

## Case Report

# A rare cause of kyphosis with low backache - “Scheuermann’s disease”: “Osteochondritis deformans juvenilis dorsi”

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## ABSTRACT

An uncommon case of Scheuermann’s disease (Apprentice’s spine) is being reported for two simple reasons- firstly, to show that it is a self-limiting disease which needs only proper observation, extension exercises & extension spinal brace and secondly, it becomes a diagnostic riddle when osteolytic lesion is seen in epiphyseal plates of adjacent vertebral bodies in an adolescent. Radiology and Imaging are needed for the early and accurate diagnosis and to differentiate it from other causes of kyphosis. Hence, it stressed the need to publish this condition not only for its rarity but also for its diagnostic puzzle to differentiate it from other conditions. Here we report a case of an adolescent male of 17 years with poor posture/slouching, fatigue, mild pain in lower thoracic area of spine (low backache), stiffness and loss of flexibility with radiological and MRI findings.

**Keywords:** Kyphosis, Adolescent kyphosis, Scheuermann’s Disease, Low backache, Osteochondritis deformans juvenilis dorsi, Skiagram, MRI-Magnetic resonance imaging

## INTRODUCTION

Kyphosis dorsalis juvenilis was described by Danish orthopaedician and radiologist Holger Werfel Scheuermann in 1921 when he noted the development of painful fixed kyphosis in 105 children.<sup>1</sup> Radiographs showed compression of the anterior vertebral borders, with a wedge deformity and irregular epiphyseal centers. Scheuermann thought it similar to the femoral head abnormality described by Calve and Perthes and named it osteochondritis deformans juvenilis dorsi.<sup>1</sup>

This entity has gone by many names, but it is most frequently called Scheuermann’s disease.

There are two types of idiopathic kyphosis; Thoracic (Type-I) & Thoraco-lumbar (Type-II). Type I is common however type II is uncommon.

Here our discussion is centered to type-II Scheuermann’s Disease. This was referred to by Sorenson as ‘Apprentice’s Spine’ & presents in adolescent age group after general skeletal maturity but not before the attainment of spinal maturity.<sup>2</sup>

Only a few cases with classical radiological and imaging findings are reported in the literature. In this case, we describe the radiographic and Magnetic Resonance Imaging (MRI) findings.

## CASE REPORT

Seventeen years old male presented to the orthopedics department of our institute with poor posture/slouching (Hunchback/Roundback), noticed by parents, initially. The patient complained of fatigue and mild pain in lower thoracic area of spine (low backache), stiffness and loss of flexibility. The pain was at the apex of curve,

aggravated by physical activity and by long periods of standing or sitting. The patient did not have any co-existent scoliosis, cardio-respiratory symptoms or any neurological symptoms such as sensations of pins and needles and numbness in bilateral lower limbs.

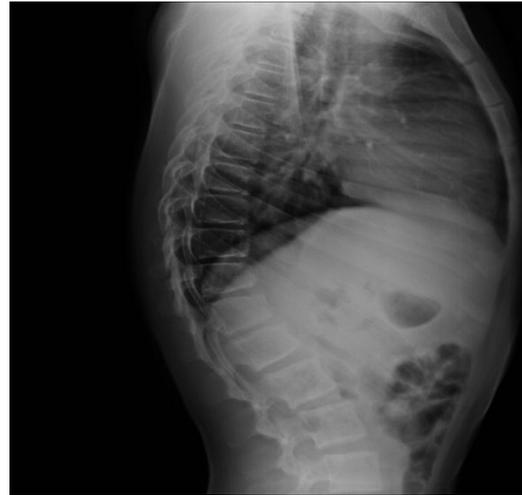
The patient underwent physical and clinical examination. The patient was a young male, moderately built and nourished & oriented to time, place and person. His general physical examination and vitals were normal. Musculoskeletal system examination revealed kyphosis, bulge and tenderness above and below the apex of kyphosis on palpation, decreased range of motion and reduced flexibility on Adam's forward bending test. Examination of other systems was unremarkable.

On radiological examination, the skiagram of dorso-lumbar spine (AP & lateral views) revealed kyphosis of lower thoracic and lumbar spines. The kyphotic curve was 34 degrees. There was evidence of anterior wedging of 9 degrees noted in 5 adjacent dorso-lumbar vertebral bodies from D9-L1. There was also an evidence of vertebral end-plate irregularity noted in D11, D12 & L1 vertebral bodies and mild narrowing of inter-vertebral disc spaces. Neural arches appeared normal. Bone density was normal and no evidence of any listhesis was noted.

MRI of dorso-lumbar spine (T1 & T2 - Sagittal axial and STIR sequences) revealed kyphotic deformity of dorso-lumbar spine. Mild anterior wedging of T8 to L1 vertebral bodies with mild irregularity of vertebral end plates is noted on both T1 and T2 weighted MRI images. T8-9 to T12-L1 disc heights are reduced with Schmorl's nodes present at articular margins of T8 to L1 vertebral bodies with evidence of early disc desiccation from D9-L1 discs. No evidence of any marrow edema, any pre or para-vertebral abscess or any listhesis noted in vertebral bodies. Thecal sac was normal. No evidence of any cord compression was noted.



**Figure 1a: Skiagram of dorso-lumbar spine (AP View) showing vertebral end-plate irregularity in D11, D12 & L1 and Schmorl's nodes at T8-L1 vertebral bodies with mild narrowing of inter-vertebral disc spaces.**



**Figure 1b: Skiagram of dorso-lumbar spine (Lateral view) showing kyphosis of lower thoracic and lumbar spines with evidence of anterior wedging of noted in five adjacent dorso-lumbar vertebral bodies from D9-L1.**



**Figure 2a: T1-weighted MRI sagittal images showing kyphotic deformity of dorso-lumbar spine and mild anterior wedging of T8 to L1 vertebral bodies with irregularity of vertebral end plates.**



**Figure 2b: T1-weighted MRI sagittal images showing kyphotic deformity of dorso-lumbar spine and mild anterior wedging of T8 to L1 vertebral bodies with irregularity of vertebral end plates.**



**Figure 3a:** T2-weighted MRI sagittal image showing reduced disc heights from T8-9 to T12-L1 with Schmorl's nodes at articular margins of T8 to L1 vertebral bodies with evidence of early disc desiccation from D9-L1 discs.



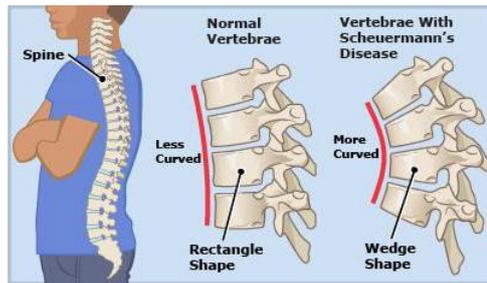
**Figure 3b:** T2-weighted MRI sagittal image showing reduced disc heights from T8-9 to T12-L1 with Schmorl's nodes at articular margins of T8 to L1 vertebral bodies with evidence of early disc desiccation from D9-L1 discs.



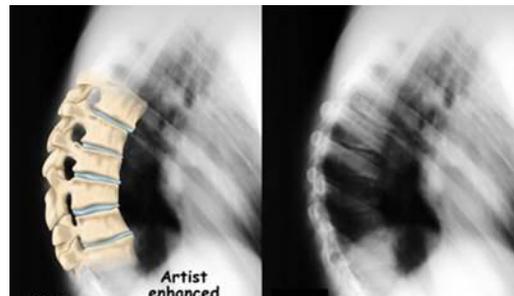
**Figure 3c:** T2 STIR-weighted MRI sagittal images.



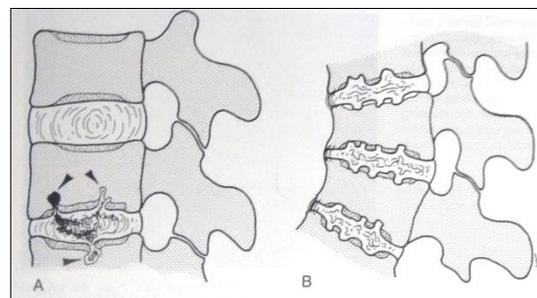
**Figure 3d:** T2 STIR-weighted MRI sagittal images.



**Figure 4a:** Hypothetical images showing wedging of vertebral bodies in Scheurmann's Disease (Image Courtesy: www.eorthopod.com).



**Figure 4b:** Hypothetical images showing wedging of vertebral bodies in Scheurmann's Disease (Image Courtesy: www.eorthopod.com).



**Figure 4c:** Hypothetical image showing intraosseous displacement of disc material (cartilaginous nodes/Schmorl's nodes) through cartilaginous endplates (arrows) in Scheurmann's Disease (Image Courtesy: Bone and Joint Imaging-3rd edition-By Resnick Kransdor).

## DISCUSSION

Scheuermann disease (also known as Scheuermann kyphosis and vertebral epiphysitis) is a common condition which results in kyphosis of the thoracic or thoracolumbar spine. The diagnosis is usually made on plain film. It is named after Danish orthopaedician and radiologist Holger Werfel Scheuermann, who first described it in 1921 as *Osteochondritis deformans juvenilis dorsii*.<sup>1</sup>

To apply the label of classical Scheuermann's disease, one needs to meet a number of radiographic criteria as described by Sorensen in 1964 (Sorensen classification).<sup>3</sup> He described Kyphosis of thoracic spine >40 degrees (normal is 25-40 degrees) or thoracolumbar spine >30 degrees (normal is 0 degrees) with at least 3 adjacent vertebral bodies demonstrating anterior wedging of > 5 degrees along with one associated sign in the lumbar region. The associated findings can include Schmorl's nodes, scoliosis (~25%), spondylolisthesis, irregularity and flattening of vertebral endplates, narrowing of intervertebral disc spaces, and antero-posterior elongation of apical vertebral bodies.<sup>3</sup> Schmorl's nodes occur when material for the nucleus pulposus extrudes through the endplate and into the vertebral body. These tend to occur centrally in classic Scheuermann's and anteriorly in the atypical lumbar form of the disorder. Radiographically, Schmorl's nodes appear as incongruous depressions of the endplate.<sup>4</sup> In Scheuermann's disease, multilevel endplate extrusions of disk material may be caused by delayed closure of endplate ossification centers. With the lumbar variant, only one or two vertebrae are involved, and they usually lie between T10 and L4.<sup>3</sup>

For most patients, Scheuermann's disease runs a fairly benign course, with resolution of symptoms at skeletal maturity. However, as described above, these individuals are more likely to have disk herniations in adulthood that may be neurologically devastating. At the apex of the kyphosis, adults often have pain along with varying degrees of degenerative spondylolysis and spondylolisthesis. The lumbar variant is more likely to be progressive in adulthood and more often symptomatic.<sup>5</sup>

Scheuermann's disease is the second most common cause of back pain in children after spondylolysis with spondylolisthesis.<sup>5</sup> It usually presents in adolescence with the insidious onset of thoracic kyphosis and back pain that is worse in the afternoons and relieved with rest. Symptoms are typically attributed to poor posture, and medical evaluation is often delayed.

This abnormality has prevalence in the general population of 4-8%, with no recognized gender predilection. There is a strong hereditary predisposition (perhaps autosomal dominant).<sup>5</sup>

Various theories have been proposed regarding the origin of this disease. Scheuermann proposed that the kyphosis

resulted from avascular necrosis of the ring apophysis of the vertebral body. Schmorl suggested that the vertebral wedging was caused by herniation of disc material into the vertebral body. Ferguson implicated the persistence of anterior vascular grooves in the vertebral bodies, which create a point of structural weakness in the vertebral body, which leads to wedging and kyphosis.<sup>6</sup> Bradford et al suggested that osteoporosis may be responsible for the development of Scheuermann disease.<sup>7</sup> Mechanical factors are common in patients who do heavy lifting or manual labour. Ippolito and Ponseti suggested that a biochemical abnormality of the collagen and matrix of the vertebral endplate cartilage.<sup>8,9</sup>

The most common endplate change observed clinically is the Schmorl's node. Exactly what causes Schmorl's nodes to form remains unknown. There seems little doubt that they begin as small defects and are therefore not always seen as often on clinical radiographs as they are at autopsy. They become more apparent radiologically as nuclear material prolapsing through the defect leads to reduced disc height and reactive new bone formation around the prolapse. Although most endplates do not show any evidence of natural perforations, Schmorl suggested that these lesions arise from focal weak spots caused by degenerated cartilage.<sup>10</sup> In a biochemical study Roberts et al. demonstrated that the composition of the cartilaginous endplate surrounding Schmorl's nodes was different from that of the normal endplate with a reduced content of proteoglycan, suggesting that there was degeneration of the cartilaginous endplate, before the formation of a Schmorl's node.<sup>11</sup> It has also been reported Schmorl's nodes is associated subchondral osteonecrosis.<sup>12</sup> Beneath the cartilage endplate, fibrosis is found within the marrow cavities with the disappearance of fat cells.<sup>12</sup> Schmorl's nodes may eventually lead to localized sclerosis of the subchondral bone plate and a fall in nutrient supply. Discs with Schmorl's nodes are more degenerate than discs without Schmorl's nodes<sup>13</sup>, and Schmorl's nodes predispose discs to degeneration at an earlier stage.<sup>14</sup> Schmorl's nodes are a common but not obligate manifestation of Scheuermann disease, which is known to enhance premature disk degeneration.<sup>15-18</sup>

There are two forms of Scheuermann's kyphosis - Type I and Type II. The classic Thoracic type (Type I) has an apex between T7 and T9 and is associated with increased lumbar lordosis. Type I (Typical) is again divided into 2 types - Type a (Thoracic) extends from T1-2 to T12-L1 and apex at T6-T8 and Type b (Thoracolumbar) extends from T4-5 to L2-3 and has apex at thoracolumbar junction. The thoracolumbar or lumbar type (Type II) has a lower apex, which frequently is associated with reduced upper thoracic kyphosis or thoracic lordosis. Type II Scheuermann's kyphosis occurs more frequently in males in a slightly older age group (15 to 18 years). This form tends to be more painful but rarely leads to progressive deformity.

Scheuermann's disease usually presents with the kyphotic deformity that is often rigid and does not disappear on lying supine or with hyperextension maneuver & pain that is usually at the apex of the deformity. Neurological examination is often normal, because kyphosis occurs gradually and over several segments. The onset is often after lifting heavy weight from a flexed position. Lumbar Scheuermann is less common and the deformity is often minimal. Rarely, thoracic disc herniation, epidural cysts, or a severe kyphosis (>100 degrees) can cause neurologic deficit in patients (usually adults) with Scheuermann's kyphosis.

Physical examination is remarkable for a fixed kyphotic deformity, most often in the thoracic region. Adam's test can be used to help differentiate from postural irregularities. When examined from the side, flexible postural abnormalities of the spine will generally smooth out when the patient is asked to bend forward. In Scheuermann's, the kyphosis will remain unchanged. Hyperlordosis of the cervical and lumbar spine is often found, along with forward head position. Up to a third of these individuals will have mild scoliosis.<sup>5</sup>

Imaging in Scheuermann's disease include AP radiographs of dorso-lumbar spine that reveal vertebral end plate irregularities, narrow disc spaces & Schmorl's nodes and Lateral radiographs of the same to measure vertebral wedging and measure Cobb's angle of deformity. A lateral radiograph should be examined for spondylolisthesis in addition to kyphosis and Schmorl's nodes. In the later stages of Scheuermann's kyphosis, radiographs may show changes of degenerative arthritis, including decreased intervertebral disc spaces, marginal osteophytes, and ankylosis. The plain skiagrams of spine are sufficient for the diagnosis. However, MRI of spine is done to rule out any posterior disc herniation.

Differential Diagnosis of the disease includes Postural Kyphosis. In forward bending test, the kyphotic deformity is accentuated, and the apex appears as a sharp angulation, in contrast to the smooth curve of a patient with postural kyphosis. A forward bending test also exposes any associated scoliosis. The hyperextension test helps the examiner understand the rigidity of the curve. A curve that is flexible or reduces significantly with hyperextension is typically postural and not Scheuermann's kyphosis, although in younger children a flexible round-back deformity may be the first sign of evolution to true Scheuermann's kyphosis. The cartilaginous nodes can accompany any disease process that weakened the end plates leading to Schmorl's nodes formation. Various diseases are included in this as - Osteoporosis, Hyperparathyroidism, Paget's disease and infections. However, the combination of kyphosis, cartilaginous nodes and irregular vertebral outline is pathognomonic of Scheuermann's disease.

Treatment options for the disease include conservative and surgical management. Conservative management

encompasses bracing as the primary modality that is effective in controlling the progression of the deformity. It is indicated in patients with kyphosis less than 1 year of onset, curves between 50-70 degrees and apex below T7. Bracing is continued for at least 18 months. Pain usually responds to NSAID's and physical therapy. While surgical management includes posterior correction with or without osteotomy and fusion.<sup>19,20</sup>

## CONCLUSION

Before labeling the case as lumbar Scheuermann's disease, it is important to exclude the possibilities of these above mentioned conditions. That means a diagnostic puzzle is required starting from good history to examination and investigations like complete hemogram, skiagrams, (Magnetic Resonance Imaging (MRI) etc. Once the diagnosis is confirmed, treatment is not a problem. The kyphosis, if present does not require surgery & in absence of spondylosis, symptoms respond well with bracing and extension exercise.<sup>5</sup> In our case there was no history of injury which excluded the possibility of fracture. Possibility of tubercular lesion was excluded by means of normal hemogram and no abnormal signal intensity on MRI. With all the radiological and imaging findings as kyphosis, anterior wedging of vertebral bodies and Schmorl's nodes, we labelled our case as Scheuermann's Disease Type-II.

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