

Case Report: Two-Stage Procedure Planning for the Surgical Cor- rection of Severe Congenital Kyphosis: A Technical Case Report



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ABSTRACT

Congenital kyphosis is a less common spinal malformation, which can progress and cause neurological deficits. The treatment of severe cases is complicated. There are several techniques of surgical intervention for the correction of kyphosis, but the selection of surgical methods is entirely dependent on the patient's condition. This article reports a 14-year-old girl with severe congenital kyphosis, who underwent hemiepiphysiodesis at age 2. Last year, her neurological deterioration began and led to paraplegia over a couple of months. To treat her, we performed 2-stage multi-level vertebral column resection (Schwab type 6). First, she underwent anterior T12-L1-L2 complete corpectomy and cord decompression. Then, the resection of posterior elements from T12 to L2 and posterior fusion with instrumentation from T8 to L5 were performed. However, concerning this kind of severe deformity, we suggest using 3D planning, which facilitates the operation.

1. Introduction

K yphosis is the term used to describe the spinal curve, which results in an abnormal round back. Kyphosis can be caused by many reasons such as trauma, inflammatory and infectious diseases, developmental anomalies, and so on [1]. James was the first who used the term "congenital kyphosis" in 1844 and divided

congenital kyphosis into two types. The first type was the failure of the formation of the vertebral body, and the second type was the failure of its segmentation [1, 2].

Later, Winter et al. described a classification that is widely accepted and used. In Winter's classification, congenital kyphosis is divided into three types. Type 1 is the most common type of congenital kyphosis and refers to the failure of vertebral body formation. Type 2 is

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the failure of vertebral body segmentation. Type 3 or the mixed type is the combination of the failure of vertebral body formation and segmentation and is the least common form of congenital kyphosis [3-5].

The etiology for congenital kyphosis is still unknown despite the different environmental and or genetic theories. Congenital kyphosis can progress and cause neurological deterioration, which occurs in 10% to 12% of patients [6]. Studies show that congenital kyphosis is progressive in the growth years of life, and surgical intervention is necessary because of the ineffectiveness of brace treatment [2, 5, 7]. However, there is no standard procedure for the correction of all types and sizes of deformities. And the surgeon must choose the method of operation based on the patient's age, type and size of the vertebral anomaly, and the presence, severity, or absence of spinal cord compression. The success of the operation depends on selecting the right method and applying it at the best time [8].

Neurologic complications are common during the interventions for kyphosis treatment [9]. Therefore, neurophysiologic monitoring of the spinal cord and normotensive anesthesia are mandatory. Various techniques of interventions in the early phase of deformity include anterior instrumentation, all-posterior approach for hemivertebrae excision, anterior and posterior hemivertebrae excision, and non-instrumented fusion [7].

Congenital kyphosis is a rare and uncommon deformity and is usually present on a single level. We report a 14-year-old girl suffering from a high magnitude multi-level congenital kyphosis. Because of the severity of her condition, the patient underwent multiple vertebral resections in two stages: anterior corpectomy of 3 levels

followed by the resection of posterior elements of 3 segments and instrumentation one week later.

2. Case Presentation

A 14-year-old girl presented with severe congenital thoracolumbar kyphosis and complete paraplegia (Figure 1). From 1 year ago, the muscle strength of her lower limb gradually and progressively reduced. Since the previous month, her condition deteriorated so much that she developed paraplegia with the loss of muscle force and sensation and urinary and bowel incontinence.

She had a history of hemiepiphysiodesis at the age of 2 that was not effective in controlling the curve progression. On physical examination, muscle strength was noted normal throughout the upper extremities. There were normal deep tendon reflexes and negative Hoffman signs bilaterally. The muscle strength of the lower extremities was reported at 0/5. Upward Babinski and exaggerated deep tendon reflex in the lower limbs were noted. Her physical exam revealed an old posterior spinal and a left lateral thoracic scar beside a severe gibbous deformity.

The authors planned a surgical procedure with two-stage Vertebral Column Resection (VCR): first, anterior T12-L1-L2 corpectomy and cord decompression and then, the resection of T12-L1-L2 posterior elements in addition to posterior fusion and instrumentation from T8 to L5 with pedicular screws.

At the first stage of the operation, the patient was under general anesthesia and turned on the right lateral decubitus. To enter the thoracoabdominal region, we made an incision over the previous incision between the 10th and

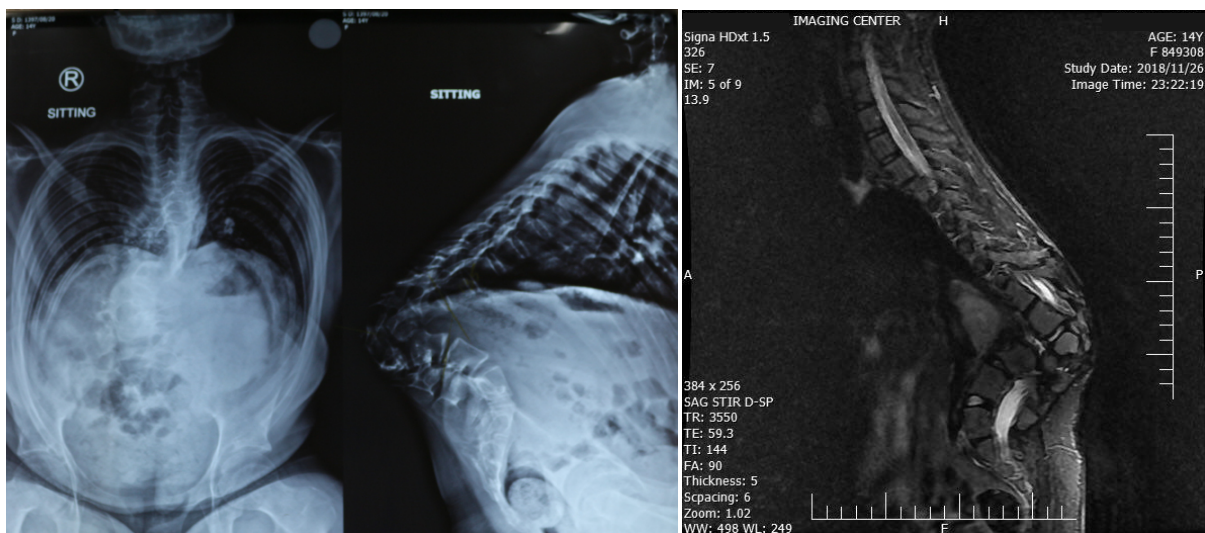
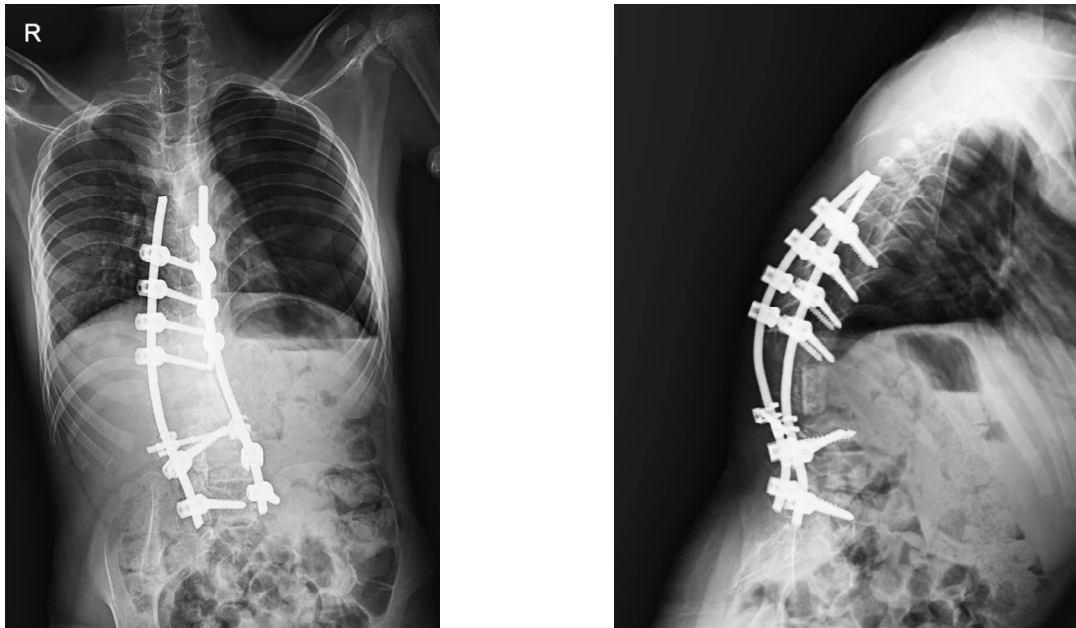


Figure 1. Radiography and preoperative MRI shows severe kyphosis



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Figure 2. AP and lateral radiography after two stage vertebral column resection and instrumentation

12th ribs. For extensive exposure of T12-L1-L2 area, the diaphragm was incised and, then, complete corpectomy and discectomy from T12 to L2 were performed. Complete decompression was performed at T12 to L3, while the dura was exposed and released. Then, a fibular structural allograft with cancellous autograft was inserted. At the end, parietal pleura and diaphragm were repaired with mesh augmentation, and the chest tube was applied. The muscles and skin were repaired. The operation took approximately 6 hours and the patient lost about 850 mL blood during this procedure.

One week later, in the second stage of the operation, a posterior midline incision was made under general anesthesia in a prone position, then paravertebral muscles were retracted bilaterally, and the spine was exposed from T7 to L5. Instrumentation consisted of pedicular screw insertion from T8 to T10 and also L3 and L5 bilaterally (Figure 2). The resection of posterior elements (lamina, facet, and pedicles) of T12-L1-L2 was done. Then, by using two temporary rods, the sequential correction was obtained.

Final rods were inserted with an appropriate bend, osteotomy site was closed, and kyphosis was corrected. A mixture of cancellous allograft and autograft was applied in the corpectomy site; then, the decortication of the fusion bed was performed, and Bone Grafts was located posterolaterally. Finally, the fascia was repaired, and the LT side chest tube was applied. Intraoperative bleeding

was approximately 1100 mL; thus, two units of packed cells were transfused during the 8-hour operation.

3. Discussion

The prevalence of spinal malformations is approximately 0.5 to 1 per 1000 people. According to various studies, congenital kyphosis is far less common than other malformations [7, 10]. Both kyphosis and scoliosis commonly exist together in congenital vertebral deformities [6]. Congenital kyphosis is one of the less common malformations, which is characterized by the impairment of the longitudinal growth of vertebral elements in the sagittal plane [11, 12]. The apex of deformity usually occurs in the thoracolumbar junction [7].

Congenital kyphosis progresses with patients' growth and can cause spinal cord compression and severe neurologic deficits. Therefore, surgical intervention is indicated for its treatment. Non-surgical approaches such as bracing prove to be non-effective in the management of congenital kyphosis [5]. Characteristics of the patient and anomaly will determine the operative approach [13]. The classification of congenital kyphosis is important to identify possible neurologic deficits [1]. For instance, type 1 deformity will lead to spinal cord compression and paraplegia if left untreated [3, 8].

According to Zhang et al. study, the one-stage operation for deformity correction is safer, but in our case, a

two-stage approach was needed because of the severity of kyphosis. However, the average blood loss in both procedures was similar to this study [14].

Pijpker et al. published a case report of 3D planning for pedicle subtraction osteotomy operation in a young girl. It seems that utilizing this 3D virtual planning can facilitate the process of operation by identifying vertebral levels and malformed vertebrae. Our case required 2 days of work. This method reduces the duration of operation [15].

Our patient with severe congenital kyphosis was presented in T12, L1, and L2 following an anterior-posterior hemiepiphysiodesis performed 12 years ago. We did an anterior T12-L1-L2 complete corpectomy and fusion, as well as posterior element resection in T12-L1-L2.

In Conclusion, the present severe case of congenital kyphosis was corrected after two long and serious operations as multi-level (T12-L1-L2) VCR. Because of the severity of this case, we need to follow up the patient to conclude the success of the procedure.

Ethical Considerations

Compliance with ethical guidelines

All ethical principles were considered in this article and the informed consent was taken from the patient.

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Authors' contributions

All authors contributed in preparing this article.

Conflict of interest

The authors declared no conflict of interest.

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