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CASE REPORT

Cervical hemivertebra resection and torticollis correction: report on two cases and literature review

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Abstract

Purpose Hemivertebra of the cervical spine is a rare but complex spinal malformation. To our knowledge, only one publication describes excision of an upper–middle cervical (between C2 and C4) hemivertebra. We present our experience with two cases of C3 hemivertebra resection and torticollis correction via a combined anterior–posterior–anterior surgical approach and short segment fixation.

Methods Two 12-year-old patients with torticollis due to congenital C3 hemivertebra underwent surgery consisting of combined anterior vertebral body osteotomy, posterior element resection with segment instrumentation and deformity correction, and iliac bone graft reconstruction and fixation via an anterior approach. During the osteotomies, the transverse process accompanied with the vertebral artery was disconnected and freed away from the spinal column. Then the deformity was corrected without touching the vertebral artery, which made the procedure safe and comfortable. The details of this technique are presented. Pre- and postoperative radiographic features, as well as clinical outcomes were evaluated.

Results The treatment process was uneventful. The patients had satisfactory clinical outcomes at a mean of 1.5 years follow-up. Head tilt and chin rotation were corrected completely. Radiographs showed favorable deformity correction, well-balanced coronal and sagittal alignment, and solid bony fusion.

Conclusion Combined anterior–posterior–anterior hemivertebra resection with short segment instrumentation is a reasonable option for the treatment of congenital cervical hemivertebra, which provided satisfactory deformity correction and good clinical outcomes.

Level of evidence 4.

Keywords Congenital deformity · Cervical scoliosis · Torticollis · Hemivertebra resection · Surgical approach

Introduction

Torticollis in children presents clinically as head and neck tilt, and rotation of the chin to the opposite side. This condition has multiple etiologies. Congenital muscular contracture is most common [1, 2], but several non-muscular causes, such as vertebral anomalies, Klippel–Feil syndrome, ocular torticollis, and posterior fossa tumors, must be considered before treatment is initiated [3]. Most non-painful, non-muscular cases of torticollis are the result of osseous malformations, which consist of Klippel–Feil syndrome [4], occipitalization of the atlas [5], hemi-atlas [6], and congenital hemivertebra [7]. Neurological deficits due to the cervical congenital hemivertebra are rare in infants and juveniles. Hence, the "twisted neck" appearance in association with facial or eye asymmetry is not noticed until parents become concerned about it as a cosmetic problem.

Compared with its incidence in the thoracic and lumbar regions, hemivertebra in the cervical spine is rare. Deburge and Briard [8] and Winter and House [9] first reported this complex spinal malformation in 1981, and Ruf et al. [5] described it in more detail years later. Cervical hemivertebra also has been described as a part of other congenital anomalies, such as Goldenhar [10] and Klippel–Feil [4, 11] syndromes. The nature history of cervical hemivertebra is unclear. But a single fully segmented hemivertebra involved in thoracolumbar spine has a progression at 1°–3.5° per annum prepuberty [12, 13]. It tends to produce cosmetic

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deformity. Since conservative treatment of this congenital malformation cannot halt its relentless progression, resection of the cervical hemivertebra is a reasonable option. Indication of the surgery is radiographically deformity progression, or significantly cosmetic issue of head inclination and chin rotation (torticollis), facial muscle and eye asymmetry, or shoulder imbalance [8]. Torticollis is generally a major reason.

For surgical treatment, a combined anterior and posterior approach is necessary, due to the path of the vertebral artery. To our knowledge, the literature contains only one report of the excision of an upper–middle cervical (between C2 and C4) hemivertebra. Resection was performed using a posterior–anterior–posterior approach. In the first posterior approach, the author dissected the vertebral artery free along its course and retracted it laterally [5]. This procedure was difficult and had a risk of vertebral artery injury. Therefore, we developed a new strategy by disconnecting the transverse process and the foramen of the vertebral artery as a whole, away from the spinal column. Then we can correct the deformity without dissect the vertebral artery. We performed this surgery using an anterior–posterior–anterior approach. The details of this technique were presented in this study, which was applied in two cases of fully segmented C3 hemivertebra resection and torticollis correction.

Case reports

Case 1

A 12-year-old boy presented to our clinic with head and neck tilt and chin rotation, as well as flattening of the left face, since the age of 10 years. The deformity had progressed in the past year. He had no pain or neurological deficit, and his neck motion was normal. Physical examination showed that the patient's head and neck were tilted to the left and his chin was rotated to the right, with facial muscle and eye asymmetry. Plagiocephaly was not observed. The patient's left shoulder was slightly higher than his right shoulder. An X-ray illustrated a C3 hemivertebra with 15° head tilt to the left (Fig. 1a). Three-dimensional computed tomography (CT) confirmed a right-side fully segmented C3 hemivertebra, and well-segmented posterior C3 elements. The Cobb angle between C2 and C4 was 30°. CT angiography provided evidence that the right vertebral artery passed through the transverse process foramen of the hemivertebra (Fig. 1c). A rapid-prototyped full-scale three-dimensional (3D) model of the cervical spine showed details of the malformation; transverse process and transverse foramen; courses of the vertebral artery, internal carotid artery, and internal jugular vein; and adjacent structures (Fig. 2).

Indication for surgery was deformity progression and cosmetic problem. The surgical strategy was aimed at resecting the hemivertebra and reestablishing stability and cervical alignment through a safe and effective procedure. The plan included complete removal of the hemivertebral body, adjacent intervertebral discs, and posterior elements to correct the scoliosis. The surgery was performed under neuromonitoring with transcranial electrical motor evoked potentials (TCeMEPs) and somatosensory evoked potentials (SSEPs). With supine position, the anterior Robinson-Smith approach was adopted first to expose the C3 vertebra. The entire hemivertebral body, including the Luschka joint, and the Y-shaped discs above and below it were removed by Kerrison rongeur, curette, and ultrasonic bone scalpel. Then, the transverse foramen and transverse process was identified, but was left free. Wound was closed, and the patient was changed to prone position.

A posterior approach was performed using the same procedure. C2 pars screws and C3–4 lateral mass screws were used for segmental fixation. The lamina of the hemivertebra was removed and the spinal cord was exposed. Then, the lateral mass, facet joints of the hemivertebra, and adjacent facets were thinned and resected by Kerrison rongeur. The pedicle of the hemivertebra was burred out around the dural sac using an ultrasonic bone scalpel until the anterior vertebral defect was reached. The nerve roots were dissected freely. Then the transverse process and the transverse foramen, which gave passage to the vertebral artery, were disconnected completely from the spinal column using the ultrasonic bone scalpel (Fig. 3g). Hence, intentional exposure the foramen and dissection the vertebral artery was not necessary. The gap that was present after removal of the hemivertebra was then closed slowly by convex-side compression instrumentation and concave-side distraction. A posterior iliac bone graft was harvested for posterior fusion; a three-cortical autograft was also harvested from this approach, but reserved for anterior reconstruction. As the anterior gap generally cannot be closed completely, the patient was turned to a supine position again. The autograft harvested from the posterior approach was used for reconstruction. Instrumentation was used to fix the anterior column.

The overall operative time was 380 min, with blood loss of 220 ml. The treatment process was uneventful. The patient was extubated postoperatively and moved back to the general ward on the same day. Torticollis was fully corrected, and patient was discharged 1 week later. Postoperative X-rays showed perfect alignment in the coronal and sagittal planes (Fig. 3a, b). X-rays taken at the 2-year follow-up showed neutrality of the head (0° tilt), with normal flexion and extension movements (Fig. 3e, f). CT demonstrated solid bony fusion (Fig. 3d, e). The Cobb angle between C2 and C4 was 0°. Clinically, the patient's head posture was neutral and

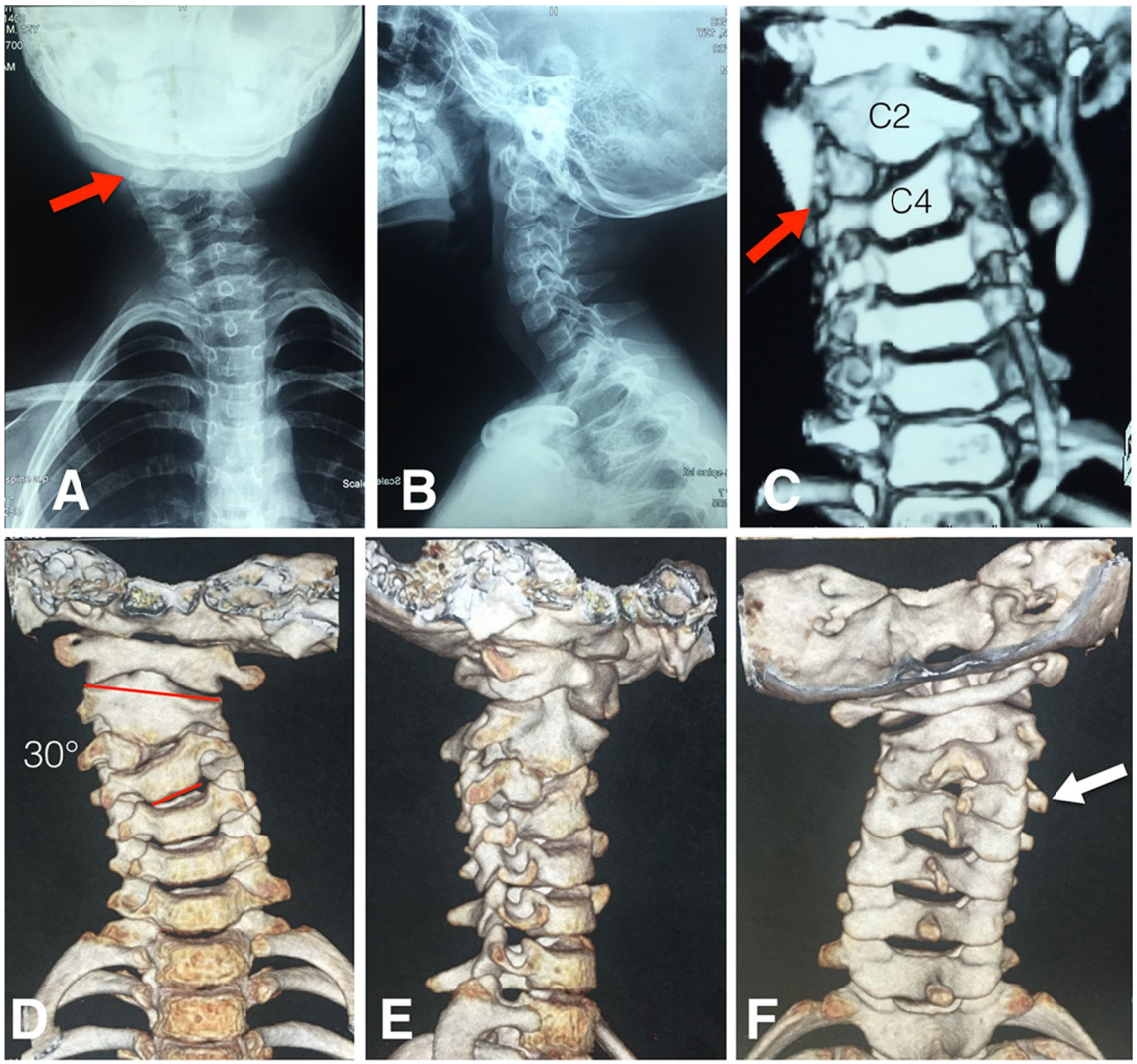

Fig. 1 Case 1, a 12-year-old boy with C3 hemivertebra. **a**, **b** Anteroposterior and lateral X-rays illustrating the C3 hemivertebra (red arrow) with 15° head tilt to the left. **c** Computed tomography (CT) angiographic image showing passage of the right vertebral artery through the transverse process foramen of the hemivertebra (red arrow). **d–f** Three-dimensional CT image showed a right-side fully segmented C3 hemivertebra (white arrow), with 30° scoliosis

his shoulders were balanced very well at the 2-year follow-up (Fig. 4).

Case 2

A 12-year-old girl presented with head tilt to the left side and chin rotation to the right side (Fig. 5a, b). She also had facial muscle and eye asymmetry. She reported no pain and had no neurological deficit. Her neck flexion and rotation were significantly limited. The patient's parents were very unhappy with the appearance of their daughter's head and neck. Indication for surgery was primarily a cosmetic problem. X-rays and CT illustrated a fully segmented C3 hemivertebra, with 12° head tilt and a 33° Cobb angle between C2 and C4 (Fig. 5c, d). In addition, the patient had multiple associated anomalies, including C5–T1 segment failure and T6 and T10 hemivertebrae (Fig. 5d, e). CT angiography showed that the right vertebral artery passed through the transverse process foramen of the C3 hemivertebra. Long-cassette anteroposterior and lateral X-rays of the full body showed major (45°) thoracic curvature, but good coronal and sagittal balance (Fig. 5f, g). Clinically, the patient's shoulder and trunk were also balanced well. Her parents were informed of the surgical plan to correct the torticollis by C3 hemivertebra resection, with observation of the cervicothoracic and thoracic curves.

Surgery was performed under intraoperative TCeMEP and SSEP neuromonitoring. With supine position, anterior C3 hemivertebra (including Luschka joint) resection and removal of the adjacent discs were performed first via

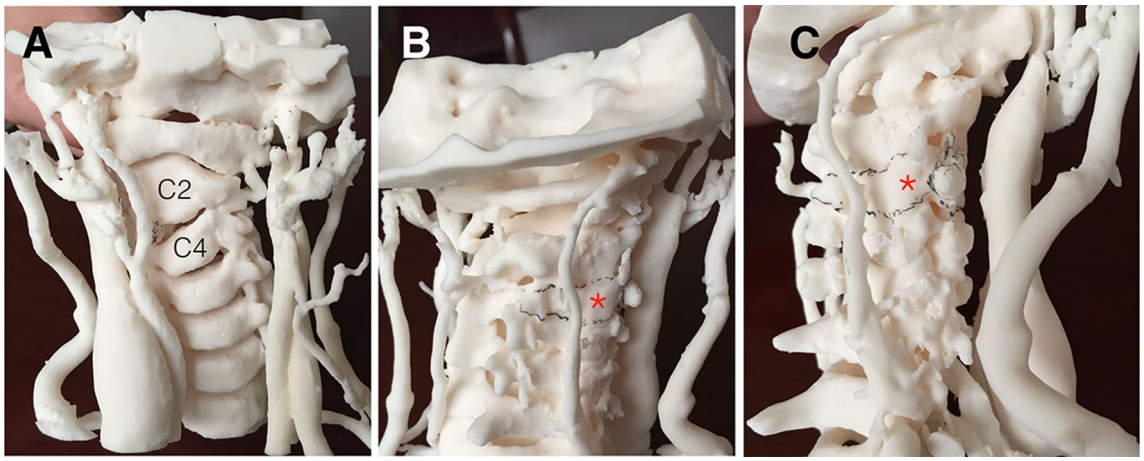

Fig. 2 **a** A rapid-prototyped three-dimensional model of the cervical spine in case 1 showing details of the hemivertebra between C2 and C4, beneath the carotid sheath. **b**, **c** Fully segmented posterior C3 elements (red asterisks) and transverse process

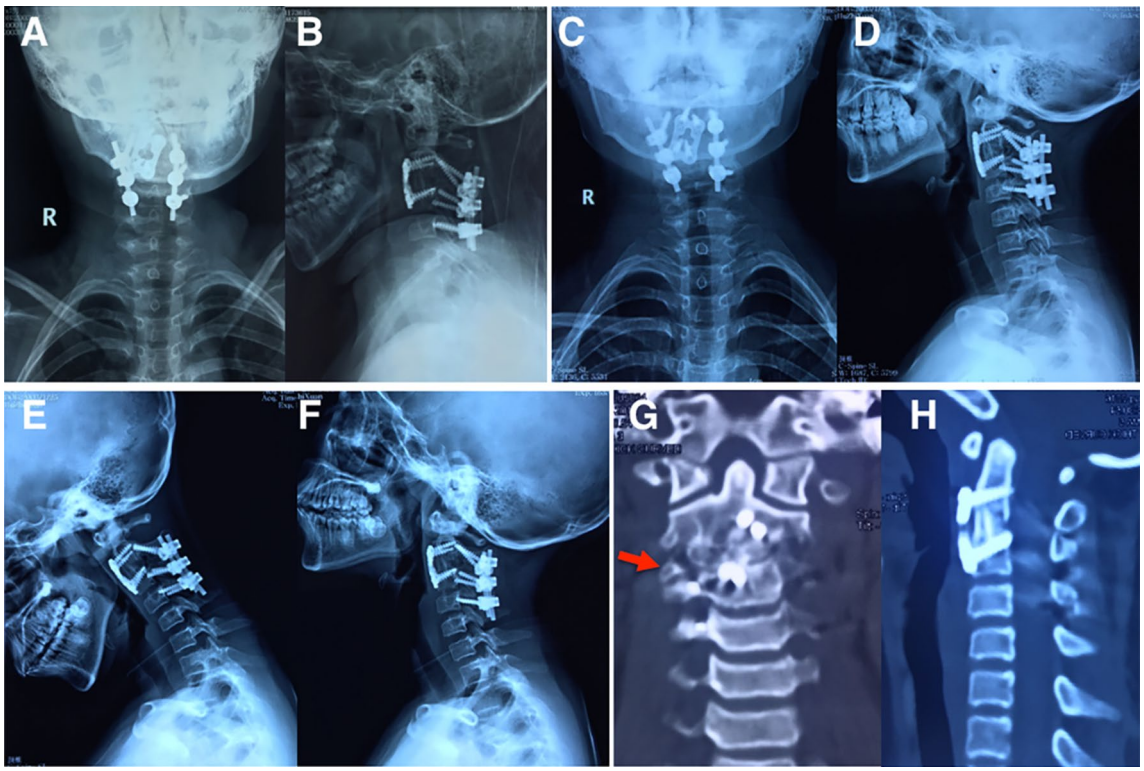

Fig. 3 Postoperative radiographs from case 1. **a**, **b** 1-week postoperative X-rays. **c**–**f** X-rays taken at the 2-year follow-up showing head neutrality (0° tilt), with normal flexion and extension movements. **g**, **h** Computed tomographic images demonstrating the sequestered transverse process and foramen (red arrow), and solid bony fusion between C2 and C4

the Robinson-Smith approach. The transverse process and transverse foramen were identified but left free. Then, the patient was turned to the prone position in the same anesthesia and a posterior approach was performed, including implant instrumentation and resection of the lamina, lateral mass, facet joints, and pedicle of the hemivertebra. Once the pedicle was burred out by an ultrasonic bone scalpel, the transverse process and foramen was disconnected freely from the spinal column (Fig. 6d); accordingly, dissection the vertebral artery was not necessary. Convex-side instrumentation compression and concave-side distraction were applied to correct the scoliosis. In this case, radical correction was not attempted. A slight (~ 10°) curve was maintained intentionally to compensate

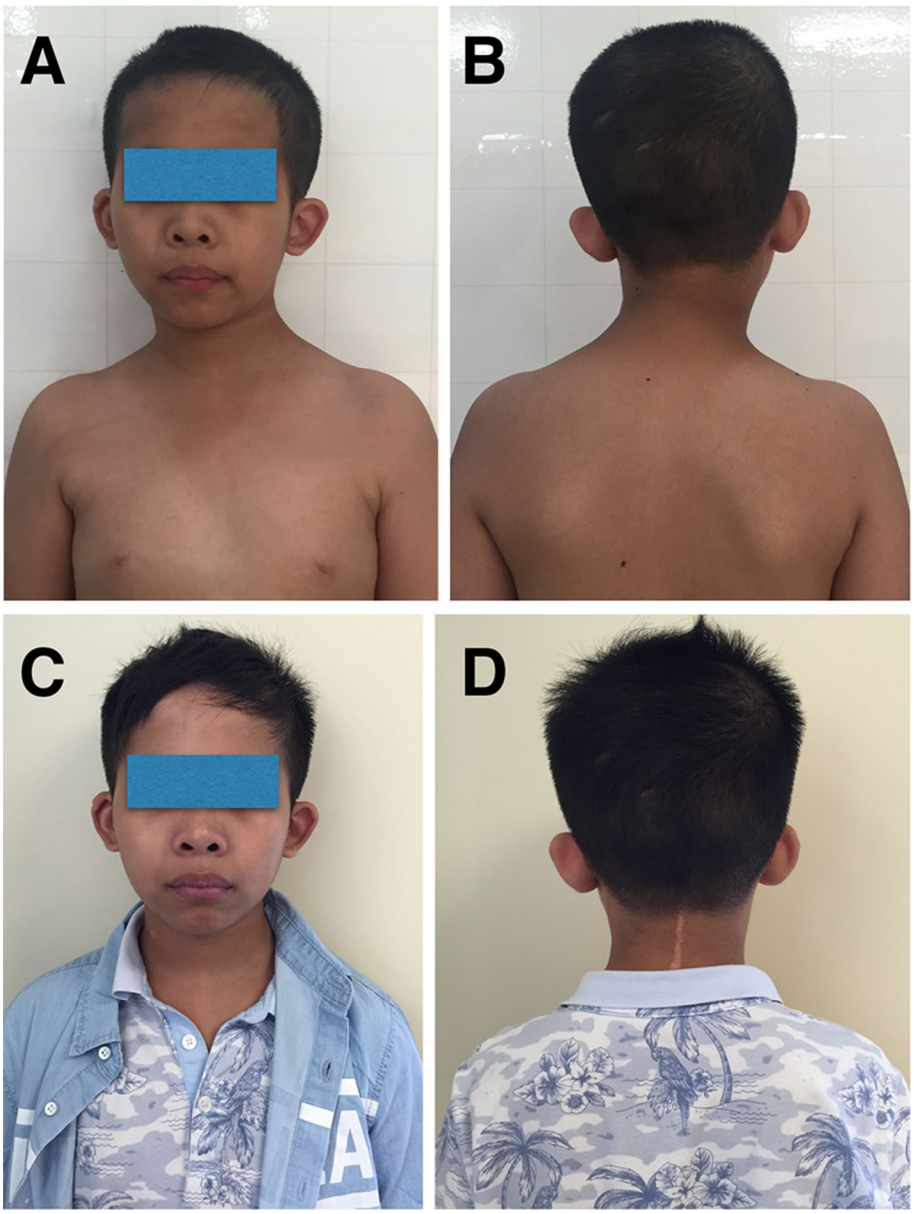

Fig. 4 Clinical images from case 1. **a**, **b** Preoperative clinical photographs. **c**, **d** Photographs taken at the 2-year follow-up, showing neutrality of the head and balance of the shoulders

the distal cervicothoracic and major thoracic curves. Finally, the patient was turned to the supine position again. Iliac bone graft reconstruction and fixation were performed using the previous anterior wound.

The overall operative time was 320 min, with blood loss of 180 ml. No complication was noted during treatment. The patient was extubated postoperatively, and the postoperative period was uneventful. The torticollis was corrected completely, with a satisfactory cosmetic result (Fig. 6a). Postoperative X-rays showed a neutral head posture (0° tilt) and normal sagittal alignment (Fig. 6b, c). At the 1-year follow-up, the patient was without complaints. CT and long-cassette X-rays demonstrated solid bony fusion between C2 and C4 (Cobb angle of 10°), and well-balanced coronal and sagittal alignment (Fig. 6d–g).

Discussion

Cervical hemivertebra deformity may have several sequelae, including shoulder imbalance, head inclination and chin rotation (torticollis), and facial muscle and eye asymmetry. Surgical indications for those patients are primarily cosmetic, and torticollis is a major reason. The goals of early surgery in children are the prevention of severe local deformities, development of second structure curves, correction of facial and eye asymmetry, and allowance of normal growth in the adjacent segment. Surgery in the cervical region requires special consideration of the osseous anomaly and neurovascular anatomy. To prepare the surgery, three-dimensional computed tomography and CT angiography are mandatory to identify the course of the vertebral artery and

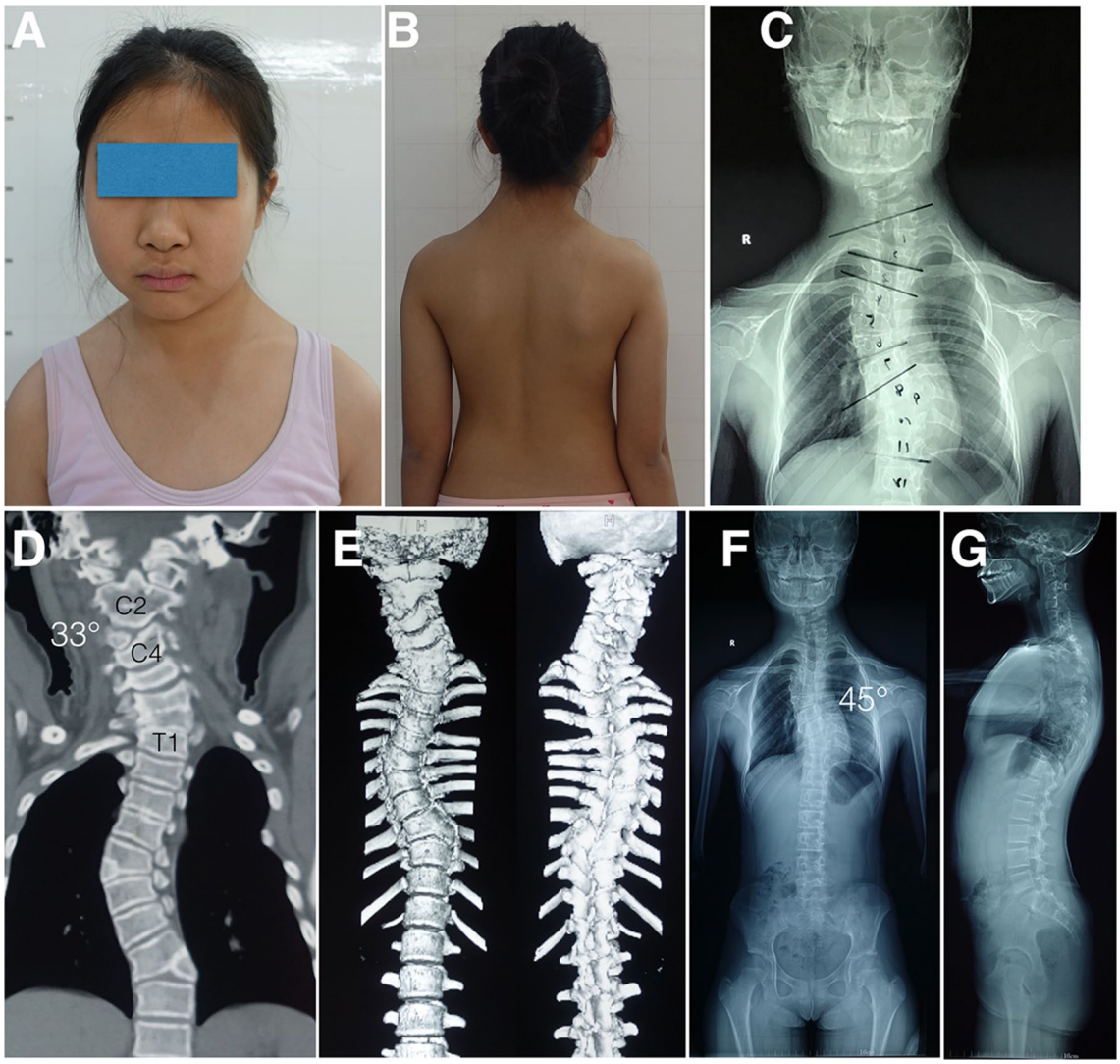

Fig. 5 Case 2, a 12-year-old girl with C3 hemivertebra. **a**, **b** Clinical photographs. **c** X-ray demonstrating the C3 hemivertebra, and major cervicothoracic and thoracic curvature. **d, e** Computed tomographic images showing a fully segmented C3 hemivertebra, and semisegmented T6 and T10 hemivertebrae. **f, g** Long-cassette anteroposterior and lateral X-rays showing a major (45°) thoracic curve

exclude any vascular variation. In these cases, a 3D-printed model is especially helpful for full characterization of the bone and vessel abnormalities.

In the first report on cervical hemivertebra resection, Deburge and Briard [8] presented the case of a 14-year-old girl with cervical-thoracic malformation, including Klippel–Feil syndrome with an associated C7 hemivertebra, block vertebra, spinal bifida, and cervical scoliosis. They used a staged anterior–posterior surgical approach to excise the C7 hemivertebra. Horner's syndrome appeared postoperatively, but then resolved fully. The deformity was corrected by manual reduction of the gap in a halo cast with the patient awake. Then, the third stage of instrumentation was performed thorough an anterior approach. Given the complex nature of the procedure and high risk of neurological deficits, Winter and House [9] and Smith [14] suggested decompression, long fusion without hemivertebra resection, and appropriate reconstruction for congenital cervical and cervicothoracic deformities. Ruf et al. [5] reported on hemivertebra resection and fusion of the adjacent vertebrae in three patients with torticollis due to cervical hemivertebra. They performed resection using a posterior–anterior(–posterior) approach. This was the first report describing the treatment of cervical hemivertebra using modern techniques. The gap that was present after complete resection of the hemivertebra was closed slowly by bending the head to the convex side. Good correction of the deformity and complete correction of the head tilt were achieved. Early operation is recommended in patients with proven or expected deterioration, to avoid the development of secondary changes such as

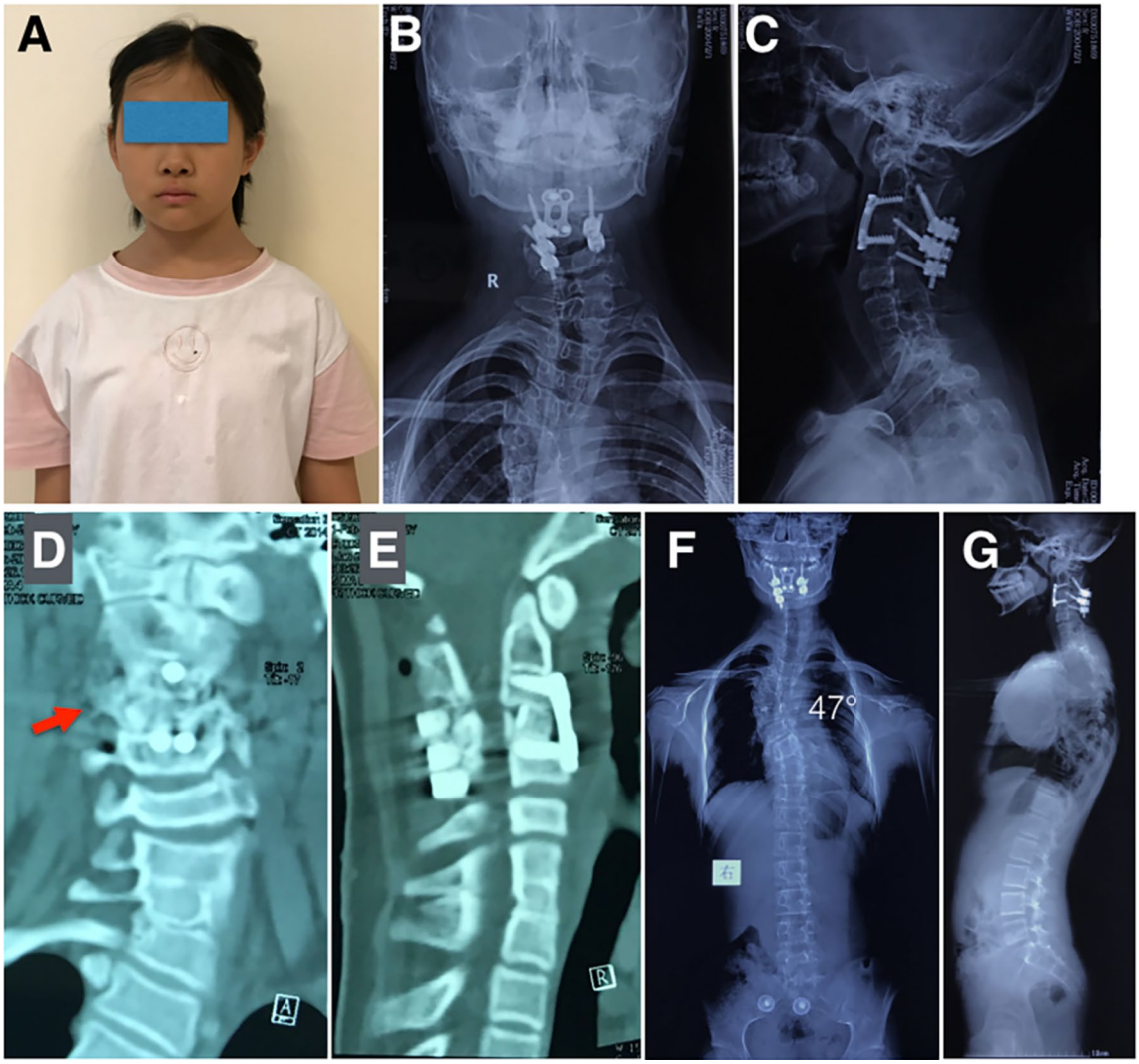

Fig. 6 Postoperative images from case 2. **a** Postoperative clinical photograph showing a neutral head posture. **b**, **c,** X-rays showing 0° head tilt. **d**, **e** Computed tomographic images obtained at the 1-year follow-up showed the remnant of transverse process and foramen (red arrow), and demonstrate solid bony fusion between C2 and C4. **f**, **g** Long-cassette X-rays showing the major (47°) thoracic curve, with good coronal and sagittal alignment

compensatory curves in the upper thoracic spine or asymmetry of the head [5].

Table 1 lists publications describing cervical hemivertebra excision (with the exception of one case [14] in which excision was not performed). In total, seven cases of cervical hemivertebra have been presented. The deformities were located at the craniocervical junction in one case [15], in the upper–middle cervical region (C2–C4) in three cases [5], and in the lower cervical region (C4–C7) in three cases [7, 8, 16, 17]. In the cervical spine, surgery is complicated by the anatomic situation and the course of the vertebral arteries. Thus, for hemivertebra resection, a combined anterior–posterior approach with carefully preparation of the spinal cord, the nerve roots, and the vertebral arteries is of great importance. Previous study reported the method of transverse foramen resection and vertebral artery dissection in the posterior approach [5]. The artery was protected and retracted laterally, which was helpful to excise the posterior part of the hemivertebra. However, expose the vertebral artery accompanied with significant bleeding and had a risk of artery injury. In our cases, we actually didn't treat the vertebral artery in the surgery. Instead of dissecting the artery, we disconnected the transverse process (which contains the artery) as a whole, and freed it away from the spinal column. Then we can correct the cervical deformity without touch the vertebral artery. The procedure is safe and comfortable. To our best knowledge, this is a new technique has never been reported before.

Another controversial is the sequence of surgical approach. Some authors proposed an initial posterior

Table 1 Publications describing cervical hemivertebra excision

Author, year	Patient's age (years)	Patient's sex	Hemivertebra location	Surgical approach
Deburge and Briard [8]	14	F	C7	Anterior-halo vest-posterior-halo vest-anterior
Ruf et al. [5]	4	F	C2a	Posterior–anterior
	8	M	C4	Posterior–anterior–posterior
	14	M	C3a, C7a	Posterior–anterior–posterior
Moghaddam et al. [14][a]	18	F	C6	Intradural lipoma excision at the C3 level
Ruf et al. [13]	42	F	C1–2	One-stage transoral, second-stage posterior
Zhuang et al. [7]	14	M	C4–6	Posterior–anterior–posterior
Otero-López et al. [15]	7	F	C4	Anterior

[a]Hemivertebra not removed in this case

approach to remove the posterior elements of the hemivertebra, then performed anterior procedures to completely excise the hemivertebra [5, 7]. As the resulting gap is difficult to close with anterior compression instrumentation, scoliosis was generally corrected by bending the head to the convex side. This maneuver, however, exerts indirect force on the cervical vertebra, which is not as strong as that achieved with direct compression and distraction by instrumentation. It is also associated with a high risk of neuro-element injury. Moreover, when the gap cannot be closed completely, additional posterior compression instrumentation is inevitable. Therefore, we've changed the sequence to an anterior–posterior–anterior approach in our patients. Following anterior–posterior complete hemivertebra resection, the gap can be closed easily and scoliosis can be corrected safely by posterior compression instrumentation on the convex side and distraction on the concave side. As strong anchors are needed before correction maneuvers are performed, we generally use a 4.0 × 20-mm pars screw in the C2 and a 3.5 × 16-mm lateral screw in the middle cervical region. The completeness of hemivertebra osteotomy and pedicle resection should be confirmed before closing the gap. Once the deformity has been corrected, the patient is turned to a supine position again to finish anterior bone graft reconstruction and fixation. With this technique, we achieved radiographic and clinical results comparable to those of Ruf et al. [5].

We encountered no complication in our cases. Neither patient required delayed extubation or was sent to the intensive care unit. No neurological deficit or evidence of instrumentation failure was observed at a mean of 1.5 years follow-up. However, complications may always occur. Surgeons should be aware that this operation is difficult and extensive. Thus, a detailed discussion with the family regarding the risks and benefits, as well as the natural history of the disease, is needed.

Conclusion

Little background information is available for this congenital condition. In our experience, combined anterior–posterior–anterior hemivertebra resection, disconnected the transverse process and foramen away to protect the vertebral artery, corrected the deformity with instrumentation and circumferential fixation is a safe and effective procedure that can restore cervical alignment and resolve the cosmetic problem. Circumferential hemivertebra excision, complete correction of the deformity, and solid fixation guarantee a good result.

Compliance with ethical statement

IRB approval statement This study was approved by the Institutional Review Board of The Second Xiangya Hospital, Central South University.

Conflict of interest The authors declare that they have no competing interests.

Ethical approval This study has been approved by the ethics and research committee of The Second Xiangya Hospital, Central South University.

Informed consent All the patients signed an informed consent prior to inclusion in this study.

References

1. Davids JR, Wenger DR, Mubarak SJ (1993) Congenital muscular torticollis: sequela of intrauterine or perinatal compartment syndrome. J Pediatr Orthop 13:141–147
2. Tatli B, Aydinli N, Caliskan M, Ozmen M, Bilir F, Acar G (2006) Congenital muscular torticollis: evaluation and classification. Pediatr Neurol 34:41–44. https://doi.org/10.1016/j.pediatrneurol.2005.06.010

3. Ballock RT, Song KM (1996) The prevalence of nonmuscular causes of torticollis in children. J Pediatr Orthop 16:500–504
4. Samartzis D, Kalluri P, Herman J, Lubicky JP, Shen FH (2011) Cervical scoliosis in the Klippel-Feil patient. Spine (Phila Pa 1976) 36:E1501–E1508. https://doi.org/10.1097/brs.0b013e31823145e4
5. Ruf M, Jensen R, Harms J (2005) Hemivertebra resection in the cervical spine. Spine (Phila Pa 1976) 30:380–385
6. Dubousset J (1986) Torticollis in children caused by congenital anomalies of the atlas. J Bone Jt Surg Am 68:178–188
7. Zhuang Q, Zhang J, Wang S, Guo J, Qiu G (2016) Multiple cervical hemivertebra resection and staged thoracic pedicle subtraction osteotomy in the treatment of complicated congenital scoliosis. Eur Spine J 25(Suppl 1):188–193. https://doi.org/10.1007/s00586-015-4352-9
8. Deburge A, Briard JL (1981) Cervical hemivertebra excision. J Bone Jt Surg Am 63:1335–1339
9. Winter RB, House JH (1981) Congenital cervical scoliosis with unilateral congenital nerve deficit in the upper extremity. Report of two cases. Spine (Phila Pa 1976) 6:341–346
10. Tsirikos AI, McMaster MJ (2006) Goldenhar-associated conditions (hemifacial microsomia) and congenital deformities of the spine. Spine (Phila Pa 1976) 31:E400–E407. https://doi.org/10.1097/01.brs.0000219876.45640.2a
11. Mesfin A, Bakhsh WR, Chuntarapas T, Riew KD (2016) Cervical scoliosis: clinical and radiographic outcomes. Glob Spine J 6:7–13. https://doi.org/10.1055/s-0035-1554776
12. Marks DS, Qaimkhani SA (2009) The natural history of congenital scoliosis and kyphosis. Spine (Phila Pa 1976) 34:1751–1755. https://doi.org/10.1097/brs.0b013e3181af1caf
13. McMaster MJ, Singh H (1999) Natural history of congenital kyphosis and kyphoscoliosis. A study of one hundred and twelve patients. J Bone Jt Surg Am 81:1367–1383
14. Smith MD (1994) Congenital scoliosis of the cervical or cervicothoracic spine. Orthop Clin N Am 25:301–310
15. Ruf M, Welk T, Merk HR, Smiszek FG, Pitzen T (2015) Resection of a hemivertebra within the craniocervical junction. Spine (Phila Pa 1976) 40:E1191–E1194. https://doi.org/10.1097/brs.0000000000001030
16. Moghaddam AM, Tanriover N, Ulu MO, Muhammedrezai S, Akar Z (2008) Cervical intradural lipoma with associated hemivertebra formation at C6 level: a case report. Turk Neurosurg 18:187–190
17. Otero-Lopez R, Rivero-Garvia M, Marquez-Rivas J, Valencia J (2016) Cervical subluxation associated with posterior cervical hemivertebra. Childs Nerv Syst 32:387–390. https://doi.org/10.1007/s00381-015-2833-0