



Case Report

Total en-bloc spondylectomy for correcting congenital kyphosis

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Study design: A case report of congenital kyphosis corrected using a total en-bloc spondylectomy.

Objectives: To report a new surgical technique for the treatment of congenital kyphosis with myelopathy.

Setting: Department of Orthopedic Surgery, Akita University School of Medicine, Akita, Japan.

Methods: A 16-year-old boy who showed a 61° angular kyphosis and a 32° scoliosis from T6 to T9 due to the failure of the vertebral bodies formation in T7 and T8 was treated with a total en-bloc spondylectomy.

Results: The kyphosis was corrected to 26° (57.3%) and the scoliosis was corrected to 5° (84.4%) postoperatively. Three years postoperatively, no loss of correction has occurred and the patient has no complaints.

Conclusions: Total en-bloc spondylectomy is one of the useful surgical procedures for correction of congenital kyphosis Type I, with a high correction rate.

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Introduction

Congenital kyphosis, while uncommon, may compromise pulmonary function and cause paraplegia. Early operation is essential particularly in Type I (absence of vertebral bodies), which is highly progressive.^{1,2} Late treatment over age 5 is difficult and the results are usually less than ideal.^{2–5}

Winter^{2–5} reported many congenital kyphosis cases and recommended combined anterior and posterior fusion in late or with more deformity. However, anterior correction with a distraction force carries the risk of paralysis. Herbert⁶ was the first to describe a wedge osteotomy and fusion for congenital kyphosis in 1965. However, there is a limit to this procedure as regards safety and degree of correction. To overcome these problems, we introduced to the correction of congenital kyphosis the total en-bloc spondylectomy, which was designed primarily for spinal tumors. This is the first report of congenital kyphosis corrected using a total en-bloc spondylectomy.^{7,8}

Case report

A 16-year-old boy complained of back pain, numbness of bilateral lower extremities, and general fatigue. There were brisk tendon reflexes in his lower extremities and mild pulmonary dysfunction. Roentgenograms in standing position showed a 61° angular kyphosis (Figure 1a) and a 32° scoliosis from T6 to T9 due to the failure of the vertebral bodies formation (Type I) in T7 and T8. Severe tethering of the spinal cord was seen at the level of the apex of the kyphosis in the myelogram and MRI (Figure 1b).

Operative technique

En-bloc laminectomy Hypoplasia of the spinous processes at T9 and T10 was observed. The electrodes (IMC-KG-102, Inter Medical Co. Ltd., Japan) for spinal cord monitoring were set in the epidural space at T1 for stimulation of the spinal cord and in the subarachnoid space at T12 for recording of the spinal cord evoked potentials.

Bilateral T7–T9 transverse processes and the proximal ends of ribs 3 cm laterally to the costo-transverse joint were resected. We used a threadwire saw (T-saw) made of stainless-steel microcable with a diameter of approximately 0.5 mm, which produces an

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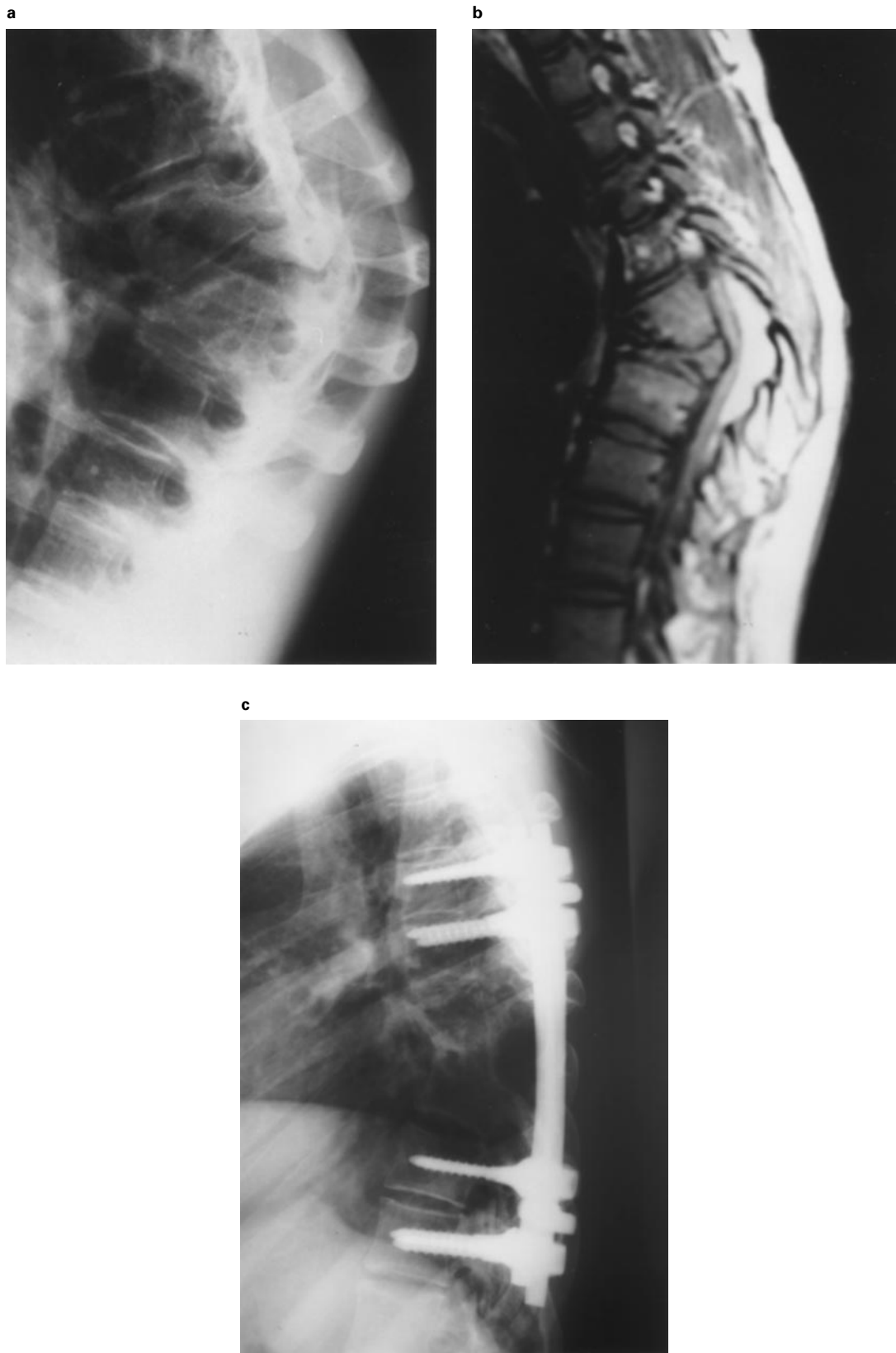


Figure 1 (a) Lateral tomogram, showing dysplasia of T7 and T8 vertebrae and angular kyphosis of 61°. (b) MR image, showing severe tethering of the spinal cord. (c) Plain lateral radiograph, showing the correction of kyphosis to 26° postoperatively

extremely thin and smooth cut with a diameter of 0.6 mm. The saw has a handle to cut the pedicle and the vertebra without injuring the dura or the spinal nerve root.⁹ After laminectomy of the lower half of the T6 lamina, the T-saw was passed through the upper and the lower intervertebral foramen around the pedicle from the spinal canal, pulled out to the lateral outside of the foramens, and clamped with a T-saw clamp at each end. With a reciprocating motion of the threadwire saw, the pedicles were cut at every vertebral segment, and then the entire posterior element from the T6 to the T8 (the spinous processes, the superior and inferior articular processes, and the pedicles) was removed in one piece. After en-bloc laminectomy, epidural vessels were coagulated thoroughly to control bleeding. To maintain stability after segmental resectioning of the anterior column, pedicle screwing using Cotrel-Dubouset (CD) pedicle screw system was performed from T4 to T11.

En-bloc corpectomy (resectioning of the anterior column of the deformed vertebrae) After the spinal branch of the segmental artery, (which runs along the nerve root) was coagulated using a bipolar coagulator, a blunt dissection of the segmental artery and vein around the vertebral body was performed bilaterally. The blunt dissection was done anteriorly on both sides through the plane between the segmental vessels with pleura and the vertebral body using fingers and a curved vertebral spatula. By continuing the dissection of both lateral sides of the vertebral body anteriorly, the aorta was simultaneously separated from the anterior aspect of the vertebral body with a spatula and the fingers. Two T-saws were inserted at the proximal and distal cutting levels of the lower end of the T6 vertebra and the upper end of the T9 vertebra. Using a cord spatula, the spinal cord was protected during the anterior osteotomy. Bilateral T7 nerve roots were cut to prevent spinal cord injury during the resectioning of the deformed vertebrae. The anterior column of the vertebra and the anterior and posterior longitudinal ligaments were carefully cut using the T-saw. However, the spinal cord evoked potentials fell suddenly flat after the cutting of the anterior column, due to the impingement of the spinal cord at the upper-posterior corner of the T9 by the anterior slippage of the pedicle screws. One side rod was inserted among the pedicle screws and correction of the slippage was performed immediately. After this procedure, the spinal cord evoked potentials quickly returned to normal. The freed anterior column was rotated around the spinal cord and removed carefully from the other side of the CD rod to avoid injury to the spinal cord (Figure 2). With this procedure, a complete anterior and posterior decompression of the spinal cord and total en-bloc resection of the vertebra were achieved.

Correction of the deformity, bone grafting, and posterior instrumentation The kyphosis was corrected through



Figure 2 Clinical photograph during surgery, showing removal of the freed anterior column

the gradual bending of the CD rods which were installed *in situ* on the deformed spinal column while observing the spinal cord evoked potentials. Local bone grafts using resected ribs were performed anteriorly between T6 to T9. After axial compression of the grafted bones, the kyphosis and scoliosis were again corrected with CD instrumentation. In addition, posterior spinal fusion from T4 to T11 using an autograft bone from the iliac crest was performed. The kyphosis was corrected to 26° (57.3%) and the scoliosis was corrected to 5° (84.4%) postoperatively (Figure 1c).

One week after the operation, standing and walking were allowed with a hard thoraco-lumbo-sacral orthosis. The orthosis was applied for 6 months. The patient acquired solid bony fusion and returned to normal activities after 6 months. Three years postoperatively, no loss of correction has occurred and the patient has no complaints.

Discussion

The best treatment for congenital kyphosis is early posterior fusion, before the curve reaches 50° and before 5 years of age. For curves greater than 50° and for patients older than 5 years, both anterior and posterior fusion are necessary.^{1-5,10}

Winter³ reported a review of 94 congenital kyphosis patients age 5 years or older, with 2 years or more follow-up in 77 of them. Combined anterior and posterior arthrodeseis gave consistently good results with an average correction of 38° (average 77° preoperatively), compared with only 21° (average 74° preoperatively) with posterior surgery alone. However, sufficient correction of the kyphosis could not be achieved even with combined anterior and posterior surgery because of the rigidity of the thorax. To achieve sufficient correction of angular kyphosis, bilateral rib resectioning at multiple levels and pushing the kyphotic spine from the posterior site to the anterior are necessary. Therefore, vertebral osteotomies and multiple rib resections from a posterior approach with posterior instrumentation have been performed and have achieved high correction rates.¹¹ Although such vertebral osteotomy provides more correction than ordinary procedures, there are problems of excessive bleeding with osteotomy in the vertebrae and a risk of spinal cord injury during osteotomy of the posterior wall of the vertebra in front of the spinal cord, especially at the apex of the kyphosis.

Total en-bloc spondylectomy that consists of a posterior en-bloc laminectomy after a bilateral pediculotomy and an en-bloc corpectomy followed by posterior instrumentation and fusion was designed primarily for patients with malignant or aggressive benign spinal tumors. Based on our experience with 10 cases of total en-bloc spondylectomy for malignant or aggressive benign spinal tumors, we have confirmed that this technique can be applied to the correction of congenital kyphosis. Total en-bloc spondylectomy for congenital kyphosis has these advantages: (1) high correction rates with removal of all elements of the anomaly, (2) safety following correction using only shortening force compared with anterior surgery with its distraction force, and (3) decreased bleeding and risk of spinal cord injury compared with vertebral osteotomy. However, total en-bloc osteotomy for congenital kyphosis has also some risks and the disadvantages: (1) pneumothorax, (2) loss of the posterior spinal column for bone grafting and stabilization, (3) injury of the segmental vessels during the blunt dissection of the anterior aspect of the vertebral body, (4) disturbance of spinal cord circulation at the level of surgery, and (5) mechanical damage to the spinal cord. To reduce the risk of nerve root and spinal cord damage, we used a thread wire saw, which has a smooth surface, to cut hard bony materials with minimal damage to the surrounding soft tissues.⁹ During dissection of the anterior aspect of the vertebral body, Kawahara *et al*¹² demonstrated that the artery must be carefully retracted anteriorly in areas caudal to T5 before manipulating the affected vertebrae. As for the circulation of the spinal cord, an abundant arterial network around the dura mater and the spinal cord may completely compensate for the ligation of one or two radicular arteries.¹³ In fact, no

neurologic complications have occurred in the spinal tumor patients undergoing total en-bloc spondylectomy in the Tomita series^{7,8} and ours.

According to Tian,¹¹ the indication of the posterior spinal osteotomy to kyphosis is from 45° to 90°. However, the limitations of total en bloc spondylectomy relating to the degree of kyphosis are still unknown. Even with severe angular kyphosis of more than 100°, total en-bloc spondylectomy is possible. However, the connection of the lordotic cephalad vertebrae to the lordotic caudal vertebrae could be difficult, and slippage of cephalad spine would occur, causing compression of the spinal cord by the supero-posterior edge of the caudal vertebra. To avoid this complication, the spine should be corrected to mild-long kyphosis in the thoracic spine by a broad posterior release of both the upper two and the caudal two levels including the facet joint and yellow ligament and be realigned with posterior instrumentation. In addition, one side fixation with a rod before complete cutting of the anterior column is essential to avoid spinal cord injury due to compression of the spinal cord at the supero-posterior edge of the caudal vertebra.

In conclusion, total en-bloc spondylectomy is one of the useful surgical procedures for correcting congenital kyphosis Type I, with a high correction rate.

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