

POLICY TITLE	VERTICAL EXPANDABLE PROSTHETIC TITANIUM RIB
POLICY NUMBER	1.136

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I. POLICY

Use of the vertical expandable prosthetic titanium rib is considered **medically necessary** in the treatment of progressive thoracic insufficiency syndrome due to rib and/or chest wall defects in infants/children between 6 months of age and skeletal maturity.

Use of the vertical expandable prosthetic titanium rib for all other conditions, including but not limited to the treatment of scoliosis in patients without thoracic insufficiency, is considered **investigational** as there is insufficient evidence to support a conclusion concerning the health outcomes or benefits associated with this procedure.

POLICY GUIDELINES

Skeletal maturity occurs at about age 14 for girls and age 16 for boys.

Given the complexity of these procedures and patients, implantation of this device should be performed in specialized centers. Preoperative evaluation requires input from a pediatric orthopedist, pulmonologist, and thoracic surgeon. In addition, preoperative evaluation of nutritional, cardiac, and pulmonary function (when possible) is required.

Cross-reference:

MP-1.120-Interventions for Progressive Scoliosis

II. PRODUCT VARIATIONS

[N] = No product variation, policy applies as stated

[Y] = Standard product coverage varies from application of this policy, see below

- [N] Capital Cares 4 Kids
- [N] PPO
- [N] HMO
- [N] SeniorBlue HMO
- [N] SeniorBlue PPO

- [N] Indemnity
- [N] SpecialCare
- [N] POS
- [Y] FEP PPO*

*Refer to FEP Medical Policy Manual MP-7.01.10 Vertical Expandable Prosthetic Titanium Rib. The FEP Medical Policy manual can be found at: www.fepblue.org

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III. DESCRIPTION/BACKGROUND

The vertical expandable prosthetic titanium rib (VEPTR) is a curved rod placed vertically in the chest that helps to shape the thoracic cavity. It is positioned either between ribs or between the ribs and either the spine or pelvis. The device is designed to be expanded every 4 to 6 months as growth occurs and also to be replaced if necessary. Some patients require multiple devices.

It is being evaluated for use in skeletally immature patients with thoracic insufficiency syndrome (TIS) and to slow or correct curve progression in pediatric patients with scoliosis without TIS.

Thoracic insufficiency syndrome (TIS) is the inability of the thorax to support normal respiration or lung growth. (1) The condition results from serious defects affecting the ribs or chest wall, such as severe scoliosis, rib fusion (which may accompany scoliosis), and various hypoplastic thorax syndromes, such as Jeune syndrome and Jarcho-Levin syndrome. Spine, chest, and lung growth are interdependent. While the coexistence of chest wall and spinal deformity is well-documented, this effect on lung growth is not completely understood.

Progressive TIS includes respiratory insufficiency, loss of chest wall mobility, worsening 3-dimensional thoracic deformity, and/or worsening pulmonary function tests. As a child grows, progressive thoracic deformity and rotation toward the concave side occurs with worsening respiratory compromise. This progression is often accompanied by a need for supplemental oxygen and can require mechanical ventilation. While spinal fusion is one approach to treatment, it may not be successful and also may limit growth (lengthening) of the spine.

Regulatory Status

A VEPTR has received approval from the U.S. Food and Drug Administration (FDA) under a humanitarian device exemption (HDE). The FDA review noted that the device is indicated for the treatment of thoracic insufficiency syndrome (TIS) in skeletally immature patients. TIS is defined as the inability of the thorax to support normal respiration or lung growth.

For the purpose of identifying potential TIS patients, the categories in which TIS patients fall are as follows:

- Flail chest syndrome
- Rib fusion and scoliosis
- Hypoplastic thorax syndrome, including:
 - Jeune’s syndrome
 - Achondroplasia
 - Jarcho-Levin syndrome

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- Ellis van Creveld syndrome

This review also indicated that the device should not be used in patients younger than 6 months.

Use of the VEPTR in pediatric patients with scoliosis without TIS is an off-label indication.

IV. RATIONALE

Thoracic insufficiency occurs in a limited patient population, and the literature on use of the vertical expandable prosthetic titanium rib (VEPTR) consists, in general, of case series from single institutions. Some series are from specialized pediatric centers. No comparative trials have been identified.

Thoracic Insufficiency Syndrome

Data submitted to the U.S. Food and Drug Administration (FDA) includes an initial feasibility study involving 33 patients and a subsequent prospective study of 224 patients (214 with baseline data) at 7 study sites. (3) Of these patients, 94 had rib fusion, 93 had hypoplastic thoracic syndrome, 46 had progressive scoliosis, and 14 had flail chest as a cause of their thoracic insufficiency syndrome (TIS). Three- and 5-year follow-up rates for the multicenter study were approximately 95%. Of the 247 patients enrolled in either study, 12 patients died (4.8%) and 2 withdrew. None of the deaths was determined by investigators to be device-related. Since standard pulmonary function testing was not possible for most of this population, an assisted ventilatory rating (AVR) was used to assess impact on respiratory status. The AVR ranged from 0 for unassisted breathing on room air to 4 for full-time ventilatory support. In the multicenter prospective study, the AVR outcome improved or stabilized for 93% of the patients. Data were not reported for the number of patients who were no longer ventilator-dependent.

Campbell, the developer of the device, and colleagues reported on 27 patients who had surgery for TIS and for whom at least 2 years of follow-up data were available; this series was based on 41 patients treated between 1990 and the acceptance of the paper. (4) Entry criteria for this study were acceptance by pediatric general surgeon, pediatric pulmonologist, and pediatric orthopedist; age 6 months to skeletal maturity; progressive TIS; more than 10% reduction in height of the concave hemithorax; and 3 or more anomalous vertebrae, with 3 or more fused ribs at the apex of the deformity. Patients were followed up for an average of 3.2 (range: 2–12) years. Prior to surgery, the mean annual rate of progression was 15 degrees per year (range: 2–50 years). Following surgery the Cobb angle (of scoliosis) improved from 74 degrees to a final value of 49 degrees. Spine growth was at the rate of 0.8 cm per year. (Normal spinal growth is 0.6 cm/year for ages 5–10 years.) The final forced vital capacity (FVC) was 49% of predicted value in the 19

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children who could complete pulmonary function tests (PFTs). Preoperatively, 1 patient required continuous positive airway pressure (CPAP), and 1 needed supplemental oxygen for ventilatory support at final follow-up.

In another series, Gadepalli et al. examined growth and pulmonary function in 26 children who received a VEPTR between October 2006 and March 2010. (5) The children underwent 29 insertions and 57 expansions, with an average of 3 surgeries per child. Each procedure required an average 0.97 days in the intensive care unit and 4.41 days in the hospital. The mean Cobb angle improved by 29% from 64.7 degrees preoperatively to 46.1 degrees postoperatively. Lung volumes measured by yearly thoracic computed tomography (CT) scans were similar when corrected for age. Pulmonary function tests were performed every 6 months in patients (n=12) who were not ventilator-dependent and could cooperate with the procedure. Pulmonary function tests showed no significant change from baseline to follow-up in percent predicted values for forced expiratory volume in 1 second (FEV1) (54.6 vs. 51.8), FVC (58.1 vs. 55.9), or residual volume (145.3 vs. 105.6 – all respectively). Reoperation was required for 14 complications, 4 for chest tube placement (pneumothorax), 1 for seroma drainage, 6 for hardware removal (for infection), and 3 for hardware repositioning (for dislodgement). Another 22 complications were treated nonoperatively.

Emans and colleagues reported results on patients with TIS who underwent the procedure at Children’s Hospital in Boston from 1999 to 2005. (6) Thirty-one patients with fused ribs and TIS were treated; 4 patients had prior spinal arthrodesis with continued progression of deformity. Before surgery, all patients showed progressive spinal deformity, progressive chest deformity, or progressive hemithoracic constriction. The mean age was 4.2 years, and mean follow-up was 2.6 years (range: 0.5 to 5.4 years). A 3-member team selected patients for surgery; cardiac function was also evaluated preoperatively. Surgery was performed using the Campbell technique for vertical expandable prosthetic titanium rib (VEPTR). Device lengthening was planned for every 4 to 6 months but often was longer due to intercurrent illness or difficulty with travel. The mean number of device lengthenings was 3.5 (range: 0–10). Six patients had device exchanges for growth. In 30 patients, the spinal deformity was controlled, and growth continued (1.2 cm/yr) in the thoracic spine during treatment at rates similar to normal children. In this study, the final FVC was 73.5% of predicted levels. Preprocedure, 2 patients were on ventilators and 3 patients required oxygen; at final follow-up, 1 patient required oxygen. Lung volume (measured by CT scan in cubic-cm) in the operated lung increased from 157 preoperatively to 326 at the final follow-up visit.

Motoyama and colleagues from Children’s Hospital in Pittsburgh reported on follow-up of 10 patients with thoracic insufficiency with follow-up as long as 33 months. (7) Using a special portable pulmonary function testing device, they reported on lung function in 10 children who had placement of VEPTR. In this population, the median age was 4.3 years (range: 1.8–9.8 years) at first test, and they followed patients an average of 22 months (range: 7–33 months). At baseline, FVC showed a moderate-to-severe decrease (69% of predicted), indicating the presence

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of significant restrictive lung defect. FVC increased significantly over time, with an average rate of 26.8% per year, similar to that of healthy children of comparative ages. In terms of percent-predicted values, FVC did not change significantly between the baseline and last test (70.3%), indicating that in most children studied, lung growth kept up with body growth.

A series of 22 patients from another Children’s Hospital was published in 2007. (8)

There are a number of additional series, some discuss weight gain following use of vertical expandable prosthetic titanium rib in thoracic insufficiency syndrome (9) while others discuss early changes in pulmonary function. (10)

Scoliosis without Thoracic Insufficiency Syndrome

White et al. reported the off-label use of spine-to-spine VEPTR to treat spinal deformity in 14 children without chest wall abnormalities. (11) The indications for the dual spine-to-spine rods were absence of a primary chest wall deformity, progression of spinal deformity to a Cobb angle of greater than 50 degrees, and migration of a previously placed proximal rib anchor or of a prior non-VEPTR growing rod to the point of loss of stable fixation. At final follow-up (24-48 months), there was an improvement in the Cobb angle from 74 to 57 degrees, an increase in T1-S1 height from 260 to 296 mm, and no significant change in kyphosis. Complications occurred in 6 of 14 patients (43%) and included 3 rod fractures in 2 patients, 3 superficial infections, and 1 case of prominent hardware that threatened skin integrity. As noted by the authors, while results are similar to those obtained with other growing rods, “the high complication rates, need for multiple procedures in growing children, and small relative gains in radiographic parameters still challenge proof of efficacy of all such treatment methods.”

Adverse Events

The complications that occur with this device need to be considered by practitioners and families as they are discussing this procedure. Information on complications is summarized using data from the FDA review and the papers by Campbell and Emans. (3, 4, 6) Up to 25% of patients may experience device migration, including rib erosion. However, there does not seem to be significant long-term consequences from this. Approximately 10% of patients had infection-related complications. Brachial plexus injury or thoracic outlet syndrome occurred in 1% to 7% of these series. Skin sloughing was reported in 4 patients (15%) in the study published by Campbell.

Ongoing Clinical Trials

A search of online site ClinicalTrials.gov in March 2012 found a multicenter trial of use of the vertical expandable prosthetic titanium rib (VEPTR) in children with early onset scoliosis without rib abnormalities (NCT00689533). The primary objective of this study is to evaluate the

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use of unilateral or bilateral VEPTR devices, with or without expansion thoracoplasty, for preventing further progression of the angle, allowing for spinal growth and improving pulmonary function in the treatment of children (18 months to 10 years) with progressive scoliosis without rib abnormalities. The study is sponsored by the Shriners Hospitals for Children in association with the Chest Wall and Spine Deformity Research Foundation. The trial began in 2008 with an estimated enrollment of 250 patients. Final data collection is expected in 2012 for the primary outcome measure, with completion of the study (follow-up until completion of spinal growth) in 2016.

Summary

No comparative trials have described the use of this device. Thoracic insufficiency occurs in a limited patient population; for example, the Boston center reported results on 31 children treated from 1999 to 2005. The natural history of progressive TIS is worsening pulmonary function and worsening pulmonary insufficiency.

Results from the series reported at different specialty centers demonstrate improvement and/or stabilization in key measures with use of this device in progressive TIS. This improvement is noted in measures related to thoracic structure (e.g., Cobb angle for those with scoliosis), growth of the thoracic spine and lung volumes, and stable or improved ventilatory status. While pulmonary function testing is very difficult in these patients, one study does demonstrate an age-specific increase in forced vital capacity (FVC), and the studies report a final FVC in the range of 50–70% of predicted value.

Given the usual disease course of worsening thoracic volume and ventilatory status, the stabilization/improvement in these measures would be highly unlikely in the absence of the intervention. Taken together, these various outcome measures demonstrate the positive impact of this procedure.

Thus, this intervention may be considered medically necessary in children with progressive thoracic insufficiency syndrome due to rib and/or chest wall defects. Given the complexity of this procedure and the patient population, use of this device should be performed in specialized centers. Preoperative evaluation requires input from a pediatric orthopedist, pulmonologist, and thoracic surgeon. In addition, preoperative evaluation of nutritional, cardiac, and pulmonary function (when possible) is required.

The VEPTR is also being evaluated for curves greater than 45 degrees in infants and juveniles without thoracic insufficiency. Limited data are available on the use of the VEPTR for early onset scoliosis without thoracic insufficiency; therefore, this is considered investigational.

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V. DEFINITIONS

NA

VI. BENEFIT VARIATIONS

The existence of this medical policy does not mean that this service is a covered benefit under the member's contract. Benefit determinations should be based in all cases on the applicable contract language. Medical policies do not constitute a description of benefits. A member's individual or group customer benefits govern which services are covered, which are excluded, and which are subject to benefit limits and which require preauthorization. Members and providers should consult the member's benefit information or contact Capital for benefit information.

VII. DISCLAIMER

Capital's medical policies are developed to assist in administering a member's benefits, do not constitute medical advice and are subject to change. Treating providers are solely responsible for medical advice and treatment of members. Members should discuss any medical policy related to their coverage or condition with their provider and consult their benefit information to determine if the service is covered. If there is a discrepancy between this medical policy and a member's benefit information, the benefit information will govern. Capital considers the information contained in this medical policy to be proprietary and it may only be disseminated as permitted by law.

VIII. REFERENCES

1. *Campbell RM, Jr., Smith MD, Mayes TC et al. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. J Bone Joint Surg Am 2003; 85-A(3):399-408.*
2. *Campbell RM, Jr. VEPTR: past experience and the future of VEPTR principles. Eur Spine J 2013; 22 Suppl 2:S106-17.*
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4. *Campbell RM, Jr., Smith MD, Mayes TC et al. The effect of opening wedge thoracostomy on thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. J Bone Joint Surg Am 2004; 86-A(8):1659-74.*
5. *Gadepalli SK, Hirschl RB, Tsai WC et al. Vertical expandable prosthetic titanium rib device insertion: does it improve pulmonary function? J Pediatr Surg 2011; 46(1):77-80.*
6. *Emans JB, Caubet JF, Ordonez CL et al. The treatment of spine and chest wall deformities with fused ribs by expansion thoracostomy and insertion of vertical*

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expandable prosthetic titanium rib: growth of thoracic spine and improvement of lung volumes. Spine (Phila Pa 1976) 2005; 30(17 Suppl):S58-68.

7. *Motoyama EK, Deeney VF, Fine GF 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(3):284-90.*
8. *Waldhausen JH, Redding GJ, Song KM. Vertical expandable prosthetic titanium rib for thoracic insufficiency syndrome: a new method to treat an old problem. J Pediatr Surg 2007; 42(1):76-80.*
9. *Skaggs DL, Sankar WN, 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(23):2530-3.*
10. *Mayer OH, 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(1):35-8.*
11. *White KK, Song KM, Frost N et al. VEPTR growing rods for early-onset neuromuscular scoliosis: feasible and effective. Clin Orthop Relat Res 2011; 469(5):1335-41.*

IX. CODING INFORMATION

Note: This list of codes may not be all-inclusive, and codes are subject to change at any time. The identification of a code in this section does not denote coverage as coverage is determined by the terms of member benefit information. In addition, not all covered services are eligible for separate reimbursement.

Covered when medically necessary:

CPT Codes®							
22899	20999						

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ICD-9-CM Diagnosis Code*	Description
737.8	Other Curvature of spine
737.30 – 737.39	Scoliosis [and kyphoscoliosis], idiopathic code range
737.40 – 737.43	Curvature of spine, unspecified code range

MEDICAL POLICY

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ICD-9-CM Diagnosis Code*	Description
754.2	Congenital musculoskeletal deformities spine
756.3 – 756.4	Other anomalies of ribs and sternum code range
756.9 -	Other and unspecified anomalies of musculoskeletal system
756.10 – 756.16	Anomaly of spine, unspecified code range
756.55	Chondroectodermal dysplasia
517.8	Lung involvement in other diseases classified elsewhere
518.82	Other pulmonary insufficiency, not elsewhere classified

*If applicable, please see Medicare LCD or NCD for additional covered diagnoses.

The following ICD-10 diagnosis codes will be effective October 1, 2014:

ICD-10-CM Diagnosis Code*	Description
J80	Acute respiratory distress syndrome
M40.13 – M40.14	Other secondary kyphosis, cervicothoracic region
M40.55	Lordosis, unspecified, thoracolumbar region
M41.00 – M41.9	Scoliosis code range
M43.8x3 – M43.8x5	Other specified deforming dorsopathies, cervicothoracic region code range
M41.03 – M41.04	Infantile idiopathic scoliosis, cervicothoracic region code range
M41.30 – M41.35	Thoracogenic scoliosis, site unspecified code range
M41.83 – M41.85	Other forms of scoliosis, cervicothoracic region
M41.113 – M41.115	Juvenile idiopathic scoliosis, cervicothoracic region code range
M41.123 – M41.125	Adolescent idiopathic scoliosis, cervicothoracic region
M96.5	Postradiation scoliosis
Q76.2	Congenital spondylolisthesis

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ICD-10-CM Diagnosis Code*	Description
Q76.3	Congenital scoliosis due to congenital bony malformation
Q76.6	Other congenital malformations of ribs (includes congenital absence of rib and congenital fusion of ribs)
Q76.425	Congenital lordosis, thoracolumbar region
Q76.49	Other congenital malformations of spine, not associated with scoliosis
Q77.6	Chondroectodermal dysplasia
Q77.2	Osteochondrodysplasia with defects of growth of tubular bones and spine; short rib syndrome (includes Asphyxiating thoracic dysplasia [Jeune])
Q79.8 – Q79.9	Other congenital malformations of musculoskeletal system
Q87.2	Congenital malformation syndromes predominantly involving limbs (includes VATER syndrome)

*If applicable, please see Medicare LCD or NCD for additional covered diagnoses.

X. POLICY HISTORY

MP 1.136	<p>CAC 10/28/12 New policy adopting BCBSA. Information regarding vertical expandable prosthetic titanium rib (VEPTR) was extracted from MP 1.120 Interventions for Progressive Scoliosis. Added additional information on VEPTR and placed in this new policy to match BCBSA. Remains medically necessary with criteria. Codes reviewed 9/20/12 klr</p>
	<p>04/05/13- Policy recoded. -skb</p>
	<p>CAC 11/26/13 Consensus review. References updated but no changes to the policy statements. Rationale added.</p>

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