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Operative management of congenital early-onset scoliosis using the vertical expandable prosthetic titanium rib (VEPTR): a case series

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ABSTRACT

To analyze surgical challenges and outcomes in patients who completed the whole journey of vertical expandable prosthetic titanium rib (VEPTR) treatment for congenital early-onset scoliosis (C-EOS), given the limited evidence available on VEPTR graduates. A retrospective review was conducted on nine consecutive patients with C-EOS and thoracic hypoplasia treated at a single tertiary care center, with assessment of clinical and radiological outcomes. At mean duration of 7.4 (range 4.3–10.5) years of VEPTR treatment, the mean coronal deformity angle measured 65° preoperatively, 50° postoperatively, and 58° at final follow-up. Mean T1–S1 length (pre-op 252 mm, final follow-up 333 mm) and T1–T12 length (preop 128 mm, final follow-up 196 mm) improved by 32% at final follow-up. Mean space available for lung was 86% (range 79%–93%) preoperatively, increasing to 90% (range 85%–95%) postoperatively and 97% (range 87%–107%) at final follow-up. Nine children had a cumulative 17 (188%) complications comprising wound problems, infection, and device migration or prominence. In five patients who underwent definitive fusion, mean coronal deformity angle and T1–S1 length improved by 17% and 11%, respectively. VEPTR is valuable in managing EOS, particularly in patients with thoracic insufficiency syndrome. However, the need for multiple surgeries, limited correction potential, and risk of partial loss of correction make it less suitable for other cases.

INTRODUCTION

Infants with congenital chest and spine abnormalities develop life-threatening extrinsic restrictive lung disease, leading to the term thoracic insufficiency syndrome (TIS); defined as the inability of the thorax to support normal respiration or lung growth.[1] The best chance to reverse or stabilize the TIS is an operative strategy that aims to achieve the largest, most symmetrical, most functional thorax by skeletal maturity.[1] This goal is accomplished through an open thoracic reconstruction and using the vertical expandable prosthetic titanium rib (VEPTR) implants to enlarge and stabilize the malformed chest (expansion thoracoplasty) along with indirect correction of the scoliosis. Though originally developed for TIS, over time the surgical strategies and VEPTR principles allowed for addressing the various themes of volume depletion deformities of the thorax, either due to absent or fused ribs or due to the hypoplastic thorax.[2]

Early and long-term studies have shown VEPTR implantation sufficiently controls scoliosis, achieves spinal lengthening, and balances pelvic obliquity well.[3 4] Gantner *et al*. demonstrated a correction in the thoracic deformity from a mean of 68° prior to surgery to a mean of 45° (p<0.001) after implantation of VEPTR.[3] Respiratory function also improves, at least initially, though due to chest wall stiffness, this is not maintained over the child's growth. Nossov *et al*. evaluated the assisted ventilation rating (AVR) to assess respiratory function and found that the AVR improved in 24% patients and remained stable in 64% patients with 16 (20%) children no longer requiring any external respiratory support.[5] The procedure is not free from complications, which include metalwork-related (migration/dislodgement of the anchors, hardware failure), infections, rib fractures, soft tissue problems, ossifications, and neurological insult including brachial plexus injury.[6–10]

Despite being in use for nearly two decades, there is scant literature published about VEPTR graduates who completed VEPTR treatment. The common VEPTR treatment end-points are final fusion, implant removal, or no further surgery after the last lengthening.[4 11 12] In January 2004, our spinal unit became the first in the UK to insert VEPTR instrumentation for early-onset scoliosis (EOS). We analyzed our single-center series of VEPTR treatment for congenital scoliosis and followed up the patients after they

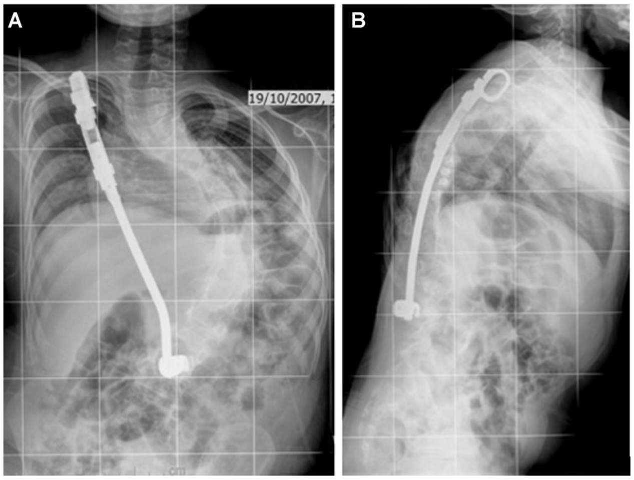

Figure 1 Antero-posterior (A) and lateral (B) view radiographs of the patient following insertion of VEPTR demonstrating the coronal deformity of congenital early-onset scoliosis and a rib to spine VEPTR on the right side. VEPTR, vertical expandable prosthetic titanium rib.

completed VEPTR treatment, attained skeletal maturity, or had final fusion surgery. The aim of the study is to have a comprehensive overview of the entire patient journey from the VEPTR implantation to final graduation, thus adding to the existing knowledge regarding managing this challenging pathology.

CASE SUMMARY

We retrospectively reviewed nine patients (6 males and 3 females) with congenital early-onset scoliosis and thoracic hypoplasia treated with VEPTR (figure 1). Six patients were treated with a hybrid construct (both rib–spine and rib–rib implant), whereas one patient each had a rib–rib construct, a rib–spine construct, and a rib–pelvis construct dependent on the curve morphology and the associated thoracic hypoplasia. Planned expansion surgery was performed at 6–12 monthly intervals as deemed necessary by regular outpatient assessments. There were no additional patients from the center who were excluded from study consideration.

Absence of extensions over 2 years was considered as the completion of VEPTR treatment. Similarly, completion of skeletal growth with no further progression of scoliosis was also deemed to be the end of VEPTR usage. Final management involved one of the following three outcomes: VEPTR removal without fusion (*n*=2); VEPTR removal followed by instrumented fusion (*n*=5); and retention of VEPTR with completion of treatment (*n*=2).

Radiological assessments on erect Postero-anterior (PA) scoliogram included main coronal curve deformity angle, coronal balance measured by amount of deviation from the C7 plumb line from the central sacral vertical line, spine and thorax lengthening (T1–S1 and T1–T12 measurements, respectively). The space available for the lung (SAL) measurement denotes the relationship between the unfolding of the concave lung space and the straightening of the spine.[1] Sagittal profile measurement included the main curve kyphotic angle. These values were measured preoperatively, immediate post-operative period, at the end of VEPTR treatment, and at final follow-up.

The mean age of the cohort at the initial VEPTR surgery was 5.4 years with age ranging between 1.1 and 8.9 years. One child had Isola growth rods converted to VEPTR at the age of 8 years.

The average follow-up period from the first surgery was 11.7±3.4 years (range 7.7–16.1 years). The average duration of VEPTR treatment was 7.4±3.1 years for the whole group (range 4.4–12.8 years), during which a mean of 9 (range 6–16) number of VEPTR expansions were performed per patient.

Radiological assessment from index treatment to latest radiographs for the entire group, irrespective of final implant outcome, was available at a mean of 11.4 years (range 8.4–14.4 years) (table 1). The main curve deformity angle measured a mean of 65° preoperatively (36–94°), 50° postoperatively, and 58° at final follow-up.

Mean T1–S1 length measured 252 mm preoperatively (205–293 mm) increasing by 12 mm (5%) postoperatively, and by 81 mm (32%) at final follow-up. A similar increase (32%) was achieved for thoracic height (T1–T12 length). Neutral coronal balance was achieved in 75% of patients postoperatively, but this decreased to 33% of patients at final follow-up. Mean % SAL was 86% preoperatively, increasing to 93% postoperatively and 97% at final follow-up. Main

Table 1 Radiographic measurements

Variables	Preoperative	Immediate postoperative	At end of VEPTR treatment	Final follow-up
Main curve coronal deformity angle	65° (36°–94°)	50° (20°–89°)	63° (17°–90°)	58° (21°–89°)
T1–S1 length, millimeters	252 (205–282)	264 (226–306)	307 (214–408)	333 (219–410)
T1–T12 length, millimeters	128 (95–161)	145 (131–159)	170 (137–203)	196 (154–238)
Main kyphosis angle	56° (16°–96°)	47° (27°–67°)	55° (37°–73°)	55° (31°–79°)
SAL concave/convex,%	86 (79–93)	90 (85–95)	96 (84–108)	97 (87–107)

Data was presented as mean (range).
SAL, space available for lungs; VEPTR, vertical expandable prosthetic titanium rib.

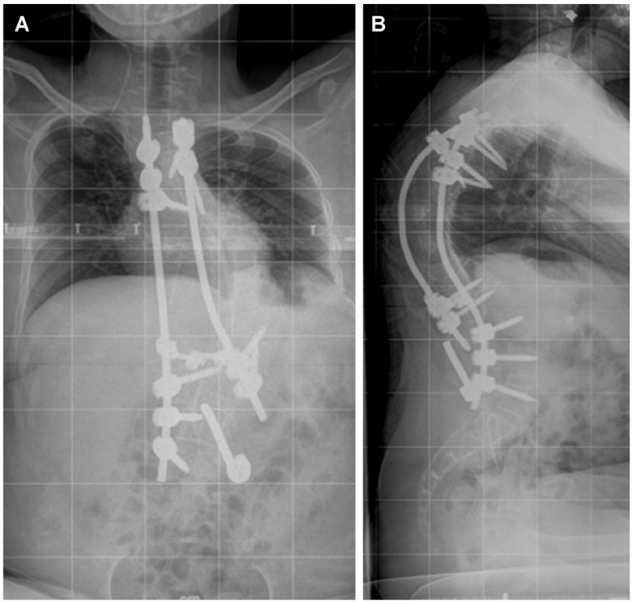

Figure 2 Antero-posterior (A) and lateral (B) view radiograph of a VEPTR graduate who went through VEPTR extensions, removal of VEPTR, halo traction, and definitive fusion in the form of posterior instrumented scoliosis correction. Note the improvement in space available for the lung. Distal section of VEPTR was left in situ. VEPTR, vertical expandable prosthetic titanium rib.

kyphotic angle, though improved immediate postoperatively, reverted back to preoperative levels (55°) at final follow-up.

Five patients had a definitive posterior spinal fusion performed after a mean of 6.4 years (range 4.4–9.2 years) of VEPTR treatment. The mean age at final fusion surgery was 13.3 years (range 11.6–14.2 years). Four of these required a staged correction, including three patients who required halo-pelvic or halo-gravity traction. All patients needed instrumentation up to or beyond the previous implant levels (figure 2). At the staged procedures, extensive ossification around the implants and the spinal column was noted in all patients, as identified on CT images preoperatively. In total, 13 staged procedures were done to achieve fusion over a period ranging from 3 weeks to 23 months in these five patients.

Compared with the end of the VEPTR treatment, the mean coronal deformity corrected from 80° to 67° (17%) and T1–S1 spine length improved from 272 mm to 301 mm (11%) following the fusion (table 2). The SAL also improved marginally over the course of treatment. However, two children had worsening of the main kyphosis, which led to reduced T1–T12 length gained. Only one child had neutral coronal balance at final follow-up. At a mean follow-up post-fusion of 4.1 years (1.1–7.8 years), two (40%) patients had wound problems, of whom one needed multiple wound-washout, removal of metalwork, and required plastics intervention. This patient had kyphoscoliosis to begin with, and unfortunately, over time, ended up with a razorback spine deformity. Four patients had a final non-fusion strategy after completion of the VEPTR treatment. The construct was left in situ following skeletal maturity in two patients. One patient died due to acute respiratory distress syndrome and respiratory failure, more than three-and-a-half years, after completion of the VEPTR treatment. In the two remaining patients, the VEPTR construct required removal, but no further surgery is planned as the curves are static. Of these in one child, significant autofusion was evident on VEPTR removal surgery, and despite halo-gravity traction, no further correction was achieved. The radiological parameters remained stable at subsequent follow-up reviews for both these patients.

Complications

Mean total number of surgical procedures per patient was 14 (10–20) of which 21% of procedures were unplanned. All nine children had a cumulative 17 complications (188%) during the course of VEPTR treatment. Five (55%) patients had wound complications, with two needing surgical debridement. Six (66%) patients required metalwork revision (excluding rod exchange) for loosening, prominence, migration, or pull-out of the hardware. One patient died more than 3 years after the last VEPTR expansion from causes not related to the surgical intervention, but due to respiratory failure. In addition, many

Table 2 Definitive fusion group (n=5) radiological assessments

Variables	Pretreatment	Prefusion	Postfusion (final)	*P* value
Coronal deformity angle	68° ± 16°	80° ± 8°	67° ± 23°	0.575
Kyphosis angle	71° ± 32°	67° ± 9°	69° ± 18°	0.707
T1–S1, millimeters	254 ± 34	272 ± 38	301 ± 64	0.565
T1–T12, millimeters	136 ± 39	163 ± 36	157 ± 50	0.866
SAL, %	88 ± 9	100 ± 15	102 ± 9	0.135
Neutral coronal balance	2 out of 5	2 out of 5	1 out of 5	0.790

Date was presented as mean ± standard deviation.
SAL, space available for lungs.

of the children had repeated respiratory infections and restrictive lung disease that required multidisciplinary input from the pulmonologists.

DISCUSSION

Length of study and coronal correction

VEPTR treatment, though initially advocated for volume depletion deformities of the thorax primarily affecting respiratory function, has now been expanded by the majority of surgeons for mainly addressing the challenges of EOS in the very young.[13 14] Longest follow-up studies reported in the literature are 5.5 years and 6.9 years,[3 4] thereby making our series with a mean follow-up of 7.4 years the longest reported duration of VEPTR treatment to our knowledge. A multicenter review by El-Hawary *et al*. of 29 patients with heterogenous EOS with 5 years follow-up of VEPTR treatment reported the main coronal curve of 69° ± 15° preoperatively to 62° ± 16° at 5-year follow-up.[4] Maximum kyphosis, however, deteriorated from a mean of 40 preoperatively to 54° at the last review. In our series, both coronal curve and kyphosis measurements were only marginally better at the completion of VEPTR treatment, despite the initial improvement after implant insertion.

Kyphosis correction

With regards to kyphosis, Gantner *et al*. found initial improvement, but the correction could not be maintained during follow-up or restored by subsequent distraction procedures.[3] In a retrospective cohort of 14 EOS cases with thoracic kyphoscoliosis, Reinker *et al*. report kyphosis worsening by 22° by the end mean of 5.8 years follow-up.[15] The hyper-kyphosis (>60° curve) contributed to reduction in the thoracic spine length and decline in the SAL, which are detrimental to the end result of the VEPTR principle. In our series, two patients had a mean kyphosis of 90° to begin with, and it improved to a mean of 76° at the end of the VEPTR treatment. Unfortunately, the kyphosis worsened further in one child despite final fusion surgery. There is recent evidence to suggest that VEPTR should be avoided in children with >70° of kyphosis.[16] It is also found that Traditional growth rod (TGR) may be a better option for correcting the EOS kyphosis compared with VEPTR,[17 18] especially in cases of severe upper thoracic kyphosis.[19]

Growth of spine

As shown in our study, spinal length (T1–S1 and T1–T12 measurements) improved consistently over time in these long-term follow-up reports. However, our study is the only one that reports ‘true spinal growth’ defined by Wijdicks *et al*.,[20] as average growth achieved after initial instrumentation and before final fusion for growth-friendly graduates.

Complications

Complications are to be expected with any growth-friendly instrumentation which aims to achieve correction while promoting growth at the same time.[19] Sankar *et al*. reported a complication rate of 2.3 per patient with TGR compared with a rate of 2.37 per VEPTR patient.[21] Our rate of 1.8 complications per patient is in keeping with other published studies. Garg *et al*. reported from a multicenter study that 18% (38/213) of patients implanted with VEPTR developed infection requiring operative debridement.[22] Infections were more likely at the implant procedure than at expansion surgery,[23] and in the majority of cases, they can be dealt without implant removal.[24] Among the device-related issues, the proximal anchor failure is the most common reported in rib-based distraction systems with rates of 19%–53% per patient.[25] Park *et al*. have suggested that the incidence is faster in patients with neuromuscular etiology or in larger (>90 degree) curves.[26] We had four issues in our nine patients regarding proximal anchor prominence, migration, or pullout.

VEPTR graduates

The literature is short on VEPTR graduates who finish their treatment and have gone through a long and challenging course of treatment that involves numerous out-patient clinic visits, hospital admissions, and repeat surgical interventions. Seven children in our series had the VEPTR implant removed after completion of treatment, and extensive soft tissue ossification was noted during the operative intervention in all. While definitive fusion was conducted in five patients, the remaining two had very stiff curves which remained static at subsequent follow-up and were left alone. In total, 13 staged procedures were required in four out of our five patients who underwent definitive fusion, including the application of halo-traction in three children. The surgical timeline for the staged fusion ranged from 3 weeks to 23 months. Lattig *et al*. reported similar findings in their series of five VEPTR graduates, where a high degree of rigidity was found in the major and compensatory curves both intraoperatively and on preoperative CT scans.[11] Studer *et al* reported that 17 VEPTR graduates who underwent final fusion surgery had 14% and 9% improvement in main scoliosis and kyphosis curves postfusion, but both these improvements declined over longer time.[12] More than half of these patients had staged fusion procedures including halo-gravity traction and had 35% revision surgery rates and 41% complication rates following the final fusion procedures. These studies of VEPTR graduates support our results that final fusion surgery is extremely challenging with very limited corrections achieved. Spontaneous soft tissue ossification and spinal autofusion are commonly seen in patients with VEPTR, adding to the rigidity of the curve.[3 10–12 27]

Limitations of the study

Our study has several limitations due to its retrospective nature, limited number of patients, and lack of pulmonary function tests (PFTs). PFTs are difficult to obtain in very young children, and though this test was available for few patients postoperatively, we did not have adequate data for the whole group to make a meaningful analysis. Despite these limitations, we believe this long-term follow-up study adequately informs the treatment challenges faced in VEPTR graduates.

In conclusion, while VEPTR forms an important part of our surgical armamentarium in the management of patients with EOS, its utilization needs to be bespoke and tailored to individual patient needs. Among the various types of growth-sparing spinal deformity correction techniques, VEPTR is a reasonable choice for patients with TIS. However, the need for multiple surgeries, limited scope of correction, and possibility of partial loss of correction make it less suitable for other types of EOS.

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Patient consent for publication Not applicable.

Ethics approval The study was conducted in accordance with the principles of the Declaration of Helsinki and applicable ethical guidelines for the use of clinical data in research. Formal ethical approval from an Institutional Review Board (IRB) was not sought, as the study did not involve any direct interaction with human participants, the data were anonymized prior to analysis, and the research posed minimal risk to individuals.

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Data availability statement Data are available upon reasonable request. Data sheets have patient identifiers. Data are protected by UK data protection rights. It can be released to the journal on request.

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