Successful treatment of a patient with congenital kyphoscoliosis associated with tethered cord

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Tethered cord is a common finding in congenital scoliosis. The most frequently advocated approach for this condition is to perform prophylactic detethering of the cord before scoliosis corrective surgery. The authors report on a 14-year-old patient with congenital thoracic kyphoscoliosis associated with a tethered cord, who developed progressive paraparesis and was successfully treated by posterior spine shortening osteotomy alone without prophylactic untethering. The patient had a 103° scoliotic curve together with a 93° kyphotic curve with an apical vertebra of T-7. Furthermore, he developed a significant progression of neurological deficits, including weakness of both legs and urinary and bowel incontinence. Pre-operative MRI revealed that the spinal cord was entrapped by the apical vertebra and the low-placed conus medullaris was at approximately L-5. A posterior vertebral column resection of T-7 was performed for the purpose of simultaneously correcting the kyphoscoliosis and releasing tension on the tethered cord without a true detethering surgery. The patient's spinal cord function recovered completely from Frankel D to Frankel E by 6 months after the procedure. Evaluation at 31 months after surgery showed maintenance of good curve correction and normal neurological function.

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KEY WORDS kyphoscoliosis; tethered cord; spinal osteotomy; neurological deficit; surgical technique

CONGENITAL scoliosis is often associated with intra-spinal anomalies such as Chiari deformity, spinal cord malformation, tethered cord, and the like.^{2,25,31} Classically, tethered cord syndrome refers to a constellation of neurological signs and symptoms resulting from longitudinal traction on the caudal end of the spinal cord. A conus medullaris located below the level of L1–2 and/or the presence of a thickened filum terminale are the typical radiological diagnostic criteria.^{4,17,31} Surgical detethering is consistently reported to be effective in treating symptomatic tethered cord syndrome.^{3,5,15,30} However, complications such as cerebrospinal fluid leakage and neurological deterioration are also common.^{19,28} To minimize these complications, Kokubun pioneered an alternative surgical procedure for the treatment of tethered cord syndrome via spine-shortening vertebral osteotomy.¹³ This procedure has the distinct advantage of reducing neural tension indirectly without violating the dura. There have been several reports of the favorable results achieved by the use

of this procedure in patients with symptomatic tethered cord.^{8,9,11,12,15,21}

Spine-shortening vertebral osteotomy has also been described in the literature as an effective approach to correct severe kyphoscoliosis.^{16,22,29} Therefore, we hypothesized that a spine-shortening vertebral osteotomy alone could simultaneously treat both kyphoscoliosis and an associated tethered cord without necessitating a detethering procedure. We are unaware of any study in the literature that addresses the utility of spine-shortening vertebral osteotomy for both of these purposes concurrently, especially in patients with neurological deficits. In this case report, we present a patient with congenital kyphoscoliosis associated with tethered cord. The patient developed progressive paraparesis and underwent posterior vertebral column resection (PVCR) for the purpose of simultaneously correcting the kyphoscoliosis and releasing tension on the tethered cord without a true detethering surgery. He made a complete neurological recovery by 6 months

ABBREVIATIONS PVCR = posterior vertebral column resection; SSEP = somatosensory evoked potential.

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after the procedure, maintaining curve correction and normal neurological function at 31 months' follow-up.

Case Report

History and Presentation

This 14-year-old boy presented to Xijing Hospital with a 6-month history of progressive weakness in both lower extremities. He had a normal gestation and development with the exception of a small hump on his back at his birth without any other abnormal skin signs such as hairy patches, dermal sinus tracts, or skin appendages. At the age of 3, he was first formally diagnosed with scoliosis and was treated by brace therapy at an outside institution. The brace therapy was not effective in halting the curve progression and was discontinued when the patient was 5 years old. The patient continued to have worsening progression of the curve until presentation at 14 years of age. At his initial presentation to us, the right lower extremity was subjectively weaker than the left. He also was prone to falling when running or jumping. He had no back pain and denied any bladder or bowel dysfunction. On physical examination, he walked unaided but with faltering steps. A prominent hump was seen in the thoracic spine; there were no abnormal cutaneous manifestations. He had no apparent lower-extremity visible abnormalities such as atrophy of the leg or foot. He had global lower-extremity weakness with 4/5 strength on the right and 4+5 on the left. Deep tendon reflexes were slightly increased and the Babinski sign was present bilaterally. No significant sensory loss was detected over the affected areas including the perineum. Long-cassette standing radiographs showed that he had a thoracic kyphoscoliosis with a 103° scoliotic curve together with a 93° kyphotic curve with an apical vertebra of T-7 resulting from an unsegmented wedge-shaped vertebra from the T-6 to the T-8 level (Fig. 1). Both curves showed minimal flexibility on dynamic radiographs. The preoperative MRI study revealed that the spinal cord was entrapped by the apical vertebra and the low-placed conus medullaris was at approximately L-5 (Figs. 2 and 3). The family refused surgical intervention initially, and the patient came back 2 months later with significant progression of his neurological deficit. At this time he was not able to walk independently (Video 1) and had difficulty in raising his right ankle.

VIDEO 1. Clip from preoperative video showing that the patient had significant lower-extremity weakness and walked slowly and unsteadily with support. Copyright Hui-Ren Tao. Published with permission. Click here to view with Media Player. Click here to view with Quicktime.

He also developed urinary and bowel incontinence. Repeat neurological examination revealed significant decrease in muscle power of both legs: tibialis anterior and the extensor hallucis longus were 3/5 and the power of all the other muscles was 3/5 to 4/5. The sensation over the perineal region was also partially lost, but the cremasteric reflex was normal. The overall spinal cord function was classified as Frankel D. The family no longer hesitated to accept the operation at this point.

Operation

A posterior vertebral column shortening osteotomy of

FIG. 1. Preoperative anteroposterior (left) and lateral (right) standing scoliosis radiographs.

T-7 was planned to simultaneously relieve the compression of the spinal cord and decrease the tension on the cord, as well as correct the kyphoscoliosis. Because monitoring of motor evoked potentials was not available when the surgery was done in 2007, somatosensory evoked potential (SSEP) monitoring was performed in conjunction with multiple wake-up tests. A long midline posterior incision from T-1 to L-2 was made and we found that the vertebrae from T-6 to T-8 were fused together. Pedicle screws were placed at T-2, T-5, T-9, L-1, and L-2 on the left side and T-2, T-3, T-4, T-5, T-9, L-1, and L-2 on the right side by free-hand technique. Posterior vertebral column resection was performed at the T-7 vertebra. After removing the processes and laminae from T-6 to T-8, a total facetectomy from T-6 to T-8 was performed. The proximal portion of the seventh rib was removed bilaterally. The lateral wall of the fused T6–8 vertebral bodies was then dissected subperiosteally. After placement of a temporary rod, vertebral column resection was carried out at T-7 by decancellation of the vertebral body using a combination of curette, pituitary rongeur, and high-speed bur. After the osteotomy was finished, there was a 3-cm gap between the osteotomy surfaces. A compressive force was then imposed through the temporary rod on the convex side, while a mild distraction was effected on the concave side to close the osteotomy site while correcting the deformity. The temporary rod was then replaced by a contoured permanent rod on the convex side before placement of the definitive concave rod. Local bone grafting was performed using bone chips from resected ribs and

FIG. 2. Preoperative sagittal T2-weighted MR images showing the apical spinal cord entrapment by T-7 and the low-placed conus medullaris that was at the level of L4/5.

FIG. 3. Preoperative axial T2-weighted MR images showing the distortion of the cord (arrows) at the apex of the kyphotic curve.

vertebrae. No abnormalities in SSEPs were noticed and all the wake-up tests were unremarkable. The total surgical time was 8 hours, and the blood loss was 2700 ml.

Postoperative Course

The patient’s postoperative course was uneventful. He recovered independent walking ability as well as bladder and bowel function in 2 months. At 6 months after the operation, he had regained full muscle strength in his lower extremities and no longer had difficulty in walking, running, or jumping (Video 2).

VIDEO 2. Clip from video obtained 6 months postoperatively showing that the patient had regained full muscle strength in his lower extremities and no longer had difficulty walking. Copyright Hui-Ren Tao. Published with permission. [Click here to view with Media Player.](#) [Click here to view with Quicktime.](#)

At 31 months’ follow-up, he still had good neurological function without any complaint. His spinal cord function improved from Frankel D to Frankel E postoperatively. The postoperative radiographs showed that the scoliosis was corrected to 67° (correction rate, 54.4%) and the local kyphosis was corrected to 35° (correction rate, 62.4%) (Fig. 4). Postoperative MRI showed complete relief of apical spinal cord entrapment (Figs. 5 and 6), but the co-

FIG. 4. Postoperative anteroposterior (left) and lateral (right) standing scoliosis radiographs.

nus medullaris was still as low as before surgery (Fig. 5). Good curve correction and trunk balance were still evident at 31 months' follow-up (Fig. 7).

Discussion

It is controversial whether there is a need for preventive detethering in the treatment of kyphoscoliosis associated with tethered cord syndrome. Some surgeons consider it unnecessary to detether the cord before patients develop symptoms.^{18,27} However, other surgeons believe that the spinal cord will be put at high risk for injury when subjected to excessive stretching during surgical correction of spinal deformity without prior detethering. As a result, they recommend that a detethering procedure be performed followed by deformity correction 3–6 months later.¹ Despite some success with this 2-stage strategy, it has inherent disadvantages, such as additional trauma, more medical cost, and additional anesthetic exposure. Furthermore, retethering might occur 3–6 months later.^{23,24,32} Therefore, some authors advocate performing spinal cord detethering and deformity correction in one stage.^{7,20,26} Whether it is performed separately or as part of a single-stage operation, detethering does entail some risk of complications such as deterioration of neurological function and wound problems as well as incomplete detethering or retethering.²⁷ Matsu-moto et al. reported on a patient who had progressive kyphoscoliosis associated with tethered cord and underwent 2-stage corrective surgery. Unfortunately, the patient developed worsening neurological deficits after the untethering procedure, including paresthetic pain in the lower extremities and bladder and bowel dysfunction.¹⁹

FIG. 5. Postoperative sagittal T2-weighted MR images showing complete relief of apical spinal cord entrapment, with the conus medullaris (arrow) still as low as preoperatively.

The rationale behind performing detethering with the use of spine-shortening osteotomy is to reduce neural tension of the spinal cord via an indirect approach.^{13,14} In a cadaver model, Grande et al. found that a shortening vertebral column subtraction osteotomy at the thoracolumbar junction significantly reduced neural tension.⁶ There have also been several reports that show the successful treatment of tethered cord syndrome by spine-shortening osteotomy.^{8,9,11,12,21}

Spine-shortening osteotomy is a technically demanding procedure which has also been shown to effectively correct fixed kyphosis or kyphoscoliosis.^{16,19} Jalanko et

FIG. 6. Postoperative axial T2-weighted MR images showing complete relief of apical spinal cord compression (arrows).

FIG. 7. Preoperative and postoperative photographs demonstrating the patient's clinical improvement. Figure is available in color online only.

al. reported that the use of hemivertebrectomy alone in the thoracolumbar/lumbar region led to neurological recovery in 4 patients.¹⁰ But, there were no reports about the successful simultaneous treatment of tethered cord and scoliosis by single-stage posterior thoracic vertebral column resection. We have employed the principle of shortening the spine to simultaneously untether a cord and correct scoliosis since 2006. Regarding this case, entrapment of the spinal cord at the apical vertebra was the main cause of the patient's progressive paraparesis. Tethering increased the tension of the spinal cord and exacerbated the neural deficit. Surgical planning should be focused on relieving both the entrapment and tethering of the spinal cord. In this report, posterior vertebral column resection (PVCR) only, without any detethering procedure, was chosen and proved to be very effective in providing good correction of the curve while simultaneously restoring the function of the spinal cord owing to its indirect detethering effect. To our knowledge, this is the first case report in the literature to elucidate that PVCR alone is an effective procedure to combine untethering and corrective surgery while treating kyphoscoliosis associated with a tethered cord, even in the setting of significant neurological dysfunction.

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Author Contributions

Conception and design: Tao. Acquisition of data: Tao, Shen. Analysis and interpretation of data: Tao. Drafting the article: Tao, Yang, Fan, Zhang. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Tao. Administrative/technical/material support: Tao. Study supervision: Tao, Luo.

Supplemental Information

Videos

Video 1, Media Player. http://mfile.akamai.com/21490/wmv/digitalwbc.download.akamai.com/21492/wm.digitalsource-na-regional/spine13-528_video_1a.asx.

Video 1, Quicktime. http://mfile.akamai.com/21488/mov/digitalwbc.download.akamai.com/21492/qt.digitalsource-global/spine13-528_video_1a.mov.

Video 2, Media Player. http://mfile.akamai.com/21490/wmv/digitalwbc.download.akamai.com/21492/wm.digitalsource-na-regional/spine13-528_video_2a.asx.

Video 2, Quicktime. http://mfile.akamai.com/21488/mov/digitalwbc.download.akamai.com/21492/qt.digitalsource-global/spine13-528_video_2a.mov.

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