The Usefulness of VEPTR in the Older Child With Complex Spine and Chest Deformity

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Abstract The vertical expandable prosthetic titanium rib (VEPTR) was originally designed to treat chest and spine deformities in young children. However, older children with complex spinal deformities may also benefit from placement of a VEPTR when vertebral column resections are deemed too risky neurologically. We report: (1) the changes in Cobb angle, T1 angle, and head tilt; and (2) the occurrence of complications in children older than 10 years of age treated with VEPTR. From a database of 214

patients treated in a Food and Drug Administration Investigational Device Exemption study of VEPTR, we identified 10 patients with assorted diagnoses who underwent surgery after age 10 and had a minimum of 24-month followup (mean, 39.6 months; range, 24–75 months). No patient sustained neurologic injury. Patients underwent an average of five lengthenings. The mean preoperative Cobb angle was 64.7° and improved to 48.4°. Head shift improved an average of 3.8 cm. Two device-related complications occurred (both in the same patient). Four patients have since undergone definitive spinal fusion. For a select group of patients 10 years of age or older, the VEPTR offers a reasonable alternative to potentially risky vertebral column resections for correcting deformities in selected patients.

Level of Evidence: Level IV, therapeutic study. See Guidelines for Authors for a complete description of levels of evidence.

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Introduction

The vertical expandable prosthetic titanium rib (VEPTR) was originally designed to treat thoracic insufficiency secondary to congenital anomalies with fused ribs, hypoplastic chest wall deformities, and early-onset scoliosis [2]. The device can be attached from the ribs to the spine or ilium to treat scoliosis and from rib to rib to treat chest wall deformity. The VEPTR with expansion thoracostomy treats both spine and chest wall deformity while allowing for continued growth [2, 4]. The greatest benefit of VEPTR placement is likely in children younger than 8 years of age because alveolar development can continue until this age [7, 10]. Although several reports suggest the device may increase thorax and lung volume [4, 5, 11], none have

documented improvement in pulmonary function. This may relate to the inability to obtain reliable pulmonary function tests in children of this age.

Correcting severe chest wall and thoracic spine deformities and improving cosmesis in patients older than 10 years of age remains a challenge. Vertebral column resections (VCRs) are one option but carry a high rate of neurologic complications and are technically demanding for most surgeons [8, 9]. We presumed the VEPTR might offer these patients improved cosmesis with low neurologic risk.

We therefore examined: (1) the correction obtained (Cobb angle, T1 angle, and head shift); (2) the growth in the spine (thoracic spine and hemithoracic height and width); and (3) the occurrence of complications using the VEPTR in children older than 10 years of age with severe chest wall and spine deformities.

Materials and Methods

We retrospectively identified 15 subjects older than 10 years of age at the time of their initial surgery with a minimum of 2-year followup from a multicenter Investigational Device Exemption study of 214 patients undergoing surgery with the vertical expandable prosthetic titanium rib (VEPTR) device. Currently, we consider VEPTR placement in children older than 10 years of age with complex spine deformity when: (1) secondary to medical comorbidities, the patient would not tolerate a lengthy fusion procedure; and/or (2) it is believed the cosmetic deformity, particularly shoulder asymmetry, may be improved by placement of the VEPTR. Among these 15 patients, we excluded one with a flail chest and one with spina bifida with chest hypoplasia who were treated for chest wall deformity only and not scoliosis. One patient died of respiratory failure at 1 year, 5 months after initial VEPTR surgery. This patient had undergone VEPTR placement at age 11 with a subsequent spinal fusion 1 year later. He died shortly after the spinal fusion from pulmonary complications. One patient had an infection postoperatively (the devices were removed and she went on to fusion); and one patient sought treatment at another

facility postoperatively and had fusion. The remaining 10 patients were followed for a minimum of 24 months (average, 39.6 months; range, 24–75 months). Four of these 10 patients had previously undergone limited spine fusion. Diagnoses included congenital scoliosis with or without fused ribs ($n = 6$), hypoplastic thorax ($n = 3$), and myelomeningocele ($n = 1$) (Table 1). The mean age at initial VEPTR surgery was 12.4 years (range, 10.2–14.1 years). In seven of 10 patients, the apex of the primary curve was in the mid- to upper thoracic spine. Preoperative Cobb angles averaged 66.5° (range, 35° – 90°).

Patients underwent placement of VEPTR(s) as previously described by Campbell and colleagues [2]. Briefly, the patient was placed in the prone position with the concave hemithorax side slightly elevated. We used a modified thoracotomy incision and after muscle dissection retracted the scapula superiorly to expose the ribs. The superior rib attachment sites were prepared (usually T2–3) and a superior cradle attached. A linear incision was made over the selected laminae (usually L1 or L2) and a downgoing lumbar hook was placed. If the ilium was used as the inferior attachment site, we made a vertical incision over the iliac crest. After subperiosteal dissection anterior and posterior to the ilium, an S hook was placed for fixation. The superior and inferior attachment sites were connected through a submuscular tunnel with the VEPTR rib sleeve and lumbar extension.

For patients with fused ribs or constricted intercostal muscles, we performed an opening wedge thoracostomy from the transverse processes of the spine to the costochondral junction. The intervals between the ribs were slowly opened using a rib spreader. Once maximal spread was achieved, a lateral rib-to-rib device was attached by placing cradles around the superior and inferior ribs and connecting them with an appropriate-sized rib sleeve. We mobilized muscular layers to provide maximal coverage of the devices. Intraoperative monitoring included somatosensory-evoked potentials and motor-evoked potentials of upper and lower motor neurons. The laterally placed rib-to-rib device placed the ipsilateral lower trunk of the brachial plexus at risk, and intrinsic hand muscle strength was tested before extubation along with assessment of lower extremity neurologic function.

Table 1. Summary of patient population

Diagnosis	Number	Average followup (months)	Average age at initial VEPTR surgery (years)	Prior treatment
Congenital scoliosis with or without fused ribs	6	38	12.6	Cervical fusion ($n = 1$); myelomeningocele with cord repair ($n = 1$)
Hypoplastic thorax	3	39	11.9	Thoracic fusion ($n = 2$); growing rod ($n = 1$)
Myelomeningocele	1	50	13.0	Thoracic fusion ($n = 1$)

As a result of the older age of this patient population, VEPTR devices were not lengthened regularly. Briefly, the lengthening procedure entails a small incision, approximately 2 cm, over the VEPTR connector. The device is then distracted between 0.5 to 1 cm and the incision closed. Most patients are discharged within 24 hours. Lengthenings were solely dependent on the growth and needs of the patient. As necessary, we replaced the central portion of the device in conjunction with a lengthening procedure. This continued until the patient was fused definitively or reached the end of growth. Lengthenings occurred an average of 4.7 times over the course of treatment. Replacement of the device resulting from growth was required in two patients.

We identified the following data: patient age at surgery, diagnosis, and length of followup. The records were reviewed for complications and whether the patient underwent a definitive spinal fusion.

We obtained routine postero-anterior/lateral plain radiographs preoperatively, postoperatively, and every 12 months thereafter. One resident (PF) measured the following radiographic parameters preoperatively, postoperatively, and throughout the course of followup: maximum Cobb angle, hemithoracic height, hemithoracic width, thoracic length, head shift, and T1 tilt angle. Head shift, or head decompensation, was defined as the distance from the center of C7 to the central sacral line [6]. The T1 tilt angle was the angle measured between a line drawn at the superior end plate of T1 and a line drawn perpendicular to the central sacral line [1]. As a result of poor radiographic visibility of all measurements, data were unavailable in two patients.

Results

Immediate postoperative Cobb angles averaged 48° (range, 17°–85°); the immediate Cobb angle correction averaged 26%. Excluding those patients who had been fused before VEPTR implantation, Cobb angle correction averaged 36%, ranging from 9% to 51%. At 2-year followup, Cobb angle correction averaged 18%. Excluding those that were fused prior, correction at 2 years averaged 32%. The

average absolute T1 tilt angle measured 17° preoperatively. Immediate postoperative absolute values measured 14° demonstrating moderate improvement. Head shift showed average correction of 3.8 cm, ranging from –0.2 to 8.9 cm pre- to postoperatively. Head shift was maintained or improved in six of eight patients (75%) (Table 2).

Measured pre- to postoperatively, thoracic spine height increased on average 1.5 cm (range, –0.2 to 3.0 cm). Excluding those who were fused prior, thoracic spine height increased an average of 2.2 cm. A positive change in thoracic spine height was seen in seven of eight patients (87.5%). Hemithoracic height and width both increased postoperatively by approximately 4.5%. Hemithoracic height improved in six of seven patients (85.7%). Hemithoracic width improved in five of eight patients (62.5%).

Six of the 10 patients experienced adverse events. Device-related adverse events included one patient with device fracture and migration (separate episodes) and one patient who experienced pain in the upper lumbar spine, which subsided without intervention. Other adverse events reported include two patients with pneumonia, one patient with respiratory issues resulting from a pneumothorax, and one patient with respiratory infection. No neurologic complications were seen in any patients.

Case Report

A 13-year-old girl with congenital scoliosis and multiple rib fusions presented with a convex left curve from T1 to T8 measuring 86° (Fig. 1A–F). On physical examination, she demonstrated moderate truncal decompensation to the left with a marked head tilt to the right. Her right shoulder was approximately 1.5 cm lower than her left. Surgical options discussed included VCR, posterior spine fusion, or VEPTR placement. It was decided to undertake the latter because it would allow for some continued growth and may obviate the need for definitive fusion. She underwent right rib-to-spine (T3/4 to L1) and rib-to-rib (T3/4 to T9/10) placement with multiple opening wedge thoracostomies.

Postoperatively, her thoracic curve measured 58°. She demonstrated improved head tilt, although her right shoulder is currently 2 cm higher than her left. She has

Table 2. Summary of results

Diagnosis	Number	Pre-/postoperative Cobb correction	2-year Cobb correction	Pre-/postoperative head shift (cm)	Definitive procedure
Congenital scoliosis with or without fused ribs	6	36.4%	32.1%	4.3	Posterior spinal fusion (n = 2); planned for fusion (n = 2); no fusion planned (n = 2)
Hypoplastic thorax	3	15.9%	–4.7%	1.8	Posterior spinal fusion (n = 1); planned for fusion (n = 2)
Myelomeningocele	1	12.7%	–16.4%	7.9	Posterior spinal fusion (n = 1)

Fig. 1A–F (A, B) A 13-year-old girl presented with congenital scoliosis and fused ribs with a convex left high thoracic curvature measuring 86° . She underwent rib-to-spine and rib-to-rib VEPTR placement with multiple opening wedge thoracostomies. (C) Three-month postoperative anteroposterior and (D) lateral radiographs demonstrate curve correction to 58° . (E, F) Six years and two routine expansions later, she continues to do well with her curve maintained at 55° .

undergone two routine VEPTR expansions. She was last seen in the clinic 4 years after her last lengthening procedure and 6 years after her initial implant. Her curvature remained stable at 55° . She has not experienced any upper back pain and has not demonstrated any other complications. She is currently 20 years old and enjoys an active lifestyle, which includes running track. She is contemplating whether to proceed with definitive spinal fusion or leave the VEPTR devices in place.

Discussion

The vertical expandable prosthetic titanium rib (VEPTR) was developed to treat chest and spine deformities in young children with considerable growth potential. However, older children with complex spinal deformities may also benefit from placement of a VEPTR when vertebral column resections are considered too risky neurologically. We therefore examined a small group of 10 patients older than 10 years of age with complex spine deformity treated with

VEPTR to determine the degree of correction and growth and to identify the complications.

Our study is limited by the small sample size and missing data in two patients owing to poor radiographic quality. Based on our preliminary results, however, our sample size is likely to increase because the indications for surgery may expand. Nonetheless, it provides preliminary data to support the use of VEPTR in carefully selected older children with complex spine deformity in whom VCR is deemed too risky.

Alternate treatment options for these patients included a posterior spine fusion with or without a VCR. A posterior spine fusion without a VCR would halt any further growth of the spine with the possibility of not attaining a satisfactory outcome. A VCR would likely improve the outcome but with substantial neurologic risk. A combined anteroposterior VCR lowers neurologic risk; however, these patients have markedly diminished pulmonary function, which precludes such an approach. Suk and colleagues [8] reported 16 patients treated with a posterior VCR and wrote “it is a highly technical procedure and

should only be performed by an experienced surgical team.” In that series, four of the 16 patients sustained a complication, including one hematoma, one hemopneumothorax, one proximal junctional kyphosis, and one complete paralysis. The beneficial effects of VEPTR in young children with congenital scoliosis and fused ribs are well documented [3–5]. In these patients, VEPTR improves the Cobb angle, increases the space available for the lung, and allows for continued growth of the spine. In addition, Campbell and colleagues [1] studied 14 patients with congenital scoliosis with fused ribs and cervical tilt. They demonstrated improved cervical tilt, head shift, and truncal decompensation with VEPTR in these patients. The mean age of their patients at the time of surgery was 4.4 years.

The surgical goals for these older patients were different than those for younger patients. These included curve control with improved cosmesis without the risks of vertebral column resection. In our series, no patients sustained neurologic injury. The other complications were similar to those previously reported for VEPTR [3–5]. Curve progression was halted with some modest correction. In addition, T1 tilt and head shift improved. Together, these radiographic improvements resulted in improved cosmesis for the patient. The numbers were too small to determine whether the number of thoracostomies correlated with cosmesis. Although four of our patients have progressed to final fusion, four patients (including our case illustration) have opted to delay definitive fusion. For these patients, the VEPTR stabilized their complex spinal deformity until they were older, likely decreasing the risks of spinal fusion. Final fusion may not be mandatory in these patients as previously thought. A recent study by Flynn and colleagues (“Are spinal fusion and implant removal needed after the completion of VEPTR treatment?”; Scoliosis Research Society annual meeting; September 8–14, 2008) shows VEPTR end point management varies by diagnosis. Although only a small percentage of children with hypoplastic chest syndromes were fused after maturity, the majority of those with congenital or progressive scoliosis have had definitive fusion at the end of treatment.

The VEPTR results in modest curve improvement, continued growth of thoracic spine height, stabilization of

hemithoracic height and width, and improved head shift. Although complications occurred in six patients, none were neurologic events. Longer followup of these patients may clarify whether, in select patients, the VEPTR may be left in.

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