BlueCross BlueShield of Alabama

## Name of Policy:
# Vertical Expandable Prosthetic Titanium Rib

Policy #: 299
Category: Surgery

Latest Review Date: April 2018
Policy Grade: C

## Background/Definitions:

*As a general rule, benefits are payable under Blue Cross and Blue Shield of Alabama health plans only in cases of medical necessity and only if services or supplies are not investigational, provided the customer group contracts have such coverage.*

*The following Association Technology Evaluation Criteria must be met for a service/supply to be considered for coverage:*

1. *The technology must have final approval from the appropriate government regulatory bodies;*
2. *The scientific evidence must permit conclusions concerning the effect of the technology on health outcomes;*
3. *The technology must improve the net health outcome;*
4. *The technology must be as beneficial as any established alternatives;*
5. *The improvement must be attainable outside the investigational setting.*

*Medical Necessity means that health care services (e.g., procedures, treatments, supplies, devices, equipment, facilities or drugs) that a physician, exercising prudent clinical judgment, would provide to a patient for the purpose of preventing, evaluating, diagnosing or treating an illness, injury or disease or its symptoms, and that are:*

1. *In accordance with generally accepted standards of medical practice; and*
2. *Clinically appropriate in terms of type, frequency, extent, site and duration and considered effective for the patient's illness, injury or disease; and*
3. *Not primarily for the convenience of the patient, physician or other health care provider; and*
4. *Not more costly than an alternative service or sequence of services at least as likely to produce equivalent therapeutic or diagnostic results as to the diagnosis or treatment of that patient's illness, injury or disease.*

## Description of Procedure or Service:

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 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 with TIS.

**Thoracic Insufficiency Syndrome**

Thoracic insufficiency syndrome (TIS) is the inability of the thorax to support normal respiration or lung growth. It results from serious defects affecting the ribs or chest wall such as severe scoliosis, with rib absence or rib fusion (which may accompany scoliosis), and various hypoplastic thorax syndromes such as Jeune's Syndrome and Jarcho-Levin syndrome. Spine, lung, and chest growth are interdependent. While the coexistence of chest wall and spinal deformity is well documented, their 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 concave side occurs with worsening respiratory compromise. This progression is often accompanied by a need for supplemental oxygen and can require mechanical ventilation.

Treatment

While spinal fusion is one approach to treatment, it may not be as successful and may also limit growth (lengthening) of the spine.

The vertical expandable prosthetic titanium rib (VEPTR, Synthes Spine Co) is a curved rod placed vertically in the chest that helps to stabilize and shape the thoracic cavity. It is positioned either between ribs or between the ribs and either the spine or pelvis. VEPTR may be described as "rib based" growth-sparing instrumentation, which is compared with "spine based" growing rods for Cobb angle correction. 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.

## Policy:

Use of the **vertical expandable prosthetic titanium rib meets** Blue Cross and Blue Shield of Alabama's medical criteria for coverage in the treatment of progressive thoracic insufficiency syndrome due to rib and/or chest wall defects in infants/children between six months of age and skeletal maturity (about age 14 for girls, 16 for boys).

**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 does not meet** Blue Cross and Blue Shield of Alabama's medical criteria for coverage and is considered **investigational**.

**Note**: 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 is required.

*Blue Cross and Blue Shield of Alabama does not approve or deny procedures, services, testing, or equipment for our members. Our decisions concern coverage only. The decision of whether or not to have a certain test, treatment or procedure is one made between the physician and his/her patient. Blue Cross and Blue Shield of Alabama administers benefits based on the member's contract and corporate medical policies. Physicians should always exercise their best medical judgment in providing the care they feel is most appropriate for their patients. Needed care should not be delayed or refused because of a coverage determination.*

## Key Points:

This policy was created in 2007 and updated periodically using the MEDLINE database. The most recent review was performed through February 5, 2018.

Evidence reviews assess the clinical evidence to determine whether the use of a technology improves the net health outcome. Broadly defined, health outcomes are length of life, quality of life, and ability to function- including benefits and harms. Every clinical condition has specific outcomes that are important to patients and to managing the course of that condition. Validated outcome measures are necessary to ascertain whether a condition improves or worsens; and whether the magnitude of that change is clinically significant. The net health outcome is a balance of benefits and harms.

To assess whether the evidence is sufficient to draw conclusions about the net health outcome of a technology, 2 domains are examined: the relevance and the quality and credibility. To be relevant, studies must represent one or more intended clinical use of the technology in the intended population and compare an effective and appropriate alternative at a comparable intensity. For some conditions, the alternative will be supportive care or surveillance. The quality and credibility of the evidence depend on study design and conduct, minimizing bias and confounding that can generate incorrect findings. The randomized controlled trial is preferred to assess efficacy; however, in some circumstances, nonrandomized studies may be adequate. Randomized controlled trials are rarely large enough or long enough to capture less common adverse events and long-term effects. Other types of studies can be used for these purposes and to assess generalizability to broader clinical populations and settings of clinical practice.

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. The following is a summary of the literature to date.

**Thoracic Insufficiency Syndrome**

Data submitted to the U.S. Food and Drug Administration (FDA) include an initial feasibility study involving 33 patients and a subsequent prospective study of 224 patients (214 with baseline data) at seven study sites. Of these, 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 two withdrew. None of the deaths was determined by investigators to be device-related. Because 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, et al 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. 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. Before surgery, the mean annual rate of progression was 15° per year (range, 2-50 years). Following surgery, the Cobb angle (of scoliosis) improved from 74° to a final value of 49°. Spine growth was at the rate of 0.8 cm per year. (Normal spinal growth is 0.6 cm/year for ages 5 to 10 years.) The final forced vital capacity (FVC) was 49% of predicted value in the 19 children who could complete pulmonary function tests. Preoperatively, one patient required continuous positive airway pressure, and one needed supplemental oxygen for ventilatory support at final follow-up. Another publication from this group reported average 40.7-month follow-up (range, 25-78 months) in 24 children with nonsyndromic congenital scoliosis. Twenty-three (95.8%) children had associated rib fusions, and the average age at surgery was 3.3 years (range, 0.7-12.5). With a mean of 5 expansion surgeries per patient (range, 1-10), the Cobb angle had improved by a mean of 8.9° and thoracic height improved by a mean of 3.41 cm. Eight of the patients (33%) had a total of 16 adverse events, all of which required surgical intervention.

In another series, Gadepalli et al examined growth and pulmonary function in 26 children who received a VEPTR between October 2006 and March 2010. 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° preoperatively to 46.1° 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 ($FEV_1$; 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 et al reported results on patients with TIS who underwent the procedure at Children's Hospital in Boston from 1999 to 2005. Thirty-one patients with fused ribs and TIS were treated;

four 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, 0.5-5.4 years). A three-member team selected patients for surgery; cardiac function was also evaluated preoperatively. Surgery was performed using the Campbell technique for 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/y) 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 centimeters) in the operated lung increased from 157 preoperatively to 326 at the final follow-up visit.

Motoyama et al from Children's Hospital in Pittsburgh reported on follow-up of 10 patients with thoracic insufficiency with follow-up as long as 33 months. 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 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. There are a number of additional series; some discuss weight gain following use of VEPTR in thoracic insufficiency syndrome while others discuss early changes in pulmonary function.

Other series have discussed weight gain after use of VEPTR in TIS10 or early changes in pulmonary function.

Section Summary: Thoracic Insufficiency Syndrome
The evidence evaluating use of VEPTR thoracoplasty to treat children with progressive TIS due to rib and/or chest wall defects consists of a few case series. TIS 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 pulmonary insufficiency. Results from the case series reported by different specialty centers have demonstrated improvement and/or stabilization in key measures with use of the VEPTR in progressive TIS. This improvement has been noted in measures related to thoracic structure (eg, 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 difficult to track in patients suffering with TIS, a study has demonstrated an age-specific increase in FVC; further still, that same study reported a final FVC in the range of 50% to 70% of predicted value. Given the usual disease course of worsening thoracic volume and ventilatory status, the stabilization and/or improvement in the clinical measures outlined above would be highly unlikely if not for the

intervention. Taken together, these outcomes demonstrate the positive impact of using the VEPTR technology.

**Scoliosis without Thoracic Insufficiency Syndrome**
In 2011, White et al reported the off-label use of spine-to-spine VEPTR to treat spinal deformity in 14 children without chest wall abnormalities. 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°, 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°, 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, three 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."

In 2014, treatment of congenital scoliosis with VEPTR (n=22) was compared with treatment with spinal fusion (n=27) or observation (n=184) based on a prospective registry. Function, pain, and mental health status were measured with the 22-item Scoliosis Research Society questionnaire. Compared with the observation group, the VEPTR group had higher total and image scores in the second and third visits and higher function scores between the third and fourth visits. Interpretation of this study is limited due to a number of confounding factors, including age at treatment, unknown comorbidities, and the rationale for the selection of treatment.

**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 articles by Campbell and Emans. Up to 25% of patients may experience device migration, including rib erosion. 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 four patients (15%) in the study published by Campbell. In one publication reporting on a single-center series reporting on complications for 65 patients treated at a single center for TIS over a 13 year period, device-related complications occurred in 22 patients.

**Summary of Evidence**
For individuals who have progressive TIS who receive VEPTR thoracoplasty the evidence includes small case series. Relevant outcomes include symptoms, morbid events, functional outcomes, treatment-related mortality, and treatment-related morbidity. Thoracic insufficiency occurs in a limited patient population; e.g., the Boston center reported results on 31 children treated from 1999 to 2005. The natural history of progressive thoracic insufficiency syndrome (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% to 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. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals with early-onset scoliosis without TIS who receive VEPTR thoracoplasty, the evidence includes small case series. Relevant outcomes include symptoms, morbid events, functional outcomes, treatment-related mortality, and treatment-related morbidity. The VEPTR is also being evaluated for curves greater than 45° in infants and juveniles without thoracic insufficiency. However, there is less certainty around the progression in disease, and thus in the risk-benefit tradeoff of VEPTR surgery. The evidence is insufficient to determine the effects of the technology on health outcomes.

**Practice Guidelines and Position Statements**
No guidelines or statements were identified.

**U.S. Preventive Services Task Force Recommendations**
Not Applicable.

## Key Words:
Vertical expandable prosthetic titanium rib (VEPTR)

## Approved by Governing Bodies:
Vertical expandable prosthetic titanium rib (VEPTR) received FDA approval for humanitarian device exemption August 23, 2004

A VEPTR initially received approval from the U.S. Food and Drug Administration (FDA) under a humanitarian device exemption (HDE) for the treatment of thoracic insufficiency syndrome (TIS) in skeletally immature patients. In 2014, FDA cleared VEPTR through the 510(k) process. The VEPTR/VEPTR II device is indicated for skeletally immature patients with severe progressive spinal deformities and/or 3-dimensional deformity of the thorax associated with or at risk of TIS. This would include patients with progressive congenital, neuromuscular, idiopathic, or syndromic scoliosis.

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
    - Ellis van Creveld syndrome

## Benefit Application:

Coverage is subject to member's specific benefits. Group specific policy will supersede this policy when applicable.
ITS: Home Policy provisions apply
FEP contracts: Special benefit consideration may apply. Refer to member's benefit plan. FEP does not consider investigational if FDA approved and will be reviewed for medical necessity.

## Coding:

CPT Codes: **22899** Unlisted procedure, spine

## References:


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5. Emans JF, Caubet JF, Ordonez CL, 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. Sep 1 2005; 30(17 Supp): S58-S68.
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11. Motoyama EK, Deeney VF, Fine FG, et al. Effects on lung function of multiple expansion thoracoplasty in children with thoracic insufficiency syndrome: A longitudinal study. Spine. Feb 1 2006; 31: 284-290.
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16. 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):1335-1341.

## Policy History:

Medical Policy Group, January 2007 **(2)**
Medical Policy Administration Committee, February 2007
Available for comment February 10-March 26, 2007
Medical Policy Group, January 2009 **(1)**
Medical Policy Group, June 2011; Updated Key Points and References
Medical Policy Group, October 2011 **(1)** Update to Key Points and References; no change in policy statement.
Medical Policy Panel, May 2012
Medical Policy Group, August 2012 **(2)**: Policy updated with literature search through March 2012; policy statement on use of vertical expandable prosthetic titanium rib for patients without thoracic insufficiency is investigational. Key Points and References updated to support policy statement.
Medical Policy Administration Committee, September 2012
Available for comments September 18 through November 1, 2012
Medical Policy Group, June 2013 **(2)**: 2013 Updates to Description, Key Points, and References
Medical Policy Panel, May 2014
Medical Policy Group, May 2014 (**4):** Updated Key Points and References. No changes to the policy at this time.
Medical Policy Panel, May 2015
Medical Policy Group, June 2015 **(2)**: 2015 Updates to Key Points, Approved by Governing Bodies, and References; no change to policy statement.
Medical Policy Group, June 2015 **(2)**: updated policy statement to include "progressive thoracic insufficiency syndrome due to rib and/or chest wall defects;" no change to intent of policy statement.

Medical Policy Group, January 2016 **(2)**: added code 22899 to coding section.
Medical Policy Panel, November 2016
Medical Policy Group, November 2016 **(7)**: 2016 Updated Key Points and References. No change in Policy Statement.
Medical Policy Panel, August 2017
Medical Policy Group, September 2017 **(7)**: 2017 Update - No new literature to review or add. No change in Policy Statement.
Medical Policy Panel, April 2018
Medical Policy Group, April 2018 **(7)**: 2018 Update to Key Points. No new literature to review or add. No change in Policy Statement.

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