Grabala Pawel. Idiopathic scoliosis in children and adolescents – a literature review. Journal of Education, Health and Sport. 2019;9(6):212-217. eISNN 2391-8306. DOI http://dx.doi.org/10.5281/zenodo.3241930
http://ojs.ukw.edu.pl/index.php/johs/article/view/7011

The journal has had 7 points in Ministry of Science and Higher Education parametric evaluation. Part B item 1223 (26/01/2017).
1223 Journal of Education, Health and Sport eISSN 2391-8306 7

The authors declare that there is no conflict of interests regarding the publication of this paper.

Received: 05.05.2019. Revised: 25.05.2019. Accepted: 09.06.2019.

Idiopathic scoliosis in children and adolescents – a literature review

Grabala Paweł

https://orcid.org/0000-0001-7059-2935
Medical University of Bialystok
The Faculty of Medicine
Department of Pediatric Orthopaedic and Traumatology
University Children's Hospital
Waszyngtona 17
15-276 Bialystok
Poland
E-mail: pgrabala@wp.pl

Background: Scoliosis is defined as a lateral deviation from the normal vertical line of the vertebral column. Idiopathic scoliosis is a term for a disease entity that is a three-dimensional curve of the vertebral column. Idiopathic scoliosis is extremely rare in the neonatal and the early childhood period, and its prevalence among adolescents up to 15 years of age is estimated between 1% and 2%. The consequences of the development of the disease may include reduced mobility of the vertebral column, spinal pain, limitation of the vital capacity of the lungs and in the psychological aspect – problems with accepting one's appearance, low self-esteem, leading to the development of depressive disorders.
Material and methods: Analysis of available literature, articles in the Google Scholar database and PubMed using keywords: scoliosis, idiopathic scoliosis, pediatric deformities.
Results: Knowledge of natural history, the prevalence of idiopathic scoliosis and the physiology and pathophysiology of the growth of the vertebral column are indispensable factors to determine if a treatment is necessary, and if so, which one. The occurrence of larger curves of above 30 degrees is estimated at between 0.15% and 0.3%. Weinstein created a table of calculations that show a decreasing prevalence with increasing curve size. The importance of these tests in the assessment of curve is that small scoliosis is common, but greater curves are much less common. Less than 10% of children with a curve value of 10 degrees or more require treatment.
Conclusions: Thoracic idiopathic scoliosis greater than 50 degrees in adult age may gradually deteriorate and potentially reduce lung function. Lumbar curves, especially those greater than 50 degrees, can also develop in adult age and lead to osteoarthritis. Curves between 30 and 50 degrees may demonstrate a slight progression in adult age, but are

generally stable. Therefore, even if cosmetic factors are not considered, it is justified to aggressively treat a child with a significant deformity of the vertebral column.

Keywords: idiopathic scoliosis, pediatric deformities, adolescents

Introduction

Scoliosis is defined as a lateral deviation from the normal vertical line of the vertebral column. Idiopathic scoliosis is a term for a disease entity that is a three-dimensional curve of the vertebral column [3,8]. Deformity refers to the coronal, sagittal and horizontal plane, and coexists with vertebral rotation [10, 13].

It is formed in the developmental age in healthy children, with the diagnosis of no other cause of deformity. According to the Scoliosis Research Society (SRS), idiopathic scoliosis is diagnosed when the Cobb angle exceeds 10 degrees in the X-ray image. Deviations below 10 degrees are considered as spinal asymmetries [19].

Epidemiology

Idiopathic scoliosis is extremely rare in the neonatal and the early childhood period, and its prevalence among adolescents up to 15 years of age is estimated between 1% and 2%. Degenerative lesions cause the occurrence of scoliosis in adults over the age of 25 of 8%, and in the age range of 60 – 90 years it increases to 68% [3, 4, 8, 10].

Etiology and pathogenesis of idiopathic scoliosis

As this disease is defined as "idiopathic", in numerous different studies the attempts have been made to demonstrate an increase in the prevalence within a family and between identical twins, but no obvious indicator was found. Modern research assumes a combination of many factors, from growth disorders to dysfunction of tissues associated with the vertebral column (muscles, ligaments, disc complexes) [2, 3, 5, 8]. However, one obvious reason has not been established yet. High prevalence of familial scoliosis has been demonstrated (6.9% to 11.1% for first-degree relatives). It is suggested that both the dominant and multifactorial pattern of inheritance has a genetic involvement in the AIS [9, 10, 12]. However, this is not confirmed by all studies.

Currently, research aimed to identify genes that cause scoliosis and its progression is conducted. The DNA genome originated from families with apparent autosomal inheritance dominating in the teen population in terms of the association with the disease, is analysed. Familial analysis may enable "tracking" of the causative genes, such as the CHD7 gene associated with idiopathic scoliosis, thus providing a better view of the aetiology of idiopathic scoliosis [17-19].

Classification of idiopathic scoliosis

According to the recommendations of the Scoliosis Research Society, idiopathic scoliosis is classified according to the age of the patient (at the time of diagnosis). Infantile scoliosis occurs in children up to 3 years of age, the juvenile one from 4 years of age up to 10 years of age, and the adolescent one between 10 years of age and reaching bone maturity. Some authors distinguish adult idiopathic scoliosis, which appears after the end of growth or scoliosis arising due to the ageing of the vertebral column [12, 13, 19].

It is necessary to distinguish between early-onset and late-onset scoliosis, because these deformities may affect the development of the cardiopulmonary system. In the simplified classification proposed by Sponseller, idiopathic scoliosis of early onset, formed before the age of 10 and late onset, formed after the age of 10, are distinguished [11, 19].

Infantile idiopathic scoliosis

Infantile idiopathic scoliosis is a structural lateral curve of the vertebral column occurring in patients under 3 years of age. James, who was the first to use the term "infantile idiopathic scoliosis", observed that the curve occurring before the age of 3 is more frequent in boys than in girls and is mainly related to the thoracic region curved to the left side [11, 12, 15]. Infantile idiopathic scoliosis is more common in Europe than in North America. Early childhood curves usually progress faster, are often difficult to treat and can lead to a significant deformity and impairment of the respiratory system, but may also undergo a spontaneous correction with age without requiring any treatment. Scoliosis with a curve size in the range of 10 – 20 degrees, usually progress in 10 – 20 % of cases. In contrast, curves in the range of above 20 degrees undergo progression in the adolescence in 70% of cases [1, 2, 3].

Juvenile idiopathic scoliosis

Juvenile idiopathic scoliosis appears between 4 and 10 years of age. There are many patterns of deformation, but usually the convexity of the thoracic curve is directed to the right. Juvenile idiopathic scoliosis accounts for 12-21% of cases of idiopathic scoliosis. In children aged 3-6, the prevalence ratio depending on the sex is 1:1. This ratio increases with age to the disadvantage of girls, in the case of children aged 6-10 years it equals 4:1, while in the case of ten-year-old children it is 8:1. The development and progression of curve in children with idiopathic scoliosis are usually slow or moderate until the growth acceleration during the puberty spurt [1-5]. Most of the childhood spinal curves are convex curves directed to the right or double curves in the thoracic region, which closely resemble those occurring in adolescent idiopathic scoliosis. A small number of patients with juvenile idiopathic scoliosis have curves in the thoracolumbar or lumbar region [1, 4, 8, 19].

Adolescent idiopathic scoliosis

Adolescent idiopathic scoliosis is the most common deformity of the vertebral column in children between 10 and 17 years of age, with a higher prevalence in females than males. The prevalence of AIS is reported to be between 0.5 and 5 per 100 adolescents. However, only 5% of these patients have a curve of more than 10 degrees and progressive to 30 degrees [12, 13, 16].

Scoliosis in adults

Deformities of the vertebral column occurring after the period of growth are most often a consequence of idiopathic scoliosis formed during the developmental period. However, adult scoliosis developed de novo due to ageing of the osteoarticular system, style and quality of life, classified as degenerative scoliosis, may also occur [8-11, 18, 19].

General recommendations for the treatment of idiopathic scoliosis

The current recommendations for the treatment of idiopathic scoliosis are as follows:

- curves from 1 to 25 degrees – observation,
- curves from 25 to 45 degrees – back brace treatment and rehabilitation,
- curves above 45-50 degrees – surgical treatment
- in children below the age of 10, surgery, when the curve exceeds 60 degrees despite aggressive conservative treatment [14 - 18].

Idiopathic scoliosis and its effects on other organs

In childhood, not only the volume of the lungs increases, but also the number of pulmonary alveoli and arterioles grows, and what is more the vascular pattern changes. From the infancy to approx 4 years old, The number of pulmonary alveoli in the bronchial tree

increases tenfold, to approximately 7-8 years of age they do not reach full development. Scoliotic deformity limits the space necessary for proper lung growth, and thus, children who have a significant curve before the age of 5, usually suffer from limiting dyspnea or cardiorespiratory failure.
Other consequences of the development of the disease may include reduced mobility of the vertebral column, spinal pain, limitation of the vital capacity of the lungs and in the psychological aspect – problems with accepting one's appearance, low self-esteem, leading to the development of depressive disorders [1 - 4].

Growth of the vertebral column and natural history of idiopathic scoliosis

Knowledge of natural history, the prevalence of idiopathic scoliosis and the physiology and pathophysiology of the growth of the vertebral column are indispensable factors to determine if a treatment is necessary, and if so, which one. The occurrence of larger curves of above 30 degrees is estimated at between 0.15% and 0.3%. Weinstein created a table of calculations that show a decreasing prevalence with increasing curve size [1 – 7].
The importance of these tests in the assessment of curve is that small scoliosis is common, but greater curves are much less common. Less than 10% of children with a curve value of 10 degrees or more require treatment.

After the diagnosis of scoliosis in the child, the curve must be evaluated for the probability of progression. Most authors define progression as an increase of 5 degrees or more, measured using Cobb measurements, during two or more visits. It is not known whether this progression will continue and what the final deformity curve will be like. Spontaneous improvement may occur in 3% of adolescents with idiopathic scoliosis, most of whom have curves below 11 degrees [1- 5]. It has been found that some factors may be related to the progression of the curve. Progression is more likely in girls than in boys. Deformities of the vertebral column in children are most noticeable during periods of growth. Therefore, it is necessary to predict the potential of the vertebral column growth and curve progression. The period of rapid growth was called the peak height velocity (PHV), which is calculated based on changes in the patient's height measurement over time and equals approximately 8 cm per year for girls and 9.5 cm per year for boys. There are two periods of rapid growth in children: the first from birth to the age of 3 and the second during adolescence. The duration of the growth spurt during adolescence can be determined by monitoring the growth velocity. The vertebral column grows unevenly, that is, during the growth spurt, the thoracic spine grows 1.2 cm per year, unlike the lumbar spine, which grows 0.6 cm per year [1 - 5]. Measuring, estimating and monitoring changes at sitting height (chest height) during growth spurt may be helpful in planning treatment. The presence of scoliosis may intensify the multifaceted distortion during growth spurts. Other related conditions, including neuromuscular disorders, that may affect the course of the curve, should also be taken into account during monitoring and treatment. The prognosis of growth during adolescence is based on physical and radiographic examinations. The growth coefficient graph is an ideal way to monitor growth, but realistically it is not always possible to obtain. Therefore, the physician should consider several different methods to assess growth and bone maturity. There are numerous methods for assessing the skeletal maturity. The most widespread, most frequently used, although not entirely reliable test is the Risser sign [8, 10, 12 - 15].

The Risser sign is a method based on the degree of ossification of the wing of ilium, with stages from 0 to 5, corresponding to the sequential ossification of the the iliac crest from the abdominal to the dorsal part. Stage 4 reflects the end of the growth of the vertebral column, and 5 reveals the connection to the ilium. Alternatively, radiograms of hands can be obtained to assess the skeletal maturity, without the need to expose the pelvis to radiation. Evidence of

progression should be obtained before the growth spurt, unless the curve is greater than 30 degrees when the juvenile patient is first observed [1, 15, 19].

The frequency of progression decreases with the age of the child. Double (bipartite) curves are at greater risk of progression than single ones, but single thoracic curves are more progressive than single lumbar ones. The frequency of progression also increases with the size of the curve. Bunnell estimated that the risk of progression for the 20 degree curve is approximately 20%, and the risk for the 50 degree curve is 90%. Lonstein and Carlson have developed a nomogram predicting the progression of the curve when the patient is first seen.

Important considerations in the surgical treatment of patients with juvenile idiopathic scoliosis are expected spine height loss and limited thoracic wall growth, as well as lung development after spinal fusion. Another important issue is the phenomenon of crankshaft. With stable posterior connection, continuous growth of the anterior portion of the vertebral bodies causes the vertebral body and discs to bulge sideways towards the convexities and rotate at the posterior stabilization, resulting in a loss of correction, increased vertebral rotation and a relapse of ribs kyphosis. Posterior fusion of the vertebral column stops the longitudinal growth in the posterior parts, but the vertebral bodies continue to grow in the forward direction [1 - 6].

Researchers demonstrated the effect of progressive curves in adults with untreated scoliosis. Five main problems in the natural history of untreated adolescent idiopathic scoliosis in adults are: back pain, reduced pulmonary function, psychosocial effects, increased mortality and progression of curve [1 – 4, 8].

A very important element before deciding about surgical treatment, is to determine if pain is associated with scoliosis . Breathing impairment is usually a restrictive lung disease and is only observed in thoracic scoliosis. There are reports that significant limitations of forced vital capacity do not occur until the curve approaches 80 – 100 degrees. Death in adult patients with idiopathic scoliosis also appears to be associated with thoracic curves greater than 100 degrees and pulmonary hypertension. In the study with a 40-year follow-up period, the mortality rate was 15%, but only in one patient pulmonary heart disease secondary to scoliosis was the cause of death [3, 4, 8].

The psychological effect of scoliosis has been investigated by many authors. Dissatisfaction with the appearance is often correlated with the deformation of the ribs, thorax and kyphosis. Middle-aged patients better tolerate psychological effects of scoliosis than teenagers, but the majority of adult patients search for a treatment for untreated adolescent idiopathic scoliosis and are most interested in the cosmetic aspects of this disease.

Despite the seemingly visible deformities caused by long-term untreated scoliosis, the majority of subjects do not have significant psychological difficulties compared to people without scoliosis.

Summary

Thoracic idiopathic scoliosis greater than 50 degrees in adult age may gradually deteriorate and potentially reduce lung function. Lumbar curves, especially those greater than 50 degrees, can also develop in adult age and lead to osteoarthritis. Curves between 30 and 50 degrees may demonstrate a slight progression in adult age, but are generally stable. Therefore, even if cosmetic factors are not considered, it is justified to aggressively treat a child with a significant deformity of the vertebral column.

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