Bilateral Rib-to-pelvis Technique for Managing Early-onset Scoliosis

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Abstract

Background Early-onset scoliosis describes progressive spinal deformity of varying etiologies in the growing child. The management of early-onset scoliosis is challenging, with many treatment options but no conclusive evidence for the best treatment method.

Questions/purposes We describe a bilateral percutaneous rib-to-pelvis technique, present our early experience with this technique in patients with early-onset scoliosis, identify adverse events, and determine whether these are comparable to those for other current techniques.

Description of Technique The VEPTR[®] device is placed through three small incisions that allow for attachment of rib hooks bilaterally at the upper end and through pelvic hooks at the distal end, providing distraction forces to correct the deformity while allowing for growth.

Patients and Methods We retrospectively reviewed all 37 patients with early-onset scoliosis treated with the bilateral rib-to-pelvis VEPTR[®] technique from 2003 and 2009. Patients were evaluated for demographics, diagnosis, curve correction, and adverse events and divided into two groups: ambulatory and nonambulatory. The 18 ambulatory

patients underwent 139 procedures and the 19 nonambulatory patients underwent 100 procedures. Average followups were 84 and 64 months in the ambulatory and nonambulatory groups, respectively.

Results The rate of adverse events per procedure was 13%. Thirty-nine percent of ambulatory patients developed a marked crouched gait over time. The rate of adverse events in the nonambulatory group was 15%.

Conclusions This technique appears a reasonable alternative to growing rods for the management of early-onset scoliosis in nonambulatory children due to the low rate of adverse events. Due to the increased incidence of crouched gait, we have abandoned this technique in ambulatory children unless there is no option to attach the distal fixation to the spine.

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

Introduction

Early-onset scoliosis (EOS) is a term used to describe progressive spinal deformity in a skeletally immature child, resulting from a broadening variety of etiologies, encompassing both idiopathic and neuromuscular curves, and includes deformity in the coronal and sagittal planes. The unifying factor in EOS curves is that, regardless of the underlying etiology, a child with substantial growth remaining has a concomitant risk of progression during growth.

The management of EOS in a child is challenging, with a myriad of treatment options and strategies but no conclusive evidence for the optimal treatment method. Treatment options include observation, bracing, casting

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Each author certifies that his or her institution approved the human protocol for this investigation, that all investigations were conducted in conformity with ethical principles of research, and that informed consent for participation in the study was obtained.

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[10, 16], traction [10], early fusion [5], and growth-preserving surgery. Surgical methods include distraction-based techniques, such as growing rods [1, 2, 14, 18], Vertical Expandable Prosthetic Titanium Rib (VEPTR®) (Synthes Inc, West Chester, PA) [6–9, 11, 15] guided growth, or growth tethers (staples) [3, 4]. Of these techniques, only the VEPTR® is approved by the US Food and Drug Administration (FDA) as a humanitarian use device under specific indications [19]. VEPTR® is approved for the treatment of thoracic insufficiency syndrome (TIS) in patients with flail chest, constrictive chest wall syndromes, including scoliosis with rib fusion, hypoplastic thorax syndromes, and progressive scoliosis of congenital or neurogenic origin without rib anomalies.

The rationale for growth preservation for children with progressive spinal deformities and substantial growth remaining is allowing continued growth in the height of the trunk while controlling progression of the spinal curve and managing TIS. TIS occurs when the thorax cannot support normal respiration and lung growth [9]. While early fusion can control curve progression during growth, the patient will often have a marked shortening of the trunk and reduced thoracic volume as an adult, producing restrictive lung disease [13, 17, 20]. There is an increasing body of evidence documenting the use of growing systems to manage EOS [1–4], but no technique appears superior to the others. However, all techniques have similar goals: preservation of spinal growth, control of the spinal deformity, allowance of pulmonary development, and minimal complications and morbidity at skeletal maturity.

From its inception, the VEPTR® device was designed as a technique to manage TIS by providing instrumentation that could simultaneously treat spine and chest wall deformities [8]. This could be achieved through the use of an expansion thoracoplasty, stabilized by an expandable rib-to-rib implant to increase the volume of the constricted hemithorax, and a rib-to-spine implant to manage the spinal curve. It is the first instrumentation system that clearly addressed the volume restriction deformities associated with these complex curves of multiple etiologies. The safety of this device was confirmed by the FDA [19].

In 2003, we began to expand the utility of the VEPTR® device by developing a new Salt Lake City percutaneous technique as an alternative to growing rods. This technique utilizes bilateral VEPTR® devices that are attached to the ribs proximally and to the pelvis distally using small incisions. The technique does not include an expansion thoracoplasty as part of the procedure.

In this article, we (1) describe our bilateral percutaneous rib-to-pelvis technique, (2) present our early experience using this technique in patients with EOS, and (3) analyze our adverse events and determine whether these are comparable to other current techniques.

Description of Technique

The patient was placed on rolls on the operating table in the prone position and carefully padded (Fig. 1). Spinal cord monitoring was used for both the upper and lower extremities. A midline incision was made from T1 to T5. Dissection was made in the midline to the tip of the spinous processes. At this level, dissection continued 5 cm laterally over the trapezius bilaterally. A small incision in the muscle was made just lateral to the spinous process, and a hemostat was used to identify the layer between the trapezius and rhomboid muscles and the erector spinae muscles. They were then divided using electrocautery in a manner allowing for later repair. Dissection was made laterally over the surface of the erector spinae muscles to their origin on the ribs. The erector spinae were then elevated from lateral to medial to the tip of the transverse process. Care was taken to preserve the periosteum of the ribs, which contributed to the blood supply of the ribs. Generally, the second through fourth rib was selected for placement of the VEPTR® rib cradles (Fig. 2). The level was confirmed using the image intensifier. Rib cradles were placed on the second and fourth ribs using a stacked rib cradle construct available with the VEPTR® 2 device (Fig. 3).

For the pelvic fixation, an oblique incision was made, beginning 4 cm proximal to the apex of the iliac crest (about 1–2 cm lateral to the posterosuperior iliac spine) and continuing along the lateral border of the paraspinal musculature. The dissection followed the lateral border of the erector spinae muscle to the top of the iliac crest apophysis. To create a space for the pelvic hook, a small incision was made by elevating the gluteus medius origin

Fig. 1 A photograph shows a patient prone on the operating table and carefully padded. A midline incision at the level of the second to fourth ribs is made for proximal rib fixation. Distal incisions 1 cm lateral to the posterosuperior iliac spine are used for pelvic fixation.

Fig. 2 A photograph shows how the second through fourth ribs are generally used for proximal rib fixation. A stacked rib cradle construct using the VEPTR[®] 2 device is preferred.

Fig. 3 A photograph illustrates how the stacked rib cradles allow for multiple points of fixation on the ribs.

off the posterior ileum. A small transverse incision was made anterior to the ileum for placement of the pelvic hook. The pelvic hook was then placed over the apex of the ileum, with the prong of the hook placed ventral over the iliac wing (Fig. 4). A long pituitary rongeur was used to make a tunnel from the proximal incision to the apex of the ileum, attempting to stay close to the posterior chest wall and beneath the paraspinous musculature. A Number 20 chest tube was then pulled from the distal incision to the proximal incision. An appropriately sized VEPTR[®] implant was selected and contoured to ensure maintenance of lumbar lordosis (Fig. 5). When sizing the implant, one must anticipate the resulting length required for the device at the completion of the initial distraction. The implant was tunneled from proximal to distal using the chest tube and then engaged using a 5.0/6.0 connector to the pelvic hook. The upper end of the rod was engaged in the proximal rib cradles and locked. A temporary c-ring was used for

Fig. 4 A photograph shows how iliac fixation is placed over the apex of the ilium, which is generally about 1 cm lateral to the posterosuperior iliac spine.

Fig. 5 A photograph shows an appropriately sized VEPTR[®] implant being selected and contoured. The surgeon should anticipate the extra length needed after insertion of the device for initial correction of the curve as demonstrated in Figure 6.

distraction of the implant until the desired correction was obtained (Fig. 6). The c-ring was then removed, and the wounds were irrigated in a standard fashion. The anesthesiologist performed a Valsalva maneuver to confirm there was no pleural violation or air leak after placement of the rib cradles. If an air leak was encountered, a small chest tube was placed to prevent a tension pneumothorax. A final confirmation of implant position and correction was made using the image intensifier. A multilayered closure was required to ensure generous muscle and soft tissue coverage of the implant. Simple dressings were applied (Fig. 7).

Patients were allowed to mobilize as tolerated. Bracing or other support was not required. Implants were typically expanded through a midline incision bilaterally at 6-month intervals. This technique required the dissection at the level of the deep fascia to go laterally to the level of the

Fig. 6A–B (A) A photograph shows the distal end of the VEPTR® device being engaged in the connector for the iliac fixation, after which a c-ring is attached and used for distraction of the implant against the pelvic fixation. (B) A photograph demonstrates the distraction technique using the 5.0/6.0 connector and the c-ring.

Fig. 7 A photograph shows the simple dressing used. No bracing is used postoperatively.

distraction lock for expansion and replacement of a new lock. This preserved a midline incision for later instrumentation and fusion at skeletal maturity. Exchange

Table 1. Summary data for ambulatory patients (n = 18) with the bilateral rib-to-pelvis technique

Variable	Preoperative	Postoperative	Improvement
Primary Cobb angle (°)	63 (0–90)	44 (0–77)	17 (–9–85)
Secondary Cobb angle (°)	15 (0–65)	7 (0–54)	8 (0–65)
Kyphosis (°)	61 (24–160)	54 (21–109)	7 (–44–102)

Values are expressed as averages, with ranges in parentheses.

procedures were required once the implant was fully expanded.

Patients and Methods

This study is an IRB-approved retrospective review of all 37 patients who underwent the bilateral percutaneous rib-to-pelvis procedure at our institution between 2003 and 2009. There were 18 boys and 19 girls with a mean age of 6.1 years (range, 1.4–11.8 years). Patients were divided into two functional groups: 18 patients were ambulatory and 19 were nonambulatory. The primary diagnosis varied within both groups, with the majority of the nonambulatory group having neuromuscular disease. These groups were sufficiently different that they warranted separate analysis.

Among the 18 ambulatory patients were nine boys and nine girls (Table 1). The primary diagnoses for this group were EOS (14), neurofibromatosis Type I (two), neuromuscular kyphosis (one), and kyphoscoliosis (one). All 18 patients were capable of independent ambulation without assistive devices. The average age at the time of initial implantation was 6 years (range, 1.5–11.7 years). The average preoperative coronal Cobb angle was 63° (range, 0°–125°) and the average sagittal Cobb angle was 61.1° (range, 24°–160°). The average coronal Cobb angle at the most recent followup was 44.3° and the sagittal Cobb angle was 54.1°. All patients underwent placement of bilateral rib-to-pelvis instrumentation using the percutaneous rib-to-pelvis technique without concomitant thoracoplasty. Minimum followup for this group was 8 months (average, 84 months; range, 8–153 months). There were 139 total procedures for the ambulatory patients, including initial implantation (18), expansions (69), implant exchanges (20), and revisions (32). One patient with neonatal Marfan's syndrome died after open heart surgery unrelated to the VEPTR® device. No patients were lost to followup.

Among the 19 nonambulatory patients were nine boys and 10 girls (Table 2). The average age at the time of initial implantation was 6.1 years (range, 1.4–10.9 years). The primary diagnoses for this group were spina bifida (six),

Table 2. Summary data for nonambulatory patients (n = 19) with the bilateral rib-to-pelvis technique

Variable	Preoperative	Postoperative	Improvement
Primary Cobb angle (°)	58 (0–125)	38 (0–88)	19 (–40–52)
Secondary Cobb angle (°)	5 (0–98)	9 (0–79)	–4 (–79–98)
Kyphosis (°)	71 (22–154)	47 (20–80)	24 (–26–74)

Values are expressed as averages, with ranges in parentheses.

Fig. 8A–D (A) A radiograph shows the spine of a 15-month-old child with Williams syndrome and kyphoscoliosis. (B) A lateral radiograph of the spine shows substantial kyphosis in the sitting position. (C) A postoperative AP radiograph of the spine shows correction of the coronal deformity with the VEPTR[®] 2 devices in place. (D) A lateral radiograph shows correction of the deformity in the sagittal plane.

myopathy (six), cerebral palsy (four), and syndromic scoliosis associated with Williams syndrome (one) (Fig. 8A–B). The primary deformities for this group were scoliosis (12), kyphosis (two), kyphoscoliosis (three), gibbus deformity (one), and lordosis (one). All 19 patients required wheelchair assistance. The average age at the time

of initial implantation was 6 years. The average preoperative coronal Cobb angle was 64° (range, 0°–125°) and the average sagittal Cobb angle was 70° (range, 24°–160°). The average coronal Cobb angle at the most recent followup was 38.3° and the sagittal Cobb angle was 47°. Patients underwent placement of bilateral rib-to-pelvis instrumentation using the percutaneous technique without concomitant thoracoplasty. The minimum followup for this group was 8 months (average, 64 months; range, 8–153 months). There were 100 total procedures for the nonambulatory group, including initial implantation (18), expansions (63), implant exchanges (8), and revisions (11). Two of the 19 patients were lost to followup at outside institutions, and one patient died of unrelated causes.

All patients were followed at 6-month intervals with physical examinations and posterior/anterior and lateral radiographs of the spine and chest. Patients were assessed clinically for skin integrity, neurologic status, and adverse events. The author reviewed all radiographs for evidence of implant migration and to make curve measurements. The curve measurements included Cobb angles in the coronal and lateral planes before initial instrumentation and at all followup visits to evaluate maintenance or improvement of correction. The variability in radiograph techniques and the lack of a radiographic marker on all patients did not allow for accurate assessment of growth. These clinical and radiographic assessments were required before a planned expansion of the existing implants. Demographic data, including age at surgery, gender, diagnosis, comorbidities, curve type, curve magnitude, curve correction, number of surgeries, major and minor adverse events, and end points, if known, were recorded. Adverse events were divided into two categories: major adverse events requiring unplanned surgeries and minor adverse events not requiring unplanned surgeries corrected at the time of scheduled device expansion.

Results

Among the ambulatory patients, we identified 18 adverse events in 139 procedures (13%). Four patients had no adverse events at latest followup. Major adverse events included infection (four) and problems with persistent pain requiring VEPTR[®] removal and instrumented fusion before skeletal maturity (one). Minor adverse events included hook migration and/or failure of instrumentation revised at the time of scheduled expansion (13). Seven of 18 ambulatory patients developed substantial crouched gait and required conversion of their rib-to-pelvic fixation to rib-to-spine fixation (39%). All patients who were converted to rib-to-spine fixation showed marked improvement in their crouched gait and sagittal balance.

Three ambulatory patients had their definitive spinal fusion and are now considered “VEPTR® graduates.” The average time for VEPTR® treatment was 43 months (range, 30–60 months). There was no evidence of spontaneous spinal fusion, as is commonly seen after treatment with growing rod constructs [12].

Among the nonambulatory patients, we identified 15 adverse events in 100 procedures (15%). Nine patients had no adverse events at latest followup. In nine patients, the adverse events were hook migration and/or failure of instrumentation (six), infection (seven), and persistent pain (two). Three nonambulatory patients had definitive spinal fusion at an average of 47 months after initial treatment with the VEPTR® device (range, 40–61 months). None of the three patients had evidence of spontaneous fusion of the spine, but we noted substantial stiffness of curves (Fig. 8C–D).

Discussion

The treatment of EOS is challenging, and while there are a number of treatment options, there is no conclusive evidence as to which is the best treatment and for which children. This study (1) describes our bilateral percutaneous rib-to-pelvis technique, (2) presents our early experience using this technique in patients with EOS, and (3) analyzes adverse events and determines whether these are comparable to other current techniques.

There are limitations to our clinical study. First, the diverse patient population and disease groups make direct comparisons within these groups difficult. Second, there are no current, validated methods to determine objective outcome scores for a pediatric population with EOS.

The bilateral percutaneous rib-to-pelvis VEPTR® technique represents one new approach to the management of EOS as an alternative to growing rods [1, 2, 14, 18] or other techniques [3, 4]. This procedure is simple, minimally invasive, and does not exclude alternative treatment if there is failure of the technique. The incidence of adverse events associated with this technique compares favorably to other growing techniques [1, 2, 14, 18]. As with all new techniques, there will be an element of a learning curve and modifications that will help delineate indications and improve outcomes. For example, in evaluating early outcomes in ambulatory children, we noted 39% of children developed a substantial crouched gait that was unsightly and began to interfere with function. Most of these children were converted to a rib-to-spine construct and their gait abnormalities were resolved. Based on these observations, we no longer use the rib-to-pelvis technique in patients who are ambulatory. We are optimistic regarding the lack of spontaneous fusion of the spine at the definitive fusion,

but a greater number of patients will be needed to confirm this observation.

This technique is effective in nonambulatory children with early-onset neuromuscular scoliosis. These patients are very difficult to manage with bracing, wheelchair modification, and supports. Early fusion is problematic because many of these children having pre-existing respiratory disease. However, complications remain problematic, as with any population of children with neuromuscular disease.

Management of EOS in children continues to be a major challenge for patient and parents and the treating surgeon. Globally, there is a substantial effort to try to improve current techniques and develop new ones to improve treatment and outcomes. Goals of current treatment include managing progressive spinal deformity while allowing for normal growth of the spine, chest, and lungs. There are a variety of treatment options with varying indications and outcomes. Casting and bracing are noninvasive options in small, flexible, early-onset curves. Nonfusion treatment techniques currently available include experimental vertebral body stapling, off-label use of growing rods in a variety of configurations, the Shila procedure, and VEPTR®. There is a general consensus that all of these procedures are preferred when compared to early fusion but remain suboptimal due to the requirement of repeated procedures to obtain growth and a considerable number of adverse events. Self-expanding technology is on the horizon but has not yet been realized.

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