Posterior Vertebral Column Resection (pVCR) for Severe Thoracolumbar Kyphosis in Patients With Achondroplasia

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Abstract

Study Design: Retrospective cohort study.

Objective: We aimed to evaluate the safety and validity of posterior vertebral column resection (pVCR) for severe thoracolumbar kyphosis (TLK) in the achondroplasia (ACH) patients.

Methods: Seven ACH patients (male: female = 6:1) who underwent pVCR procedures due to severe TLK from December 2008 to December 2017 in the authors’ hospital were included in this retrospective study. Their mean follow-up duration was 67 ± 35 months. Their clinical characteristics, radiologic characteristics, surgical characteristics and surgical complications were reviewed.

Results: A total of 8 vertebrae were removed with an average of 5 ± 2 levels of decompression and 9 ± 2 segments instrumented. The mean correction rates of TLKs and the main curves were 73 ± 15% and 87 ± 6%, respectively. Five patients (71%) had preoperative neurological symptoms with a mean Japanese Orthopedic Association (JOA) score of 8 ± 3 points. Their neurological functions were all improved, with a recovery rate of 78 ± 32% for the JOA score at the last follow-up. Four patients (57%) suffered from surgical complications, including rod breakages (43%), neurological complications (28%), dural tears (14%), cerebrospinal fluid leaks (14%) and proximal junction kyphosis (14%).

Conclusions: pVCR can offer a good correction for TLK and improve neurological function with extensive laminectomies in ACH patients. But the morbidity of surgical complications is relatively high. Therefore, it is a reserved surgical option for severe TLK in ACH patients by experienced spinal surgeons, especially with apical markedly hypoplastic vertebrae.

Keywords

thoracolumbar kyphosis (TLK), achondroplasia (ACH), posterior vertebral column resection (pVCR), complications

Introduction

Achondroplasia (ACH), first described by Parrott in 1878, is the most common skeletal dysplasia resulting in disproportionate short stature (dwarfism).¹,² The prevalence was reported to be approximately 1:25,000-1:30,000, affecting approximately 250,000 persons worldwide.²,⁴ ACH is caused by an autosomal dominant inherited mutation in the fibroblast growth factor receptor-3 (FGFR3) gene on the short arm of chromosome 4, arising from a mistranslation (Gly380Arg) in the FGFR3 protein, which affects the maturation of chondrocytes in the growth plate.⁵ The major clinical characteristics include rhizomelic shortening of the limbs, macrocephaly, and characteristic facial features with frontal bossing and midface retrusion, exaggerated lumbar lordosis, limitation of elbow extension and rotation, genu varum, brachydactyly, and trident appearance of the hands.⁵

In addition, the spinal columns of ACH patients are also often involved, manifesting thoracolumbar kyphosis (TLK),

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scoliosis and spinal stenosis. The prevalence of TLK and scoliosis were reported to be 50 ~ 79% and 60%, respectively, in the ACH patients, which is much higher than those in the general population. The prevalence of severe TLK and scoliosis were approximately 19% and 1%, respectively. Severe TLK will cause chronic pain, cosmetic deformity, difficulty sitting and lying flat, and even further aggravate the neurologic symptoms of spinal stenosis, including paresthesia, inability to walk, and neurologic incontinence. Therefore, surgical correction of TLK should be considered for fixed kyphosis with a Cobb angle above 30° to prevent further sequelae.

Vertebral column resection (VCR), first reported by MacLennan in 1922, is the most powerful surgical technique for severe spinal deformity. It can be performed through a combined anterior and posterior approach (apVCR) or a posterior approach alone (pVCR). To date, only 4 reported ACH cases have undergone VCR operations for severe TLK with short-term follow-up. In this study, we will evaluate the safety and effectiveness of the pVCR operation for severe fixed TLK in a larger series of ACH patients by a retrospective study in our institution.

Methods

Following approval by the institutional ethic review board (No. S-K1118), we conducted a retrospective review of a series of ACH patients who underwent pVCR surgeries for TLK between December 2008 and December 2017. Informed consent was obtained from all patients/parents. All ACH patients were diagnosed based on their characteristic clinical and radiographic findings according to Pauli’s suggestions (Figure 1). Genetic testing was only performed on voluntary patients. Referring to Khan’s study, TLK was defined as a kyphotic curvature of greater than 10° at the thoracolumbar junction with an apex between T11 and L2 on the lateral radiograph. The inclusion criteria included the following: ① patients with a diagnosis of ACH and TLK; ② patients who received pVCR surgical treatment; and ③ patients with at least 2 years of clinical follow-up. The exclusion criteria included the following: ① patients who could not be diagnosed with ACH or TLK; ② patients who only accept spinal canal decompression; ③ patients who only accepted other spinal osteotomy surgeries, such as Ponte’s osteotomy and pedicle subtraction osteotomy (PSO); ④ less than 2 years of follow-up.

Preoperative Evaluation

Clinical data, including age, sex, height and neurological symptoms (numbness of lower extremities, weakness of lower extremities and bladder dysfunction) was obtained through retrospective chart review. The preoperative Japanese Orthopedic Association (JOA) scores (11 points) based on the patients’ neurologic status of the spinal cord, including the motor function of the lower extremity (4 points), the sensory functions of trunk (2 points), the lower extremity (2 points), and the bladder (3 points), were also recorded. The Cobb angle of TLK was measured based on X-rays before surgery. A computed tomography (CT) scan was performed to check whether the apical vertebral body was markedly hypoplastic and the diameters of the vertebral pedicles were fit for pedicle screws. Magnetic resonance imaging (MRI) was performed to observe the extent of spinal cord compression.

Surgical Procedures

The surgical plans were decided by all spine experts in the spinal center based on the clinical symptoms, signs, and images of the patients. The indication of pVCR surgery was progressive fixed TLK of more than 40° with apical markedly hypoplastic vertebrae and/or significant neurological symptoms in ACH patients. The apical markedly hypoplastic vertebrae were removed to correct the kyphosis and restore the trunk balance. The range of the laminectomy was decided based on the symptoms and MRI. The fusion range was at least 3 vertebral above and below the resection vertebrae according to the spine deformity, containing the segments of spinal canal decompression.

The patient was placed in the prone position on a radiolucent operating table after general anesthesia. After the spine was subperiosteally exposed through a midline incision, the pedicle screws were implanted by free hand. The pVCR procedures were performed by the same spine surgeon as Suk’s description. In brief, the posterior elements (spinal process, lamina, transverse processes and facet joints) were first removed. For a thoracic vertebra, the rib heads were then resected, and the pleura was carefully detached from the vertebra. Finally, the pedicles, vertebral body, and adjacent discs were resected with an osteotome. A titanium mesh cage with autologous and allogenic bone was used to reconstruct the anterior column to avoid excessive shortening of the spinal cord. The combination of autologous and allogenic bone was also grafted to the posterolateral spine. Intraoperative neurophysiologic monitoring (IONM) of the spinal cord, including motor-evoked potentials (MEPs) elicited by multipulse transcranial stimulation of the motor cortex, was performed throughout the operation. A loss of 80% of the baseline amplitude in MEP was considered significant enough to alert the surgeon to a potential change in spinal cord function. Intraoperative blood cell salvage was routinely applied. All the patients were advised to wear plastic braces for more than 6 months after the operation.

Surgical data, including the location of the resected vertebrae, the decompression range, bleeding, allogeneic blood transfusion, changes of IONM and intraoperative complications, was retrospectively reviewed.

Postoperative Evaluation

All the ACH patients were followed up at 3, 6, and 12 months after the operation. CT scans were performed at 12 months after the operation to observe bone fusion. After that, the patients were routinely followed up every year. The symptoms, signs, JOA scores and X-rays were retrospectively reviewed.
Means, standard deviations, and proportions were used to describe the results. The recovery rate of JOA scores was calculated based on the preoperative and last follow-up scores according to Matsumoto’s report. The correction rate of TLK was calculated based on the preoperative and postoperative Cobb angles and the corresponding segmental standard values.
Results

General Characteristics

As shown in Table 1, a total of 7 ACH patients with severe fixed TLK, including 1 female and 6 male patients, were enrolled in this study. Their mean age was 21.9 ± 13.2 years/old. Two patients (Case 6 & 7) underwent genetic testing with the identification of the mutation in FGFR3 (c.1138G > A; p.Gly380Arg). Among them, there were 3 skeletally immature patients (Cases 1, 5 & 7) and 4 skeletally mature patients (Cases 2, 3, 4 & 6). The mean height was 124 ± 14 cm for the skeletally mature patients. The mean follow-up duration was 67 ± 35 months for all patients. A total of 9 vertebrae were resected with the means of 9 ± 2 fusion levels and 5 ± 2 decompression levels. Four of 7 patients (57\%) underwent extensive laminectomies (C2-15 levels) because of spinal canal stenosis. The average bleeding was 1800 ± 1000 ml. The average patient received allogeneic blood transfusions with 4 ± 2 U RBC and 629 ± 496 ml plasma.

Neurological Characteristics

As shown in Table 2, 71\% of patients (5/7) had preoperative neurological symptoms, mainly including numbness of the lower extremities (57\%, 4/7), weakness of the lower extremities (71\%, 5/7) and bladder dysfunction (43\%, 3/7). The mean JOA score was 8 ± 3 points before surgery. At the last follow-up, the mean JOA score was improved to 10 ± 2 points. For 5 patients with neurological defects, the average recovery rate of the JOA score was 78 ± 32\%. Two skeletally immature patients did not have any significant neurological symptoms, but they both had progressive TLKs even after brace treatment.

Corrections

As shown in Table 3, the mean Cobb angle of TLK was corrected to 31 ± 26° (2°-78°) immediately after surgery from 94 ± 42° (41°-142°) before surgery, yielding a mean correction rate of 73 ± 15\% (42%-96\%). In addition, there were 6 patients with mild to moderate scoliosis. After surgery, the mean Cobb angle of the major curves was also corrected to 4 ± 2° (2°-7°) from 31 ± 18° (11°-58°), yielding a mean correction rate of 87 ± 6\% (81%-94\%). There was no apparent loss of correction observed at the last follow-up.

Complications

There were 4 patients (57\%) with at least 1 operative complication (Table 3). The most common complications were rob

| Table 1. General Characteristics. |
|-----------------------------------|
| No. | Sex | Age (years/old) | Height (cm) | pVCR | Laminectomy (levels) | Fusion (levels) | Bleeding (ml) | RBC (U) | Plasma (ml) | Follow-up (months) |
|-----|-----|-----------------|-------------|------|----------------------|---------------|---------------|--------|--------------|--------------------|
| 1   | Female | 13 | 143 | T12 | 3 | 7 | 1300 | 4 | 400 | 130 |
| 2   | Male | 28 | 118 | T11&12 | 6 | 11 | 3000 | 4 | 800 | 85 |
| 3   | Male | 42 | 126 | L1 | 6 | 10 | 2000 | 6 | 600 | 72 |
| 4   | Male | 34 | 125 | L1&2 | 6 | 11 | 3000 | 8 | 1600 | 64 |
| 5   | Male | 14 | 127 | L1 | 5 | 7 | 1000 | 2 | 400 | 62 |
| 6   | Male | 17 | 130 | L1 | 3 | 10 | 2000 | 6 | 600 | 33 |
| 7   | Male | 5 | 96 | T12 | 3 | 8 | 300 | 1 | 0 | 24 |
| Mean | ± SD# | N/A* | 22 ± 13 | 126 ± 4\| | N/A* | 5 ± 2 | 9 ± 2 | 1800 ± 1008 | 4 ± 2 | 629 ± 496 | 67 ± 35 |

* N/A = Not available. # SD = standard deviation. \(^*\) For skeletally mature patients (No. 2, 3, 4 & 6).

| Table 2. Neurological Characteristics. |
|---------------------------------------|
| No. | Numbness of lower extremities | Weakness of lower extremities | Bladder dysfunction | JOA Score |
|-----|-----------------------------|-----------------------------|-------------------|----------|
|     | Preoperation | Last follow-up | Recovery rate (%) |          |
| 1   | No | Yes | Yes | 9 | 11 | 100 |
| 2   | Yes | Yes | Yes | 4 | 6 | 29 |
| 3   | Yes | Yes | No | 8 | 11 | 100 |
| 4   | Yes | Yes | Yes | 6 | 9 | 60 |
| 5   | No | No | No | 11 | 11 | N/A* |
| 6   | Yes | Yes | No | 9 | 11 | 100 |
| 7   | No | No | No | 11 | 11 | N/A* |
| Mean | ± SD# | N/A* | N/A* | 8 ± 3 | 10 ± 2 | 78 ± 32 |

* N/A = Not available.
breakages (3 patients, 43%), followed by neurological complications (2 patients, 28%), dural tears (1 patient, 14%), cerebrospinal fluid (CSF) leaks (1 patient, 14%) and proximal adding-on phenomenon (1 patient, 14%, Figure 2H&I). There were 2 patients (Cases 2 & 6) with more than 1 complication. Postoperative CSF leak was secondary to intraoperative dural tear (Case 2). Wound drainage was removed after keeping in a flat position for 4 days.

Both 2 patients with many complications had neurological complications. One patient had decreased muscle strength for the lower extremities from Grade IV to Grade III immediately after the operation but recovered to the preoperative level at 7 days after the operation and Grade V at the last follow-up (Case 6). His JOA score improved to 11 points from 9 points at the last follow-up. The other patient (Case 2, Figures 2&3) lost motor evoked potential (MEP) signals when closing the

Table 3. Correction of Spinal Deformity and Complications.

| No. | Cobb Angle of TLK (°) | Correction Rate (%) | Last Follow-up | Cobb Angle of Major Curve (°) | Correction Rate (%) | Last Follow-up | Complications |
|-----|-----------------------|---------------------|---------------|-----------------------------|---------------------|---------------|--------------|
| 1   | Preoperation | Postoperation | | Preoperation | Postoperation | | |
| 2   | 41         | 5               | 96            | 6               | N/A*         | N/A*         | No           |
| 3   | 97         | 28              | 71            | 29              | 11           | 2             | CSF# leak after operation |
| 4   | 142        | 78              | 42            | 75              | 16           | 3             | Aggravated neurological dysfunction after operation and recovery to preoperative level at 1 year after operation |
| 5   | 42         | 2               | 89            | 3               | 27           | 3             | Rod breakage at 1 year after operation |
| 6   | 132        | 28              | 73            | 29              | 58           | 4             | Rod breakage at 1 year after operation |
| 7   | 79         | 26              | 73            | 26              | 44           | 7             | Rod breakage at 1.5 years after operation |
| 8   | N/A        | N/A             | N/A*          | N/A*            | N/A*         | N/A*         | No           |
| Mean ± SD# | 94 ± 42 | 31 ± 26 | 73 ± 15 | 30 ± 24 | 31 ± 18 | 4 ± 2 | 87 ± 6 | 4 ± 2 | PJK* at 2 years after operation |

*N/A = not available; #CSF = cerebrospinal fluid; *PJK = proximal junction kyphosis.
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osteotomy gap because the spinal cord was compressed by the lamina. The MEP signals reappeared at approximately 1 hour after stopping the correction and removing more lamina. His muscle strength of the lower extremity decreased to Grade 0 immediately after the operation from preoperative Grade III with a JOA score of 2 points (4 points before the operation). He recovered to be able to walk with the support of his relatives and urinate by himself after rehabilitation training for 1 year. The JOA score improved to 6 points (recovery rate = 29%) at the last follow-up. To date, no patient has undergone a revision surgery due to neurologic complications.

All 3 rod breakages (75%) occurred in the skeletally mature patients (4 patients) at 12-18 months after surgery. Fortunately, no patient suffered from aggregated neurological dysfunction secondary to the rod breakage. Then, every patient underwent 1 revision surgery. To date, no more rod breakage has occurred again in these patients.

Discussions

Endochondral ossification, which is impaired because of FGFR3-activating mutations in ACH patients, plays an important role in the development of the fetal spine from the thoracolumbar junction to crania and cauda [31, 32]. Three ossification centers are separately located in the vertebral centrum and the anterior parts of both pedicles. Therefore, the vertebral size and the cross area of the spinal canal will be affected in ACH patients, especially for the thoracolumbar junction. TLK, shorter pedicles and smaller spinal canals are characteristic features of ACH patients.5,21

Figure 2. Images of a 28-year-old male patient with thoracolumbar kyphosis for achondroplasia (Case 2). He underwent a pVCR procedure at T11 and T12 with the decompression level of T10-L3 and the fusion level of T5-L3. A&B: Preoperative coronal and sagittal radiographs show that the Cobb’s angles are 123° and 32° for kyphosis and scoliosis, respectively; C: preoperative sagittal MRI image shows the compression of spinal cord at the apex; D&E: postoperative coronal and sagittal radiographs show that the Cobb’s angles are 48° and 2° with correction rates of 64% and 94% for kyphosis and scoliosis, respectively; F&G: coronal and sagittal radiographs at 1 year after operation show both rod breakages; H&I: coronal and sagittal radiographs immediately after revision surgery; J&K: coronal and sagittal radiographs at last follow-up.
Most infantile TLK cases can be spontaneously resolved after they can stand and ambulate, decreasing from 94% in infantile patients (<1 year old) to 11% in children patients (5-10 years old). Thus, early brace treatments are first suggested for these infantile and children ACH patients with TLKs of more than 25° – 30° or the early sign of vertebral wedging. When the patients enter adolescence, these unresolved TLKs will often be fixed. Ten to 15 percent of TLKs will persist and even progress into severe fixed TLKs without the appropriate treatments. The risk factors of progressive TLKs include developmental motor delay, apical vertebral translation, and apical vertebral wedging. Otherwise, the spinal cord or cauda equina in the abnormally narrowed spinal canal will be further compressed by progressive TLK to cause neurologic sequelae. Degenerative spondylosis and ligamentum flavum hypertrophy will also aggravate neurologic sequelae in adult ACH patients. Surgical interventions should be considered if the patients have progressive TLKs or significant neurological symptoms. In this study, most ACH patients (71%) had neurological symptoms with an average JOA score of 8 points and 100% for adult patients. Only 2 minor patients (29%) underwent operations because of progressive TLKs even with brace treatment.

There are 10 ~ 25% ACH patients with spinal stenosis, who require surgical treatment for neurological progression. Laminectomy is first suggested to decompress the spinal canal for TLK and spinal stenosis and in ACH patients. However, simple laminectomy may further increase spinal instability and aggregate TLK. Although the anatomies of pedicles in ACH patients, which are shorter and thicker, are different from those in the general population, the safety of pedicle instrumentation has been confirmed with the developments of pedicle screws, pedicle morphological study and surgical techniques (navigation and others) in the last 2 decades. In this study, no pedicle screw-related complications were observed in these 7 ACH patients. Therefore, posterior fixations via pedicles were suggested after decompression by surgeons.

Failure to correct severe TLK will likely impede neurological functional recovery and cosmetic improvement and cause long-term deformity progression. Thus, an anterior release is once suggested with the combination of posterior decompression and fixation to correct the deformity if the Cobb angle of TLK is more than 50°. However, there is a potential risk of elongated and compressed spinal cord. With the development of spinal osteotomy, pedicle subtraction osteotomies (PSO) and pVCR have been advocated to treat severe TLK in the ACH patients. pVCR is limited for the TLK of more than 40° with apical markedly hypoplastic vertebrae. Therefore, these 7 patients underwent pVCR in this study.
pVCR can provide the most powerful correction for severe spinal deformity. It creates a segmental spinal defect to separate the spinal columns into 2 limbs completely.\textsuperscript{32} Spinal surgeons can handle anterior and posterior column to perform a 3-D spinal correction at the same time, including shaft and rotation.\textsuperscript{33} In these ACH patients, the correction rates were 73\% and 87\% for TLK and the major curve, respectively. They were as good as those reported before, 63\% and 61\% for the kyphosis and scoliosis, respectively.\textsuperscript{34}

pVCR is a 3-column circumferential vertebral osteotomy and that can achieve a 360\degree decompression of the spinal cord at the apex. Symptomatic spinal stenosis often occurs in the thoracolumbar spine and lumbar spine, because the cross-sectional area of the spinal canal in ACH patients is reduced by 27\%-50\%, decreasing from T12 to L4 and is exacerbated by TLK or lumbar hyperlordosis.\textsuperscript{25,30,35,36} Thus, the range of laminectomy should be more extensive than normal pVCR procedures to decompress spinal stenosis at the same time (decompression levels $\geq$5: 57\% of patients). If not sufficiently decompressed, a disastrous neurological complication might occur when closing the osteotomy gap to correct the deformity (Case 2). Conversely, pVCR combined with extensive laminectomies could offer good relief of neurological symptoms. The preoperative neurological symptoms of all these 5 patients (100\%) were alleviated to some degree with the improvement of the JOA score (recovery rate = 78\%). Otherwise, the recovery rates of the patients with higher JOA scores (Cases 1, 3 & 6) were significantly higher than those of the patients with lower JOA scores (Cases 2 & 4). A similar result could also be observed in Matsumoto’s report.\textsuperscript{15} It suggested that surgical intervention should be performed as soon as possible after the onset of neurological symptoms.

Although pVCR could offer a good clinical effect for ACH patients with TLK, there was a high risk of surgical complications (57\%). The morbidity of complications was similar to those for pVCR in the general population (14\%–54\%) and laminectomy in ACH patients (61\%).\textsuperscript{34,37}

Neurological complications are still a major and catastrophic complication of pVCR. The reported independent risk factors included preoperative neurologic dysfunction, intraspinal anomalies, scoliosis with thoracic hyperkyphosis and multiple vertebrae resection.\textsuperscript{38} In this series of patients, most had preoperative neurologic dysfunctions (71\%). Their spinal cord was thick relative to the contractile spinal canal in the ACH patients, which requires more laminectomy to avoid compressing the spinal cord by the lamina during spinal cord shortening. Both 2 patients with neurological complications had severe TLK and apparent scoliosis. These reasons explained why the morbidity of neurological complications in this study (28\%) was higher than that in a previous report (8\%).\textsuperscript{35} IONM was very useful for detecting changes in spinal cord function promptly to prevent permanent spinal cord injury, such as in Case 2.

Rod breakage was another major complication in this study (43\%). They all occurred in skeletally mature patients at 12-18 months before achieving osseous fusion. As we reported before, ACH is an independent risk factor for rod breakage for pVCR in adult deformity patients.\textsuperscript{39} Impaired endochondral ossification might influence bone formation in ACH patients. The lack of anterior support or fusion was the most important reason for instrumentation failures. Thus, the important bone grafts should be emphasized, especially for the anterior column and the application of bone morphogenetic proteins (BMPs) should be encouraged if available. A multi-rod construct can reduce the morbidity of rod breakage and effectively.\textsuperscript{40} It should also be considered for these ACH patients. Different from normal pVCR procedures, more lamina might be removed because of spinal stenosis in the ACH patients, which would further decrease the stability of the posterior column and increase difficulty in posterolateral fusion outside the osteotomy area. This might be the reason why the rods were still broken outside the osteotomy area in case 6 (Figure 1) when using 2 short satellite rods. Therefore, we suggested that the lengths of the satellite rods should be at least longer than the range of the laminectomy.

Dural tears are another common complication in ACH patients. There are several reasons to explain it, including extensive laminectomies, exceedingly thin and friable dura mater, decreased epidural fat and dense adhesion to the hyperplastic ligamentum flavum.\textsuperscript{25} In addition, these risk factors would be aggravated at the apex by severe kyphosis. Therefore, the laminae should be removed very carefully, especially at the apex. Thinning the laminae first was advised to expose the ligamentum flavum.\textsuperscript{11}

Conclusions

pVCR can offer a good correction for TLK and improved neurological function in combination with extensive laminectomies in ACH patients. However, the morbidity of surgical complications for pVCR is relatively high, including rod breakages, neurological complications and dural tears. Fortunately, disastrous complications can be controlled with IONM by experienced spinal surgeons. Therefore, pVCR is a reserved surgical option for severe TLK in ACH patients, especially with apical markedly hypoplastic vertebrae. Long satellite rods were suggested for use in adult ACH patients.

Declaration of Conflicting Interests

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