



The Curvilinear Chine-Scheuermann's Disease



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Preface

Scheuermann's disease is a hyper-kyphosis of the spinal vertebrae and intervertebral discs characterized by anterior wedging of $\geq 5^\circ$ within three or more adjacent vertebral bodies. The thoracic spine is commonly implicated although thoracolumbar or lumbar region can manifest the disease. Scheuermann's disease is additionally denominated as Scheuermann's kyphosis, juvenile kyphosis or juvenile discogenic disease.

Disease Characteristics

Disease Characteristics of obscure aetiology, the disease is commonly discerned within adolescents of 12 years to 17 years and is exceptional in children below 10 years. Scheuermann's disease is the commonest cause of kyphotic deformity in adolescents [1,2]. Mode of disease transmission remains undetermined although a hereditary component contributes to genesis of the disease. Disease incidence is enhanced within monozygotic twins, in contrast to dizygotic twins [1,2].

Disease prevalence appears at around 1% to 8%. A male predominance is observed with a male to female proportion of 2:1 [1,2]. Scheuermann's disease is categorized as Type I or Classic subtype wherein the thoracic spine is incriminated with curve apex of kyphosis from thoracic vertebral body 7 to vertebral body 9 (T7 to T9). Type II wherein the thoracic and lumbar spine is implicated, and curve apex appears at thoracic vertebral body 10 to vertebral body 12 (T10 to T12). Type II is likely to progress and engender the disease in adulthood [1,2]. Sorenson et al categorized Scheuermann's disease as anterior wedging of three or more adjacent vertebrae of around $\geq 5^\circ$ with an absence of concurrent congenital, infectious, or traumatic disorders of the spine [1,2].

Disease Pathogenesis

Disease Pathogenesis of obscure pathogenesis, Scheuermann's disease demonstrates a genetic inheritance. It is posited that

mineralization and ossification of vertebral endplate may be discordant during skeletal growth with disproportionate expansion of vertebrae and consequent occurrence of classic wedge-shaped vertebral bodies engendering kyphosis [3,4]. Additional findings include aberrant collagen to proteoglycan ratio, cysts of the dura, osteoporosis occurring during childhood, hypersecretion of growth hormone and appearance of biomechanical stressors as tight hamstring muscles [3,4]. Alternatively, mechanical, metabolic and endocrinologic factors are posited to contribute to occurrence of Scheuermann's disease [3,4].

Clinical Elucidation

Scheuermann's disease demonstrates a structural deformity of vertebral bodies and spine wherein kyphosis of thoracic region appears at around 45° to 75° along with vertebral wedging $\geq 5^\circ$ of 3 or more adjacent vertebrae. Wedge shaped vertebral bodies engender the characteristic, rigid hyper-kyphosis which accentuates upon forward bending and is not relieved with extension or adopting a prone or supine position. However, aforesaid hyper-kyphosis can be counteracted by lumbar and cervical hyper-lordosis [3,4]. Cervical lordosis can be enhanced by protrusion of head wherein shoulders are positioned anteriorly. Mild to moderate scoliosis is an accompaniment. Tight hamstring muscles are exemplified along with muscle stiffness, fatigue, and decreased flexibility of the torso. Neurologic deficits are uncommon [3,4]. A cosmetic or postural deformity or a "hunchback" appearance is observed. Pain can arise within hyper-kyphotic vertebral region. Hyper-kyphosis of the thoracic spine, irregular vertebral endplates, occurrence of Schmorl's nodes and decimated height of intervertebral disc space, discernible on sagittal imaging, are commonly encountered [3,4].

Implicated adolescents manifest a cosmetic or postural deformity and subacute pain within the thoracic region which worsens upon activity and ameliorates with rest. Typically,

deformity of spine appears within early to mid-teens [3,4]. Subjects depict physical disability with inability to exercise, work and complete daily activities on account of pain or accompanying physical deformity. Cutaneous pigmentation upon the greatest curvature of hyper-kyphosis can appear due to friction with chairs or seating manifestos. Individuals with Scheuermann's disease are well muscled [3,4]. Subjects with Scheuermann's disease demonstrate an enhanced possibility of chronic back pain. Typically, pain within the affected region

ameliorates following skeletal maturity [3,4]. Severe instances are exceptionally associated with impaired cardiac and pulmonary function along with serious neurological symptoms [3,4].

Histological Elucidation

On microscopy, aberrant cartilage of the vertebral endplate, foci of irregular bony mineralization, altered endochondral ossification, reduced collagen to proteoglycan ratio with enhanced proteoglycan values are discerned [5] (Figures 1-6).

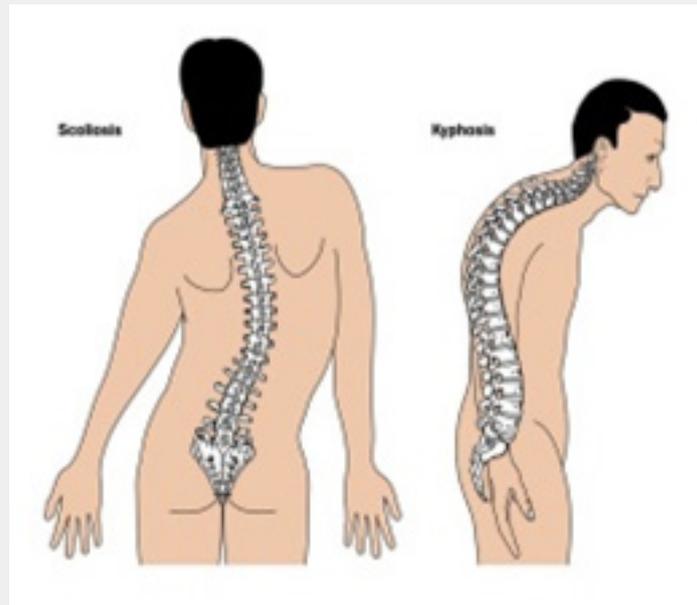


Figure 1: Scheuermann's disease representing a hyper-kyphotic spine with a hunch-backed appearance [9].

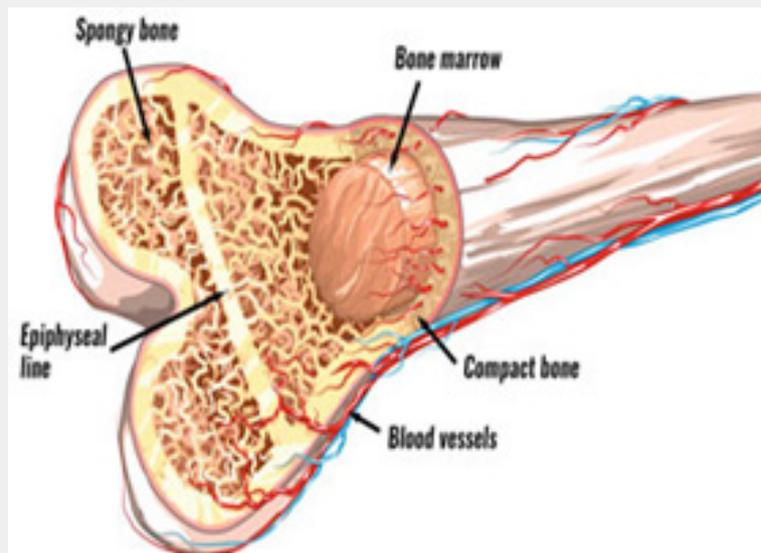


Figure 2: Scheuermann's disease depicting a prominence of spongy bone with abundant bone marrow [10].

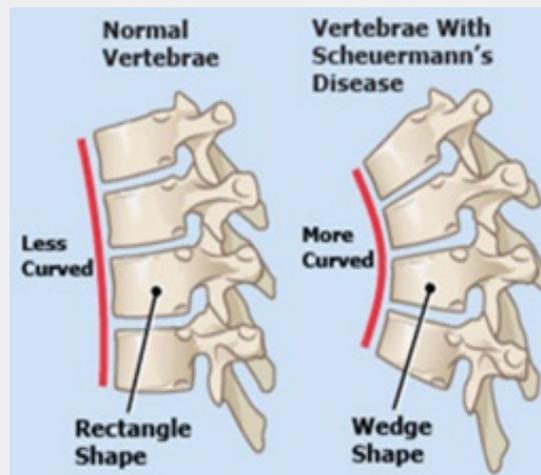


Figure 3: Scheuermann's disease delineating a hyper-kyphotic spine with absent height of intervertebral disc and anterior wedging exceeding 5° [11].

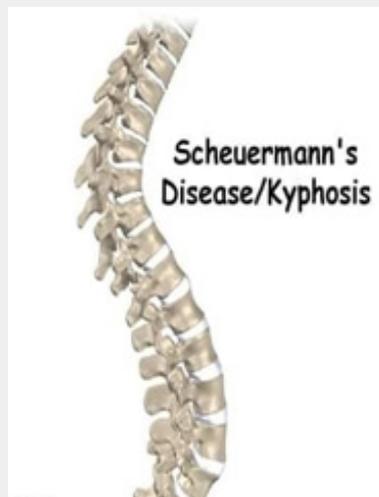


Figure 4: Scheuermann's disease demonstrating hyper-kyphotic spine with vertebral wedging [12].

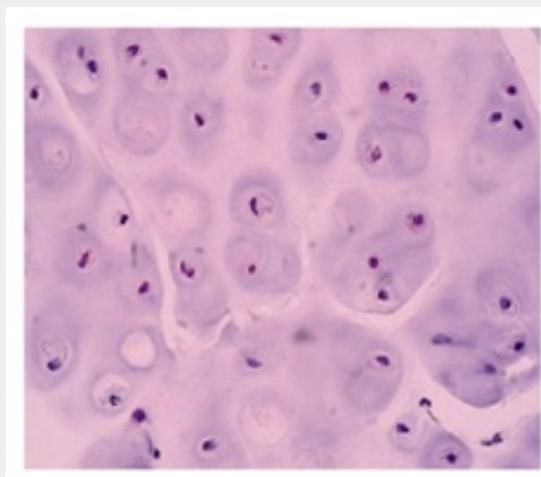


Figure 5: Scheuermann's disease exhibiting irregular bone mineralization with altered endochondral ossification and reduced collagen [13].

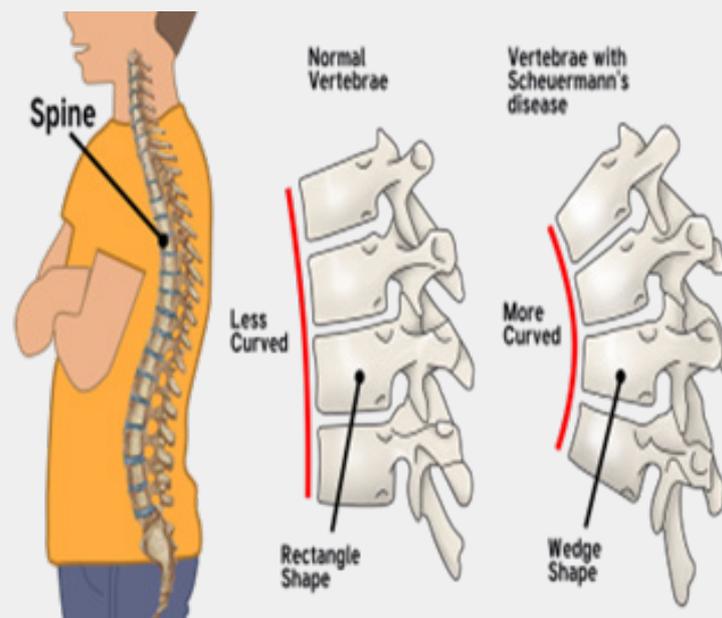


Figure 6: Scheuermann's disease exemplifying a hyper-kyphotic spine with excessive curvature and vertebral wedging [14].

Differential Diagnosis

Scheuermann's disease necessitates a segregation from conditions such as postural kyphosis with flexible postural deformity hyper-kyphosis contingent to secondary disease postsurgical kyphosis ankylosing spondylitis scoliosis [5,6].

Investigative Assay

Upon physical examination, a rigid curve of hyper-kyphosis is observed which accentuates upon forward bending. Hyper-kyphosis is unrelieved upon extension, prone or supine position thereby implying a "rigid" deformity. Additionally, cervical, or lumbar lordosis, scoliosis and contracted hamstrings are observed. Although neurologic symptoms are infrequent, a competent neurologic examination is mandated [6,7]. Physical examination of Scheuermann's disease is comprised of postural assessment with anterior, posterior, and lateral view along with neurological screening to exclude impending paraplegia with clonus and hyperreflexia. Associated scoliosis can be assessed with Adam's forward-bend test. Evaluation of muscle length, especially of pectorals, hamstrings, sub-occipitals, and flexors of hip joint along with possible contractures of anterior shoulder and hip is necessitated. Evaluation of muscle strength of abdominal, core, gluteal muscles and extensors of trunk is mandated [6,7].

Evaluation of spinal range of motion is necessitated with assessment of planes such as flexion/extension, right/left lateral bending and right/left spinal rotation. Percentage of hyper-kyphosis requires assessment upon serial imaging to evaluate degree of disease progression. Investigating the functional range

of motion along with degree of deformity aids in appropriate therapeutic intervention and assaying prognostic outcomes [7,8].

Plain radiographs such as antero-posterior or lateral radiographs are essential to evaluate Scheuermann's disease. Lateral radiographs can appropriately categorize diagnostic criteria such as rigid hyper-kyphosis exceeding 40° and anterior wedging $\geq 5^\circ$ in three or more adjacent vertebral bodies [7,8].

Additionally, antero-posterior, or lateral radiographs display irregular vertebral endplates, Schmorl's nodes, lack of height of intervertebral disc space, scoliosis, spondylolysis or spondylolisthesis and herniation of intervertebral disc [7,8]. Magnetic resonance imaging (MRI) can evaluate anatomic alterations or be employed for pre-operative planning. Computerized tomography (CT) is usually unnecessary as are non-diagnostic haematological or biochemical parameters or cogent histological features [7,8].

Therapeutic Options

Scheuermann's disease can be appropriately managed with non-operative techniques such as stretching, lifestyle modification, physical therapy, and administration of non-steroidal anti-inflammatory drugs (NSAIDs). Anti-inflammatory agents are employed for a short duration in young individuals and extended duration in adult subjects with continuous pain [6,8]. Non-invasive therapy is advantageous in asymptomatic individuals or subjects with kyphosis $<60^\circ$ [6,8]. Extension bracing is adopted for kyphosis between 60° to 80° and symptomatic individuals. It is typically required for a period of 12 months to 24 months and delays progression of kyphosis. Skeletally immature individuals

are benefitted, and bracing is efficaciously adopted prior to skeletal maturity at around 14 years. Ideally, a brace is applied at puberty for approximately 2 years and removed at skeletal maturity [7,8]. Diverse categories of braces such as Milwaukee brace, kyphologic brace and thoraco-lumbosacral orthosis-style Boston brace can be employed. Treatment with brace is advantageous in kyphosis between 55° and 80° , especially in skeletally immature subjects [7,8].

Adjuvant physiotherapy is recommended in combination with bracing. Physiotherapy is preferably adopted to assist with pain, appropriate spinal posturing, assessment and therapy of muscular imbalance which affects mechanical pull upon the spine [7,8]. Surgical methodologies such as spinal fusion are indicated when kyphosis exceeding $> 75^{\circ}$ engenders an unacceptable deformity, neurologic deficit, spinal cord compression and severe refractory pain. Symptomatic amelioration as well as melioration of curve deformity is delineated [7,8]. Conservative management is optimal for majority of incriminated subjects. Pain within implicated region ameliorates with emergence of skeletal maturity. However, an enhanced possible occurrence of chronic back pain is observed. Skeletally mature subjects with a kyphotic curve $< 60^{\circ}$ are devoid of long-term sequelae [7,8]. Scheuermann's disease is associated with complications such as progressive cosmetic deformity, chronic back pain, neurological deficits, and spinal cord compression. Additionally, postoperative complications such as pseudo-arthritis and persistent pain may ensue [7,8].

Extension-based stretching, stretching of hamstrings, muscle strengthening, adequate body posture and body mechanic techniques are advantageous for treating Scheuermann's disease. Appropriate employment and handling of braces is recommended. Bracing can be adopted for pain relief in adults where surgical treatment is not recommended [7,8]. Neurologic deficits require an exclusion while treating hyper-kyphosis unresponsive to

conservative management. Subjects may depict compromised pulmonary function tests on account of kyphosis [7,8].

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9. Image 1 Courtesy: Spine universe.com
10. Image 2 Courtesy: Virtual sports injury clinic.
11. Image 3 Courtesy: Scoliosis and spine associates.
12. Image 4 Courtesy: Concord orthopaedics.
13. Image 5 Courtesy: Hindawi.com
14. Image 6 Courtesy: Align Brace.com.



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