

## AN OVERVIEW OF IDIOPATHIC SCOLIOSIS DIAGNOSIS AND MANAGEMENT APPROACH: LITERATURE REVIEW

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### ARTICLE INFO

**Received:**  
20 Jul 2020  
**Received in revised form:**  
27 Oct 2020  
**Accepted:**  
07 Nov 2020  
**Available online:**  
28 Dec 2020

**Keywords:** Scoliosis, idiopathic scoliosis, Diagnosis, Clinical Features, Management.

### ABSTRACT .

**Background:** Scoliosis is a deformity of the spine and trunk where the three-dimensions of the bones alignment are altered. This deformity can affect people of all ages, children, and adults, but the adolescent is the most commonly affected. Idiopathic scoliosis affects between 0.47 and up to 5.2% of the general population. Also, females are more affected by this condition with a 1.4 to 1 ratio when compared to males in mild severity scoliosis. **Objectives:** We aimed to review the literature reviewing the pathophysiology of scoliosis, clinical features, risk factors, diagnosis, and management of this disease. **Methodology:** PubMed database was used for articles selection, papers were obtained and reviewed. **Conclusion:** Scoliosis is one of the main spinal deformities seen in the general population, which may be primary or reflect underlying pathology. Thus, a physician's clinical suspicion, good history taking, and physical examination skills are paramount to help in diagnosing this deformity. The importance of early diagnosis and management is immeasurable since multiple treatment options are available and long term complications can be avoided. Nevertheless, a new basis for treatment based on new braces types and new surgical techniques are being developed and studied heavily which may provide more personalized therapy for these patients.

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**To Cite This Article:** Abdulaziz Marzouq Aloatibi, Saeed Abdulaziz Alghamdi, Dina Abdullah Asiri, Musaad Abdullah Alotaibi, Ziyad Mansour Alsadhan, Abdulallah Lafi Almoutairi and *et al.*, (2020), "An Overview of Idiopathic scoliosis Diagnosis and Management Approach: Literature Review", *Pharmacophore*, 11(6), 36-40.

### Introduction

Scoliosis is a deformity of the spine and trunk where the three-dimensions of the bones alignment are altered. This deformity can affect people of all ages, children, and adults, but the adolescent is the most commonly affected. This disease can have multiple causes, however, the most common variety is idiopathic with 80% of all cases. Many theories have been suggested and studied regarding the pathogenesis, and possible risk factors of idiopathic scoliosis. In terms of prevalence, idiopathic scoliosis affects between 0.47 and up to 5.2% of the general population. Nevertheless, a prevalence of 2 to 3% among the general population is more accepted among recent papers. Also, females are more affected by this condition with a 1.4 to 1 ratio when compared to males in mild severity scoliosis. Severe scoliosis cases are more notable in women as well, with the ratio increasing to 7.2 to 1 when compared to males. [1, 2] In this paper, we will review this disease with a focus on the idiopathic variety, discussing its pathophysiology, risk factors, clinical features, diagnosis, and management.

### Methodology

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PubMed database was used for articles selection, and the following keys used in the mesh ((“Scoliosis”[Mesh]) AND (“Diagnosis”[Mesh] OR “Management”[Mesh])). In regards to the inclusion criteria, the articles were selected based on the inclusion of one of the following topics; Scoliosis’ evaluation, management, and diagnosis. Exclusion criteria were all other articles that did not have one of these topics as their primary endpoint.

### Review

Scoliosis is a term that has many similar conditions which all will result in changes in the position and shape of the spine, trunk, and thorax. Scoliosis is a word that means curved or crooked, and it reflects the abnormal lateral spinal curvature. Overall, scoliosis can be divided into primary structural scoliosis and functional scoliosis. Functional scoliosis is any spinal abnormal curvature secondary to another extra spinal known cause. The most notable causes include the shortening of a lower limb and paraspinal muscle tone asymmetry. Nevertheless, once the secondary pathology is treated the scoliosis is usually reduced and in some cases completely disappear. Clinically, this can be notable in some patients where their scoliosis disappears in a recumbent position. As the name suggests, no underlying cause can be identified in these patients, but –unfortunately- it can still progress and get severe over time (e.g. concerning growth). Generally, idiopathic scoliosis is present as a single deformity, but in some cases, it can be a sign of a syndrome. In the latter situation, further testing and investigations may show significant other subclinical signs. [3]

In mechanical terms, the torsional deformity of the spine is seen in these patients usually these regions are joined together by a junctional zone. Many lordotic vertebrae can be seen in every region, which is usually rotated to the same side. When observing the middle sagittal plane of the central scoliotic part (known as apex), flat back (morphological lordotization) can be noticed. This is related to the secondary –relative- anterior spinal overgrowth. However, on the latero-lateral radiograph, showing the middle sagittal plane of the patient, many variables can be seen regarding the geometry of the spine. The back asymmetry and trunk deformity observed in these patients usually correlate with the existing spinal deformity, but some cases have significant discrepancies. Another important mechanical aspect is the Cobb angle which is vital to determine the severity and management of the patients. In anteroposterior radiographs in an upright position (frontal plane), the curvature is limited by two vertebrae, one is on the upper end, and the second on the lower end. The angle between these two is calculated and is the reference behind measuring the Cobb angle. Generally, idiopathic scoliosis can be classified based on the age group upon diagnosis into infantile, juvenile, adolescent, and adult. Another classification is based on angular deformity calculated by Cobb angle into low, moderate, severe, and very severe. The last popular classification of this disease is based on the location and it divides it into cervical, cervicothoracic, thoracic, thoracolumbar, and lumbar. [4]

### Etiology:

Several hypotheses have been suggested to explain this condition, and generally, now it is accepted as a multifactorial clinical problem [5-7]. Many theories suggested congenital and/or acquired problems as a possible cause behind this deformity of the vertebral structure. Moreover, many cases will have coexisting issues like sensory and balance impairment, asymmetrical structure of the brain stem, disorders of blood platelet, and collagen function defects. Thus, the role of genetic factors in the development of scoliosis has been emphasized by research and a higher prevalence of the disease in the same family line has been established. A theory of a hereditary disorder of oestrogen receptor structure and function have been suggested and researched heavily. Many researchers suggested that scoliosis may be a result of melatonin synthesis, mucopolysaccharide, and/or lipoprotein synthesis disorders. [8] However, recent studies only attribute a minimal role to melatonin in the pathogenesis of this disease, and a connection with the age of menarche in girls is hypothesized. Recent studies have attributed the association of scoliosis to calmodulin instead of melatonin. The secondary role of melatonin is due to its interaction with calmodulin which is a protein with calcium receptors. This has been hypothesized to influence the contractility of skeletal muscles and was found in higher levels in the blood platelets of scoliosis patients when compared to normal patients. Other theories involved the gene variant of matrix metalloproteinase-3 (MMP-3) and IL-6 promoter polymorphisms as possible causes of this disease. Moreover, a genetic predisposition of high BNC2 expression has been suggested and implicated in the etiology of scoliosis. Nevertheless, the exact etiology of scoliosis is not fully understood and a multifactorial origin is the mainstay opinion in regards to this topic. On the other hand, regarding the higher female ratio of severe scoliosis, relation to their more slender and narrow vertebral bodies is hypothesized. A mechanical model suggested by Schultz implicates that the lateral curve progression is proportional to the height of the spine and inversely proportional to the thickness. Thus, tall and slim spines (which are notable in females more) are more likely to buckle than thicker and shorter ones. Nevertheless, if this theory applies to the reason behind the increased prevalence of scoliosis among females is still controversial. [9]

### Risk Factors:

Genetics have been implicated as the main risk factors in this disease as discussed earlier. The estimated rate of having scoliosis in a heritability fashion is up to 38% and some papers reported a 1.5 higher odds ratio for idiopathic scoliosis if the mother had scoliosis. Additionally, some papers have associated positive family history with higher rates of curves that needed treatment. [10] Moreover, certain sports activities have been associated with scoliosis including swimming, dancing, and gymnastics, especially in adolescents. This can be associated with wrong techniques and asymmetrical load on the spine which may place the thoracic area in a lordotic position. Thus, accelerating existing scoliosis and further compromise the

spine in cases with other associated risk factors. People who have any form of joints hypermobility are associated with a higher risk to develop scoliosis. Height is another point that was associated with this condition were taller people tend to have more spinal deformities overall. However, growth in itself has been a controversial point regarding the etiology but its effect on the overall scoliosis process is certain. High BMI has been associated with scoliosis especially when paired with early menarche. On the other hand, low BMI with delayed menarche is also found to be associated. Sunlight, melatonin secretion, and leptin levels were all found to have an association with the development of idiopathic scoliosis. [11-13]

#### **Clinical Features:**

Patients usually present to the clinic with an established spinal deformity which may or may not hinder their daily tasks. A physician shall acquire a detailed history from the patient, especially their family history. Moreover, asking about possible risk factors mentioned earlier is vital since it may alter the management plan. For example, any sports activity done regularly to establish better technique, and are important. Another important example is acquiring about the maturity in the adolescent cases because patients usually have their growth spurt at puberty, which is the same period where the developing spinal curvature changes are the most rapid. The clinician shall also inquire about any deteriorations and/or complications, along with the full detailed natural history of the disease along with a full systematic review. This may reveal other symptoms that may indicate other possible general syndromes that are associated with scoliosis. Moreover, a sudden worsening of scoliosis in adulthood may have subtle causes like progressive osseous deformities and collapsing of the spine. In physical examination, the clinician may notice visible external abnormalities, these may include lateral deviation of the spinous processes, with or without asymmetry of the scapulae, shoulders, hips, and/or waistline. The physician shall assess the patient for any lateral imbalances of the trunk, any humps in the lumbar region and/or rib cage, and possible disturbances in physiologic kyphosis and lordosis. Generally, these findings are easier to detect in the early stages of scoliosis. If the clinician still not sure about the diagnosis and wants further confirmation radiological modalities are indicated. [3, 4, 14]

#### **Diagnosis:**

The clinician shall be able to suspect patients with scoliosis based on physical examination and history. The next step shall be ascertaining the diagnosis which is achievable by radiological modalities. The golden standard in cases of idiopathic scoliosis is an X-ray of the spine with multiple views. According to the Scoliosis Research Society (SRS), the diagnoses are confirmed once the patient has a Cobb angle of 10° or more, mainly in the frontal plane, plus a recognizable axial rotation. The axial rotation is measured at its maximum at the apical vertebra. Nevertheless, clinically clinicians may notice structural scoliosis can be seen even when the Cobb angle is under 10°, and the patient may deteriorate later on. [14]

Regarding screening, an x-ray is not used due to radiation exposure and its associated risks. Generally, a clinical examination suffices as a screening method for these diseases. In cases where a low degree of trunk deformity is picked up and the angle of rotation is less not enough to diagnose the case, a rescreening of the patient shall be done in 4–12 months. Recent studies advocated using the sitting forward-bending position in screening. This position reveals the real trunk asymmetry, more stable posture, and better surface-spinal deformity in comparison to standing position. It also eliminates the effect of any unequal leg length and levels the pelvis. Another method of screening is via surface topography which evaluates the external body contour. Many techniques are used, nowadays, the most common ones are raster stereography and light beam body scanning. The early diagnosis is vital to prevent any long term complication and/or worsening of scoliosis. This is more significant in adolescent girls since females tend to have higher progression rates. This may result in trunk deformities limiting their chest capacity, resulting in less exercise capacity, fitness, and impair their quality of life daily. Clinically, once the scoliosis is higher than 30° the risk of progression is increased and once it is more than 50° progression of the disease into adulthood along with complications are almost always there. [15]

#### **Management:**

The aims for management for clinicians regarding this disease vary according to the severity at the time of presentation. However, reducing the progression of deformity, restoring normal quality of life, and delay or even avoid the need for invasive management options like surgery are the main goals for treatment. As in many orthopedic diseases, there are surgical and non-surgical options, and the choice between them is based on each patient's case. The treatment in secondary scoliosis cases is based on treating the underlying problem and addressing them which will solve the scoliosis problem. [16] One of the most important non-surgical management options is physiotherapy where the patient is started on scoliosis-specific exercises. This option can be used if the patient has a mild curve, supportive therapy for braces in moderate curves, and if the scoliosis curve is higher than a certain threshold in adulthood. Mild cases are regarded as having less than 25° Cobb angle, and physiotherapy will focus on exercises specific to the three-dimensional spine position, pelvis, and rib cage which helps to maintain the curve and rib hump under control. Moreover, Scoliosis Orthopaedic and Rehabilitation Treatment (SOSORT) group recommend physiotherapy to be used in moderate cases (less than 45° Cobb angle) along with braces as supportive therapy. [8] In these cases, physiotherapy reduces the side effects of braces which include muscle weakness, rigidity, flat back, moreover, it reduces spinal stiffness, improves mobility resulting in an overall better correction and outcome. Also, this management option has been suggested to prevent further curve progression, prevent and/or treat any resultant respiratory dysfunction, treat and avoid spinal pain syndrome, improve patients' aesthetics with postural correction, and reduce the need for further treatment (especially surgery). Other treatment options of physiotherapy are

steroid and/or local anesthetic injections in the muscle, joints, or spinal canal. And in some cases where the neurological leg pain is a persistent epidural, nerve block, and/or facet injection may be indicated. These injections can provide therapeutic as well as diagnostic value in these patients. [17, 18]

Braces are used as well as a treating option in these patients, however, they may be indicated for usage once the Cobb angle is higher than 25°. The types of braces developed through the year and it related to the material used and the overall design. The main examples include Boston, Cheneau, and the more recent German Gensingen model. The latter has a reduced amount of plastic in it when compared with the others which improve the overall corrective effects. Moreover, compliance of patients with this modern type is higher due to the less time required for the brace to be worn. Due to the same reason, the overall side effects are less and thus the overall outcome is better. Thoracolumbosacral Orthosis (TLSO) is another example that has been recently developed as well. This model covers the whole or part of thoracic, lumbar, and sacral portions of the spines. There are many side effects associated with bracing in addition to the aforementioned patients report stress, discomfort, limitation inactivity, fear of injury, and overall reduced quality of life especially in young patients. Overall, the bracing treatment option is very beneficial, especially when combined with physiotherapy with success rates ranging between 70% (in Boston brace) and up to 90% (in Cheneau for example). As a result, bracing is used as the last step before surgery in patients with moderate to severe curves. [4, 8, 19]

Surgery is the definitive treatment in scoliosis and is indicated for patients once they exceed 45° Cobb angle. The surgical approach in such correction operation has developed through the year, with the posterior, anterior approach, and vertebral body stapling and tethering as the mainstay techniques. While anterior approach surgery was developed more recently compared to posterior but it has a higher risk of compromising the pulmonary function. Moreover, the thoracoscopic discectomy process is more difficult, with higher corrective forces and higher fusion rates of the screws constructed. [20] The posterior approach went through many modifications and improvements over the years. They used rods with anchors or hooks, but recently the pedicle screws are more in use especially in thoracic and lumbar curves. Pedicle screws offer a better pullout strength, and better three-column fixation, thus ensuring improved stability and better control in three panels. Both of the aforementioned techniques can be combined in special cases. [21] Vertebral body stapling and tethering are the most recently developed techniques aiming to modulate the vertebral body growth on the side of the curve convexity. Stapling involves the physical endplates and the discs next to the vertebra which results in modulation of the growth plate. This surgery is indicated in less severe cases where the curve is less than 35° (if thoracic curve) or 45° (if lumbar curve) and did not comply with bracing. Tethering is indicated in more severe cases with Cobb angle ranging from 30° to 65° and 50% curve reduction in bend X-ray. Nevertheless, the rates of re-operation in these cases are quite high with some papers reporting rates exceeding 50% within two years of surgery. Overall, it is accepted that patients with a Cobb angle of more than 45° are a candidate for surgery. However, the selection of the operative approach and the instruments used will depend on the curve location, magnitude, flexibility, and sagittal alignment. In very severe cases, which present with large and rigid curves the anterior-posterior approach is used and shows the best results. [22-24]

When comparing surgery to bracing, there is no significant difference in the quality of life in the long term. And in both therapies the curve did not increase significantly, moreover, the surgical complications rate was not high. Surgical patients had better lumbar muscle endurance and mobility which resulted in better overall mobility and physical function. Also, patients treated with bracing reported higher rates of low back pain in the long term. Finally, the vital capacity and respiratory function were significantly better in both types of treatment. [16, 25]

## Conclusion

Scoliosis is one of the main spinal deformities seen in the general population, which may be primary or reflect underlying pathology. This disease does not have an immediate pathology and the etiology is multifactorial and it can even be a part of another syndrome. Thus, a physician's clinical suspicion, good history taking, and physical examination skills are paramount to help in diagnosing this deformity. The importance of early diagnosis and management is immeasurable since multiple treatment options are available and long term complications can be avoided. Nevertheless, a new basis for treatment based on new braces types and new surgical techniques are being developed and studied heavily which may provide more personalized therapy for these patients.

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