

Surgical treatment of achondroplasia with thoracolumbar kyphosis and spinal stenosis—a case report

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A 13-year-old boy suffered from low back weakness and bilateral lower extremity numbness for many years. In June 2003, intermittent claudication appeared, the boy could only walk for 5 min, and he was seen at our clinic. Physical examination revealed a short stature of only 126 cm, prominent frontal bossing, nose-bridge depression, short limbs with long trunk, and thoracolumbar kyphosis (Figure 1). No neurological deficit was noted. Radiographs revealed scoliosis and reduced interpedicular distance of the lumbar spine (Figure 2). An anterior wedged deformity of the L1 body with prominent thoracolumbar kyphosis was observed with a T12-L2 kyphosis angle of 55° (Figure 3). MRI of the thoracolumbar spine identified a spinal canal stenosis (diameter 10 mm) below the T11 level (Figure 4). One stage anterior L1 corpectomy with interbody fusion was performed, followed by posterior decompression with pedicle screw



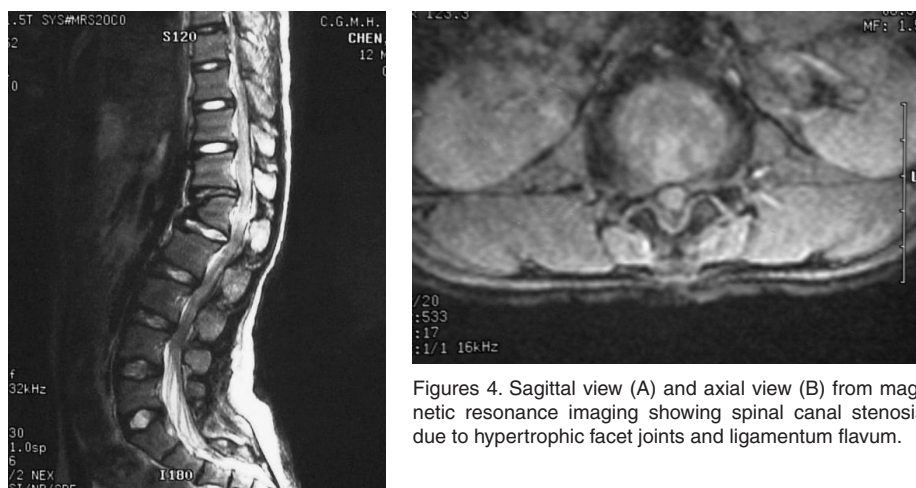
Figure 2. Anteroposterior radiographs of the thoracolumbar spine demonstrate mild thoracic scoliosis, with decreased lumbar inter-pedicle distance.



Figure 1. Preoperative image of a 13-year-old boy with prominent frontal bossing, depressed nose-bridge, and persistent thoracolumbar kyphosis.



Figure 3. Lateral radiographs of thoracolumbar spine showing L1 anterior wedge deformity and thoracolumbar kyphosis (55°, T12–L2).



Figures 4. Sagittal view (A) and axial view (B) from magnetic resonance imaging showing spinal canal stenosis due to hypertrophic facet joints and ligamentum flavum.

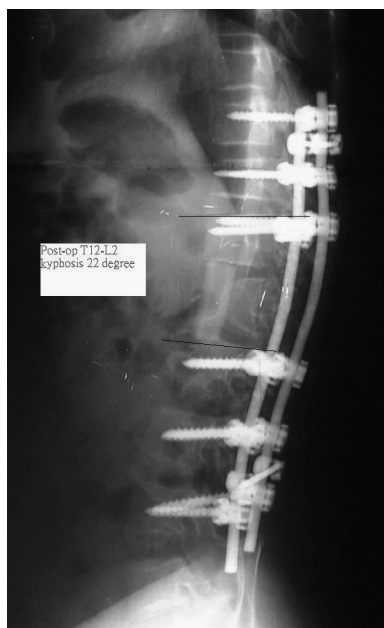


Figure 5. Postoperative lateral radiographic images showing kyphosis 22° (T12–L2).

instrumentation. The patient was placed in the right decubitus position, and L1 corpectomy and T12–L2 interbody fusion with fibula allograft and rib autograft were performed first via the left extra-pleura-retroperitoneal approach. The patient then was turned over to a prone position and the laminectomy was extended longitudinally to include all the stenosis segments (T12–L4), but facet joints were preserved. Posterolateral fusion with pedicle screwing (T10–L4) was then performed. The pos-

terior procedures were monitored via somatosensory evoked potential (SSEP).

The patient was encouraged to ambulate on the second day after operation and was discharged 5 days later. Postoperative radiographs revealed T12–L2 kyphosis of 22° (Figure 5). 1 year postoperatively, the patient was completely relieved of neurological symptoms and had resumed his previous daily activities.

Discussion

Achondroplasia was first described by Parrot in 1878 (Thomeer and van Dijk 2002). It is the most prevalent form of dwarfism, with an incidence of approximately 1 in 25,000 (Pauli et al. 1997). Owing to the endochondral ossification defect, limb and spine growth disturbances occur in achondroplasia (Thomeer and van Dijk 2002). The clinical features of achondroplasia include short limbs, rhizomelic dwarfism, and a relatively long trunk. There are two spinal problems related to achondroplasia: thoracolumbar kyphosis and lumbar spinal stenosis. Most infants with achondroplasia display thoracolumbar kyphosis, and muscle hypotonia has been hypothesized to be responsible for its development (Kopits 1976). At the age of 12–18 months, the trunk develops strength and the child can extend the trunk. Anterior vertebral wedging in achondroplasia becomes resolved in 90% of patients (Hall 1988).

Early bracing is another method of preventing fixed kyphosis (Bethem et al. 1981, Pauli et

al. 1997). However, approximately 10% of such transient kyphosis cases become fixed, and angular kyphosis is associated with vertebral wedging or hypoplasia (Bethem et al. 1981). For the few children in whom thoracolumbar kyphosis does not resolve, anterior and posterior fusion with posterior wiring of kyphotic segments has been proposed (Tolo 1999). Lumbar spinal stenosis may be the main etiology of neurological claudication in achondroplasia, but generally occurs only after early adulthood (Bethem et al. 1981). The short pedicle and reduced inter-pedicle distance reduce the size of the spinal canal, both anteroposteriorly and transversely (Kahanovitz et al. 1982, Pyeritz et al. 1987, Fortuna et al. 1989, Savini et al. 1991, Thomeer and van Dijk 2002). Factors related to ageing, such as herniation of the intervertebral disc, increasing lumbar lordosis, degenerative facet joint, and progressive thoracolumbar kyphosis, contribute to the development of neural symptoms (Savini et al. 1991). The third and fourth decades of life are the most common age for development of thoracolumbar spinal stenosis syndrome in achondroplasia. The mean age on admission with spinal stenosis is 31 (22–37) years (Fortuna et al. 1989). At the age of 13, the mean sagittal diameter of the lumbar spine is 20 mm (Hinck et al. 1965), but the sagittal intraspinal canal diameter of our patient was 10 mm below the T11 level. Midsagittal lumbar canal diameters of less than 10 mm represent absolute stenosis (Verbiest 1979). Multi-level laminectomy is indicated if the clinical features mainly result from a narrow canal. The good long-term results of multi-laminectomy for spinal stenosis are related to the short duration of preoperative symptoms (Pyeritz et al. 1987). Thomeer and van Dijk (2002) reported the results of 36 patients with achondroplasia dwarfism who received laminectomy for spinal stenosis. The mean age in their study was 38 years and the youngest patient was 10 years old, but he underwent laminectomy only without any fusion or instrumentation procedure. Shikata et al. (1988) reported 3 cases of achondroplasia dwarfism with paraplegia, treated by laminectomy (the youngest patient in that report being only 19 years old). Simultaneous posterolateral spondylodesis using Harrington instrumentation was attempted for one of the patients.

The indication, timing, and procedure for surgical treatment of spinal canal stenosis with thoracolumbar kyphosis in achondroplasia have not been well established. Anterior decompression with anterior interbody fusion and posterior decompression with posterolateral fusion are indicated for spinal stenosis over 3 disc levels with kyphosis > 40° (Shikata et al. 1988). The posterior instrumentation can stabilize the spine and enhance posterolateral fusion following laminectomy, but the hook system is contraindicated because of the narrow canal in achondroplasia (Lonstein 1988). Neurological damage intraoperatively by Harrington compression instrumentation in an achondroplasia patient has been reported (Savini et al. 1991). This surgical complication can be prevented by using pedicle screw instrumentation in achondroplasia.

To our knowledge, this patient is the youngest achondroplasia patient to have received one stage of anterior fusion and posterior decompression, and posterolateral fusion with pedicle screw instrumentation for thoracolumbar kyphosis and spinal stenosis. This method is safe and effective.

Contributions of authors

J-CL wrote the manuscript. W-JC did the operation and helped writing the manuscript. P-LL corrected the English. L-HC prepared the photographs.

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