

## Spinal cord impingement by a displaced rib in scoliosis due to neurofibromatosis

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Neurofibromatosis is often associated with a dysplastic progressive scoliosis.<sup>1,2</sup> Enlargement of the neural foramina by erosion of adjacent neurofibromas occasionally predisposes to the adjacent attached rib entering the spinal canal.<sup>3-8</sup> We describe such a case in which partial rib excision and later posterior spinal fusion resulted in complete recovery.

### Case report

A 10-year-old boy was referred for evaluation of unusual backache with associated scoliosis. His back pain would be accentuated by lying on his stomach watching TV. When he rolled onto his right side, his right leg would get weak and he often felt a “shock” that would radiate to his toes. Neurofibromatosis type I had been diagnosed when he was 6 years old, when the skin lesions began to appear, but no spinal deformity was noted. When he presented to our scoliosis clinic, a right-sided prominent rib hump associated with a 76° thoracic scoliosis was seen. There was no evidence of any permanent neurologic impairment.

Radiologic examination demonstrated a right thoracic curve from T4 to T12, measuring 76° by the Cobb method (Fig. 1). His curve had progressed from 55° to 76° over the preceding year.

MRI showed no tumours intracranially or within the spinal canal. Three-dimensional CT and CT-myelography

revealed detachment and translocation of the cephalad end of the ninth rib on the convex side of the curve through an enlarged intervertebral foramen into the spinal canal, where it was severely compressing the spinal cord, even though the cord lay against the concave side of the spinal canal (Fig. 2, left).

After thorough evaluation by both orthopedic and neurologic surgical teams, it was elected to perform a 2-stage procedure in an attempt to minimize postoperative complications such as paraparesis and paraplegia described in other reported cases.<sup>3-8</sup> The first stage comprised rib excision with decompression of the protruding right ninth rib into the thoracic canal. We had planned to excise the whole posterior third of the rib. However, attempts to remove the rib from the canal resulted in a loss of signal by spinal cord monitoring. The rib was adherent to the cord. Because of this, the small intraspinal segment of the rib was left in situ, and a 5-cm segment starting at the nerve-root foramina was excised with the rib periosteum. The rib could then not exert a lever effect on the cord, and we hoped the remaining intraspinal portion would eventually be resorbed, similar to intraspinal bone fragments after burst fractures. After the procedure was completed, a wake-up test was performed, and the boy moved his feet well. Postoperatively, he was ambulatory the next day and exhibited no change in neurologic

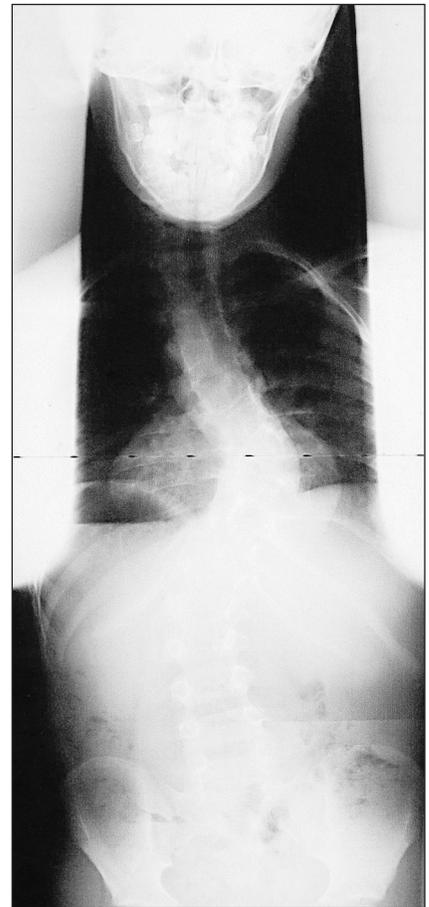


FIG. 1. Presenting radiograph of the spine in a 10-year-old boy shows a short, sharp thoracic curve measuring 76° by the Cobb method.

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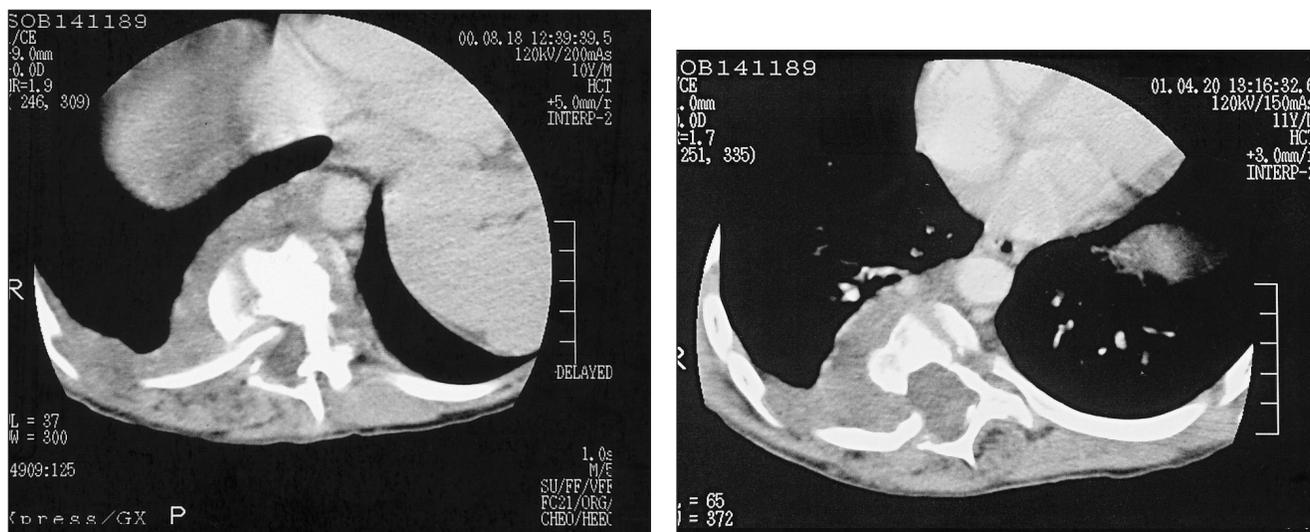


FIG. 2. Left: protrusion of the ninth rib on the convex side of the curve impinging on the spinal cord. Right: CT 2 years postoperatively shows retraction of the ninth rib with no residual spinal cord compression.

signs or symptoms. He was discharged home on postoperative day 2. The second stage consisted of an in-situ spinal fusion performed from T6 to L1. A 2-year follow-up radiograph demonstrated a solid posterior fusion mass with no further spinal cord impingement of the head of the dysplastic right ninth rib (Fig. 2, right) and no progression of his scoliosis.

### Discussion

To our knowledge, only 8 children (ranging in age from 5–16 yr) with rib encroachment of the spinal canal have been reported in the English-language literature.<sup>3–8</sup> All rib impingements occurred on the convex side of the curve. The apex of the curve, where maximal rotation of the vertebral bodies is present, was the site of the rib displacement in 6 patients. Three of the reported patients were asymptomatic,<sup>6,8</sup> and 2 had mild long-tract signs and symptoms.<sup>3,5</sup> Two had paraparesis.<sup>4,7</sup> One patient had loss of sensation below the waist with inability to move the lower limbs after a fall on the rib hump. Symptoms resolved spontaneously after a few minutes.<sup>8</sup> The patient with paraplegia underwent a 2-stage combined posterior and anterior spinal fusion with resection of the rib, after which neurologic function returned to normal.<sup>4</sup>

In neurofibromatosis patients with scoliosis, rib penetration into the spinal canal is rare but may be more commonly identified with modern imaging techniques. This protrusion has the potential to cause paraplegia or paraparesis with or without a traumatic episode during spinal surgery or even postoperatively. These risks of neurologic deficit can be avoided by excising the intracanal rib segment or by leaving the proximal penetrating part of the rib in place but removing a 5-cm segment of its lever arm. In treating patients with dystrophic spinal curves, rib penetration should be sought with CT and MRI imaging of the apex of the curve before any planned spinal surgery or in the presence of any neurologic cord deficit.

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