

# Atypical Scheuermann's Disease with Severe Kyphosis and Negative Sagittal Balance in the Thoracolumbar Region; A Case Report and Literature Review

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## INTRODUCTION

Scheuermann's kyphosis is a structural deformity of the spine that affects roughly 2.2% of the global population ranging from 0.4 to 10% [1, 2]. Scheuermann's kyphosis results in progressive thoracic and/or thoracolumbar hyperkyphosis in the spinal structure; consequently, ends in severe back pain in the afflicted population [3]. The structural deformity root in osteonecrosis of vertebral apophyses resulting in wedging of at least three adjacent vertebrae [1]. Another theory for the pathophysiology of this disease is alterations in endochondral ossification [4].

Patients with Scheuermann's kyphosis are classified into typical and atypical. Typical cases have thoracic spine involvement, while atypical Scheuermann's kyphosis cases have wedging in lumbar or thoracolumbar regions [5, 6]. The management of atypical cases initiate with conservative treatments such as bracing and analgesics, but for thoracolumbar curves greater than 25 to 30 degrees, surgical interventions are indicated, whereas the cutoff value for surgical interventions in thoracic Scheuermann's kyphosis is curvatures greater than 70 to 75 degrees [7]. However, surgical treatment of atypical Scheuermann's kyphosis is associated with multiple adverse effects, most importantly, junctional kyphosis [8]. It has been shown that spinal sagittal balance does not change significantly after surgical interventions, and is not correlated with preoperative thoracolumbar kyphosis [9, 10]. In this article, we present a case of atypical Scheuermann's kyphosis who underwent surgical intervention with anterior release and posterior fusion with a 2-year follow-up and excellent outcome without any complications or adjacent segment disease.

## CASE PRESENTATION

A 17-year-old boy from Shiraz, Iran, presented to our outpatient clinic with a kyphotic deformity in the lumbar region, which had begun developing two years beforehand. The patient also mentioned enuresis and non-significant low back pain in the upper lumbar region, which was intermittent and not radicular. The patient had no weakness, paresthesia, hyposthesia, numbness, or gait problems. The past medical, allergy, drug, and surgical history was unremarkable. The patient had left school and worked as a mechanic. In terms of family history, he had a sister who died of hepatic failure at the age of 13. The patient did not use tobacco, opium, or any specific substances. No domestic, social, financial or psychological problems were reported.

On physical examination, the patient had normal cranial nerves, normal muscle power (all 5/5), normal deep tendon reflexes (all 2+), bilateral downward plantar reflex, negative Hoffman sign, no sensory level,

no saddle hypoesthesia, and no incontinency or hesitancy. A kyphotic gait was observed. The preoperative radiographic evaluation of the patient revealed a lumbar lordosis (LL) of  $49^\circ$ , sacral slope (SS) of  $21^\circ$ , pelvic incidence (PI) of  $33^\circ$ , and pelvic tilt (PT) of  $11^\circ$  (Fig. 1A). The patient had a sagittal vertical axis (SVA) of 1.21cm (Fig. 1B), a thoracic kyphosis (TK) of  $70^\circ$  and thoracic local kyphosis (TLK) of  $85^\circ$  (Fig. 1C). The preoperative CT-scan of the thoracic space revealed several Schmorl's nodules and sclerotic endplates (Fig. 1D). The magnetic resonance imaging (MRI) revealed wedging of the three adjacent endplates, and disc space narrowing in thoracic spine (Fig. 1E, F). As the TK was less than  $75^\circ$ , the patient was treated with a thoracic lumbar sacral orthoses (TLSO) brace for six months, which had no positive effects on his symptoms. Thus, he was scheduled for an elective surgical fixation and correction of the thoracolumbar kyphosis. A preoperative diagnosis of kyphotic deformation in the sagittal plane with  $93^\circ$  Cobb's angle was made. The patient underwent surgery with intraoperative neuromonitoring through posterior-only approach. Bilateral pedicular screw fixation of T3 to L3 (except T10) was performed along with removal of inferior facet and spinous process and part of laminae of T5 to L2 (Smith-Peterson osteotomy). T10 pedicle subtraction osteotomy was also done for better correction. Two rods were applied bilaterally and open reduction of deformity was performed. The patient had an uneventful postoperative course and was discharged on fourth postoperative day with a TLSO brace. The postoperative imaging revealed a LL of  $60^\circ$ , SS of  $35^\circ$ , PI of  $49^\circ$ , and PT of  $14^\circ$ , all improved compared to preoperative parameters (Fig. 2A). The TK and TLK decreased to  $40^\circ$  and  $9^\circ$ , respectively (Fig. 2B). The SVA also increased to 5.06cm (Fig. 2C). Table 1 compares the sagittal balance parameters of the patient pre- and postoperatively.

The remarkable change in the physical examination was a normal gait, while enuresis was also relieved. In follow-up, he had normal lower extremity motor and sensory function with improved function of the sphincters. At 6-month postoperative visit, he had normal motor, sensory, sphincter, and gate function with no sign of adjacent segment disease was found. The patient was doing well without any complications at 1-year follow-up.

## DISCUSSION

We herein, reported a 17-year-old boy with atypical Scheuermann's kyphosis who underwent Smith-Peterson Osteotomy and posterior fixation surgery, and unlike most cases, had a remarkable improvement in his signs and symptoms with no significant complications. Usually, the management of Scheuermann's kyphosis is based on conservative treatment of the patient's symptoms, and surgical interventions are reserved for severe cases [11]. The current concept regarding the management of the patients with Scheuermann's kyphosis is based on the TK degree. Patients with less than  $60^\circ$  TK are managed conservatively with analgesics, physical therapy, and sport medicine. In patients with TK of  $60$ - $75^\circ$ , hyperextension brace is administered with the aim of decreasing  $15^\circ$  of the TK. In those, with TK of more than  $75^\circ$ , surgery is indicated [11]. Surgery is also indicated in those with intractable pain, neurological impairment, and progressive curve [4, 12].

In the surgical approach to typical Scheuermann's kyphosis, two methods are well established; the first one is the combination of anterior release and posterior fusion, and the second one is the posterior-only approach. Since anterior release and posterior fusion in Scheuermann's kyphosis composes of two consecutive interventions, it is accompanied by more complications, duration of operation, and blood loss. It is also accompanied by higher rates of neurologic, cardiologic, and pulmonary complications [12, 13]. The aim of the surgery is not only the correction of the kyphosis, but also correction of the all-sagittal balance parameters [13]. In order to improve the pain and the neurological symptoms, the cervical, thoracic and lumbar parameters should be corrected during the surgery [8, 10]. The stable vertebra should be detected preoperatively in order to design a perfect construct for correction of the spine parameters [13, 14]. The most common long-term complications of the surgery remains the distal junctional disease compared to the proximal disease [15, 16]. In this case report, we performed a posterior-only approach for correction of the kyphosis in this young boy. The posterior-only approach along with Smith-Peterson osteotomy, successfully improved the TK and all the sagittal balance parameters. We experienced no intraoperative and postoperative complication and the patient's sign and symptoms relieved successfully. Thus, we recommend evaluation of the sagittal spine parameters preoperatively in order to perform a successful posterior-only approach for

correction of the TK and improving the patient’s sign and symptoms.

There are various differences between the typical and atypical Scheuermann’s kyphosis. The typical Scheuermann’s kyphosis is mid-thoracic and encompass the radiological signs of the disease such as Schmorl’s nodule, vertebral wedging and disc narrowing more frequently [5]. The atypical Scheuermann’s kyphosis is usually thoracolumbar and is more frequently symptomatic compared to the typical disease [1, 6, 11]. In addition, patient with atypical symptoms have less frequently the radiological characteristics of the disease [4, 7]. The adult Scheuermann’s kyphosis is also accompanied by a 25% rate of scoliosis that makes the entity more complex [1, 11].

Weiss et al. showed that a holistic approach to the rehabilitative programs directed toward patients with thoracic and thoracolumbar Scheuermann’s kyphosis can significantly decrease the pain in the patients [17]. Previously, combined posterior and anterior approaches were the gold standard of treatment in surgical interventions, but recently, posterior-only approaches are brought into attention due to fewer postoperative complications and better outcomes [7]. Dai et al. reported a series of atypical lumbar Scheuermann’s kyphosis associated with scoliosis in a family and concluded that this condition might have genetic predisposing factors. The patient underwent surgical fixation and in the three-month follow-up of the patient, no major complication was found [18].

Wang et al. also reported a case of atypical Scheuermann’s kyphosis in a patient with heterozygous mutation in COL1A2 gene. Consequently, the patient was diagnosed with osteogenesis imperfecta. An association might exist between atypical Scheuermann’s kyphosis and osteogenesis imperfecta, but since patients with atypical lumbar Scheuermann’s kyphosis do not regularly undergo genetic sequencing, such associations are unclear [19]. The patient in that study did not undergo operation and the comparison of results is not possible. Nasto and colleagues performed a study on 64 patients with Scheuermann’s kyphosis and 33 controls. The patients underwent surgery and were followed-up for six months. They evaluated and compared the post-op cervical sagittal alignment in patients with thoracic and thoracolumbar Scheuermann’s kyphosis. They found that the cervical lordosis increased in patients with thoracolumbar Scheuermann’s kyphosis, whereas the patients with thoracic Scheuermann’s kyphosis had a decrease in their cervical lordosis [20].

Van Loon and colleagues performed pedicle subtraction osteotomy on eleven patients with atypical Scheuermann’s kyphosis and the patients were followed-up for about 3.5 years. They showed that this procedure was significantly effective in decreasing the pain of the participants. They concluded that this intervention could be used as an add-on option to previously established treatments of atypical thoracolumbar Scheuermann’s kyphosis [21]. Tsirikos reported two cases with atypical thoracolumbar Scheuermann’s kyphosis who had spontaneous fusion and ankylosis in the kyphotic vertebrae. The author concluded that in patients with anteroposterior fusion of the discs, an anterior release of the vertebrae and a posterior fusion might be the intervention of choice [22].

In a case report by Lamartina, a 27-year-old man with severe kyphosis was operated on by a posterior approach for fusion of T3-L2 vertebrae. No data regarding the follow-up of the case is available, but this approach is speculated to have less post-operative neurologic complications since the spinal cord is less likely to be manipulated [14]. In our case, in a follow-up period of two years, no neurologic deficit was seen and no further surgical intervention was needed. In a study by Heegaard et al. performed on 22 cases, the authors reported that the complication rate in the surgical treatment of Scheuermann’s kyphosis was as high as 77% [16]; however, the authors did not evaluate the complications based on the surgical approach. In a much larger study by Coe et al. performed on The Scoliosis Research Society morbidity and mortality database of 683 patients, the authors found that the complication rate for surgical repair of Scheuermann’s kyphosis was 14%. Only 1.9% of the patients suffered from acute neurologic complications. They also compared the posterior-only approach to the patients with combined anterior and posterior approach and found that although the complication rate in posterior-only approach was lower than in combined anterior and posterior approach, this difference was not statistically significant [15].

Lonner and colleagues reported the surgical outcomes in 78 typical Scheuermann’s kyphosis patients and

compared the posterior-only and combined anterior and posterior approaches. They found that patients undergoing posterior-only surgical interventions had better outcomes; however, in that study, the preoperative lordosis angle, kyphosis angle, and greatest Cob angle in the two groups differed significantly; consequently, no definite conclusion could be made based on this study's results [23]. Etemadifar et al. compared the complications and outcomes of the two approaches in 30 patients with Scheuermann's kyphosis. They concluded that the surgical outcomes of the two approaches do not differ significantly; however, the complications of posterior-only surgical approach were much lower than the combined anterior and posterior approach; consequently, posterior-only approach is usually the first recommended surgical intervention to treat this condition [12].

Some rare complications have also been shown to be associated with anterior release and posterior fusion surgery. For instance, Daniels et al. reported acute celiac artery compression syndrome as a complication of this intervention [24]. Moreover, Soares do Borito et al. encountered anterior sternoclavicular dislocation after this surgery [25]. In our case with atypical thoracolumbar Scheuermann's kyphosis, anterior release and posterior fusion was performed and no complication, either acute or chronic, mild or severe, was encountered in the two-year follow-up period. We have summarized the summary of the previously reported cases in Table 2.

## CONCLUSION

Atypical Scheuermann's kyphosis with neurological impairment and progressive kyphosis should be treated with surgical intervention. Considering the sagittal parameters of the spine, the surgical intervention should be designed to correct the kyphosis and the other indices of the sagittal balance. Posterior-only approach is safe and effective method for correction of the TK and improving the signs and symptoms of the patients. Further studies with larger populations are required to shed light on the subject.

## ABBREVIATION

**CT:** Computed Tomography

**LL:** Lumbar Lordosis

**MRI:** Magnetic Resonance Imaging

**PI:** Pelvic Incidence

**PT:** Pelvic Tilt

**SS:** Sacral Slope

**SVA:** Sagittal Vertical Axis

**TK:** Thoracic Kyphosis

**TLK:** Thoracic Local Kyphosis

**TLSO:** Thoracic Lumbar Sacral Orthoses

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