Scheuermann's kyphosis: update on pathophysiology and surgical treatment

Amer Sebaaly1,2, Sarah Farjallah2, Khalil Kharrat2, Gaby Kreichati1,2 and Mohammad Daher1

1School of Medicine, Saint Joseph University, Beirut, Lebanon
2Orthopedic Department, Spine Unit, Hotel Dieu de France Hospital, Beirut, Lebanon

• Scheuermann's Kyphosis (SK) is a rigid spinal kyphosis. Several theories have been proposed concerning its pathogenesis, but it is, to this day, still unknown.
• It has a prevalence of 0.4–8.3% in the population with a higher incidence in females.
• Clinical examination with x-rays is needed to differentiate and confirm this diagnosis.
• Non-surgical management is reserved for smaller deformities and in skeletally immature patients, whereas surgery is recommended for higher deformities.
• Combined anterior and posterior approach was considered the gold standard for the surgical treatment of this disease, but there is an increasing trend toward posterior-only approaches especially with use of segmental fixation.
• This study reviews the pathophysiology of SK while proposing a treatment algorithm for its management.

Introduction

Scheuermann's Kyphosis (SK), first described by Horgel Welfer Scheuermann in 1920, is a rigid spinal kyphosis usually involving the thoracic or thoracolumbar area (1). Albeit several theories, its etiology is still unknown (2). SK is usually separated into two groups: typical and atypical SK. Typical SK is the more common type and has a mid-thoracic (T7–T9) apex in its deformity. It is associated with a hyperlordosis of both the cervical and lumbar spine (3). The atypical group is also known as 'Apprentice Kyphosis', and most commonly presents in athletic adolescent males or heavy lifters, the deformity’s apex is in the thoraco-lumbar or lumbar spine and this form is the most likely to progress (4). Sorensen’s criteria defined as anterior wedging of 5° or more in at least three vertebral bodies alongside endplate narrowing, and Schmorl nodes (5) are always met in the typical form but not necessarily in the atypical form (6).

Treatment for SK includes both conservative and operative treatment (2). The purpose of this narrative review is to provide a treatment algorithm for this disease after reviewing its physiopathology.

Etiology and pathophysiology

Until now, the exact etiology of SK is still unknown (7). Several theories were proposed concerning its pathophysiology. The first one was by Scheuermann et al. stating that an osteonecrosis of the ring apophysis in the vertebral bodies caused an arrest in anterior growth, thus causing this disease (8). This theory was rapidly rejected by Bick et al. showing that the ring apophysis has no effect on anterior longitudinal growth since it is not a part of the cartilaginous physis (9). On the other hand, Schmorl et al. postulated that herniated disks cause a loss of anterior disk height and result in growth disturbance and ultimately SK (7). This theory is not proven till now, but it is likely to be erroneous due to the fact that not only Schmorl nodes are common among normal people but also they are present in areas that are not involved in the deformity (7). Finally, Bradford et al. proposed a third theory that is based on the idea that osteoporotic vertebra are more prone to collapse under compression (10). Studies showed no significative difference in bone density between people with SK and the normal population resulting in a rejection of this theory (11).

The most accepted theory is the biomechanical theory. Ogden et al. suggested that there is a biomechanical process behind this kyphosis implying that biomechanical stresses alter the remodeling response in the vertebral bodies resulting in increased compressive forces on the anterior part of the vertebral body which will stop the growth and cause SK (7). Other biomechanical theories came out such as the presence of tight hamstrings in
patient with SK increasing the pelvic tilt when bending forward (12, 13). A smaller sternum was also associated with SK due to it increasing compression in the anterior part of the thoracic vertebral bodies leading to kyphosis (14). Bracing supports the mechanical origin of this disease by being a successful way of management (15).

Genetics was also used as a way to explain this deformity (16). Halal et al. showed an autosomal dominant transmission with high penetration of this disease but with variable expression (16). Other authors supported the presence of a genetic basis behind this deformity implying that family history of hyperkyphosis should be taken into consideration to exclude the possibility of SK (17). Candidate genes linked to SK such as IHH, SOX9 and PAX 1 were identified by Zaidman et al. (18). Damborg et al. showed a higher concordance by pairs and probands for monozygotic twins than for dizygotic twins indicating a higher genetic contribution to Scheuermann's disease (19). Findlay et al. also described a family where SK was present in three consecutive generations with a male to male transmission (16).

Other theories were suggested as an etiology of SK such as high levels of growth hormone, juvenile idiopathic osteoporosis, hypovitaminosis D, dural cysts, spondylolysis, infections, spinal malformations, etc. (20). It is possible that Scheuermann's disease is the result of the simultaneous action of several factors (21).

Natural history

Prevalence

The prevalence of SK in the population ranges from 0.4 to 8.3% (7). Some studies showed SK being more frequent in men (15, 22). In fact, the most accepted male to female ratio is between 2:1 and 7:1 (23). The age onset of SK is between 10 and 12 years old but an adulthood onset is not unheard of (24). Although the kyphosis angle was not influenced by sex, it was positively correlated to the age (25).

Evolution

The evolution of SK is benign. SK may only mildly affect the quality of life (QOL) of the patients affected by the disease compared to the general population (7). Patients presenting with a kyphosis lower than 60° have good clinical outcomes (22). To add to that, Murray et al. reported that patients with curves lower than 85° reported little concern with their physical appearance and had similar QOL to patients with curves lower than 60° (work absence due to back pain, interference of pain with daily activities, recreational activities, self-esteem and self-consciousness) (22). Ristolainen et al. followed patients with mild thoracic kyphosis for 46 years and reported an increase of a mean of 14° (from 46° to 60°) with no correlation between the extent of progression of the kyphosis and function (26).

In other studies, untreated SK resulted in ‘severe thoracic spine pain’ in nearly 50% of the cases (27, 28). Such discrepancies may be explained by the fact that these studies had patients with greater deformities (22). All in all the evolution of Scheuermann disease is still unknown since it is less studied than other spinal deformities like adolescent idiopathic scoliosis (7).

Clinical examination

The most common symptom at presentation is pain at the apex of the deformity in the pediatric population, whereas esthetic deformity is the most common symptom in the adult population (7). On examination, the patient demonstrates a varyingly flexible thoracic hyperkyphosis associated to an increased lumbar and cervical lordosis compensating for the thoracic deformity (7). These associated compensations may be a cause of pain (15, 29). The neck and head adopt an anterior position with a forward protrusion aspect (‘goose-neck deformity’) (Fig. 1). This characteristic posture leads to aesthetic disturbances and dissatisfaction with the patient’s own body image (30). Lumbar hyperlordosis produces an added increase in stress on the pars interarticularis, which could explain the increase in the incidence of spondylolysis, up to 11% (31). If a patient with SK presents with pain in the lumbosacral region, spondylolysis must be ruled out (32). Tightness in the iliopsoas, hamstrings, pectoralis and the anterior shoulder may also be present (33). When inspecting the back, a cutaneous pigmentation may be present at the apex of the kyphosis caused by skin friction due to the protruding spinous process (22, 34).

Figure 1

Clinical presentation of a 15-year-old boy with SK. Note the ‘goose-neck deformity’ and the angular kyphosis with Adam’s forward bending test.
Scoliosis may be associated to the hyperkyphosis. This may be caused by the irregularities at the vertebral endplates of every level of the kyphosis with a presence of a lateral wedging (19). The present information is not enough to confirm this hypothesis, but it is important to note that if it is correct, then the characteristics of this scoliosis could be identified and it should be not confused with an idiopathic scoliosis in order to efficiently correct the deformities in both planes (19).

Associated neurologic abnormalities are rare but if present, an MRI of the thoracic spine must be ordered (7). The present neurological symptoms may be due disk herniation, severe kyphosis or dural cysts (35). Restriction of pulmonary function is rare and is usually present in cases where kyphotic curves surpasses 100° (22). Finally, SK may be a risk factor for ossification of the ligamentum flavum and thoracic disk herniation, but it is not associated to an ossification of the posterior longitudinal ligament (36). As a matter of fact, a recent study found that 95.2% of thoracic disc herniation occurs in patients with typical or atypical SK (37).

**Differential diagnosis**

Parents and general practitioners sometimes attribute the onset of kyphosis to poor posture, resulting in delayed diagnosis and treatment (15, 34, 38, 39). It is very important to differentiate Scheuermann’s disease from curved dorsothoracic or postural kyphosis. Adam’s forward bending test may be helpful in differentiating the two entities (Fig. 1). With postural kyphosis, the deformity disappears on forward bending, whereas with SK, the deformity is increased alongside a transition in the thoracolumbar region (7). Other conditions such as osteochondral dystrophies and spondyloepiphyseal dysplasias, congenital kyphosis, spondylodiscitis, sequelae of vertebral compression fractures, post-laminectomy kyphosis and neoplasms should also be considered (7, 15, 40).

**Imaging**

**X-ray**

Initial imaging should include a standing postero-anterior and lateral radiographs of the spine. Cobb angle is measured on the lateral radiograph to assess the degree of kyphosis. The classical normal of kyphosis was thought to be between 25° and 45° (41). A recent study found that the thoracic kyphosis changes with the PI with a nearly constant upper arch of the kyphosis (41). Other signs include a round back (Fig. 2) on lateral view alongside vertebral wedging. Sorensen’s signs must also be searched for and assessment for Schmorl nodes (Figs. 3 and 4). Scoliosis, increased lumbar lordosis and spondylolysis may also be present (4). C7 plumb line may be found lying behind the sacral promontory instead of being within 2 cm of it (39, 42) showing a negative sagittal balance (Fig. 2) (7).

If surgical management is considered, flexibility of this deformity should be assessed and the patient is positioned placing a bolster under the thoracic spine to have a hyperextension lateral radiograph of the deformity (7).

**MRI**

Authors recommend ordering a pre-operative MRI for several reasons. First, a normal neurological exam can coexist with an abnormal MRI, with the rate of abnormal findings not reported in the literature (5, 43). Secondly, MRI is used to rule out abnormalities of the neural axis such as Chiari malformations, syrinx, stenosis, spinal cord impingement and findings that can alter the operative plan. In fact, Lonner et al. found a 19.7% chance of anomalies in patients to be operated for SK: low-lying conus (2.3%), syrinx without Chiari malformations (17.4%) posterior disc herniations and spondylolysis (8.1%). An interesting fact is that 4.7% of the cases had the operative plan changed as a result of the pre-operative MRI due to either neural compression, disc herniation or to a spinal cord draped over the apex (5). Thus, the probability of causing spinal cord damage during corrective surgery of
Management

The management of SK, whether conservative or surgical, can be very challenging. There is no clear consensus on treatment indications, guidelines and protocols, and treatment strategies are controversial (Table 1) (2, 45, 46, 47).

Non-surgical management

In 2010, the International Society on Scoliosis Orthopaedic and Rehabilitation Treatment SOSORT has evaluated the use of non-operative techniques along with specific braces and physiotherapy techniques in the treatment of SK. Non-operative management includes exercises and lifestyle modification, pain medication and bracing (48). The indication for non-operative management varies for each treatment depending on the kyphosis angle, rigidity and symptoms (49). When kyphosis is <60° and asymptomatic, physical therapy and lifestyle modification with NSAID are recommended (50). The main rehabilitation techniques are extension stretching/strengthening, hamstring stretching and proper self-postural control. This protocol should be repeated at home daily for 20 min (45). Exercises and lifestyle modification are also indicated in rigid kyphosis because they increase the trunk’s range of motion. This leads to reduced plaster series, better adaptation to braces and better correction (51).

The main indications to bracing are 60°–80° kyphosis, pain and rigidity. Wearing a brace can prevent the collapse of the anterior wall of the vertebral body by decreasing mechanical stress on the anterior wall of the vertebral body (50). They are mostly effective in skeletally immature patients, in which the threshold for implementing the brace is lower than 60°, because brace impedes progression of curve. The best time of application of the brace is at the onset of puberty. Braces should be worn for about 12–24 months and removed at the end of growth (45). Brace wear is recommended for 16–23 h per day until apical wedging is corrected (2). Brace model and wearing time differ depending on the type of SK and the age of the patient:

- For thoracic SK: Most experts suggested the use of 4–5-point bracing systems which must be worn all night and for part of the day for a total of 22 h (2). Five-point system is better used in the case of muscular imbalance (51).
- For a Thoraco-lumbar SK: The brace must be worn during the day in the sitting position and the ideal brace is a 4-point system.
- For a juvenile SK: The brace must be worn part time with a total of 16 h, and the ideal brace is the Milwaukee brace (45).
The adult age, the two-stage anteroposterior fusion is ideal with more rigid curves (51). Two-stage approach is the method of choice when deformation is significant with more rigid curves. Two-stage approach was found to be more efficient at reducing the kyphotic deformity, as indicated by a greater correction rate, bolster bending correction index (BBCI), and gain in spinal height (46). The two-stage anteroposterior fusion was also associated with reduced incidence of junctional failure when using various forms of instrumentation, including Cotrel-Dubousset instrumentation and Luque rods (55). This lead for the recommendation of the two-staged approach as the gold standard in the surgical treatment of SK. However, with the advancement of the surgical techniques (Ponte osteotomies) and instrumentation (multi-segmental posterior pedicle screws), there is a shift toward posterior-only approach (Fig. 5) (56).

In the 80s and 90s, recommendation for severe curve correction was dual approach (38). Since Bradford et al. carried out the first report on posterior fusion in the treatment of SK, the surgical indications, as well as the operating techniques, have altered significantly (57), and modern single-stage posterior-only fusion now typically incorporates a Smith-Petersen or Ponte osteotomy (58).

Considering the controversy surrounding the best form of surgical treatment, many studies compared the results of treatment using the dual approach (DA) with treatment using posterior approach (PO). In Yun’s et al. systematic literature review and meta-analysis, outcomes of Scheuermann's disease after DA and PO approaches were compared. Blood loss, length of surgery time, PJK/DJK and return to surgery seemed to be less in the PO group than that in the AP group (59). The sagittal vertical axis was the only significant spinopelvic parameter in favor of combined anteroposterior in Mcdonnell’s retrospective review on two-stage anterior and posterior fusion versus one-stage posterior fusion in patients with SK.

Levels of fusion

Dubousset and Guillaumat in 1987 recommended that the lower limit should be the lumbar vertebra just above the horizontal disc on the hyperextension lateral Xrays (60). The most appropriate lowest instrumented vertebra (LIV) was the level below the first lordotic disk (FLD) (55). The FLD is defined as the most proximal thoracolumbar or lumbar disk below the level of the kyphosis with ≥5° of anterior opening. Because of hyperlordosis in the lumbar spine in patients with SK, determining the FLD can sometimes be difficult, thus leading to fusion short of the true FLD and subsequent DJK (2). The sagittal stable vertebra (SSV) was defined by Lenke as the most proximal touched vertebra by the posterior sacral vertical line (61). In a matter of fact,

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**Table 1  Management techniques for Scheuermann’s Kyphosis (SK).**

<table>
<thead>
<tr>
<th>Conservative management</th>
<th>Surgical management</th>
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<tbody>
<tr>
<td><strong>Lifestyle modification</strong></td>
<td>Two-stage anterior and posterior fusion (56)</td>
</tr>
<tr>
<td><strong>Physical therapy</strong></td>
<td>UIV: T2 (66)</td>
</tr>
<tr>
<td><strong>Extension stretching/strengthening</strong></td>
<td>LIV: FLD (56) or the SSV (63). However, the SSV is better since it decreases the risk of DJK when compared to FLD (63)</td>
</tr>
<tr>
<td><strong>Hamstring stretching</strong></td>
<td>One stage posterior-only fusion (57)</td>
</tr>
<tr>
<td><strong>Proper self-postural control daily for 20 min (46)</strong></td>
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<tr>
<td><strong>Pain medication (49):</strong></td>
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<tr>
<td><strong>Anti-inflammatory drugs (46)</strong></td>
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<tr>
<td><strong>Muscle relaxant (46)</strong></td>
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<tr>
<td><strong>Bracing (49):</strong></td>
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<tr>
<td><strong>Thoracic SK: 4-5-point bracing for 22 h/day</strong></td>
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<tr>
<td><strong>(must be worn all night) (2), or 5-point system in case of muscular imbalance (52).</strong></td>
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<tr>
<td><strong>Thoraco-lumbar SK: 4-point bracing worn during the day in sitting position (51).</strong></td>
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<tr>
<td><strong>Juvenile SK: Milwaukee brace worn part time for 16 h/day (46).</strong></td>
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</table>

FLD, first lordotic vertebra; LIV, lower instrumented vertebra; SSV, sagittal stable vertebra; UIV, upper instrumented vertebra.

Older patients are also likely to have less flexible curves that cannot be corrected with external pressure alone. While skeletal maturity may not necessarily be a contraindication to bracing, it is generally thought that skeletally mature patients are poorer candidates for bracing and more appropriately treated with surgery (52). Historically, operative intervention in younger patients with remaining growth potential was associated with high risk of failure given the crankshaft phenomenon (47). Nonetheless, this complication has decreased significantly with patients with newer segmental instrumentations (47).

Surgical management

The main indication for surgical management of SK is (1) painful kyphotic deformity that does not respond to non-surgical treatment, (2) cosmetically displeasing deformity, (3) functionally progressive curve or (4) neurologic deficit/spinal cord compression (7). Traditionally, surgery has been offered when the curve magnitude exceeds 65°–80° degrees, especially in adults (53). Cardiopulmonary indications for surgical intervention are rare and appear only in patients with curves >100° (22). In the context of progressive deformity, older patients are likely to have more severe and less flexible curvature amenable only to operative correction (54).

Correction may be achieved using one of two surgical strategies: a two-stage anterior and posterior fusion or a one-stage posterior-only fusion. It is still controversial the better strategy for better outcome (31). Age should be taken in consideration when choosing the surgical strategy: during the period of growth, posterior arthrodesis is the preferred may be the method of choice, while at
the FLD is more proximal than the sagittal stable vertebra (SSV). According to Cho et al., choosing the SSV instead of the FLD decreases the incidence of DJK (61). Patients who were fused at or below the SSV using posterior-only construct were found to have markedly greater lordotic disk angles below the LIV and lower revision surgery rates for DJK compared to patients who had an LIV proximal to the SSV (5% vs 36.3%) at the expense of incorporating additional motion segments (62). The choice of the LIV is important to decrease the occurrence of DJK. A recent meta-analysis found a DJK incidence in patients fused to the FLV to be 43.6% compared to 5.9% in the SSV group with a risk reduction of 86% (63). The latest recommendation by Ames & Lenke in 2019 is using the SSV concept for distal level selection: ‘If the selected SSV is just barely touched by the sacral vertical line, the adjacent disk space should be evaluated further. If the proximal disk space is lordotic, the “barely touched” SSV is still a safe choice’ (2).

After the distal fusion level is selected, maintaining symmetry of the construct overall is recommended. Proximally, a general consensus exists in recommending the inclusion of the proximal end vertebra (which is generally T2) in the construct (64). The fusion should extend roughly the same extent from the apex proximally and distally, with some consideration for adding one additional proximal fusion level to ensure that the proximal end vertebra is also included in the fusion construct (2).

Complications
Among non-operatively managed patients, loss of correction occurred in at least 30% of patients once the brace is removed, even if they were compliant to brace wear (30).

Among operatively treated patients, the most common complications, mechanical mainly, were hardware failure and proximal or distal junctional kyphosis. Combined anterior-posterior procedures were additionally associated with neural, pulmonary and cardiovascular complications.

Figure 5
Case of a 15-year-old male who presented with a 100° kyphosis (A). A fulcrum test showed that the most horizontal disc is L3-L4 (B). He was operated with multilevel Ponte osteotomies (C). The postoperative x-ray showed a thoracic kyphosis of 50°.

Figure 6
Proximal junctional failure (A) with proximal hooks pullout. (B) Distal junctional failure with screw pullout.
and greater incidence of infection with two-stage anteroposterior fusion (45).

PJK/DJK (Fig. 6) risk factors include short fusion, greater pre-operative sagittal imbalance, correction of sagittal deformity, more posterior LIV plumb line, low bone mineral density and a smaller postoperative kyphosis (65, 66). Hypercorrection is one of the most common risk factor for PJK/DJK and final kyphosis should be tailored to patient's pelvic incidence (41, 57). There are conflicting data whether age at the time of presentation is risk factor for DJK with some authors finding younger age to be a risk factor (66), whereas other found increased age to be risk factor (65).

The overall complication rate specifically for combined-approach surgeries was 19.6%, and posterior-only procedures had a 9.9% ate (67) However, hardware failure is more important in posterior fusion (47).

Treatment algorithm
Based on this review, we propose the following algorithm for the treatment of SK (Fig. 7). The main criteria for the selection of optimal treatment are the patient’s age, the Cobb angle magnitude and clinical impact of the disease. In summary of this algorithm, non-operative treatment is the most indicated at when curve magnitude is less than 60°. When SK Cobb angle is between 60° and 80°, the decision depends on the age of the patient. If the patient is skeletally immature (before or at puberty), bracing is the best option. The type of the brace and time for wearing it depend on the type of SK. on the other hand, if the patient is skeletally mature, the two options are either surgery or physical therapy. Surgery is indicated in the setting of neurological impairment, progressive curve or bad cosmesis. In other cases, physical therapy should be recommended and posterior instrumentation with multilevel osteotomies for the surgical cases.

Conclusion
In conclusion, SK is a disease whose etiology is still unknown. It has a wide spectrum and a lot of differential diagnoses. It usually presents with pain and most of the times imaging techniques along the clinical exam are needed to diagnose it. An MRI is usually needed before undergoing surgery due to the high rates of association with intracanal anomalies. Indications for surgical or conservative treatment are still unclear.
ICMJE Conflict of Interest Statement
The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of the research reported.

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