Scheuermann's kyphosis: Etiology and diagnosis

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Introduction

Over a century has passed since Holder Scheuermann described a kyphotic deformity with anterior wedging of the vertebral bodies and characteristic irregularities of the vertebral endplates [1].

The first signs usually appear during the growth spurt at the start of puberty and no radiographic signs have been reported before the age of 10 years.

Scheuermann's kyphosis affects around 0.4 to 10% of adolescents aged between 10 and 14 years old. When even minor irregularities of the ossification of endplates are considered, the prevalence increases to 40%.

Pathophysiology

1. Histology

Scheuermann suggested a form of aseptic necrosis of the growth plate at the level of the lateral vertebral margins. Schmorl and Junghans considered endplate herniations as typical lesions found in Scheuermann's disease and hypothesized that alterations in the consistency of the cartilaginous endplates may be the cause of Scheuermann's disease [2]. However, the occurrence of such irregularities may be seen outside of the kyphotic area and even in patients without Scheuermann's disease.

Histologic studies have revealed anomalies of the vertebral endplates and growth plates suggesting that the etiology may be an alteration of endochondral ossification. Aufdermaur and Spycher [3], and Ippolito and Ponseti [4] identified similar anomalies with a lack of osteoporosis or avascular necrosis of the lateral margins. In fact, Aufdermaur and Spycher [3] suggest that the primary histologic lesion may be an alteration of the connective tissue fibrils of the vertebral endplates that are disrupted, irregular, and may be fragmented, which lead to weakening of the connective sheath of the vertebral endplates. Ascani and Montanaro suggested different elements [5], with the primary anomaly being mosaic ≈ alterations of the cells and the extracellular matrix of the vertebral growth plates and endplates. Rarefaction and thinning of the collagenous fibers and excessive proteoglycans characterize this pathological extracellular matrix. The cellular phenomenon of endochondral ossification is altered, slowed, or absent with bone formation taking place directly from cartilage without

the process of physiological differentiation. These phenomena alter the longitudinal growth of the vertebra.

Scheuermann's disease is, in fact, secondary to a "slowing in growth" rather than a process of destruction. This finding contrasts with the fact that the vertebral endplates lying outside of the deformed zone are not affected by this pathological process. In fact, these zones are characterized by accelerated growth and hyperplasia of the lateral margins, thus leading to a progressive increase in vertebral wedging. According to Pierre Stagnara, this could be due to defective adaptation of the spine to the standing or sitting position [6]. Studies showing similar anomalies in large primates who are quadrupedal but only occasionally bipedal, conform to this hypothesis [7]. Other studies measuring the bone mineral density in patients with Scheuermann's disease have shown that a kyphosis superior to 45° is accompanied by notable osteoporosis [8], leading to possible therapeutic options.

2. Genetics

A high incidence of Scheuermann's disease has been noted within families. Halal et al. studied the genetic transmission of Scheuermann's disease in 5 families with high incidences of this pathology and suggest an autosomal mode of transmission with a high degree of penetrance and variable expressivity [9]. The existence of this anomaly in the homozygous twin brother of a patient with Scheuermann's kyphosis is observed in almost 75% of cases. Within a 36-member family in whom signs of Scheuermann's kyphosis have been identified, the previously described observations have been found in 30% of the members, while only 4% are found in control subjects [10]. This anomaly may be seen in both males and females without an established sex ratio, although some authors have shown that boys are twice as likely to be affected than girls [10].

3. Mechanical

Patients with Scheuermann's disease are larger and heavier with a higher body mass index than control subjects [11]. Nevertheless, this finding is not correlated to the severity of the deformity and may correspond to a hormonal profile that is associated rather than causal. In fact, bone age is higher for chronological age in patients with Scheuermann's disease and the role of transitory growth hormone hypersecretion has been hypothesized but not proven [12].

Moreover, vertebral overloading with repetitive microtrauma has been incriminated in the development of the lumbar type of Scheuermann's disease, especially in male subjects.

Conclusions on the pathophysiology

The actual etiology of Scheuermann's disease remains unknown with variable and sometimes contradictory data resulting from histological studies of resected specimens. Typical disruption of the vertebral endochondral ossification may be the result rather than the cause of this pathology. Nonetheless, an evident genetic context and possible mechanical factors exist that could predispose to or aggravate the disease.

Functional signs

Primary reasons for consultation are twofold, with the first being obviously kyphotic deformity. This kyphosis is generally uniform in the upright position and appears angular when the patient bends forward (Adams forward bending test) (figure 1).

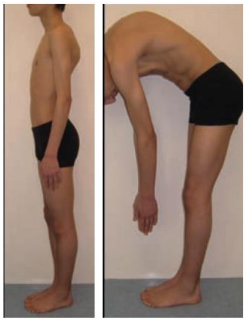

Figure 1: *uniform thoracic kyphosis in the upright position that becomes angular when the patient bends forward (Adams forward bending test) – The fingertip-to-floor distance is reduced and is probably due to hamstring tightness.*

The kyphosis in Scheuermann's disease is associated with a significant limitation in sagittal spinal mobility both in flexion and in extension [13].

Pain is also a frequent reason for consultation. Pain at the apex of the deformity may be secondary to degenerative phenomena of the vertebra endplates and the intervertebral discs, as may be seen on MRI [14]. The rigidity of the thoracic kyphosis may explain the excess mobility of the adjacent cervicothoracic and thoracolumbar junctions [11]. The increased cervical and lumbar lordosis restoring the sagittal balance on either end of the thoracic kyphosis (figure 2) may in and of itself be the cause of the pain due to posterior interarticular impingement. The pain often radiates to the paraspinal area and distal to the level of the deformity [15]. Patients with Scheuermann's disease of the thoracolumbar spine, more specifically the lumbar spine, are especially prone to present with pain from a disrupted sagittal alignment.

The disordered global sagittal alignment of the spine with anterior translation of the head leads to muscle contractions that may damage certain muscles, such as the pectorals or the hamstrings [16] (figure 1).

Physical exam

Similar to all spinal deformities, the physical exam starts by measuring the patient's length in the standing position, then only the head and trunk segments in the seated position. Clinical measurement of the sagittal curves may be done using a plumb line: distance between the plumb line running tangent to the apex of the kyphosis and the protrusion of C7, the apex of the lumbar lordosis, and the convexity of the sacrum (figure 2).

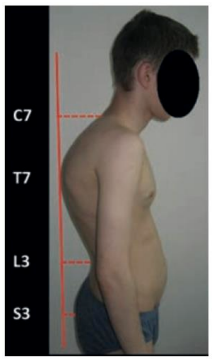

***Figure 2**: Exaggeration of cervical and lumbar lordosis on either end of the thoracic kyphosis restoring the sagittal balance – the plumb line allows for the measurement of the sagittal curves (C7: 7th cervical vertebra, T7: 7th thoracic vertebra, L3: 3rd lumbar vertebra, S3: 3rd sacral vertebra)*

These measurements quantify the deformity and assess the results of the suggested treatment. In severe forms, the plumb line that runs tangent to the apex of the deformity lies greater than 2cm than the clinical contour of the sacrum (figure 2). A minor gibbosity (rib hump) may be found indicating a small scoliotic curve, often of the lumbar spine, that is often a non-structural deformity. Moderate pectus excavatum with a caved-in appearance of the thorax beneath the mammary glands, induced at least partially by the patient collapsing into kyphosis, accompanies quite often such kyphotic deformities (figure 3).

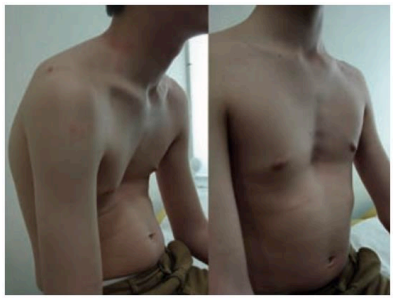

***Figure 3**: Moderate pectus excavatum with caved-in appearance beneath the mammary glands induced by the collapse into kyphosis.*

Pectus carinatum is also possible but rare. Tightening of the hamstrings (figure 1) may be noted and evaluated with the popliteal angle. In some cases, posterior horizontal stretch marks may be seen at the apex of the kyphosis (figure 4), thought to be due to the increased straing on the cutaneous and subcutaneous tissues.

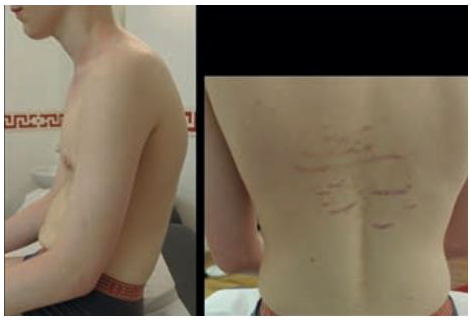

Figure 4*:* posterior horizontal stretch marks at the apex of the kyphosis.

Examining the limbs, the overlying skin, and a complete neuro-orthopedic exam complete the physical assessment.

Finally, the psychological burden of the disease felt by the child must be assessed, who is often introverted. This global attitude of collapsing in oneself may be the consequence of the deformity but may also be an element exacerbating the kyphosis.

Radiographic signs

Radiographic diagnosis is based on whole-spine radiographs in the standing position using the EOS® system with measurement of the sagittal Cobb angle (between the tangent to the superior vertebral endplate of the upper end vertebra and the inferior vertebral endplate of the lower end vertebra) (figure 5). A sagittal Cobb angle superior to 40° at the level of the thoracic spine is deemed as pathological [17]. Pierre Stagnara considered that this cutoff value may be too stringent, suggesting instead that every patient has their own, personal sagittal alignment [6]. The sagittal Cobb angle is an indicator of the functional prognosis and is considered of poor prognosis if it is superior to 75°. Sagittal angular alterations are generally less important in patients with thoracolumbar or lumbar deformities with a limited wedging of the vertebra, giving way instead to Schmorl nodes. The Cobb angles of adjacent lordotic curves as well as the variable differences in sagittal balance allow a complete analysis of the deformity (figure 5).

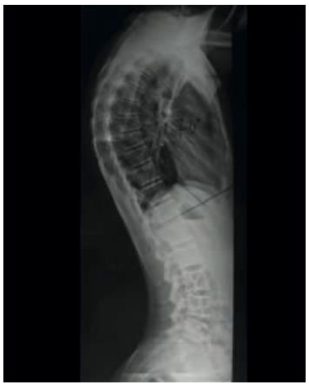

Figure 5*: Sagittal spinal radiograph of a subject with Scheuermann's disease of the thoracic spine: Wedging of the vertebral bodies around the apex of the kyphosis with vertebral endplate irregularities.*

The radiographic diagnosis of Scheuermann's disease requires four criteria established by Sôrensen (18):

Anterior wedging across 3 consecutive vertebrae around the apex superior to 5°, vertebral endplate irregularities, intervertebral disc space narrowing, and intravertebral disc herniations. All of the above criteria must not necessarily be met for the diagnosis to be made. Schmorl nodes correspond to a depression of the vertebral endplate and result from the penetration of the nucleus pulposus into adjacent cancellous bone. This finding is not specific to Scheuermann's disease and may be found even in subjects without vertebral pathology. Separation of the anterior vertebral margins (limbus vertebrae) at the level of the lumbar spine is secondary to the same pathophysiologic mechanism [19] (figure 6).

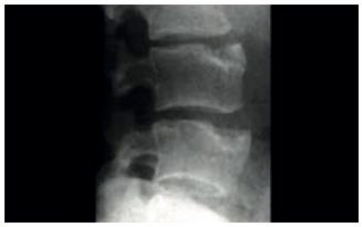

Figure 6*: Schmorl node and separation of the anterior vertebral margins (limbus vertebrae) at the level of the lumbar spine.*

Differential diagnosis

Postural kyphosis differs from Scheuermann's disease by a reducible deformity both passively and actively (figure 7), and by the absence of vertebral wedging and other radiographic typical signs of Scheuermann's kyphosis. Kharrat and Dubousset described a type of kyphosis, probably congenital in nature, with progressive anterior fusion of the intervertebral discs which, prior to the appearance of these anomalies, could be falsely labeled as Scheuermann's disease [21]. Other types of kyphosis (spondyloepiphyseal dysplasia, type I neurofibromatosis, dysraphism) are characterized by distinct radiographic signs, relevant history and clinical elements allowing to rapidly establish the correct diagnosis.

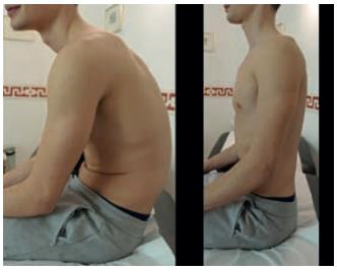

Figure 7*: Actively reducible [ostural thoracic kyphosis.*

In sum, Scheuermann's kyphosis is a frequently encountered deformity that may lead to functional decline. The consideration of certain elements as well as knowledge of the natural history of the disease will guide management.

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