Journal of Pediatric Surgery (2011) **46**, 77–80

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Vertical expandable prosthetic titanium rib device insertion: does it improve pulmonary function?

Samir K. Gadepalli*, Ronald B. Hirschl, Wan C. Tsai, Michelle S. Caird, Kelly L. Vanderhave, Peter J. Strouse, Robert A. Drongowski, Frances A. Farley

C.S. Mott Children's Hospital, University of Michigan, Ann Arbor, MI 48105, USA

Received 15 September 2010; accepted 30 September 2010

Key words:
VEPTR;
Thoracic insufficiency syndrome;
Scoliosis Research Society;
Pulmonary function tests;
Congenital scoliosis;
Infantile scoliosis;
Early-onset scoliosis;
Jeune syndrome;
Neuromuscular scoliosis;
Vertical expandable prosthetic titanium rib;
Lung volume;
Lung growth;
Children;
Pediatric;
Forced vital capacity (FVC);
Forced expiratory volume (FEV1);
Residual volume;
Lung reconstruction;
Spinal deformity

Abstract

Purpose: Vertical expandable prosthetic titanium rib (VEPTR) insertion and expansion has been advocated to increase thoracic volume and pulmonary function in patients with thoracic insufficiency syndrome. We reviewed our experience with VEPTR implantation to determine if lung function and growth is augmented, to determine the children's functional status, and if the scoliosis is controlled.

Methods: From 2006 to 2010, 29 insertions and 57 expansions were performed in 26 patients at our institution. Demographic data were reviewed in conjunction with complications, scoliosis angles, pulmonary function tests (PFTs), and computed tomography–guided 3D reconstructions to determine lung volumes; and quality of life scores were determined using a modified Scoliosis Research Society (SRS) questionnaire preoperatively and postoperatively. The groups were also stratified by age (because of lung growth potential), disease (congenital or infantile scoliosis, Jeune syndrome, neuromuscular, other structural thoracic disorders), and sex. Analyses using SPSS (SPSS, Chicago, Ill) were performed with $P < .05$ considered significant.

Results: Each patient underwent 3.03 ± 1.8 surgeries, spending 0.97 ± 1.8 days in the intensive care unit and 4.41 ± 6 days in the hospital for each procedure. Mean age was 90.7 ± 41 months. Of the 36 complications, most were because of infection (12), half requiring operative repair (hardware removal). The average PFT percent predicted values for forced expiratory volume in 1 second, forced vital capacity, and RV were 54.6 ± 22, 58.1 ± 24, and 145.3 ± 112, respectively, preoperatively and 51.8 ± 20, 55.9 ± 20, and 105.6 ± 31, respectively, postoperatively. The lung volumes measured by computed tomography when corrected for age do not increase significantly postoperatively. The mean Cobb measurement for the preoperative major curves was 64.7° and postoperatively was 46.1° for those curves measured preoperatively, for a 29% curve improvement. All postoperative curves had a mean of 56.4° and 58.1° at final follow-up, a 3% curve increase. The SRS scores for patients remained unchanged and no statistical difference was seen from preoperative to postoperative values. No statistically significant difference was seen in complications, PFT (forced expiratory volume in 1 second, forced vital capacity, RV), lung volumes, scoliosis angles, and SRS scores between sex, age, and disease categories.

* Corresponding author. Department of Pediatric Surgery, C.S. Mott Children's Hospital, University of Michigan, Ann Arbor, MI 48105, USA. Tel.: +1 734 764 4151; fax: +1 734 936 9784.

E-mail address: samirg@med.umich.edu (S.K. Gadepalli).

0022-3468/$ – see front matter
doi:10.1016/j.jpedsurg.2010.09.070

Conclusion: There was mild improvement in scoliosis angles but no improvement in lung function and volume. Scoliosis Research Society scores indicate that the children have near normal function both before and after VEPTR placement. Pulmonary function, lung volume, and patient subjective assessments did not increase dramatically after VEPTR placement, although scoliosis angles improved.

In patients with congenital scoliosis, an alternative to early spinal fusion to prevent a worsening curve is expansion thoracoplasty using vertical expandable prosthetic titanium rib (VEPTR; Synthes Spine Co, West Chester, Pa) insertion and serial lengthening, first described in 1989 [1]. Other indications for VEPTR insertion have later been introduced including thoracic insufficiency syndrome [2,3], where abnormal thoracic structure affects lung development, limiting a patient's functional capacity. Although designed to increase the thoracic volume and lung growth, VEPTR insertion has failed to show improvement using pulmonary function tests (PFTs) in short-term follow-up [2,4-6].

As useful adjuncts to PFTs, 3-dimensional reconstructions of thoracic CT scans (3DCTR) can be used to evaluate lung volumes after thoracic spinal reconstruction procedures [7]. Using a combination of 3DCTR and PFT values, preoperative and postoperative changes can be carefully monitored to determine the effect of VEPTR insertion on pulmonary function. Improvements in ventilation after VEPTR insertion, especially in younger patients, have been described using VQ scans [8], although studies using 3DCTR are limited.

Quality of life variables however do not always reflect the changes seen on these objective measures; therefore, a functional survey was created and refined by the Scoliosis Research Society (SRS) [9,10]. The SRS questionnaire measures a patient's subjective self-assessment in the following areas: function, pain, self-image, mental health, and satisfaction. In this report, we present outcomes after VEPTR insertion and lengthening in our experience, from a prospectively collected database, including complications, changes in scoliosis angles, PFT, volumes from 3DCTR, and results of a modified SRS questionnaire.

2. Methods

A prospectively collected database was established to follow patients with placement of VEPTR device after institutional review board approval. Vertical expandable prosthetic titanium rib insertion was performed by a pediatric surgeon and orthopedic surgeon acting in conjunction using a thoracotomy for placement of the device. Subsequent lengthening procedures were performed serially at an average of 6 months from the previous procedure.

A total of 26 patients (16 male and 10 female) underwent 86 procedures between October 2006 and March 2010 at the University of Michigan in Ann Arbor, Mich. The patients, with an average age of 91 months, underwent 29 insertions (21 bilateral) and 57 expansions. The patients were also evenly grouped above and below 84 months or 7 years of age. The patients were categorized into 4 diagnoses (congenital or infantile scoliosis, Jeune syndrome, neuromuscular scoliosis, and unspecified structural thoracic disorder). Almost half of these patients had congenital or infantile scoliosis (12), whereas the other half was mostly split between neuromuscular scoliosis (5) and unspecified structural thoracic disorder (7). Only 2 patients with Jeune syndrome comprised our VEPTR experience.

Outcome measures included PFT preoperatively and every 6 months in all patients (12) who were not ventilator-dependent and can cooperate with the procedure, lung volumes in patients who consented to 3DCTR preoperatively and yearly (10), a modified SRS-22 questionnaire, and scoliosis angles measured preoperatively and at each postoperative visit in all patients, with 23 of 26 patients having adequate follow-up for analysis of these values. To measure scoliosis angles, standing PA and lateral spine radiographs were obtained. The Cobb measurement was obtained from the largest curve preoperatively, postoperatively, and at final follow-up. The mean Cobb angle measurement was calculated and compared at the different periods. In addition, the lengths of stay, complication rates including return to operating room, PFT results, and lung volumes by 3DCTR were compared among sex, age, and diagnoses groups.

SPSS software (SPSS, Chicago, Ill) was used to analyze all data; and Student t test, analysis of variance, and χ^2 analyses were performed with $P < .05$ considered significant.

3. Results

The 29 VEPTR insertions averaged 6.72 days in the hospital postoperatively, including 2.72 days in the ICU, whereas the 57 lengthening procedures averaged 2.44 days in the hospital, including 0.26 days in the ICU. In all, 36 complications occurred with 22 of 36 treated nonoperatively: 3 transfusions for bleeding, 6 infections treated with antibiotics, 9 neurologic (pain or numbness), 2 hardware dislodgements, and 2 pleural effusions. Reoperation was required in 4 for chest tube placement (pneumothorax), 1 for seroma drainage, 6 for hardware removal (for infection), and 3 for hardware repositioning (for dislodgement).

No statistical difference was noted between preoperative and postoperative values of the pulmonary outcomes for PFT

Table 1 Pulmonary outcomes

		Preoperative	Postoperative	*P*
PFT	FEV_1	54.6 ± 22	51.8 ± 20	.63
	FVC	58.1 ± 24	55.9 ± 20	.62
	RV	145.3 ± 112	105.6 ± 31	.34
3DCTR	Total volume	944.2 ± 450	1042.1 ± 311	.19

FEV_1 indicates forced expiratory volume in 1 second; FVC, forced vital capacity.

and 3DCTR (see Table 1). When the CT volumes were plotted on a previously described scoliosis lung volume table [7], no increase in the age-corrected volume was seen.

The mean Cobb measurement preoperatively for major curves was 64.7° and decreased by 29%. Postoperatively, all curves had a mean of 56° and at final follow-up was 58.1°, which was only a 3% increase.

Functional outcomes by a modified SRS-22 survey show near-normal values in all categories both preoperatively and postoperatively (see Table 2).

4. Discussion

Vertical expandable prosthetic titanium rib implantation and expansion has been used to treat thoracic insufficiency syndrome in young children with varied results using PFTs and CT lung volumes [2-6]. In our database, we found no difference between preoperative and postoperative values, highlighting the need for further studies to support the use of VEPTR for improving lung function. One argument for the lack of pulmonary function improvement might be the increased age of our patient pool. Because the potential for lung growth is greater [7,11] and a larger improvement in lung function after VEPTR insertion was noted in younger patients [6,8], we used 84 months as a criteria to separate our groups in equal proportions. No difference was noted however even in the younger children using PFT values, 3DCTR lung volumes, and functional scores. Our data confirmed improvement in Cobb angle measurements in the older children as demonstrated previously [12], although no difference was found in the pulmonary and functional outcomes.

Measured spine angles showed modest improvement over time and postoperatively after insertion, that the device can be used to maintain the scoliosis during growth. When subjective assessment using a modified SRS-22 survey was used, the children had nearly normal function both before and after the VEPTR insertion. Of the variables tested, self-image improved the most, although not statistically significant.

Pulmonary function tests and CT lung volumes may not fully reflect the changes after VEPTR insertion. Hemoglobin levels [13] and weight gain [14] after VEPTR can be used to demonstrate improved lung function. In our population, postoperative hemoglobin levels were not routinely performed in the outpatient setting and weights were not clearly documented in both the preoperative and postoperative setting to be used as markers.

Finally, quality of life is significantly affected by thoracic insufficiency syndrome for the patient and family [15]. In studying the effect of VEPTR insertion, we used the modified SRS-22 survey [9,10] preoperatively and postoperatively at each visit. The overall results were positive, although no statistically significant difference was seen between preoperative and postoperative values. Of the variables tested, self-image seemed to improve the most, although not statistically significant. In fact, many parents noted on follow-up visits that their children's interest in sports and other activities increased after the VEPTR insertion, possibly secondary to their improved self-confidence.

Table 2 Functional survey results

	Preoperative	Postoperative	*P*
Function	4.01 ± 0.66	3.93 ± 0.60	.48
Pain	3.99 ± 1.04	3.82 ± 0.89	.45
Self-image	4.00 ± 0.44	4.16 ± 0.47	.17
Mental health	4.39 ± 0.46	4.38 ± 0.45	.78
Satisfaction	4.08 ± 0.69	4.03 ± 0.76	.78

5. Conclusion

A modest improvement was noted using mean Cobb measurements; however, there was no improvement in lung function, volume, or SRS scores between the pre- and postoperative period. The SRS-22 survey was stable after VEPTR placement. Our data suggest that VEPTR placement does not enhance pulmonary status by these measures.

References

[1] Hell A, Campbell R, Hefti F. The vertical expandable prosthetic titanium rib implant for the treatment of thoracic insufficiency syndrome associated with congenital and neuromuscular scoliosis in young children. J Pediatr Orthop B 2005;14:287-93.
[2] Mayer O, Redding G. Early changes in pulmonary function after vertical expandable prosthetic titanium rib insertion in children with thoracic insufficiency syndrome. J Pediatr Orthop 2009;29:35-8.
[3] Hasler C, Mehrkens A, Hefti F. Efficacy and safety of VEPTR instrumentation for progressive spine deformities in young children without rib fusions. Eur Spine J 2010;19:400-8.
[4] Motoyama E, Deeney V, Fine G, et al. Effects on lung function of multiple expansion thoracoplasty in children with thoracic insufficiency syndrome: a longitudinal study. Spine (Phila Pa 1976) 2006;31:284-90.
[5] Emans J, Caubet J, Ordonez C, et al. The treatment of spine and chest wall deformities with fused ribs by expansion thoracostomy and insertion of vertical expandable prosthetic titanium rib: growth of thoracic spine and improvement of lung volumes. Spine (Phila Pa 1976) 2005;30:S58-68.
[6] Motoyama E, Yang C, Deeney V. Thoracic malformation with early-onset scoliosis: effect of serial VEPTR expansion thoracoplasty on lung growth and function in children. Paediatr Respir Rev 2009;10:12-7.

[7] Gollogly S, Smith J, White S, et al. The volume of lung parenchyma as a function of age: a review of 1050 normal CT scans of the chest with three-dimensional volumetric reconstruction of the pulmonary system. Spine (Phila Pa 1976) 2004;29:2061-6.
[8] Waldhausen J, Redding G, Song K. Vertical expandable prosthetic titanium rib for thoracic insufficiency syndrome: a new method to treat an old problem. J Pediatr Surg 2007;42:76-80.
[9] Asher M, Min Lai S, Burton D. Further development and validation of the Scoliosis Research Society (SRS) outcomes instrument. Spine (Phila Pa 1976) 2000;25:2381-6.
[10] Asher M, Lai S, Glattes R, et al. Refinement of the SRS-22 Health-Related Quality of Life questionnaire Function domain. Spine (Phila Pa 1976) 2006;31:593-7.
[11] Dimeglio A. Growth in pediatric orthopaedics. J Pediatr Orthop 2001;21:549-55.
[12] Samdani A, St Hilaire T, Emans J, et al. The usefulness of VEPTR in the older child with complex spine and chest deformity. Clin Orthop Relat Res 2010;468:700-4.
[13] Caubet J, Emans J, Smith J, et al. Increased hemoglobin levels in patients with early onset scoliosis: prevalence and effect of a treatment with Vertical Expandable Prosthetic Titanium Rib (VEPTR). Spine (Phila Pa 1976) 2009;34:2534-6.
[14] Skaggs D, Sankar W, Albrektson J, et al. Weight gain following vertical expandable prosthetic titanium ribs surgery in children with thoracic insufficiency syndrome. Spine (Phila Pa 1976) 2009;34:2530-3.
[15] Vitale M, Matsumoto H, Roye DJ, et al. Health-related quality of life in children with thoracic insufficiency syndrome. J Pediatr Orthop 2008;28: 239-43.

Discussion

Unidentified Speaker: So, if I understand, and I want to—you guys collected a lot of data here, very thorough in the follow-up, tough group of patients, but there was really no significant difference in any of the measured outcomes. The naïve person could look at this and ask should you be doing this at all.

Dr Gadepalli: Actually we felt the same thing and one of the thoughts that we had was it is possible we missed any significant variables and we would collect these for a while longer and see if maybe this was a type B error. The other possibility is that there is not any significance but since it does improve angles we can still use these devices. We just can't say it is used for lung volume improvement. The final thing would be we may be using the wrong variables to measure improvement in lung function. There are some new data coming out in terms of just using weight gain or using hemoglobin levels as a way of monitoring lung function.

John Waldhausen, MD (Seattle, Wash)

Dr Waldhausen: If you look at the thoracic insufficiency group, specifically the Jeune's children who were on the ventilators, Greg Redding, who is in Seattle, a pulmonologist, if you look at a national consortium of data, actually shows that a lot of those kids do have improved lung function and either stabilize on the ventilator or potential get off. Did you separate your data and look at those kids specifically?

I also wondered about your complication rate because that seemed relatively high compared to other series that have been reported. I was wondering how much attention did you pay to nutritional management preoperatively so that you had adequate skin and soft tissue coverage?

Dr Gadepalli: Thank you for your questions. The first question regarding Redding and Waldhausen's data looking at the case series using VQ scans and showing improvement in younger children, essentially we split our data base into sex, age, diagnoses to see if there was any difference with any of these different modalities, and we found no difference in any of them. One problem is that the numbers become so small we only had two Jeune's patients and that is the case with Seattle as well. I contacted them to get their data. I think that unless you have a collaboration with several centers you will not have enough data for Jeune's data.

Regarding the preoperative nutritional management, all these patients received preoperative nutrition management in order to effectively have—I can't explain why we had so many complications. There is a huge learning and a lot of our complications occurred early. I think we learned lessons to help avoid these in the future.

Harsh Grewal, MD (Philadelphia, Pa)

Dr Grewal: Thanks for your paper. A couple of comments. One comment is I have been doing this for about 10 years and we have done about 150 patients. The caveat is, sort of like Dr Waldhausen mentioned, the largest group that is having these inserted nationwide are scoliosis with complex chest wall deformities, fused ribs, etc. In that group, the reason to insert the VEPTR is not to improve lung function. It is to correct the scoliosis and the chest wall deformity without fusing the spine, because these are growing kids. I think the question asked then is not really relevant to that group. The question then is relevant to the group in which you have Jeune syndrome, for example. In that group, there definitely is anecdotal experience starting from when Campbell first did this procedure that they come off the ventilator. The question is in that group that are ventilator dependent that have asphyxiating chondral dystrophies the PFTs are very inaccurate. In fact, in almost 50% of those patients the pulmonologist will say this PFT is not adequate. Having those two caveats, how would you then address the question that you asked?
Thank you.

Dr Gadepalli: Thank you for your comment. In terms of Jeune's and measuring lung function over time, I think the data are still unclear and we just have to wait over time and see what happens.