Expert's comment concerning Grand Rounds case entitled “Surgical treatment of a 180° thoracolumbar fixed kyphosis in a young achondroplastic patient: a one stage ‘in situ’ combined fusion and spinal cord translocation” (by J. C. Aurégan, T. Odent, M. Zerah, J.-P. Padovani and C. Glorion)

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Abstract

An expert comment is provided for the case of an 18-year-old male achondroplastic patient with a severe thoracolumbar kyphosis and spinal stenosis managed with a five level hemilaminotomy, a decancellation osteotomy of the three apical vertebrae and circumferential fusion. A review incidence, presenting symptoms and treatment options for thoracolumbar kyphosis in adults with achondroplasia, is provided.

Keywords Achondroplasia · Spinal stenosis · Kyphosis · Osteotomy

The authors present a case of an 18-year-old male achondroplastic patient with a severe thoracolumbar kyphosis (TLK) who developed progressive neurogenic claudication of the lower extremities. An MRI demonstrated congenital narrowing of the spinal canal and abnormal spinal cord signal intensity at the apex of the kyphosis. The patient was managed with a five level hemilaminotomy, a decancellation osteotomy of the three apical vertebrae, circumferential fusion with placement of a cast for 3 months followed by a TLSO for 6 months. At 3-year follow-up, the claudication symptoms had resolved. There was no worsening of the TLK on radiographic imaging. The patient's only complaint was of violent electric shock sensations in the lower limbs with external sudden pressure on the posterior thoracic region in the area without bony covering.

Spinal disorders are quite common in persons with achondroplasia. In a recent study of 437 adults with achondroplasia, 41% reported chronic back problems, 30% reported having a spinal deformity, and 20% noted weakness in upper or lower limbs [9]. The development of TLK is a common deformity in achondroplasia [3]. The incidence of TLK has been reported to be as high as 94% in patients less than 1 year of age, gradually declines to 11% by 10 years of age, and then increases 35% by 30–50 years age [3, 5] Approximately, 15% of adults with achondroplasia will have a fixed kyphosis at the thoracolumbar junction of sufficient severity to result in neurological symptoms [3, 5, 8, 10].

Traditionally, the surgical treatment of both spinal stenosis and TLK in patients with achondroplasia has emphasized spinal canal decompression by laminectomy. Both active deformity correction and the used of posterior spinal instrumentation were discouraged due to the risk of neurological injury [8, 10, 13]. Surgically important differences in the different angles and diameters of thoracic

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and lumbar pedicles of patients with achondroplasia compared with those of healthy people have been defined [6]. Despite the anatomic differences, more recent series describing management of spinal disorders in patients with achondroplasia have demonstrated the safety and efficacy of spinal instrumentation [1, 4].

Laminectomy without fusion has historically been used to manage spinal stenosis in patients with achondroplasia neurological deficits. The development of post-laminectomy kyphosis is increasingly recognized to be a consequence of posterior spinal decompression, especially in the skeletally mature patient [2]. Unlike the spinal stenosis commonly encountered in the elderly, the spinal stenosis associated with TLK in achondroplasia has the principle spinal compression on ventral aspect of the spinal cord. Laminectomy alone frequently fails to address the cause of neural impingement in these patients. Either laminectomy alone or laminectomy and noninstrumented spinal fusion risk a progression of kyphotic deformity. Increased kyphosis can exacerbate spinal cord compression or increase traction and strain within the spinal cord. Both Qi and Sarlak [11, 12] have recently reported direct resection of the apex of deformity in TLK cases using thoracic pedicle subtraction or vertebral column resection techniques and spinal instrumentation with improvement of neurological symptoms. Despite the recognized technical demands of these surgical procedures, the neurological and functional outcomes were excellent. Resection of the apex of the kyphosis with correction of the sagittal deformity with spinal instrumentation has the additional benefits of improving cosmesis and reducing lumbar hyperlordosis that can both exacerbate lumbar spinal canal stenosis and be a contributing factor for chronic lower back pain. Spinal instrumentation can frequently obviate the need for casting or bracing following surgery. Casting and bracing with a severe kyphosis risks skin compromise in the area of maximal deformity.

The authors describe the case of an 18 year old who initially presented with aesthetic concerns related to the spinal deformity that subsequently developed neurological symptoms. Any spinal decompression and fusion procedure for the management of TLK with neurological symptoms should ideally resolve the signs and symptoms caused by neurological compromise, achieve a solid fusion, improve functional and cosmetic outcome and limit the long-term consequences of the procedure such as progression of deformity above or below the site of fusion.

The surgical procedure reported used a mix of both newer and more traditional surgical techniques. In this reviewer's experience, vertebral column resection or decancellation osteotomy is the appropriate technique for management of severe TLK in the achondroplastic patient with neurological symptoms. The posterolateral costovertebral approach

permits a more direct circumferential decompression of the spinal cord compared with anterior transthoracic approaches. The use of a unilateral approach as performed in this report can risk inadequate decompression of the entire spinal cord compared with a circumferential vertebral column resection technique. A bilateral approach for vertebral resection/decancellation provides more predictable results in achieving spinal decompression and deformity correction [7, 14].

There are several well-recognized intraoperative risks with thoracic osteotomy procedures including vertebral column displacement at the osteotomy site and excessive shortening of the spinal cord resulting in the risk for neurological compromise. The use of pedicle screws and a temporary rod permits much greater control while both performing the osteotomy and during the active correction of the spinal deformity. If a concern that neurological compromise is occurring on neurophysiological monitoring, the ability to correct any spinal displacement or reduce correction forces are better with spinal instrumentation in place. In the postoperative period, the danger of spinal column displacement, dislodgement or subsidence of the strut graft or loss of spinal alignment are lower with the presence of spinal instrumentation. Despite the smaller tolerances for accurate pedicle screw placement in the achondroplastic patient, the benefits of screw placement generally outweigh the risks. Prominence of spinal instrumentation can be a concern in any patients with compromised skin quality but a substantial correction of the TLK usually leaves abundant tissue of implant coverage.

In the case presented, the primary goal of improving the neurological signs and symptoms was achieved without complication. The failure to obtain any improvement the severe TLK deformity will likely diminish secondary aspects of the procedure such as functional recovery, limit cosmetic improvement and long-term deformity progression through changes at adjacent segments. This case demonstrates the balance between a more conservative surgical approach performed because of concerns about catastrophic complications and a more aggressive approach with the potential for better results but at a higher risk. Larger series with objective pain, functional and cosmetic outcome measures are needed to better define the best approach in this patient population.

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