Pulmonary and Radiographic Outcomes of VEPTR (Vertical Expandable Prosthetic Titanium Rib) Treatment in Early-Onset Scoliosis

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Background: VEPTR (vertical expandable prosthetic titanium rib) expansion thoracoplasty is used to manage thoracic insufficiency syndrome in early-onset scoliosis. Literature regarding the effects of this technique on pulmonary function is scarce. The aim of this study was to report the intermediate-term results of VEPTR expansion thoracoplasty.

Methods: Twenty-one children with thoracic insufficiency syndrome underwent VEPTR expansion thoracoplasty from 2002 to 2012 and had complete chart data, preoperative and follow-up radiographs, and pulmonary function tests performed at the index implantation, first expansion, and last expansion. Pulmonary function tests with forced and passive deflation techniques developed for children under general anesthesia were performed prior to the index implantation and each expansion surgery under the same anesthetic conditions. Pulmonary and radiographic parameters were analyzed longitudinally.

Results: Mean follow-up was six years, and mean age at implantation was 4.8 years. The mean number of expansion procedures per patient was eleven, and the mean number of pulmonary function tests was ten. The mean interval between surgical procedures was 6.4 months. Mean forced vital capacity (FVC) increased from 0.65 to 0.96 L (p < 0.0001). However, the percentage of the predicted FVC decreased from 77% to 58%. Respiratory system compliance normalized on the basis of body weight, Crs/kg, decreased by 39%, from 1.4 to 0.86 mL/cm H2O/kg. The mean Cobb angle before treatment was 80°, and the mean maximum thoracic kyphosis angle was 57° (range, 7° to 107°). The initial coronal correction was maintained at the time of final follow-up (67°); however, there was a trend toward a decrease in the maximum thoracic kyphosis angle (to 66°, p = 0.08). Clinically apparent proximal thoracic kyphosis occurred in four patients, and spinal imbalance occurred in seven. The mean gain in T1-T12 height during the treatment period was 18 mm (2.9 mm/year).

Conclusions: FVC improved over time; however, this increase in lung volume did not keep up with the growth of the child, as the percentage of the predicted FVC decreased, and the chest wall stiffness increased. Coronal correction was maintained, but the increase in proximal thoracic kyphosis is concerning.

Level of Evidence: Therapeutic Level IV. See Instructions for Authors for a complete description of levels of evidence.

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insufficiency syndrome. In order to clearly understand how much of this goal could be achieved, recent studies have focused on estimating changes in lung volume by means of computed tomography (CT) scans or measuring lung volume by means of pulmonary function tests. However, the effects of the VEPTR technique on pulmonary function and chest growth are still not fully understood.

VEPTR patient selection criteria, indications, and design have been evolving as more studies and longer follow-up data become available. The aim of the present study was to investigate the intermediate-term pulmonary and radiographic outcomes of VEPTR expansion thoracoplasty in children with thoracic insufficiency syndrome.

**Materials and Methods**

This study was conducted with institutional review board approval. Since 2002, we have been measuring lung volume (especially forced vital capacity, or FVC) and total respiratory system compliance (Crs) under general anesthesia. Forty-nine children with early-onset scoliosis underwent VEPTR treatment at our institution from 2002 to 2012. The inclusion criteria for the study were (1) availability of preimplantation and consecutive postimplantation pulmonary function tests, (2) full-length spinal radiographs, and (3) a follow-up duration of more than twenty-four months. Twenty-one children with thoracic insufficiency syndrome who underwent VEPTR expansion thoracoplasty surgery, including the index implantation and consecutive lengthenings, at our institution met the inclusion criteria. Pulmonary function tests were performed under general anesthesia with muscle relaxants immediately prior to each VEPTR implantation and most of the expansion procedures. The timing of the radiographs was similar to that of the pulmonary function tests.

**Pulmonary Function Tests**

Pulmonary function testing was performed prior to the index implantation and each expansion surgery under the same anesthetic conditions. Testing was performed under general endotracheal anesthesia by means of forced and passive deflation techniques with use of a mobile unit as described previously (see Appendix). For children over six years of age, predicted FVC values were taken from Schoenberg et al. For younger children, normative values developed at the pulmonology laboratory at our institution were used. Arm span was used instead of height for calculation of body-size-adjusted predicted values because of the decreased torso height of the children.

**Radiographic Measurements**

The coronal Cobb angle, maximum thoracic kyphosis, global thoracic kyphosis (T2-T12), lumbar lordosis, spinal balance, T1-T12 height, and space available for the lung (SAL) ratio were measured on radiographs. The T1-T12 height was measured on posteroanterior whole-spine radiographs between the midpoint of the T1 upper vertebral end plate and the midpoint of the T12 lower end plate. Height comparisons were made between radiographs made after the implantation surgery and the last lengthening to avoid overestimating the spinal growth. The SAL ratio was determined by measuring the height of each hemithorax.
from the middle of the first rib to the dome of the hemidiaphragm, then dividing the height of the hemithorax on the concave side by that of the convex side. The timing of the radiography with regard to inspiration or expiration was not controlled reliably.

Hospital charts were reviewed for the number and type of surgical procedures and complications. An “unplanned surgery” was defined as any spinal surgical procedure performed at a time other than that of the planned expansion procedures. Rod or other implant adjustments and exchanges made during the planned expansions were not included as unplanned surgical procedures.

Statistical Analysis

Descriptive statistics were calculated for the radiographic and pulmonary variables. Matched-pair t tests were performed to determine whether measurements made at different time periods differed significantly. The Pearson correlation coefficient was used to assess associations between the variables of interest at each time period. Linear mixed models were used for longitudinal subset analyses. A p value of <0.05 was considered significant.

Source of Funding

No external funding was received for the study.

Results

Demographics (Table I)

Ten of the subjects were male and eleven were female. The mean age at the index procedure was fifty-eight months (range, twenty-four to 131 months), and the mean duration of follow-up was seventy-two months (range, thirty-eight to 103 months). The etiology of the scoliosis was syndromic in six patients, congenital in eleven, neuromuscular in three, and idiopathic in one (Table I). The mean number of expansion procedures per patient was eleven, and the mean interval between expansions was 6.4 months. Two hundred and forty-five surgical procedures were performed; 171 were performed purely for expansion, sixty-three were for rod exchanges, and eleven were unplanned. The mean number of pulmonary function tests per patient was ten, resulting in a total of 210 tests performed during the treatment period.

Construct Types

Fifteen patients had a distraction device between a rib and the iliac wing or a lumbar vertebra (with or without a rib-to-rib implant), five patients had only unilateral rib-to-rib constructs, and one patient had only bilateral rib-to-rib constructs. A thoracotomy was performed at the time of initial implantation in thirteen patients; the decision to perform a thoracotomy was made on the basis of the presence of fused ribs and the severity of the deformity.

Complications

There were no neural or vascular complications. Eleven implant-related complications required unplanned surgical procedures. Five of these were performed for implant complications such as implant dislodgement or rod breakage. The remaining six were performed for wound revisions; three of these were performed for deep infection and three for skin breakdown secondary to implant prominence.

Three patients underwent VEPTR device removal without replacement. In one of these patients, a pelvis-to-rib device was removed because a CT scan showed extensive spontaneous fusion and the device was deemed no longer necessary. However,
despite the observed fusion, the patient showed increased coronal imbalance after device removal. In the second patient, a rib-to-rib device was removed because of skin compromise. In the third patient, a pelvis-to-rib device was removed to eradicate an implant-related infection. All three patients had worsening of deformity after removal. One of these patients has already undergone posterior instrumented arthrodesis, and definitive posterior instrumented arthrodesis is planned for the other two patients.

**Pulmonary Function (Table II)**

FVC increased from the preoperative mean of 0.65 L to 0.96 L (p < 0.001) at the time of the last follow-up (Fig. 1). The increase in FVC continued throughout the treatment period in almost all patients (see Appendix). The percentage of the predicted FVC based on arm span decreased significantly from 77% to 58% (p = 0.0001) (Fig. 2; see Appendix). Although the absolute Crs value increased from 18.1 to 20.2 mL/cm H$_2$O (p = 0.08), the compliance normalized for body weight, Crs/kg, decreased by 39% from the initial mean of 1.4 to 0.86 mL/cm H$_2$O/kg; this decrease was significant (p = 0.0006) (Fig. 3). Plots of the normalized Crs values of almost all individual patients over time showed a similar trend (see Appendix).

As the patient cohort was very heterogeneous in many aspects, subset analyses were performed to better understand the data. A subset analysis utilizing linear mixed models was performed to compare the largest two etiology subgroups, congenital and syndromic; no significant differences in the Cobb angle, maximum thoracic kyphosis angle, normalized Crs value, or percentage of the predicted FVC value were found.

We also categorized and compared long and short congenital deformities. Congenital abnormalities involving fewer than five vertebrae were arbitrarily defined as short deformities. Patients with short deformities had significantly higher FVC and normalized Crs values initially (p < 0.05), but none of the pulmonary variables differed significantly between patients with short and long congenital deformities at subsequent times. Coronal and sagittal-plane deformities did not differ significantly in magnitude between long and short congenital deformities. We did not perform a subset analysis to compare different constructs because of the small number of patients with solely rib-to-rib constructs. A
comparison of patients with and without a thoracotomy revealed no significant difference in any parameter. There were no significant correlations between age at the index implantation surgery and any pulmonary outcome parameter (p > 0.05).

Associations between deformity magnitude and pulmonary parameters were also investigated. The Cobb angle was not correlated with any of the pulmonary parameters at any time period. Significant inverse correlations were found between the maximum thoracic kyphosis and the percentage of the predicted FVC at two time periods, before implantation (p = 0.0074) and before the first lengthening (p = 0.03). However, the correlation between a lower kyphosis magnitude and a higher percentage of the predicted FVC did not reach significance at the time of the last follow-up (p = 0.0525).

**Radiographic Findings (Table II)**
Prior to the initial implantation, the mean primary Cobb angle was 80° (range, 16° to 127°), and the mean maximum thoracic kyphosis angle was 57° (range, 7° to 107°). The modest initial coronal correction, which averaged slightly more than 10° (p = 0.002), was maintained at the time of the last follow-up (Cobb angle, 67°) (Fig. 4). Initial instrumentation provided approximately 7° of correction of thoracic kyphosis (p = 0.03). However, thoracic kyphosis subsequently deteriorated again to a mean of 66° at the time of the last follow-up, a value that did not differ significantly from the preoperative value (p = 0.08) (Fig. 5). Maintenance of the initial deformity correction was achieved in most patients (Fig. 6). Global thoracic kyphosis followed the same pattern as the maximum kyphotic deformity, although it was less severe. Increased proximal thoracic kyphosis occurred in four patients (see Appendix); VEPTR expansion in these children was halted, and definitive posterior arthrodesis surgery is planned.

The mean gain in T1-T12 height between the implantation surgery and the last lengthening was 18 mm, or 2.9 mm/year. The mean increase in overall standing height was 23 cm over the mean follow-up period of 6.0 years. On the convex side of the deformity, the mean height of the hemithorax (as measured from the dome of the diaphragm to the first rib) increased from 113 mm preoperatively to 128.1 mm at the time of the last follow-up. On the concave side of the deformity, the mean height of the hemithorax increased from 86.7 to 111.1 mm. The SAL ratio improved from 0.77 on the preoperative radiographs to 0.87 at the time of the last lengthening.
The SAL ratio was not correlated with the FVC at any time period ($p > 0.81$). The sum of the values for the two hemithoraces, however, was positively correlated with the FVC value at each time period ($r > 0.75$, $p < 0.0001$).

Eighteen of the twenty-one patients had some degree of coronal imbalance prior to the initial implantation (mean, 35 mm; range, 7 to 114 mm), two patients were not able to stand or sit and were therefore not included in coronal and sagittal balance measurements, and only one patient had neutral coronal alignment preoperatively. Mean coronal balance improved slightly at the time of the last follow-up (to 31 mm, $p = 0.7$). Mean sagittal balance was positive by 31 mm before implantation and by 23 mm ($p = 0.5$) at the time of the last follow-up. The mean changes in coronal and sagittal balance were not significant.

Discussion

This study evaluated the pulmonary and radiographic outcomes of serial VEPTR expansions at a mean of six years of follow-up in children with thoracic insufficiency syndrome. The results revealed that thoracoplasty with VEPTR instrumentation resulted in modest correction of the Cobb angle with a tendency toward increasing kyphosis during the treatment period. Pulmonary function studies showed that the FVC increased significantly with time but that the rate of increase lagged behind body growth, as indicated by a decrease in the percentage of the predicted FVC value. We also observed a general increase in chest wall stiffness, as indicated by decreased respiratory system compliance.

Despite the previously proposed benefits of the VEPTR device, recent literature suggests that serious problems may be associated with this treatment method and long-term outcomes may be less optimistic than those reported in the earlier literature.

Campbell et al. reported data on twenty-seven patients with thoracic insufficiency syndrome, but only three of the patients were old enough to cooperate with preoperative pulmonary function testing. Vital capacity improved in these three patients, but the percentage of the predicted value declined. Emans et al. reported increased chest volume on postoperative CT scans. In contrast, Mayer and Redding reported no change in absolute FVC and a significant decrease in the percentage of the predicted FVC eight months after VEPTR treatment in a large cohort of older patients with thoracic insufficiency syndrome (fifty-three patients; mean age at insertion, 9.1 years). However, the spirometry technique used before and after the VEPTR treatment depends on a patient’s active inhalation and exhalation, therefore requiring cooperation, and repeatability may be low. A recent study comparing preoperative and follow-up pulmonary function tests also failed to demonstrate any significant improvements in lung volume and function in a group of twenty-six patients with early-onset scoliosis.

Most studies reporting pulmonary function have used conventional pulmonary function testing in older children. However, children who undergo VEPTR treatment for early-onset scoliosis and thoracic insufficiency syndrome are often not mature enough or mentally and physically competent to cooperate with such studies. Our measurement technique differed from those in the aforementioned studies in that we measured lung volumes under general endotracheal anesthesia. This method provided consistent and highly reproducible lung volume measurements, including FVC, inspiratory capacity (IC), and flow function indices in patients of all ages. Similar to the findings of Emans et al. and Campbell et al., our results demonstrate that absolute lung volume increased as a result of VEPTR expansion thoracoplasty treatment. Our data additionally imply that the improvement continued during the treatment period, with further increases in FVC after the initial implantation (see Appendix). This increase in FVC may be the result of the improved physical capacity of the chest as a result of the VEPTR treatment or a result of the anatomical growth of the child. As the study did not include a control group, it is not possible to determine the contribution of the VEPTR treatment. The increase in FVC did not keep up with overall patient growth, as the percentage of the predicted volume declined gradually over time (see Appendix). This percentage based on arm span decreased over time since little thoracic growth occurred and arm growth was relatively normal in these children. The clinical importance of the increase in absolute FVC along with the decrease in the percentage of the predicted value is a subject of debate.

Another interesting finding of the study involves the SAL ratio, which has been used in the early-onset scoliosis literature. Although the SAL ratio in the present study was not correlated with the FVC, the sum of the heights on the concave and convex sides was correlated with the FVC and may therefore be more useful.

We also measured the Crs value, indicating the compliance of the respiratory system, with the passive deflation technique. This value represents the compliance of the lung and that of the thoracic wall. Although the lung size of most of the patients in the present study was relatively small, airway function was nearly normal in most patients, with the exception of a few with mild increases in airway resistance. The decreasing normalized Crs values over time in the present study, therefore, apparently resulted from increasing stiffness of the thorax.

Subgroup analyses were performed to enhance the understanding of the effects of VEPTR on deformities with different causes; however, the number in each subgroup was small, and the results must therefore be interpreted with caution as it is possible that the significance of results may change if larger groups are analyzed. According to our analyses, addition of a thoracotomy did not worsen the pulmonary function parameters compared with those in the subgroup without a thoracotomy. We believe this to be valuable information, as it shows that thoracotomy may not be necessarily detrimental to long-term pulmonary function in this patient population. Subgroup analysis also showed that patients with smaller congenital anomalies fared better in terms of pulmonary function. However, this should be expected, since an increase in the number of segments affected by a congenital anomaly will likely increase the stiffness resulting from the deformity. One surprising finding of the subset analysis...
is the lack of differences between the subgroups with congenital and syndromic deformities. However, the number of patients in each subgroup was small, and larger patient series with more homogeneous diagnoses will be required to draw more definitive conclusions regarding the pulmonary effects of VEPTR treatment.

Earlier studies showed initial correction of coronal deformity after VEPTR implantation followed by maintenance of the correction with reported corrections of up to 40%. A recent multicenter study by Flynn et al. reported a mean of 8.9° of coronal Cobb angle improvement in a group of twenty-four patients with nonsyndromic congenital scoliosis; our findings were similar. Although there is general agreement in the literature that maintenance of the spinal deformity correction occurs with VEPTR treatment, Lattig et al. reported worsening coronal balance as well as increased rigidity of the curves secondary to spontaneous fusions, which in turn rendered the final arthrodesis surgery more extensive and the end result less satisfactory. They also noted development of high thoracic kyphosis. Spontaneous fusions of ribs and vertebrae have been reported by other authors. At least one group reported increased kyphosis magnitude and rigidity with VEPTR treatment. Four patients in the present study had a substantial increase in proximal thoracic kyphosis, with the final maximum value exceeding 100° in three of the patients. This agrees with the findings of Lattig et al. and underlines the concern regarding increasing proximal thoracic kyphosis and a difficult definitive arthrodesis procedure in these children. Four patients in the present study had worsening of coronal imbalance to >4 cm, and five had worsening of sagittal imbalance to >4 cm. Therefore, we also found that loss of spinal balance may become an issue during the longer-term follow-up in such patients.

Data regarding the growth of the spine during VEPTR treatment in children with complex spinal deformities have been scarce. Previous studies on patients with scoliosis indicated growth of the thoracic spine at a rate similar to normal. However, the height gain in the thoracic spine in the present study occurred more slowly than normal (2.9 mm/yr; mean gain, 18 mm). The minimal height gain in the thoracic spine may have been due to congenital vertebral anomalies. Additionally, because of the three-dimensional deformity, evaluation of vertebral growth in a deformed spine is difficult. Since the lung volume increased, the change in T1-T12 height in anteroposterior spinal radiographs may not be a valid proxy for spinal growth in children with a complex spinal deformity.

In summary, the findings of this study raise questions about the utility of VEPTR treatment in early-onset scoliosis. Absolute FVC increased over the treatment period; however, the percentage of the predicted FVC decreased and chest stiffness increased. Although VEPTR treatment did not result in definite gains in pulmonary function as measured by the percentage of the predicted FVC, it may have acted to prevent further worsening. The retrospective nature of the study and the lack of a control group prevent us from identifying how much change in pulmonary function was secondary to natural growth and disease progression and how much was caused by the VEPTR treatment. Worsening of deformity with long-term treatment is a concern and should be weighed against the potential gains from repeated surgical expansion procedures.

Appendix

A more detailed description of the pulmonary function testing as well as figures showing changes in FVC and normalized Crs values over time in the individual patients and serial radiographs of a patient with postoperative thoracic kyphosis and spinal imbalance are available with the online version of this article as a data supplement at jbjs.org.

References

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