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Review Article

Scheuermann's disease: Current diagnosis and treatment approach

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Abstract.
OBJECTIVES: To summarize the current knowledge relating to diagnosing and treating Scheuermann's disease. Scheuermann's disease is the most common cause of structural kyphosis in adolescence.
METHODS: A literature-based narrative review of English language medical literature.
RESULTS AND CONCLUSIONS: Recent studies have revealed a major genetic contribution (a dominant autosomal inheritance pattern with high penetrance and variable expressivity) to the etiology of Scheuermann kyphosis with a smaller environmental component (most probably mechanical factors). The natural history of Scheuermann kyphosis remains controversial, with conflicting reports as to the severity of pain and physical disability. Since we cannot predict which kyphotic curves will progress, we are unable to determine effectiveness of brace treatment. Physical therapy is scarcely mentioned in the literature as an effective treatment for Scheuermann kyphosis. Although there is little evidence that physical therapy alone can alter the natural history of Scheuermann's disease, it is often used as the first choice of treatment. Brace treatment appears to be more effective if an early diagnosis is made, prior to the curvature angle exceeding 50° in patients continuing to grow. Surgical treatment is rarely indicated for severe kyphosis ($>75^\circ$) with curve progression, refractory pain, or a neurologic deficit. Rigorous methodology clinical trials are essential to evaluate the efficacy of conservative interventions, especially different exercises and manual therapies and their combinations with braces.

Keywords: Scheuermann's disease, etiology, review, spine, treatment

1. Introduction

Scheuermann's disease is the most common cause of hyperkyphosis of the thoracic and thoracolumbar spine during adolescence. After idiopathic scoliosis, it is the most common disorder in patients with a deformed spine [1,2]. This condition is characterized by vertebral body wedging, vertebral endplate irregularity, diminished anterior vertebral growth, Schmorl's nodes, narrowing of the intervertebral disk spaces and premature disks degeneration.

Scheuermann kyphosis develops prior to puberty, after ossification of the vertebral ring apophysis, and appears most prominently during the adolescent growth

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spurt, as a structural kyphotic deformity of the thoracic or thoracolumbar spine. The disease onset typically occurs from the late preschoolers to 16 years of age, most commonly between ages 12 and 15 [3–6].

The criteria for diagnosing Scheuermann's disease, its etiology and natural history remain controversial. Therefore, we attempt herein to summarize the present knowledge relating to prevalence, etiology, diagnosis and treatment of Scheuermann's disease.

2. Methods

PubMed, Ovid, Google Scholar and PEDro databases were searched using predefined strategy from inception until January 2013 for the keywords: "spine", "spinal deformities", "kyphosis", " juvenile osteochondrosis", "Scheuermann", "Scheuermann's disease", "hyperkyphosis". All relevant articles in English were reviewed. Pertinent secondary references were also retrieved. We also consulted experts in pediatric orthopedic surgery, rheumatology and Schroth method practitioners to produce this narrative review on current diagnosis and treatment approaches to Scheuermann's disease.

We critically analyzed all published material. We are aware that this traditional approach to narrative reviews has much more potential for bias than systematic reviews or meta-analyses; however, we have endeavored to be inclusive and open-minded.

2.1. *Qualification of searchers*

Literature was searched and screened by two authors (TB and LK). TB, master in the physical therapy was trained by LK, physical therapists and PhD, experienced in writing reviews on spinal pathologies [7–10].

3. Results

3.1. *Definition*

Scheuermann's disease was initially described in 1920, by Holger Scheuermann, as a rigid thoracic and thoracolumbar kyphosis associated with vertebral body wedging in late childhood [11]. This definition has been developed and modified over the years. In 1964, Sorenson described specific criteria of three or more adjacent apical vertebrae each wedged a minimum of 5° [12]. Currently, most studies use Sorenson's definition. Other authors have proposed different criteria. In 1987, Drummond suggested that two or more adjacent apical vertebrae are sufficient for diagnosis [13]. In 1987, Sachs reduced the requirement to at least one vertebra wedged a minimum of 5° and a thoracic kyphosis >45° (T3-T12) [14].

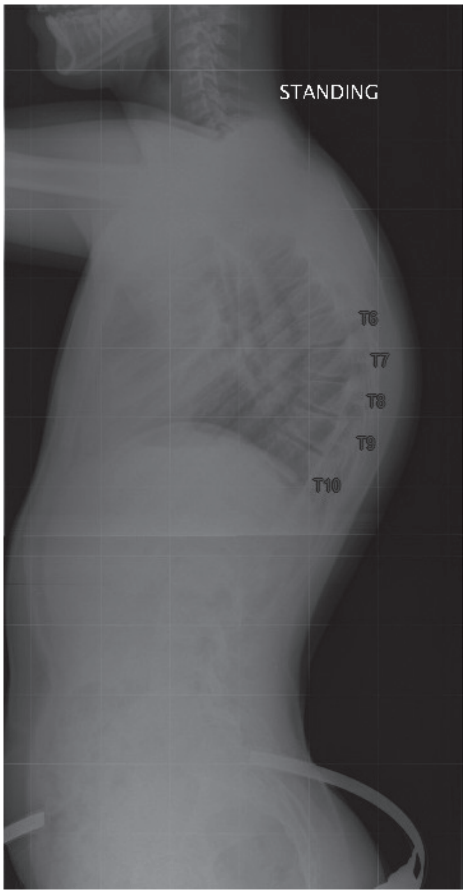

Fig. 1. The thoracic pattern. Note the excessive anterior wedging of T6-T10 vertebral bodies.

3.2. *Classification*

Two different curve patterns have been described. The thoracic pattern (Fig. 1) is the most common and is associated with a nonstructural hyperlordosis of the lumbar and cervical spine [15]. The thoracolumbar pattern (Fig. 2) is rare but thought to be the most likely to progress during adulthood. Lumbar, Type 2 Scheuermann's, or "Apprentice kyphosis" are other names used to describe this disease [16–18]. This condition, most commonly seen in athletically active adolescent males or those involved in heavy lifting, manifests itself by localized back pain and radiographic vertebral changes

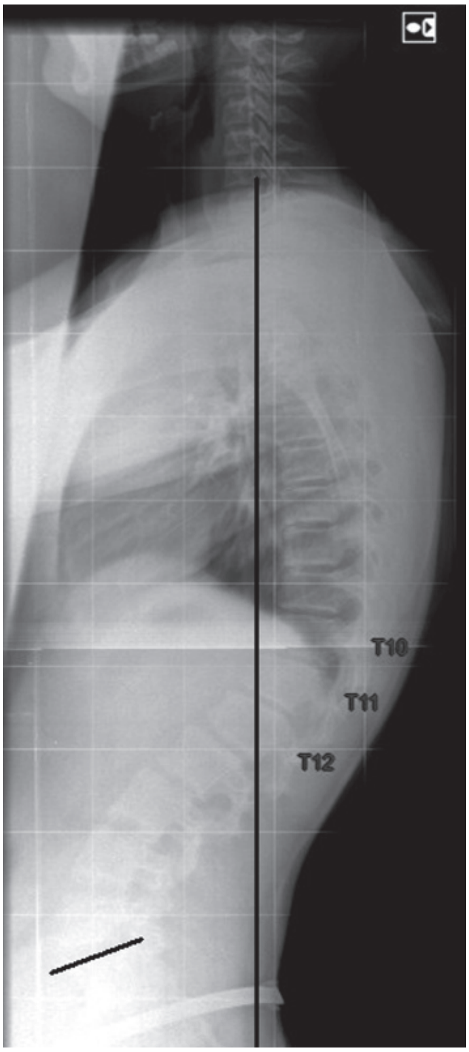

Fig. 2. The thoracolumbar pattern. Note the excessive anterior wedging of T10-T12 vertebral bodies. The C7 vertebral body vertical axis (vertical line) passes behind the sacral promontory (lower oblique line), resulting in negative sagittal balance.

at the thoracolumbar junction, and is not typically associated with significant clinical kyphosis [4]. Unlike classic thoracic Scheuermann kyphosis, treatment of lumbar Scheuermann's disease is not controversial, as its course is non-progressive and symptoms resolve with rest, activity modification and time [16,18].

3.3. Etiology

The etiology of Scheuermann's disease is still under debate. Several theories have been suggested, including an increased release of growth hormone, defective formation of collagen fibrils with subsequent weakening of the vertebral end plate, juvenile osteoporosis, trauma, vitamin A deficiency, poliomyelitis and epiphysitis [4,6,13,19].

A study by Damhborg et al. [20] of 35,000 symptomatic twins with Scheuermann kyphosis indicated a major gene effect in etiology of Scheuermann kyphosis with a smaller environmental component. A dominant autosomal inheritance pattern with high penetrance and variable expressivity has been suggested for Scheuermann's disease etiology [20–22].

Most investigators agree that mechanical factors also play a significant role in the pathogenesis of Scheuermann kyphosis. For example, repetitive activities involving axial loading of the immature spine was mentioned as a factor in the development of Scheuermann kyphosis. It has been reported that children with Scheuermann's disease are heavier and taller compared to healthy children [4,6,16,19,23–26]. Fotiadis et al. [3] suggested that those parameters do not affect the magnitude of the kyphotic curve and therefore not part of pathogenetic mechanism of Scheuermann's disease. The increased weight and height may be the secondary result of other disturbances (i.e. hormonal), which may play more crucial role in Scheuermann's disease pathogenesis [3].

Several studies have shown decreased bone mineral density (BMD) in Scheuermann's patients [27–29]. Bradford et al. in 1976 have noted decreased BMD in 12 Scheuermann's patients [27]. Lopez et al. in 1988 compared BMD of 10 Scheuermann's patients and seven controls [28] and also found significantly lower BMD in Scheuermann's patients. More recent study [29] found decreased bone density in children prior to sexual maturity (Tanner stages 1–4), but not in mature subjects (Tanner stage 5). However, Gilsanz et al. in 1989 found no significant differences in bone density between 20 Scheuermann's patients and control [30]. Popko et al. [31] evaluated total and L2-L4 BMD in 24 Scheuermann's patients and compared it to reference population. They found lower BMD level in nine patients. Authors concluded that low BMD in these patients may be caused by decreased physical activity due to vertebral pain. The cross-sectional design of all aforementioned studies, not allows determining unequivocally the causal relationships between decreased BMD and Scheuermann disease. Additional, follow-up studies are needed to clarify this question. If indeed low BMD is part of the etiology of Scheuermann disease, new prevention and treatment strategies could be developed.

The radiological changes of the disease may be expressions of an altered remodeling response to abnormal biomechanical stresses. The anterior vertebral body wedging is secondary to the increased anterior forces due to the first occurring kyphosis [25]. Degenerative changes in the disks are seen on magnetic resonance images (MRI) in more than half of the Scheuermann patients [32]. Whether those changes are the cause for Scheuermann or the result of it, is still unclear [33].

Another radiological sign that was correlated with Scheuermann disease pathogenesis is the length of the sternum. A smaller length of the sternum has been associated with Scheuermann disease [34]. Authors explained this finding by primary mechanical forces. The smaller length of the sternum increases the compressive forces in the anterior part of the thoracic vertebra bodies, which leads to a kyphotic deformity.

3.4. Prevalence

Prevalence of Scheuermann kyphosis has been reported to range from 0.4% to 8% of the population [4, 12,19,25,35,36], although its true incidence is probably understated since the diagnosis is often either missed or attributed to poor posture [20,37,38].

Diverse male to female ratios have been reported, 1:2 [39], 1:1 [40,41], 2:1 [6], or 7.3:1 [11]. In a recent study based on 11,436 pairs of twins, male prevalence was found to be almost twice that of female prevalence. The estimated prevalence in males and females based on the hereditability, the threshold and regression coefficient for gender, presented a calculated male prevalence of 4% and a calculated female prevalence of 2% [42].

3.5. Radiographic findings

Initial radiographs of the patients include a standing postero-anterior (PA) and lateral view of the spine. The degree of kyphosis on the lateral films is measured using the modified Cobb method. The thoracic Cobb angle is measured from the T3 superior endplate to the T12 inferior endplate. In addition to the increased measurable round back on lateral view, vertebral wedging is used to clarify the diagnosis. Associated findings of scoliosis and spondylolysis can occur with Scheuermann kyphosis, but are usually minor and do not alter treatment decisions [43,44].

The normal range of thoracic kyphosis (T3-T12) is 25° to 45° as measured by the Cobb method on a standing lateral radiograph with arms at a 45° angle below horizontal line [45,46].

Normal kyphosis increases with age and is slightly greater in women. Lumbar lordosis varies considerably ranging from 36° to 56°, which is considered normal. The transitional zone between the thoracic and lumbar spine includes T10-L2 and is normally slightly lordotic (0° to 10°) [14,45].

When the spine is balanced in the sagittal plane, the C7 vertebral body vertical axis (C7 plumb line) should lie vertically within 2 cm of the sacral promontory, which is in contrast to patients with Scheuermann's disease where the spine tends to be negatively balanced when compared to the general population, and the C7 vertebral body vertical axis C7 plumb line lies behind the sacral promontory (Fig. 2) [38,47].

In summary, the two main radiographic diagnostic findings are thoracic kyphosis >45 degrees (T3-T12), and at least one vertebra wedged at a minimum of 5° [14].

3.6. Clinical presentations and comorbidities

Deformity is the most common complaint of patients with Scheuermann's disease, and is typically the primary reason younger patients seek medical attention [39]. Unfortunately, the likelihood of progression of a kyphotic curve at any given degree of severity is currently unknown [48].

Pain, when present, is usually mild, brought on by prolonged periods of sitting or exercise and is usually located near the apex of the deformity. Usually the pain subsides with the cessation of growth. Occasionally, low back pain may be associated with spondylolisthesis, noted with an increased incidence in patients with Scheuermann's disease [43].

The long-term prognosis of patients with Scheuermann kyphosis is largely unknown. Murrey et al. [6] in their 32 year follow up study, reported that patients with Scheuermann's disease experienced more pain than the control group. The pain symptoms, however, did not interfere with activities of daily living or employment.

A recent study by Ristolainen et al. [36] demonstrated that Scheuermann's patients had a higher risk for back pain compared to controls. In addition, these patients reported a lower quality of life and poorer general health. Risk of disabilities while performing activities of daily living was more prevalent in Scheuermann's disease patients than the controls. However, no correlation between the degree of kyphosis and self-reported quality of life, health or back pain was found. In addition to the kyphosis of the thoracic spine, these patients demonstrated varying degrees of hyperlordosis of the lumbar and cervical spines and a forward protrusion of the head related to a high thoracic kyphosis. The lumbar and cervical deformities are usually flexible. Varying degrees of structural scoliosis are noted in approximately 33% of the patients. Most of these curves are minor.

The kyphosis may be thoracic or thoracolumbar and fixed, thus visible on hyperextension of the spine. When viewed from the side in the forward-bending position (Adams' test), the deformity is sharply angulated. In addition to the spinal deformity, there is often tightness of the anterior shoulder girdle, the hamstring and iliopsoas muscles [49].

An interesting clinical sign is a cutaneous skin pigmentation appearing at the most protruding spinous process at the apex of the kyphosis, due to skin friction at the back of the chair [1,4,6].

Neurologic deficits are rare, including gradual and progressive lower paraparesis. These deficits are usually secondary to thoracic disk herniation, kyphotic angulation, tenting of the spinal cord, extradural spinal cysts, osteoporotic compression fractures and vascular injury to the anterior spinal artery due to compression of the spinal artery of Adamkiewicz [50,51].

The effect of kyphotic deformity on pulmonary functions is unclear. Normal or above normal averages for pulmonary function were found when the kyphosis was $<100^\circ$. Patients in whom the kyphosis was $> 100^\circ$ and the apex of the curve was in the 1^{st} to 8^{th} thoracic segments, were diagnosed with restrictive lung disease [6].

3.7. *Differential diagnosis*

Scheuermann's disease is characterized by a rigid or relatively rigid thoracic or thoracolumbar deformity with specific radiographic findings. It is essential to differentiate Scheuermann's disease from postural kyphosis. The latter is a benign condition secondary to the patient's bad posture. Thoracic postural kyphosis is uniformly rounded and clinically flexible. Scheuermann's specific imaging findings (wedging vertebral bodies and disc degeneration) are not observed [52].

In the forward bending test, a patient with postural kyphosis presents with a smooth, flexible, and symmetric contour, while patients with Scheuermann's disease display an area of angulation in a fixed kyphotic curve. For differential diagnosis the pathologies that must be excluded are idiopathic kyphosis, specific or not spondylitis osteochondral dystrophies, spondyloepiphyseal dysplasias, and congenital kyphosis [4, 53].

3.8. *Conservative treatment*

Managing treatment of Scheuermann's disease is based on the severity of the deformity, presence of pain, and the patient's age. The treatment is primarily non-operative. Adolescents whose kyphosis remains $<60^\circ$ are usually treated only by exercises to increase flexibility. They are then periodically followed up by x-rays until skeletal maturity [47].

Soo et al. [53] suggested appropriate treatment for Scheuermann kyphosis based on the patient's age, spinal deformity, and severity of back pain. Generally, skeletally immature patients with a kyphotic curve of 45° to $>50^\circ$, and radiographic findings diagnosing Scheuermann's disease, may be candidates for physical therapy and bracing treatment. Intensive physiotherapy exercise programs for postural improvement have been tried for many years but without any conclusive data that physical therapy alone can assist in kyphotic improvement [4,54].

On the other hand, the majority of adults with untreated Scheuermann's disease responded satisfactorily to physical therapy and a back exercise program provided that the kyphosis was not severe ($<60^\circ$) [47].

According to Rachbauer et al. [55], extension sports such as gymnastics, aerobic, swimming, basketball, cycling and hyperextension exercises are advised. However, sports associated with jumping, marked stress and functional overuse of the back, especially in patients with thoracolumbar and lumbar Scheuermann's kyphosis, should be discouraged [55,56].

Several case reports [57,58] and our clinical experience, demonstrated the effectiveness of Schroth therapy in preventing and improving the thoracic curve angle in Scheuermann's patients, however, efficacy and effectiveness of this method should be examined in a controlled clinical research.

In adolescent patients with Scheuermann's disease whose kyphosis is $<60^\circ$, an exercise programs with or without partial time bracing is recommended [47]. In patients with kyphosis $>60^\circ$, a brace program of >20 hours per day should be considered [14,40].

The few available studies on the efficacy of brace treatment are retrospective, have different inclusion criteria, and no control group. Perhaps due to incomplete understanding of the natural history, the indications for brace treatment are not well defined and differ between studies. Weiss et al. [57] reported their results of long-term physical therapy, osteopathy, manual therapy, exercise programs, and psychological therapy for a group of 351 patients (17–21 years of age) with painful Scheuermann kyphosis. At the end of treatment, both the visual analogic and numbered scales showed a reduction in pain between 16% and 32%, which was significant in all cases.

Montgomery et al. [40] reviewed 39 patients with Scheuermann kyphosis treated with a modified Milwaukee brace for an average of 18 months. Prior to treatment, the kyphosis averaged 62° and at completion averaged 41°. Follow up of >18 months after completion of brace wear showed an average of a 15° loss of correction, resulting in an overall average correction of 6°. Wedging of the vertebral bodies improved the kyphosis from 7.9 to 6.8° with brace treatment. Brace treatment was successful in improving kyphosis >75° in several cases.

Sach et al. [14] reported on the long-term results of brace treatment of 120 patients with Scheuermann kyphosis. Patients were treated with a Milwaukee brace at the Twin Cities Scoliosis Center. All patients were observed for at least 5 years after completion of treatment. Of the patients who consistently wore the brace, 76 showed kyphosis improvement between the initial and follow up evaluations; 24 exhibited some worsening, and 10 were unchanged. The patients showed an initial improvement of 50% followed by a gradual loss of correction. The authors noted that 4/14 patients with kyphosis of >75°, subsequently required a spinal fusion.

Patients with thoracic kyphosis with an apex above T7 are best managed with a Milwaukee brace, whereas those with low thoracic or thoracolumbar kyphosis can usually be managed with an underarm orthosis with anterior infraclavicular outriggers [39,40]. Both stretching and strengthening exercises should also be prescribed for trunk as well as tight hamstrings and pectoral musculature [57].

Partial reversal of anterior wedging of vertebral bodies is often noted after 12 to 18 months of successful brace treatment when full passive correction has been achieved. At that time, part-time (12 hours per day) brace treatment can be continued until complete skeletal maturity is achieved (i.e., fusion of iliac and vertebral ring apophysis). Although initial improvement is often significant, a 20 to 30% loss of correction with time usually occurs, therefore, only a modest overall long-term correction of the pre-brace deformity can be expected [14,40].

Unfortunately, these studies do not indicate whether brace treatment actually corrects the deformity caused by Scheuermann kyphosis. In addition, as noted above, there are no available data allowing us to predict which kyphotic curves are at significant risk for progression. Despite these shortcomings, bracing is widely regarded as efficacious in the treatment of Scheuermann kyphosis in the skeletally immature patient [19,24,41].

Bracing has been used primarily in treating the cosmetic aspect of deformities and therefore published results of treatment focused on kyphosis improvement. Data regarding the effect of brace treatment on pain have not been published.

3.9. Surgical treatment

Surgery is usually considered in adolescents with Scheuermann's disease only when the deformity is severe (>80°) and cannot be controlled with brace treatment. Operative treatment should also be considered in adults with kyphosis >75°, who have persistent pain uncontrolled by non-operative management or an unacceptable cosmetic deformity [19,59]. Biomechanical principles of correction of kyphosis secondary to Scheuermann's disease include lengthening of the anterior column (anterior release), providing anterior support (interbody fusion), and shortening and stabilizing the posterior column (compression instrumentation and arthrodesis) [60].

In the skeletally immature patient, these goals can usually be met with a posterior fusion alone since the remaining anterior growth provides anterior column stability. In adolescents, as well as adults, an anterior release and fusion is usually performed first, followed by a posterior fusion with segmental spinal instrumentation [47].

Both efficacy and the potential complications of a combined anterior/posterior fusion for Scheuermann kyphosis are well documented in the literature. Hemothorax, pneumothorax, pleural effusion, postoperative wound infections, and permanent paraplegia are noted in the literature. Pulmonary function is also known to be negatively impacted by anterior surgery even at the 2 year follow up [60]. The major complication after surgical treatment is junctional kyphosis, usually related to inappropriate fusion levels or overcorrection of the deformity.

Literature on the surgical management of Scheuermann kyphosis consists primarily of retrospective case series with different inclusion criteria for surgery and no control groups. The indications for surgery remain unclear since the natural history in Scheuermann kyphosis remains controversial as regards to pain, disability, self-esteem, and deformity progression. A decision for surgery needs to be an individual one between the surgeon and patient based on several components, including the patient's symptoms and self-perception [52].

4. Conclusions

Scheuermann's disease is the most common cause of structural kyphosis in adolescence. A review of the literature reveals many shortcomings and gaps in the scientific knowledge and evidence. The etiology and even the criteria for diagnosis remain unclear. Recent studies have revealed a major genetic contribution (a dominant autosomal inheritance pattern with high penetrance and variable expressivity) to the etiology of Scheuermann kyphosis with a smaller environmental component (most probably mechanical factors).

The natural history of Scheuermann kyphosis remains controversial, with conflicting reports as to the severity of pain and physical disability. Since we cannot predict which kyphotic curves will progress, we are unable to determine effectiveness of brace treatment.

Physical therapy is scarcely mentioned in the literature as an effective treatment for Scheuermann kyphosis. Although there is little evidence that physical therapy alone can alter the natural history of Scheuermann's disease, it is often used as the first choice of treatment. Brace treatment appears to be more effective if an early diagnosis is made, prior to the curvature angle exceeding 50° in patients continuing to grow. Surgical treatment is rarely indicated for severe kyphosis (>75°) with curve progression, refractory pain, or a neurologic deficit.

Future research should focus on the natural progression and risk of deterioration in patients with Scheuermann's disease. Prospective studies with longer follow-up duration and control groups, using valid outcome measures are needed to provide the clinicians with more accurate tools for treating patients with Scheuermann's disease. Rigorous methodology clinical trials are essential to evaluate the efficacy of conservative interventions, especially different exercises and manual therapies and their combinations with braces.

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