Treatment of Scheuermann's disease

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Introduction

Holger Werfel Scheuermann, a Swedish physician, described in 1920 a painful thoracic hyperkyphosis which he linked to the "apprentice kyphosis", a deformity previously described by Schanz in 1891 [1]. Scheuermann's disease is a disorder affecting the growth of the vertebral bodies. It corresponds to a spinal osteochondritis in children, appropriately named osteochondritis deformans juvenilis dorsi. The prevalence of this disease in the general population varies from 2 to 10% [2]. However, these estimates are generally for the typical form of Scheuermann's kyphosis, thereby underestimating its prevalence. Furthermore, some studies show a higher prevalence in males compared to females [2].

Scheuermann's kyphosis is a disco-vertebral pathology due to the bipedal nature and erect spine of human beings. It occurs at the onset of puberty, at around 11 years of age in girls and 13 in boys. The vertebral bodies at this age are more fragile and vulnerable, and mechanical load leads to irregularities at the bony contours and to vertebral deformities [2]. The etiology of Scheuermann's disease is not well understood and is believed to be multifactorial. A multitude of factors have been suggested: Genetic predisposition; repetitive microtrauma (for example intensive practice in certain types of sports), especially between 8 and 12 years old (i.e. at the midpoint through the growth spurt); sagittal balance alterations leading to microtraumatic events with repetitive load on the anterior part of the vertebral bodies [2-4]. Growth is thus delayed at the anterior portion of the vertebral bodies and remains normal posteriorly, thus causing an increasingly hyperkyphotic spine during growth. The resulting cartilaginous and vertebral lesions are irreversible, even though the progression of the disease is halted at the end of growth. Patients may experience pain during adulthood secondary either to the vertebral deformities or to the appearance of intervertebral disc herniations that are probably due to the disease process itself.

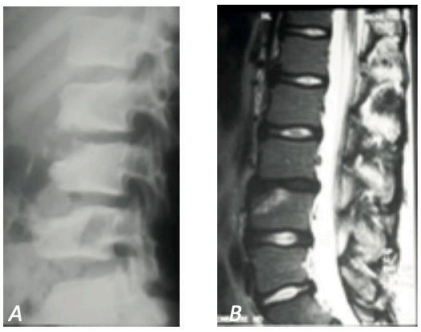
Scheuermann's kyphosis is characterized by a thoracic kyphosis superior to 45°. Sorensen [5] described, in 1964, the radiographic characteristics of the disease. Of note, Anterior wedging across 3 consecutive vertebrae superior to 5° associated with a T5-T12 kyphosis superior to 45°. The concept of Scheuermann's disease, which was initially strictly thoracic in its topography, was later extended to other spinal segments, which are deemed atypical. The latter are distinguished from the typical form either by a different location of the deformity (i.e. thoraco-lumbar or lumbar), or by the presence of distinct radiographic features. As a result, new radiographic criteria were described by Cleveland in 1981 [6]. In these atypical

forms, the presence of kyphosis is no longer a requirement. Actually, Scheuermann's disease includes the aforementioned typical and atypical forms, and is characterized by the presence of at least two of the following radiographic criteria: Anterior wedging across 3 consecutive vertebrae superior to 5° , T5-T12 thoracic kyphosis superior to 45°, or endplate irregularities.

Scheuermann's disease should ideally be treated in its early forms in order to benefit from the potential residual growth of the spine and limit the development of hyperkyphosis. Unfortunately, patients tend to present for consultation with a spine that is already stiff and painful. Physiotherapy may beneficial, especially in the preservation of mobility, and surgery is generally not indicated except in severe forms.

1. What is the natural history of Scheuermann's disease?

It is estimated that 26% of adults have sequalae of Scheuermann's disease [7] and 76% of the general population have Schmorl nodes [8]. The earlier the onset of Scheuermann's disease, the higher the risk of disease progression [4]. Less progressive forms are known to be benign. In the absence of marked degradation in sagittal alignment and severe inter-vertebral disc disease, the pain encountered during adolescence decreases with skeletal maturity and is rarely carried over into adulthood [9-10]. Nevertheless, it is difficult to imagine that a painful, deforming, and stiffening disease of adolescence could be asymptomatic during adulthood. Scheuermann's disease may be associated with persistent back pain [11-12] and spondylolysis [13]. Intervertebral disc disease is irreversible and may be exacerbated at the onset of adolescence [14-16] (figures 1A and A). Thoracic kyphosis superior to 70° generally leads to poorer functional results [17].Thoracolumbar and lumbar kyphosis (low pelvic incidence and lumbar lordosis) are not well tolerated and deteriorate faster than kyphosis purely of the thoracic spine. During adulthood, patients rarely complain of the aesthetic appearance of their backs but have a tendency to work less physically demanding jobs than their counterparts. Restrictive lung disease may also be present, but only in severe kyphosis superior to 85°. Some neurological complications secondary to kyphosis are possible but exceptional (sensory deficits at the level of the trunk, nerve root or spinal cord compression by herniated discs, and myelopathy at the apex of the kyphosis) [18].



Figures 1A and B*: Scheuermann's disease of the lumbar spine with degenerations of the intervertebral discs and the anterior margins of the vertebrae, confirmed by MRI.*

Recent studies classify the different types of backs according to the magnitude of the sagittal curves and spinopelvic parameters. The primary classification differentiating between varying types of backs is that of Roussouly [19] (figure 2). Roussouly's classification is based on the

sacral slope (SS) and pelvic incidence (PI) and classifies backs into 4 different morphotypes. Sacral slope is the angle formed between the horizontal and the sacral plate. Pelvic incidence is the angle formed between a line perpendicular to the sacral plate at its midpoint and a line connecting this point to the femoral head axis. The advantage of pelvic incidence is that it is relatively stable in a given individual and varies only slightly with age. A type 1 back is defined as a SS <35°, with a low PI and a short and disharmonious lumbar lordosis (called a junctional kyphosis). Type 2 backs also have a SS <35° and a low PI and a more prolonged and harmonious lumbar lordosis. Type 3 backs have a SS between 35 and 45° with a good sagittal alignment. Type 4 backs have a SS >45° and a high PI, which characterize a spine with marked thoracic and lumbar curves. The lower the SS or the PI, the stiffer and flatter the spine, due to a lower lumbar lordosis. Contrarily, the higher these parameters, the higher the lumbar lordosis. Backs with a Roussouly type 1 and 2 are considered as flat backs, and types 3 and 4 as having harmonious curves. From this classification, certain at-risk situations may be identified. In fact, "flat" backs do not allow appropriate adaptation to situations that are highly stressful on the spine. This is especially true when excessive axial load is placed on the spine, as well as during hyperextension of the spine. In the general population, Roussouly types 1 and 2 are at increased risk of developing Scheuermann's disease, independently from their participation in physical exercise [20-21]. This was shown by Jiang who compared 55 adolescents with Scheuermann's disease to 60 control subjects [22]. Adolescents with Scheuermann's disease had a tendency to present a significantly lower pelvic incidence compared to control subjects (32 vs. 45°, respectively).

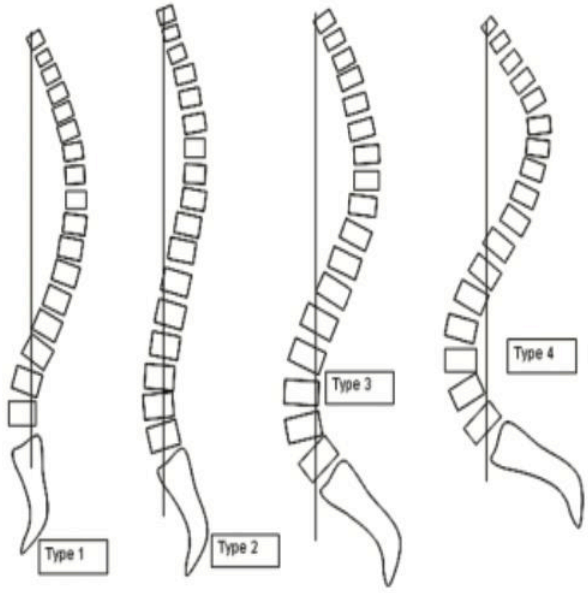


***Figure 2**: Roussouly's classification.*

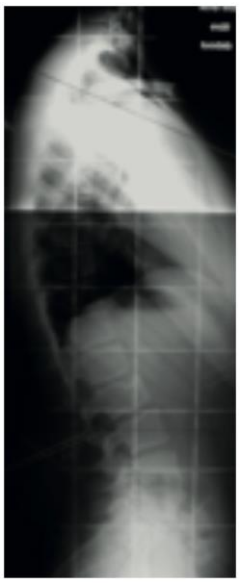


Figure 3*: Junctional kyphosis (Roussouly type 1).*

2. Can the natural history of Scheuermann's disease be prevented?

The natural history of Scheuermann's disease may be only partially prevented. By focusing on aspects that are secondary to repetitive microtrauma, it may be possible to slow the progression of the deformity. In fact, the only strategies having shown favorable outcomes in this regard are corrective orthopedic and surgical treatments.

2.1. Prevention

Screening of at-risk individuals is paramount, including subjects with family history of Scheuermann's disease, and especially patients with low pelvic incidence (predisposing lumbar and thoracolumbar osteochondritis, which are less well tolerated than the thoracic forms). Preventive strategies in children with kyphosis must be undertaken at the youngest possible age. This includes postural exercises with shoulder retropulsion.

The following general preventive measures must be adhered to:

- Adaptation of the profession: Prolonged sitting increases constraints on the anterior portion of the spine. As a result, seated positions with a curved back must be avoided as much as possible, and patients with Scheuermann's disease must favor non-seated professions.
- Adaptation of the work environment (chairs, tables, workstations): Ergonomic kneeling chairs (figure 4) may be used during prolonged sitting (homework, video games, etc…). This would force the child into a straighter posture. Although no studies have shown its effectiveness, ergotherapy may allow proper adaptation to the professional environment.
- Adaptation of practiced sports: Allowed types of sports include those with limited jumps, shocks, blows or falls.
- Avoiding overweight with the help of nutritionists if necessary.
- Avoiding wearing heavy items on back (backpacks).

Figure 4*: Ergonomic kneeling chair. The patient's weight is supported by the knees, the chair is lightly inclined. As a result, the patient is forced to maintain an upright posture.*

2.2. What types of sports may be safely practiced?

Physical education at school is rarely contra-indicated. If required, the physician may provide a written note partially excusing the patient from sports placing high stressors on the spine according to the official bulletin of the ministry of education. Physical exercise is authorized in a recreational rather than performance capacity in order to reduce microtrauma. Activity levels must be monitored and limited, sports unloading the spine preferred, sufficient recovery time provided, and a healthy lifestyle promoted [23]. Scheuermann's disease and high-performance sports are not always compatible and the link between the two have been well established [23]. As a rule, thoracic kyphosis and lumbar lordosis have a tendency to increase with the number of hours per week of physical exercise [24,27]. The prevalence of Scheuermann's disease of the lumbar spine increases with the intensity of training, especially in boys carrying heavy weights [25].

In his thesis, Cubillé discussed Scheuermann's disease and its associated factors in 97 young high-performance skiers [26]. His study showed an increased prevalence of back pain and Scheuermann's disease in this population. Partaking in intense, competition-level alpine ski seems to be the principal driving force behind the appearance of this pathology. The mechanical loads placed on the spine during these activities are extremely high, thus placing all professional skiers at risk [26].

Furthermore, intensive gymnastics increases thoracic kyphosis, especially in male athletes [27]. A specialist should be consulted in order to ascertain each patient's actual risk factors. Certain sports to avoid in an intensive manner are those which place high strains on the spine, such as activities requiring flexion of the spine thereby increasing the thoracic kyphosis. This includes horseback riding [28], wrestling, rugby, football (soccer), judo, field hockey, diving, parachuting, skiing [26,29], tennis, table tennis, and hockey [30] among others. Sports that theoretically should be recommended instead include those that extend spine, including dancing [31], rhythmic gymnastics, fitness, volleyball, handball, and backstroke swimming among others. Nevertheless, there is no consensus between the different studies as to the effects that football (soccer) [32], volleyball [33], handball [34], and tennis [35] have on Scheuermann's disease. Swimming increases muscle strength while reducing the load on the spine, but intensive swimming, especially butterfly strokes, may increase Scheuermann's disease of the thoracic spine [30].

During the painful stages of the disease, sports is contra-indicated for an extended period of time. If the pain has completely disappeared and radiographic signs have stabilized for at least 3 months, progressive return to sports may be allowed.

When no contra-indications to physical exercise have been found during a consultation, preventive strategies must be highlighted. Care must be taken when physical activity surpasses 8 to 10 hours per week, or when joining a school sports team, sports training facilities or a national team. Adjustments to the volume or intensity of training may be suggested if they are judged to be unsuitable or poorly experienced. Children requesting permission for “playing-up” require a specific medical exam.

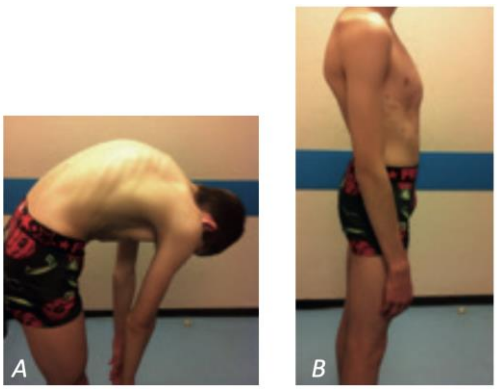
3. Functional treatment

3.1. Symptomatic treatment

Even though hyperkyphosis is localized to the thoracic spine, pain is generally located outside of the stiff area, especially at the thoracolumbar junction and at the lumbar segment. The pain is persistent, mechanical in nature with occasional inflammatory episodes, and may progress in flares that may be disabling. Ergotherapy, physiotherapy, shockwave therapy, infrared therapy, and electrotherapy may provide pain relief. Consultation with a pediatric pain specialist may be particularly helpful.

3.2. Rehabilitation

According to a recent meta-analysis [36], rehabilitation has a positive effect on thoracic kyphosis. strength training in particular should be preferred: abdominal, lumbar and erector spinae muscles, and postural correction [29]. Effects are much less tangible in terms of lumbar lordosis, for which strength training and stretching must be simultaneously undertaken (posterior chain and iliopsoas). Hamstring tightness are a known risk factor for sagittal decompensation [37,38] (figures 5A and B). Postural exercises counteract posterior shift of the spine, pelvic retroversion, shoulder antepulsion, and anterior shift of the head [4]. Unfortunately, these exercises do not prevent the progression of the deformity, but may decrease the stiffness of the spine, especially when associated with bracing.



***Figures 5A and B**: Adolescent with thoracic kyphosis and hamstring tightness.*

4. Bracing

Nonoperative treatment has two goals:

- Limit progression of the disease and preserve the intervertebral discs by reducing the load on the anterior portion of the spine
- Control the structural deformity of the thoracic kyphosis and prevent the appearance of compensatory curves at the lumbar and cervical segments [36].

In patients with higher remaining potential for spinal growth (Risser <2), correction of the deformity is generally more durable if spinal hyperextension during treatment allowed for compensatory growth of the anterior portion of the spine [39].

There is actually no consensus on the optimal type of brace and length of treatment. The effectiveness of treatment is assessed based on the restauration of acceptable sagittal alignment, correction of the anterior wedging of the vertebral bodies, and the appearance of signs of healing in case of a fracture of the marginal borders of the vertebra. Complete correction or ad integrum restitution of the kyphotic deformity is generally not possible, except for some very early cases. Intervertebral disks generally remain deformed and only moderate correction of the vertebral wedging is possible [40]. Bracing is generally necessary for a long period of time and maintained until skeletal maturity, which tends to discourage adolescents who are often unable to tolerate it. Even though the effectiveness of bracing has been established by some authors, the overall outcomes are deceiving. After weaning of the brace, loss of correction is expected (between 5 and 20°), suggesting that, even though bracing expands the anterior intervertebral space, its effects are temporary [14]. Effects are even less significant in the treatment of lumbar disease. Treatment must therefore be undertaken in the early stages of the disease, before wedging of the vertebral bodies has taken place.

4.1. Immobilization

Temporary immobilization is indicated only in acute, painful episodes with the aim of relieving pain, and is not recommended for prolonged use, as it may lead to atrophy of the paravertebral muscles.

4.2. Corrective treatment

When bracing is considered, a multitude of options exist, all of which must include a posterior support at the apex of the deformity, and 2 anterior supports parasternally in order to retropulse the shoulders.

4.2.1. Corrective cast brace

The use of a cast brace has the ultimate goal of reducing thoracic hyperkyphosis, thereby decreasing the mechanical load on the anterior portion of the vertebral bodies. This is meant restore the normal growth of the anterior part of the vertebrae and reduce wedging. The cast brace is inspired by the elongation-derotation-flexion (EDF) brace used in scoliotic patients. The indication for orthopedic treatment is generally progressive kyphosis >60° with residual

potential growth (and thereby the potential exacerbation in the absence of treatment) [9]. The spine must be relatively flexible in order to undertake this treatment (supine fulcrum extension test). This treatment modality is undertaken in association with physiotherapy and postural exercises.

The cast is molded on a Cotrel traction table [41]. The primary posterior support must be situated beneath the apex of the kyphosis, on both sides of the spinous processes. Two anterior supports opposing the previous one are placed at the level of the pelvis and the manubrium sterni. Flexion of the hips during cast molding reduces lumbar lordosis by retroverting the pelvis. Windows allowing for the expansions of the rib cage and the abdominal cavity must be conceived. The anterior thoracic window must be relatively large allowing for the expansion of the rib cage during breathing. Depending on brace tolerance, sequential casting is undertaken at 2- to 3-week intervals until correction is obtained. Occipital-mandibular support is not recommended in this case and should be reserved for patients with a deformed hyperkyphosis.

Cutaneous complications may develop at the level of the cast-skin points of contact. At the level of the pelvis, the window must also be large enough to allow flexion of the hips and avoid entrapment of the lateral femoral cutaneous nerve in the seated position.

4.2.2. Progressive correction by the addition of felt

Correction is obtained by progressively increasing pressure at the zones of contact with the addition of layers of felt to the initial cast, 2 to 3 weeks after initiation of treatment. An extra layer of felt is added on a weekly basis with surveillance of the skin at the zones of contact. Radiographic follow-up is necessary in order to assess the effectiveness of the correction.

4.2.3. Milwaukee brace

The principal of the Milwaukee brace may be tempting by its active auto-elongation effect on the trunk which tends to decrease the spinal curves in the sagittal plane. It is more frequently reserved for young children, although some authors have utilized it in adolescents. It may be undertaken as a standalone treatment or as a continuity of correction by casting [41]. Unfortunately, it is often badly tolerated by the patient and discontinued by almost 50% patients, especially those who are a little older [41]. Some authors have recommended the use of a thoraco-lumbo-sacral orthosis (TLSO) instead of the Milwaukee brace [42].

4.2.4. Rigid brace (figure 6)

Since the Milwaukee brace is often badly tolerated, it may be replaced by a bivalve brace [13]. This type of brace is molded on the trunk. During the molding process, the patient must be in a standing position with the hips partially flexed in order to decrease lumbar lordosis. Once more, cutaneous complications at the contact points must be monitored. Brace-wear must be continued during the entire period of residual growth of the trunk. A diurnal-only use of the brace may be allowed. A hypercorrective night-time brace may also be prescribed, although it places excessive stress on the spine and may not be well tolerated.

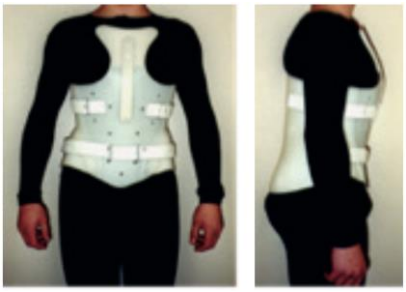


Figure 6*: Bivalve rigid brace*

5. Operative treatment

The need for operative treatment in Scheuermann's disease remains exceptional and controversial. A detailed chapter has been consecrated to this end (see "Is there a place for surgical management in adolescents with Scheuermann's disease?" by Antoine Hamel). Surgery is considered as the final recourse for pediatric patients who are at the end of their growth or in adults. It may be indicated in patients with severe kyphosis (>70°), back pain, neurological symptoms, and resistance to all other forms of treatment [37,38,41]. The surgical approach may include posterior or combined spinal fusion, with or without osteotomies of the apex of the deformity. Instrumentation may be hybrid or an all-screw construct. Fusion must be extensive, include 10-12 vertebrae, and must include the first lordotic intervertebral segment (often T2/T3 to L2/L3). Shorter constructs lead to post-operative loss of correction with the appearance of badly tolerated junctional kyphosis. In the setting of preoperative assessment, an MRI is ordered in search of spinal cord anomalies, herniated discs, or spinal cord compression at the apex of the deformity since the major risk of this surgery is the development of paraplegia. Nevertheless, in order minimize neurological complications, it is recommended not to overcorrect the kyphosis.

6. Indications

Indications for the choice of treatment depend primarily on the age of the child, stiffness of the deformity, location and number of vertebrae implicated, and refractory pain [42]. Thoracolumbar and lumbar disease often lead to moderate angular deformities which are more often painful due to the presence of Schmorl nodes; These are generally treated by conservative management, except in rare cases where neurological signs may be present, in which case surgical spinal cord decompression along with posterior spinal fusion would be necessary. In younger patients with thoracic disease, the deformity is rarely severe, is always flexible, and is manageable by conservative treatment. Rehabilitation is extremely effective in improving the posture, especially when the thoracic spine is flexible, and the sagittal curve is not extreme (45-55°). The rehabilitation strategy should concentrate on hamstring and pectoral stretching, as well strengthening the extensors of the spine. When the curve magnitude is superior to 50-55°, a full-time brace may be indicated (TLSO or Milwaukee). Rehabilitation may be associated with brace-wear but is in no way sufficient by itself as a means of correcting an already-structuralized kyphotic deformity. Conservative treatment must be pursued until the end of growth in order to prevent any loss of correction [12-13]. In adolescents, the majority of authors agree on the effectiveness of conservative treatment in flexible forms or in patients with a Risser<3. For patients with a stiff deformity, serial

corrective casting may be attempted as an initial treatment modality. A loss of correction of 10-20° after discontinuation of conservative treatment has been reported in at least 30% of patients [11]. For patients with a kyphosis >70°, functional results are less satisfactory [17]. Severe postural alterations that accompany more advanced forms of Scheuermann's disease are sources of discogenic pain at the junction between the structuralized, stiff deformity and the mobile segments [14]. Moreover, compensatory curves, such as lumbar and cervical hyperlordosis, are also sources of painful mechanical spinous process impingement. Scheuermann's disease tends to resolve in males between 16 and 18 years of age, with possible radiographic sequellae that may not be painful.

Conclusion

The optimal treatment modality in Scheuermann's disease is prevention. However, the majority of patients present at advanced stages where bony deformity has already been established and is almost impossible to reverse. The primary goal of treatment is to attempt to delay progression of the deformity. Conservative treatment by bracing is protracted and often badly tolerated by adolescents. Surgical management is exceptional and may be indicdated at the end of growth and only as a last resort.

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