JUVENILE KYPHOSIS (SCHEUERMANN DISEASE)

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ABSTRACT

Juvenile Kyphosis is the most frequent cause of structural kyphosis in the adolescence period. Many theories have been proposed for the etiology. These theories are avascular necrosis, herniation of disc material through the growth plate, repetitive strains, mechanical and static deforming forces, juvenile osteoporosis and congenital collagen aggregation disorders. Juvenile kyphosis shows very high familiar occurrence.

The characteristic feature is a wedge-shaped deformity of middle or lower thoracic vertebrae. The clinical onset is around puberty. The complaints are poor posture, fatigue or pain. Rarely, the symptoms of spinal cord compression may be seen. Radiographic findings include three adjacent vertebrae with wedging of each vertebra of 5 degrees or more. Hyperextension x-rays must be taken in all cases to determine rigidity.

Biomechanically, replacing the center of gravity which results from acute angular deformity, is led to increase deforming forces on the anterior elements. The treatment of Juvenile Kyphosis is based on the severity of the deformity, the presence of pain and the age of the patient. Before skeletal maturity, bracing is the most effective method in treatment. When the deformity can not be controlled by bracing, surgery is indicated. The success of operative treatment is based on the choice of the surgical approach.

Key Words: *Juvenile Kyphosis, Biomechanics, Treatment.*

Juvenile kyphosis or Scheuermann disease is the most frequent cause of structural kyphosis in adolescence period (3, 9, 10, 11, 19). It is a fixed deformity and develop around the puberty period. The characteristic feature is a wedge- shaped deformity of vertebrae that are demonstrated with conventional radiographies. This wedging was first described by Holger Scheuermann (15) in 1920, and he found three abnormal vertebrae in all of his cases. In 1936, he reported that the number of wedge-shaped vertebrae might vary from one to five. In 1964, Sorensen (19) suggested that the definition of Scheuermann disease should be a kyphosis including at least three adjacent vertebrae with wedging of each vertebra of 5 degrees or more. The exact prevalence of Juvenile Kyhosis is not known. According to Sorensen (19), it has been reported in 0.4 to 8 per cent of the general population. Ascani and associates (19) found an incidence of about 1 per cent. Sorensen suggested that there are no significant differences between two sexes, however Bradford (3) reported a higher incidence in the female and confirmed by Ascani.

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Etiopathogenesis

Many theories have been proposed for the etiology of the Scheuermann disease in the literature, but the cause is still unknown. Initially, in 1920, Scheuermann considered the disease caused by avascular necrosis of the ring apophysis that led to an arrest of growth, and resulting in wedging of the anterior portion of the vertebral bodies. He labeled the disease as "Osteochondritis deformans juvenilis dorsi" (15).

This theory was refuted by Copel and Bick (2) who demonstrated that the ring apophysis does not contribute to the longitudinal growth of the vertebral body. Any change that occur in the ring apophyses does not necessarily effect the growth potential of the vertebral body. Lambrinudi (19) proposed that the damage of the ring apophyses caused by inflammation or infection. This infection theory was confirmed by recent reports (6). However, this theory could not be acceptable because of the inflammatory changes could not be demonstrated with pathologic studies (1, 19).

In 1930, Schmorl implicated herniation of the intervertebral disc material through the growth plate is a causative factor. The bulged discs would herniate in to the spongiosa with congenital or traumatic tears in the end plates and longitudinal growth would inhibit. Schmorl claimed that these changes were not present in the types of kyphosis other than the juvenile one. His theory was challenged by the finding of such changes in vertebra outsides the confines of the deformity and even in persons who did not have Juvenile Kyphosis (19).

Mechanical and static forces have been considered to be of etiologic importance (8, 16). Heavy physical activity and recurrent injuries were accused but effects on pathogenesis did not reveal. Repetitive mechanical stress on a growing spine can damage vertebral end-plates both in the articular cartilage and in the growth-plate cartilage. Although, histologic changes of vertebrae that damaged by repetitive mechanical stress show similarity to Scheuermann disease (13). Taut hamstring and contracture of the iliopsoas muscle have been proposed as an etiologic factor. Some authors implicated the anterior vascular groove as a causative factor. They occur between vertebral bone-marrow spaces and the intervertebral discs. However, the blood vessels do not extend from the cartilage into the intact disc of juvenile spines (19). Pathologic studies showed that disc tissue within vascular channels or vascular scars are not characteristic histologic findings of patients with Juvenile Kyphosis (1).

Juvenile osteoporosis has been advanced as an etiologic factor. It is possible that the supportive cancellous bone could be weakened secondary to an episode of osteopenia occuring during the juvenile period. The extrusion of disc material into the spongios bone would than be a secondary phenomenon (3, 4, 12). Recently, measurement of vertebral bone density with quantitative computed tomography showed that density of vertebral bone is not significantly different between normal and Juvenile Kyphosis (7). Also, single-photon absorptiometric analysis showed that no evidence of osteoporosis was noted in the Juvenile Kyphosis compared with a normal individual (16). On the other hand, signs of Juvenile Kyphosis compared with a normal individual (16). On the other hand, signs of Juvenile Kyphosis are absent in porotic disorders.

A possible relationship between extradural cysts and Juvenile Kyhosis has been evaluated by some authors. Extradural cysts are very rare and compared to the incidence of Juvenile Kyphosis, they could not be an important factor in most cases of Juvenile Kyphosis (18).

Histologic studies of vertebral end-plates have been showed that the relationship between some translucent areas without collagen fibers and Juvenile Kyphosis. This translucent area presents in every specimen but they are much larger and more frequent in spines with Juvenile Kyphosis. Translucent areas might be related to a developmental error in collagen aggregation. Such a developmental defect would explain the familiar incidence of Juvenile Kyhosis (1).

Juvenile Kyphosis shows very high familiar occurrence. Some reports support autosomal dominant inheritance of this disease, but defective genes and it's role in pathogenesis has not been described yet (11).

The Biomechanical Features of Kyphotic Deformities

The spinal column may be divided into two parts anatomically, anterior elements and posterior elements. Posterior longitudinal ligament and everything anterior are called the anterior elements and all component's posterior to it are called the posterior element. At the anterior site, anterior and posterior longitudinal ligament, vertebral body and intervertebral discs; at the posterior site lamina and the yellow ligament are stabilize the vertebral column biomechanically. The muscles that apply loads aid the stabilization with these static forces. The interspinous ligaments are not so active there and the facet joint capsules particularly in the thoracic region are not well developed structures. For that reason these structures do not effect the stability significantly.

The thoracic spine is already in a kyphotic posture and the center of gravity is anterior to it. Therefore, the posterior elements are primarily resisting tensile forces and the anterior elements are mainly resisting compressive forces. When these components that resist tension and compression are disrupted kyphotic deformity occurs (23). Increased physiologic load, increased moment arm or weakening of posterior and/or anterior elements lead to kyphosis. A decrease in the anterior part of the vertebral body height results in angulation of the spinal column. Decreasing the anterior bone support caused by the vertebral body wedging is primary factor in Juvenile Kyphosis. Replacing the center of gravity anteriorly due to vertebral wedging

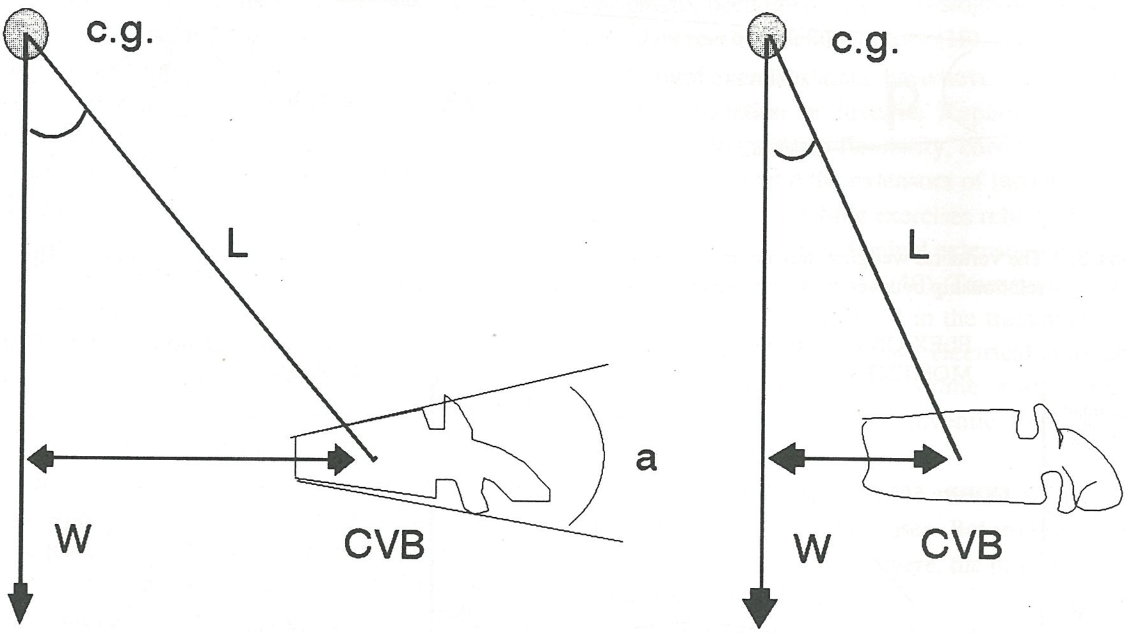

Figure 1. A model to analyze the effect of vertebral wedging on the bending moment in the thoracic spine. Intact vertebra is demonstrated on fig. A. Effect of wedge angle on the moment arm is demonstrated on fig. B, (23).

more than 5 degrees leads to increasing the moment arm and flexion loading (23). Figure 1.

Cobb angle measurement technique is used for evaluate the thoracic kyphosis and the normal range reported by the literature is 20 to 50 degrees (23). Although Cobb's method alone is inadequate to give an idea about kyhotic curve. Measurements of thoracic kyphosis with Cobb's method could be same in smooth or acute angle curves. In spite of this, flexion bending moment and moment arm is more increased in acute angle curves, for example vertebral wedging (Figure-2). Wedge angle and anterior body height ratio is helpful to evaluate the acute angle curves. These can be directly measured from an x-ray and are not affected by radiographic magnification (22, 23) (Figure-3).

The flexion bending moment of spinal column increases as a result of vertebral wedging and its proportional with wedge angle. The relationships between flexion bending moment and wedge angle are shown in figure 4-A. Increasing the wedge angle leads to augmentation the flexion bending moment. When comparing the less and more kyphotic curve, flexion bending

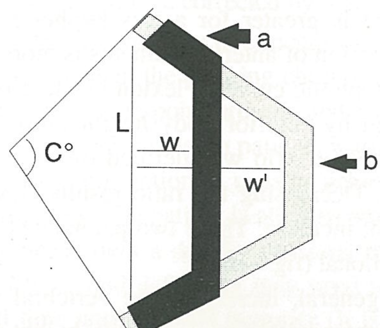

Figure 2. Although two curves a and b are identical in curve magnitude measured by Cobb's method, width of the curves and moment arms (W and W') are different. Thus, biomechanically clinical deformity is more severe on acute angle curve b. (21).

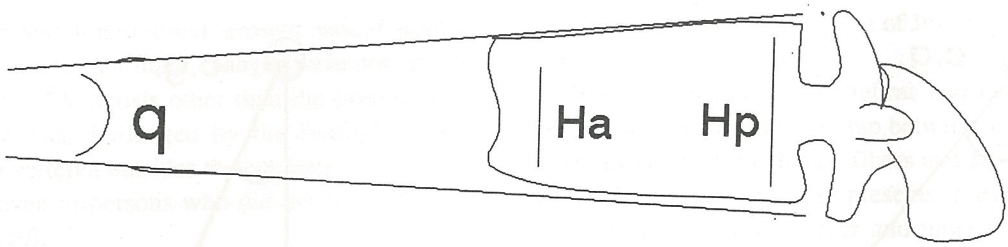

Figure 3. The vertebral wedging may be defined by the wedge angle (q) or by the anterior body height ratio (Ha/Hp). The relationship between two parameters is represented by this schematic drawing (23).

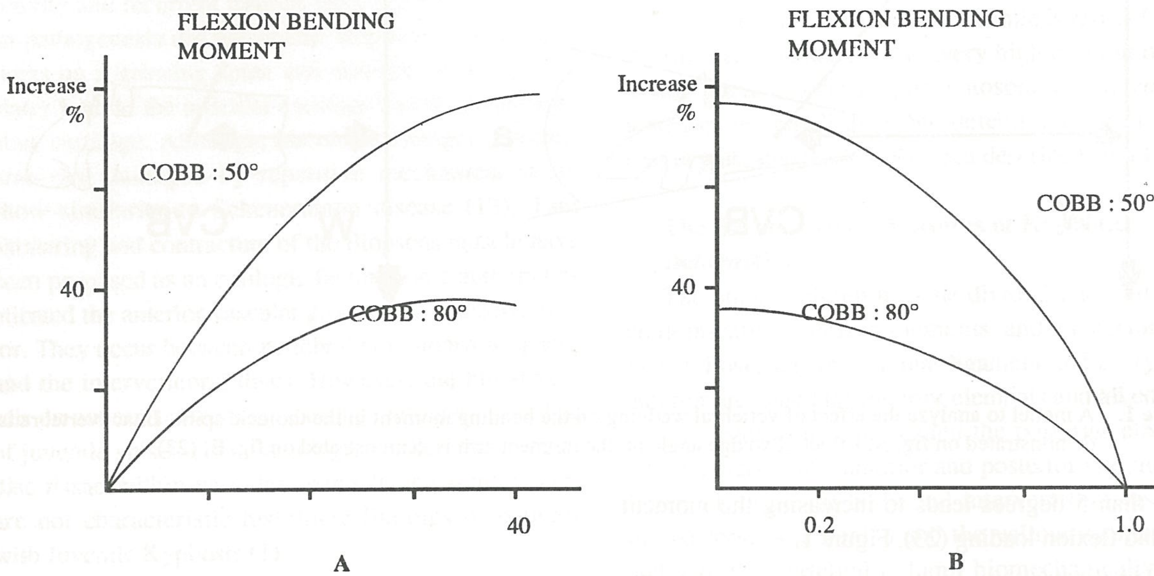

Figure 4. The relationship between flexion bending moment and wedge angle or anterior body height ratio demonstrated on these graphics. The upper curve : more straight, lower curve : more kyphotic spine (23).

moment is greater for a less kyphotic spine. Thus, compression of anterior elements is more excessive in large kyphotic curves. Flexion bending moments also affected by anterior body height ratio. The anterior body heihgt ratio was defined on the anterior body height. Decreasing the ratio results flexion bending moment increase. These two parameters are inversely proportional (fig 4-b).

In general, increasing the vertebral wedge angle and decreasing the anterior body height ratio leads to augmentation of flexion bending moment. These parameters both result in increased kyphotic angulation.

Clinical Features

The age of clinical onset is varying from 13 to 17 years around puberty. The patient is usually complaint of poor postre. Fatigue or pain in the region of the kyphosis may be present. The pain is aggravated after standing and relieved by lying down.

The physical findings depend on the location of the deformity. About three fourths of the patients the deformity is purely thoracic and Type I Scheuermann disease. Other one forth of the patient is Type II Scheuermann disease, which is located on thoracolumbar and lumbar region (5).

Kyphotic deformity in Scheuermann disease, typically, is acute angular. The cervical and lumbar lordosis increases to compensate thoracic hyperkyphosis. In pure thoracic kyphosis, increased lumbar lordosis results a protuberant abdomen. As a results of acute angular deformity, the center of gravity falls behind the sacrum. Normal pelvic tilt is exaggerated and shoulder protruding anteriorly.

Thoracolumbar kyphosis exposes long kyphotic curve in lateral view, and pure lumbar kyphosis shows straight back defortmity.

In the beginning, the postural defect may be corrected by hyperextension but in a very shot time deformity becomes fixed and could not be corrected both actively and passively.

Compression of the spinal cord in Scheuermann's kyphosis is rare. These neurological pictures consists of neurological dysfunction associated with severe angular kyphotic deformities and neurologic compression resulting from intervertebral disc displacement. Some authors also noted that an extradural spinal cysts may be seen and produces symptoms of impingement of the spinal cord. It is manifest by spastic paraparesis and hyperreflexia. Conservative alternatives in treatment are inadequate. Posterior surgical procedures could not decompress the medullary canal perfectly. Recent reports have suggested that anterior decompression is the most safe and effective procedure (18, 19).

Planning of Patient Management

Complete history and careful physical examinations are very important in planning the treatment. Next step in management is radiological examination. X-rays are obtained with the patient standing in the lateral plane with the arms outstretched parallel to the floor. A hyperextension x-ray is taken in the supine position. The kyphotic angles, vertebral wedging and the anterior vertebral body height ratio are measured and compared. Radiological imaging of pelvis are taken and Risser sign are detected.

The thoracic curves, that Cobb's angle more than 50 degrees assumed thoracic kyphosis. If there is less than 15 or 20 degress of mobility on hyperextension, it is accepted as a rigid deformity (4).

Treatment Considerations

The treatment of Juvenile kyphosis is based on the severity of the deformity, the presence of pain and the age of the patient. If there is no neurological complication, the basic principles will be to correct the deformity and prevent progression. Spinal balance will be achieved by reducing deforming forces, if indicated fusion techniques and implants are used. Adolescent who have a mildly increased kyphosis of less than 50 degrees without evidence of progression or neurologic involvement, need only periodic radiographic follow-up until they reach skeletal maturity (10).

Postural exercises alone have never been shown to provide correction in Juvenile Kyphosis. It is prescibed to help maintain flexibility, correct lumbar lordosis and strengthen the extensors of the spine with a brace treatment. Stretching exercises relieves tightness of hamstring muscles and spinal extensors thus, could be helpful for relieving pain (10). Transcutaneus muscle stimulation has been used in the treatment of kyphosis. In spite of this, surface electrical stimulation can not be recommended at this time as an alternative form of treatment for active Juvenile Kyphosis (10, 23).

Juvenile Kyphosis could be treated with bracing successfully in appropriate cases. Before skeletal maturity, if kyphosis is not so severe, the primary alternative in treatment is bracing.

Skeletal maturation is noted by closure of the iliac apophysis. It should be remembered that correction is still possible even though the iliac apophises may be closed as long as the ring apophises of the vertebral bodies are still open. Severe degrees of kyphosis (greater than 70 degrees), and severe vertebral wedging (greater than 10-15 degrees) may limit the correction of deformity with bracing (4, 10, 19). Sachs et al. treated 274 cases with Milwaukee brace and found that a kyphosis of more than 74 degrees was associated with a higher percentage of unsatisfactory results (14). Rigid deformity could not be corrected by bracing.

Milwaukee brace is an effective means of managing Juvenile Kyphosis in the growing child or adolescent. It is a dynamic three-point orthosis and promotes extension of the thoracic spine. In patients who have a flexible deformity, correction is usually achieved in six to twelve months. The patient is allowed out of the brace two to three hours a day for personal hygiene and bathing. More rigid deformity may need to wear the brace full time until skeletal maturity (It is determined by closure of the ring apophyses of the wedge vertebra), (4, 10, 19).

Some authors have reported excellent results with use of serial hyperextension body cast. Although it has not been preferred because of problems with the skin, restriction of physical activity, and need for frequent chances of the cast.

Operative Treatment

In Juvenile Kyphosis, when the deformity can not be controlled by bracing, the operative treatment should be considered. The patients who have a kyphotic curve more than 70 degrees or wedge angle more than 10 degrees at the apical vertebra needs to an operation at the first choice. In adults enough criteria for operation are kyphosis of 60 degrees or more, persistent pain and cosmetic appearance. Very rarely, operative treatment may be indicated in adolescent or adults because of paraparesis secondary to compression of the cord related tosevere angular deformity or to herniation of a thoracic disc (4, 14, 17).

Several reports concerning the surgical management of Juvenile Kyphosis have recommended combined anterior and posterior procedures. Some authors have stressed that posterior fusion alone for kyphosis greater than 70 degrees is usually inadequate. Significant complications and loss of correction with instrumentation failure are more frequent in posterior procedures alone. These problems were related to the fact that the posterior arthrodesis was done on the tension side of the spine, thus, the bending movements and distraction forces on the fusion mass led to pseudoarthrosis and bending of the fusion mass. They proposed that the two-stage procedure which consist of anterior and posterior approach is most useful for correcting kyphotic deformities secondary to Juvenile Kyphosis and the results are more satisfactory (4, 10). In spite of this, some authors suggest that results of posterior fusion is satisfactory. Exclude the exceptional cases, posterior fusion alone are acceptable, provided the fusion area is long enough (12, 20).

As a conclusion, the treatment of Juvenile Kphosis in patients with a curve of less than 60 degrees is certainly not surgical. In the skeletally immature patients which has spinal flexibility more than 20 degrees, correction and stabilization is achieved by posterior fusion alone. Loss of correction on the postoperative period results from the selection of incorrect fusion level. Therefore, the technique of posterior fusion and instrumentation is important. The fusion level must extend over the whole length of kyphosis.

Posterior compression should be achieved with effective instrumentation.

For skeletally mature patients with rigid curves combined anterior and posterior surgery offers the best results. The anterior fusion should be carried out over the apical five to seven vertebral levels or the most rigid portion of the kyphosis to be fused. Spinal column supported by anterior fusion and loss of correction minimize. A few weeks after anterior procedure, a posterior spinal fusion and instrumentation could be performed. Flexion bending moment and deforming forces is decreased by anterior fusion with bicortical solid bony graft after anterior distraction. Anterior release of contracted tissues and effective posterior compression after facet joint resection will provide excellent correction. Center of gravity will replace posteriorly to basic anatomic position. Compressive and tensile forces at anterior and posterior side would balance and this leads to prevent loss of correction.

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