

# Scheuermann's Kyphosis

## Scheuermann Kifoza

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### Öz

Hastalık ilk olarak Scheuermann tarafından, 1920 yılında, juvenilin dorsal kifoza olarak tanımlanmıştır. Scheuermann hastalığı adölesan çağda torakal hiperkifoza en sık nedenidir. İki alt tibi bulunmaktadır: Torakal ve torakolomber/lomber. Histolojik açıdan vertebral son plakalardaki düzensizlikler incelendiğinde, kollejen fibrillerde azalma veya kaybolma görülmektedir. Bunun sonucunda son plak devamlılığı bozulmakta ve disk materyalinin korpus içine protrüde olmasıyla Schmorl nodülleri ortaya çıkmaktadır. Bozulmalar vertebra korpusunda kamalaşmaya neden olmakta, bunun sonucunda dar açılanma gösteren kifotik deformite ortaya çıkmaktadır. Prevalans açısından bakıldığında, büyük serilerde %8'e varan oran görülmekte, hastalığın epidemiyolojisinde ise genetik geçiş ve mekanik stres faktörleri ön plana çıkmaktadır. Hastalığın semptomatik olması durumunda, ağrı deformiteye eşlik edebilir. Hastalığın klasik radyolojik tanımlaması, düzensiz vertebral son plak varlığı, 45 dereceyi geçen kifotik deformite ve en az 3 vertebrada bulunan ve 5 dereceyi geçen vertebra korpusunda olan kamalaşmayı içermektedir. Bazı olgularda kifoz ile minimal skolyoz birlikteliği görülebilmektedir. Tedavi ve takipte 50 derecenin altında kifozu olan olgularda periyodik radyolojik takip önerilmektedir. 75 dereceye kadar kifozu olan olgularda eğer iskelet matüritesi tamamlanmamış ise breys kullanımını uygundur. 75 dereceyi geçen olgularda ise; adölesanlarda breys uygulamasının başarısız olduğu durumlarda, erişkinlerde ise belirgin şekil bozukluğu olan ve konvansiyonel tedavi yöntemlerine rağmen ağrı şikayeti düzelmeyen olgularda cerrahi tedavi uygulanması düşünülmelidir.

**Anahtar Kelimeler:** Scheuermann kifoza, omurga, şekil bozukluğu, sagittal denge

### Abstract

The disease was first described by Scheuermann as the dorsal kyphosis of juveniles in 1920. Scheuermann's disease is the most common cause of thoracic hyperkyphosis in adolescence. There are two sub-types: thoracic and thoracolumbar / lumbar. Histologically, when the irregularities in the vertebral end plates are examined, collagen fibrils are seen to decrease or vanish. As a result, end-plate continuity deteriorates and Schmorl nodules appear when the disc material protrudes into the corpus. Deformations cause wedging in the vertebral corpus, resulting in narrow angled kyphotic deformity. In the large series, the prevalence is up to 8%, and the genetic transition and mechanical stress come into prominence in the epidemiology of the disease. Pain can be accompanied by deformity in symptomatic state. Classical radiological features include irregular vertebral endplates and kyphosis more than 45 degrees with 5 degrees of wedging in 3 or more vertebrae. In some cases, minimal scoliosis may accompany to kyphosis. Periodic radiologic follow-up is recommended for patients below 50 degrees of kyphosis. Bracing is appropriate in uncompleted skeletal maturation with kyphosis up to 75 degrees. In cases above 75 degrees, it is appropriate to perform surgical treatment when bracing is unsuccessful in adolescence and presence of exceeding pain despite the conventional methods and obvious deformity in adults.

**Keywords:** Scheuermann's kyphosis, spine, deformity, sagittal balance

### INTRODUCTION

Scheuermann's disease is the most prevalent cause of hyperkyphosis in the adolescent thoracic spine. It is the second common disease that causes spinal deformity after idiopathic scoliosis (1-3). In this disease, narrow angled kyphotic angulation of the spine, vertebral corpus wedging, vertebral end plate irregularity, Schmorl's nodule, intervertebral disc disorders and disc space narrowing are observed (3-7). Although the disease begins to develop before puberty, it becomes evident in the adolescent period in growth spurt (8).

The initial definitions focused on the thoracic kyphosis variant of the disease (9,10). It was later revealed that thoracolumbar or lumbar variants of the disease were

also present (1,3,11). The deformity is progressive and can become symptomatic in the period of rapid growth (4,8). In symptomatic state, deformity can be accompanied by pain (1,12). Minimal scoliosis may be combined with kyphotic deformity. However, this situation usually does not alter the treatment approach (5,13,14). Classical radiological features includes irregular vertebral endplates, kyphosis more than 45 degrees with 5 degrees of wedging in 3 or more vertebrae (3-5,7,14,15).

### History

The disease was first described by Scheuermann in 1920 (9). In this definition, the correlation between thoracic vertebral corpus changes and clinical juvenile kyphosis is shown on radiography. Scheuermann

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emphasized that the wedging on the anterior part of the vertebral corpus caused spinal deformity. The definition made by Sorenson in 1964 yielded that anterior wedging of 5 degrees or higher in three adjacent vertebrae to be included in the diagnostic criteria of Scheuermann's kyphosis (10).

### **Classification**

Scheuermann's disease predictably follows two patterns based on the apex of the deformity: Thoracic and thoracolumbar (1,3). Thoracic pattern is widespread and incorporated with lumbar and cervical hyperlordosis (16). This situation is mostly seen in adolescents. The thoracolumbar or lumbar pattern is less frequent (1,3,11). It is seen in adults who are involved in heavy sports activities and load lifting (11). In this disease, which occurs with localized pain and vertebral irregularities in the thoracolumbar junction, clinically significant kyphosis is not apparent as in the thoracic pattern (1,11).

### **Histology**

Electron microscopy reveals that the collagen fibers are weakened or vanished at the cartilaginous end plates of the vertebral corpus (17). This deterioration leads to the deformation of the vertebral bodies by loss of the growth zone (17,18). Significant irregularity occurs at vertebral end plates. The continuity of the end plates is disrupted by protrusion of the disc material into the corpus (19).

### **Epidemiology**

Current studies show the prevalence between 2.8% and 8% (20,21). This was demonstrated in a study on twins as 2.8% in 34,000 people (20). And 8% in another study conducted on radiological examinations with 10,000 individuals over 50 years old (21). There is a predisposition to males with Scheuermann's disease and this is seen in the study of 34,000 twins as 2:1 (20).

### **Etiology**

The studies on the last decades have focused on the relevance of the disease to the genetic transition. In a Danish-based study of 35,000 twins born over a 50-year period, 2.8% of the participants had Scheuermann's disease, and the heritability (familial genetic transition) was found to be 74% (22). In the same study, when monozygotic twins (31%) and dizygotic twins (19%) were evaluated in terms of concordance of the disease, it was found that concordance was significantly higher in monozygotic twins. In another genetic study, the autosomal dominant genetic inheritance of the disease and its association with three genes (Duffy,

COL1A1, COL1A2) were investigated (23). In that study, it was found that there was not any linkage with Scheuermann's disease. A mutation in COL2A1 was discovered to be interrelated with spondyloepiphyseal dysplasia similar to Scheuermann's kyphosis in serious osteoarthritis (24). The COL9A3 gene has been shown to be associated with radiographic Scheuermann's disease signs in patients with low back pain (25).

The other factor focused on is mechanical stress. Studies in this scope concentrate on the opinion that repetitive and non-daily intense loads cause deformation on the spine, resulting to Scheuermann's disease findings. This thesis is supported by Scheuermann, who first described the disease. The prevalence of the disease has been shown to increase in the workers who started to do heavy work at an early age (7). Studies in those participating in heavy sports activities also support this theory. In a study which 120 elite skiing athletes under the age of 17 were compared with their non-sporting peers demonstrated that end plate irregularities and Schmorl nodules were significantly higher in the athletes group (26). Also in a study conducted on Elite gymnasts, it was found that disc abnormalities, end plate irregularities and Schmorl nodes were more common than control group (27). Eventually, it has been shown that repetitive strain on the rats' tails for 2 hours per day for 2 months leads to histological changes similar to Scheuermann's disease (28). In another study conducted with twin sisters presents radiological findings of lumbar Scheuermann's disease were more prominent in who participated in heavy sports activities. Those patients were also complaining of pain (29). This study supports the theory that disease has the genetic basis and becomes evident by mechanical factors.

There are studies showing that increased body mass index is associated with Scheuermann kyphosis. Two studies with the control group found that patients with Scheuermann's disease had higher body mass index than the control group (30,31). In the cross-sectional case-control study of 10,000 students, the patients diagnosed with Scheuermann disease were found to be significantly higher in terms of height, weight, and body mass index than the control group. However, there was no correlation between weight, height, body mass index, and severity of deformity (31). In the study which 92 Scheuermann's kyphosis cases was matched with the control group, kyphosis group showed a significant increased body mass

index. In the same study, contrary to the other one, the body mass index and maximum kyphosis angle were correlated with each other (30). In this study, it was emphasized that increased body mass index made the diagnosis difficult and the deformity increased due to delayed diagnosis.

### Clinical Presentations

The most common complaint in Scheuermann's disease is deformity of the spine (13). There is a typical appearance in patients with thoracic Scheuermann's disease. The head is positioned ahead. Cervical and lumbar hyperlordosis develops to compensate for increased thoracic kyphosis. Similarly, the shoulders are positioned forward (Figure 1A). The kyphosis is fixed and becomes prominent by hyperflexion of the spine. When the patient is tilted forward, sharply angulated deformity is observed when viewed from the lateral side (Adams' Test-Figure 1B).

In the case of pain, this complaint is usually localized to the area of the deformity. Pain increases with physical activity and ameliorates with rest (1,8). In a 32-year follow-up study comparing the normal population with individuals with Scheuermann kyphosis, it was concluded that pain was more experienced in the patient group. However, there was no difference in the effect of pain on daily living activities. Also, there was a significant difference in

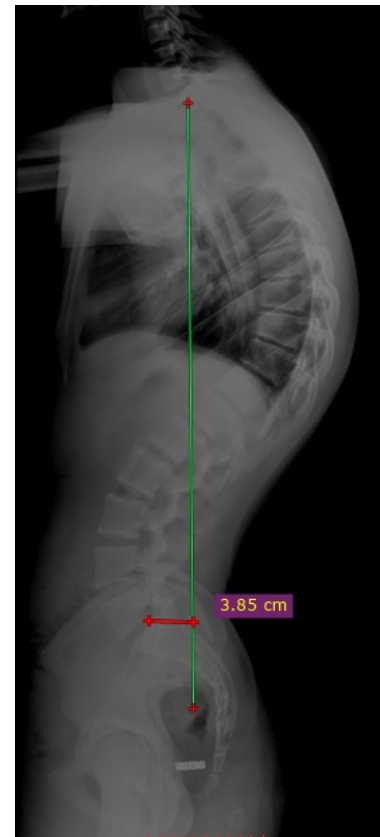


**Figure 1A.** The laterally sight of the patient showing the posterior sagittal balance, obvious kyphotic deformity, lumbar hyperlordosis, and the stooped shoulder position

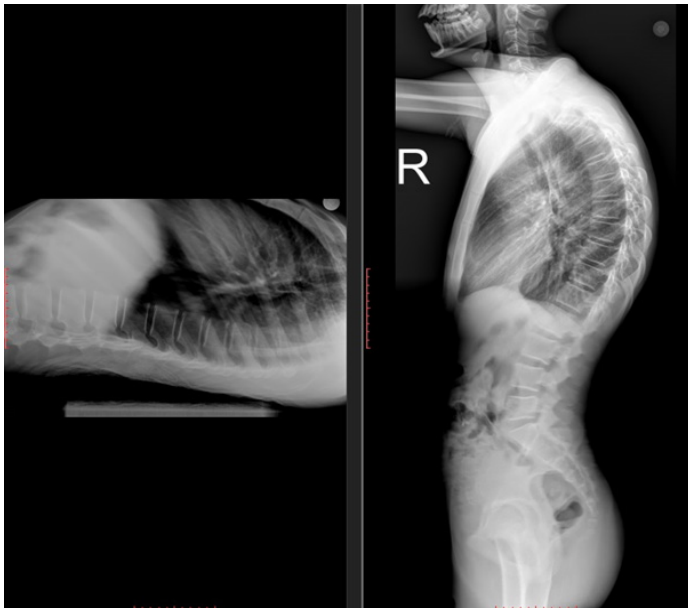


**Figure 1B.** Adams' forward bending test showing the steep thoracic kyphotic deformity of the patient.

localization of pain. In the patient group, pain was more common in the deformity region. In the same study, the patient population had less leg pain than the normal population (8). If the pain is localized to the lumbar region and the patients should be investigated



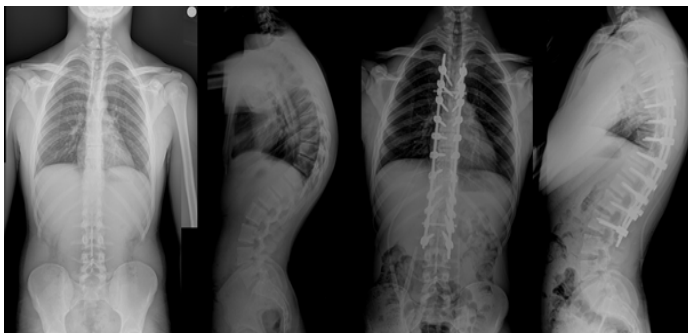
**Figure 2.** Lateral xray showing the posteriorly shifted sagittal balance of the patient.



**Figure 3.** A: Lateral fulcrum radiograph showing the correction of the sagittal deformity. B: Lateral xray of the same patient showing the round back deformity.

in terms of spondylolysis. Increased lumbar lordosis may increase the strain on the pars interarticularis and cause fatigue fractures (32).

Neurological complications are extremely rare in Scheuermann's disease. It has been described that this condition is caused by compression of cord in the apex of the kyphosis in severe cases and thoracic herniated disc or extradural spinal cysts in this region (33-35). In cases with severe kyphosis, a decrease in lung capacity may occur due to compression on the chest. It has been shown that restrictive lung disease develops in cases with kyphotic angulation of more



**Figure 4.** Preoperative and early postoperative anteroposterior and lateral X-rays of a patient with Scheuermann's kyphosis who underwent posterior correction and instrumentation of the deformity.



**Figure 5.** The lateral xray of a patient with Scheuermann's kyphosis who developed proximal junctional kyphosis after the posterior correction and instrumentation of the deformity.

than 100 degrees and where the kyphotic apex is between the first and eighth thoracic vertebrae. In the same study, in cases below 100 degrees, normal or above-normal lung capacities were observed (8).

#### **Radiological Findings**

The Spinal Deformity Study Group defines the thoracic kyphosis from the superior end plate of T2 to the inferior endplate of T12 utilizing the Cobb Method (36,37). The accepted normal range of kyphosis is 10 to 40 degrees (36). However, Fon et al. described that the upper limit was higher and justified that limit was 45 degrees on a study of 316 normal subjects (38). As described, Scheuermann Kyphosis is diagnosed by lateral radiograph of entire spine. The lateral spinal images should be taken in clavicular position (Standing position with both arms are holding the contralateral clavicle) (39). As previously described, radiological diagnosis is accomplished with presence of kyphosis over 45 degrees, wedging of three or more vertebrae by 5 degrees, existence of irregular

end plates with Schmorl nodules and narrowing of disc spaces.

Patients with Scheuermann's disease commonly have negative sagittal balance. Normally, a line drawn down from the corpus of C7 vertebrae, called C7 plumb line, passes 2 cm in front of the sacral promontory. In patients with Scheuermann's disease, this line passes behind the promontorium (40) (Figure 2). The pelvic incidence is lower in Scheuermann's disease than the normal population (41). This is a direct result of pelvic retroversion to compensate for the negative sagittal balance. In Scheuermann's disease, the possibility of degeneration of the discs rises. MRI studies revealed that the Schmorl nodules, narrowed discs and the thoracic disc bulging increased significantly (6,33). Furthermore, performing MRI can be discussed if surgery is required in Scheuermann's disease. A study conducted on 56 cases of Scheuermann's disease, the rate of spinal anomaly was found to be around 30%. In the same study, although the MRI examination minimally reduced the possibility of neurological changes, it changed the operation plan in 4.7% of cases (42). So it can be recommended to regard performing MRI in operative Scheuermann's disease cases. Computed tomography imaging may help to better demonstrate changes in bone structure and may lead to earlier diagnosis.

#### **Differential Diagnosis**

Scheuermann's disease is mostly confused with postural kyphosis. Postural kyphosis can be differentiated from Scheuermann's disease by dispersion of angulation to more vertebrae, lack of vertebral wedging, and loss of structural kyphosis on hyperextension or fulcrum lateral radiographs (4) (Figure 3). Adam's forward bending test is also effective in differential diagnosis. While narrow angulated kyphotic deformity is observed in Scheuermann kyphosis, a broader angled kyphosis appears in postural kyphosis.

#### **Treatment**

Determining the treatment method in Scheuermann's disease is based on the degree of the angular deformity, the age of the applicant and the associated symptoms (pain, neurological deficit). In adolescent cases with kyphosis less than 50 degrees, periodic radiological examinations should be performed for the evaluation of progression (43). In the absence of associated symptoms and progression of angular deformity, this follow-up is appropriate until bone maturity is completed. Treatment should be initiated in cases with kyphotic angulation of 50

degrees or more and progression is detected during follow-ups. There is no particular method, and many treatment methods are advocated for different conditions.

#### **-Rehabilitation**

Postural exercise treatments in Scheuermann's disease are aimed at strengthening paraspinal muscles, and mechanically correcting kyphosis and hyperlordosis. In addition hamstring stretching and balance exercises can be done. However, the effect of the exercise on the correction of kyphosis in Scheuermann disease has never been demonstrated, but the pain reduction can be achieved by rehabilitation. In a study, the results of treatment with physiotherapy, manual therapy and osteopathy combination in 312 patients showed a decrease of 16% to 32% in pain scoring (44).

#### **-Bracing**

Bracing is a considered treatment method in kyphosis less than 75 degrees and uncompleted skeletal maturity. In a study conducted on 274 patients, it was shown that bracing was unsuccessful in patients with kyphosis more than 75 degrees and surgical intervention was required (13). During bracing, there is a possibility that the treatment fails and the kyphotic deformity is likely to progress. The same study suggested that surgical intervention should be performed if bracing failed and progression of kyphosis was observed with angulation more than 60 degrees (13). It is thought that the use of brace will not be beneficial in adults when the skeletal maturation is complete.

There are many braces available for Scheuermann's disease. The most popular one of these is the Jewett corset. This brace forms the thoracolumbar spine in extension with three-point pressure. Also it can be modified according to the thoracolumbar curves. It has been shown to be effective in correcting the wedging in kyphotic deformity with full-time use (22 hours per day) for 12-18 months followed by part time use (12 hours per day) (13,45). However, it is very difficult to tolerate the use of brace for so long. It has been argued that the removal of brace during the sport activities for 2-4 hours a day increases the acceptability of brace without reducing its effectiveness (46).

#### **-Surgery**

In adolescents surgical treatment should be taken into consideration in cases of severe deformity (>75 degrees) and when it cannot be controlled by brace treatment (Figure 4). In adults, it should be preferred if there is a marked cosmetic deformity and the pain

cannot be controlled by conservative methods. In Scheuermann's disease the thoracolumbar spine is shortened in the anterior column. And the spinal cord has been adapted to this shortening. Correction of the kyphosis angle will also cause the anterior column to become elongated. This situation causes the risk of developing neurological damage due to prolongation of medulla spinalis. Therefore, cord tension should be avoided by providing reduction of posterior vertebral column with performing osteotomies in the apex of the deformity (5,14,15). The first publications about the surgical treatment of the disease include surgical approaches with Harrington and Luque instrumentation systems. Using the Harrington instrumentation system by Bradford et al., deformity correction was achieved at the beginning, especially in advanced kyphotic angulations. In this system, deformity was tried to be corrected by posterior compression. However, this improvement was worsened in follow-up (47). Luque instrumentation systems provided segmental fixation reducing the nonunion rates and iteration rates of the deformity. Sublaminar wiring was required in this method, thus increasing the risk of neurological deficit (48).

Recent studies have focused on the comparison of anterior/posterior combined approach and posterior only approach. In a study by Hosman et al (49), no significant difference was found in the correction of the deformity when the combined approach was compared to the posterior only approach. In a comparative study by Lee et al (50), posterior intervention was found to be more beneficial. In the combined approach group, the hybrid instrumentation system was used while the posterior approach group was stabilized by pedicle screws with the Smith-Peterson osteotomy. Correction rate of the deformity was higher in the posterior approach group but no complication was observed in this group. In addition, complications were seen including junctional deformities (Figure 5), temporary paraplegia, and wound infections in the combined group. In the study conducted by Lonner et al (51), there was no difference in terms of correction. Hybrid instrumentation was used in the combined group and all-screw stabilization was performed in the posterior group. While there was no complication in the posterior group, complications such as neurogenic bladder, renal failure, pneumothorax, and pulmonary embolism were observed in the combined group. In a study by Johnston et al (52), hybrid instrumentation was used in the combined group and closing wedge laminar resection was performed

in the posterior group. There was no difference in terms of correction of deformity. In a study by Geck et al (53), using only posterior approach with Ponte osteotomies was shown that the kyphotic deformity of 75 degrees was corrected to 38 degrees. While no neurological complication was observed in this study which conducted on 17 cases, one case each had junctional kyphosis and late infection.

## CONCLUSION

Scheuermann's disease is endemic in the community and may occur in different clinical presentations. In most of the cases, the deformity is the main complaint, but sometimes pain can accompany this complaint. The disease is diagnosed by lateral radiographs of the spine, and the Adam's test may be helpful in the diagnosis. Although the etiology of the disease is still controversial, it can be argued that the main reason is due to increased mechanical stress on the genetic basis. The major factors affecting the choice of treatment are the state of the skeletal maturation of the spine, severity of the deformity and its association with pain. Although conventional treatment methods such as periodic radiographic follow-up, rehabilitation and bracing are adequate in most cases, surgical treatment may be necessary in severe deformity. In the choice of surgical method, posterior only approaches with the osteotomies has become more popular due to their better correction results and lower complication rates.

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