Medicine®

CLINICAL CASE REPORT

OPEN

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-Il Suk, MD

Abstract: The etiology of congenital scoliosis and its development remains unclear and has not yet been fully identified, even there are theories that 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. We reported a case of long-term follow-up after posterior vertebral column resection (PVCR) in both identical twins with similar congenital kyphoscoliosis at thoracolumbar levels. Twin I had been noticed by his parents to have asymmetry of his back at age 5 years, but no treatment was given. Twin II was first noticed to have a spinal problem at 11 years of age by his parents. Overtime, spine of both twins 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.

(*Medicine* 95(17):e3499)

Abbreviation: PVCR = posterior vertebral column resection.

Editor: Girish Bhatt.
Received: November 17, 2015; revised: March 31, 2016; accepted: April 4, 2016.
From the Department of Orthopaedic Surgery (D-GC, J-HK, S-IS), Sanggye Paik Hospital, College of Medicine, The Inje University; Department of Orthopaedic Surgery (J-HY, S-WS), Korea University Guro-Hospital, College of Medicine, The Korea University; Department of Orthopaedic Surgery (J-HL), Kyung Hee Hospital, College of Medicine, The Kyung Hee University; and Department of Orthopaedic Surgery (Y-HK, K-YH), Seoul St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Seoul, Korea.
Correspondence: Se-Il Suk, Department of Orthopaedic Surgery, Inje University Sanggye Paik Hospital, Sanggye-Dong, Nowon-Ku, Seoul, Korea (e-mail: seilsuk@unitel.co.kr).
D-GC and J-HY equally contributed to this study.
The authors have no funding and conflicts of interest to disclose.Institutional Review Board approval of Inje University Sanggye Paik Hospital (IRB No. 2015-02-019) was obtained before data collection and analysis.

ISSN: 0025-7974
DOI: 10.1097/MD.0000000000003499

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.[1–11] Developmental imbalance of the vertebral anomalies during the growth may cause various types of kyphosis, scoliosis, or kyphoscoliosis.[6–16]

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 by a private clinic physician after a radiograph had shown congenital anomalies in the thoracolumbar spine at 13 years of age. The height and weight of Twin I was 155 cm and 45 kg. Trunk shift to right was 15 mm and left thoracolumbar hump 40 mm.

Twin II was first noticed to have a spinal problem at 11 years of age by his parents. Overtime, his spine also became further deviated to the left with a severe kyphotic deformity, and he was referred to our hospital with twin I at 13 years of age. The height and weight was 158 cm and 55 kg, respectively. Trunk shift to right was 10 mm and left thoracolumbar hump was 28 mm.

On the clinical examination, both twins were noted to have left thoracolumbar scoliosis and severe kyphosis. Examinations were otherwise within normal limits. Neurological examination and magnetic resonance imaging (MRI) were unremarkable. Both twins had normal electrocardiography (EKG), normal heart function without cardiac anomaly in echocardiogram, and normal pulmonary function (Twin I: FVC: 3.19 L, FEV1: 2.92 L, FEV1/FVC = 86%, Twin II: FVC: 3.31 L, FEV1: 3.03 L, FEV1/FVC: 86%) in spirometry before surgery.

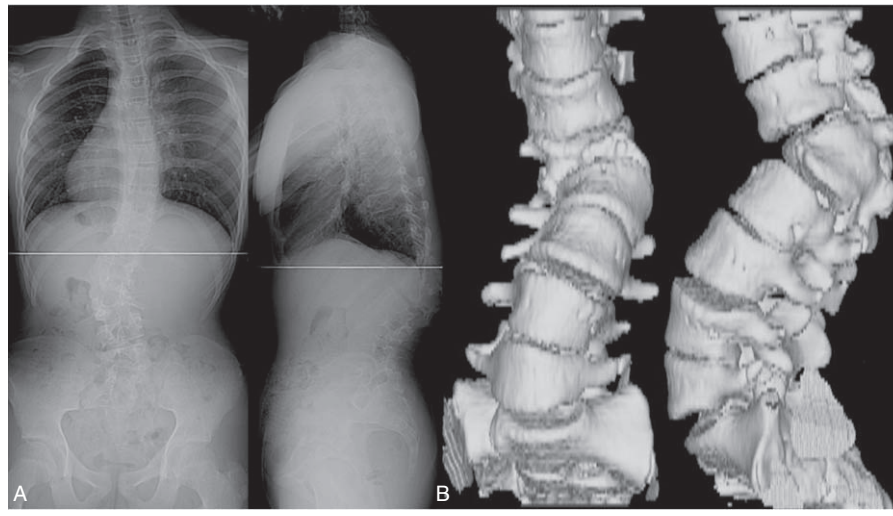

FIGURE 1. Twin I at age 14 years. (A) Twin I had congenital hemivertebra of T12 and L1 with 50° of left thoraco-lumbar scoliosis with segmental kyphosis of 52°. (B) 3D reconstruction CT (computed tomography) scan. CT = computed tomography.

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, hemivertbra 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).

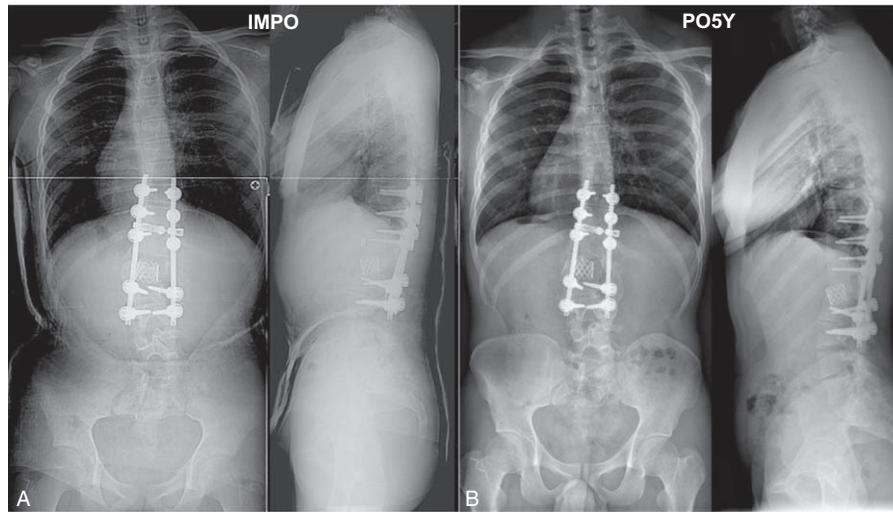

FIGURE 2. Twin I radiographs. (A) Immediate postoperative radiographs showed that the main curve of scoliosis improved to 13° and segmental angle of kyphosis to 12° with satisfactory deformity correction. (B) Five-year follow-up radiographs showed that main curve of scoliosis maintained to 17° and segmental angle of kyphosis to 14° without curve progression.

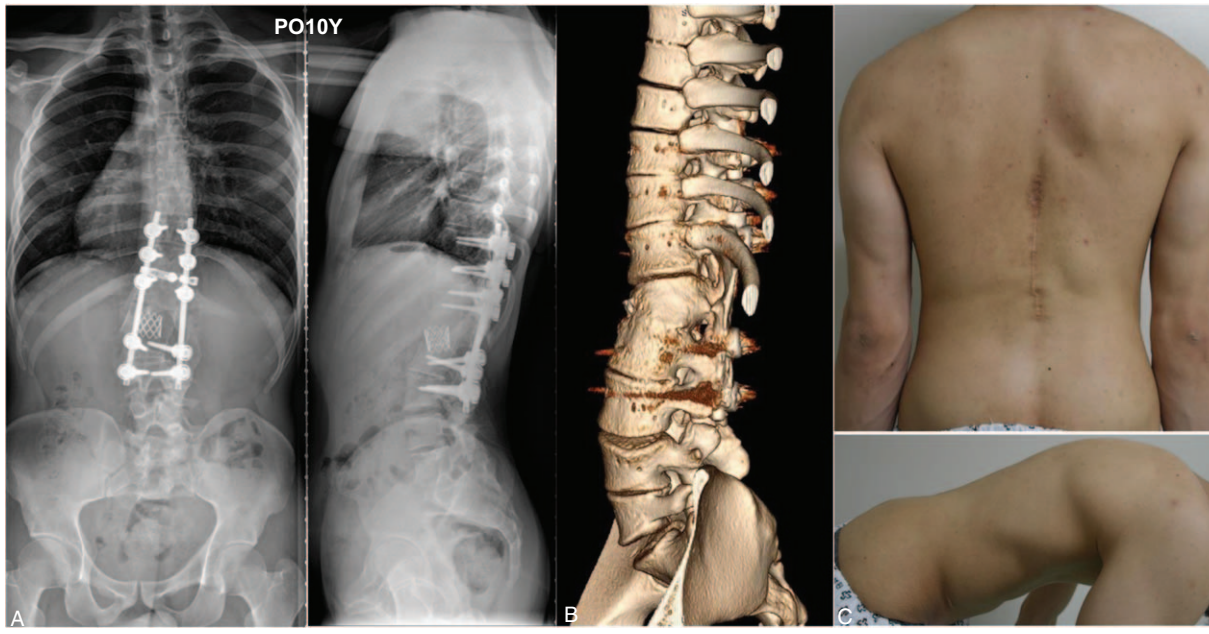

FIGURE 3. Twin I radiographs and clinical photographs. (A) Ten-year follow-up radiographs showed that the main curve of scoliosis well maintained to 18° and segmental angle of kyphosis to 10° without curve progression. (B) Ten-year follow-up 3D CT. (C) Ten-year follow-up clinical photographs. CT = computed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

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.

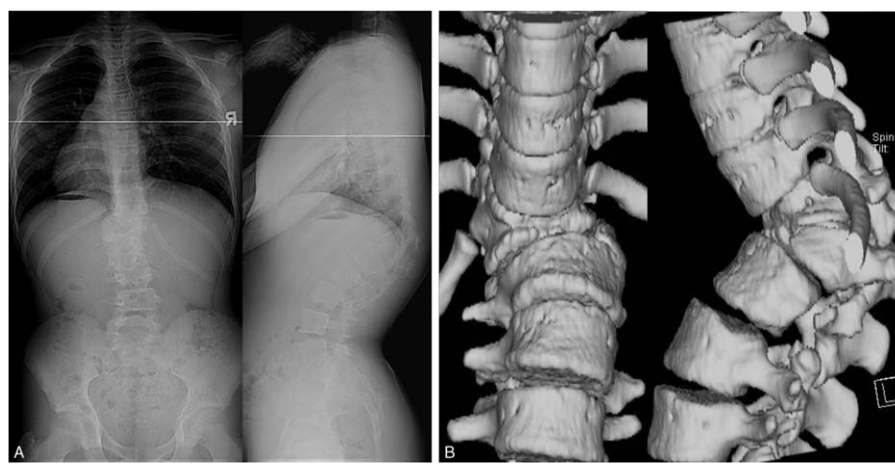

FIGURE 4. Twin II at age 14 years. (A) Twin II had congenital hemivertebra of T11 and T12 with 28° of left thoraco-lumbar scoliosis with severe segmental kyphosis of 86°. (B) 3D reconstruction CT scan. CT = computed tomography.

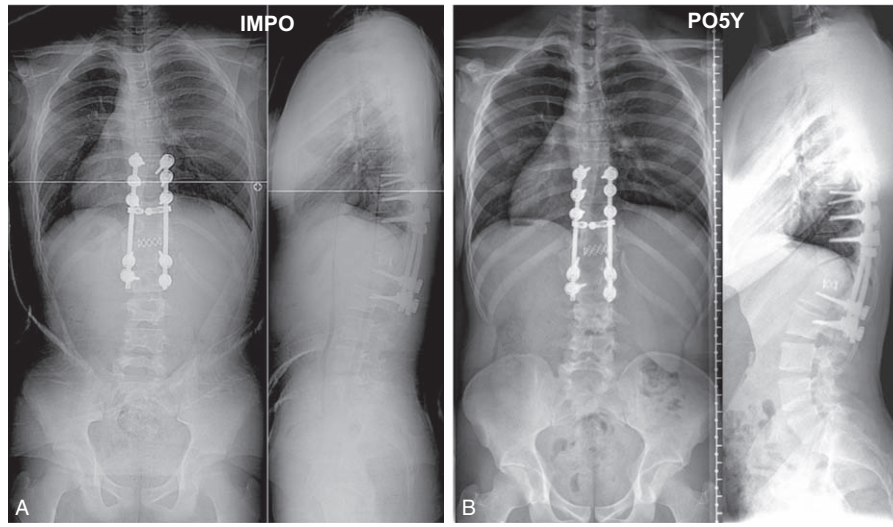

FIGURE 5. Twin II radiographs. (A) Immediate postoperative radiographs showed that the main curve of scoliosis improved to 6° and segmental angle of kyphosis to 35° with satisfactory results. (B) Five-year follow-up radiographs showed that main curve of scoliosis was to 7° and segmental angle of kyphosis to 33° and well maintained during follow-up period without curve progression.

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.[26]

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.[24–26] 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.[26,27]

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.[27]

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.[24,25] There are 2 different main principles of surgery: (1) prophylactic

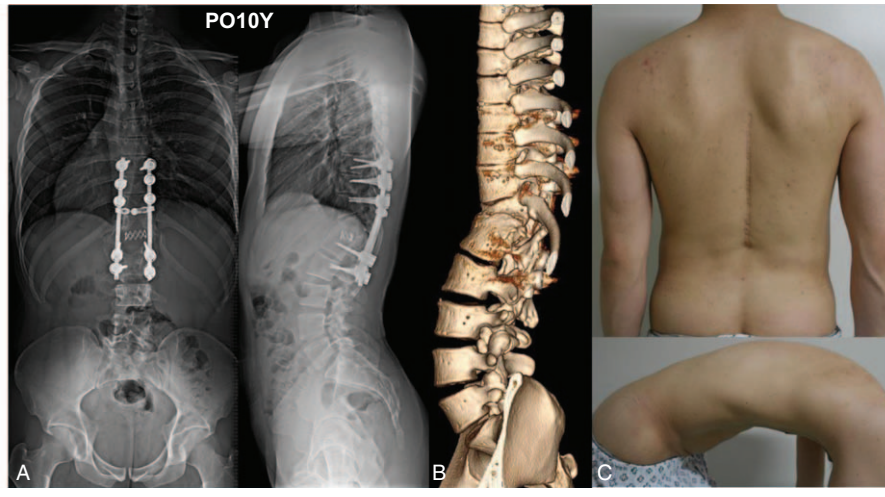

FIGURE 6. Twin II radiographs and clinical photographs. (A) Ten-year follow-up radiographs showed that the main curve of scoliosis well maintained to 5° and segmental angle of kyphosis to 35° without curve progression. (B) Ten-year follow-up 3D CT. (C) Ten-year follow-up clinical photographs. CT = computed tomography.

surgeries and (2) corrective surgeries.[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.[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.[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.[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.[11] However, little information exists regarding the relative contribution of each of factors to the development of congenital scoliosis.[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. Grauers 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.[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.

ACKNOWLEDGMENTS

The authors thank Dr P William Haake for English-language editing and Eui-Jong Lee for data collection of this paper.

REFERENCES

1. McMaster MJ, Ohtsuka K. The natural history of congenital scoliosis. A study of two hundred fifty-one patients. *J Bone Joint Surg Am.* 1982;64:1128–1147.
2. Giampietro PF, Raggio CL, Blank RD, et al. Clinical, genetic and environmental factors associated with congenital vertebral malformations. *Mol Syndromol.* 2013;4:94–105.
3. Zeng Y, Chen Z, Qi Q, et al. Posterior surgical correction of congenital kyphosis and kyphoscoliosis: 23 cases with minimum 2 years follow-up. *Eur Spine J.* 2013;22:372–378.
4. Chang DG, Kim JH, Ha KY, et al. Posterior hemivertebra resection and short segment fusion with pedicle screw fixation for congenital scoliosis in children younger than 10 years. *Spine.* 2015;40: E484–E491.
5. Chang DG, Suk SI, Kim JH, et al. Surgical outcomes by the age at the time of surgery in the treatment of congenital scoliosis in children under 10 years. *Spine J.* 2015;15:1783–1795.
6. Goldstein I, Makhoul IR, Weissman A, et al. Hemivertebra: prenatal diagnosis, incidence and characteristics. *Fetal Diagn Ther.* 2005;20:121–126.
7. Ruf M, Jensen R, Letko L, et al. Hemivertebra resection and osteotomies in congenital spine deformity. *Spine.* 2009;34: 1791–1799.
8. Pool RD. Congenital scoliosis in monozygotic twins. Genetically determined or acquired in utero? *J Bone Joint Surg Br.* 1986;68: 194–196.
9. Sturm PF, Chung R, Bomze SR. Hemivertebra in monozygotic twins. *Spine.* 2001;26:1389–1391.
10. Akbarnia BA, Heydarian K, Ganjavian MS. Concordant congenital spine deformity in monozygotic twins. *J Pediatr Orthop.* 1983;3:502–504.
11. Bonicoli F, Delvecchio E. Scoliosis in monochorial twins (pathogenetic considerations). *Chir Organi Mov.* 1968;57:178–186.
12. Brewer C, Holloway S, Zawalnyski P, et al. A chromosomal deletion map of human malformations. *Am J Hum Genet.* 1998;63: 1153–1159.
13. Giampietro PF, Blank RD, Raggio CL, et al. Congenital and idiopathic scoliosis: clinical and genetic aspects. *Clin Med Res.* 2003;1:125–136.
14. Tredwell SJ, Smith DF, Macleod PJ, et al. Cervical spine anomalies in fetal alcohol syndrome. *Spine.* 1982;7:331–334.
15. Bantz EW. Valproic acid and congenital malformations. A case report. *Clin Pediatr (Phila).* 1984;23:352–353.
16. Kaspiris A, Grivas TB, Weiss HR. Congenital scoliosis in monozygotic twins: case report and review of possible factors contributing to its development. *Scoliosis.* 2008;3:17.
17. Chang DG, Yang JH, Lee JH, et al. Congenital scoliosis with posterior vertebral column resection in patients younger than 18 years: longer than 10-year follow-up. *J Neurosurg Spine.* 2016:1–9[Epub ahead of print].
18. Suk SI, Chung ER, Lee SM, et al. Posterior vertebral column resection in fixed lumbosacral deformity. *Spine.* 2005;30: E703–E710.
19. Suk SI, Chung ER, Kim JH, et al. Posterior vertebral column resection for severe rigid scoliosis. *Spine.* 2005;30:1682–1687.
20. Ruf M, Harms J. Pedicle screws in 1- and 2-year-old children: technique, complications, and effect on further growth. *Spine.* 2002;27:E460–E466.
21. Ruf M, Harms J. Hemivertebra resection by a posterior approach: innovative operative technique and first results. *Spine.* 2002;27:1116–1123.
22. Winter RB. Congenital scoliosis. *Orthop Clin North Am.* 1988;19:395–408.
23. Winter RB, Moe JH, Lonstein JE. Posterior spinal arthrodesis for congenital scoliosis. An analysis of the cases of two hundred and

ninety patients, five to nineteen years old. *J Bone Joint Surg Am.* 1984;66:1188–1197.

24. Hedequist D, Emans J. Congenital scoliosis: a review and update. *J Pediatr Orthop.* 2007;27:106–116.
25. Modi HN, Suh SW, Fernandez H, et al. Accuracy and safety of pedicle screw placement in neuromuscular scoliosis with free-hand technique. *Eur Spine J.* 2008;17:1686–1696.
26. Arlet V, Odent T, Aebi M. Congenital scoliosis. *Eur Spine J.* 2003;12:456–463.
27. Hedequist D, Emans J. Congenital scoliosis. *J Am Acad Surg.* 2004;12:266–275.
28. Lee SM, Suk SI, Chung ER, et al. Direct vertebral rotation: a new technique of three-dimensional deformity correction with segmental pedicle screw fixation in adolescent idiopathic scoliosis. *Spine.* 2004;29:343–349.
29. Grauers A, Rahman I, Gerdhem P, et al. Heritability of scoliosis. *Eur Spine J.* 2012;21:1069–1074.