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Case Report

Severe scoliosis in an adolescent, benefits of early detection: a case report

Motssim S. Halawani*

Department of Family Medicine, University of Jeddah, Kingdom of Saudi Arabia

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***Correspondence:**
Dr. Motssim S. Halawani,
E-mail: mhalawani@uj.edu.sa

ABSTRACT

Presentations of scoliosis to primary health care physicians are not rare. Challenges may arise when it comes to screening and who require treatment once diagnosed. Idiopathic scoliosis is a three-dimensional spine and trunk deformity that is considered the most common form of scoliosis in children. It commonly does not cause symptoms and may be overlooked. However, if there was progressive deformity and it remained untreated, it may cause serious complications. Apart from medical concerns, emotional and cosmetic worries due to visible deformity may lead to psychological and social effects. Despite advances in understanding scoliosis, there are still controversies when it comes to optimal screening and treatment of this condition. This case is about a thirteen years old female who presented with intermittent back symptoms and was found to have a severe form of idiopathic thoracolumbar scoliosis. Through this case, author will be discussing different aspects of scoliosis (prevalence, screening, diagnosis, and treatment options) with emphasis on psychological support and guidance to the physician on how to overcome this challenge.

Keywords: Adolescent idiopathic scoliosis, Back deformity, Psychosocial, School screening

INTRODUCTION

Scoliosis is a term that is derived from the Greek word "skolios", which means curved.[1] It is a description when the spine is curved laterally in excess of 10 degrees measured by Cobb method on standing radiographs.[2] It is a three-dimensional deformity of the growing spine causing lateral curvature accompanied by rotation of vertebrae within the curve.[3] Many people do have some degree of curvature that is less than 10 degrees and is considered insignificant as it is within normal variation. Scoliosis is the most common spinal disorder within paediatric and adolescent age groups.[4]

There are different types of scoliosis according to various characteristics such as age of onset, severity, shape and pattern of spine deformity.[3] These are classified into two major groups which are non-idiopathic (secondary) and idiopathic scoliosis.[4] Non idiopathic is further classified into congenital, mesenchymal and neuromuscular sub-groups.[5] It may be associated with some conditions such as cerebral palsy, spina bifida, muscular dystrophies, Marfan's syndrome, osteogenesis imperfecta and many more. Red flags that raise suspicion of a non-idiopathic cause are illustrated in (Table 1).[4] Idiopathic on the other hand is classified by age of onset into infantile (between birth and 3 years), juvenile (between 4 and 10 years), adolescent (between 11-18 years) and adult scoliosis.[1] Approximately 85% of scoliosis cases are idiopathic.[6] The diagnosis of idiopathic scoliosis lies in the exclusion of non-idiopathic causes.[7] Primary health care physicians require a good understanding of the condition to be able

to differentiate the stable patient who may be observed compared to those who have severe scoliosis or are at risk of progression and require referral for treatment. Lack of such knowledge may lead to unnecessary referrals or treatments for those who have minor scoliosis and add financial burden or even marked anxiety for the patient and their family. In contrast, delayed referrals of those who are at increased risk of progression may lead to increase morbidity.[8]

Table 1: Red flags suggestive of possible non idiopathic cause.

Red flags
Sever pain
Having abnormal neurological signs
Presence of a left sided thoracic curve
Sudden rapid progression in previously stable curve
Stigmata of other clinical syndromes associated with non-idiopathic causes

This report was on a young female who presented with intermittent non-specific back discomfort for a few months. Upon physical examination and imaging, it was confirmed that she had a sever thoracolumbar scoliosis of double curve cobb angle 88 degrees in thoracic and 56 degrees in lumbar regions. The purpose of this case is to review and discuss the approach to such presentations, provide insight on the different aspects of adolescent idiopathic scoliosis (AIS) with focus on the importance of early detection and the role of psychological intervention for the patient and their family when it comes to management. Informed consent has been obtained from the patient to conduct this case report.

Idiopathic adolescent scoliosis (AIS)

Adolescent idiopathic scoliosis (AIS) is considered the most common form, affecting up to 2-3% of adolescents.[9] Approximately, 10% of diagnosed AIS progress and require medical intervention.[5] Although the male to female ratio for developing scoliosis is roughly similar for minor scoliosis, females have up to 10-fold chance of having progressive disease.[10] AIS appears in otherwise healthy individuals, the exact underlying pathophysiological cause is unknown (Table 2). Nevertheless, a genetic role is generally accepted.[3] Studies have shown a higher prevalence of developing and or having progressive scoliosis compared to the general population if both parents had the condition.[11] However, there is still lack of understanding and evidence as to the exact mode of inheritance. More studies are required to this regard and genetic testing is not recommended.

In many cases of AIS, there are no specific symptoms and it may easily be overlooked. Physical signs may be more obvious as the scoliosis progresses. School screening for early detection have long been implemented for asymptomatic school aged children at ages between 10-14.[3] Most significant curvatures can be detected at the peak of adolescent growth by age of 10. The aim of early detection is to treat and reduce progression of spine curvature during adolescent growth period.[8]

Table 2: Adolescent idiopathic scoliosis facts.

Facts
Underlying cause is unknown (idiopathic)
It affects 2-3% of adolescents
Most significant curves can be detected at peak of growth by 10 years of age
90% of thoracic curves are to the right
Rapid curve progression is an increase by 10º or more in few months
Females have 10x risk of curve progression
Approximately, 10% of diagnosed have curve progression requiring intervention
It may lead to psychological and social effects
Factors for curve progression- magnitude of curve on presentation, female gender, and future growth potential
Studies have shown curves of less than 30º at bone maturity are unlikely to progress.[12]

Screening is mainly carried out by preforming a physical examination using the 'forward bending test' also known as the Adams's bending test.[3] It is a simple test that aims to detect visible external abnormalities such as asymmetry in the spine, shoulders, scapula, waistline, and hips, lumbar or rib humps (Figure 1).[5]

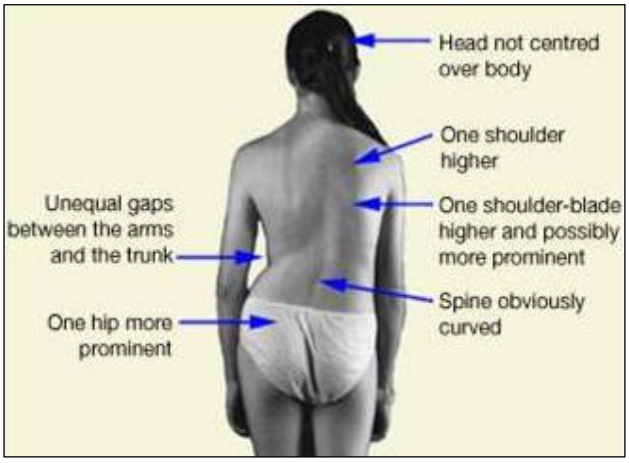

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Figure 1: Common physical signs of scoliosis.

In conjunction, the use a scoliometer to help quantify the angle of trunk rotation is of benefit.[13] Values between 5-9 degrees prompts re-examination within 6 months, but a value of 10 degrees or more indicate a need for radiograph evaluation (Table 3).[14] Both of these tools guide clinical decision making at this stage. Once an abnormality is detected, the patient is then referred to his

primary care physician for further evaluation which may include an initial spine X-rays to confirm the diagnosis.

Table 3: Referral for spine X-rays.

Indications
Symptomatic patients
Presence of obvious sever spine deformity
Presence of rib hump, asymmetry
Scoliometer measurement of greater than 10 degrees

Evaluation is by a single standing posteroanterior view of full spine X-ray with cobb angle and Risser grade measurements.[5,16]

Although school screening for scoliosis is considered a simple, inexpensive, non-invasive, easily accessible and tolerated by patients, in recent years there have been many controversies on its efficacy. Concerns included the financial burden of unnecessary referrals, over radiation exposure, patient anxiety and overtreatment in insignificant mild forms.[4,8,9] In the United States for example, screening for scoliosis is not implemented in all states and is now similar in other countries. The U.S. Preventative Services Task Force (USPSTF) current recommendations concluded that "there is insufficient evidence to support screening efficacy."[8] Saying that, various approaches to screening are implemented depending on the American state, some recommend, legislate while others do not recommend screening. The same is applied worldwide, some countries have no formal screening programs and others mandate it by law such as Japan.[17]

Scoliosis diagnosis is confirmed by preforming a standing posteroanterior spine X-rays.[16] The extent of severity is determined by the degree of the curvature with a Cobb angle measurement. That is conducted by marking the most tilted vertebra above and below the apex of the curve, drawing two perpendicular lines to the top and lower vertebra, intersecting angle is the cobb angle.[16] Mild scoliosis is when the angles are between 10 and 20 degrees. Cobb angle of more than 25 degrees or 10-20 degrees that have progressed within 6 months is considered moderate. Sever scoliosis is classified as angles of over 40 degrees or those with rapidly progressive curvatures.[3] If there were any red flags such as a left sided thoracic curve, magnetic resonance imaging (MRI) is indicated to exclude non idiopathic causes.

Factors to consider when it comes to the potential for scoliosis progression include severity of curvature on presentation (higher cobb angle), sex (female) and growth potential.[4,5] Growth potential can be assessed by the Risser grade and the Tanner stage. The Risser grade is radiographic measurement of the ossification of iliac apophysis. It is calculated on a 0-5 scale, the higher the grade meaning the less growth potential (Table 4).[5,16] The usual progression is from stages 1-5 during a two year period.[16] The time of maximum progression coincides with Tanner stages 2 and 3, that is after the onset of pubertal growth spurt.[6] The Risser grade is considered to be a better predictor of growth potential.

Table 4: Risser grade for growth potential.

Risser grade	Estimation of bony fusion at iliac apophysis
0	No ossification (0%)
1	25% ossification
2	26-50% ossification
3	51-75% ossification
4	76-100% Ossification
5	Complete ossification (100%)

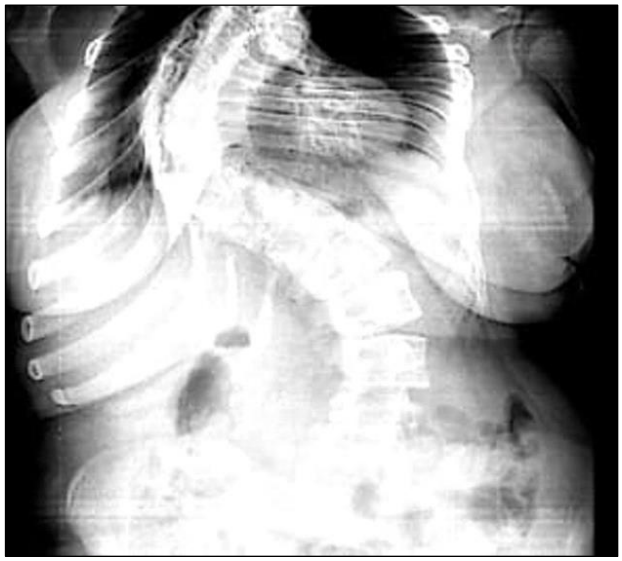
Principles of treatment of scoliosis depend on the severity and the primary goal is to prevent curve progression.[4-6,16] Spinal surgery and bracing are treatments proven to alter the curvature and natural history of scoliosis while less evidence is associated with chiropractor care and specific physical therapy. Mild forms (cobb angle of less than 20º) of non-progression deformities are usually benign and treated by physiotherapy and watchful observation. Moderate cases (cobb angle of more than 25º or have progressive curve) are treated with conservative measures which include physiotherapy and bracing. Few reported studies have shown success with spinal manipulation therapy in moderate scoliosis, however it remains controversial and lacking evidence.[18] Sever scoliosis (cobb angle of more than 40º) and those with rapidly progressive curvatures usually require surgical correction. For optimal management, psychological support for the adolescent is of vital importance through all cases of scoliosis. Depending on the severity, the psychological and social effects of the illness can be significant.[8]

CASE REPORT

Thirteen-year-old girl presented with intermittent vague back discomfort for a few months. It was described as an "ache" rather than pain, mainly in the lumbar region. However, few weeks prior to her presentation, she found it a bit uncomfortable to sleep on her back and preferred to sleep on her left side instead. While her mother inspected her back, she questioned whether there was a shoulder asymmetry and hence their presentation. The patient denied any previous injuries, neurological symptoms or other red flags.

Upon presentation, she was one-year post menarche and was morbidly obese. Her past medical history was uneventful apart from congenital hypothyroidism for which she has been taking oral thyroxine replacement since birth. She had no previous surgical history. She was physically active and participated in few sport activities. There was no family history of scoliosis or other significant medical conditions. There was no history of emotional or psychological distress. She was developing well with good performance in school, interpersonal skills and had supportive family and friends.

On examination, she seemed anxious but a healthy appearing adolescent otherwise. Vitals were all within normal limits, BMI 31. Upon inspection, her gait seemed lurched to the left side. There was asymmetry between her shoulders with more elevation on the right side and minimal waistline asymmetry. No bony tenderness. Adams bending test confirmed a prominent scoliotic spine deformity and a right thoracic hump. Neurological examination grossly intact. Cardiovascular and respiratory examination were also unremarkable.



Initial radiograph confirming diagnosis: double curve scoliosis with thoracic cobb angle 88 degrees and 56 degrees in lumbar region.

Figure 2: Spinal radiograph.

She was referred for spine X-rays to further evaluate the prominent scoliotic signs. This confirmed a right sided S shaped thoracic curve between T2-12 of 88º cobb angle and a left lumbar curve between L1-L4 with cobb angle 48º (Figure 2).

The patient and her family were very anxious about the diagnosis and the possibility of having a major surgery. As the patient's symptoms were mild and non-specific, they accounted it to muscular origin or growth related. They had no previous knowledge of scoliosis, no screening was conducted at school or self-detection awareness for scoliosis was ever heard of. They kept asking "why does she have this illness?" and "how come it was not picked up at an earlier age?". The emotional distress was evident and there was a need for ongoing counselling. This was an important aspect of the patient's management through to recovery. Due to the extent of her thoracic curvature, a respiratory complication may arise due to the anatomic disruption. Her lung function test confirmed a mild restrictive pattern. Initially she denied any respiratory symptoms but upon further questioning, she did admit of occasionally finding it difficult to breath but didn't think much of it. All her other pre-operative assessments were unremarkable. Thorough explanation of the illness was provided in the form of verbal and written documentation. Discussions raised with the patient and their family regarding the need for surgery and agreed for an orthopaedic surgeon referral. She ultimately underwent spine fusion surgery with good outcome of alignment (Figure 3). To help her recovery, she also received ongoing physiotherapy, psychological therapy together with lifestyle modification strategies to further tackle her obesity.

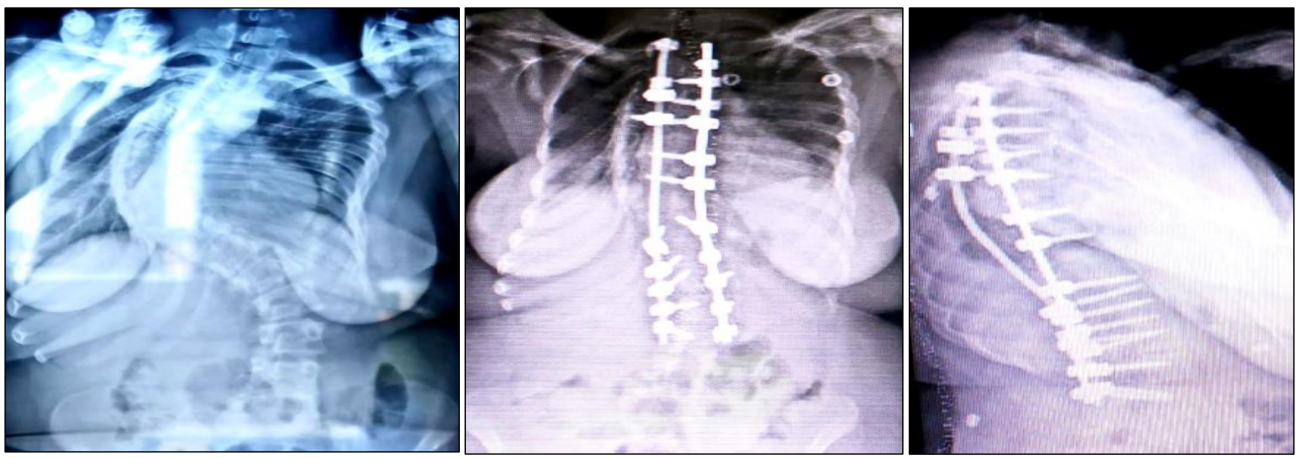


Figure 3: Pre and post-operative radiographs. A) Pre-operative scoliosis, B) Post-spinal fusion surgery.

DISCUSSION

This patient is considered one of the unlucky 10% of cases being diagnosed with AIS at such severity. It was very severe that it was clear, surgical intervention was mandated for her recovery. If left untreated, there were risks for further curvature progression, pulmonary complications and psychological illness related to the

physical deformity. Undergoing such invasive surgery is not easy and leading to her recovery the psychological impact of such journey had an enormous effect not only on the adolescent patient but also her loved ones. Careful attention to this anxiety is a skill that carries the patient to a faster recovery and improves their well-being. Without doubt, most likely she had unnoticed scoliosis features that along the last two years had rapidly progressed and only symptoms prompted her presentation. The question is, what could have been done to detect her illness earlier and yield an overall better outcome?

Thoracic cage deformity is associated with spinal deformity in AIS.[19] The higher the magnitude of the curvature, the more anatomical alterations of the thoracic cavity. This may lead to lung compression resulting in less volume and compliance. Pulmonary impairment due to spinal deformity in itself is an indication for surgery.[19] Many studies have shown a relationship between progressive sever scoliosis and decreased pulmonary function.[20] However, researchers have found more correlation between the degree of the thoracic cobb angle and pulmonary function in early-onset scoliosis and not AIS.[21] When curves are at 100º or greater, life threatening effects on pulmonary functions may occur.[22] As this patient had a thoracic cobb angle of close to 90º and due to the risk of postoperative pulmonary complications, lung function tests were included in the preoperative assessments, showing only a mild restrictive pattern.

Although invasive surgical intervention is the treatment for AIS, it carries negative psychological consequences itself.[4,9,23] AIS patients are at higher risk of developing psychological conflicts such as having negative body image, low self-esteem and mental disorders including anxiety.[23] Studies have shown that even after surgical intervention and correction of the spinal deformity, some patients experience lower body image persisting for few years.[24] Rullander AC et al, examined stress symptoms within AIS patients pre and post-surgery and its correlation with post-operative pain.[25] It has concluded that although anxiety and depression rates were higher in the pre-operative stage than 6 months post-surgery, preoperative stress correlated with post-operative pain. This signified the importance of pre-operative stress reduction for better outcomes.[25]

Adolescents after spinal surgery are unable to engage back in their normal activities for up to a year or even longer in some cases. Limitation of physical activity such as playing sports carries further anxiety for the adolescent as they feel set apart from their peers.[3] Social impairment and negative body image are concerns in the postoperative period. Although both genders are affected, females are more prone. Noonan et al reported that post-surgical treatment period, there were high rates of negative body image and depression within this group.[26]

Early detection of scoliosis is of paramount importance for a healthy future. As it is shown by this case, if this patient had screening by age of 10 and 12 years of age or if there was any awareness about the condition, early detection and use of conservative treatments could have proved a better outcome. This also might have reduced the psychological burden associated with going through invasive treatment. Despite controversies and discouragement of school scoliosis screening, this is still practiced in some areas worldwide.[27]

Proposals addressing this issue are contraindicatory. Existing recommendations in support of screening are centred on moderate evidence while recommendations against it are grounded on lower quality evidence.[27] Screening by the Adams forward test alone have shown to be inadequate due to the low specificity in certain curves and led to increased referrals, hence the addition of scoliometer measurement is considered more practical.[4,6,9,27] Moreover, Grivas TB et al, reported a more efficient screening by the use of a sitting forward-bending position, providing a better assessment of trunk asymmetry with a more stable posture demonstrating a better surface spinal deformity.[28] To reduce the cost effectiveness and inappropriate radiation exposure, suggestions have been laid out to minimize the risks. Leone A et al, proposed a two-step approach to school screening providing improved sensitivity and specificity.[29]

Awareness programs are of benefit as they provide education on the illness and target the individual and their family by familiarizing them on possible AIS signs and when to seek medical review. In Australia for example, after the screening program was abandoned, The Spine Society of Australia introduced the 'National Self-Detection Program' for Scoliosis, which is endorsed by the government and the Royal Australian College of Physicians.[30] This program works by direct distribution of a scoliosis fact sheet in the schools for the target age groups in years 5 and 7 (10-12 years of age). Moreover, in corporation to this method, further educational programs are implemented for family physicians and radiologists.[30]

CONCLUSION

AIS is the most common form of scoliosis. If diagnosed late, progressive deformity may lead to both medical and psychological complications. Practice of medicine nowadays aims at prevention more than treatment. Early detection of AIS is of paramount importance, the target group being adolescents at 10-14 years of age. It provides early conservative treatment options aiming at reducing curvature progression and better overall well-being. Optimal management regardless of the stage of scoliosis, requires an individualized approach and exploration of the psychological impact of the illness on the patient and their family. Despite controversies with school screening programs, modifications to such programs and or implementation of other awareness strategies do help.

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