Convex Short Segment Instrumentation and Hemi-Chevron Osteotomies for Putti Type 1 Thoracic Hemivertebrae

A Simple Treatment Option for Patients Under 5 Years Old

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Study Design: A case series depicting the results of a novel surgical technique.

Objective: To prove that a minimally invasive surgical technique can effectively control and even correct congenital scoliosis caused by a fully segmented hemivertebra.

Summary of Background Data: Congenital hemivertebrae have been treated by anterior and posterior growth arrest with/without fusion, anterior and posterior hemivertebrectomy, transpedicular hemivertebra excision, and transpedicular hemiepiphysiodesis. These approaches are complex and require experience. There is a need for a simple treatment method to treat these deformities.

Methods: Twelve patients under 5 years of age with Putti type1 hemivertebrae were treated by posterior convex short segment instrumentation, partial chevron osteotomies, and fusion. Scoliosis, segmental scoliosis, kyphosis, segmental kyphosis, trunk shift were measured both preoperatively and postoperatively.

Results: The mean correction of the segmental curve was 6 degrees (21%) which was maintained at the latest follow-up. The average final correction of the main curve was 23%. The trunk shift was 1.8 cm (range, 1–3 cm) preoperative and 1.4 cm (range, 0–2.5 cm) at the latest follow-up. The segmental angle of kyphosis averaged 11 degrees (range, −12 to 20 degrees) preoperative, and 14 degrees (range, 0–29 degrees) at the latest follow-up assessment. The values of the total thoracic kyphosis (T2–T12) were 29.5 degrees (range, 10–46 degrees) preoperative, 31 degrees (range, 10–44 degrees) postoperative, and 32 degrees (range, 16–45 degrees) at the last follow-up resulting in a mean improvement of 2 degrees. This improvement continued at the latest follow-up with a mean increase of 3 degrees.

Conclusions: Transpedicular instrumentation is ideal for early correction in young children. The new posterior approach is much less invasive than the combined approaches or other posterior vertebrectomies and is well tolerated even in very young patients. The fusion segment is kept short. The deformities seem to stop progressing and this can avoid development of severe local deformities and secondary curves.

Key Words: hemivertebra, transpedicular instrumentation, chevron osteotomy, congenital scoliosis

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Congenital spine deformities causing scoliosis may be classified into failures of formation, failures of segmentation, or mixed deformities. The most common anomaly caused by failure of formation is a hemivertebra.[1–4] In 1910 Putti[5] described 3 types of hemivertebra: (1) the full segmented, (2) the semisegmented, (3) the nonsegmented. A hemivertebra (fully segmented one) creates a wedge-shaped deformity which progresses during spinal growth. The rate of progression and the ultimate curve angle depends on the type of anomaly and the site at which it occurs. Because of the poor natural history of hemivertebrae, surgical treatment is required in most cases.

The indications for surgical treatment of hemivertebrae are not clearly definable. However, the documented progression of a curve caused by an isolated hemivertebra that will lead to truncal imbalance is an indication for surgical treatment.

Numerous surgical strategies for the treatment of congenital scoliosis have been developed. Spinal surgeons have successfully treated congenital hemivertebrae using anterior and posterior growth arrest with/without concomitant fusion, anterior and posterior hemivertebra

TABLE 1. Patient Data

Patient No.	Age at Surgery	Sex	Abnormality	Follow-up	Operation Time (min)	Blood Loss
1	2.5 y	M	T8 L	18	30	10
2	5 y	F	T10 R	16	35	110
3	9 mo	F	T8 L	40	55	15
4	3 y	M	T9 L	12	45	60
5	4.5 y	F	T7 R	14	35	30
6	4 y	F	T8 L	24	40	20
7	2 y	F	T7 L	30	30	20
8	4 y	F	T10R	32	35	25
9	3 y	F	T6 L	38	45	30
10	2.5 y	M	T7 L	44	35	50
11	5 y	M	T9 R	27	30	65
12	2 y	F	T8 L	34	40	45

F indicates female; M, male.

excision, transpedicular hemivertebra excision, and transpedicular hemiepiphysiodesis,[5–15] which stopped or even reversed the deformity associated with congenital scoliosis, provided there is proper adherence to the prerequisites.

However, these approaches require experience and their learning curve is shallow. New methods of treatment like growing rod instrumentations and expansion thoracoplasty were developed to address some of these patients but these require expensive hardware and repeated surgeries.[6–8] There is a need for a simple treatment method which may stop (if not correct) the deformities of these patients.

The purpose of this study is to evaluate the efficacy of posterior short segment instrumentation and hemichevron osteotomy in congenital scoliosis because of Putti type 1 thoracal hemivertebra in children under 5 years of age. This, to our knowledge is a new surgical technique which has not been reported before.

MATERIALS AND METHODS

We retrospectively reviewed 12 consecutive patients with Putti type 1 hemivertebra treated by posterior convex short segment instrumentation, partial chevron osteotomies, and posterior fusion under age 5 years (Table 1). A total of 12 patients, 4 males and 8 females, underwent operative treatment between 2006 and 2010. The average follow-up period was 2 years and 3 months (range, 12–44 mo). Average age of the patients at surgery was 3 years and 2 months (range, 9 mo to 5 y). All 12 patients had a single nonincarcerated hemivertebra and locations of the hemivertebra were T6 in 1 patient, T7 in 3, T8 in 4, T9 in 2, and T10 in 2 patients. Posterior instrumentation was used for correction of the deformity in all patients. We used a 3 mm rod with polyaxial posterior cervical lateral mass screws of 3.5 mm diameter in 5 patients whose ages were below 3 years and we used 5 mm rod with 4.0 mm pediatric posterior instrumentation screws in the remaining 7 (Tasarim Med, Istanbul, Turkey).

Of the 12 patients, 3 had genitourinary, 2 had auditory, 1 had central nervous system, and 3 had cardiopulmonary system anomalies. None of the patients had undergone a prior operation. None had a neurologic deficit or cord anomaly. Surgery was indicated by proved or expected deterioration of the deformity. Deterioration was proved in 4 cases by radiographs taken more than 6 months before surgery.

Before surgery, a complete neurologic evaluation and a survey for coexisting congenital anomalies were performed. Radiographic evaluations were conducted on the preoperative, postoperative, and follow-up standing posteroanterior and lateral radiographs. Magnitude of the frontal (scoliosis) and sagittal (kyphosis) deformities were measured with the Cobb method. Operative reports were reviewed to determine the presence of any intraoperative complications. Medical records were reviewed to identify any complications in the perioperative and follow-up periods.

Measurements were taken from standing long cassette anterior-posterior and lateral radiographs (Figs. 1, 2). On the coronal plane, the segmental scoliosis was measured from the upper endplate of the vertebra above the hemivertebra to the lower endplate of the vertebra below the hemivertebra, and the whole curve Cobb angle was also measured and recorded. On the sagittal plane, the segmental kyphosis was measured in the same way as was the segmental scoliosis on the coronal plane. Thoracic kyphosis (T5–T12) was also measured and recorded. Trunk shift is defined as the perpendicular distance (mm) from the sacrum center to the plumb line drawn from the midpoint of the C7 vertebral body.

Operative Technique

The patient is positioned prone on the Relton-Hall 4 poster frame with the abdomen relieved of pressure. A standard midline incision was made, and subperiosteal dissection was performed only at the convex side to expose the hemivertebra and the vertebrae just above and below, including the lamina, transverse processes, and facet joints. Unilateral pedicle screws were inserted into the vertebra above, the hemivertebra, and the vertebra below using the classical freehand technique. Fluoroscopy was used to confirm the hemivertebra and the positions of the screws. Then a hemi-chevron osteotomy was done

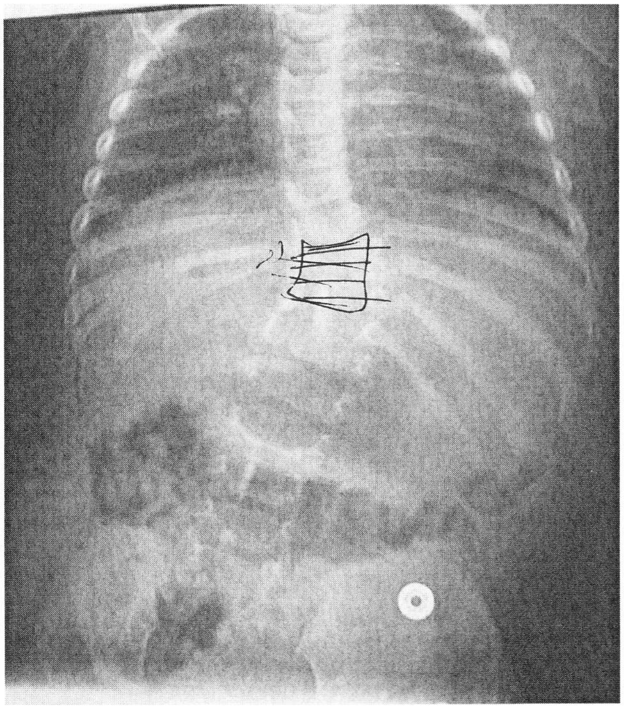

FIGURE 1. Anteroposterior x-ray of a 9-month-old female patient with a T8 hemivertebra and contralateral unsegmented bar. Segmental Cobb is 34 and total Cobb angle is 40 degrees. The patient was diagnosed as Jarcho Levin syndrome.

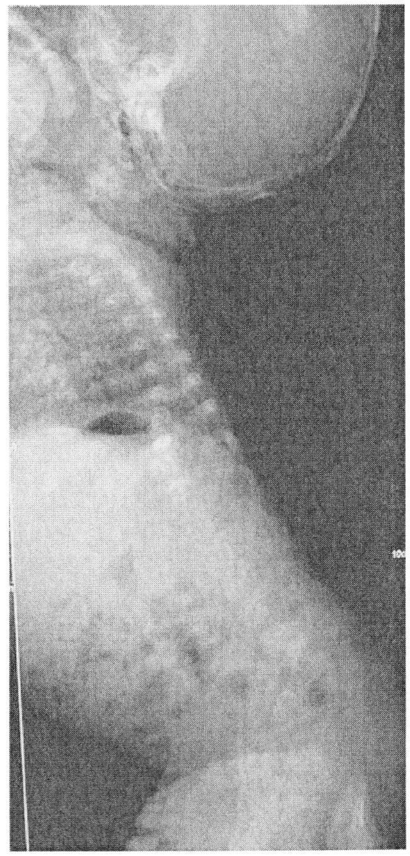

FIGURE 2. Lateral x-ray of the same patient. There is thoracic segmental lordosis (12 degrees). Total thoracic kyphosis is 12 degrees.

above and below the hemivertebra. First, inferior half of the lamina above was excised including the inferior articular process, and then, the superior articular process of the hemivertebra was excised. This was followed by excision of the inferior articular process of the hemivertebra and the superior articular process of the vertebra below. Then a rod was placed and the system was locked under compression by a compressing device. This procedure helped reduce the angle of the curve. Then the laminae and the spinous processes of the instrumented levels were decorticated and the bones retrieved during osteotomy were used as graft material for fusion of the posterior elements. Anteroposterior and lateral radiographs are obtained to check correction of the deformity (Figs. 3, 4). Spinal cord monitoring is performed throughout the procedure to check neurologic status. No patient had an intraoperative wake-up test. All patients were allowed to walk the day after surgery. Five patients under age 3 wore a brace for 1 month and 1 patient (9 mo of age) was put in a pelvipedal cast again for a month. Six patients did not wear any braces (Figs. 5–8).

RESULTS

The intraoperative blood loss averaged 40 mL (range, 10–110 mL), and the operating time averaged 38 minutes (range, 30–55 min). No intraoperative complications were noticed. There were no neurologic complications. There was 1 superficial wound infection which healed with regular dressing changes and antibiotic treatment. None needed implant removal and no implant failure was found at the final radiographic evaluations. There were 2 pedicle fractures. No rod migration, fracture, or correction loss was noted in the following observation.

Correction of Main Curve and Trunk Shift

The segmental Cobb (Fig. 1) averaged of 31 degrees before surgery (range, 17–35 degrees). After surgery, the mean angle measured 25 degrees (range, 14–30 degrees) (Fig. 3). At the last follow-up assessment, the mean angle was 25 degrees (range, 20–32 degrees). Thus, the mean correction of the segmental curve was 6 degrees (21%) and this was maintained at the latest follow-up.

In terms of the total main curve, the following results were obtained. The mean Cobb angle was 26 degrees (range, 18–40 degrees) before surgery, 19 degrees (range, 17–22 degrees) after surgery, and 20 degrees (range, 14–24 degrees) at the latest follow-up assessment. The average correction was 6 degrees (26.07%) after surgery. At the latest follow-up there was 4% (1 degree) loss of correction and the average final correction when compared with preoperative Cobb was 23% (Table 2).

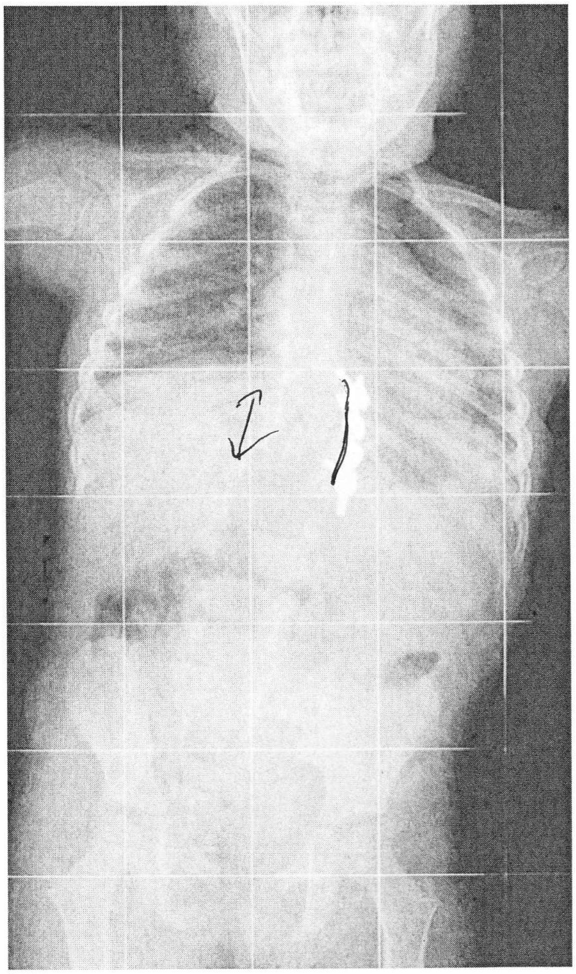

FIGURE 3. Anteroposterior x-ray of the same patient after 40 months follow-up. Segmental Cobb is 20 degrees and the total Cobb angle is 14 degrees.

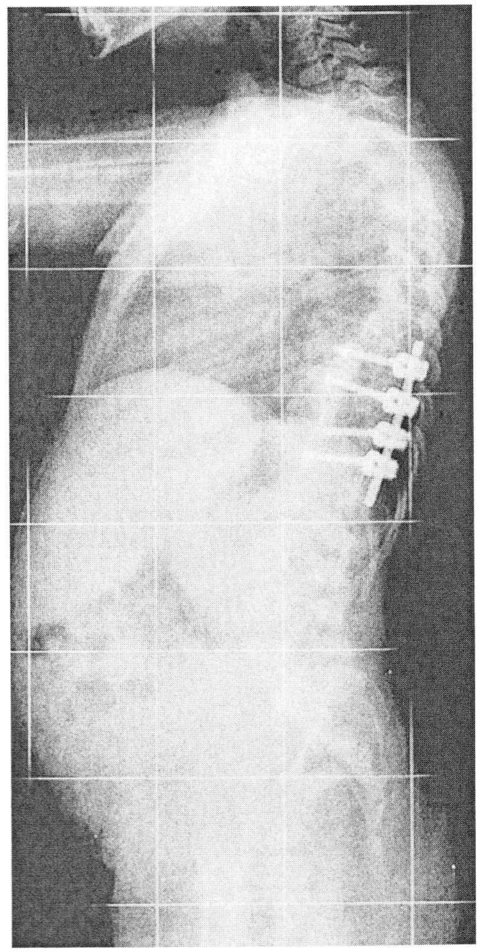

FIGURE 4. Lateral x-ray of the same patient showing good sagittal alignment of the spine. There is 0 degree segmental kyphosis and 24 degrees total kyphosis.

The trunk shift was 1.8 cm (range, 1–3 cm) before surgery, 1.4 cm (range, 0–2.5 cm) after surgery, and 1.4 cm (range, 0–2.5 cm) at the latest follow-up assessment.

Correction of the Sagittal Plane

The segmental angle of kyphosis averaged 11 degrees (range, −12 to 20 degrees) before surgery, 13 degrees (range, 5–28 degrees) after surgery, and 14 degrees (range, 0–29 degrees) at the latest follow-up assessment.

The values of the total thoracic kyphosis (T2–T12) were 29.5 degrees (range, 10–46 degrees) before surgery, 31 degrees (range, 10–44 degrees) after surgery, and 32 degrees (range, 16–45 degrees) at the last follow-up assessment resulting in a mean improvement of 2 degrees postoperatively and this improvement continued at the latest follow-up with a mean increase of 3 degrees.

DISCUSSION

The natural history of hemivertebrae are not fully understood. Some studies outlined the natural history of congenital scoliosis and demonstrated that progressive congenital scoliosis requires surgery.[1–4] Winter and Lonstein[9] reported that spontaneous curve improvement is rare, but possible and recommended careful observation with careful measurement, warning against inappropriate early, aggressive surgery. Surgery was recommended before the age of 10 because the untreated congenital curve could lead to a larger globally stiff curve by adolescence which could require longer fusion segments. In our series, the whole patient population was under 5 years of age.

In the past, several studies reported the results of different surgical techniques for the treatment of hemivertebrae. Among these are growth modulation (stapling, rib procedures, anterior and posterior convex hemiepiphysiodesis, growing rod, vertical expandable prosthetic titanium rib) and hemivertebra excision (anterior, posterior, combined).

As the great majority of congenital spinal deformities are progressive and the rigidity of the curves poses difficulty during correction, spine surgeons have generally

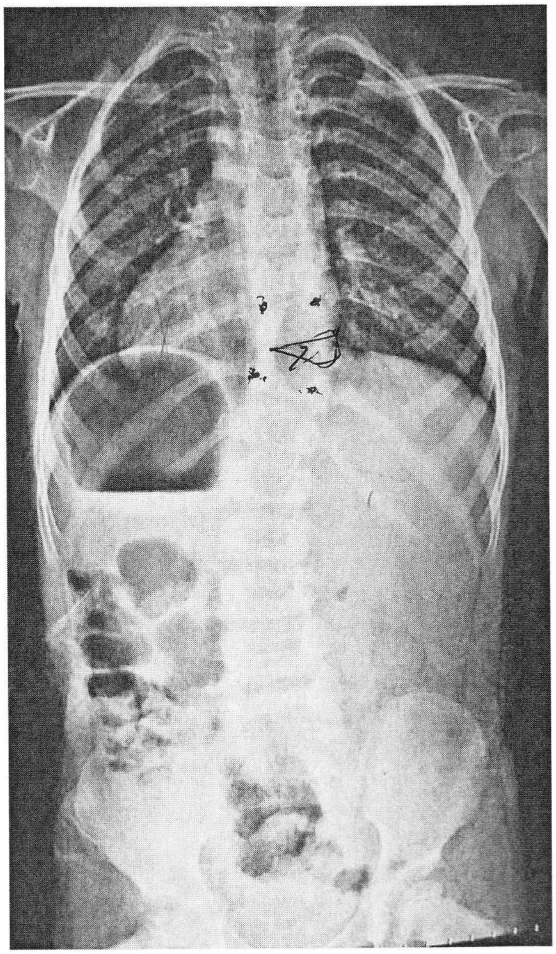

FIGURE 5. Anteroposterior x-ray of a 5-year-old female patient with a T10 hemivertebra. Segmental Cobb is 32 degrees and total Cobb angle is 20 degrees.

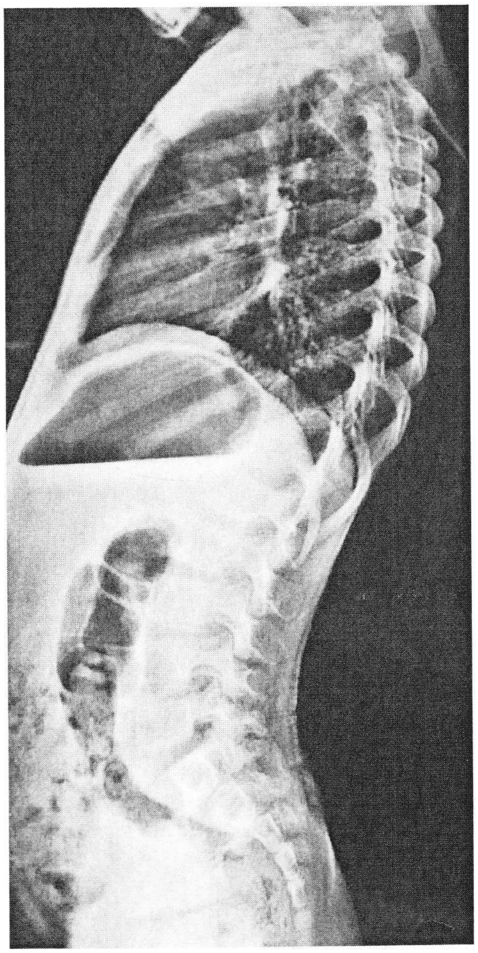

FIGURE 6. Lateral x-ray of the same patient. With 20 degrees segmental and 44 degrees total kyphosis.

preferred to stop the deformity before it becomes severe. In congenital spinal deformities the growth potential is asymmetric, due either to formation or segmentation defects. The control of the growth of the convex side, which is relatively longer than the concavity, not only stops progression but also leads to spontaneous correction. The career growth arrest (CGA) procedure based on this concept has been popularized because of its safety, efficacy, and simplicity compared with other surgical alternatives.[11–13,16–19] Several studies have documented an epiphysiodesis effect in the majority of patients with control of deformity, whereas the correction effect is observed less frequently. The procedure is easily performed, with a known potential of biologic correction with time. The reported epiphysiodesis effect ranges from 20% to 77%, fusion effect from 17% to 70%, and progression between 0% and 21%. In a series, 32 patients with congenital scoliosis underwent convex anterior and posterior hemiepiphysiodesis at an average age of 29 months and were followed for a minimum of 2 years. Their patients had no major or minor neurologic complications. However, in 3 patients, weaning of the chest tube was prolonged; 2 other patients had atelectasis, which was treated conservatively; and in 1 patient pneumonia developed. Overall, 41% of the patients had epiphysiodesis, 47% had fusion effect, and 12% of patients deteriorated. An 88% success rate was comparable with that of the aforementioned studies on CGA. The behavior of the curve after the procedure is unpredictable. Therefore, some questions need to be answered, such as why some patients responded well to the treatment and others did not. To delineate this issue, all the well-defined parameters such as curve localization, length, magnitude, patient age, and so forth should be analyzed in detail. However, the limited number of patients treated with CGA in the literature precludes drawing definitive conclusions easily.[15,16] In our series, as the procedure was an posterior-only surgery, there were no problems regarding the lung functions.

Unilateral epiphysiodesis was first described by Roaf in 1963, when 35 patients with a mean age of 7.8 years at surgery were treated. An average correction of only 13.6 degrees was obtained.[10] Hemiepiphysiodesis has

Epiphysiodesis - surgical procedure where growth plate is fused

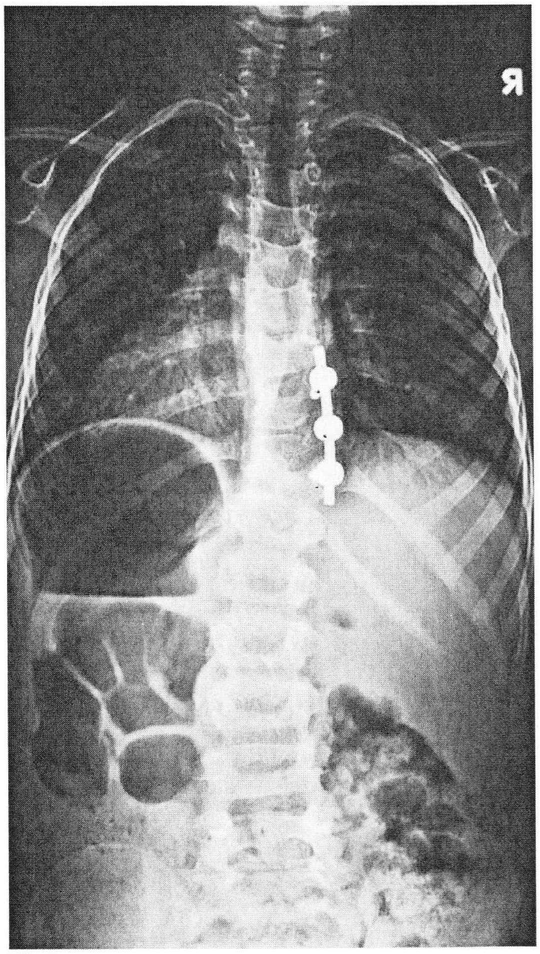

FIGURE 7. Anteroposterior x-ray of the same patient after 16 months follow-up. Segmental Cobb is 30 degrees and the total Cobb angle is 18 degrees.

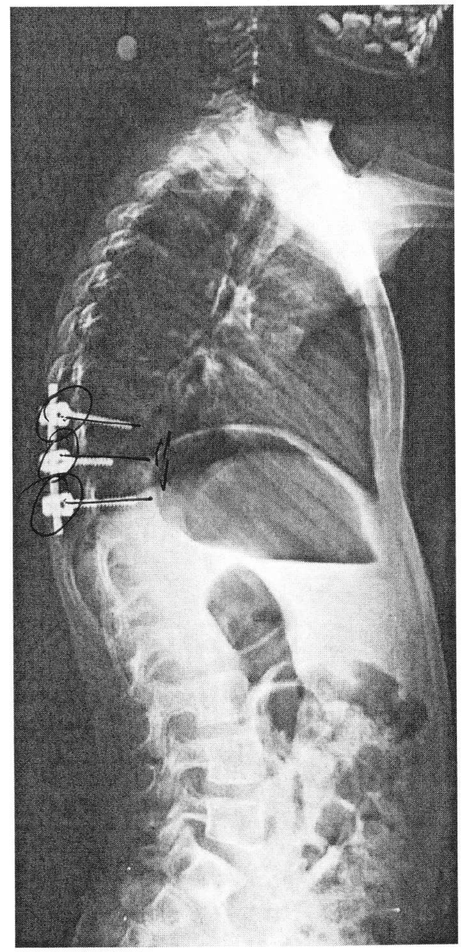

FIGURE 8. Lateral x-ray of the same patient showing good sagittal alignment of the spine with a 14 degrees segmental and 30 degrees total kyphosis.

then been studied by several authors. Good results have been reported after open anterior and posterior hemiepiphysiodesis and fusion.[11,12] Winter[11] demonstrated success with an anterior and posterior convex hemiepiphysiodesis and short segment fusion using a separate transthoracic or retroperitoneal approach to perform the anterior portion of the procedure. Keller et al[13] and King et al[14] described a limited posterior approach and a transpedicular technique to perform the hemiepiphysiodesis. Their curve improvement/arrest rates were 79% and 100%, respectively. The best results of epiphysiodesis have been obtained in patients younger than 5 years with a single lumbar hemivertebra.[7–10] The main disadvantage of this technique is that unilateral epiphysiodesis relies on growth potential on the contralateral side, which can be variable. Moreover, the entire convex hemivertebra from anterior to posterior must undergo arrest to avoid deformities in the sagittal plane. If the arrest is not complete, continued growth of the vertebrae may cause a bending of the posterior fusion mass and a worsening rotational deformity.[7] We believe that, using a transpedicular screw system which anchors into the anterior structures, we have caused growth arrest of both the anterior structures and the posterior structures. In our series, the deformities of 2 out of 12 patients (16.6%) have progressed.

Theoretically, hemivertebra resection can remove the pathology and correct the deformity completely. Shono et al[20] reported on hemivertebra resection through a single posterior approach with a mean correction rate of 63.3% in 12 patients. Nakamura et al[21] reported 5 cases of posterior hemivertebra resection with an improvement of 55.1% for 3 thoracolumbar hemivertebrae, 31.4% for 2 lumbosacral hemivertebrae on the coronal plane, and 67.4% for regional kyphosis. Ruf and Harms reported hemivertebra resection by the posterior-only procedure. In 2 of their studies, the reported coronal plane correction was between 63.4% and 80.5%. In patients without bar formation, there was a correction rate of 80.5% of the main curve and 63.6% of the kyphosis. In patients with bar formation, the improvement of the main curve was 66.7%, and the kyphosis improvement was 62.5%. The

TABLE 2. Cobb Angle Measurements (Degrees) and Trunk Shift (cm)

	Segmental Cobb			Total Cobb			Segmental Kyphosis			Total Kyphosis			Trunk Shift (cm)		
No.	Preop	Postop	FF	Preop	Postop	FF	Preop	Postop	FF	Preop	Postop	FF	Preop	Postop	FF
1	28	28	22	20	20	18	16	28	16	35	44	45	2	2	2
2	32	28	30	20	20	18	20	14	14	44	40	30	1	1	1
3	34	20	20	40	18	14	−12	5	0	12	15	24	2	0	0
4	17	14	22	18	18	20	11	12	16	29	33	35	1	1.5	1
5	34	28	30	19	17	23	21	16	19	38	40	33	2.5	2	2
6	30	22	22	24	18	19	14	12	5	19	18	26	2	1.5	2
7	32	30	24	23	20	19	20	20	29	41	39	45	1.5	1	1
8	32	26	28	20	18	18	10	14	26	46	40	35	2.5	2	2
9	35	22	24	30	19	22	−8	0	9	10	10	16	2	2	2
10	28	28	22	24	17	18	19	12	18	29	33	36	1	0	0
11	35	30	32	33	20	24	20	16	19	40	40	33	3	2.5	2.5
12	34	20	20	36	22	24	0	5	0	11	19	23	1.5	1	1

FF indicates final follow-up; preop, preoperative; postop, postoperative.

complications included 3 implant failures, 3 convex pedicle breakages, and 2 progressed deformities after the surgery. Revision surgeries were performed on these cases. Their study published in 2009 showed a tendency of better correction and shorter fusion when earlier surgical intervention was performed.[22,23] There had been no adverse effect of pedicle screws concerning further growth in their series.

The procedures to remove the hemivertebra completely, are lengthy operations (averaging 4.5 h) which also cause significant blood loss (average 600 mL). Also these highly demanding techniques may cause neurological complications. The common neurological complications were transient muscle weakness of the lower limb with a rate of 9.1%–20.5%. Pseudarthrosis, wound infection, loosening of internal fixation, implant bulging were also reported complications of these procedures. In our study we did not have any neurological complications. In combined approaches, complications related to the anterior procedure like pneumothorax, pleural effusion requiring drainage, and abdominal hernia were also reported.[20–24]

To prevent screw-related complications, selection of the proper instrumentation system is important. In most cases a pediatric spinal instrumentation system with 4.5 mm rods and in some very young children, a cervical lateral mass screw system with 3.0 mm rods can be used. In our series, 2 of our patients had pedicle fractures both at the level of hemivertebra and in those patients a cervical lateral mass screw system comprising of 3.5 mm diameter screws and a 3 mm rod were used instead of 4.5 mm pediatric pedicle screws with a 5 mm rod. None of our patients had instrumentation failures. None were revised. This may be because of preserving spinal stability by not doing a corpectomy.

One of the shortcomings of our study is the relatively short duration of follow-up (2 y, 3 mo). As these patients are young and have further growth potential, studies with longer term follow-up are needed to draw definite conclusions.

CONCLUSIONS

The natural history of congenital scoliosis is still unknown. There is no treatment algorithm because of the variability of surgical solutions. The coronal and sagittal deformities, as well as the patient age and type and location of the anomaly should be taken into consideration. There is no study in the literature (regarding either CGA or vertebral resection) in which the study group is homogeneous in regard of age and type and location of the anomaly.

Transpedicular instrumentation is an ideal procedure for early correction in young children. The new posterior approach is much less invasive than the combined approaches or other posterior vertebrectomies described before and is well tolerated even in very young patients. The fusion segment can be kept short. The deformities seem to stop progressing and this can avoid development of severe local deformities and secondary curves. With this technique, full excision of the hemivertebrae may not be required to treat these patients.

REFERENCES

1. McMaster MJ, Ohtsuka K. The natural history of congenital scoliosis. A study of two hundred and fifty-one patients. *J Bone Joint Surg Am*. 1982;64:1128–1147.
2. Nasca RJ, Stelling FH, Steel HH. Progression of congenital scoliosis due to hemivertebrae and hemivertebrae with bars. *J Bone Joint Surg Am*. 1975;57:456–466.
3. Winter RB, Moe JH, Eilers VE. Congenital scoliosis: a study of 234 patients treated and untreated. Part I. Natural history. *J Bone Joint Surg Am*. 1968;50:1–15.
4. Winter RB, Moe JH, Eilers VE. Congenital scoliosis: a study of 234 patients treated and untreated. Part II. Treatment. *J Bone Joint Surg Am*. 1968;50:15–47.
5. Putti V. Die angeborenen deformintaten der wirbelsaule. *Fortschr Geb Rontgenstr*. 1910;15:65–76.
6. Akbarnia BA, Marks DS, Boachie-Adjei O, et al. Dual growing rod technique for the treatment of progressive early-onset scoliosis: a multicenter study. *Spine (Phila Pa 1976)*. 2005;30(suppl 17):S46–S57.
7. Elsebai HB, Yazici M, Thompson GH, et al. Safety and efficacy of growing rod technique for pediatric congenital spinal deformities. *J Pediatr Orthop*. 2011;31:1–5.