

Use of Rib-Based Distraction in the Treatment of Early-Onset Scoliosis Associated With Neurofibromatosis Type 1 in the Young Child

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Abstract

Study Design: This study was an institutional review board–approved retrospective review of a prospectively collected multicenter database for rib-based distraction systems used in the treatment of young children with early-onset scoliosis associated with neurofibromatosis type 1 (NF-1).

Objective: To evaluate the effectiveness of rib-based distraction and associated complications in managing scoliosis in the growing child with NF-1.

Summary of Background Data: Patients with NF-1 commonly have scoliosis with curves that can be dysplastic and progressive and respond poorly to bracing. Rib-based distraction systems have recently been described in the management of these complex patients. The efficacy and complication rate using these systems have not previously been reported.

Methods: Twelve children with NF-1 and scoliosis who were treated with rib-based distraction systems were identified from a prospectively collected multicenter registry. Preoperative and postoperative Cobb angle and T1–S1 spine height were measured from posteroanterior radiographs. The number of lengthenings, age at implantation, years of follow-up, and complications were also acquired from the registry or patient charts.

Results: Mean age at implantation of the Vertical Expandable Prosthetic Titanium Rib device was 6.34 years. Mean preoperative Cobb angle was 66.3°. Average follow-up was 5.2 years. Mean postoperative Cobb angle was 60.8° after an average of 7.75 lengthening procedures (range, 2–16 procedures). T1–S1 height increased in all patients. There were 17 complications in 8 patients (device migration in 6, wound dehiscence in 3, rod breakage in 2, medical issues in 5, and 1 revision for progression of curve). Of the 17 complications, 10 were grade I, 1 was grade II, and 6 were grade IIA; there were no grade III complications.

Conclusions: The use of rib-based distraction is an effective and relatively safe method of stabilizing curve progression through growth in severe dysplastic scoliosis associated with NF-1.

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Keywords: Neurofibromatosis; Scoliosis; VEPTR; Complications

Introduction

Neurofibromatosis type 1 (NF-1) is a multisystemic disease that affects neural tissue, soft tissues, bones, and

skin. It has a variety of presentations and can range from mild pigmentation abnormalities to extreme and debilitating or even life-threatening systemic issues. Skeletal abnormalities are often seen in patients with NF-1 and can present significant treatment challenges [1].

Scoliosis is the most common osseous defect associated with NF-1 [1–3] and may account for as much as 2% of patients seen in orthopedic clinics for spinal deformity [4]. Curves can vary in severity from mild to severe. More troublesome than the high prevalence in NF-1 is the aggressive nature many of these deformities exhibit. Often these curves are dysplastic and

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progressive, have associated rib anomalies, and typically respond poorly to bracing [3–8]. For children with early-onset deformity, the management can be challenging. Many of these children are young with early and progressive curves that make definitive fusion procedures an undesirable option [9,10]. Poor local bone quality, dysplastic pedicles, and excessive kyphosis often make the use of more traditional spine-based growing rods difficult and in some cases impossible [11] (Fig. 1A, B). As an alternative, the current authors have used rib-based distraction in some of these more difficult early-onset patients with NF-1, using the Vertical Expandable Prosthetic Titanium Rib (VEPTR; Synthes Spine, West Chester, PA) (Fig. 2A, B).

In 2004, Hell et al. [12,13] described the use of the VEPTR device for the treatment of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. The VEPTR has been used as a treatment method for a variety of conditions, including progressive scoliosis without fused ribs, neuromuscular scoliosis in young children, and other exotic forms of scoliosis such as spinal muscular atrophy and myelodysplasia. The purposes of this

study were to determine the effectiveness of rib-based distraction for managing scoliosis in the growing child with neurofibromatosis and to report the incidence of complications when using rib-based distraction systems.

Methods

This study was designed as an institutional review board–approved retrospective review. A multicenter, institutional review board–approved early-onset scoliosis patient registry was queried for patients with progressive spinal deformity associated with NF-1. Patients included in the study were those who had previously undergone or were currently being treated for progressive early-onset scoliosis related to the disease. For inclusion in the study, patients treated with rib-based distraction systems who had previously failed nonsurgical management, including observation and bracing, were considered. Patients treated with previous surgery were excluded. Also excluded were patients who had achieved skeletal maturity before completing 2 years of rib-based distraction and those with less than 2 years of follow-up, as well as patients with

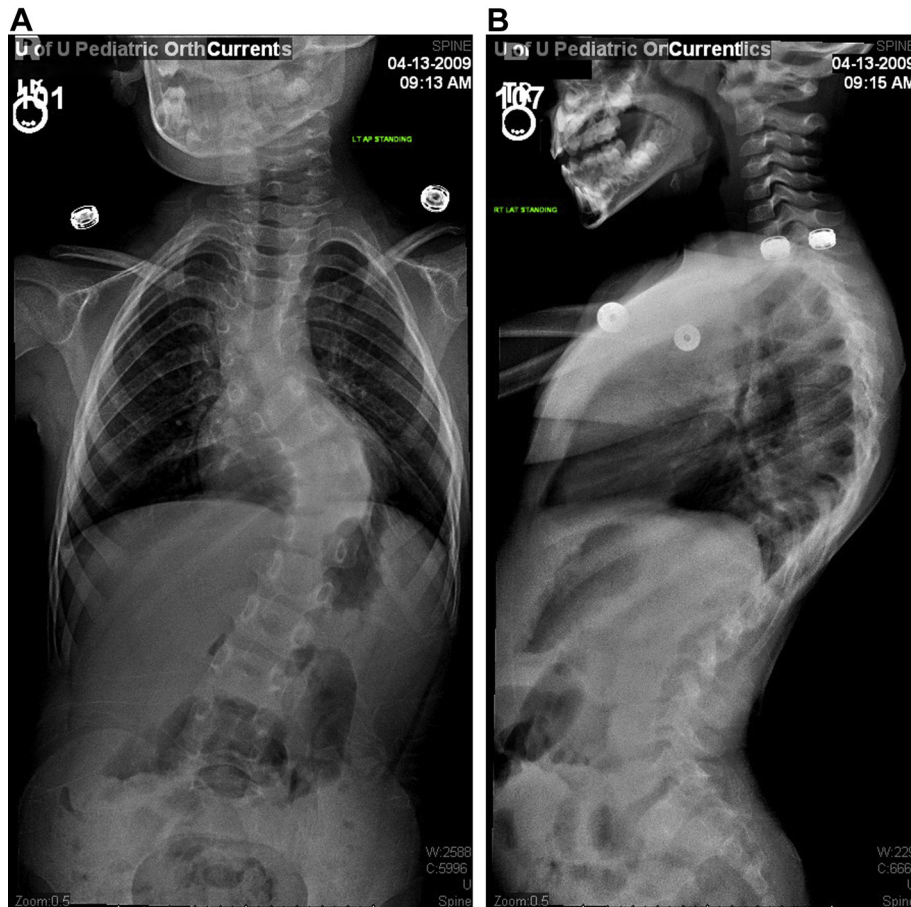


Fig. 1. (A) Preoperative standing posteroanterior radiograph of the spine of a 3-year-old boy with a dystrophic spinal deformity. The 2 thoracic curves measured 62° and 74° . Because of the small and dysplastic nature of the upper thoracic pedicles and the significant kyphosis (67°), it was thought that traditional growing rods would not be the best option. The Vertical Expandable Prosthetic Titanium Rib was chosen to manage the deformity. (B) Preoperative lateral X-ray of the same patient.

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