

Scheuermann's Kyphosis

Thomas G. Lowe, MD

Woodridge Spine Center, PC, 3550 Lutheran Parkway West, Suite 201, Wheat Ridge, CO 80033–6014, USA

In 1920, Scheuermann [1] described rigid kyphosis of the thoracic or thoracolumbar spine occurring in adolescents. The disease, characterized by vertebral body wedging and whose etiology remains unknown, continues to be the most common cause of kyphosis in adolescence today. Its reported incidence ranges from 1% to 8% of the general population, although its true incidence is probably understated because it is missed or attributed to poor posture [2,3]. It is generally thought that the prevalence of Scheuermann's disease is approximately equal in male and female adolescents [3–6]. At the present time, as in idiopathic scoliosis, no conclusive evidence has been found that defines the true etiology of Scheuermann's disease. It has been noted by several investigators that there seems to be an increased familial incidence of Scheuermann's disease. The mode of inheritance has not been determined at present but is most likely multifactorial.

The histopathologic findings in Scheuermann's disease have been the subject of investigation in recent years. Disorganized enchondral ossification similar to that in Blount's disease and a reduction in collagen and an increase in mucopolysaccharide in the vertebral end plates have been noted in patients with Scheuermann's disease [7,8]. Whether these changes are primary or are secondary to abnormal mechanical loading of the kyphotic spine is merely speculative. Other investigators have noted thickening of the anterior longitudinal ligament as well as partial reversal of the vertebral wedging with brace treatment, lending further credence to mechanical factors having a primary role in the pathogenesis [9–12]. It is certainly possible that several factors are operational in the pathogenesis of Scheuermann's disease, but

further investigation is needed before any definite conclusions can be reached.

Clinical findings

The onset of Scheuermann's disease usually appears just before puberty after ossification of the ring apophysis as a structural kyphotic deformity of the thoracic or thoracolumbar spine. Often, the deformity is attributed to poor posture, which may result in a delay in diagnosis as well as in treatment. Pain, when present, usually is mild, is brought on by prolonged periods of sitting or exercise, and is usually located near the apex of the deformity. Usually, the pain subsides with the cessation of growth.

In addition to the sharply angulated kyphosis of the thoracic spine, these patients demonstrate varying degrees of hyperlordosis of the lumbar spine and a forward protrusion of the head related to relative cervical kyphosis. The lumbar and cervical deformities are usually flexible. The kyphosis may be thoracic or thoracolumbar. The kyphosis is fixed and remains visible on hyperextension of the spine. When viewed from the side in the forward bending position (Adam's test), the deformity is sharply angulated, as noted in Fig. 1, when compared with a patient with postural kyphosis in which there is a uniform rounding of the entire spine, as seen in Fig. 2. In addition to the spinal deformity, patients with Scheuermann's disease often have tightness of the anterior shoulder girdle musculature as well as of the hamstring and iliopsoas muscles [13].

Natural history

There is little written about the epidemiology of Scheuermann's disease. It is generally accepted

E-mail address: woodridgespine@aol.com

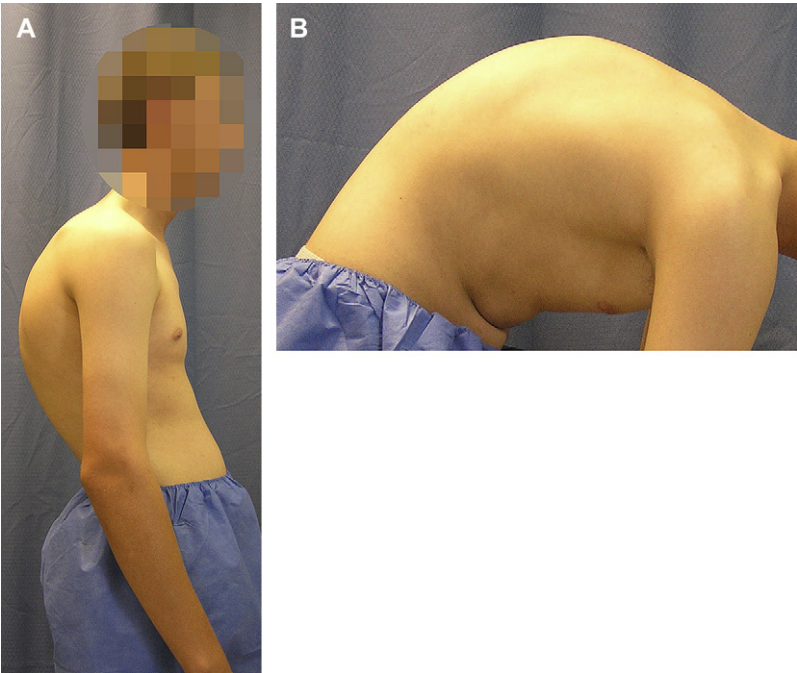


Fig. 1. (A) Patient with a kyphotic thoracic spine secondary to Scheuermann's disease. (B) Note the sharp angular deformity, especially in the forward-bending position.

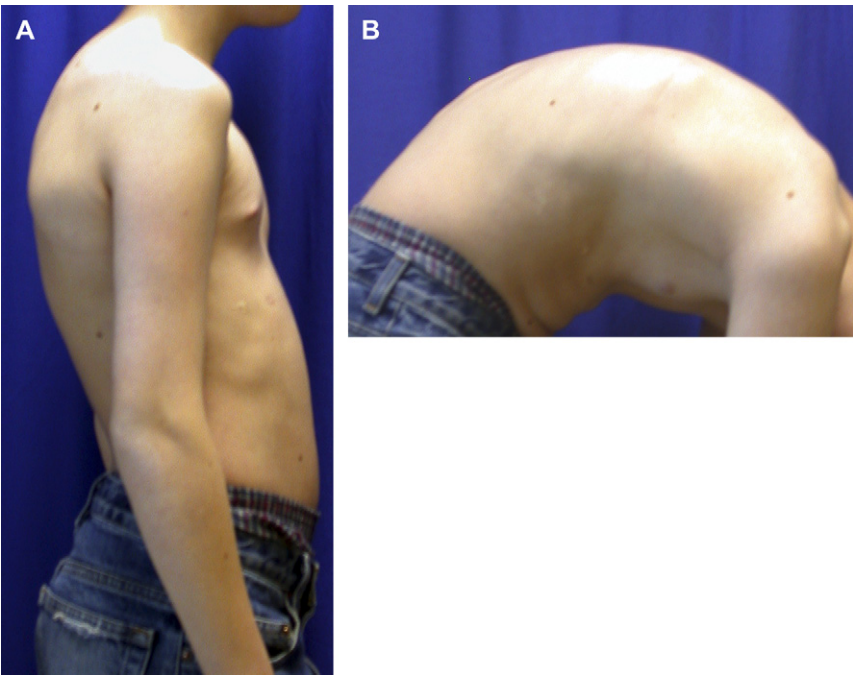


Fig. 2. (A) Patient with postural kyphosis. (B) Note rounded deformity, which disappears in the forward-bending position.

that Scheuermann's disease most often has a benign course, resulting in little deformity and few symptoms. Back pain and fatigue are common complaints during growth but usually disappear with skeletal maturity. If the resultant deformity remains less than 75° , there should be no long-term difficulties other than varying degrees of back pain, which are usually nondisabling. Other patients with more significant kyphosis ($>75^\circ$) who have untreated Scheuermann's disease may seek medical attention as adults because of progressive deformity or chronic back pain [3,5,9]. Neurologic complications secondary to severe kyphosis, dural cysts, or thoracic disc herniation have been described in a small number of patients with Scheuermann's disease [14–16].

A study by Murray and colleagues [5] detailing a long-term follow-up of 67 patients with Scheuermann's disease for an average of 32 years noted that they worked in lighter jobs than controls, had more severe back pain, and were more concerned about their appearance but did not seem to be limited by their symptoms, although there were a few patients with severe deformities ($>80^\circ$) in the study. Untreated high-magnitude kyphosis in the adult may progress, although the true incidence of progression is unknown. The pain that develops in adults is frequently attributable to degenerative spondylosis, which is often a sequela of untreated Scheuermann's disease and may be resistant to nonoperative treatment. In general, the thoracolumbar pattern of Scheuermann's disease is more likely to result in painful progressive kyphosis in the adult than is the thoracic pattern [4].

Radiographic findings

Routine radiographic studies should include erect 36-in posteroanterior (PA) and lateral radiographs as well as a supine hyperextension lateral radiograph over a bolster, which demonstrates the structural degree of the kyphosis. The normal range of thoracic kyphosis is from 20° to 45° , as measured by the Cobb method on a standing lateral radiograph with the arms positioned 60° below the horizontal [17,18]. Normal kyphosis increases with age and is slightly greater in women than in men [18]. There is considerable variation in lumbar lordosis, and ranges between 40° and 65° are considered normal [17]. The transitional zone between the thoracic and lumbar spine includes T10 to L2 and is normally slightly lordotic

(0° – 10°). When the spine is balanced in the sagittal plane, the C7 vertebral body vertical axis should lie horizontally within 2 cm of the sacral promontory. Patients with Scheuermann's disease tend to be "negatively" balanced when compared with the general population [19].

The radiographic criteria for Scheuermann's disease as described by Sorenson [20] include greater than 5° of anterior wedging of at least three adjacent vertebral bodies. Associated radiographic findings include Schmorl's nodes, irregularity and flattening of the vertebral end plates, narrowing of the intervertebral disc spaces, and anteroposterior elongation of the apical vertebral bodies, as shown in Fig. 3.

Varying degrees of scoliosis are seen in approximately one third of the patients. Spondylolysis or spondylolisthesis is known to occur more frequently in patients with Scheuermann's disease and may be a source of low back pain. In adults, varying degrees of degenerative spondylosis are frequently seen primarily in the region of the apex of the kyphosis and are the major source of back pain [21,22].

Two different curve patterns have been noted in Scheuermann's disease [6]. The thoracic pattern noted in Fig. 4A is the most common, but the thoracolumbar pattern seen in Fig. 4B is the most likely to progress after skeletal maturity and the most likely to become symptomatic in adult life. Postural kyphosis is readily differentiated from Scheuermann's disease radiographically because of the presence of a less acutely angulated deformity that is nonstructural and the absence of wedging of vertebral bodies. Fig. 5 comprises radiographs of a patient with postural kyphosis.

Nonoperative treatment

The need for treatment of Scheuermann's disease is based on the severity of the deformity, location of the deformity, presence of pain, and age of the patient. The treatment of Scheuermann's disease is largely nonoperative.

Adolescents in whom the kyphosis remains mild need only an exercise program to increase flexibility and periodic radiographs until skeletal maturity. Adolescent patients with Scheuermann's disease whose kyphosis is greater than 55° in the thoracic spine or 40° in the thoracolumbar spine should be placed in a combined brace and exercise program. Initial brace treatment should be full time (>20 hours per day) until full passive

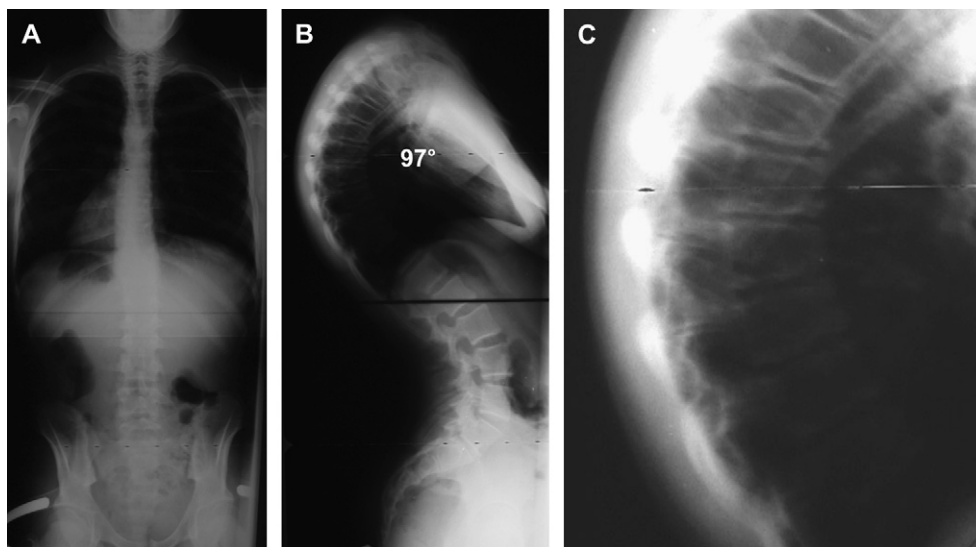


Fig. 3. (A) Anteroposterior, (B) lateral, and (C) focused radiographs of a patient with Scheuermann's disease. (C) Note the sharp angular kyphosis and wedging of the apical vertebrae and narrowing of intervertebral discs.

correction of the kyphosis in the brace has occurred, including a partial reversal of vertebral body wedging. At that point, brace treatment can usually be reduced to 12 to 14 hours per day. Part-time brace treatment needs to be continued for 1 year after the fusion of the iliac apophysis. Most adolescents with Scheuermann's disease can be managed with a thoracolumbosacral orthosis

(TLSO) type brace with anterior infraclavicular outriggers, as shown in Fig. 6. This brace is usually well tolerated and is cosmetically acceptable to most adolescents because it can be hidden under clothing. Stretching and strengthening exercises should be prescribed for trunk as well as tight hamstring and pectoral musculature. Although the initial improvement in kyphosis may be significant, there is often a gradual loss of correction within the first few years of discontinuance of the brace, with only a modest overall long-term correction of the prebrace deformity [11,12]. Fig. 7 depicts serial radiographs of a patient with Scheuermann's disease undergoing brace treatment.

As long as the deformity is less than 75° to 80°, most adults with untreated Scheuermann's disease should respond to a combination of physical therapy and an aerobic and torso strengthening exercise program [4,19].

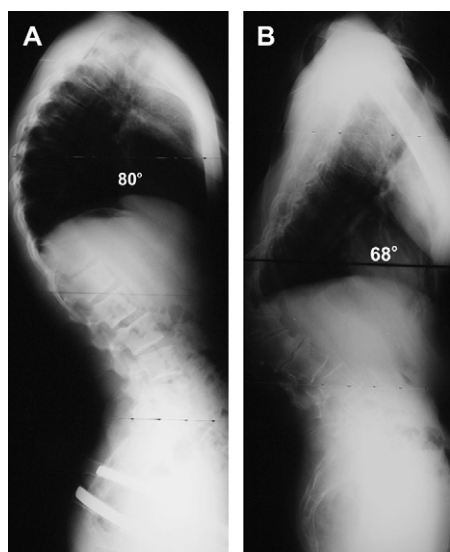


Fig. 4. (A, B) Radiographs of the two patterns of kyphosis secondary to Scheuermann's disease.

Operative treatment

Indications for surgical treatment in the adolescent include a symptomatic kyphotic deformity of 80° in the thoracic spine or 65° in the thoracolumbar spine not controlled by nonoperative measures or patients with significant sagittal imbalance secondary to the kyphotic deformity [4,19,23].

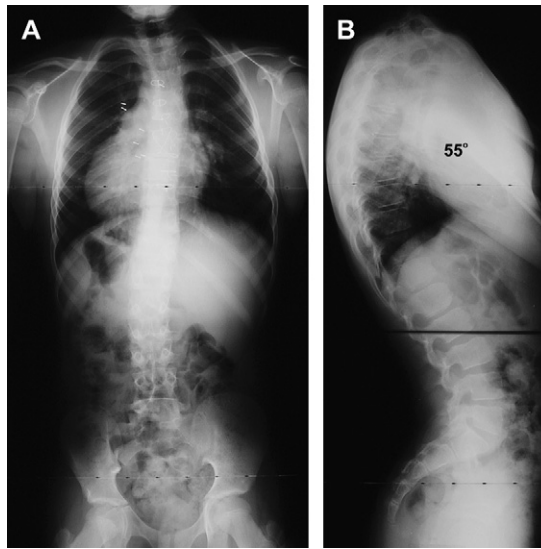


Fig. 5. (A) Anteroposterior and (B) lateral radiographs of a patient with postural kyphosis demonstrate a uniform rounding of the thoracic spine without vertebral or disc abnormalities.

Surgical technique

Posterior instrumentation and fusion

Neuromonitoring (myogenic motor evoked potentials [MMEP] and somatosensory evoked

potential [SSEP]) should be used throughout the surgical procedure. Posterior instrumentation and fusion of the entire kyphotic deformity constitute the basic method of surgical treatment for Scheuermann's disease in adolescents and adults. A thoracic MRI scan should be obtained before surgery because of the thoracic disc herniation that is occasionally present in Scheuermann's disease and may result in cord compression, as shown in Fig. 8. Stainless-steel instrumentation should normally be used because it holds its profile better than titanium and is less likely to fail because it is not "notch sensitive" with in situ bending. The proximal end of the construct should include the upper sagittal Cobb level, and the distal end of the construct should include, at a minimum, the distal sagittal Cobb level and the first lordotic level, which is normally one or two levels beyond the distal Cobb level. Recommended instrumentation levels are shown in Fig. 9. Instrumentation short of these recommended levels frequently results in proximal or distal junctional kyphosis. The two most commonly used constructs consist of pedicle screws throughout the construct or pedicle screws distally in the lumbar spine and hooks in the thoracic spine, as shown in Figs. 10 and 11. When an all-pedicle screw construct is used, the proximal and distal three levels should all be instrumented. If the kyphosis is flexible on a hyperextension lateral radiograph ($\leq 60^\circ$), pedicle screws can be staggered



Fig. 6. (A, B) Thoracolumbosacral orthosis brace with anterior infraclavicular outriggers for the treatment of kyphosis secondary to Scheuermann's disease.

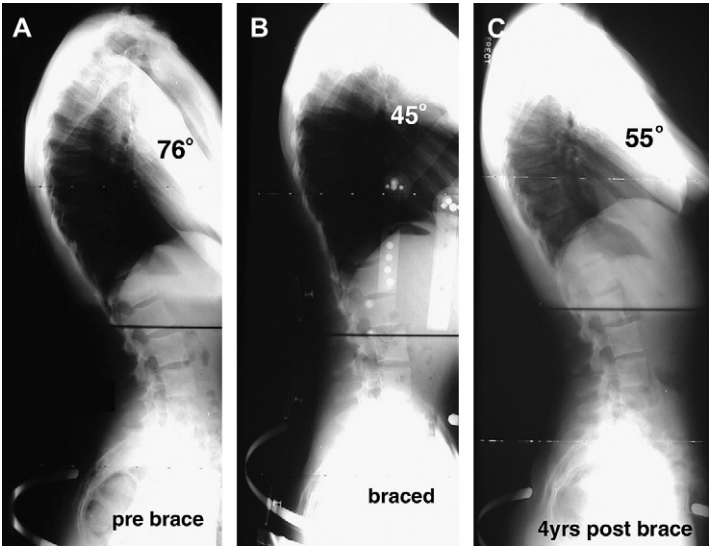


Fig. 7. (A) Patient undergoing brace treatment for Scheuermann's disease. Note the improvement (B) with the brace and the slight loss of correction at (C) final follow-up.

on each side in between. If the deformity is rigid ($>60^\circ$) on the hyperextension lateral radiograph, pedicle screws should be considered at all levels. When a hybrid construct is used (hooks

proximally and screws distally), two double-level pedicle hook–transverse process claw configurations on each side should be used for maximum grip strength. Centrally, a pedicle hook just below



Fig. 8. T2 sagittal MRI scan of a patient with Scheuermann's disease with thoracic disc herniation.

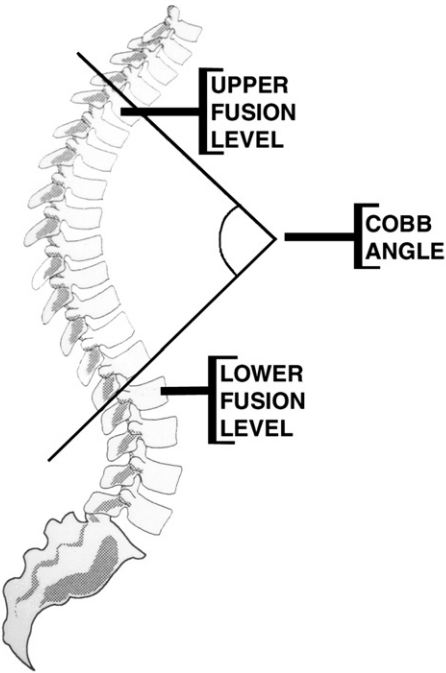


Fig. 9. Diagram of recommended posterior instrumentation levels for kyphosis secondary to Scheuermann's disease.

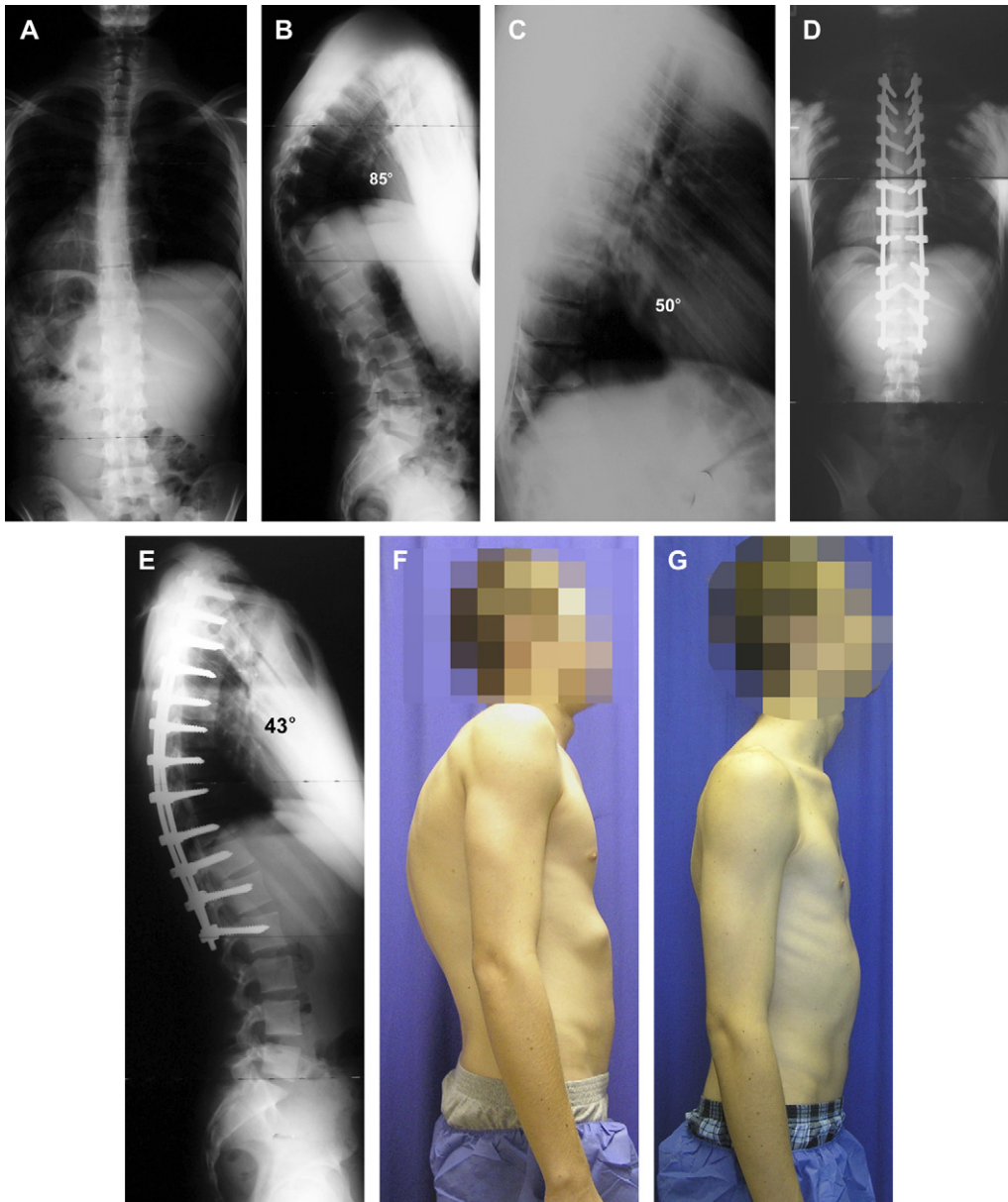


Fig. 10. (A) Anteroposterior, (B) lateral, and (C) hyperextension radiographs demonstrate the pedicle screw construct (D, E) for Scheuermann's disease as well as (F) pre- and (G) postoperative clinical photographs.

the apical level should be placed on both sides for additional fixation. After insertion of the anchors, partial facetectomies should be performed at all instrumented levels. For rigid kyphotic deformities, three or four periapical complete facetectomies, as described by Ponte and Siccardi [24], provide additional flexibility. Correction is first achieved by underbending kyphosis into the rods based on the hyperextension lateral radiograph.

With the pedicle screw construct, correction beyond what is obtained on the hyperextension lateral radiograph can always be achieved. Correction is achieved by cantilever and compression maneuvers. The rods are first inserted into the proximal screws and then gradually cantilevered sequentially into the distal screws. After insertion of both rods, compression is first applied at the apex and then extended to the caudal and

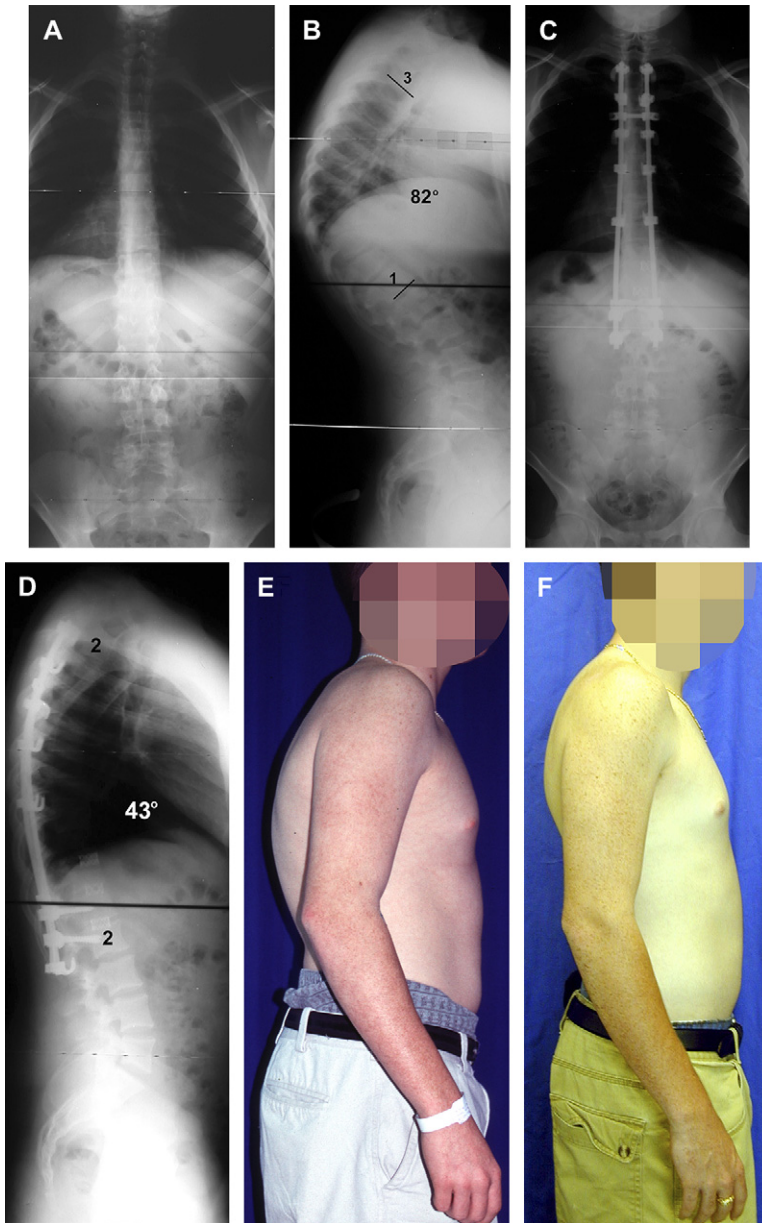


Fig. 11. (A) Anteroposterior, (B) lateral, and (C) hyperextension radiographs demonstrate the hybrid construct (C, D) for Scheuermann's disease as well as (E) pre- and (F) postoperative clinical photographs.

rostral ends of the construct sequentially. Care should be taken to avoid correction beyond 50% of the preoperative magnitude of the kyphosis to avoid junctional kyphosis proximally or distally. Transverse connectors are then inserted proximally and distally. The fusion is completed by decortication and bone grafting. For severe fixed kyphosis ($>100^\circ$), periapical pedicle subtraction

osteotomies can be considered, thus avoiding the need for anterior surgery.

Postoperative management

The patients are out of bed and walking on the second postoperative day and are generally discharged on day 6 or 7. No postoperative

orthosis is needed. Patients are on a walking program for the first 6 weeks and are then phased into a light aerobic exercise program that includes swimming, stretching, and strengthening as well as the use of an exercise bike or treadmill, and they are generally back to regular activities in 6 months.

Complications

The risk of neurologic injury after surgery for Scheuermann's disease is probably less than 2%. A preoperative MRI scan should be considered because of the possibility of thoracic disc herniation. The use of neurologic monitoring is essential during the entire surgical procedure, however. Evoked potential monitoring, motor and sensory, should be used on all patients. An increase in latency in motor or sensory evoked potentials of more than 10% or an amplitude drop of greater than 50% is an indication for an immediate wake-up test during surgery. If the patient has a motor deficit after surgery, instrumentation should immediately be removed and an emergency MRI or CT-myelogram should be obtained to rule out the presence of a compressive lesion as a possible cause of cord injury.

The incidence of postoperative wound infections in adolescents undergoing major surgery with instrumentation is approximately 3%.

Prophylactic antibiotics should always be given for 24 hours after surgery. Traffic in operative suites should be kept to an absolute minimum during surgery, and a normal body temperature should be maintained.

Probably the major instrumentation-related complication after the surgical correction of kyphotic deformities is the development of junctional kyphosis occurring above or below the primary kyphotic deformity [6]. In patients undergoing posterior instrumentation and fusion, failure to incorporate all levels of the kyphosis as well as the first lordotic segment distally may result in proximal or distal junctional kyphosis, as shown in Figs. 12 and 13. Junctional kyphosis may also occur if correction of the kyphosis exceeds 50% of the preoperative kyphosis magnitude, which may interfere with global sagittal balance, resulting in compensatory junctional kyphosis.

Summary

Scheuermann's disease is the most common cause of structural kyphosis in adolescents. The kyphotic deformity is frequently attributed to "poor posture," resulting in delayed diagnosis and treatment. Indications for treatment remain somewhat debated, because the true natural history of the disease has not been clearly defined.

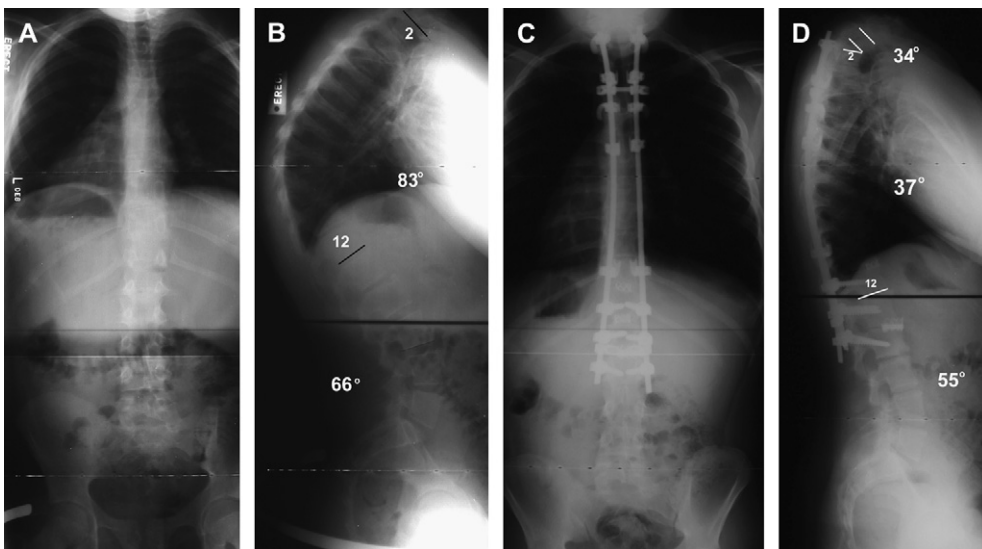


Fig. 12. (A–D) Pre- and postoperative radiographs demonstrate proximal junctional kyphosis related to excessive correction of kyphosis (>50%).

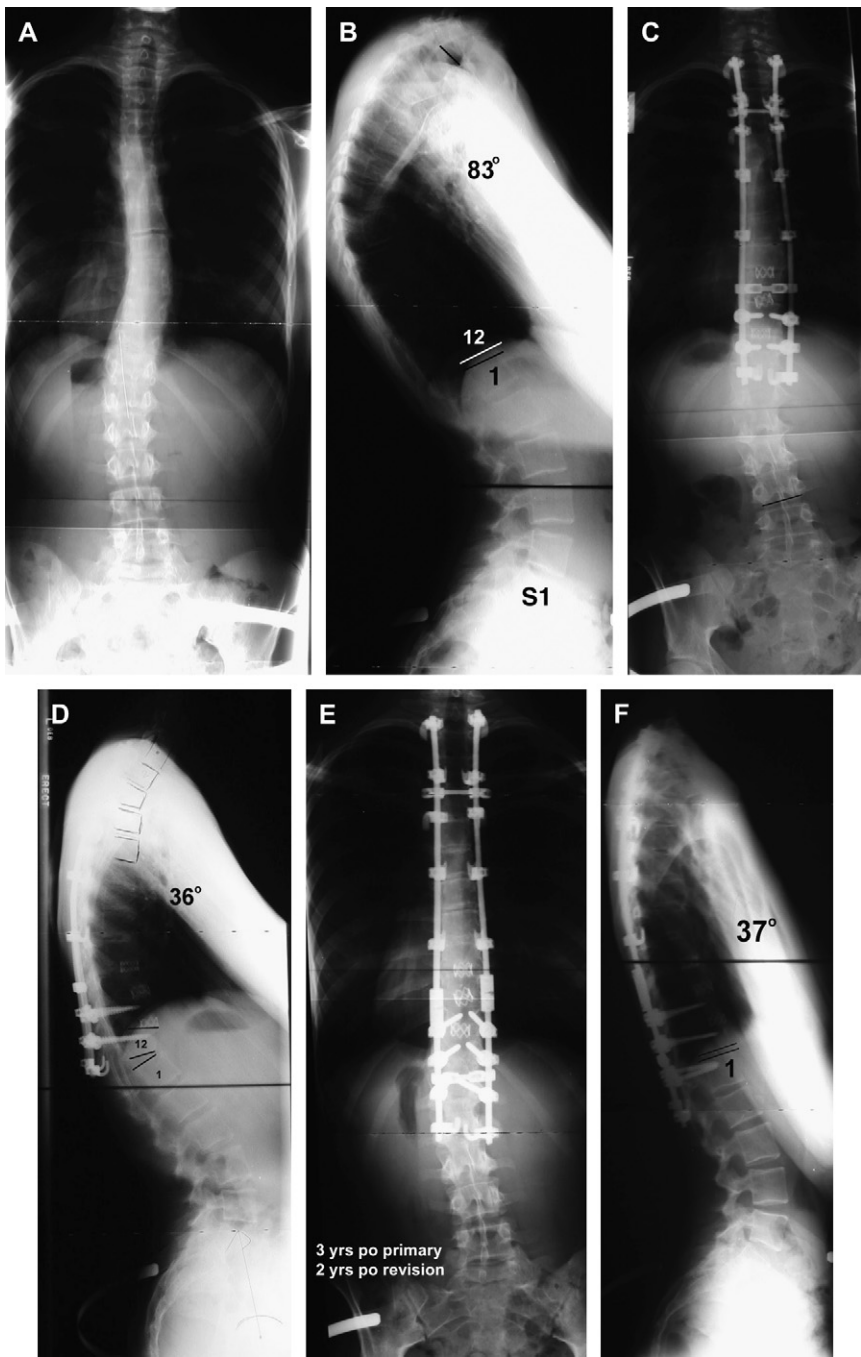


Fig. 13. (A–F) Radiographs demonstrate distal junctional kyphosis related to not including the first lordotic level distally. The instrumentation was extended one level distally with resolution of the distal junctional kyphosis.

Brace treatment is almost always successful in patients with kyphosis between 55° and 80° if the diagnosis is made before skeletal maturity. Kyphosis greater than 80° in the thoracic spine or 65° in the thoracolumbar spine is almost never treated successfully without surgery in symptomatic patients. Surgical treatment in adolescents and young adults should be considered if there is documented progression, refractory pain, loss of sagittal balance, or neurologic deficit. The major postoperative complication after surgical treatment is junctional kyphosis proximally or distally, which is usually related to not including all levels of the kyphosis or overcorrection of the deformity (> 50%). With proper patient selection, excellent outcomes can be expected with nonoperative or operative treatment in patients with Scheuermann's disease.

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