlying etiologies for their child's presentation![2]

Management of Scoliosis:

Screening

Although a thorough history, physical exam, and imaging can aid us in identifying whether a child may be presenting with scoliosis, there is controversy regarding the need for regular screening. Some feel that it may be a useful tool that is part of preventative care and may reduce the need for invasive intervention if scoliosis is identified early enough.[2] Others feel that since most children with scoliosis can be managed conservatively, it may lead to a high rate of false positive referrals to specialists.[2] The Canadian Task Force on Preventive Health Care does not recommend screening for idiopathic adolescent scoliosis.[7] It is thought that since asymptomatic patients often have a milder disease course, subjecting patients to excess evaluations and interventions may not have significant impacts on their quality of life.[7]

Conservative Management

Mild cases of scoliosis, defined as a Cobb angle under 20 degrees, often don't require any active intervention.[2] Instead, serial observation and monitoring is indicated.

Curves between 30 to 50 degrees in an adolescent with remaining growth, often correlating to a Risser stage of 0 or 1, may require bracing.[2] Bracing can reduce the risk of progression during the adolescent growth spurt, although it does not necessarily eliminate the risk of surgical intervention altogether.[2] It is thought that the curve advances the most during early adolescence, during what we call the curve acceleration phase.[1] Prior to this, curves are thought to progress by 0.2 degrees per month.[1] It is quite rare to see bracing in adolescents at a Risser stage greater than 3, or in females who are 6 months post-menarche.[2] Braces must be worn for at least 13 hours per day, although the best results are found when worn for at least 18 hours.[2] This often means that children must wear them to school on a daily basis. Radiographs are done to evaluate for progression every 4 to 6 months.[2] Of note, bracing is often less effective in children with neuromuscular and congenital scoliosis.[4,6]

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Surgical Management
For congenital scoliosis, young children often require surgical interventions.[4] Additionally, if an older child has a severe curve and a high risk of progression, surgery may be indicated.[3] Surgery does not aim to straighten the spine, but rather attempts to prevent progression.[2]

The type of surgical options typically depend on the child's age and type of scoliosis. Toddlers and younger children may be eligible for growing rods.[4] This is a surgery that involves fixing temporary implants to the spine with screws to control the curvature.[4] The implants are lengthened every 6 months with subsequent surgeries.[4]

Adolescent idiopathic scoliosis and neuromuscular scoliosis are most often treated with a posterior spinal fusion.[8] In this procedure, metal is attached to the curved spine.[9] Bone grafts are then placed over the spine with the hope that they will grow with the vertebrae and allow for better positioning.[9] Recovery can take anywhere from 6 to 12 months and once fusion has occurred, further curve progression will not take place.[8] Most patients with adolescent idiopathic scoliosis do not require further bracing after this surgery.[8] Children with congenital scoliosis often undergo a similar fusion surgery, but from an anterior approach.[8] In cases of congenital scoliosis, children often require bracing during healing after their surgery.[8] While most patients are able to return to school about 3-4 weeks after spinal fusion surgery, it is often advised that they avoid sports for 2 to 6 months.[9]

Newer technologies are consistently being introduced for scoliosis management. This includes the Magnetic Expansion Control system surgery, which involves placing adjustable growing rods that are lengthened remotely using magnetic technology.[4] This avoids the need for repeat surgeries and is becoming more widespread for children between 2-10 years of age across Canada.[10]

Other Management Considerations
Having scoliosis can take a significant emotional toll on a growing child, and may influence their self-esteem.[3] For this reason, it's important to think about the child's wellbeing holistically and consider involving counsellors or including psychotherapy in the management of scoliosis.[3]

Back to Our Case:
Let's return to our clinical case. You return to Ellie with your preceptor for further evaluation. You work with your preceptor to guide Ellie through an Adam's forward bend test, where you recognize asymmetry of her trunk. Your preceptor also confirms that Ellie has not yet reached menarche. You talk to Ellie and her mother about your concern, highlighting that Ellie may have scoliosis. When Sandra questions whether Ellie's poor posture while watching TV caused her to develop scoliosis, you reassure her that Ellie's condition is most likely idiopathic in nature. Ellie is referred for imaging of her thoracic and lumbar spine, with follow up scheduled to occur in 2 weeks.

Upon return of Ellie's radiographic imaging, you note that her Cobb angle is 27 degrees. She is referred to orthopedic surgery, with the preface that she is likely to require bracing to control the progression of her curve. You note that Ellie is visibly upset with this news, and

you assure her that, if she is interested, you can put her in contact with a counsellor to talk through strategies to cope with the diagnosis. Ellie and her mother are happy to be connected to further supports as she works to manage her concerns moving forward.

Conclusion and Key Take-Aways:

To wrap up our discussion on scoliosis, let's go through a quick summary of what we covered today. Scoliosis is an abnormal lateral curvature of the spine, often categorized as congenital, neuromuscular or idiopathic in nature. Of these, adolescent idiopathic scoliosis is most common. Key physical exam maneuvers include an Adam's Forward Bend Test if there is suspicion of scoliosis. Additionally, further investigation with imaging can determine the magnitude of a spinal curvature with the Cobb angle. A Cobb angle under 20 degrees is considered mild scoliosis, an angle between 20-40 degrees is considered moderate, and an angle over 40 degrees is severe. Understanding the Cobb angle and skeletal maturity of the patient can guide management, as more mild cases can be observed or may benefit from bracing. On the other hand, severe cases that may pose risk of progression and respiratory complications often benefit from surgical intervention. Thank you for listening to this podcast about scoliosis! Stay tuned for more podcasts from PedsCases!

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