REVIEW

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Idiopathic scoliosis and epidemiology: a narrative review

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ABSTRACT

Idiopathic scoliosis is a problem that affects a great number of patients of all ages causing pain, imbalance and prohibiting the development of the angular curve. Idiopathic scoliosis causes a number of alterations in the spine that can result in cardio- respiratory failure and mobility issues. In this review, we study the latest literature publications regarding the starting age, the types of scoliosis and the pathogenesis of the disease. We also analyze the epidemiological factors of the syndrome according to the current literature.

KEY WORDS: Idiopathic Scoliosis, Epidemiology, Pathogenesis

Introduction

Scoliosis is the pathological curvature of the spine. The primary scoliosis is not restricted to the curvature (left or right) but is also combined with a slight rotation of one vertebra towards the other. The main characteristic of scoliosis is the angle that the spine shows in the frontal plane in combination with the torque. The back of a patient with scoliosis resembles an S or C due to the curvatures. However, the most common type of Scoliosis is the idiopathic (the exact cause remains unknown) [2], [3].

Idiopathic scoliosis can be encountered in infants and in early childhood, but the majority of the cases occur in children of 10 years old until their full skeletal maturation. Even if the certain cause of scoliosis is unknown in the majority of cases, it is well known that can be hereditary.

If scoliosis is evident at birth (congenital scoliosis), it is likely to be accompanied by other developmental abnormalities such as malignancies of the chest wall, malignancies in the urogenital tract and cardiovascular comorbidities. It is also possible that scoliosis can be accompanied with a neuromuscular disorder (neuropathic scoliosis in children with poliomyelitis or cerebral palsy). Other diseases that could cause scoliosis are bone malignancies, tumors in the spinal cord and intervertebral hernias [5], [6].

Starting Age

There are 4 types of idiopathic scoliosis when we classify the syndrome according to age. Firstly, the infant scoliosis (0-3 years old). Secondly, the childhood scoliosis (4-9 years old). Thirdly, the adolescent scoliosis (10 years old after the convergence of the pineal glands) and finally the adult scoliosis. The most commonly occurring idiopathic scoliosis is the adolescent scoliosis. Furthermore, the term "early onset scoliosis" is used to describe the pathological situation in which the spinal curve is fully developed before the age of 5 while the term "late-onset scoliosis" is used to describe the pathological situation when the spinal

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curve is fully developed after the age of 5 years old [15].

The size of the Curve

The size of the spinal curve is measured using the Cobb method, in typical front-lateral spinal x-rays. The measurement of the angle with the Cobb method is based on the vertebra with the greatest inclination and the vertebra with the greatest curve. The accuracy of the method is 3-5 degrees. In a measured Cobb angle of 10 degrees we cannot characterize the spine as a spine with scoliosis according to the literature. A scoliosis of 11-25 degrees is characterized as slight scoliosis, 25-45 degrees as medium scoliosis and a measured angle greater than 45 degrees allows us to characterize the scoliosis as severe scoliosis. The Cobb angle is directly proportional to the rotation of the vertebra and usually increases with the increment of the surface inclination along the spine. The appropriate therapy is selected according to the Cobb angle and the child's state of skeletal maturity (Risser Sign) [8].

Type of Curves for non-surgical Treatment

In order to classify the type of curvature we use two types of classifications: the Lehnert - Schroth and the Rigo methods [30], [37]. The Lehnert – Schroth classification is based on the three "curves and blocks" standard. More specifically these are the shoulder, the torso and the lumbar-pelvic block that divert from one another in the frontal level. The Lehnert-Schroth classification further divides the lumbar – pelvic block in lumbar and pelvic blocks to assess their diversion and their turning points. We must also underline that the pelvic block represents the reversed curve and is defined as the 4th curve [37].

In the Rigo classification we define certain corrections that must be made in order to successfully construct the spinal braces based on the clinical and x-ray findings. Radiology criteria are used to discern 5 types of curves: non – balanced chest curve, double curve (4 curves), balanced chest curve and false double curve (non-3, non-4) and single long curve [30].

Types of Curves for the surgical treatment of scoliosis

The King-Moe classification defines 5 types of curves (the S curve, the S curve where the curves are crossed by the central sacral vertical line, the main thoracic curve, the long thoracic curve with C shape and the double thoracic curve) [21]. However, there are also two additional types of classification: the Lenke classification (defines 6 types of curves) [17] and the Peking Union Medical College classification (defines 3 types of major and 13 types of secondary curves) [28].

All three classifications provide further details about the secondary characterization of the curves based on static and/or dynamic x-rays thus allowing the clinicians to better design and establish the levels of spinal fusion on the curve.

The King-Moe classification is rarely used today, however, we must mention it in our review, because there are publications that uses this certain classification to describe surgical operation's results, based on Harrington instrumentation. The classification that is used with greater frequency is the Lenke classification [17].

In Lenke classification, a modifying factor of the lumbar curve is added to the six types of curves. This modifying factor is defined by the position of the CVSL in the apical vertebra of the lumbar curve (Figure 1). Lenke introduced three lumbar modifiers [17]:

- Modifier A: when CSVL passes between the pedicles of the apical vertebra
- Modifier B: when CVSL touches the pedicle of the apical vertebrae
- Modifier C: when CVSL does not touch the apical lumbar vertebrae

In the Lenke classification a modifier of the thoracic (plain) profile is also added. The thoracic curve is measured between the fifth and twelfth thoracic vertebrae and is defined as +(plus) when the curve is > 40 degrees, N (normal) when the curve is measured between 10 and 40 degrees and – (minus) when the curve is measured <10 degrees [17].

The Lenke curves of type 1 to 5 can be assessed with anterior or posterior approaches. Type 2, 3, 4 and 6 can be assessed with posterior approach. In patients with lumbar modifiers A or B the selective thoracic fusion is recommended, in order to preserve the mobility of the lumbar vertebrae [21], [17], [18], [28].

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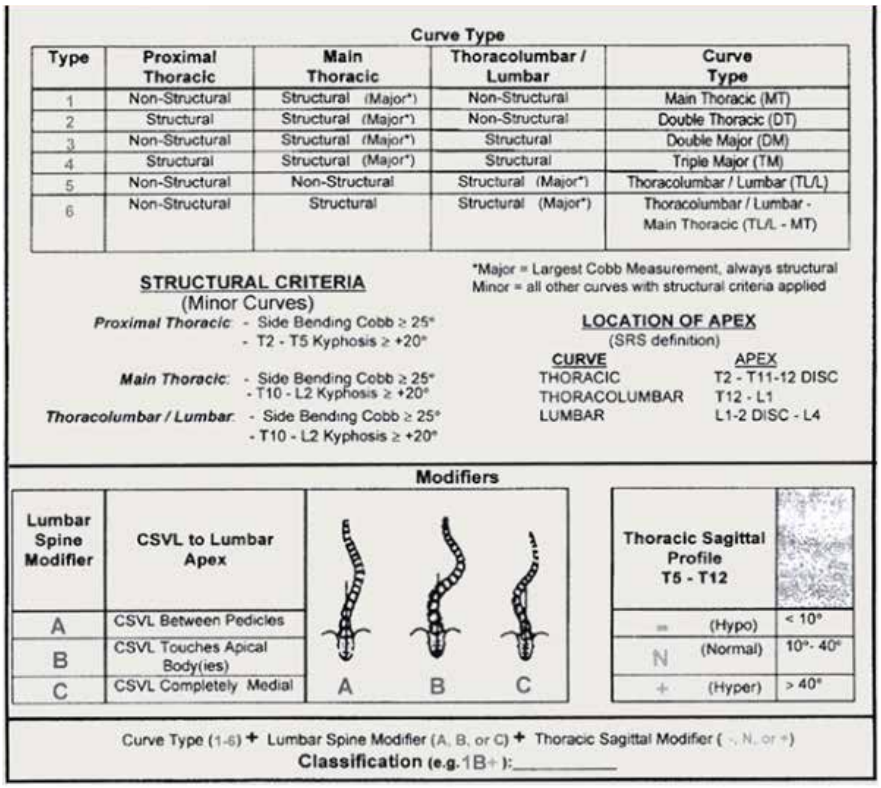

Figure 1. *The Lenke classification of scoliosis.*

The three-dimensional nature of the curve

In recent studies, stereo radiographic images were provided to measure the distortion of the lumbar curve. The measurements that were used for that analysis were the Cobb angle, the upper vertebra, the swivel axially of the upper vertebra and the orientation of the upper vertebra in relation to the sagittal plane. These parameters allow us to further estimate the distortion of the spine in three plains (the sagittal, the frontal and the transverse). New technologies such as the EOS system, that is able to provide 3D reconstruction of the spine can establish a three-dimensional classification of the spine and new types of treatment strategies [25]. Existing classifications which are based on the three dimensional morphology of the curve are: The Poncet Classification which is based on three different standards of geometrical torque in the main curve [27]. Furthermore, the Negrini classification which is based on the direction, the dislocation and the phase of the curve [25]. Moreover, the Stokes classification [35] and the Illes classification [14].

The upward trend of the imaging technology will allow the scientific community to establish a three dimensional classification based on the geometrical morphology of the main curve [25], [35], [32], [14].

Epidemiology

The prevalence of idiopathic scoliosis is directly related to geography. The syndrome is more widespread in countries located in higher northern geographical latitude in comparison to countries located in smaller geographical latitude [13].

In a meta- analysis including 36 studies from 17 countries, the global aggregated prevalence of idiopathic scoliosis of the spinal cord with Cobb angle >10 degrees was 1,94% (95% CI: 0,98-1,70%) [9].

For over 50 years, the question regarding the cause of idiopathic scoliosis still exists, but, unfortunately, there is no clear answer. The causes are multimodal including genetics, hormonal, metabolic, biochemical, neurological and asymmetric development [16].

The genetics as a cause of idiopathic scoliosis are commonly accepted by the scientific community. Studies show that 11% of the first-degree relatives

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are affected but relatives of second and third degree are also affected by 2,4% and 1,4 % correspondingly. Similar percentages have been observed in identical twins [29], [39], [31], [7], [20]. In a recent study it was concluded that adolescents that had a relative with idiopathic scoliosis had a 51% chance to develop idiopathic scoliosis, which further strengthens the genetically modal of the disease. A great effort is being made to identify specific genes responsible for the syndrome [10], [12].

Idiopathic scoliosis affects the 0,2%-0,6% of the general population and a recent study proved that the percentage of idiopathic scoliosis in adolescents is between 0,47 and 5,2%. However, the 70-90% of all the scoliosis cases is classified as idiopathic scoliosis. The prevalence of scoliosis is also greater in women than in men [1], [4], [34].

Another factor of the development of idiopathic scoliosis could be the interaction of genetic and environmental factors, on the grounds that different predisposition to genes can modify or deteriorate further the progress of the disease. In the starting phase, genetics possibly play a more important role than the environmental factors, while the environment contributes further in the deterioration of the curve [22].

Fat body distribution is also likely to contribute to the natural history of the disease, as a decrease in fat that leads to a decrease in the baseline levels of leptin. This fact can also lead to an abnormal development of the central nervous system. This theory links the decrease in the leptin levels with the start of the abnormal neural and bone development across the neural axis. In early developmental stages this leads to alterations in the normal development of the spine in the sagittal plain [33].

Conclusions

There is a vast amount of literature available for the study of idiopathic scoliosis regarding patients of all ages. The knowledge that is associated with idiopathic scoliosis is changing along the scientific progress. However, the current literature mainly focuses in defining the molecular and genetic mechanisms for the pathogenesis of the disease. With this regard, it is possible that in the future, effective preventative methods will be developed to inhibit the development of idiopathic scoliosis at an early stage. Meanwhile, the comprehension of physiology, diagnosis and management of the syndrome will help the scientific community and the clinical physicians to reduce the prevalence of idiopathic scoliosis and its associated complications in the overall population.

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