Congenital Kyphoscoliosis in Monozygotic Twins: Ten-Year Follow-up Treated by Posterior Vertebral Column Resection (PVCR)

A Case Report

Dong-Gune Chang, MD, Jae Hyuk Yang, MD, Jung-Hee Lee, MD, Young-Hoon Kim, MD, Jin-Hyok Kim, MD, Seung-Woo Suh, MD, Kee-Yong Ha, MD, and Se-II Suk, MD

INTRODUCTION

Congenital scoliosis could be derived from the failure of formation or failure of segmentation, which are etiologically heterogeneous with genetic, epigenetic, and environmental factors contributing to their occurrence. Developmental imbalance of the vertebral anomalies during the growth may cause various types of kyphosis, scoliosis, or kyphoscoliosis. There are very few reports of congenital scoliosis in monozygotic twins. Furthermore, there have been no reports in the literature of long-term follow-up after surgery following posterior vertebral column resection (PVCR), anterior support with a mesh cage, fusion and fixation using pedicle screws almost simultaneously in 2 monozygotic twins with analogous congenital scoliosis.

CASE REPORT

Two male monozygotic twins, born in 1991, following a full-term pregnancy by cesarean section. The pregnancy was uneventful. Birth weights were 1870 g (twin I) and 2800 g (twin II), respectively, and twin I was treated for low birth-weight in the incubator for 1 month. There was no history of maternal toxin, drug, or smoking exposure during the pregnancy. The family history did not contain any cases of congenital spinal anomalies.

Twin I had been noticed by his parents to have asymmetry of his back at age 5 years, but no treatment was given. Overtime, his spine became further deviated to the left with kyphosis and was referred to our hospital. Both monozygotic twins were treated by PVCR and satisfactory results were demonstrated at 10-year follow-up.

This case is the first report on the surgical treatment with PVCR, almost simultaneously in both identical twins who had similar congenital vertebral anomalies causing kyphoscoliosis. Both identical twins with congenital kyphoscoliosis had undergone surgical correction by PVCR, anterior support with a mesh cage and posterior fusion using pedicle screws at the age of 14 years and achieved a satisfactory correction and a stable spine without curve progression with 10-year follow-up.

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Abbreviation: PVCR = posterior vertebral column resection.
Surgery was conducted with somatosensory evoked potential (SSEP) monitoring, and we checked motor/sensory after surgery and during postoperative care. Motor evoked potential (MEP) monitoring was not available in early years of our study because MEP was introduced in 2007 in our hospital. Surgical procedure was performed with PVCR and posterior fusion.17–19

Twin I had a left thoracolumbar scoliosis of 50° (T11-L2) and segmental kyphosis of 52° (T11-L2) with T12 and L1 hemivertebra (Figures 1A and B). At the age of 14 years, Twin I underwent a PVCR of L1, hemivertebra resection of T12, anterior support with a mesh cage and posterior fusion with pedicle screws from T9 to L3 (Figure 2A). The preoperative scoliosis of 50° improved to 13° postoperatively and maintained to 17° at 5-year follow-up (Figure 2B) and 18° at 10-year follow-up (Figures 3A and B). The segmental angle of kyphosis of 52° before surgery improved to 12° postoperatively and maintained to 10° at 10-year follow-up (Figure 3C).

Twin II had a left thoracolumbar scoliosis of 28° (T11-L2) with severe kyphosis of 86° (T10-L1) due to T11 and T12 hemivertebra (Figures 4A and B). One week after the operation of twin I, twin II underwent a PVCR at T11-T12, anterior support with a mesh cage, and posterior fusion with pedicle screws from T7 to L2 (Figure 5A), a similar procedure as in twin I. The preoperative scoliosis of 28° improved to 6° postoperatively and maintained to 7° at 5-year follow-up (Figure 5B) and 5° at 10-year follow-up (Figures 6A and B). The segmental angle of kyphosis of 86° before surgery improve to 35° postoperatively and 33° at 5-year follow-up and 35° at 10-year follow-up (Figure 6C).
Both twins were mobilized 2 weeks after surgery in a localizer cast that was worn for 4 months and then a thoraco-lumbo-sacral orthosis (TLSO) for 6 months.

**DISCUSSION**

The etiology of congenital scoliosis and its development remains unclear and has not yet been fully identified.1–16 The risk of deformity progression and its subsequent severity is affected by various factors including the number, type, and location of vertebral anomalies.1–3 There are very few reports of congenital scoliosis in monozygotic twins. Furthermore, no reports have been described in the literature about surgical treatment with PVCR almost simultaneously in identical twins with a long-term follow-up.20–25 Associated malformations should be looked for clinically and with an appropriate work-up. Thorough physical examination including foot or leg asymmetry, craniofacial malformation, cardiac and urinary malformations should be performed as well as neurological examination including abdominal reflexes and root tension signs.24–27 Both twins in our study showed normal physical and neurological examinations. Our case of monozygotic twins with analogous congenital spine anomalies was presented in both with significant kyphoscoliotic deformity at the thoracolumbar junction.

**FIGURE 3.** Twin I radiographs and clinical photographs. (A) Ten-year follow-up radiographs showed that the main curve of scoliosis well maintained to 18° and segmental angle of kyphosis to 10° without curve progression. (B) Ten-year follow-up 3D CT. (C) Ten-year follow-up clinical photographs. CT = computed tomography.

**FIGURE 4.** Twin II at age 14 years. (A) Twin II had congenital hemivertebra of T11 and T12 with 28° of left thoraco-lumbar scoliosis with severe segmental kyphosis of 86°. (B) 3D reconstruction CT scan. CT = computed tomography.
However, there were some differences of clinical features between the twins. Twin I had low height and weight and was more severe trunk shift and thoracolumbar hump compared with Twin II.

Diagnosis of congenital scoliosis/kyphosis is confirmed with whole spine posterior–anterior and lateral radiographs and clinical findings, which were essential for the diagnosis of congenital scoliosis. Computed tomography (CT) with thin slices and reconstruction is useful, and MRI provides high-quality pictures of the cartilage end plates, possibly giving the best information on growth potential.

The treatment goal of congenital spine deformities is to obtain a balanced trunk and spine while maintaining as much normal spinal growth as possible and preventing neural deficit. Complications that could occur in untreated congenital kyphoscoliosis include progression of curve, shoulder imbalance, trunk shift, pulmonary compromise, or directly damage the neurologic elements by growing in the canal.

Conservative treatment with bracing or localizer cast is not well indicated because braces usually do not affect progression of the congenital curve even if braces sometimes could be used to control compensatory curves that develop above or below the congenital curve.

The principle of surgery would give rise to allowing best management of severe complex pediatric deformities. Surgery is performed when progression of the curve is documented or an anomaly is predicted to have high risk of progress. There are 2 different main principles of surgery: (1) prophylactic...
surgery and (2) corrective surgeries.\textsuperscript{24–26} Surgical treatment should be selected for correcting the deformity, preventing curve progression, and reducing the influence of adjacent vertebral growth.

Surgery is usually recommended for patients with severe spinal deformity, but the surgical treatment for severe spinal deformities in children is extremely challenging. There are various types of surgical procedure in the treatment of congenital kyphoscoliosis. However, the definitive treatment of the congenital kyphoscoliosis due to hemivertebrae should include removal of hemivertebrae. Recently, hemivertebrae excision through posterior approach and PVCR in pediatrics have been reported as the treatment of choice with successful results.\textsuperscript{17} The indication of PVCR was congenital spinal deformities with a curve magnitude $>30^\circ$ with fast progression. This included documented progression of curve with $>10^\circ$ in a 1-year period, failure of the conservative treatment, or both.

Congenital kyphoscoliosis is a complex 3-dimensional deformities with rotation.\textsuperscript{28} PVCR could enable translational correction as well as rotational correction of the spine with a controlled manipulation of the both spinal columns simultaneously via a single posterior approach.\textsuperscript{17–19}

In our case, monozygotic twins with analogous spinal anomalies was described. Differences of vertebral anomalies at birth and differences of deformity type with different curve patterns during the growth between twins may be associated with complex interactions mediated by genetic, epigenetic, and environmental factors during the development of congenital scoliosis.\textsuperscript{11} However, little information exists regarding the relative contribution of each of factors to the development of congenital scoliosis.\textsuperscript{1–3}

The limitation of the study is that this case reports represents a rarity for the very fact that the anomalies occurred in monozygotic twins or that genetic plays a role in linking these twins. Grauer et al reported the findings on heritability of scoliosis following a survey of 64,578 twins in the Swedish twin registry and concluded that genetic factors were responsible for 38\% of scoliosis cases as compared to 62\% environmental association with the development of scoliosis.\textsuperscript{29} In fact the hemivertebrae stated in this study are not identical. There are many phenotypes which are identical in twins for many pathologies in spinal disorder, but this case does not seem to be one of them.

To our knowledge, this is the first report on the surgical treatment with PVCR, almost simultaneously, in both identical twins who had similar congenital vertebral anomalies causing kyphoscoliosis. Both identical twins with congenital kyphoscoliosis had undergone surgical correction by PVCR, anterior support with a mesh cage, and posterior fusion using pedicle screws at the age of 14 years and achieved a satisfactory correction and a stable spine without curve progression with 10-year follow-up.

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