ISSN: 2755-9890

Journal of Spine Research & Reports

SCIENTIFIC Research and Community

Case Report

Open Access

When Convex Growth Arrest in the Thoracic Spine fails to control progression of a congenital scoliosis: Case Report

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Abstract

Congenital scoliosis are one type of Early-Onset Scoliosis (EOS), a paediatric spinal deformity that progresses rapidly before the age of 10. Due to the high risk of spinal curvature aggravation and subsequent thoracic constraints during the critical period for lung development early in life, these young children with EOS may develop impaired pulmonary function. Some of these patients are at risk of developing Thoracic Insufficiency Syndrome (TIS) due to Volume Depletion Deformities (VDD), particularly the Type II (with rib synostosis). In the past, early spinal fusion of the thoracic spine was the standard surgical treatment but resulted in iatrogenic restrictive lung disease due to shortening of thorax "height". For some of the congenital cases, Convex Growth Arrest (CGA or hemiepiphysiodesis) of the main spinal curvature is a recognized growth-guided surgical technique, appropriate to deal with this type of condition, but the outcome can be rather unreliable and its failure can lead to severe residual chest and spinal deformities that may compromise pulmonary function in the same way.

Case Report: This is a 2 year old boy with Goldenhar Syndrome, a congenital condition with several spine and rib deformities like hemivertebrae, butterfly vertebrae, fused ribs and congenital scoliosis. With all these anomalies present, the thoracic scoliosis progressed to 75º by two years of age and he underwent a CGA in order to stop curve deterioration and hopefully to improve the deformity. However, despite this procedure the scoliosis got progressively worse and at 5y of age had a cobb angle of 114º. Upon discussion of the several alternatives to improve the spinal deformity, a VEPTR (spine to rib) was inserted at the of age five with subsequent repeated distraction program that successfully controlled the deformity. The boy at the age of ten had the curvature almost fully corrected but due to the hospital's waiting list, the final procedure was only performed by the age of thirteen (with progression of the curvature). He reached skeletal maturity with definitive posterior spinal instrumented fusion in a stable condition with preserved respiratory function leading a normal life, able to play tennis.

Conclusions: For patients at risk of TIS due to congenital EOS and rib synostosis, expandable thoracic distraction device is an alternative growth sparing tool in the armamentarium to correct a severe thoracic scoliosis while preserving thoracic spine lengthening and optimizing pulmonary growth development. Failure of hemiepiphysiodesis often requires more invasive salvage techniques to manage aggressive curve progression. Clinical success should be assessed by the stabilization of respiratory condition and preservation of vertical trunk growth in a balanced spine, rather than by correction of the Cobb angle only.

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Received: May 07, 2026; **Accepted:** May 11, 2026; **Published:** May 20, 2026

Keywords: Early-Onset Scoliosis, Congenital Scoliosis, Convex Growth Arrest, Hemiephysiodesis, VEPTR, Thoracic Insufficiency Syndrome, Complications, Expandable Thoracic Distraction System

Introduction

Congenital scoliosis is one type of Early-onset scoliosis (EOS), a spinal deformity that usually progresses rapidly before the age of 10, regardless of the aetiology and can be one of the most complex challenges in paediatric orthopaedics. As a rapidly progressive deformity it can compromise not only the spinal growth but also lung development. In many cases the congenital scoliosis is only part of a general condition that may involve many other organs as in syndromic cases. As a consequence of this aggravation compromising growth of the chest, the Thoracic Insufficiency Syndrome (TIS) defined as the inability of the chest to support normal breathing or lung growth, can be one of the major complications associated to Volume Depletion Deformities (VDD). These are three-dimensional distortions of the chest wall resulting in thoracic volume loss, classified as rib absence (Type I), rib fusion (Type II), and global hypoplastic chest (Type III), leading to segmental hypoplasia of that hemithorax. However,

as growth of the lung is triggered by volume expansion of the thorax and pulmonary tissue hypertrophy, segmental hypoplasia may compromise thoracic volume development and pulmonary function [1-4].

Over the years, early spinal fusion was the standard treatment, effectively correcting the Cobb angle but compromising not only the spinal growth and its longitudinal lengthening but also inhibiting vertical trunk growth—the primary determinant of intrathoracic volume [4]. A severe restrictive respiratory failure later on in life was very often the end result of this type of surgical treatment, exchanging a skeletal deformity for an iatrogenic lung disease.

However, some syndromic congenital scoliosis associated with rib synostosis (VDD type II) also have a devastating natural history. Such a rapidly progressive deformity at this young age can compromise the skeletal axial development but subsequently, the concave hemithorax becomes longitudinally constricted and shortened, threatening lung development through alveolar hypoplasia and decreased multiplication that usually occurs before the age of eight years. To manage this, growth-guided procedures in the spine such as CGA have been used with unreliable results; its failure often leads to a marked spinal fusion mass with loss of bony landmarks, making subsequent reconstructive salvage procedures extremely difficult and risky. The main objective of a CGA is to modulate spinal growth, inhibiting it on the convex side of the curvature and allowing the concave aspect to grow, thus correcting the curvature gradually. However, the post-operative behavior of these curvatures can be quite unpredictable with less-than-optimal outcome and therefore other more reliable alternatives have been developed [3-6].

The development of the Vertical Expandable Prosthetic Titanium Rib (VEPTR) by Campbell presented a significant advancement in the treatment of these EOS with VDD. It was conceived for the direct treatment of patients at risk of TIS, with emphasis on Type II VDD. The Opening Wedge Thoracoplasty may on one hand facilitate the insertion of the device between ribs to mechanically expand and stabilize the rigid constricted chest wall but at the same time, another piece of the instrumentation inserted between the spine and the ribs (or rib to rib) distracts the concavity of the spinal curvature. This dual mechanism is a completely new concept to deal with a spinal deformity. By instrumenting the chest wall, it aims to progressively restore the thoracic volume to allow proper lung growth, while correcting the scoliosis simultaneously, fulfilling the growth-guided strategy of preserving spinal growth and maximizing pulmonary development [2-7].

In this context, we present a case of a rapidly progressive Congenital Syndromic Scoliosis that failed CGA, in which the use of Expandable Thoracic Distraction device proved to be effective as a salvage procedure in the strategy to correct a rigid fused thoracic curvature in a young child.

Case Presentation

We present a 2-year-old boy diagnosed with a congenital syndromic scoliosis (Goldenhar syndrome) and Type II VDD (rib fusion), with an initial Cobb angle of 45º (Figure 1).

Goldenhar Syndrome is a rare congenital condition with several spine and rib deformities like hemivertebrae, butterfly vertebrae, fused ribs and congenital scoliosis and affecting other anatomical areas of the body.

Due to the scoliosis curve progression to 75º by age three (Figure 2), the patient underwent anterior and posterior CGA. Notwithstanding this procedure, the curve continued to progress to 90º at age four and reached a critical 114º at age five (Figure 3).

This aggressive worsening of the spinal deformity, indicating an imminent risk of TIS, required a solution that was not easy: posterior three column osteotomy of the fused thoracic spine was an alternative but risky and difficult to correct a large segment of the fused thoracic spine. So we have decided to apply the Wolff's principle in this young bone by distracting the concavity of the rigid curvature. At five years of age, the patient underwent a salvage procedure, consisting of a VEPTR device (rib to spine fixation) to manage both the spinal and the thoracic deformity (Figure 4).

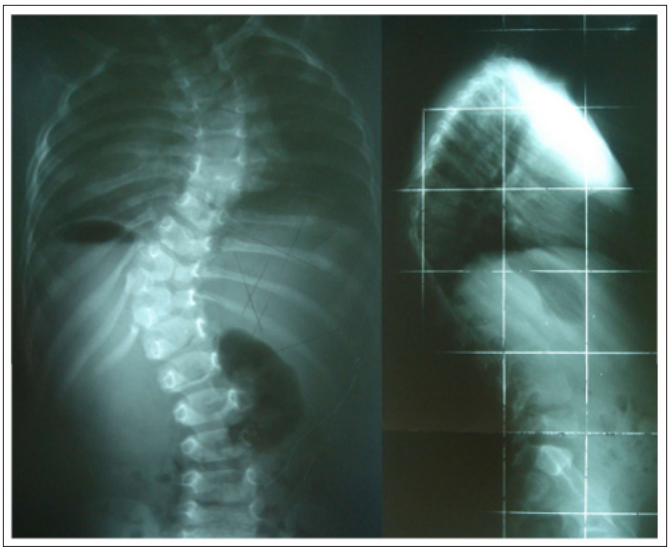


Figure 1: AP and Lateral X-Ray views of the Spine at Two Years of Age – Hemivertebrae, Butterfly Vertebrae and RIB fusion and a Main Cobb Angle of 45º

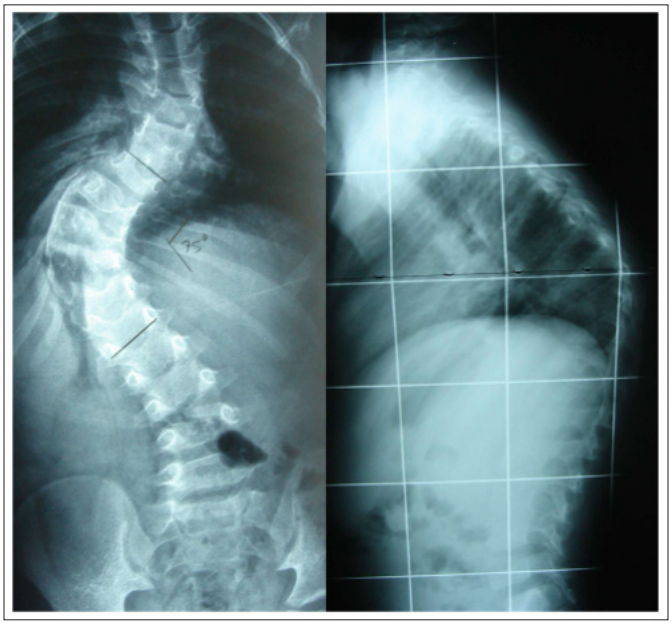


Figure 2: AP and Lateral X-Ray Views of the Spine at Three Years Old with a Cobb Angle of 75º

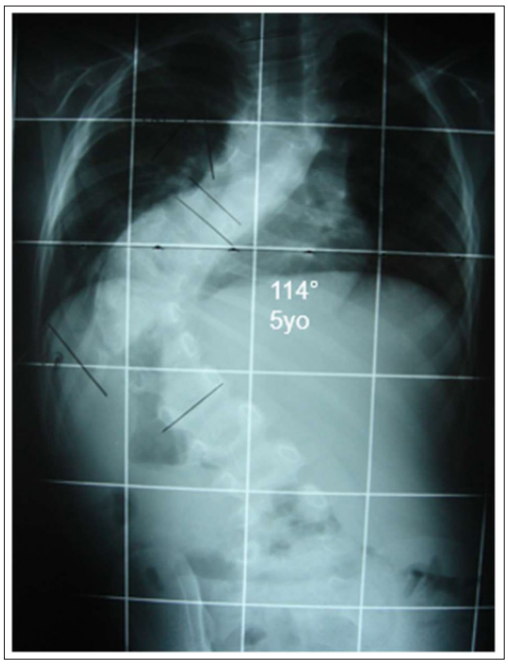


Figure 3: AP X-Ray view of the Spine at Five Years Old with a Cobb Angle of 114º

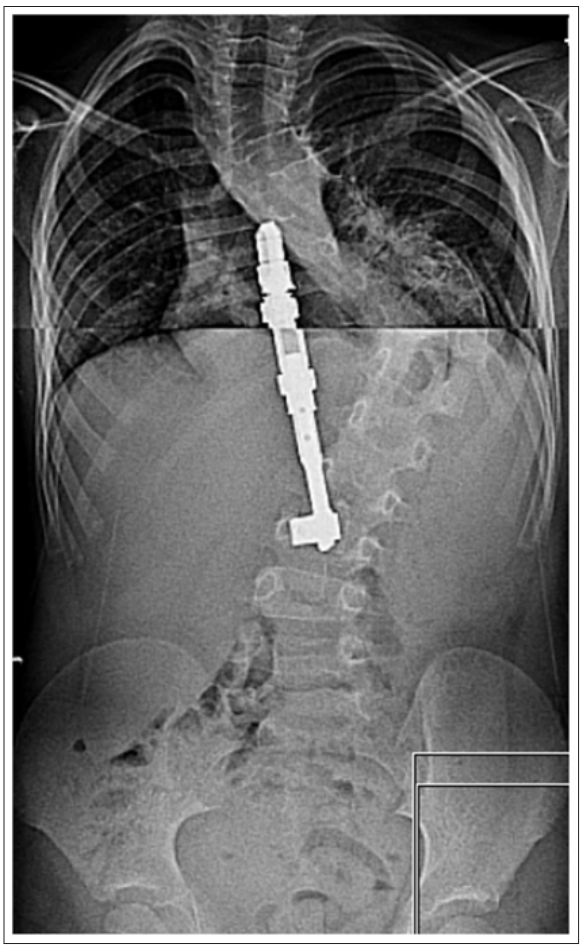


Figure 4: AP X-Ray view of the Spine at Five Years Old after VEPTR Insertion

Upon sequential distraction of the implanted device, we were able to progressively correct the spinal deformity and by ten years of age, the patient exhibited almost a complete correction of the scoliosis and reached the end of the growth-sparing phase (Figure 5). Although eligible for definitive spinal fusion, surgical waitlist restrictions delayed the final procedure until age 13, highlighting challenges in the timely transition to definitive posterior instrumented fusion (Figure 6) in severe scoliosis in this age group. Current clinical photographs at age 22, shows an excellent maintenance of trunk symmetry, spinal alignment and correction of the rib hump, reflecting a successful long-term functional and aesthetic outcome (Figures 7) with the strategy chosen.

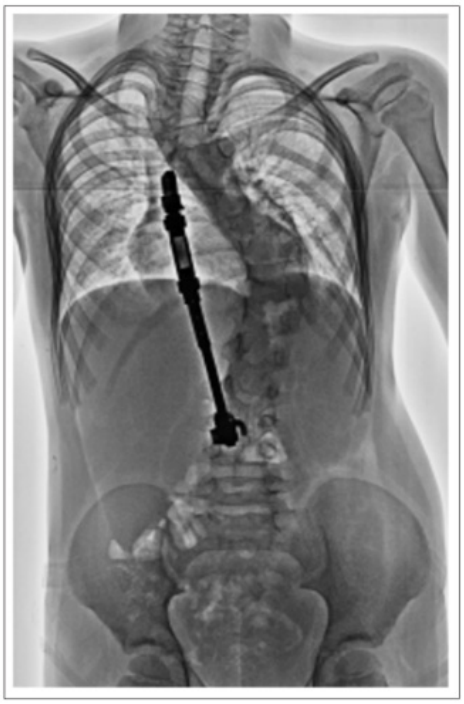


Figure 5B

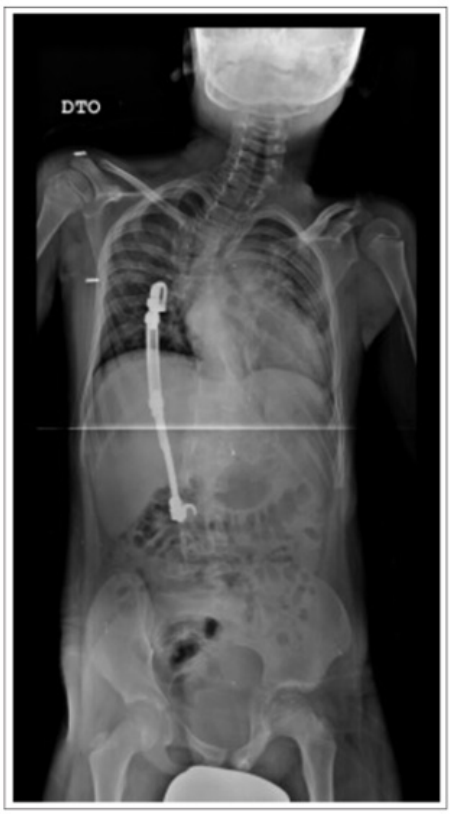


Figure 5C

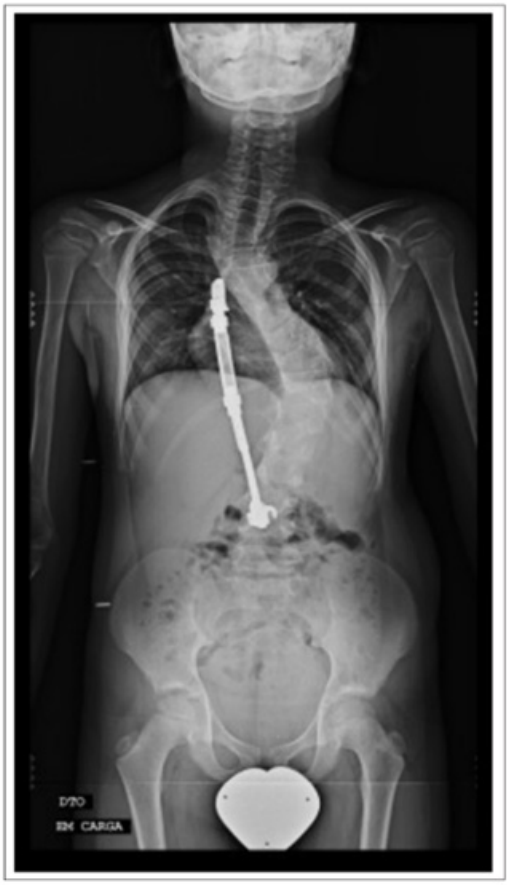


Figure 5A: AP X-Ray view of the Spine at Nine Years Old (A), at Ten Years Old (B) and at Twelve Years Old (C), after Several VEPTR Lengthenings

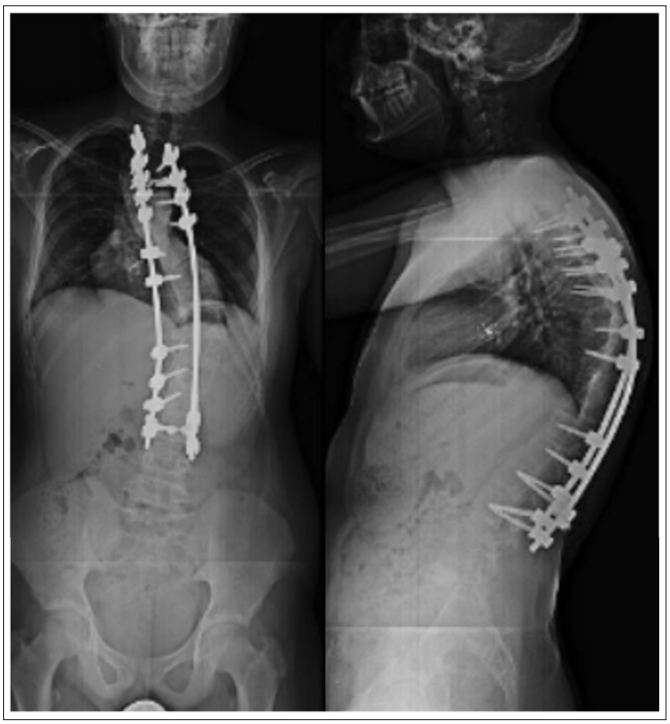
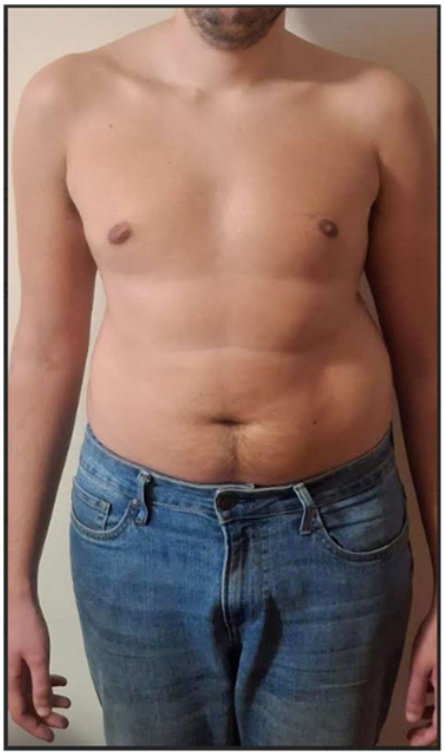


Figure 6: AP and Lateral X-Ray Views of the Spine at Five years after Posterior Instrumented Spinal Fusion



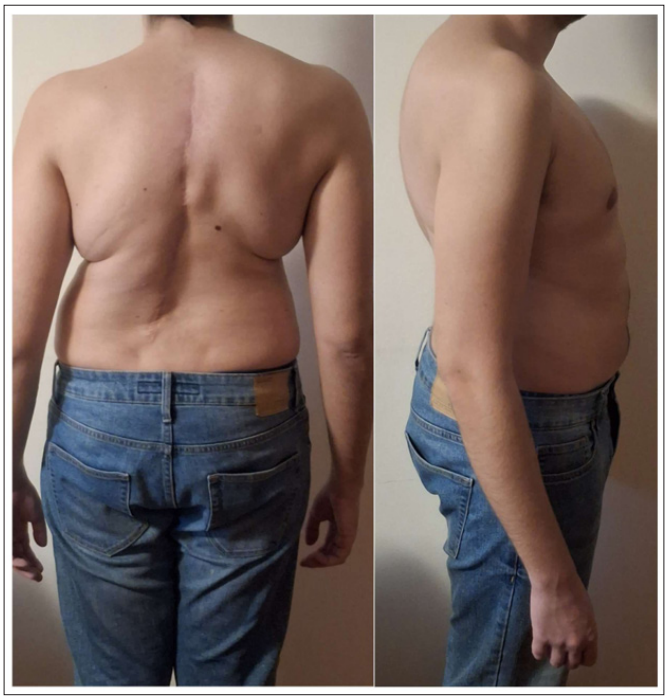


Figure 7: Clinical Photograph at Age 22, Showing Final Spinal Alignment, Thoracic / Waist Symmetry and Balanced Thoracic Kyphosis.

Discussion

The EOS case reported here, clearly shows how this congenital syndromic scoliosis can progress so rapidly during this early period of life and these are very difficult conditions to manage. Although the scoliosis is only a skeletal deformity, it can severely compromise the thoracic development and respiratory function, shortening life expectancy of these children.

This case report highlights the significance of VEPTR in managing these patients at risk of TIS, particularly in the presence of rapidly progressive congenital scoliosis with rib synostosis (VDD Type II) [3,7]. This approach serves as a viable alternative to early fusion, which has historically been associated with iatrogenic respiratory failure due to the restriction of thoracic growth [2].

A critical observation in this case was the failure of initial growth-guided strategy. Convex Growth Arrest (CGA) is often attempted as a preliminary measure to enhance gradual curve correction in certain congenital scoliosis; however, on many occasions its failure leads to very poor results that are difficult to handle [7]. As reported by Tauchi et al, the development of a marked fusion mass and the subsequent loss of bony landmarks after failed hemiepiphysiodesis make reconstructive salvage surgery extremely difficult. In our patient, the transition to concave thoracic distraction was not merely a secondary option but a necessary salvage procedure to address aggressive curve progression that initial CGA could not control, being aware that bone is dynamic tissue that alters its internal architecture and external shape in response to mechanical loading [7].

In the surgical armamentarium to deal with EOS, the traditional surgical procedures varied among the different types of spine-based instrumentation, such as traditional growing rods (TGR) or the more recent Magnetically Controlled Growing Rods (MCGR) but a new concept to correct spinal deformity through thoracic instrumentation was introduced by Robert Campbell [4]. VEPTR was developed to prevent the risk of TIS because its primary mechanism requires direct expansion and stabilization of the rib cage through concave thoracoplasty, thereby increasing intrathoracic volume. This direct thoracic expansion is not achieved with the same efficacy by MCGR or any other growing rod system, which focuses primarily on spinal curvature correction. While MCGR may be an alternative to manage idiopathic EOS with the advantage of reducing repetitive surgeries, VEPTR remains the gold standard for primary chest wall deformities like VDD Type II [8-15].

The success of this salvage procedure in this young boy with solid thoracic spine fusion is largely supported by the high plasticity of growing bone. As seen in reported case, skeletal/bone plasticity allows the rib cage and the spine to adapt structurally to mechanical loading and expansion by remodeling its shape, density, and structure according to the Wolff principle [16]. This biological adaptability is crucial when applying extrinsic forces to a constricted hemithorax, enabling not only space for lung development but also the structural reconfiguration of the thoracic cage and thoracic spine during this period of skeletal growth [4].

Despite the benefits in controlling spinal deformity and preserving lung potential, the high rate of complications remains a concern. The repetitive nature of growth-preserving surgeries requires periodic lengthening under general anesthetics [10,11]. Literature reports complication rates as high as 137% per patient or 40% per surgery [11]. VEPTR and traditional growing rods (TGR) exhibit comparable incidences of implant failure per surgery (approximately 4.3% and 4.0%, respectively), with more severe stiffness of both segments of the skeleton (chest wall and instrumented spine) with repeated distraction. Although the distractive device used in this case was “hybrid”, not pure spine to spine or rib to rib, we believe that it could have been the reason for the distraction device to work throughout several years, relying more on mechanical loading of the growing bone to remodel the shape and structure of this segment of the spine [10-13].

Although during the whole process we did not have any mechanical failures, implant failures in this type of instrumentation are frequent and more likely to occur early in the distraction process, when resistance is high but bone and soft tissues are not strong enough as the anchor sites. However it does highlight the need for multi-stage interventions to prevent progression of the scoliosis during the different stages of spinal growth, achieving a definitive posterior instrumented fusion during the adolescent growth spurt before skeletal maturity [14]. Although in this case, the optimal curve reduction was reached by age ten, surgical waitlist restrictions delayed final fusion until age 13. This delay was clinically significant, as a timely transition is crucial to prevent curve progression and long-term implant complications and it explains why there was a curvature aggravation between the age of ten and thirteen when he underwent final spinal procedure [13-14].

This paper has limitations, including its retrospective nature and being a case report. Furthermore, while follow-up extended to skeletal maturity, longitudinal spirometry data was unavailable and conventional radiographs 20 years ago did not allow us to measure spinal lengthening between the different procedures.

Conclusion

The clinical case presented demonstrate the essential role of VEPTR in the management of rapidly progressive congenital scoliosis in patients at risk of TIS. In the context of these complex EOS, VEPTR can be a useful tool to enhance lung growth potential, avoiding the deleterious consequences of premature thoracic spine fusion.

As evidenced by the overall progression of the case reported, the clinical success of this approach should not be assessed solely by the correction of the Cobb angle. When initial growth-sparing techniques like CGA fail, concave thoracic distraction can be part of a vital salvage strategy at this stage of pubescent growth. The primary measurement of success should be based on the stabilization and preservation of respiratory function, the maintenance of vertical trunk growth and the control of spinal deformity without compromising thoracic capacity.

In summary, VEPTR instrumentation represents a paradigm shift in how to deal with certain complex paediatric spinal deformities. Its focus extends beyond the correction of skeletal deformity; it is fundamentally about protecting cardiorespiratory potential and preserving spinal lengthening and long-term quality of life for children with aggressive spinal deformities of the thoracic spine.

Acknowledgments

None Declared. The authors report no conflicts of interest.

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