

Clinical Study

Evaluation of a combined approach to the correction of congenital cervical or cervicothoracic scoliosis

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Abstract

BACKGROUND CONTEXT: An anteroposterior combined approach has been used for the treatment of congenital cervical or cervicothoracic scoliosis. However, its outcomes and surgical risks have not been clarified.

PURPOSE: We analyzed the characteristics of congenital cervical and cervicothoracic scoliosis and evaluated the outcomes of an anteroposterior combined approach for its treatment.

STUDY DESIGN: We carried out a retrospective clinical study of prospectively collected data.

PATIENT SAMPLE: Sixteen patients were treated between 2009 and 2013. Their average age was 9.2 years.

OUTCOME MEASURES: Radiographic and surgical outcomes were measured and recorded. We compared morphological parameters and preoperative and postoperative results.

METHODS: All patients underwent surgery with a combined approach. The following radiographic parameters were measured: head tilt (HT), mandible incline (MI), shoulder balance (SB), structural and compensatory curves, cervical lordosis, C7 central sacral vertical line (C7–CSVL) ratio, C7 sagittal vertical axis (C7–SVA) ratio, C2–C7 SVA ratio, the angle between the upper endplate of the T2 vertebra and a horizontal line (T2 tilt), gravity line ratio. Demographic and surgical data were also collected.

RESULTS: On average, the duration of follow-up was 68.0 months, surgical blood loss was 675 mL, and the duration of surgery was 400.5 minutes. The average correction rate was 64.9% in the structural curve and 29.5% in the compensatory curve. Statistical analysis showed that MI significantly correlated with HT and SB ($p < .05$). The C7–CSVL ratio correlated with the HT, MI, and SB ($p < .05$). The C7–SVA ratio correlated with the structural curve and cervical lordosis ($p < .05$), and the gravity line ratio correlated with the structural and compensatory curve, cervical lordosis, and C7–SVA ratio ($p < .05$). Moreover, there were correlations between the structural and compensatory curves as well as between the structural curve and cervical lordosis ($p < .05$). There were significant differences before and after surgery in HT, MI, and structural and compensatory curves. Four patients developed nerve root palsy after surgical correction and totally recovered by 6 months of follow-up.

CONCLUSION: The combined approach is an effective surgical option for congenital cervical or cervicothoracic scoliosis. The resection of the hemivertebra cannot only improve head-neck aesthetic appearance but can also maintain the growth potential of the neck. © 2018 Published by Elsevier Inc.

Keywords:

Aesthetic evaluation; Combined approach; Congenital hemivertebral deformity; Cervical scoliosis; Head tilt; Mandible incline.

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Introduction

Congenital cervical or cervicothoracic scoliosis is usually caused by multiple defects of segmentation or formation, examples of which include block vertebrae, hemivertebrae, and unilateral bar. Such deformities can increase the risk of undesired posture, including head tilt (HT), shoulder imbalance, and mandibular deviation [1,2], which often trouble patients and their families.

Coronal deformity is usually accompanied by a sagittal malformation known as Klippel-Feil syndrome (KFS), which is defined by the presence of a congenital synostosis of two or more cervical vertebrae. In a study of 22 patients with KFS, Guille et al. [3] found that the mean scoliotic curve angle ranged from 20° to 31° on average, and that larger curves are always associated with proximal thoracic compensatory curve. The prevalence of cervical scoliosis, with multiple contiguous, congenitally fused cervical segments, is as high as 70% in KFS [4]. The extraskelatal malformations usually include cardiac or genitourinary abnormalities, accessory spleen, renal agenesis, congenital anal atresia, cleft palate, and hearing deficiencies. Skeletal anomalies may take several forms, including cervical rib and Sprengel deformity [5]. KFS occurs in an estimated 1 in 40,000 to 1 in 42,000 births [5], with slight female predominance of about 3:2 [6].

The complexity of congenital cervical or cervicothoracic scoliosis and of structures such as the vertebral artery can greatly increase difficulties in surgical correction, which was for a long time limited to posterior in situ fusion [7,8]. Then, in 2004, Ruf et al. [9] reported the introduction of a combined surgical approach. The purpose of our research was to prospectively investigate in children (1) the therapeutic and radiological outcomes of surgical correction of congenital cervical or cervicothoracic scoliosis by a combined anteroposterior (AP) approach and (2) the aesthetic and clinical characteristics of the cervical and cervicothoracic regions in these patients.

Materials and methods

Study participants

The criterion for inclusion in our study was the presence of torticollis caused by congenital cervical or cervicothoracic scoliosis (defined as a Cobb angle of >10°). These children presented with symptoms that included a twisted and tilted neck, neck stiffness, neck pain, restricted neck rotation, asymmetrical facial features, and unequal eye slant. The indication for such surgery is clinically apparent deformity or an oblique HT.

A cohort of 16 patients (6 males and 10 females) with an average age of 9.2 years (range 4–16 years) was recruited prospectively from 2009 to 2013 in our orthopedic center after we obtained review board approval from Peking University Third Hospital. All patients were treated using an AP combined surgical approach.

The locations of the deformities were classified as upper (C0–C2), upper-middle (C0–C5), middle (C3–C5), middle-lower (C3–T1), and lower (C6–T1). Exclusion criteria included muscular torticollis, cervical kyphosis because of neurofibromatosis, atlantoaxial joint deformity, and other sagittal disorders. Neither bracing nor previous surgery was considered exclusionary (Fig. 1). Follow-up was intended to continue for at least 5 years.

Radiographic parameters of spinal alignment

Radiographic parameters of coronal and sagittal spinal alignments (Figs. 2 and 3, respectively) were measured independently by two surgeons before surgery, immediately after surgery, and at the final follow-up evaluation. We recorded the average of each measurement.

Preoperative computed tomography assessment of bone quality and pedicle size

Bone quality and size of the cervical pedicles were assessed before surgery. Categorization was based mostly on the patients' developmental stage and bone quality, considering that some of the young patients were undernourished, along with a desire to spare the motion segments and discs as much as possible.

Assessment of the spinal cord was conducted using magnetic resonance imaging to identify deformities such as spina bifida, meningocele, and syringomyelia. Preoperative

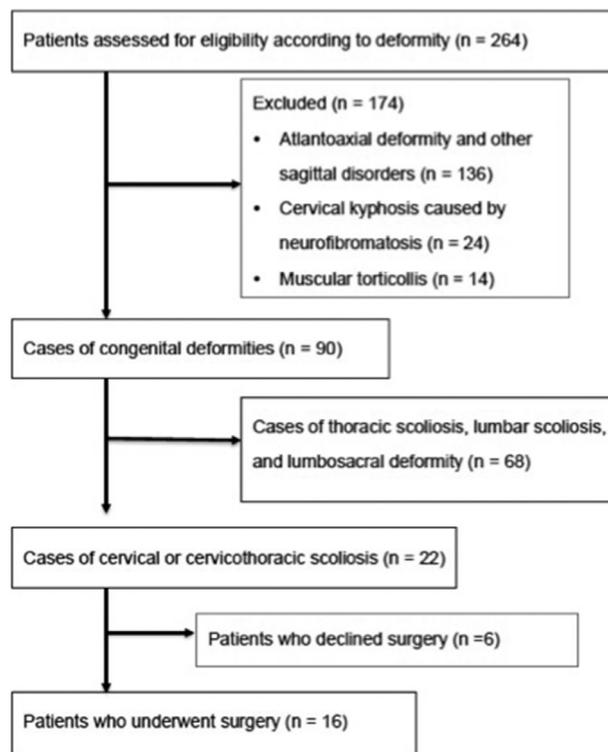


Fig. 1. Flow diagram of participant selection for surgery for cervical or cervicothoracic scoliosis.

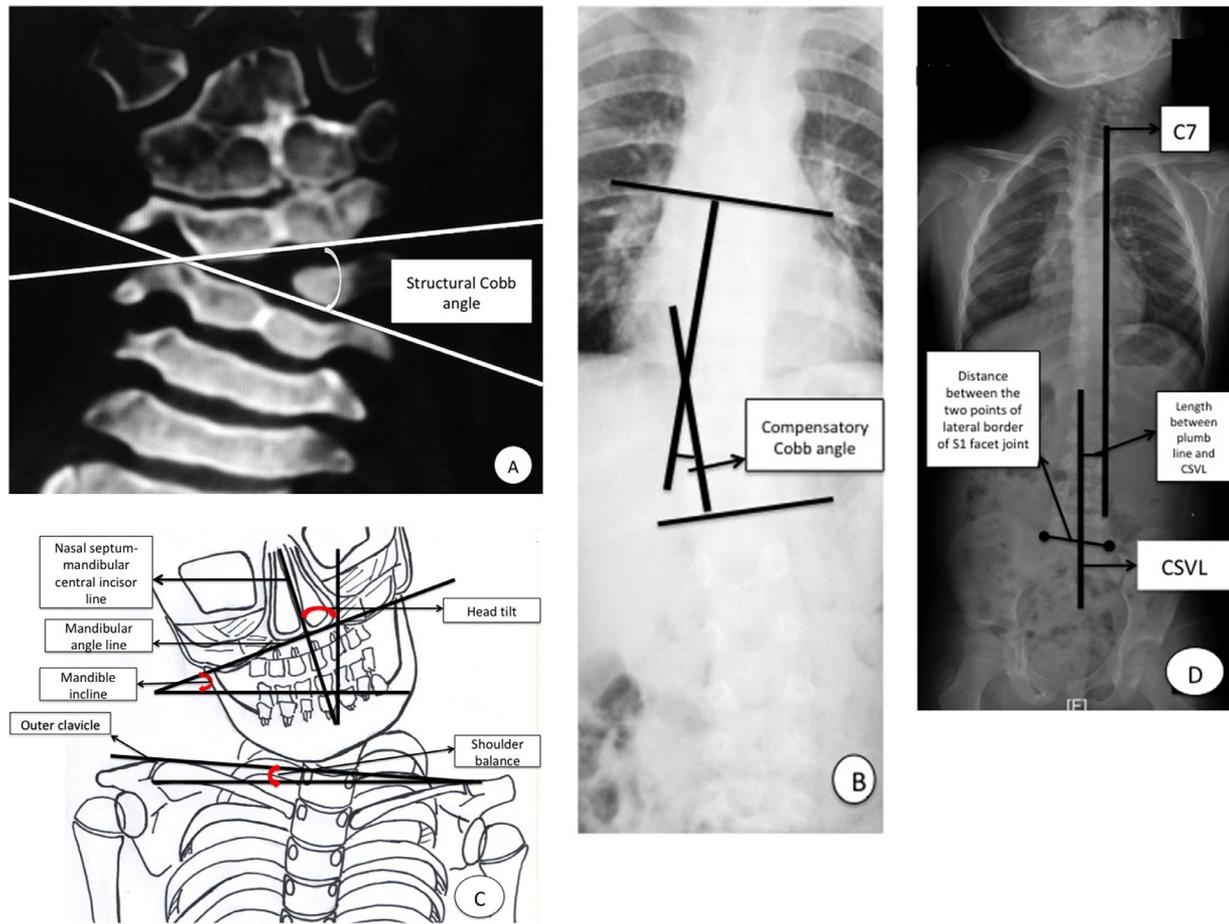


Fig. 2. Definitions of coronal radiographic parameters of spinal alignment. All lengths and distances were measured in millimeters. Ratios between some parameters were measured to adjust for a possible effect of age. (A) Structural Cobb angle: the large curve in the segments of congenital deformity that causes clinical asymmetry or head deviation, needs to be surgically corrected, and is shown as the angle between the lines drawn parallel to the inferior endplate of the most cranial vertebra and to the superior endplate of the most caudal vertebra in the curve. (Image is from participant 2.) (B) Compensatory Cobb angle: the small curve in the segments without vertebral deformities. It is compensatory to the structural curve, and it is shown as the angle between the lines drawn parallel to the inferior endplate of the most cranial vertebra and to the superior endplate of the most caudal vertebra in the curve. (Image is from participant 2.) (C) Head tilt: the angle of the deviation between the nasal septum–mandibular central incisor line from a vertical line. Mandible incline: the deviation of the mandibular angle line from a horizontal line. Shoulder balance: the angle between the outer clavicle and a horizontal line [23]. (Image is from participant 8.) (D) C7–CSVL (central sacral vertical line) ratio: the length of a plumb line dropped from vertebra C7 to the CSVL, divided by the distance between the two points of the lateral border of the S1 superior facet joint in the coronal plane. (Image is from participant 7.)

computed tomography angiography of the vertebral artery was used to determine the abnormality.

Surgical techniques

Resection of the cervical or cervicothoracic hemivertebra was performed using an AP or anteroposterior-anterior combined approach. A sterilized three-dimensional (3D) printed model was prepared to assist the surgeon in recognizing anatomical malformations during surgery. Neurophysiological monitoring of the spinal cord was done throughout the procedure by measuring somatosensory-evoked and motor-evoked potentials.

Patients were placed in the supine position and then given general anesthesia. The hemivertebra and adjacent discs were entirely dissected through an anterior approach.

Then a nerve hook was used to identify the anterior part of the transverse process, which was resected laterally as much as possible. The vertebral artery was anteriorly exposed. If the hemivertebra was nonsegmented or semi-segmented, a wedge was shaped out of the consolidated bony mass (Fig. 4A). In hemivertebral resections, the surgeon identified vertebral arteries on both sides and then concluded the procedure. For patients with a wedge vertebra, the surgeon performed a discectomy.

Then during the same procedure, patients were placed in the prone position for a posterior approach. The lamina of the hemivertebra was resected, and the spinal cord was exposed. In patients with fused lamina, a wedge-shaped portion of the consolidated bony mass was resected (Fig. 4B). The adjacent segmental nerve roots were also exposed so as to prevent surgical injuries during compression or distraction. Preoperative

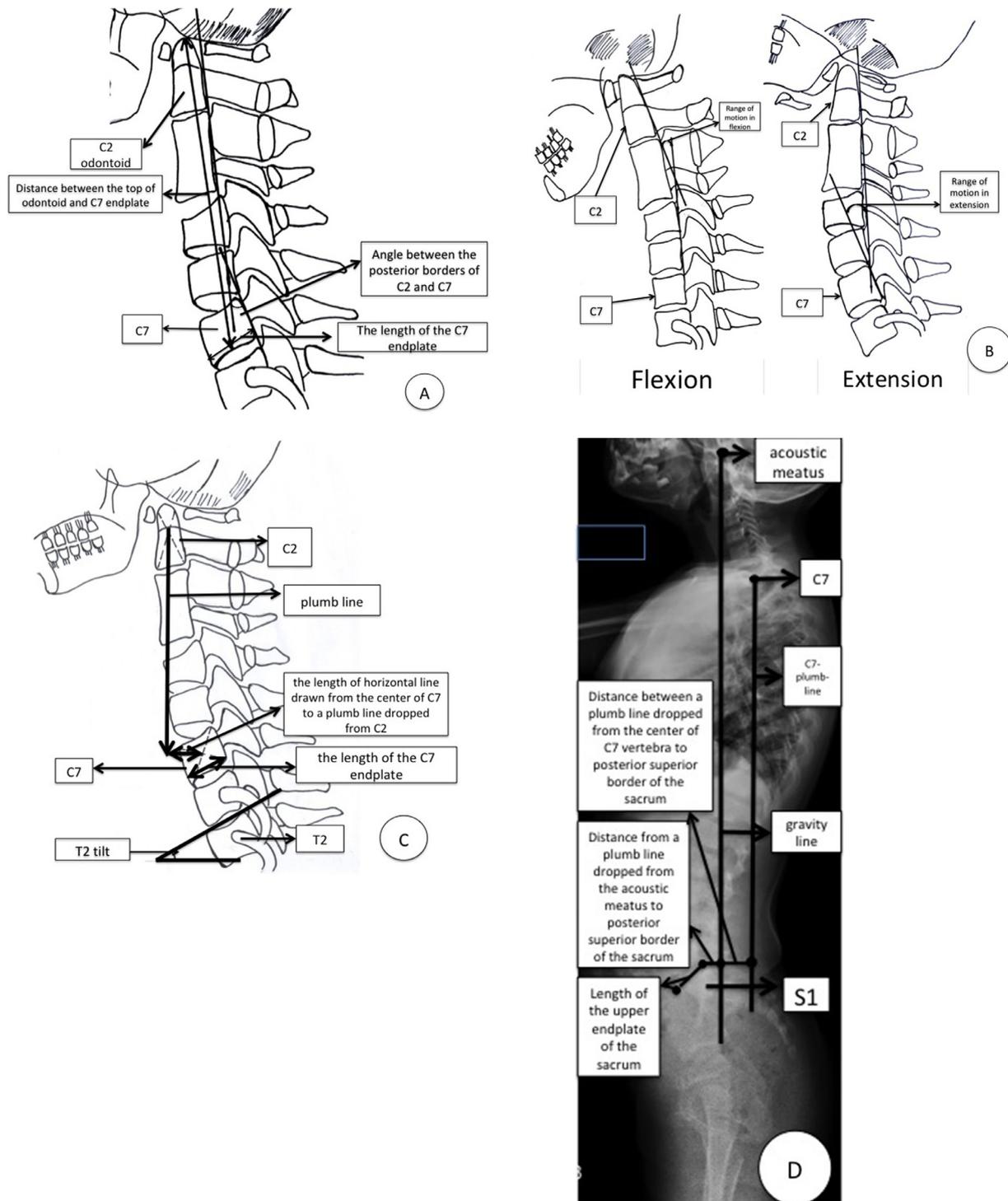


Fig. 3. Definitions of sagittal radiographic parameters of spinal alignment. All distances and lengths were measured in millimeters. Ratios between some parameters were measured to adjust for a possible effect of age. (A) Cervical lordosis: the angle between the posterior borders of C2 and There is a space missing between these words. Please insert one. length ratio: the distance between the midpoint at the top of the C2 odontoid and the inferior endplate of C7, divided by the length of the superior of C7 endplate. (Image is from participant 7.) (B) Cervical range of motion: the sum of the angles measured between the posterior borders of C2 and C7 in flexion plus that in extension. (Image is from participant 7.) (C) T2 tilt: the angle between the upper endplate of the T2 vertebra and a horizontal line. C2–C7 SVA (sagittal vertical axis) ratio: the length of a horizontal line drawn from the center of vertebra C7 to a plumb line dropped from vertebra C2, divided by the length of the inferior endplate of the C7 vertebra. (Image is from participant 7.) (D) C7–SVA ratio: the horizontal distance between a plumb line dropped from the center of the C7 vertebra to the posterosuperior border of the sacrum, divided by the length of the upper endplate of the sacrum. (Image is from participant 11.) Gravity line (GL) ratio: the horizontal distance from a plumb line dropped from the acoustic meatus to the posterosuperior border of the sacrum, divided by the length of the upper endplate of the sacrum [23]. (Image is from participant 11.)

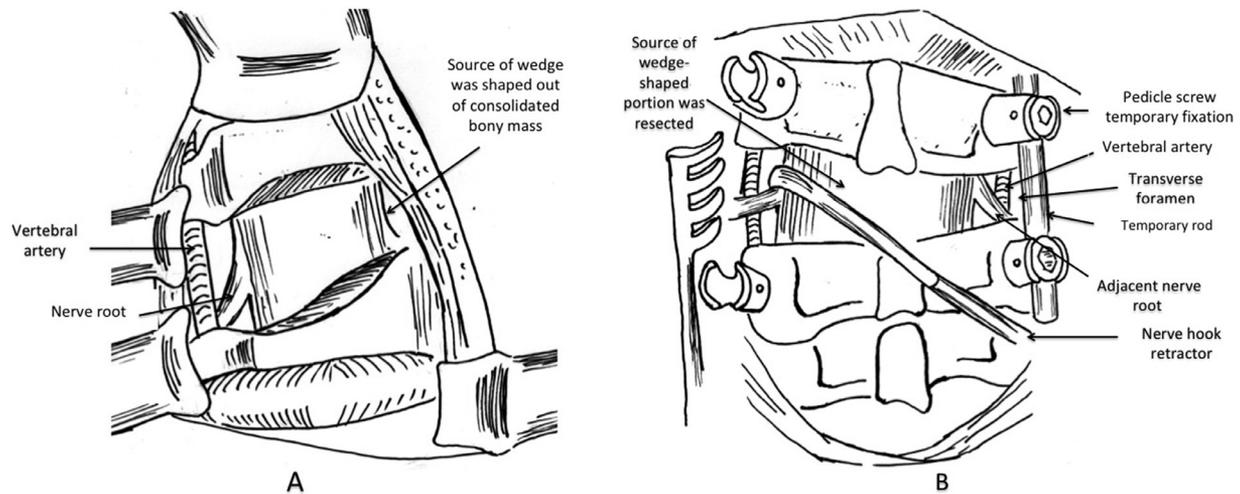


Fig. 4. (A) In the anterior approach in patients in whom the hemivertebra was unsegmented or semisegmented, a wedge-shaped portion was shaped out of the consolidated bony mass. (B) In the posterior approach in patients with fused lamina, the wedge-shaped portion of the consolidated bony mass was resected.

assessment of bone quality and the size of the cervical pedicles, complexity of the deformity, and surgical judgment as to the likelihood of successful implantation guided implantation of pedicle screws at one or at two levels. Participants who were undernourished underwent implantation at two levels. Implantation was done on both cranial and caudal sides (cervical pedicle screw diameter, 3.5 mm; rod diameter, 4.5 mm).

Temporary fixation on the concave side was carried out to prevent intraoperative neurological injury. First, the transverse foramen and the vertebral artery were identified, and then the residual bone structure was completely resected. The resected bone was retained for possible use in case the patient underwent a subsequent anterior approach. The vertebral artery was completely released along its course between the nerve roots, and the residual pedicle was resected along its medial wall. The gap between the adjacent levels after resection was thereafter slowly closed by bending the head to the convex side under Mayfield traction by an assistant on the cranial side and by compression from pedicle screws placed between the superior and inferior levels.

Intraoperative fluoroscopy was done to determine whether closure was incomplete and whether there were still gaps at the osteotomy site. If there were still gaps, then the patient was placed in the supine position for anterior fusion with plate fixation. Bone harvested via the posterior approach was preferred to bone harvested from the iliac crest cortical bone. The bone, preferably contained in a polyetheretherketone cage to provide additional support, was grafted into the intervertebral space, above which a titanium plate was then placed.

Patient satisfaction

At the final follow-up evaluation, patients or their parents rated their satisfaction with surgery outcome as

“very satisfied,” “satisfied,” “neither satisfied nor dissatisfied,” “dissatisfied,” or “very dissatisfied” (Fig. 5).

Statistical analysis

An adaptation of the Kolmogorov-Smirnov test was used to examine whether the data were normally distributed. Nominal relationships were assessed with the chi-square test. Preoperative and postoperative interval variables were assessed with the Mann-Whitney *U* test. Descriptive statistics in the form of median (minimum, maximum) for all parameters were provided for all patients. Measurements of radiographic parameters of spinal alignment by two surgeons were assessed for interobserver agreement with the Pearson rank correlation coefficient test. Statistical significance was set at .05. Data were analyzed with SPSS Statistics software, version 20.0 (IBM, Armonk, NY, USA).

Study approval, informed consent, and funding

The study protocol was approved by the hospital’s institutional review board, and written evidence of informed consent was obtained from patients’ parents.

Results

Interobserver correlation for all radiographic parameters was 0.97. Of the 264 patients assessed for study eligibility, 84 underwent surgery for congenital deformities, of whom 16 (19%) underwent surgery for cervical or cervicothoracic scoliosis.

Characteristics of congenital cervical and cervicothoracic scoliosis

Demographics for patients with cervical or cervicothoracic deformity are shown in Table 1, and surgical details and outcomes are shown in Table 2. Participants 9 and 10 had a surgical history of sternocleidomastoid muscle release, but

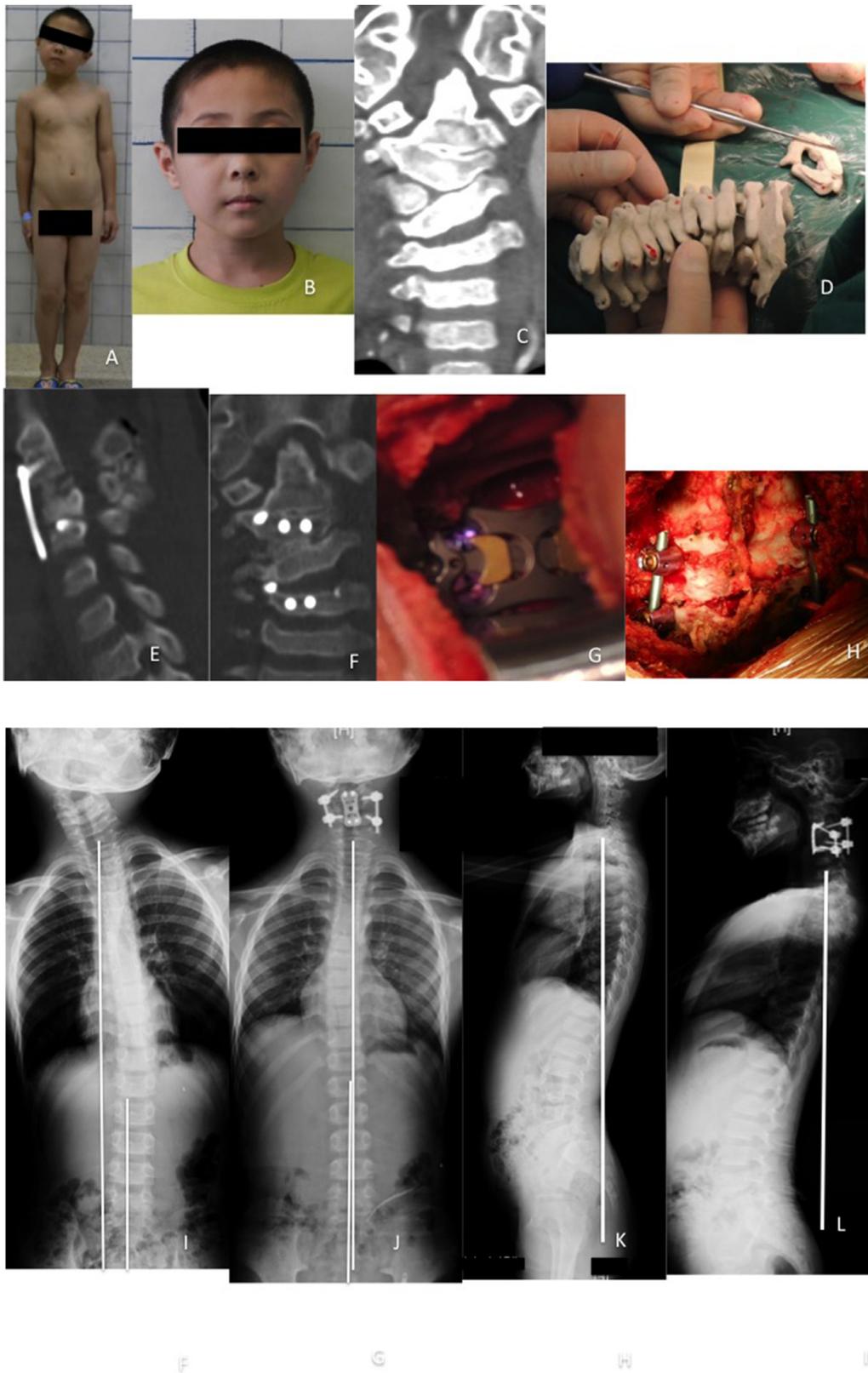


Fig. 5. A 9-year-old boy (patient 4) had a tilted neck. He could no longer tolerate bracing after several months. (A) A preoperative photograph shows an oblique eye slant and head tilt. (B) A photograph taken 3 years after surgery shows improved appearance. (C) C2–C3 unsegmented and C4 incarcerated hemivertebrae can be seen on a preoperative computed tomography scan in the coronal plane. (D) A sterilized three-dimensional printed model was used for pinpointing anatomical malformations during surgery from a posterior approach. Computed tomography scans show solid fusion on coronal (E) and sagittal (F) planes 3 years after surgery. (G) A photograph shows an anterior hemivertebrectomy and bone fusion with plate instrumentation during the second surgery, via an anterior approach. (H) A photograph shows a posterior laminar and facet joint resection with pedicle fixation. (I) A global coronal image shows the right shift of the C7–CSVL (C7 central sacral vertical line). (J) Satisfactory balance was achieved. (K, L) The sagittal plane maintaining balance before (K) and after (L).

Table 1
Demographics for patients with cervical or cervicothoracic scoliosis

Anteroposterior approach								
Patient no.	Type of deformity	Age at diagnosis (years)	Age at surgery (years)	Primary curve	Compensatory curve	Other skeletal malformation	Extraskeletal malformation	Klippel-Feil syndrome
5	C3 semisegment HVB	4	6	C2–C4	C4–T1	Occipitalization	None	None
12	C2–C5 unilateral bar; C3–C5 unilateral semi-segmented HVB; C6–C7 block vertebra	Newborn	8	C2–C5	C7–L3	None	None	Type III
1	C3 semisegmented HVB; C6–C7 wedge vertebra	6	10	C2–C4	C7–T5	Occipitalization	None	None
3	C2–C4 block vertebra; C5 fully segmented HVB	Newborn	14	C2–C5	C5–T5	None	Polycystic kidney	None
9	C2–C3 wedge vertebra	Newborn	15	C2–C3	C3–T5	Occipitalization	None	None
Anteroposterior–anterior approach								
Patient no.	Type of deformity	Age at diagnosis	Age at surgery	Primary curve	Compensatory curve	Other skeletal malformation	Extraskeletal malformation	Klippel-Feil syndrome
16	C5–C6–C7 wedge vertebrae; C4 unsegmented HVB	6	11	C3–C7	C7–T5	None	Accessory spleen	None
11	C2–C4 right unilateral bar; left C2–C4 semisegmented HBV; C3 fully segmented HVB	Newborn	4	C1–C4	C4–T4	Occipitalization	None	Type I
2	C2–C4 block vertebra; C5 semisegmented HBV	2	5	C2–C6	C6–T10	Sprengel deformity and cervical rib	None	None
10	C1–C4 right unilateral bar; C3 left unsegmented HBV	Newborn	6	C1–C4	C4–T3	Occipitalization	None	Type II
14	C2–C4 block vertebrae; C5–C6 block vertebra; C7–T4 right unilateral bar; T1 semisegmented left HVB; left L4 semi-segmented HVB	Newborn	7	C2–C4; C4–C7; C7–T5	T5–T12	Sprengel deformity and cervical rib	None	Type III
6	C2–C3 block vertebra; C4 incarcerated HV	Newborn	8	C2–C5	C5–T4; T10–L1	None	None	Type III
7	C2–C3 block vertebra; C4 nonsegmented HVB	5	9	C2–C5	C5–T11	None	None	None
4	C2–C4 left unilateral bar; C3 right nonsegmented hemivertebra	6	9	C2–C4	C4–T5	None	None	None
13	C2–C3 block vertebra; C5–T3 right unilateral bar; T2 left nonsegmented HBV	Newborn	11	C2–C3; C5–T3	T3–T10	Sprengel deformity	None	Type III
8	C5–C6 wedge vertebra	6	12	C5–C6	C6–T4	None	None	None
15	C6–C7 to T1–T2 right unilateral bar; C7–T1 left fully segmented HBV	1	15	C0–C3; T1–T4	C6–C7; T7–L5	None	None	Type III

HVB, hemivertebra.

Table 2

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<i>Anteroposterior approach</i>							
Patient no.	Type of deformity	Surgical options	Surgical time (min)	Blood loss (mL)	Complications	Duration of follow-up	Satisfaction
5	C3 semisegmented HVB	AP A: C3 hemivertebrectomy P: C2–C4 laminectomy, pedicle screw fixation, and fusion	430	700	None	59	Very satisfied
12	C2–C5 unilateral bar; C3–C5 unilateral semisegmented HVB; C6–C7 block vertebra	AP A: C4 hemivertebrectomy; C3–C5 bone fusion and plate fixation P: C3–C5 laminectomy; left C3–C5 facetectomy and transversectomy; C3–C5 pedicle screw fixation and fusion	510	500	None	44	Satisfied
1	C3 semisegmented HVB, C6–C7 wedge vertebra	AP A: C3 hemivertebrectomy; P: C3 laminectomy; C2–C4 pedicle screw fixation and fusion	423	900	None	83	Neither satisfied nor dissatisfied
3	C2–C4 block vertebra; C5 fully segmented HVB	AP A: C5 hemivertebrectomy P: Left C4–C6 laminectomy, pedicle screw fixation, and fusion	110	50	None	95	Very satisfied
9	C2–C3 wedge vertebra	AP A: C2–C3 discectomy with fusion P: right C2–C3 laminectomy and facetectomy; right C2–C3 pedicle screw fixation and fusion	150	270	None	58	Very satisfied
<i>Anteroposterior–anterior approach</i>							
Patient no.	Type of deformity	Surgical options	Surgical time (min)	Blood loss (mL)	Complications	Duration of follow-up	Satisfaction
16	C5–C6–C7 wedge vertebrae; C4 unsegmented HVB	APA A: C7 hemivertebrectomy P: C7 laminectomy; right C6–C7 facetectomy; left C5–T1–T2 pedicle screw fixation and fusion; right C5–C6–T1–T2 pedicle screw fixation and fusion A: C6–T1 bone fusion with plate fixation	465	1000	C7 nerve root palsy	59	Neither satisfied nor dissatisfied
11	C2–C4 right unilateral bar; left C2–C4 semisegmented HBV; C3 fully segmented HVB	APA A: C3 hemivertebrectomy P: C2–C4–C5 laminectomy with pedicle screw fixation and fusion; A: C2–C4 bone fusion with plate fixation	523	1000	None	59	Very satisfied
2	C2–C4 block vertebra; C5 semisegmented HBV	APA A: C5 hemivertebrectomy P: C4–C6 laminectomy with pedicle screw fixation and fusion A: C4–C6 bone fusion with plate fixation	400	500	None	84	Very satisfied
10	C1–C4 right unilateral bar; C3 left unsegmented HBV	APA A: C3 hemivertebrectomy; C4 partial corpectomy P: C3–C5 facetectomy and transversectomy; C2–C6 laminectomy, pedicle screw fixation, and fusion A: C3–C6 bone fusion with plate fixation	523	1000	None	60	Satisfied

Table 2 (Continued)

<i>Anteroposterior approach</i>							
Patient no.	Type of deformity	Surgical options	Surgical time (min)	Blood loss (mL)	Complications	Duration of follow-up	Satisfaction
14	C2–C4 block vertebrae; C5–C6 block vertebra; C7–T4 right unilateral bar; T1 semisegmented left HVB; left L4 semisegmented HVB	APA A: T1 hemivertebrectomy P: Left C7-T2 facetectomy; T1 laminectomy; C7–T2 pedicle screw fixation and fusion A: C7–T2 bone fusion with plating	401	650	T1 nerve root palsy	58	Very satisfied
6	C2–C3 block vertebra; C4 incarcerated HVB	APA A: C4 hemivertebrectomy P: C4 laminectomy and transversectomy; C3–C5 pedicle screw fixation and fusion A: C3–C5 bone fusion with plate fixation	360	1000	None	70	Very satisfied
7	C2–C3 block vertebra; C4 nonsegmented HVB	APA A: C4 hemivertebrectomy P: C3–C5 laminectomy; right C2–C6 pedicle screw fixation and fusion; left C3–C6 pedicle screw fixation and fusion A: C3–C5 bone fusion with plate fixation	360	700	C5 nerve root palsy	72	Satisfied
4	C2–C4 left unilateral bar; C3 right nonsegmented hemivertebra	APA A: C3 hemi-vertebrectomy P: C3 laminectomy; C2–C4 partial facetectomy, C2–C4 pedicle screw fixation and fusion A: C2–C4 bone fusion and plating	513	2500	C5 nerve root palsy	112	Satisfied
13	C2–C3 block vertebra; C5–T3 right unilateral bar; T2 left nonsegmented HBV	APA A: T2 hemivertebrectomy P: T1–T3 facetectomy; T1–T4 pedicle screw fixation and fusion A: T1–T3 bone fusion and plate fixation	326	510	None	60	Neither satisfied nor dissatisfied
8	C5–C6 wedge vertebra	APA A: C5–C6 discectomy; C5 partial corpectomy P: C5–C6 laminectomy and facetectomy; C5–C7 pedicle screw fixation and fusion A: C5–C6 bone fusion with plate fixation	240	100	None	60	Very satisfied
15	C6–C7 to T1–T2 right unilateral bar; C7–T1 left fully segmented HBV	APA A: C7 hemivertebrectomy P: C7 laminectomy; C5–T2 pedicle screw fixation and fusion A: C6–T1 bone fusion with plate fixation	347	400	None	58	Dissatisfied

A, anterior; AP, anteroposterior; APA, anteroposterior-anterior; HVB, hemivertebra; P, posterior.

this procedure did not ameliorate torticollis because cervical scoliosis had been misdiagnosed as muscular torticollis. None of 16 participants was lost to follow-up.

Cervical and cervicothoracic deformities were diagnosed at birth in 8 patients, before age 5 years in 3 patients, and after age 5 years in 5 patients. The number and distribution of deformity locations were as follows: in the upper and middle spine (C0–C5) in 13 patients, in the middle spine (C3–C5) in 1 patient, and in the middle and lower spine (C3–C7) in 2 patients. Other skeletal malformations were found in 8 patients, and extraskeletal malformations were found in 2 patients. KFS was diagnosed in 7 patients. The incidence of KFS was 43.8% (7/16) in this cohort: type 1, 1 patient; type 2, 1 patient; type 3, 5 patients [10].

Clinical outcomes of surgery

Surgical data are as follows. The average duration of follow-up was 68.0 months (range, 44–112 months). The average blood loss during surgery was 675.0 mL (range, 50–2500 mL). The average duration of surgery was 400.5 minutes (range, 110–523 minutes).

Twelve of the 16 participants or their parents rated patient satisfaction as “very satisfied” or “satisfied,” and the other 4 rated it as “neither satisfied nor dissatisfied” or “dissatisfied” (Table 2).

There were some complications: 4 patients developed a postoperative complication of nerve root palsy, including 2 at the level of C5, 1 at C7, and 1 at T1. By 6 months after surgery, all of these patients had totally recovered. Neither vertebral artery injuries nor tears of the dural sac were observed either during surgery or during the follow-up period in this cohort.

The correlation between radiological parameters of spinal alignment before surgery

Regarding relationships between preoperative parameters (Table 3), there were significant correlations between mandible incline (MI) and HT, and between MI and shoulder balance (SB) (p<.05). The C7 central sacral vertical line (C7–CSVL) ratio significantly correlated with HT, MI, and SB (p<.05), and the C7 sagittal vertical axis (C7–SVA) ratio significantly correlated with structural curve and cervical lordosis (p<.05). There were additional correlations between structural and compensatory curves as well as between structural curve and cervical lordosis (p<.05). Moreover, there were correlations between the gravity line (GL) ratio and structural curve, compensatory curve, cervical lordosis, and the C7–SVA ratio (p<.05).

Comparisons between preoperative and postoperative radiographic parameters of spinal alignment

There was a significant difference in structural and compensatory curves between the preoperative and immediate postoperative Cobb angles (p<.001), but none between

Table 3
Pearson correlation coefficients between parameters of spinal alignment

Parameter	Parameter (r, p)												
	SCA	CCA	HT	M	SB	C7–CSVL ratio	CL	CLR	CROM	C7–SVA ratio	GL ratio	T2 Tilt	C2–C7 SVA ratio
SCA													
CCA	0.51; 0.03												
HT	0.04; 0.88	0.08; 0.74											
MI	0.02; 0.95	-0.06; 0.82	0.71; 0.001										
SB	-0.04; 0.89	0.08; 0.77	-0.45; 0.06	-0.59; 0.047									
C7–CSVL ratio	0.15; 0.56	-0.02; 0.93	-0.477; 0.046	-0.68; 0.002	0.68; 0.002								
CL	-0.58; 0.01	-0.39; 0.11;	-0.09; 0.73	0.1; 0.69	0.18; 0.47	0.07; 0.78							
CLR	0.2; 0.43	0.02; 0.95;	-0.17; 0.49	-0.55; 0.02	0.57; 0.01	0.62; 0.01	0.04; 0.86						
CROM;	-0.28; 0.26	0.09; 0.71;	-0.15; 0.56	-0.34; 0.17	0.23; 0.37	0.02; 0.98	0.07; 0.79	0.3; 0.23					
C7–SVA ratio	-0.61; 0.008	-0.38; 0.12	-0.13; 0.58	0.06; 0.81	0.3; 0.23	-0.04; 0.89	0.62; 0.007	-0.13; 0.62	-0.07; 0.79				
GL ratio	-0.61; 0.008	-0.52; 0.03	-0.04; 0.88	0.09; 0.72	0.23; 0.35	-0.07; 0.77	0.5; 0.03	-0.13; 0.62	-0.03; 0.92	0.93; 0			
T2 tilt	-0.42; 0.09	-0.41; 0.9;	0.31; 0.21	0.2; 0.43	-0.03; 0.92	-0.1; 0.7	0.34; 0.17	-0.37; 0.13	-0.13; 0.61	0.17; 0.51	0.36; 0.14		
C2–7 SVA ratio	0.36; 0.14	0.2; 0.43	0.34; 0.17	0.06; 0.82	0.16; 0.52	-0.1; 0.7	0.35; 0.15	0.2; 0.42	0.18; 0.47	-0.15; 0.57	-0.04; 0.87	0.22; 0.38	

SCA, structural Cobb angle; CCA compensatory Cobb angle; CL, cervical lordosis; CLR, cervical length ratio; CROM, cervical range of motion; CSVL, central sacral vertical line; GL, gravity line; HT, head tilt; MI, mandible incline; p, probability value; r, Pearson correlation coefficient; SB, shoulder balance; SVA, sagittal vertical axis.

immediate postoperative and final postoperative Cobb angles ($p=.68$ and $.44$). The average correction rates were 68.5% (range, 13.9%–97.3%) for structural curves and 29.5% (range, 42.9%–78.1%) for compensatory curves. Preoperative cervical length had increased significantly by the time of the final follow-up examination ($p<.001$). Moreover, the values for HT and MI were found to be statistically significantly different before versus after surgery ($p=.04$ and $.03$, respectively). Nevertheless, no statistically significant differences were observed for cervical lordosis, range of motion, C7–CSVL ratio, C7–SVA ratio, C2–C7SVA ratio, the angle between the upper endplate of the T2 vertebra and a horizontal line (T2 tilt), GL ratio, or SB between presurgery values and final follow-up values.

Discussion

Congenital cervical spinal anomalies are usually detected incidentally and radiographs are taken only when the patients develop either a rotatory or laterally decompensated HT that proves recalcitrant to stretching and physical therapy maneuvers [11]. The incidence of coexistence of mandibular deviation and cervical scoliosis is about 9.8%, and a linear correlation has been proved to exist between the degree of mandibular deviation and that of scoliosis, as well as between the imbalance of the trunk and that of the shoulder [12]. The curves in the frontal plane contribute to the development of different dentofacial asymmetries, with the head tilting sideways because the cervical spine and muscles serve to maintain head posture and play an important role in the head's complex and diverse movements [13]. Therefore, the main reason for medical consultations in our cohort was aesthetic asymmetries noticed by the patients' parents. The parents were concerned because exposure to degrading comments from the children's peers about their appearance could lead to the development of psychosocial problems.

As shown in Table 3, MI was significantly correlated with HT and SB, which indicates that vertical parameters are more sensitive than horizontal ones in manifesting cervical scoliosis. However, the relationship between the C7–CSVL ratio and all aesthetic parameters, including HT, MI, and SB, suggests that global coronal balance is influenced by the head-shoulder alignment. Moreover, the correlation between the C7–SVA ratio and the structural curve, as well as cervical lordosis, indicates that coronal and sagittal alignment interact to affect deformity status, and an abnormal cervical curve could affect thoracolumbar curvature in scoliosis [14], because the C7–SVA ratio is believed to reflect the modification of thoracic and lumbar alignment. Besides, the GL ratio seems to be more sensitive than the C7–SVA ratio to reflect both coronal and sagittal parameters. Yagi et al. verified this advantage of the GL over the C7plumb line in adults with spinal deformity [15]. Meanwhile, cervical lordosis was correlated with structural curve, which proves that the coupled motion principle may

also be appropriate in the cervical spine despite the high incidence of multiple defects of segmentation and formation that reduces the motion of these segments. Consequently, the correlation of slight Cobb angles and asymmetrical facial appearance indicates that cervical coronal deformity is liable to cause obvious aesthetic problems, because there are relatively fewer segments adjacent to the deformity that form a compensatory curve to balance the existing structure.

In our cohort, 2 patients underwent sternocleidomastoid muscle release before the operation that we performed. However, the ineffectiveness of such manipulation leads us to consider spinal deformity as a possible cause of torticollis, to which muscular and osseous factors could make similar contributions.

For all patients with scoliosis, the complexities of both the deformity and the vertebral artery make it a great challenge to determine the most effective surgical strategy. Therefore, a combined surgical approach is needed.

Bracing is of relatively limited usefulness in congenital cervical scoliosis. The first 2 patients (case 1 and 2) in our cohort were given braces at their first consultations, but the progression of the deformity regarding appearance caused the parents and physicians to end that treatment. Moreover, the unsatisfying results of halo-traction in our cohort (cases 4, 5, and 6) convinced us to refer the patients directly to surgeons.

The first case of cervical hemivertebra resection with a combined approach was reported in 1981 by Deburge and Briard [16]. However, serial reports have been rare since then except for the one by Ruf et al. [9].

In surgery, anterior release was indicated with deformity resection on the convex side regardless of the types of defects in segmentation or formation. Even in nonsegmented deformities, minor motion could still have been achieved if the intervertebral disc and surrounding soft tissues were removed. Nevertheless, the anterior hemivertebrectomy was quite limited because of the obstacle of the longus colli muscle. Therefore, posterior resection of the residual transverse part in the lateral side is a vital supplement to the anterior approach. Angiography was necessary before surgery to avoid injuring the vertebral artery. Posterior facet joint resection during the procedure allowed for clear exposure of the artery and the nerve root. Exposure of the vertebral artery and instrumentation with pedicle screws are the two most crucial steps of the procedure. Abnormal development of cervical pedicle always puts patients at risk of intraoperative injury because of its tiny diameter. To compensate, we included a reference to the fixation of the adjacent level in all surgery plans after measuring and calculating pedicle diameter on computed tomography scans.

Moreover, a 3D printed model aids identification of abnormal anatomical landmarks during surgery when there is no navigation system. The remaining anterior transverse process on the ventral side of the nerve root does not hinder compression between the screws after resection of the major anatomical structures of the column.

Table 4
Comparisons between presurgical and postsurgical radiographic parameters of spinal alignment

Parameter	Before surgery	Immediately after surgery	At final follow-up evaluation
SCA (°)	38.2a (20.6, 67.2)	11.3a (0.0, 42.7)	12.8 (0.3, 42.0)
CCA (°)	19.8b (7.5, 37.0)	13.1b (1.5, 32.0)	12.9 (3.7, 38.0)
HT (°)	6.4c (−14.2, 22.9)	0.1c (−3.9, 8.8)	0.0 (−4.1, 8.7)
MI (°)	5.8d (−6.3, 25.1)	0.8d (−2.7, 6.0)	0.9 (−2.6, 6.2)
SB (°)	−0.95 (−7.5, 5.5)	0.12 (−4.3, 3.8)	−0.8 (−8.3, 6.6)
C7–CSVL ratio	−12.7 (−36.1, 28.1)	−8.2 (−36.2, 19.2)	−6.9 (−37.8, 17.8)
CL (°)	2.75 (−16.9, 42.3)	4.9 (−8.5, 25.5)	10.2 (−12.1, 51.7)
CLR	92.4e (58, 133.1)	106.9e (63.2, 140.1)	107.3 (62.6, 138.2)
CROM (°)	35.4 (8.0, 81.9)	35.5 (6.8, 59.2)	35.3 (6.4, 58.8)
C7–SVA ratio	15.5 (−1.6, 36.6)	−8.3 (−45.3, 30.1)	−10.7 (−60.5, 32.6)
GL ratio	−9.6 (−62.3, 94.0)	−9.5 (−31.2, 28.4)	−9.7 (−29.3, 32.9)
T2 tilt (°)	18.8 (7.9, 29.4)	18.4 (5.6, 28.3)	18.3 (4.0, 29.2)
C2–C7 SVA ratio	13.5 (−1.6, 36.6)	13.2 (−26.9, 34.1)	14.9 (−29.3, 32.9)

Nonparametric test: ap < .001; bp < .001; cp = .04, dp = .03, and ep = .00.

CCA, compensatory Cobb angle; CL, cervical lordosis; CLR, cervical length ratio; CROM, cervical range of motion; CSVL, central sacral vertical line; GL, gravity line; HT, head tilt; MI, mandible incline; SB, shoulder balance; SCA, structural Cobb angle; SVA, sagittal vertical axis.

Initially we adopted an AP approach for patients with block vertebrae or a unilateral bar. However, because of concerns about the stability of the three columns, we performed additional anterior procedures after the posterior ones to effect solid fusion. As a result, anterior bone fusion with plating was achieved within 6 to 12 months of follow-up even without external protection such as a Philadelphia collar. Consequently, we believe that anterior column bone support is necessary more often in the cervical spine rather than in thoracic or lumbar segments because of its larger flexibility.

The significant differences between preoperative and postoperative parameters shown in Table 4 suggest that not only structural and compensatory curves but also head-neck postures, which are complex in both the vertical and horizontal planes, are greatly ameliorated by surgery. Moreover, cervical length was not influenced by segmental fusion, probably because the procedures were performed before our patients entered puberty. The residual growth potential of the unfused segments accounts for the ongoing development of cervical spine. However, range of motion was slightly restricted because of multilevel fusion. Besides, the position of the deformities, which were mostly located in upper and middle cervical segments, contributed not only to a decrease in range of motion but also to the

restoration of cervical lordosis after correction [4,17]. In addition, neither global spine balance nor regional cervicothoracic equilibration was influenced by local deformity correction, because there were no significant changes in the values of C7–CSVL, C7–SVA, or C2–C7 SVA and in GL ratios after surgery.

Regarding complications, all patients with postoperative nerve root palsy completely recovered by 6 months of follow-up. Because of this, we believe that intraoperative traction or transient ischemia of the nerve root could be the main cause for this complication.

Pedicle screws provide an ideal closure after resection of the hemivertebra, and a combined approach could generate better clinical results [18,19]. In our cohort, the correction rate was approximately 70%, with an average blood loss of 675 mL and surgery duration of 400 minutes; for compensatory curves, the correction rate reached about 30% by the time of the final follow-up evaluation.

Compared with the results of the study by Ruf et al. [19], our correction rate is lower (68.5% vs. 80%, Table 5), but the blood loss and duration of surgery are similar. The difference can be attributed to our patients' higher incidence of multiple defects in segmentation and formation (Table 1), which reduced cervical flexibility tremendously. Moreover, correction rate is always lower when there is nonsegmented

Table 5
Anteroposterior approach for hemivertebra resection in different segments

	Location	Structural curve correction (%)	Compensatory curve correction (%)	Blood loss (mL)	Surgery time (min)
Jalanko et al. [20]	T & L	71		258	258
Bollini et al. [18]	T & L	69		720	280
Ruf et al. [19]	T & L	81/70	81–84/67–73	309/272	187/272
Zhuang et al. [22]	L & S	83	57	235	207
Ruf et al. [9]	Cervical	80		617	363
Our study	Cervical	68.5	29.5	675	400

L & S, lumbosacral spine; T & L, thoracolumbar spine.

hemivertebra, a unilateral bar with hemivertebra, and block vertebrae. In comparing thoracic or lumbar hemivertebra resection performed with the combined approach in [20–22] Table 5, we found that the correction rate in curves with bar formation was about 70%, and the compensatory correction rate was between 67% and 73%, which was almost 10% lower than for curves without bars. These results show that multiple nonsegmented deformities, whether in the thoracic, lumbar, or cervical spine, reduce flexibility even with adequate release on both sides. Meanwhile, cervical hemivertebra resection always causes more blood loss and a longer duration of surgery despite adequate presurgery preparation. We think the reasons maybe related to the delicate manipulation of pedicle instrumentation, release of the vertebral artery, and resections around the spinal cord.

Our study had some limitations. First, our cohort included only certain types of deformities and thus was not fully representative of the complexity of cervical scoliosis. Additionally, given the young age of our patients, more studies with a longer follow-up period are necessary to determine true clinical success.

In summary, the AP combined approach in hemivertebrectomy can produce satisfactory clinical results in the correction of congenital cervical or cervicothoracic scoliosis.

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References

- [1] Ferro F, Spinella P, Lama N. Transverse maxillary arch form and mandibular asymmetry in patients with posterior unilateral crossbite. *Am J Dentofacial Orthop* 2011;140:828–38.
- [2] Chung KR, Kim SH, et al. Distalization of the mandibular dentition with mini-implants to correct a Class III malocclusion with a midline deviation. *Am J Orthod Dentofacial Orthop* 2010;137:135–46.
- [3] Guille J, Miller A, Bowen R, et al. The natural history of Klippel-Feil syndrome: clinical, roentgenographic, and magnetic resonance imaging findings as adulthood. *J Pediatr Orthop* 1995;15:617–26.
- [4] Samartzis D, Kalluri P, Herman J, et al. Cervical scoliosis in the Klippel-Feil patient. *Spine* 2011;36:E1501–8.
- [5] Thomsen M, Schneider U, Weber M, et al. Scoliosis and congenital anomalies associated with Klippel-Feil syndrome types I–III. *Spine* 1997;22:396–401.
- [6] Warner WC. Pediatric cervical spine. ed. In: Canale ST, ed. *Campbell's Operative Orthopaedics*. St. Louis, PA: Mosby 1999. p. 2815–47.
- [7] 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;66:1188–97.
- [8] Liew SM, Simmons ED Jr. Cervical deformity: rationale for selecting the appropriate fusion technique (anterior, posterior, and 360 degree). *Orthop Clin North Am* 1998;29:779–86.
- [9] Ruf M, Jensen R, Harms J. Hemivertebra resection in the cervical spine. *Spine* 2005;30:380–8.
- [10] Samartzis D, Herman J, Lubicky J, et al. Classification of congenitally fused cervical patterns in Klippel-Feil patients. *Spine* 2006;31:E798–804.
- [11] Canale ST, Griffin DW, Hubbard CN. Congenital muscular torticollis. A long-term follow-up. *J Bone Joint Surg Am* 1982;64:810–6.
- [12] Zhou SC, Yan JJ, Da H, et al. A correlational study of scoliosis and trunk balance in adult patients with mandibular deviation. *PLoS One* 2013;8:e59929.
- [13] Wakano S, Takeda T, Nakajima K, et al. Effect of experimental horizontal mandibular deviation on dynamic balance. *J Prosthodont Res* 2011;55:228–33.
- [14] Yu M, Silvestre C, Roussouly P, et al. Analysis of the cervical spine sagittal alignment in young idiopathic scoliosis: a morphological classification of 120 cases. *Eur Spine J* 2013;22:2372–81.
- [15] Yagi M, Takeda K, Machida M, et al. Discordance of gravity line and C7PL in patient with adult spinal deformity—factors affecting the occiput–trunk sagittal discordance. *Spine Journal* 2015; 15:213–22.
- [16] Deburge A, Briard JL. Cervical hemivertebra excision: report of a case. *J Bone Joint Surg Am* 1981;63:1335–8.
- [17] Samartzis D, Kalluri P, Herman J, et al. 2008 Young Investigator Award: the role of congenitally fused cervical segments upon the space available for the cord and associated symptoms in Klippel-Feil patients. *Spine* 2008;33:1442–5.
- [18] Bollini G, Docquier PL, Viehweger E, et al. Lumbosacral hemivertebrae resection by combined approach: medium- and long-term follow-up. *Spine* 2006;31:1232–9.
- [19] Ruf M, Jensen R, Harms J, et al. Hemivertebra resection and osteotomies in congenital spine deformity. *Spine* 2009;34:1791–9.
- [20] Jalanko T, Rintala R, Puisto V, et al. Hemivertebra resection for congenital scoliosis in young children: comparison of clinical, radiographic, and health-related quality of life outcomes between the anteroposterior and posterolateral approaches. *Spine* 2011;36:41–9.
- [21] Bollini G, Docquier PL, Viehweger E, et al. Thoracolumbar hemivertebrae resection by double approach in a single procedure: long-term follow-up. *Spine* 2006;31:1745–57.
- [22] Zhuang QY, Zhang JG, Li SG, et al. One-stage posterior only lumbosacral hemivertebra resection with short segmental fusion: a more than 2-year follow-up. *Eur Spine J* 2016;25:1567–74.
- [23] Kuklo T, Lenke L, Graham E, et al. Correlation of radiographic, clinical, and patient assessment of shoulder balance following fusion versus nonfusion of the proximal thoracic curve in adolescent idiopathic scoliosis. *Spine* 2002;27:2013–20.