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Case Report

An unusual case of congenital scoliosis associated with rib agenesis in the upper part of the concavity treated by VEPTR vertebra to vertebra

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Abstract

BACKGROUND CONTEXT: Rib agenesis in congenital scoliosis is rarely encountered, and its disposal in the application area of the proximal vertical expandable prosthetic titanium rib (VEPTR) module is a challenge to the orthopedic surgeon.

PURPOSE: To present a case in which known treatment methods in early-onset scoliosis were not possible to apply.

STUDY DESIGN: Case report.

METHODS: A patient aged 1 year and 10 months, presenting a congenital scoliosis with the following characteristics: left T3 hemisegmented hemivertebra, T5–T6–T7 hemivertebral segment, T9, T10 trapezoidal vertebrae, right side I–IV rib agenesis with T1–T2–T4 hemivertebral hypoplasia (T3 agenesis) and bilateral XIIth rib agenesis, and V–VI and VII–VIII–IX fused ribs on the right side. We applied a standard VEPTR in a new construct, vertebra to vertebra.

RESULTS: The VEPTR vertebra to vertebra proved to be an efficient and stabile construct after 1.5 years of follow-up and three device distractions in a row. The curve corrected from 100 to 58 Cobb degrees.

CONCLUSIONS: We believe that the vertebra-to-vertebra construct with eventual modifications may be a solution in the treatment of early-onset scoliosis needing surgery, which associate rib agenesis in the area where the proximal module has to be applied.

Keywords: Congenital scoliosis; Rib agenesis and hypoplasia; Longitudinal bar; Thoracic insufficiency syndrome; VEPTR vertebra to vertebra

Introduction

Congenital scoliosis with unique or multiple rib fusions, in association with rib agenesis, presents a highly increased progressive potential, inducing respiratory failure because of a thoracic insufficiency syndrome (TIS), obvious in the first years of life [1]. R. Campbell Jr, first described TIS in 1992 [1]. The vertical expandable prosthetic titanium rib (VEPTR) is a surgically implanted device used to treat TIS in pediatric patients, to straighten the spine and separate ribs so that the lungs can grow and fill with enough air to breathe [2]. Rib agenesis is rarely present, and its disposal on the site, where the VEPTR proximal module should be applied, represents a challenge to the surgeon.

Our experience with VEPTR is based on 26 patients, excluding the presented case, with 27 VEPTR devices in standard constructs, during 2009 to 2011. This report is about the only congenital scoliosis case, to our knowledge, with rib agenesis in the upper part of the concavity.

Case report

A 3-month-year-old boy presented to the Neonatal Intensive Care Unit, transferred from a maternity, because

FDA device/drug status: Approved (VEPTR).

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of TIS, boosted by a respiratory infection episode. The clinical examination revealed signs and symptoms because of TIS (tachypnea, wheezing, cyanosis when feeding) in the presence of a severe chest deformity and an axial deviation of the spine in the coronal plane.

Spinal X-ray established the diagnosis of congenital scoliosis with a 70° Cobb curve associated with multiple rib malformations. The presence of hemivertebrae was noticed. Three bony blocks probably united by an osteocartilaginous bridge on the concave side of the curve was interpreted as a longitudinal bar (Fig. 1).

Respiratory symptoms regressed after oxygen therapy, mineral salt perfusion, antibiotics, vitamins, and symptomatic medicines. The patient started conservative treatment after this episode, consisting of physiotherapy and a plaster cast "bed," replaced by a thoracolumbar brace at the age of 8 months.

Thoracic insufficiency syndrome became more obvious during growth and the respiratory symptoms more severe, accompanied by a significant three-dimensional (3D) deformity of the chest, affecting the somatic development and the current physical activities (after walking a few steps, he asked his mother to pick him up). The X-ray showed Cobb angle progression.

The rapid progression of the respiratory symptoms and the fast developing deformity, which was obviously clinical and on medical imaging, imposed the necessity of a surgical treatment at the age of 8 months, but the surgery had to be postponed because of social and economic issues.

X-rays (Fig. 2), computed tomography (CT), and 3D CT scans were performed at the age of 1 year and 4 months. The increased progressive potential of the scoliosis curve has been considered maximal: the Cobb angle increased from 70° to 100°. The 3D CT scan established a certain diagnosis of all vertebral and costal bony malformations (Fig. 3): left T3 hemisegmented hemivertebra, T5–T6–T7 segmented hemivertebral block, T9 and T10 trapezoidal vertebrae, I–IV rib agenesis on the right with T1–T2–T4 hemivertebral hypoplasia (T3 agenesis), bilateral XIIth rib agenesis, and V–VI and VII–VIII–IX fused ribs on the right side.

The magnetic resonance scan of the coronal plane showed a right diminished in volume hemithorax and a synostotic osteocartilaginous block on the concave side of the spine, being interpreted as a longitudinal bar extended from T4 to T10. The medulla was discretely marked by the hemivertebral block on the concave side.

The clinical examination performed preoperatively and the thoracoabdominal dysmorphic aspects are suggestive

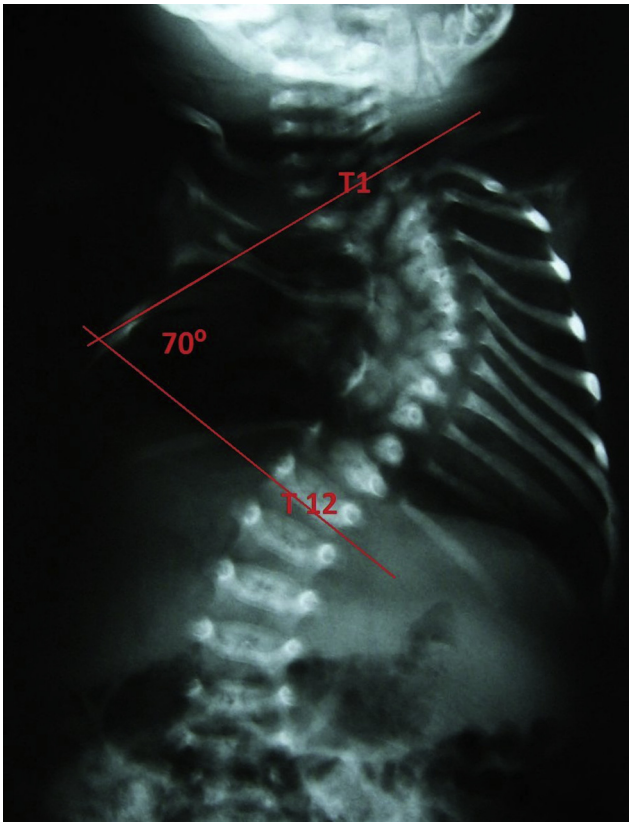

Fig. 1. The first X-ray at the age of 3 weeks: thoracic T1–T12 scoliosis with a 70° curve and multiple rib agenesis, hypoplasia, and fusions.

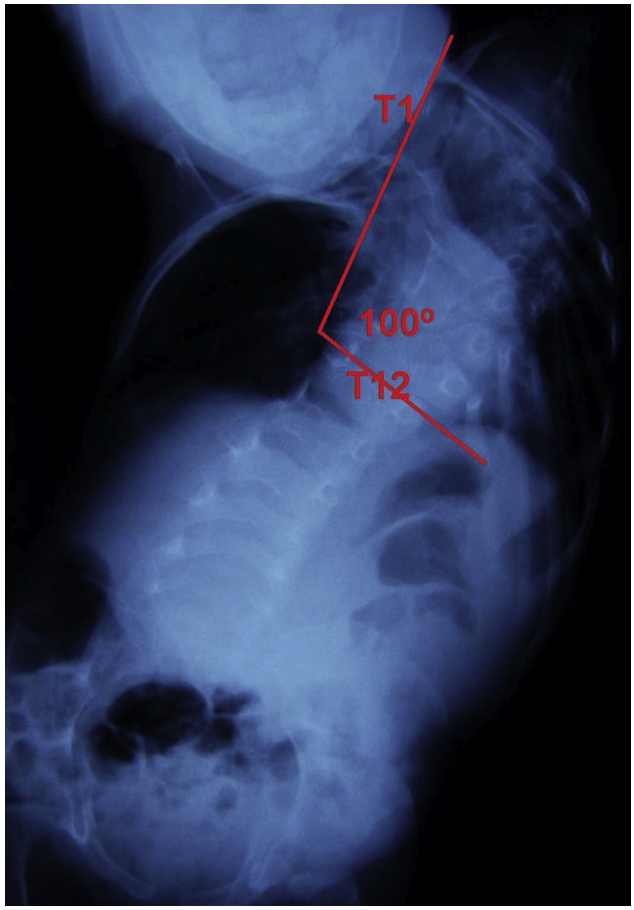

Fig. 2. X-ray at the age of 1 year and 4 months: a scoliotic curve of 100°.