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Concave Thoracic Distraction System as Salvage Procedure for Progressive Congenital Scoliosis at Risk of Thoracic Insufficiency Syndrome: Report of Two Cases

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ABSTRACT

Early-Onset Scoliosis (EOS) is a complex pediatric spinal deformity that progresses rapidly. Young children with EOS may develop impaired pulmonary function due to the high risk of aggravation of spinal curvatures and thoracic constraints during a critical period for lung development. 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 these cases, aggressive curve progression and thoracic constriction can compromise severe lung development at this early stage of life. Historically, early spinal fusion of the thoracic spine resulted in iatrogenic restrictive lung disease due to shortening of thorax "height" and its consequences. Although spinal hemiepiphysiodesis is a recognized growth-guided technique, appropriate for this type of condition, the outcome can be rather unreliable and its failure can lead to severe residual chest and spinal deformities that raises significant reconstructive challenges.

Case Reports: *We present two cases of Congenital Scoliosis with Rib Synostosis at risk of TIS, treated with expandable thoracic distraction until final posterior instrumented fusion in adolescence. Case 1 is a two-year-old boy with a 68° thoracic scoliosis. Following a hemiepiphysiodesis, the deformity progressed to 93° by age six, requiring a salvage procedure. He was treated with an Expandable Thoracic Distraction System, later converted to VEPTR with subsequent repeated distraction program, resulting in a final Cobb angle of 55°. Case 2 is a 2 year old boy with Goldenhar Syndrome whose thoracic curve progressed to 114° despite a convex growth arrest at 2 years of age. VEPTR implantation at age five with subsequent repeated distraction program successfully controlled the deformity. In both cases patients reached skeletal maturity for definitive spinal fusion in a stable condition with preserved respiratory function.*

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 these more invasive salvage distraction techniques to manage aggressive curve progression. Clinical success should be assessed by the stabilization of respiratory condition and preservation of vertical trunk growth, rather than by Cobb angle correction only.*

Keywords
Early-onset Scoliosis, Congenital Scoliosis, VEPTR, Thoracic Insufficiency Syndrome, Complications, Volume Depletion Deformities, Expandable Thoracic Distraction System.

Introduction
Early-onset scoliosis (EOS) is referred to any spinal deformity that is present before the age of 10 [1,2], regardless of the etiology and it can be one of the most complex challenges in pediatric orthopedics [1]. As a rapidly progressive deformity it can compromise not only the spinal growth but also lung development. As a consequence of this condition, the Thoracic Insufficiency Syndrome (TIS) defined as the inability of the chest to support normal breathing or lung growth [2], can be one of the major complications associated to Volume Depletion Deformities (VDD) [3,4]. These are three-dimensional distortions of the chest wall resulting in volume loss, classified as rib absence (Type I), rib fusion (Type II), and global hypoplastic chest (Type III) [1], leading to segmental hypoplasia of that hemithorax. However as growth of the lung is triggered by volume expansion and tissue hypertrophy, segmental hypoplasia may compromise thoracic volume development and pulmonary function.

Historically, early spinal fusion was the standard treatment, effectively correcting the Cobb angle but inhibiting vertical trunk growth—the primary determinant of intrathoracic volume [4]. This often resulted in severe restrictive respiratory failure later on in life, exchanging a skeletal deformity for an iatrogenic lung disease.

However, some congenital scoliosis associated with rib synostosis (VDD type II) also has a devastating natural history. Such a rapidly progressive deformity at this young age can compromise not only skeletal axial development but as the concave hemithorax becomes longitudinally constricted and shortened, it threatens lung development due to alveolar hypoplasia and decreased multiplication [4]. To manage this, growth-guided procedures in the spine such as hemiepiphysiodesis have been used with unreliable results [5]; its failure often leads to a marked spinal fusion mass with loss of bony landmarks, making subsequent reconstructive salvage surgery extremely difficult [3] and risky. The main objective of a convex growth arrest is to modulate spinal growth, inhibiting on the convex side of the curvature and allowing the concave aspect to grow, thus correcting the curvature progressively. However, the post-operative behavior of these curvatures can be quite unpredictable [5] with less than optimal outcome and therefore other more reliable alternatives have been developed [6].

The development of the Vertical Expandable Prosthetic Titanium Rib (VEPTR) by Campbell presented a significant advancement [4,6] in the treatment of these EOS with VDD. It was conceived for the direct treatment of patients at risk of TIS [2,6], with emphasis on Type II VDD [6]. The Opening Wedge Thoracoplasty facilitates the insertion of this device to mechanically expand and stabilize the rigid constricted rib cage [4,5] or the chest wall. This dual mechanism aims to progressively restore the thoracic volume to allow proper lung growth, while correcting the scoliosis [4,6,7] simultaneously, fulfilling the growth-guided strategy of preserving spinal growth and maximizing pulmonary development [7].

In this context, we present two cases of Congenital Scoliosis with rib fusion at risk of developing TIS, in which the use of Expandable Thoracic Distraction devices proved to be effective as a salvage procedure in the strategy to correct a rigid fused thoracic curvature.

Case Presentation
Case 1 concerns a 2 year old boy diagnosed with congenital thoracic scoliosis (concave unsegmented bar with convex hemivertebrae), presenting an initial Cobb angle of 68o and right rib cage synostosis (Figure 1). At age three years and eleven months, the patient underwent anterior and posterior hemiepiphysiodesis of the convex half of the thoracic curve. However, the spinal deformity progressed aggressively (Figure 2), reaching 93º by age six years and five months (Figure 3). This marked progression was associated with a high risk of Thoracic Insufficiency Syndrome (TIS) due to Type II Volume Depletion Deformity (VDD), signaling the failure of the initial procedure.

In response to this failure and the technical challenges of a post-hemiepiphysiodesis residual deformity [3] that was gradually progressing, a salvage growth-sparing approach was chosen. The patient underwent an open concave wedge thoracoplasty and implantation of an Expandable Thoracic Distraction System (rib-to-rib) (Figure 4). As the VEPTR device was not yet commercially available in Portugal at that time, an adapted system was initially inserted, that was subsequently revised to a VEPTR at age seven (Figure 5). Six subsequent lengthenings were performed in accordance with the child's growth (Figure 6).

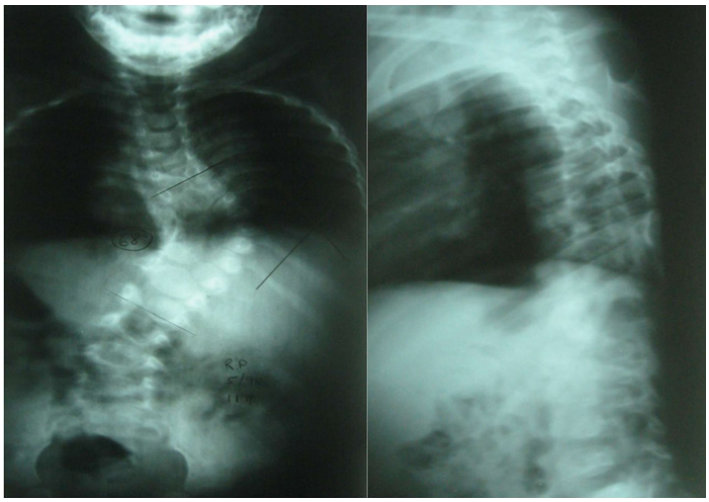


Figure 1: Case 1 - AP and lateral X-Ray views of case 1 patient's spine at 2 years old with a Cobb angle of 68º.

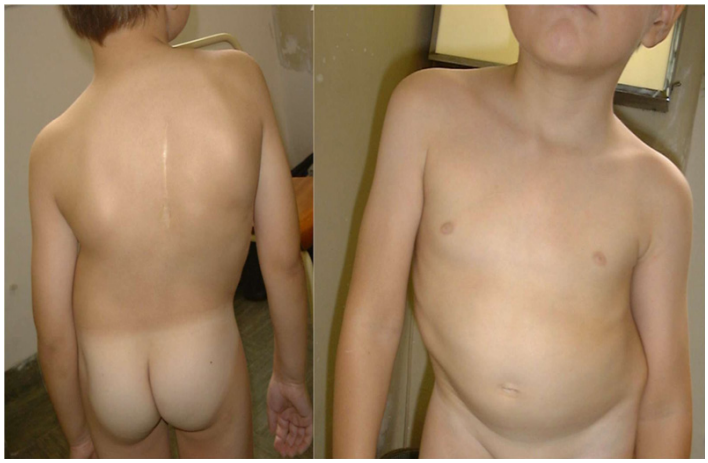


Figure 2: Case 1 - spinal deformity

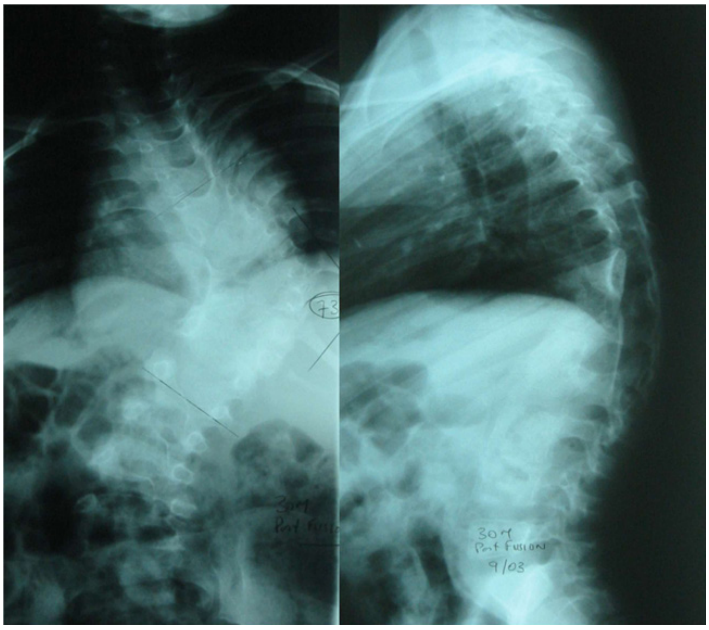


Figure 3: Case 1 - AP and lateral X-Ray views of the spine at six years and five months with a Cobb angle of 93º.

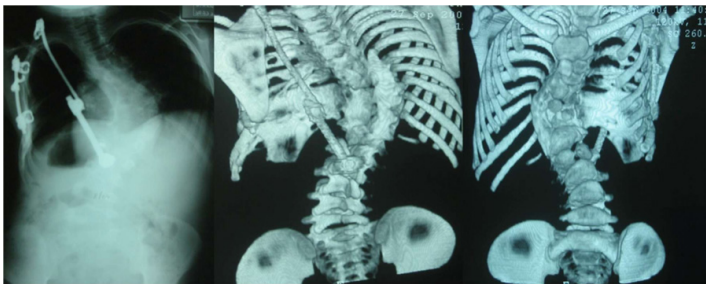


Figure 4: Case 1 - AP X-Ray and 3D CT-scan spine reconstruction after thoracoplasty and implantation of an Expandable Thoracic Distraction System (rib-to-rib).

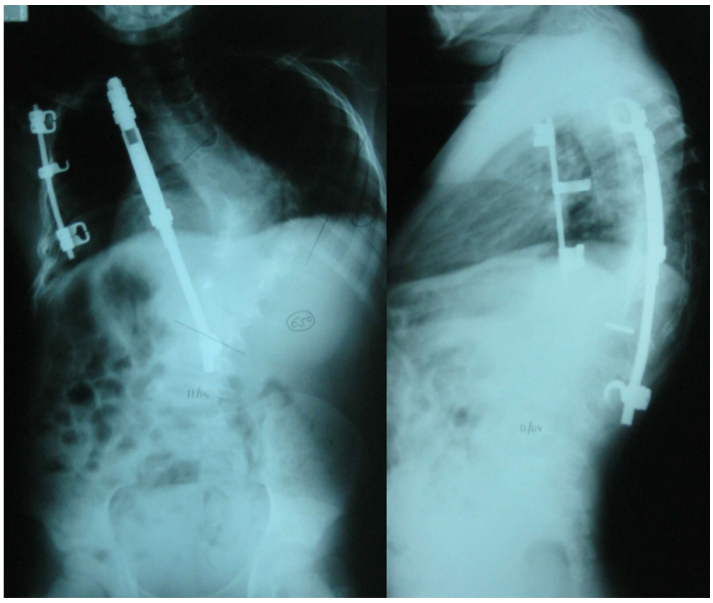


Figure 5: Case 1 - AP and lateral X-Ray views of the spine at 7 years after new thoracoplasty and VEPTR implantation.

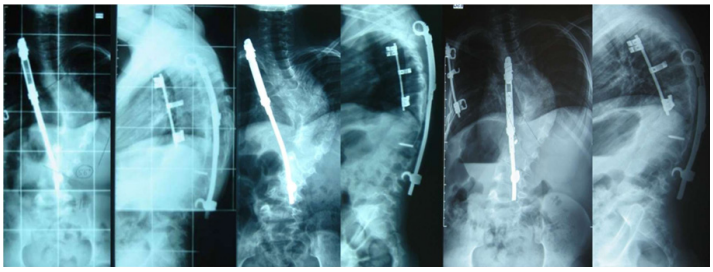


Figure 6: Case 1 - AP and lateral X-Ray views of the spine after VEPTR lengthenings.

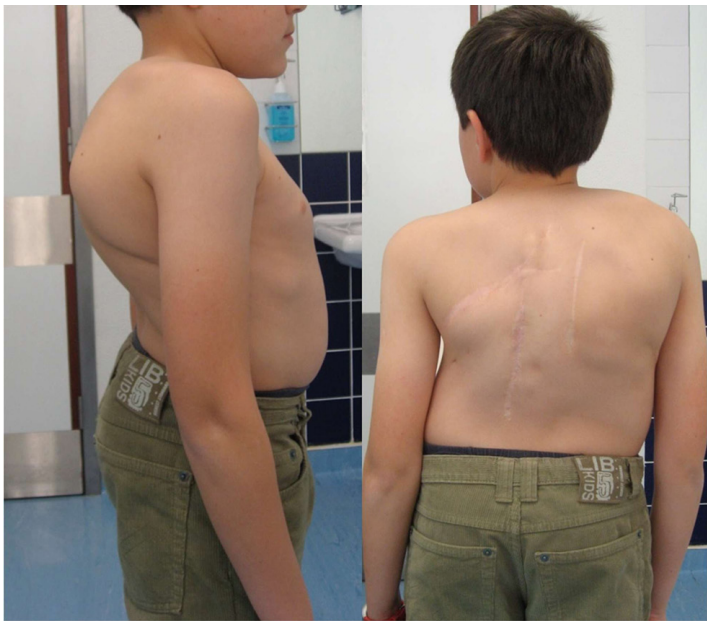


Figure 7: Case 1 – spinal deformity after VEPTR lengthenings.

At the most recent follow up, 17y of age, 4 years post- posterior definitive instrumented fusion, radiograph showed a significant cobb angle correction to 55º (Figure 6). Clinical and respiratory outcomes were highly satisfactory, with the curve controlled and the patient remaining respiratory asymptomatic (Figure 7). Despite two additional procedures to manage distal fixation complications (hook dislocation), the outcome remained favorable. Final procedure was removal of VEPTR device and posterior instrumented fusion with several posterior column osteotomies at age of 13 (Figures 8 and 9), marking the end of the growth-sparing phase with preservation of thoracic function.

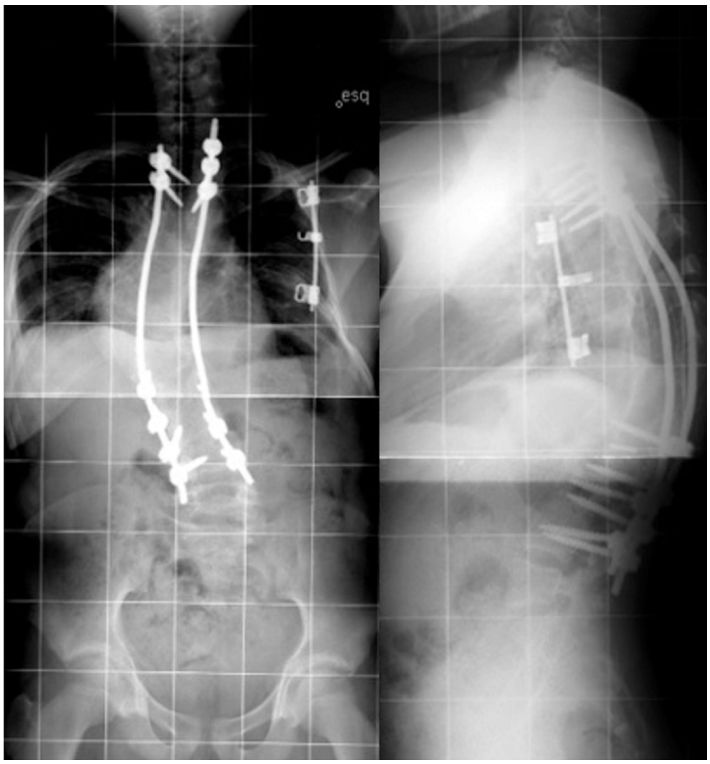


Figure 8: Case 1 - AP and lateral X-Ray views of the spine after posterior fusion.

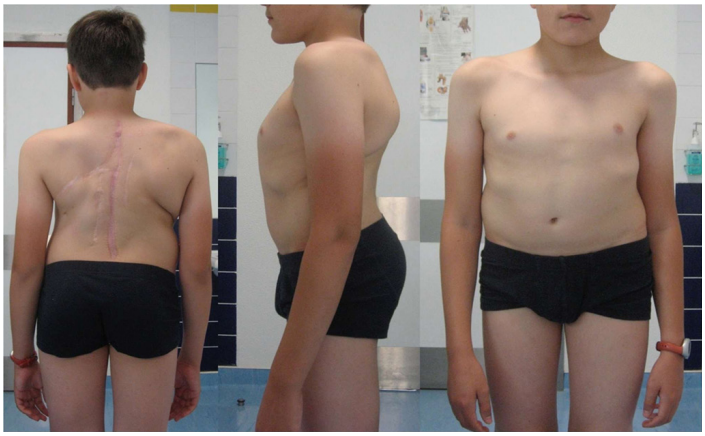


Figure 9: Case 1 – Spinal deformity after posterior fusion.

Case 2 concerns 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 10). Due to curve progression to 75º by age three (Figure 11), the patient underwent anterior and posterior hemiepiphysiodesis. Notwithstanding this procedure, the curve continued to progress to 90º at age four and reached a critical 114º at age five (Figure 12).

This aggressive progression, indicating an imminent risk of TIS, required concave thoracic expansion. At age five, the patient underwent salvage surgery consisting of a VEPTR device (rib to spine fixation) to manage both the spinal and the thoracic deformity (Figure 13).

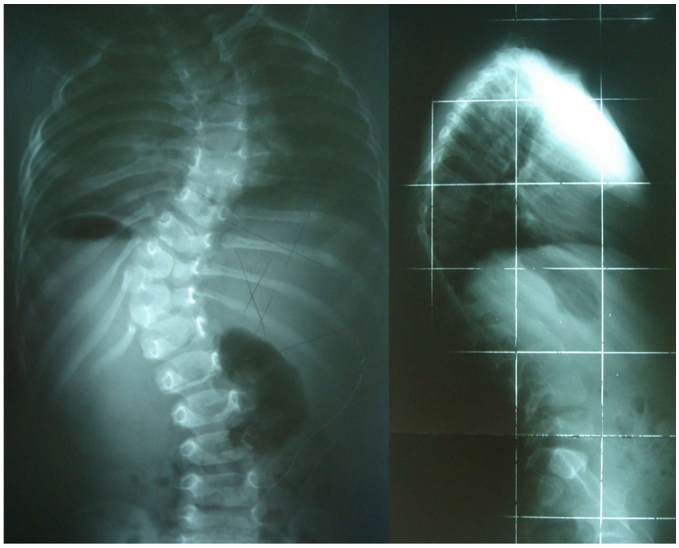


Figure 10: Case 2 - AP and lateral X-Ray views of the spine at two years of age – hemivertebrae, butterfly vertebrae and rib fusion and a main Cobb angle of 45º.

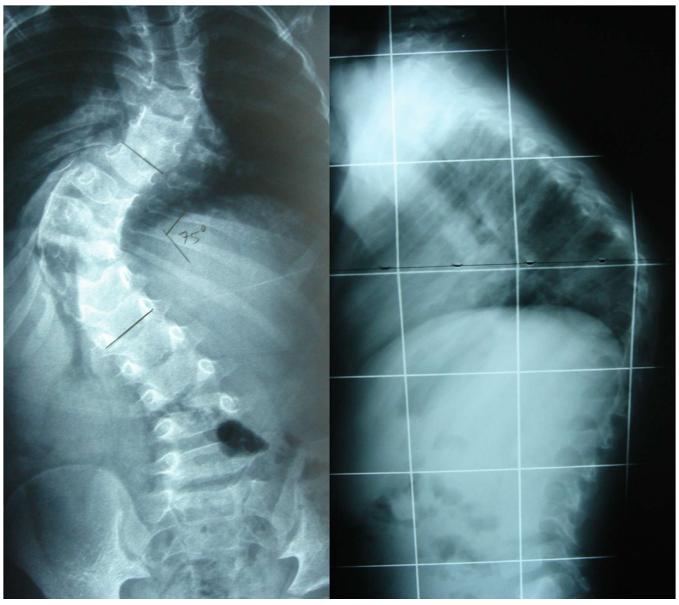


Figure 11: Case 2 - AP and lateral X-Ray views of the spine at three years old with a Cobb angle of 75º.

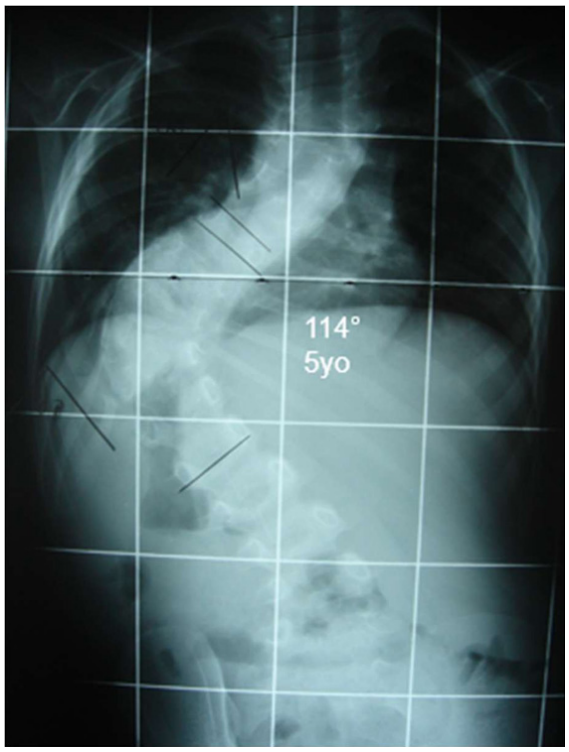


Figure 12: Case 2 - AP X-Ray view of the spine at five years old with a Cobb angle of 114º.

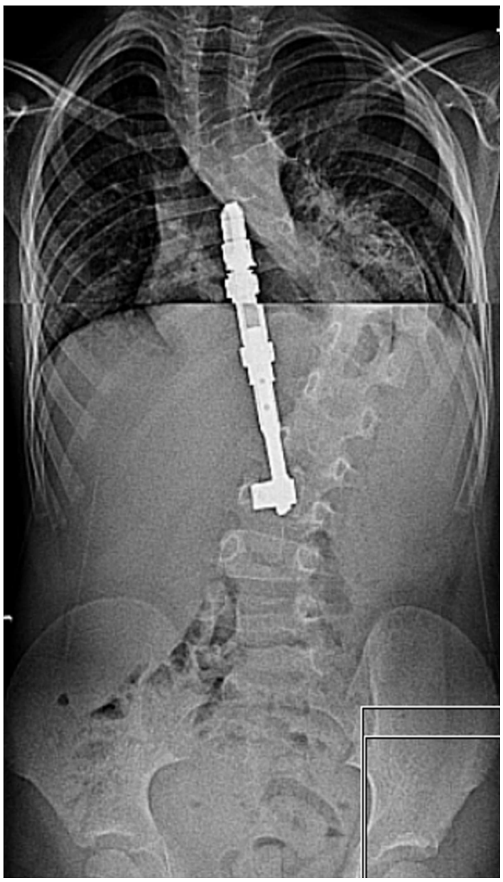
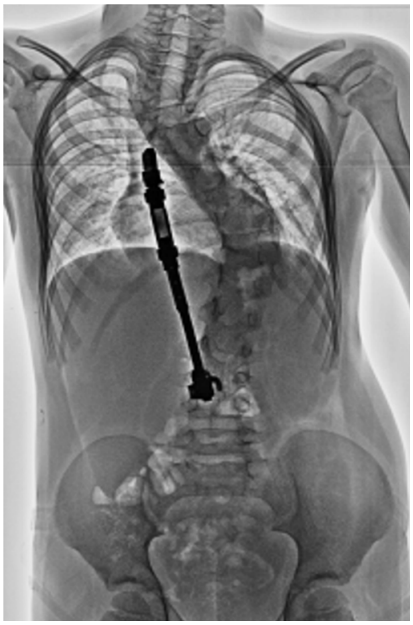
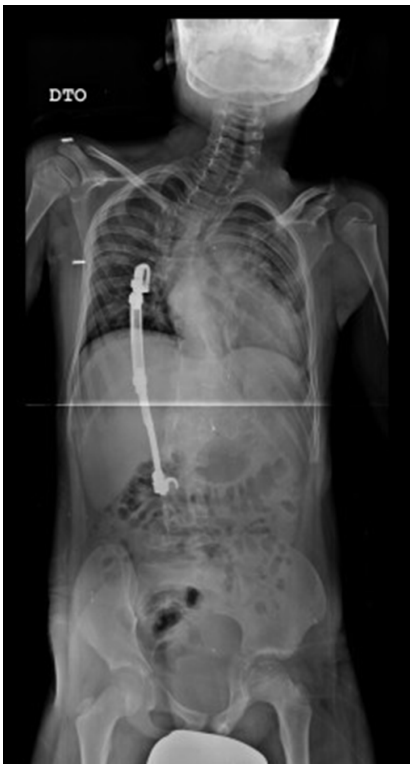


Figure 13: Case 2 - AP X-Ray view of the spine at five years old after thoracoplasty and VEPTR insertion.

By ten years of age, the patient exhibited excellent curve correction and reached the end of the growth-sparing phase (Figure 14). 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 15). 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 16) with the strategy chosen.





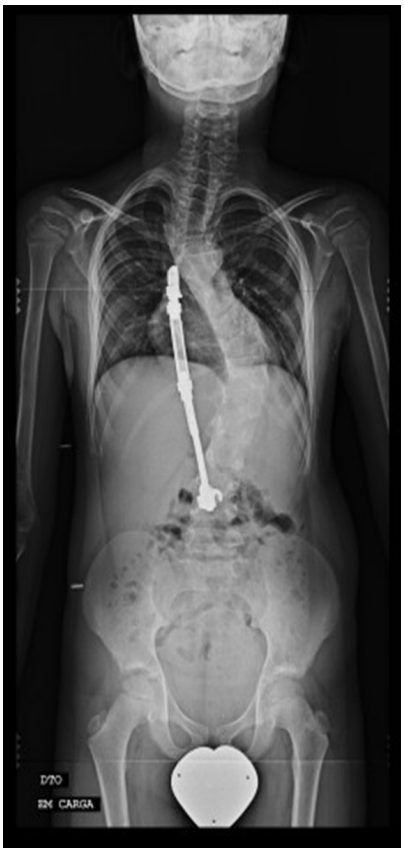


Figure 14: Case 2 - AP X-Ray view of the spine at nine years old (left), at ten years old (middle) and at twelve years old (right), after several VEPTR lengthenings.

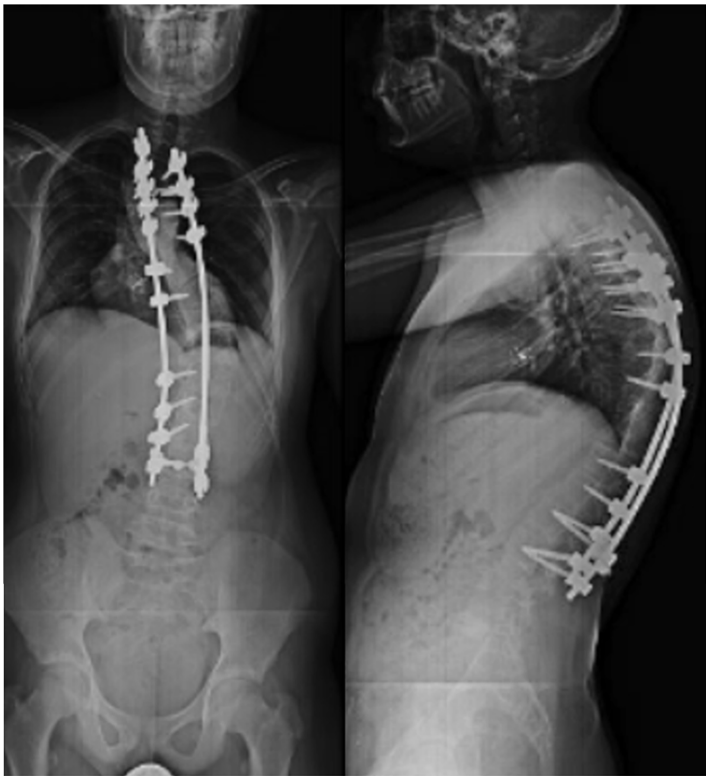
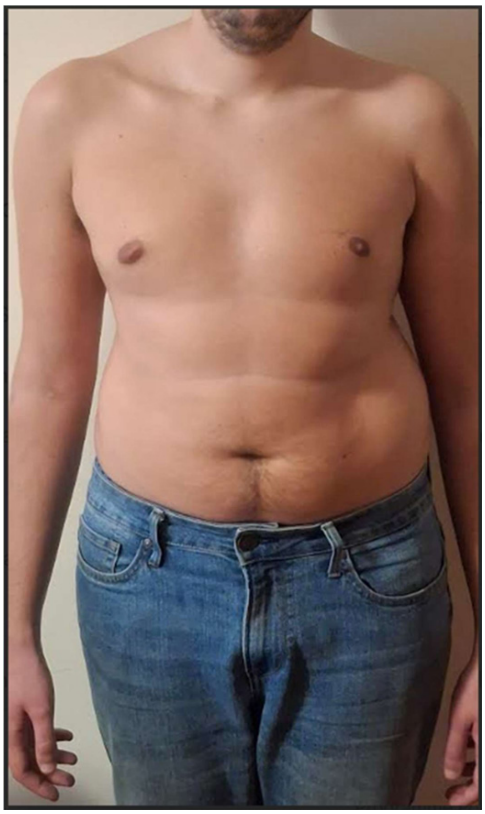


Figure 15: Case 2 - AP and lateral X-Ray views of the spine at five years after posterior instrumented spinal fusion.



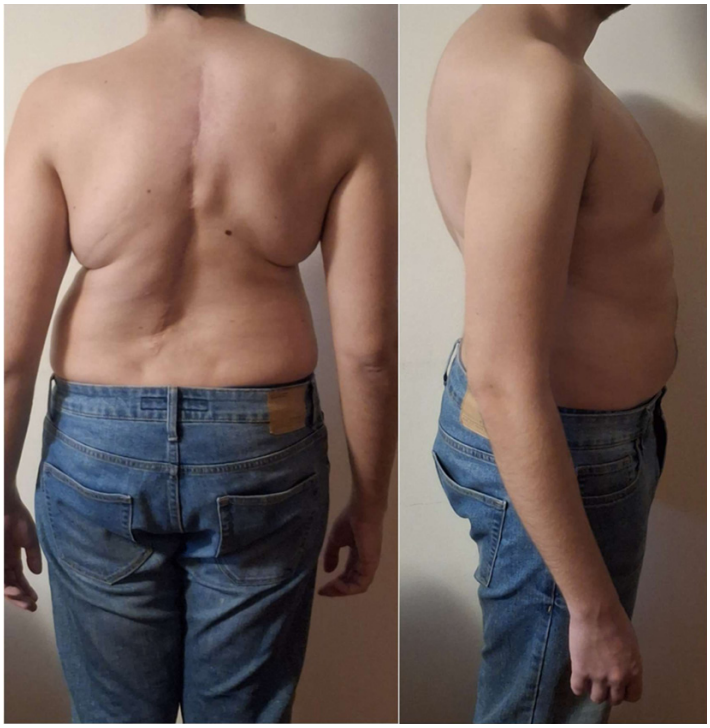


Figure 16: Case 2 – Clinical photograph at age 22, showing final spinal alignment, thoracic / waist symmetry and balanced thoracic kyphosis.

Discussion

The two cases of EOS reported here, highlight 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 both cases 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, its failure leads to significant clinical and technical challenges [7]. As reported by Tauchi et al. [7], 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 patients, 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 [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) [7] 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 [4,8]. This direct thoracic expansion is not achieved with the same efficacy by MCGR or any other growing rod systems, which focuses primarily on spinal curvature correction [9,15]. 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 [2,9,15]. The success of this salvage procedure in these two young boys with solid thoracic spine fusions is largely supported by the high plasticity of growing bone. As seen in Case 2, 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-Virchow principle. 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) [12,13], with more severe stiffness of both segments of the skeleton (chest wall and instrumented spine) with repeated distraction. Although the distractive devices used in the two cases were “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 on mechanical loading of the growing bone to remodel the shape and structure of this segment of the spine.

The two additional procedures for relocation of the distal fixation in case 1, reinforces evidence that implant failures in this type of instrumentation are more likely to occur early in the distraction process, when bone and soft tissue resistance are high [13]. Both cases highlight the necessity of multi-stage interventions to prevent rapid progression of the scoliosis during the different stages of spinal growth, culminating in definitive posterior instrumented fusion during the adolescent growth spurt before achieving skeletal maturity [14]. In Case 2, although optimal curve reduction was reached by age ten, surgical waitlist restrictions delayed final fusion until age 13. This delay is clinically significant, as a timely transition is crucial to prevent curve progression and long-term implant complications [14].

This report has limitations, including its retrospective nature and the small, heterogeneous sample size. 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. Nevertheless, these cases underscore the role of VEPTR as a life-altering salvage strategy for children with rapidly progressive congenital EOS at high respiratory risk.

Conclusion

The two cases presented demonstrate the essential role of VEPTR in the management of rapidly progressive congenital scoliosis in patients at risk of Thoracic Insufficiency Syndrome (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 these cases, the clinical success of this approach should not be assessed solely by the correction of the Cobb angle. When initial growth-sparing techniques like hemiepiphysiodesis 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.

To conclude, VEPTR instrumentation represents a paradigm shift in how to deal with certain complex pediatric 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 thoracic-spinal deformities.

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