Clinical Case Report

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Fractional curve progression with maintenance of fusion mass in congenital scoliosis

An 18-year follow-up of a case report

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Abstract

Rationale: The management of congenital scoliosis concentrates on early diagnosis and proper surgical treatment before the development of severe deformities. Decision making regarding the appropriate fusion levels, proper surgical treatment, and reduction amount of kyphoscoliosis is very important but difficult in the treatment of congenital scoliosis, especially in young children.

Patient concerns: We report an 11-year follow-up of revision surgery for fractional curve progression after combined anterior and posterior fusion without hemivertebra resection using pedicle screw fixation (PSF) in congenital kyphoscoliosis at age 4 years (a total 18-year follow-up). A T12 hemivertebra was documented in a 4-year-old girl and was treated by combined anterior and posterior fusion in two stages with PSF. The fusion mass was maintained but the distal compensatory curve progressed during the follow-up period. The patient underwent a posterior vertebral column resection (PVCR) with extended posterior fusion at the age of 11, 7 years after initial surgery.

Outcomes: Eleven years after the revision surgery with PVCR, the patient showed satisfactory results and her spine was well balanced.

Lessons: The cause of revision surgery for the curve progression may include inappropriate fusion level, incomplete hemivertebra resection, or failure of anterior and posterior fusion. Especially, inappropriate fusion level may result in deterioration of the compensatory curve even without progression of the fusion mass.

Conclusion: Appropriate selection of fusion levels, complete resection of hemivertebra, and satisfactory reduction of scoliosis and kyphosis are important factors for deformity correction and prevention of progression of both main and compensatory curves (adding-on of structural curve or progression of compensatory curve) as well as reducing the influence of adjacent vertebral growth using as short a fusion as possible.

Abbreviations: PSF = pedicle screw fixation, PVCR = posterior vertebral column resection, TLSO = thoraco-lumbo-sacral orthosis.

Keywords: congenital scoliosis, curve progression, fusion level, hemivertebra, posterior vertebral column resection

Editor: Vasile Valeriu Lupu.

J-HY and J-HL have equally contributed to this study

Institutional Review Board approval of Inje University Sanggye Paik Hospital (IRB No. 2015-07-014) was obtained before data collection and analysis.

The authors have no funding and conflicts of interest to disclose.

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Medicine (2017) 96:36(e7746)

Received: 21 July 2016 / Received in final form: 21 January 2017 / Accepted: 18 July 2017

http://dx.doi.org/10.1097/MD.0000000000007746

1. Introduction

Congenital scoliosis is caused by failure of formation or segmentation of vertebral elements that may lead to asymmetric growth of the spine.[1] Any hemivertebra can have a variable course, but most have growth potential, and those in scoliosis create wedge-shaped deformities that progress during growth[10]; compensatory curve develops in attempt to preserve trunk balance.[2–14]

The rate of progression and severity of the curve depends on the age of the patient, type of congenital anomalies, size of the deformities, and site at which the anomalies occur.[10] Therefore, decision making regarding the timing of treatment, type of surgery, reduction amount of kyphoscoliosis, and appropriate fusion levels is very important but difficult in the treatment of congenital scoliosis at a young age because natural evolution of the hemivertebra does not always lead to severe deformity, although worsening of the spinal deformity could occur during growth, usually in the form of a severe scoliosis curve rather than a local malformative curve.[11,12]

We report a case of fractional curve progression after combined anterior and posterior fusion without hemivertebra resection using pedicle screw fixation (PSF) that was treated by revision surgery with posterior vertebral column resection (PVCR) and extended posterior fusion in congenital scoliosis in a young patient.

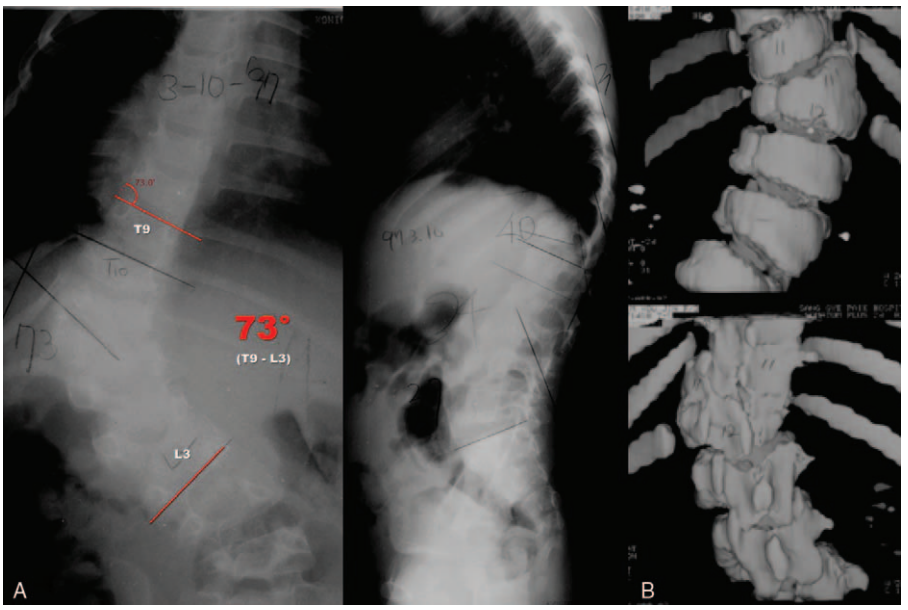

Figure 1. (A) Radiographs taken before the initial surgery showing a congenital hemivertebra at T12 with 73° of left thoraco-lumbar scoliosis and segmental kyphosis of 40°. (B) 3D reconstruction computed tomography scan before surgery.

2. Case report

A female was born in 1993, following a full-term normal pregnancy and vaginal delivery. There was no history of drug use, maternal toxin, or smoking exposure during the pregnancy. The family history did not include any cases of congenital spinal anomalies.

The patient had been noticed by her parents to have an asymmetry in her back at the age of 2 months and was referred to our hospital by a private physician after a radiograph showed congenital anomalies at T12 and a left thoracolumbar scoliosis with segmental kyphosis.

The physical and neurological examinations were otherwise within normal limits, and magnetic resonance imaging was unremarkable. Associated malformations were ruled out during a clinical work-up including investigation of craniofacial malformation, cardiac and urinary malformations, and foot or leg asymmetry, with unremarkable findings. At the first visit to our hospital, the patient was prescribed a thoraco-lumbo-sacral orthosis (TLSO).

The patient had a left thoracolumbar scoliosis of 73° (T9–L3) and segmental kyphosis of 40° (T10–L2) due to a T12 hemivertebra (Fig. 1A and B). Because of the known progression of this vertebral anomaly, surgical treatment was performed. At the age of 4 years, the patient underwent combined anterior and posterior fusion without hemivertebra resection using PSF from T10 to L2. The preoperative scoliosis of 73° improved to 42° postoperatively and the segmental angle of kyphosis of 40° to 20° (Fig. 2A). The patient had no subjective symptoms and did well after surgery. One year after surgery, the main curve (T9–L3: 44°)

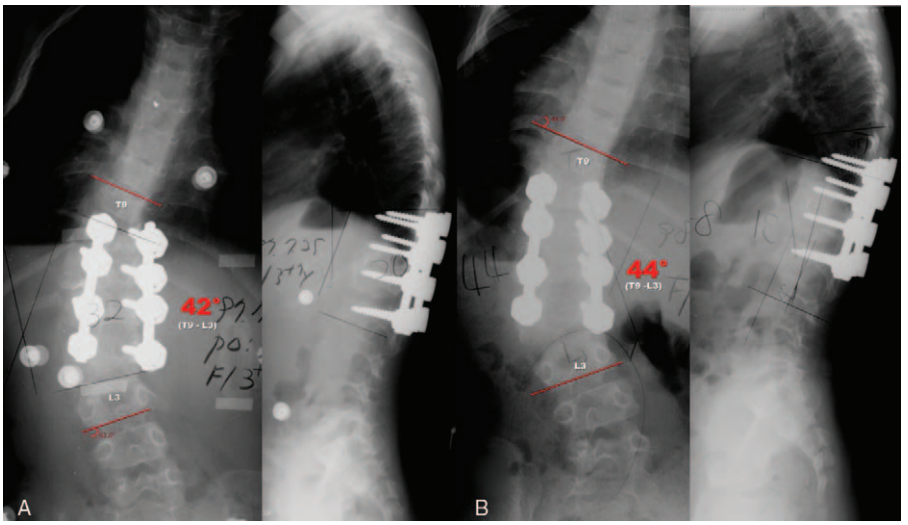

Figure 2. (A) Radiographs taken 3 months after the initial surgery showing that the main curve of scoliosis improved to 42° and segmental angle of kyphosis to 20°. (B) Sixteen-month follow-up radiographs showing that main curve (T9–L3: 44°) and fusion mass (T10–L2: 33°) were well maintained with slight progression of the distal compensatory curve from 14° to 18°.

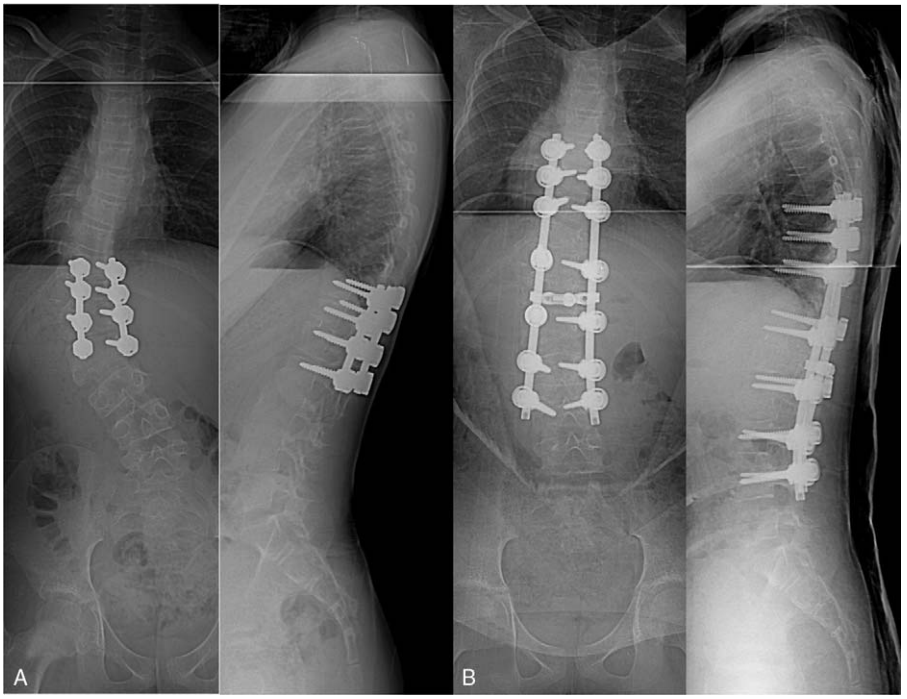

Figure 3. (A) Seven-year follow-up radiographs after the initial surgery showing that the main scoliotic curve deteriorated to 54° and distal compensatory curve has progressed to 33°. (B) Immediate postoperative radiograph after revision surgery with posterior vertebral column resection showing that the main curve of scoliosis improved to 14° with distal compensatory curve of 2°.

and the fusion mass (T10–L2: 33°) were well maintained with slightly progression of the distal compensatory curve from 14° to 18° (Fig. 2B). However, at the age of 11 years (7 years after surgery), the left thoracic scoliosis increased to 54°, and the distal compensatory curve has progressed to 33° (Fig. 3A). A PVCR of a T12 hemivertebra with extended posterior fusion using pedicle screws from T7 to L4 was performed. The main curve of scoliosis and the distal compensatory curve improved to 14° and 2° postoperatively, respectively (Fig. 3B), and were 20° and 9° at 11 years post-PVCR (Fig. 4A and B) with a well-balanced spine.

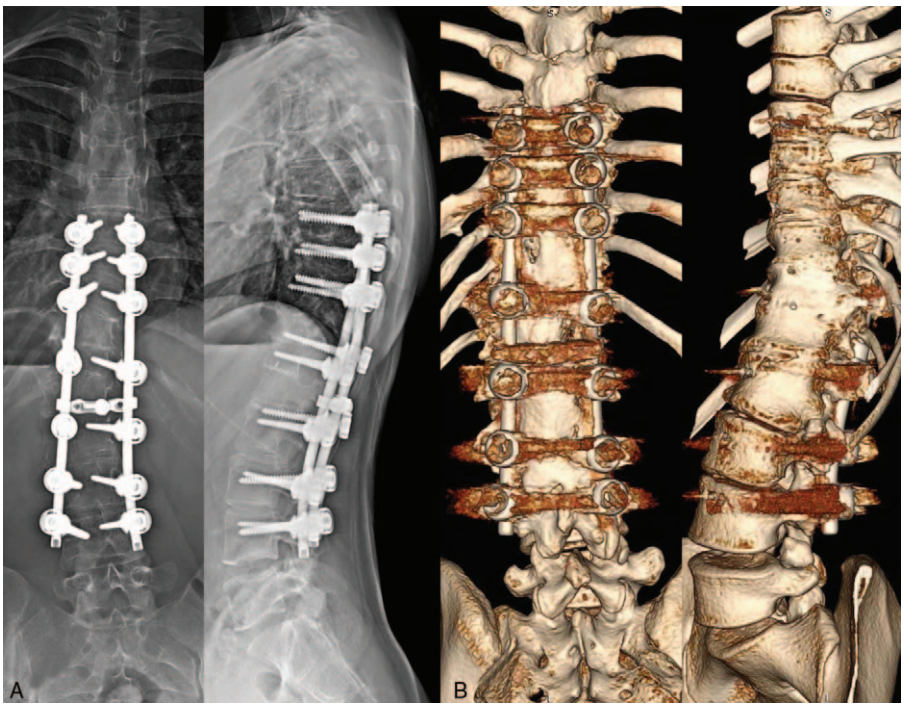

Figure 4. (A) Eleven-year follow-up radiographs after PVCR showing the main scoliotic curve well maintained at 20° and distal compensatory curve at 9°. (B) Eleven-year follow-up 3D reconstruction computed tomography scan after PVCR. PVCR = posterior vertebral column resection.

The patient was mobilized 2 weeks after PVCR in a localizer cast that was worn for 4 months and then a TLSO for 6 months.

3. Discussion

The management of congenital scoliosis concentrates on early diagnosis and proper surgical treatment before the development of severe deformity of the primary and compensatory curves.[10,13–16] Curative treatment of congenital hemivertebra should include removal of the hemivertebra.[17–20] The usual method of hemivertebra resection has been combined with anterior and posterior surgery in 1 or 2 stages.[1–6,9] Recently, posterior hemivertebra resection or PVCR in pediatrics has been reported with satisfactory results.[11–16]

The cause of revision surgery for the curve progression may be due to inappropriate fusion levels, incomplete hemivertebra resection, and/or failure of anterior/posterior fusion.[17–20] In our case, progression of the distal compensatory curve was not associated with increased curvature of the main curve was fused with the segmental PSF because of decompensation below the fusion mass rather than of the main curve. This suggests that the cause of curve progression may be due to inappropriate selection of fusion levels (short fusion) and incomplete resection of hemivertebra with insufficient reduction of the deformity.

Complete hemivertebra resection, satisfactory reduction of kyphoscoliosis, and appropriate selection of fusion levels are important for clinical outcomes, which mean proper surgical treatment, reduction amount, and selection of fusion levels could influence the amount of reduction. Initially, our patient underwent combined anterior and posterior fusion without hemivertebra resection using PSF from T10 to T12, which may have caused unsatisfactory deformity correction, which was the first mistake in our treatment. The curative treatment of congenital scoliosis due to hemivertebra should include removal of the hemivertebra.[20–23] We overlooked this process and the effect of the remaining hemivertebra growth potential. Therefore, we performed revision surgery using PVCR, which can correct deformity corrections through sufficient shortening of the posterior column and can achieve more bony union through bone to bone contact of vertebral bodies.[20,21] The successful revision surgery of the patient likely resulted from recognition of failure factors such as inadequate surgical correction of congenital scoliosis and reduction condition.

In our case, the curve of the fusion mass (T10–L2) had not increased and was maintained at about 32° to 33° during the follow-up period, but deformity progression occurred in the distal compensatory curve. This means that the hemivertebra was not the only cause of the increase in the distal compensatory curve. Although the discs and endplates above and below the hemivertebra were removed in the initial surgery, the hemivertebra and pedicle were not. Remnant of the hemivertebra after surgery can cause deterioration of the main curve as well as the compensatory curves.[20–23]

There is a limitation to our case. Although many cases present with similar symptoms, definite conclusions cannot be drawn from a single case report. However, there are very few previously published case reports on long-term follow-up after PVCR for fractional curve progression after combined anterior and posterior fusion with PSF in congenital scoliosis at a young age, so this case is valuable. Second, our X-rays included old-scanned films (Figs. 1 and 2); therefore, we could not delete the previous hand drawing on X-rays. To overcome this shortcoming, the X-ray radiograph was digitized using the Picture Archiving and Communication System, and we added a new digital drawing of the main curve of the scoliosis (T9–L3) over the previous X-ray radiograph for better visibility and precision.

In conclusion, determination of the appropriate fusion level, proper surgical treatment, and reduction amount of kyphoscoliosis are very important but difficult in the treatment of congenital scoliosis, especially in young children. Therefore, appropriate selection of fusion levels, complete resection of hemivertebra, and satisfactory reduction of scoliosis and kyphosis are important factors for deformity correction and prevention of progression of both main and compensatory curves (adding-on of structural curve or progression of compensatory curve), as well as reducing the influence of adjacent vertebral growth using as short a fusion as possible.

Acknowledgments

The authors thank Dr P. William Haake for English-language editing and Yeon-Seok Jeong for data collection of this paper.

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