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Weight Gain Following Vertical Expandable Prosthetic Titanium Ribs Surgery in Children With Thoracic Insufficiency Syndrome

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Study Design. Retrospective review of patients from a multicenter database.

Objective. To evaluate the nutritional status of children with thoracic insufficiency syndrome (TIS) and to determine if treatment with vertical expandable prosthetic titanium ribs (VEPTR) leads to improvements in weight percentile.

Summary of Background Data. Children with pulmonary insufficiency characteristically have poor nutrition as the energy expenditure from the extra work of breathing approaches the nutritional gain of eating. To our knowledge, no previous studies have examined the relation between VEPTR and potential nutritional improvements in children with TIS.

Methods. Seventy-six patients at 7 different institutions underwent placement of VEPTR devices for treatment or prevention of TIS. Mean age at surgery was 3.7 years (range, 8 months–14 years). All patients were observed for a minimum of 24 months with an average follow-up of 3.3 years (range, 2–6 years). Before surgery and at each postoperative visit, patients were weighed and the Cobb angle was measured. All weights were converted to normative percentiles based on the patient's age.

Results. Overall, we found a significant increase in the percentile of patients' weights after VEPTR surgery ($P = 0.0004$). Of the 76 patients in our series, 60 (79%) were $\leq$5 percentile in weight before surgery. Of these most nutritionally depleted patients, 40% (24/60) had increase in percentile weight after surgery; more subjects may have improved, but due to the basement effect $\leq$5 percentile any improvement in which the final weight was under 5 percentile may not have been detected. Of the 16 patients who were $>$5 percentile weight before surgery, 50% (8/16) had increased weight percentiles after surgery. For most patients, the majority of weight gain occurred between 4 and 8 months after surgery; weight gain continued up to 48 months after surgery. The change in Cobb angle had no relation to the change in weight percentile.

Conclusion. A total of 79% of patients with TIS were less than 5 percentile in weight, thus meeting the criteria for "failure to thrive." Our study demonstrates a significant improvement in the nutritional status of these children after VEPTR surgery, which is an important outcome measure in this population.

Key words: VEPTR, thoracic insufficiency syndrome, weight gain, nutrition. **Spine 2009;34:2530–2533**

Thoracic insufficiency syndrome (TIS) is the inability of the thorax to support normal respiration or lung growth.[1] TIS may occur when a complex chest and/or spine deformity compromises thoracic volume and function, and consequently negatively impacts the growth, development, and function of the lungs. It is well known that children with pulmonary insufficiency are often nutritionally deficient as the energy expenditure in the extra work of breathing approaches the nutritional gain of eating.[2,3]

Because of this association, nutritional consultation is considered an essential part of the pulmonary care of these children. For growth and development, it is important to keep caloric intake greater than energy demands. Dimeglio estimates that weight increases 20-fold from birth to adulthood, one-third of which occurs during the first 5 years of life.[4]

Children with TIS may be treated with the vertical expandable prosthetic titanium rib (VEPTR) device. The technical goal of surgery is to enlarge the thorax acutely and over time with surgical expansion of the VEPTR device. It is hoped that this will control the spine deformity and improve pulmonary capacity and function. Previous work has demonstrated that VEPTR treatment enlarges the thorax, but no studies to date have unequivocally proved an improvement in pulmonary function.[5,6] To our knowledge, no previous studies have examined the relation between VEPTR and potential nutritional improvements in children with TIS.

The purpose of our study was to determine the nutritional status of children with TIS and to determine the change in percentile weight for these children after treatment with VEPTR.

Materials and Methods

A prospectively collected multicenter VEPTR database of 214 patients was queried to find those patients with a minimum follow-up of 24 months. We identified a subcohort of 76 subjects who satisfied this criteria. The sample included patients from 7 sites: Christus Santa Rosa Children's Hospital, Chil-

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dren's Hospital Los Angeles, Children's Hospital and Regional Medical Center Seattle, Primary Children's Medical Center, Salt Lake City, Children's Hospital of Pittsburgh, Shriner's Hospital for Children, in Philadelphia, and Children's Hospital of Boston.

Patient's age at initial surgery, diagnosis, and length of follow-up were extracted from the database. Subjects were evaluated before surgery, and after surgery at 4-month intervals until 24 months after surgery. After this point, patients were seen once a year. Weight measurements and Cobb angles were recorded from each visit. All weights were converted to normative age-adjusted percentiles. It is important to note that the normative data has a basement effect ≤5 percentile. This means that a change in percentile from 2 percentile to 5 percentile would not be detected. Similarly, a change from 2 percentile to 6 percentile would only show a change of 1 percentile. Thus in children whose weights are ≤5 percentile before surgery, a subsequent gain in weight can be underestimated.

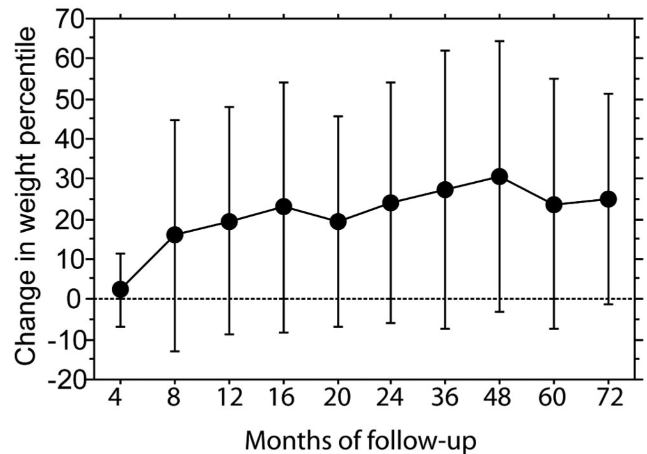

Figure 1. Postoperative change in weight percentile (mean ± SD) *versus* time in subjects who increased or decreased weight percentile (N = 36). Gains in weight percentile seem to increase up to 48 months after surgery.

Statistical Methods

Paired Student *t* tests were used to compare preoperative and postoperative weight percentiles. Simple linear regression was used to examine the relationship between changes in Cobb angle (dependent variable) and changes in weight percentile (independent variable).

■ Results

At the time of the initial surgery, the average age of the patients in our series was 3.7 years (range, 8 months–14 years). All patients were observed for a minimum of 24 months with an average follow-up of 3.3 years (range, 2–6 years).

Diagnoses included Jarcho-Levin syndrome (n = 22), Jeune syndrome (n = 18), thoracogenic scoliosis (n = 11), VACTERL syndrome (n = 6), Goldenhar syndrome (n = 3), Pierre-Robin syndrome (n = 2), cerebro-costo-mandibular syndrome (n = 2), and a multitude of single diagnoses (n = 12).

The mean preoperative weight of our study population was 12.4 kg (range, 4.8–44.3 kg). The mean weight at final follow-up was 20.9 kg (range, 8.4–65.6 kg). Seventy-five of 76 patients (98.6%) gained weight after surgery. The average absolute weight gain over a 12-month period was 2.7 kg. Only one patient lost weight after treatment with VEPTR, dropping from 14.3 kg to 13.9 kg over a 4 year period.

Of our 76 patients, 60 children (79%) were ≤5 percentile in weight before VEPTR surgery. Of these most nutritionally depleted patients, 40% (24/60) showed increased weight percentiles at final follow-up (mean percentile improvement was 27; range, 5–90 percentile). In the 36 children who did not experience improvements, their weight percentile at final follow-up remained less than 5 percentile. The vast majority of these children (35/36) still gained absolute weight after surgery, but this was not sufficient to cause an increase in their weight percentile. It is possible that a change in percentile weight may have occurred, but would not be recognized due to the basement effect of the ≤5 normative percentile.

Of the 16 patients who were more than 5 percentile weight before surgery, 50% (8/16) showed an increase in weight percentile at final follow-up (mean percentile increase was 25; range, 5–50 percentile). Four of these 16 patients had no change in their weight percentile; 4 patients lost weight percentile after surgery. In total, 42% (32/76) of our patients had increases in weight percentile after VEPTR surgery (mean increase = 26 percentile). The mean improvement in weight percentile for our entire study population (76 patients) was statistically significant ($P = 0.0004$).

In the 32 patients who gained weight percentile after treatment with VEPTR, the largest increase was seen between 4 and 8 months after surgery. Gains seemed to increase up to 48 months after surgery (Figure 1).

Before surgery, the average initial Cobb angle for the patients in our series was 52° (range, 0°–110°). Sixty-four of our patients had substantial scoliosis as part of their thoracic deformity (Cobb angle >20°). After surgery, Cobb angle decreased on an average of 5.4°. For those patients with an initial Cobb angle >20°, there was no evident relationship between the change in Cobb angle and change in weight percentile ($P = 0.99$) (Figure 2).

■ Discussion

Adequate nutrition is essential for normal wound healing and reduced postoperative complications such as infection and prolonged hospitalization. For VEPTR surgery, nutrition is especially important as up to 4 devices require soft tissue coverage and repeated lengthenings occur on average twice a year. (Figures 3A, B) For these reasons, a central part of each patient's preoperative preparation was maximization of nutrition status. Each patient was seen by a team including an orthopedic surgeon, a general

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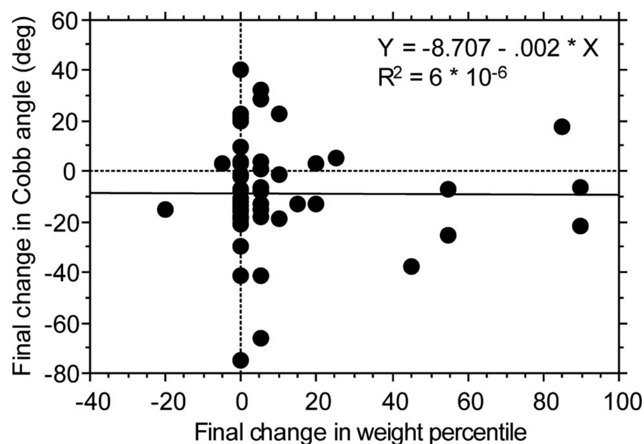

Figure 2. Change in Cobb angle *versus* final change in weight percentile showing no relation between the two in subjects with substantial scoliosis before surgery (cobb angle >20°; N = 64).

surgeon, and a pulmonologist. The patient was not scheduled for surgery until it was felt by the team that nutritional status had been maximized. The exact criteria for this was a team decision, and not objective. Thus, the post-VEPTR weight gain demonstrated is not likely due to nutritional intervention, but most likely a reflection of the VEPTR treatment.

According to the Center for Disease Control Pediatric and Pregnancy Nutrition Surveillance System, underweight is defined as weight-for-age less than fifth percentile and body mass index (BMI)-for-age less than fifth percentile for children 2 to 20 years of age based on the CDC gender-specific BMI-for-age reference.[7] For children with a spine or thoracic deformity, BMI is not a reliable measure of nutrition as it relies on the height measurement. The term "failure to thrive" has also been applied to children less than fifth percentile for body weight.[8]

Children with TIS have complex chest and/or spine deformities that compromise pulmonary function. Due to the increased work of breathing necessary for survival, nutritional depletion can approach the nutritional gain of eating.[2,3] In addition, some children do not have enough pulmonary reserve to hold their breath long enough to swallow while eating repeatedly; this can further contribute to nutritional depletion and "failure to thrive."

In our study, 42% of children had improved weight percentiles following VEPTR surgery. In those that were considered underweight (≤5 percentile), 40% showed an increase after surgery, but this is likely an underestimate. In children who were ≤5 weight percentile before surgery, an improvement in weight percentile may have occurred but might not be recognized due to the basement effect of normative percentiles. For example, a child may have increased from the first percentile to the fifth percentile but still has to be lumped together with all those children less than 5 percentile.

Pulmonary improvement after VEPTR surgery has been difficult to quantify due in part to the limited availability of appropriate pulmonary function tests in children. The extra work required to breathe requires additional calories which makes it difficult for the child to gain weight. It may be hypothesized that improved weight percentiles following VEPTR surgery may indicate better pulmonary function. However, further study is necessary to investigate this possibility.

An inherent limitation in this case series with prospectively collected data are the lack of a control group. As most of these children are sick, and getting

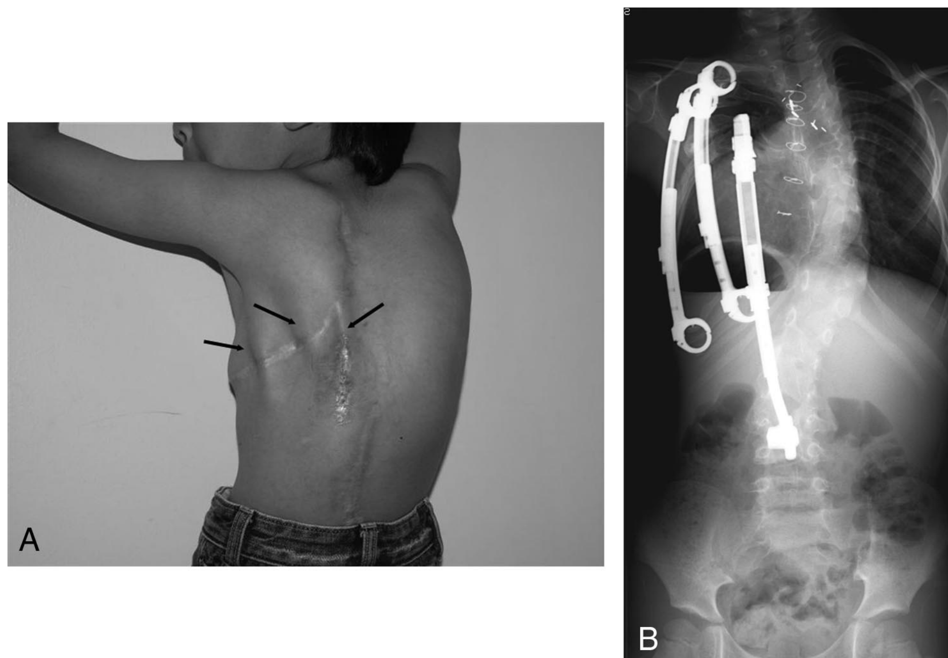

Figure 3. **A, B**: Thin body habitus of a typical VEPTR patient (**A**) with arrows pointing out the subcutaneous location of the VEPTR device seen in the accompanying radiograph (**B**).

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sicker, withholding clinical care was not an option. Likewise, it would be impossible to compare VEPTR to another treatment for this population as there is no commonly accepted alternative treatment for such disparate diagnoses as Jarcho-Levin Syndrome and Jeunes Syndrome. In addition, there is at least one confounding variable that should be considered when interpreting our data. It is possible that the close attention given to these children by parents and health care providers during the postoperative period could result in a better diet and weight gain independent of VEPTR insertion. Improvements in weight, however, appear to persist up to 48 months after surgery which is long after the "acute" postoperative period where the child would receive the most attention.

A further limitation of this study is that while all children have TIS, there are underlying diagnoses with insufficient numbers to examine the efficacy of VEPTR for each cause of TIS. Furthermore, this study can only show an association of weight gain with VEPTR treatment, and not causation. Although unlikely, it is theoretically possibly that these patients of variable ages were all just about to undergo a growth spurt at the start of VEPTR treatment. This is the first study we are aware of, which demonstrates an improved objective outcome measure following VEPTR surgery.

This study highlights the very poor nutritional status of children with TIS, with 79% of patients before surgery being ≤5 percentile in weight. Our data also supports an improvement in weight percentile of children with TIS after VEPTR surgery, which is a critically important outcome measure in this population. Weight gain after VEPTR surgery may be secondary to improved pulmonary function, although further studies are needed to test this hypothesis.

■ Key Points

- The majority of patients with TIS are less than 5 percentile in weight, meeting the criteria for "failure to thrive."
- The use of VEPTR in this patient population results in a statistically significant increase in weight percentile after surgery.
- Weight gain continued up to 48 months after surgery.
- Change in Cobb angle had no relation to the change in weight percentile.

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