Eur Spine J (2010) 19:1807–1811
DOI 10.1007/s00586-010-1530-7

GRAND ROUNDS

Surgical treatment of a 180° thoracolumbar fixed kyphosis in a young achondroplastic patient: a one-stage "in situ" combined fusion and spinal cord translocation

J. C. Aurégan · T. Odent · M. Zerah · J. P. Padovani · C. Glorion

Received: 16 November 2009 / Revised: 4 April 2010 / Accepted: 15 July 2010 / Published online: 17 August 2010

Abstract

An achondroplastic patient with a thoracolumbar kyphosis was first seen at the age of 16 at our institution. His only concern at that time was the aesthetic implication of his deformity. His physical examination was normal except for loss of the neurologic reflexes in the lower limbs. The radiographs showed a fixed 180° thoracolumbar kyphosis with correct frontal and sagittal balances. No spinal cord anomaly was found on MRI. Two years later, he developed a progressive neurogenic claudication of the lower limbs. He was still neurologically intact at rest. The MRI showed an abnormal central spinal cord signal in front of the apex of the kyphosis associated with the narrow congenital spinal canal. In regards to this progressive neurological worsening, a surgical treatment was decided. We decided to perform a front and back arthrodesis combined with a spinal cord decompression without reduction of the deformity. A five-level hemilaminotomy was performed with a posterior approach at the kyphosis deformity. The spinal cord was individualised onto 10 cm and the left nerve roots were isolated. A decancellation osteotomy of the three apex vertebrae and a disc excision were performed. The posterior aspect of the vertebral body was then translated forward 2 cm and in association with the spinal cord. Two nerve roots were severed laterally to approach the anterior part of the kyphosis and a peroneal strut graft was inlayed anterolaterally. A complementary anterior and a right posterolateral fusion was made with cancellous bone. The patient was immobilised in a cast for 3 months relayed by a thoracolumbosacral orthosis for 6 months. At 3 years follow-up, the neurogenic claudication had disappeared. No worsening of the kyphosis was observed. His only complaint is violent electric shock in the lower limbs with any external sudden pressure on the spinal cord in the area uncovered by bone.

Keywords Achondroplasia · Thoracolumbar kyphosis · Spinal stenosis

J. C. Aurégan (✉) · T. Odent · J. P. Padovani · C. Glorion
Department of Pediatric Orthopaedic Surgery,
Hôpital Necker-Enfants-Malades, Assistance Publique-Hôpitaux de Paris, University Paris V, 161 rue de Sèvres,
75743 Paris Cedex 15, France
e-mail: aureganjc@yahoo.fr

M. Zerah
Department of Pediatric Neurosurgery,
Hôpital Necker-Enfants-Malades, Assistance Publique-Hôpitaux de Paris, University Paris V, 161 rue de Sèvres,
75743 Paris Cedex 15, France

Case presentation

An achondroplastic patient with a thoracolumbar kyphosis was first seen at the age of 16 at our institution. He was 102 cm tall when standing, 65 cm tall when sitting and

27 kg in weight. Past medical history was unremarkable. The family history was negative for achondroplasia. He had regular follow-up with a general practitioner in his country (North Africa). His only concern was the aesthetic implication of his deformity. At this time, his physical examination was normal except for loss of the Achillean and patellar reflexes in the lower limbs. He had no sphincter disturbance or pyramidal signs. The radiographs showed a 180° thoracolumbar kyphosis with no reduction on bolster test. The frontal and sagittal balances were normal with an odontoid process plumb line passing through the L5–S1 disc. The MRI did not reveal an abnormal spinal cord signal or nerve root compression. At our department conference it was decided to observe him every 6 months. At each consultation, a physical examination was done followed by frontal and sagittal X-rays. No physical or radiological worsening was noticed in the subsequent 2 years. At 2 years, he developed a progressive neurogenic claudication of the lower limb appearing at after 500 m of walking. At rest, the neurological exam was unchanged. No apparent worsening of the deformity was noticed in the frontal and sagittal X-rays. A new MRI revealed an abnormal central spinal cord signal in front of the apex of the kyphosis associated with a narrow spinal canal without vertebral wedging or disc protrusion. With regard to this progressive neurological worsening, a surgical treatment was decided to prevent neurological worsening.

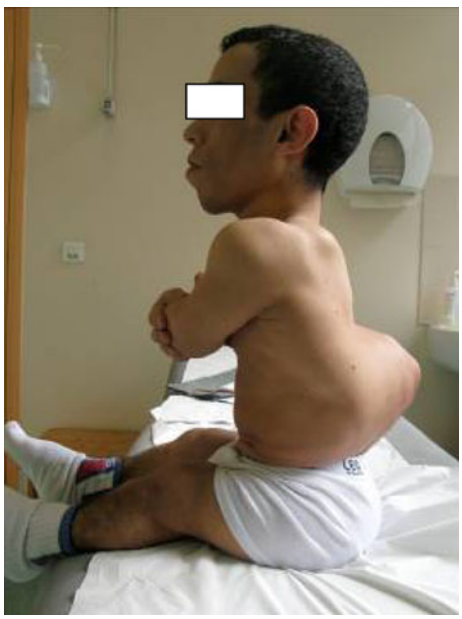


Fig. 1 Pre-operative clinical picture of the patient

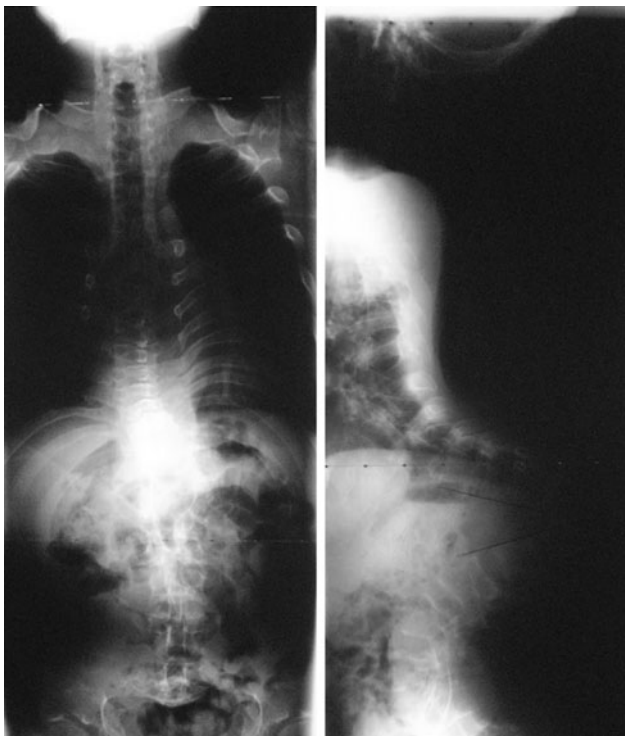


Fig. 2 Pre-operative frontal and lateral X-rays

Diagnostic imaging section

1. Pre-operative clinical picture of the patient (Fig. 1).
2. Pre-operative frontal and lateral X-rays (Fig. 2).

Historical review

Achondroplasia is a rare genetic disease. Its incidence is estimated at 1/15,000 births. Most cases arise as spontaneous genetic mutations in the FGFR 3 receptor. It leads to a dysplasia with predominantly metaphyseal involvement. It is responsible for short-limbed dwarfism and skeletal deformities with orofacial anomalies [1].

Thoracolumbar deformity in patients with achondroplasia is a recurrent problem. Prevalence is estimated around 94% in children younger than 1 year of age. The treatment in early childhood is to prevent the development of a fixed deformity by sitting modification, physiotherapy and bracing [2]. Without early conservative management, approximately 11% of patients developed important and progressive deformity with a significant risk of spinal cord compression with neurological impairment during growth [3].

In the case of thoracolumbar kyphosis surgery is either indicated when the deformity is progressing or in the presence of a spinal cord compression with neurological symptoms as bladder dysfunction or lower limb weakness.

Rationale for treatment and evidence-based literature

Various surgical treatments were proposed to manage the neurogenic complications associated with kyphotic deformity in achondroplasia. Most of the reported neurogenic symptoms are progressive claudication and incontinence. They are related to both deformity and narrowed spinal canal with a risk of unexpected paraplegia and inconsistent recovery after treatment.

In immature patients, a single posterior decompression or in association with non-instrumented posterior fusion leads systematically to an increase of the deformity [4], which results in ongoing pain with neurological worsening.

In case of thoracolumbar kyphosis below approximately 30°, the association of an in situ instrumentation fusion to the posterior decompressive laminectomy and foraminotomy may be performed with good outcome [5].

For kyphosis above approximately 30°, front and back fusion is the recommended treatment [5]. Many authors emphasise the neurological risk in case of rigid deformity, which is increased by the congenitally narrowed spinal canal [6, 7]. An important kyphotic angle or an anteriorly compromised spinal cord at the level of the deformity requires sometimes an anterior release [8]. When an anterior approach had been chosen in the presence of a very narrow canal, doing the disc excision first may be harmful to the already compromised spinal cord because of the anterior spinal manipulation along with the oedema induced by the surgery [9]. Therefore, in case of important kyphosis, it could be safe to decompress posteriorly the spinal cord first or at the same time when a two-stage procedure had been chosen [10–12].

To the best of our knowledge, no such deformity as this one had been reported. As to the late onset of neurologic symptoms of our patient, surgical treatment was required. Due to the importance of the deformity and the acceptable sagittal balance of the patient, our option was to avoid any reduction of the deformity to minimise the neurological risks. This being considered, our major concerns were to achieve a decompression of the spinal cord along with a stable fusion.

From a biomechanical point of view, one of the options was to fix the vertebrae by an anterior strut graft completed with a posterolateral fusion with autologous bone graft. The first concern with such a deformity was the surgical approach. Obviously, a classical anterior approach of the spine was not possible. The only way to approach the anterior aspect of the kyphosis for us was posterolaterally. The second concern was the spinal cord liberation and protection. The spinal cord needed to be decompressed mainly anteriorly. With the amount of deformity that was present in the patient, performing the vertebral body resection during the posterior approach was the only option. This could be done safely after the laminectomy. To minimise bleeding and respect the anterior spinal cord vascularisation, a vertebral body decancellation osteotomy was chosen. The third concern was to obtain a good acute and long-term stabilisation of the deformity. Hooks and wires were not recommended due to the diminished size of the spinal canal. The pedicle screws were used successfully in the achondroplastic spine. But in this case, a posterior fixation required a wide extension of the fusion area, a threat for the cutaneous covering and poor mechanic efficiency. For these reasons, we decided to stabilise the spine postoperatively only with the cast during the time required for the bone graft to heal.

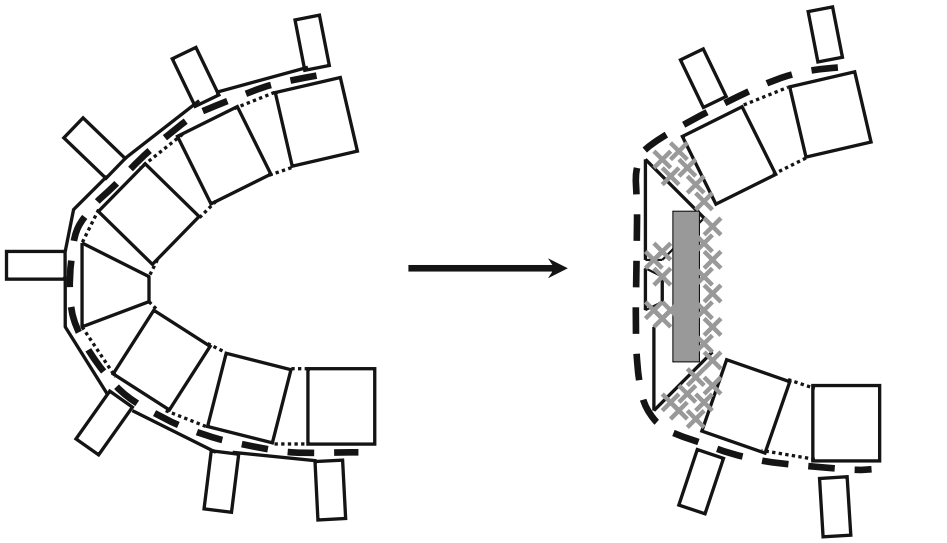
Procedure

The patient was positioned prone on the operating table. A midline skin incision was made to expose the posterior elements of the spine from T8 to S1. A five-level left hemilaminotomy was performed at the kyphosis deformity apex. We removed the spinous processes and laminae of T12, L1 and L2. The spinal cord was exposed. The spinal cord was skeletonised onto 10 cm and the left nerve roots were isolated. The bone excision was continued laterally with the pedicle removed. Through the pedicle start point, a decancellation osteotomy of the three vertebral bodies located at the apex of the deformity and the corresponding disc excision was performed. The posterior aspect of the vertebral body was translated forward by 2 cm. After that, we released the spinal cord of its tissue attachment in order to translate it. Two ribs and their corresponding nerve roots (T11 and T12) were severed laterally to approach the anterior part of the kyphosis. To avoid significant bleeding, the anterior aspect of the spine was exposed in an extraperiosteal fashion. A fibula strut graft was inlayed anterolaterally into holes that were made in the anterolateral aspect of the vertebral body. A complementary anterior and a right posterolateral fusion was made with cancellous bone to enhance bone mass fusion.

The patient was immobilised in a cast for 3 months and relayed by a thoracolumbosacral orthosis (TLSO) for 6 months.

Procedure imaging section

1. Drawing of the procedure (Fig. 3).
2. Frontal, transversal and sagittal CT-scan views (the under skin position of the spinal cord and the inlayed fibula strut graft can be noticed) (Fig. 4).



Procedure:
- 5 level left hemilaminotomy,
- Removal of spinous processes and laminae of T12, L1 and L2,
- Spinal cord and left nerve roots releasing onto 10 centimetres,
- Decancellation osteotomy and a disk excision of the 4 apex vertebras,
- Translation of the posterior aspect of the vertebral body,
- Spinal cord translation,
- Section of two ribs and their correspondent nerve roots (T11 and T12),
- Fibula strut graft inlayed anterolaterally,
- Complementary anterior and a right postero-lateral cancellous bone grafting.

Legend:
- : spinal cord
- : Fibula strut graft
- : cancellous bone graft

Fig. 3 Drawing of the procedure

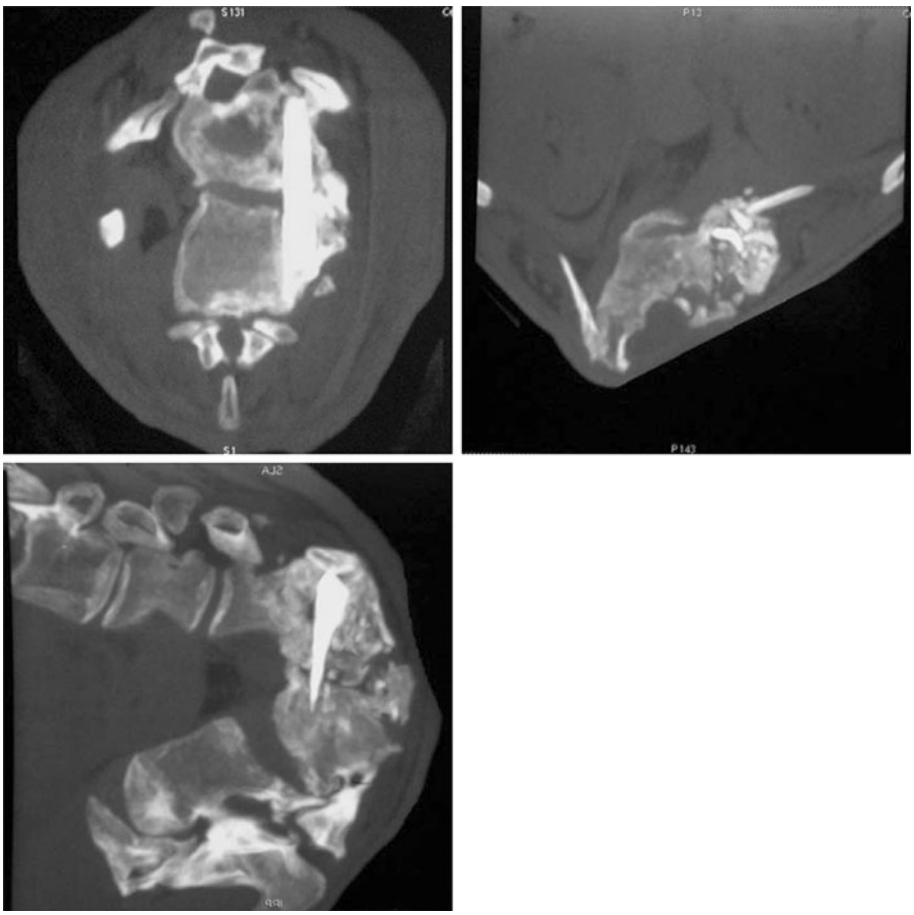


Fig. 4 Frontal, transversal, and sagital CT-Scan views (the under skin position of the spinal cord and the inlayed fibula strut graft can be noticed)

Outcome and follow-up

No complications occurred during or after the surgery. The post-operative time had been normal and the patient started to ambulate on day 3. The two severed nerve roots resulted in a left metameric thoracic anaesthesia without any pain. The patient supported well even the cast and the TLSO. We followed him at 6 weeks, 3 months and every 6 months until 2 years. Then we decided to follow him once a year. At each consultation, a physical examination followed by frontal and sagittal X-rays had been performed.

At 3 years follow-up, his neurological claudication had disappeared. No worsening of the kyphosis was noticed. His only complaint is violent electric shock in the lower limbs with any external sudden pressure on the spinal cord in the area uncovered by bone.

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