Idiopathic Scoliosis

ABSTRACT

Scoliosis refers to curves exceeding 10 degrees observed through posteroanterior direct radiography. In fact, the diagnosis for idiopathic scoliosis is accepted to exclude already available causes. The aim of this paper was to review the etiopathogenesis, classification systems and the treatment management of idiopathic scoliosis. A search in the National Library of Medicine (PubMed) database using the key words ‘idiopathic’ and ‘scoliosis’ was performed. For the literature review, papers concerning the etiopathogenesis, classification and treatment were selected among these articles. A search in the National Library of Medicine (PubMed) database using the key words ‘idiopathic’ and ‘scoliosis’ yielded 4518 articles published between 1947 and 2013. The main hypothesis put forward included genetic factors, hormonal factors, bone and connective tissue anomalies. King, Lenke, Coonrad and Peking Union Medical College (PUMC) classifications were the main classification systems for idiopathic scoliosis. Exercise, bracing and anterior, posterior or combined surgery when indicated are the choices for the treatment. Every idiopathic scoliosis case has to be managed to its own characteristics. It is the post-operative appearance that the surgeons are perhaps the least interested but the adolescent patients the most interested in. The aim of scoliosis surgery is to restore the spine without neurological deficit.

KEYWORDS: Idiopathic, Scoliosis, Review

INTRODUCTION

Scoliosis refers to curves exceeding 10 degrees observed through posteroanterior direct radiography (16). Scoliosis forms a complex curve that leads to deformities not only in the coronal plane but also in all three planes, which is caused by the self-rotating movement of the spine. Idiopathic scoliosis corresponds to around 80% of structural coronal deformities (35). In fact, the diagnosis for idiopathic scoliosis is accepted to exclude already available causes. It is divided into three sub-groups according to the age as infantile (age 0-3), juvenile (age 4-9) and adolescent (age 10 up to maturity).

While the incidence rate for curves of 10 degrees and above varies between 1 and 3 %, the incidence rate decreases to 0.15 to 0.3% when it comes to curves of 30 degrees and above, which require treatment. The female/male ratio is 1.4/1 in curves of 10 degrees and above, whereas it increases to 5/1 for curves of more than 30 degrees (34).

A search in the National Library of Medicine (Pubmed) database using key words ‘idiopathic’ and ‘scoliosis’ yielded 4518 papers published between 1947 and 2013. For the literature review, papers concerning the etiopathogenesis, classification and treatment were selected. A total of 348 papers that did not comply with the literature review criteria were excluded.

ETIOPATHOGENESIS

A search using the key words ‘idiopathic’, ‘scoliosis’ and ‘etiology’ yielded 1211 articles, and this indicates that causes of scoliosis have not yet been definitely established and the search for causes are still continuing. The main hypothesis put forward includes genetic factors, hormonal factors, bone
and connective tissue anomalies and autonomous nervous system dysfunctions. All the above mentioned causes are interconnected with mutual effects on each other.

**Genetic-Hormonal Causes**

A search using the key words ‘idiopathic,’ ‘scoliosis’ and ‘gene’ yielded 166 papers. Articles regarding the genetic causes (COL1A1, COL1A2, COL2A1, FBN1, elastine) of Marfan or Ehler-Danlos triggered by disorders of the extracellular matrix, neuromuscular disorders (Friedreich Ataxia, muscular dystrophies, osteochondrodystrophies (osteogenesis imperfecta, neurofibromatosis), and disorders that co-exist with segmentation anomalies of the vertebrae were excluded from the literature review, leaving 86 articles to be included.

Idiopathic scoliosis observed in identical twins indicates that the causes may be of genetic origin (34). Studies aimed at determining how the disorder is handed down to generations have reported that idiopathic scoliosis is not inherited in an autosomal dominant or X-linked dominant manner except for a couple of familial forms (10, 20, 28, 30, 83, 85, 105). It was discovered that idiopathic scoliosis has multi-factorial hereditary characteristic and multiple genes are affected for the development of the disease. Genes found to be linked with idiopathic scoliosis include SNTG1 (gamma-1-syntrophin) in 8q11.22, ESR1 (estrogen receptor-1) in 6q25.1, CHD7 (chromodin helicase DNA binding protein 7) in 8q12.1 (7, 104). Matrilin-1 is a non-collagenous protein secreted by chondrocytes. Matrilin-1 (MATN1) protein ensures the distribution of chondrocytes within the growth plane. Montanaro discovered that MATN1 single nucleotide polymorphism in 1p35 led to a disorder of chondrocyte distribution and caused scoliosis (61), whereas single nucleotide polymorphism at the Matrilin receptor level caused idiopathic scoliosis MTNR1B in 11q21-q22. Furthermore, an association between IS and Calmodulin1 (CALM1) gene polymorphisms has been reported (106). Esposito et al. showed that steroid binding protein polymorphisms were effective in IS development (26). IS related zones were spotted in 6q, 10q and 18q, 17p11.2, 19p13.3, 8q11, Xq23-26.1, 17q13.2-q21.3, 17q25.3-qtel chromosomes (4, 7, 14, 38, 58, 68, 87, 103).

ACTB and GAPDH β-actin protein account for the protective molecules of the cell that are responsible for glyceraldehyde-3-phosphate-dehydrogenase, and glycolysis secretion (90). Lack of protective molecules was found in familial idiopathic scoliosis (97).

Lebouf et al. indicated that estrogen had some impact not on the development of idiopathic scoliosis but on the degree of the curve. Later on it was discovered that such impact was more due to the insufficiency at the receptor level rather than decreasing levels of hormone secretion (53). Core receptors in bone cells, namely estrogen receptor alpha and beta, were shown to have an impact on AIS curves (29, 77).

The melatonin hormone causes an increase in osteoblasts and a decrease in osteoclasts. Thillard reported scoliosis development after removal of the pineal glands (95). Decreasing melatonin levels were held responsible (6, 23, 51). Melatonin receptor-2 (MT-2) levels were observed to be lower in AIS patients with accompanying lower melatonin responses. The co-effect of 17-beta-estradiol and melatonin on osteoblastic cells obtained from AIS patients was shown to be higher than their individual effects (47). Wang et al. indicated that tryptophan hydroxylase 1 (TPH1) was one of the enzymes playing a role in melatonin synthesis. Polymorphism in the TPH-1 gene results in decreased levels of melatonin, which in turn causes idiopathic scoliosis development (98).

Calmodulin is the secondary indicator for melatonin and has an impact on muscle contraction. Relatively higher levels of calmodulin were detected in the muscles on the convex side compared to the concave side in adolescent idiopathic scoliosis (1).

Park et al. discovered that the osteogenic differentiation capabilities of mesenchymal stem cells decreased in idiopathic scoliosis patients (71). It is known that osteopenia develops in the spinal bone structure during idiopathic scoliosis. One reason for the osteopenia is the Bsm I polymorphism in the Vitamin D receptor (VDR) gene (93). (Table I provides an overview of articles concerning hormonal and genetic causes of idiopathic scoliosis and related remarks).

**Biomechanical Causes**

Adams discovered that an existing deformity increased by bending forward (2). The spine has a fixed rotational direction. Posterior elements make an attempt to rotate towards the concave side, and they try to make the shortest advancement just like an athlete running at the extreme interior lane of a marathon course. Therefore, the perpendicular distance is shorter on the posterior side of the vertebrae compared to the relevant distance on the anterior side (91).

In a normal spine, the rotational axis crosses the thoracic region from the anterior. This prevents bending of the thoracic region when compressed. However, lordosis development in this area forces the vertebrae to transcend the rotational axis, whereby the area is made susceptible to bending. This is the reason why patients have an increased deformity by bending forward. A vertebra shall move in two ways under compression. Either kyphosis or lordo-scoliosis will develop. Furthermore, biomechanical studies have shown that in order to ease the load, the loaded vertebra shall: 1) increase the existing curve 2) extend its length and 3) increase its inner load (59).

Anatomical studies have shown that the corpus vertebrae develop a deformity at the axial sections of T4-T9 due to the descending thoracic aorta, as a result of which the thoracic curves bend towards the right side (27).

**CLASSIFICATION of IDIOPATHIC SCOLIOSIS**

A search on the classification of idiopathic scoliosis yielded 310 papers. Since the SRS, Schwab and Aebi classifications are about classifications for adult scoliosis, they were excluded...
Table 1: An Overview of Papers Concerning Hormonal and Genetic Causes of Idiopathic Scoliosis and Related Remarks

<table>
<thead>
<tr>
<th>Reference Paper</th>
<th>Conclusion-Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wise et al. (2000) (103)</td>
<td>The idiopathic scoliosis gene loci are present on chromosomes 6, 10, 18.</td>
</tr>
<tr>
<td>Salehi et al. (2002) (87)</td>
<td>The idiopathic scoliosis associated gene locus is present on chromosome 17p11.</td>
</tr>
<tr>
<td>Chan et al. (2002) (14)</td>
<td>The familial idiopathic scoliosis associated gene locus is present on chromosome 2 and 19p13.3.</td>
</tr>
<tr>
<td>Miller et al. (2012) (58)</td>
<td>The familial idiopathic scoliosis associated gene locus is present on chromosomes 9q31.3–q34.3 and 16p12.3-q22.2.</td>
</tr>
<tr>
<td>Ocaka et al. (2008) (68)</td>
<td>The familial adolescent idiopathic scoliosis gene locus is present on chromosomes 9q31.2–q34.2, 17q25.3-qtel.</td>
</tr>
<tr>
<td>Edery et al. (2011) (24)</td>
<td>The IS associated gene locus is present on 5q13–q14 and 3q11–q13</td>
</tr>
<tr>
<td>Chan et al. (2009) (-)</td>
<td>rs1149048 polymorphism in MATN1 causes a progression of the curve in AIS.</td>
</tr>
<tr>
<td>Jiang et al. (2012) (37)</td>
<td>Matrix metalloproteinases (MMPs) and activator inhibitors (TIMP-2) play a role in endochondral ossification. SNP (rs8179090) in gene TIMP-2 causes an increase in the thoracic curve of AIS.</td>
</tr>
<tr>
<td>Kou et al. (2013) (42)</td>
<td>GPR126 (encoding G protein-coupled receptor 126) is secreted from the cartilage. Suppression of GPR126 causes delays in spine development. SNP in 6q24.1</td>
</tr>
<tr>
<td>Lee et al. (2010) (44)</td>
<td>IL6-572 G→C polymorphism decreases bone density in the lumbar region and it is associated with the AIS.</td>
</tr>
<tr>
<td>Morocz et al. (2011) (63)</td>
<td>SNPs in bone morphogenetic protein 4 (BMP4), interleukin-6 (IL6), leptin, matrix metalloproteinase-3 (MMP3) and melatonin 1B receptor (MTNR1B) genes causes AIS.</td>
</tr>
<tr>
<td>Qiu et al. (2012) (78)</td>
<td>Polymorphism in Neutrophin-3 (NTF3) gene has an impact on the curve caused by AIS.</td>
</tr>
<tr>
<td>Peng et al. (2012) (72)</td>
<td>SNPs in GPER receptor (rs3808351, rs10269151 and rs426655s3) genes have an impact on the degree of the curve in AIS.</td>
</tr>
<tr>
<td>Waller et al. (2013) (97)</td>
<td>ACTB and GAPDH protective molecules are associated with the familial idiopathic scoliosis.</td>
</tr>
<tr>
<td>Wang et al. (2008) (98)</td>
<td>Polymorphism of the triptophan hydroxylase (TPH1) with a role in melatonin synthesis is associated with AIS.</td>
</tr>
<tr>
<td>Zhao et al. (2009) (106)</td>
<td>Polymorphism of Calmodulin1 (CALM1) and estrogen receptor-α genes is associated with AIS.</td>
</tr>
<tr>
<td>Acaroglu et al. (2009) (1)</td>
<td>Higher levels of calmodulin were detected in the muscles on the convex side compared to the concave side, in adolescent idiopathic scoliosis.</td>
</tr>
<tr>
<td>Zhou et al. (2012) (107)</td>
<td>An association has been discovered between the polymorphism of IL-17RC gene and AIS.</td>
</tr>
<tr>
<td>Esposito et al. (2009) (26)</td>
<td>Polymorphism of steroid binding protein has an influence on IS development.</td>
</tr>
<tr>
<td>Leboeuf et al. (2009) (43)</td>
<td>Estrogen hormone has an influence on the degree of the curve in idiopathic scoliosis.</td>
</tr>
<tr>
<td>Letellier et al. (2008) (47)</td>
<td>17-beta-estradiol and melatonin were observed to have a higher influence on the AIS osteoblast cells.</td>
</tr>
<tr>
<td>Man et al. (2011) (55)</td>
<td>Lower levels of Melatonin 2 (MT2) receptors were discovered in AIS.</td>
</tr>
<tr>
<td>Suh et al. (2010) (93)</td>
<td>Bsm I polymorphism in Vitamin D receptor (VDR) gene has an influence on AIS development.</td>
</tr>
<tr>
<td>Park et al. (2009) (71)</td>
<td>It was discovered that osteogenic differentiation capabilities of mesenchymal stem cells degraded in idiopathic scoliosis.</td>
</tr>
</tbody>
</table>

**SNP:** single nucleotide polymorphism.
While the structural curve is considered as the biggest (major) curves and it did not consider the sagittal alignment disorder which was poor in terms of reproducibility and reliability since it did not explain thoracolumbar, lumbar, double major and triple major spinal imbalance once segmental instrumentation was applied (86). Cummings in 1998 and later Behensky et al. also found that King type II and III patients developed with a lumbar curve of 40 degrees and above (81). Roye et al. discovered that spinal imbalance developed in the post-surgery period of selective thoracic fusion in patients with a lumbar curve of 40 degrees and above (81). Roye et al. also found that King type II and III patients developed major spinal imbalance once segmental instrumentation was applied (86). Cummings in 1998 and later Behensky et al. in 2002 discussed the reproducibility and reliability of the King Classification in their papers and stated that the classification was poor in terms of reproducibility and reliability since it did not explain thoracolumbar, lumbar, double major and triple curves and it did not consider the sagittal alignment disorder (9, 21).

King Classification (two-dimensional classification)

While the structural curve is considered as the biggest (major) curve, the other two curves are considered minor. There are some criteria according to which one can determine whether the minor curves are structural (Table III). In addition to the 6 types of curves (I-VI), the Lenke classification also provides for definitions of lumbar spine modifier and sagittal spine modifier (Table IV) (45). Lenke Type I curve features a major curve in the lumbar region. Curves in proximal thoracic (PT) and lombothoracic (LT/L) regions are minor and non-structural. Lenke Type II curve features a double thoracic curve. Curves in proximal thoracic (PT) and main thoracic (MT) regions are structural, but the curve in the lombothoracic region is non-structural. Lenke Type III curve features a major curve in the main thoracic region and a structural curve in the thoracolumbar region. Triple curves are defined at Lenke Type IV. In Lenke Type V, major curve is in the thoracolumbar (TL) junction. Curves in the proximal thoracic (PT) and main thoracic (MT) regions are structural. Lenke Type VI features a structural curve in the main thoracic (MT) and thoracolumbar/lumbar regions. The curve in TL/L is bigger than the curve in the MT.

Lenke et al. assessed the reliability of their own classification system and rated it as 93% reliable (46). Five different surgeons conducted a comparative analysis of Lenke classification and King classification, the results of which indicated that Lenke classification had a reliability of 85%, with a kappa value of 0.83, whereas King classification had a reliability of 69% with a kappa value of 0.69. Lenke classification was reported to have an interobserver reliability of 85%. Lenke et al. conducted in association with several surgeons indicated a reliability level of 84 to 90% for the new classification (46). Ogden stated that Lenke classification was more reliable than the King classification, but it involved some problems concerning the assessment of upper thoracic and lumbar curves (69). Nimeyer et al. stated that both King and Lenke classifications were reliable (67). Harms Working Group reviewed 1281 cases of Lenke et al. retrospectively, and indicated that Lenke did not comply with his own classification in 192 (15%) cases. The ’violation of the rule’ was most frequently observed for Lenke Type 3. Newton et al. reviewed 203 cases of Lenke Type IB and Type IC post-fusion (Lenke classification suggests that only the structural curve should be included in the fusion, while stating that the lumbar region should not be included in the fusion). The common feature of non-selective cases

**Table II: King Classification: Scoliosis is Divided into 5 Sub-Groups According to the Main Curve and Compensatory Curve (41)**

<table>
<thead>
<tr>
<th>Type</th>
<th>Primary Curve</th>
<th>Secondary Curve</th>
<th>Lateral Bending</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Lumbar, crossing the midline</td>
<td>Thoracic, crossing the midline</td>
<td>Lumbar curve is larger</td>
</tr>
<tr>
<td>II</td>
<td>Thoracic, crossing the midline</td>
<td>Lumbar, crossing the midline</td>
<td>Thoracic curve is larger</td>
</tr>
<tr>
<td>III</td>
<td>Thoracic</td>
<td>Lumbar, not crossing the midline</td>
<td>-</td>
</tr>
<tr>
<td>IV</td>
<td>Long Thoracic</td>
<td>Where L5 is centered over the sacrum</td>
<td>-</td>
</tr>
<tr>
<td>V</td>
<td>Double Thoracic</td>
<td>T1 is tilted to the upper thoracic curve</td>
<td>-</td>
</tr>
</tbody>
</table>

**Table III: Structural Criteria for the Minor Curve**

<table>
<thead>
<tr>
<th>Structural Criteria (for minor curves)</th>
<th>Cobb &gt; 25° at lateral bending</th>
<th>T2-5 kyphosis &gt; +20°</th>
</tr>
</thead>
<tbody>
<tr>
<td>Proximal Thoracic</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Main Thoracic</td>
<td>Cobb &gt; 25° at lateral bending</td>
<td>T10-L2 kyphosis &gt; +20°</td>
</tr>
<tr>
<td>Thoracolumbar/lumbar</td>
<td>Cobb &gt; 25° at lateral bending</td>
<td>T10-L2 kyphosis &gt; +20°</td>
</tr>
</tbody>
</table>
Idiopathic Scoliosis has an influence on intraoperative surgical decision-making. The Lenke classification has a weakness of not regarding the rotation of lumbar region before the surgical decision is made. While treating the curve, surgeons tend to violate the basic rules of Lenke Classification by 15%. The Lenke Classification does not clearly state the lowest and highest limit of fusion; neither does it indicate the extent to which the structural curve should be included in the fusion. The classification does not consider shoulder imbalance, patient maturity or body balance. However, compared to the King Classification, the Lenke Classification is useful for the surgeon to use a common vocabulary while describing the curve.

Coonrad Classification
Coonrad et al. reviewed 2000 cases of idiopathic scoliosis and described 21 curve types (18). The author indicated 98.7% and 100% for the interobserver and intraobserver reliability of the Coonrad Classification, respectively. Behensky et al. reported an interobserver reliability level of kappa 0.38 for Coonrad Classification. The use of this classification has not been widespread as it only considered the coronal plane (9).

PUMC Classification System (Peking Union Medical College)
Qiu et al. defined the PUMC Classification system in 2005. This method is useful to determine the surgical approach and define the related fusion levels, and there are 3 main

<table>
<thead>
<tr>
<th>Lumbar Spine Modifier</th>
<th>Type A</th>
<th>CSVL passes between pedicles of apical lumbar vertebrae.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type B</td>
<td>CSVL touches pedicle of apical lumbar vertebrae.</td>
<td></td>
</tr>
<tr>
<td>Type C</td>
<td>CSVL does not touch apical lumbar vertebrae.</td>
<td></td>
</tr>
<tr>
<td>Thoracic Spine Modifier</td>
<td>Sagittal Cobb Angle at T5 to T12</td>
<td></td>
</tr>
<tr>
<td>Hypo (-)</td>
<td>&lt; 10 degrees</td>
<td></td>
</tr>
<tr>
<td>Normo N</td>
<td>10-40 degrees</td>
<td></td>
</tr>
<tr>
<td>Hyper (+)</td>
<td>&gt; 40 degrees</td>
<td></td>
</tr>
</tbody>
</table>

Table IV: Lumbar Spine Modifier Is Defined According to the Central Sacral Vertical Line (CSVL). Types A, B and C are Determined Depending on the Central Sacral Vertical Line Position to the Apical Vertebrae. Thoracic Spine Modifier Is Determined As Hypokyphotic, Normokyphotic or Hyperkyphotic According to the Cobb Angle at the Sagittal Plane

Table V: Papers and Remarks about the Classification Systems for Idiopathic Scoliosis

<table>
<thead>
<tr>
<th>Reference</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Richards BS (1992) (82)</td>
<td>Spinal imbalance develops after selective thoracic fusion in King Type II patients with a lumbar curve greater than 40 degrees.</td>
</tr>
<tr>
<td>Roye et al. (1992) (86)</td>
<td>Selective thoracic fusion causes significant spinal imbalance in King Type II and Type III patients.</td>
</tr>
<tr>
<td>Cummings et al. (1998) (21)</td>
<td>Reliability and reproducibility of the King Classification System is poor since it does not explain thoracolumbar, lumbar, double major and triple curves and take into account the sagittal alignment disorder.</td>
</tr>
<tr>
<td>Behensky et al. (2002) (9)</td>
<td>Reliability of the Coonrad and King classification systems is poor. Both systems are inadequate in classifying upper thoracic and lumbar curves.</td>
</tr>
<tr>
<td>Lenke et al. (1998) (46)</td>
<td>The reliability of the King Classification system in identifying the curve type is poor; it is therefore rarely used to guide the treatment approach.</td>
</tr>
<tr>
<td>Lenke et al. (2001) (45)</td>
<td>Many surgeons can have a consensus by 84-90% on the curve type identified and its degree by using the Lenke Classification system.</td>
</tr>
<tr>
<td>Niemeyer et al. (2006) (67)</td>
<td>King and Lenke classification systems are both reliable.</td>
</tr>
<tr>
<td>Ogon et al. (2002) (69)</td>
<td>The Lenke Classification system is more reliable than the King classification system; however, it causes some issues in evaluating the upper thoracic and lumbar curves.</td>
</tr>
<tr>
<td>Newton et al. (2003) (66)</td>
<td>There is a rule violation by 32% in Lenke Type IC (The non-structural lumbar region was contained in the fusion). The Lenke classification cannot indicate the fusion levels clearly.</td>
</tr>
<tr>
<td>Qui et al. (2008) (80)</td>
<td>When the PUMC and Lenke classification systems are compared, PUMC appears to be relatively simpler and to cause less confusion in surgical planning.</td>
</tr>
</tbody>
</table>
categories as Type I (single curve), Type II (double curve) and Type III (triple curve) with a total of 13 sub-types (79).

Comparing the PUMC and Lenke Classification systems, Qui et al. stated that the PUMC Classification system is relatively simpler and the inter- and intra-observer variability caused less confusion in surgical planning (80).

None of the existing classification systems is ideal to diagnose and treat scoliosis. Table V shows brief information about the papers published on the reproducibility and reliability of the concerned classification systems.

**Natural History**

It is important to know the natural history of idiopathic scoliosis for treatment planning. There must be 5 to 10 degrees increase in the curves to define progression. The degree of the curve and the remaining growth potential of the patient should be known prior to the treatment planning. The Risser sign (39, 48, 84) and the shape of the distal phalanx epiphysis provide information about the maturation process (88). Peak height velocity in girls should particularly be taken into account. This period corresponds to the time of 6 months before menarche in girls. This is the period when the progression in scoliosis is the fastest. The peak growth velocity terminates with the menarche and growth decelerates gradually following this period.

According to Bunnel and Lonstein, 70% of the patients with Risser sign 0, and curves of 20-30 degrees progressed 5 degrees or more. Weinstein stated that the curve could still progress although the patients he had followed-up for 40 years completed their maturation process (102). Nachemson reported that the curve might progress by up to 6 degrees in 66% of the patients with a curve between 20 to 30 degrees (64). Large curves (30-40 degrees) progress more than the minor curves (20-29 degrees) (12, 75).

The curve type has an impact on progression as much as the curve degree does. Double curves progress more than single curves. The least progression is in the curves in the lumbar region (49). The curve progression rate in girls is higher than that in the boys (12, 102).

**TREATMENT**

**Exercise Therapy**

The search on the Pubmed electronic search engine with the key words ‘adolescent idiopathic scoliosis’ ‘physiotherapy’ or ‘exercise therapy’ or ‘rehabilitation’ resulted in a total of 169 papers in English by June 2013. Articles at level 3 or below for evidence-based medicine, where there was no follow-up by measurements, and that did not have at least one-year follow-up were excluded from this review. 9 papers were included in total (Table VI).

Negrini et al. divided 74 patients into two groups and reported the outcomes of the patients they followed for 12.1 years (65). Patients in the first group were treated by a scientific exercise specific to scoliosis whereas the patients in the 2nd group were treated by physiotherapy. The clinical course of the patients was also followed with respect to the need for bracing and Cobb angles. 6.1% of the patients performing the scientific exercise specific to scoliosis needed bracing while this rate

**Table VI:** Papers on the Efficacy of Exercise Therapy in Idiopathic Scoliosis and Their Conclusions

<table>
<thead>
<tr>
<th>Reference</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Negrini et al. (2003)</td>
<td>Need for bracing decreased in patients performing scientific exercise specific to scoliosis, and the Cobb angle was preserved.</td>
</tr>
<tr>
<td>McIntire et al. (2008)</td>
<td>Curve progression was avoided for 8 months in patients (Cobb angle at 20-40 degrees) undergoing body rotation and traction through the MedX Rotary Torso.</td>
</tr>
<tr>
<td>Weiss et al. (2006)</td>
<td>A decrease in the lateral deviation and surface rotation was observed in patients treated by Scoliosis Intensive Rehabilitation (SIR).</td>
</tr>
<tr>
<td>Weiss et al. (2003)</td>
<td>Curve progression was found to decrease significantly in the group of patients treated by Scoliosis Intensive Rehabilitation (SIR) compared to the control group.</td>
</tr>
<tr>
<td>Weiss (1992)</td>
<td>Cobb angles decrease due to the Schroth method compared to the pre-treatment values.</td>
</tr>
<tr>
<td>Mooney et al. (2003)</td>
<td>Cobb angles were observed to decrease in 16 out of 20 patients treated by MedX Rotary Torso.</td>
</tr>
<tr>
<td>Otman et al. (2005)</td>
<td>Cobb angles were observed to decrease in a year in patients treated by Schroth’s 3D exercise therapy.</td>
</tr>
<tr>
<td>Mamyama et al. (2002)</td>
<td>No progression was observed in the Cobb angles when side shift exercise was practiced by patients with idiopathic scoliosis who reached full maturity.</td>
</tr>
<tr>
<td>Dobosiewicz et al. (2002)</td>
<td>208 AIS patients with a median age of 14.2 years were treated by asymmetrical body mobilization. Cobb angles were observed to decrease (no statistical data).</td>
</tr>
</tbody>
</table>
was found to be 25% in the patients undergoing the classic physiotherapy. The mean Cobb angle of the patients in the 1st group remained the same (-0.67 degrees), whereas the Cobb angle of the patients undergoing the classic physiotherapy worsened (+1.38 degrees).

McIntire et al. treated 15 patients with a median age of 13.9 years, with Cobb angles ranging between 20-60 degrees, Risser sign III and below with physiotherapy by means of the MedX Rotary Torso Machine. It was reported that six patients with curves between 20 to 40 degrees did not progress during the 8-month follow-up period but progression could not be avoided by the end of 24-month follow-up (57). On the other hand, Mooney and Brigham who used the same method found a reduction in the Cobb angles in 16 out of 20 patients in the age group of 11-17 years (62).

Weiss et al. followed the lateral deviation surface rotation and kyphotic angle of a patient group by applying the 4-week Scoliosis Intensive Rehabilitation (SIR) program. It was observed that the lateral deviation and surface rotation decreased in the treatment group compared to the control group (99). In another group of patients who underwent the same program, progression was observed in the curves of the control group compared to the control group (100). It was stated that the Cobb angles of 107 patients treated by Schroth method fell from 43.06 to 38.96 degrees compared to the pre-treatment values (101). Cobb angles were reported to fall from 26.10 degrees to 18.85 degrees in a year in another series of patients treated by the same method (70).

Mamyama et al. treated 69 patients with idiopathic scoliosis, who reached full maturity, with the side shift exercise for 4.4 years and reported that Cobb angle fell from 31.5 degrees to 30.3 degrees (54). Dobosiewicz et al. treated 208 AIS patients with a median age of 14.2 years with asymmetrical body mobilization and reported that Cobb angles decreased by 33.6% in single curves, by 22.8% in double curves in the thoracic region, and by 26.1% in the lumbar region; however, this study has a lower value as no statistical data was used in the study (23).

**Bracing**

For the literature review, ‘idiopathic scoliosis’ and ‘bracing’ were used as the key words to search in the Pubmed search engine and 329 papers were found as a result. Papers on bracing for other scoliosis types, papers on conservative treatment, genetic issues and irrelevant papers were excluded from this section. 81 papers were analyzed.

For successful bracing, the existing curve should be preserved below 45 degrees until the patient reaches full maturity (81, 89). Richards et al. suggested bracing for patients with Risser sign from 0 to 2, and current curve at 25-40 degrees. Surgery is suggested for patients reaching their full maturity and with curves greater than 45 degrees (81). Lonstein found a reduction by 1-4 degrees in 78% of 1020 patients with adolescence idiopathic scoliosis using Milwaukee braces while it was reported that 22% of the cases required surgery (49, 50). Bassett treated 75 cases with a curve at 20-39 degrees, Risser sign from 0 to 1 and using Wilmington braces for 2.5 years and stated that the degree of curves in patients decreased by 50%. Only 11% of the cases in this series required surgical intervention (8). In the Katz series where 51 cases with curves between 36 to 45 degrees and using Boston braces, bracing was successful in 61% of the patients while 31% of the cases required surgery (40). Coillard et al. treated 170 patients by using Spine-Cor braces and reported 59% success rate (17). 23% of the cases required surgery.

Milwaukee, Wilmington, Spine-Cor and Boston braces appear to be superior to the other types of braces. Price reported that the outcomes of 98 patients using Charleston braces were excellent by 63%. He reported that the major curves were corrected by 85% and the minor curves by 33% (74). In another series using the same braces, curve progression was stopped by 60% in the patients with Risser sign from 0 to 1 (96).

Braces should be used for 18-23 hours a day on average. Braces were reported to be effective for patients with curves less than 35 degrees when used part-time or at night (74). Braces should be used full-time by patients with curves equal to or greater than 35 degrees. Braces should be used until the end of full growth. Bracing should be continued for 18-24 months by the end of 6th month following the end of growth, up to Risser sign 4 in girls, Risser type 5 in boys, and following the menarche in girls (89).

**SURGICAL TREATMENT**

**Indications for Surgical Treatment**

In general, curves greater than 45 and 50 degrees should be treated by surgery. It was found in the studies that curves greater than 50 degrees still progressed even after reaching full maturity (102). Edgar et al. applied a non-surgical approach for the treatment of the patients with thoracic curves between 50 to 75 degrees, Risser sign 4 and 5 for 40 years and reported a mean increase by 29.4 degrees in the curves during this period (25). In another study where patients were followed for 50 years, it was reported that the thoracic curves progressed from 60.5 degrees to 84.5 degrees on average. One should not wait for the curve to progress since surgical treatment of greater degrees of curves would increase complication rates and cause more intraoperative blood loss.

**1. Fusion Surgery**

**Posterior instrumentation**

Instrumentation was first used by Paul Harrington in the surgical treatment of scoliosis (32). Harrington attempted to correct the curves by distraction from the concave side using the rods. The second generation instrumentation system was developed by Cotrel and Dobousett. They tried to achieve correction using rod rotation manoeuvres (19). Spine can be attached tightly to the rods by pedicle screws or hybrid systems thanks to today’s advanced technology.
Suk used the pedicle screws in the surgical treatment of deformities. He reduced the curves from 51 to 16 degrees on average (69% correction) (94). Asher et al. reported to have achieved 63% correction by the hybrid system they applied with hooks and pedicle screws (5). Cheng et al. compared the hooks and transpedicular screws and reported no difference between both systems in terms of correction rates (15).

**Anterior instrumentation**

Anterior approach can be preferred as it can achieve correction with shorter fusion levels in the scoliotic thoracolumbar and lumbar regions. Postoperative pain and scar formation decrease in patients with the introduction of the video-assisted thoracoscopic surgery (76, 92). Potter compared anterior and posterior fusion for thoracic curves and reported that posterior approach achieved better correction compared to the anterior approach (73). Hee et al. reported no difference in the coronal plane between the anterior instrumentation and pedicle screws in patients with adolescence idiopathic scoliosis (33).

**Determining the Fusion Levels**

**Right Thoracic Curves (Lenke Type 1: Main Thoracic Curve)**

This is the most frequent curve type. There is a major curve in the main thoracic region. Curves in the proximal thoracic (PT) region and lumbothoracic (LT/L) region are minor and non-structural. All curves in this type can be treated by posterior instrumentation and fusion. T3, T4 or T5 that will be instrumented as the upper vertebrae while lower instrumented vertebrae (LIV) depends principally on the lumbar spine modifier (45).

Lower instrumented vertebrae (LIV) of a patient with the lumbar spine modifier A should be the one that the central sacral vertical line (CSVL) intersects in the TL/L region. This is usually one level below the lower end vertebrae (LEV) of MT.

The same technique shall apply to the patients with 1B curves. Lenke suggested for patients with curve modifier B that some residual tilt should be preserved in the LIV due to the lumbar apical deviation of curves (45).

It is suggested to go down to the stable vertebrae (SV) for patients with 1C modifier curves. This should be T11 or T12. Sagittal balance should also be taken into account in determining the lowest level of the fusion. It should ensured that the junctional kyphosis was also contained in the fusion to maintain the sagittal balance. Selective thoracic fusion for patients with 1C curves would be useful to preserve the lumbar range of motion of the patients (45).

**Double Thoracic Curve (Lenke Type 2)**

Proximal thoracic (PT) curves and main thoracic (MT) curves are the structural curves. However, lumbothoracic curves are non-structural. The upper instrumented vertebrae (UIV) should be T2 or T3. On the other hand, LIV should be selected according to the procedure applied to Lenke Type I depending on the lumbar spine modifier. One of the basic rules to take into account in PT curves is to preserve the shoulder balance. Left shoulder is usually higher than the right one (in right MT, left PT curves). In order to achieve the shoulder balance, it is necessary to apply compression forces on the convex side of the PT and distraction forces on the concave side of the PT (45).

Although rarely observed, UIV should also be T2 or T3 for Lenke Type 2 curves where right shoulder is higher. Shoulder balance should be achieved in correcting the MT curves. Left shoulder should not be lifted too high while achieving the shoulder balance in selected patients for whom the fusion level starts at T4 and T5 and PT is not contained in the fusion (45).

**Double Major Curve (Lenke Type 3)**

There is a major curve in the main thoracic region and a structural curve in the thoracolumbar junction. The UIV should be T3, T4 or T5 depending on the non-structural PT curve with shoulder imbalance just like in Lenke Type 1. LIV should usually be extended to L3 or L4 (45). If the apex of the TL/L curve is on or lower than L2, L3-4 disc is convex or open on the convex side of the TL/L curve and L4 has a rotation of 1 degree or greater according to Nash-Moe classification, the fusion should be extended to L4 (45).

If the apex of the TL/L curve is on or higher than L1-2, L3-4 disc is neutral or closed on the convex side of the TL/L curve and if L4 has a rotation of 1.5 degrees and lower according to Nash-Moe classification, L3 should be selected as the LIV. Type 3 curves usually have C spine modifier. Curves with A and B spine modifier have wide-angle MT curves, which makes the TL/L curve structural in the lateral bending x-rays.

**Triple Major Curve (Lenke Type 4)**

Since the proximal thoracic (PT), main thoracic (MT) and thoracolumbar/lumbar (TL/L) curves are structural, three curves should be contained in the fusion. UIV should be selected as in Lenke Type 2 while LIV should be selected as in Lenke Type 3. Fusion level usually ranges between T2, T3 and L3, L4 (45).

**Thoracolumbar and Lumbar Curves (Lenke Type 5)**

Main curve is in the thoracolumbar (TL) junction. Curves in proximal thoracic (PT) and main thoracic (MT) regions are non-structural. UIV should be the vertebrae one level or two level above the USV, whereas the vertebrae one level or two level below the LSV should be selected as the LIV.

**Main Thoracolumbar / Lumbar Curve (Lenke Type 6)**

The curve in the proximal thoracic region is non-structural. The curve in the main thoracic region is structural. The curve in the thoracolumbar/lumbar region is larger than the one in the main thoracic region. Both curves should be contained in the fusion (Figure 8). The surgical limits should be determined according to the principles applying to the Type 3 curves.
2. Non-Fusion Surgery

Non-fusion surgery is another option in order to control growth in the treatment of idiopathic scoliosis. Progression of the curve might be avoided by means of instrumented or non-instrumented epiphysiodesis on the convex side of the curve.

Betz et al. stated that preventing the growth in front of the vertebral column will also prevent the progression of adolescence idiopathic scoliosis whereas Marks argued that preventing the anterior and posterior growth in infantile scoliosis alone cannot stop the progression of the deformity (11, 56).

After the fusion surgery at younger ages, the body remains shorter than the limbs. Shorter body prevents the development of lungs. Some techniques for the correction of the existing curve while also allowing the growth of the spine have been developed (31, 60). The upper and lower parts of the curve can be fixed by Isola double rod system developed by Akbarnia, attached to the rods and the rods are attached to one another with an additional rod. The rods are extended in 6-month follow-up. After reaching the full growth, fusion is completed with instrumentation. The degree of the curves which was preoperatively 82 on average was reduced to 38 degrees and then 36 degrees following an average 6.6 extension surgery as a result of this method applied on 23 patients between 1993 and 2001 (3).

Vertical expandable prosthetic titanium ribs (VEPTR) were developed to treat the thoracic insufficiency syndrome that is caused by combination of ribs and curves (13). The deformity can be corrected acutely by means of VEPTR following the wedge thoracostomy. VEPTR device is expanded in 4-6 months time. Seventy two patients with a mean age of 3.2 years were treated with this device for 5.7 years and it was reported that the vital capacity increased and also the curve degree was reduced from 72 on average to 49 (13).

CONCLUSION

It is the post-operative appearance that the surgeons are perhaps the least interested but the adolescence patients are the most interested. It is essential to avoid breast asymmetry and achieve shoulder balance for girls. Rib humps that can be recognized by the adolescent or her peers over the clothing should be reduced to acceptable surgical limits postoperatively.

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