



BlueCross BlueShield
of Alabama

Name of Policy:

Vertical Expandable Prosthetic Titanium Rib

Policy #: 299
Category: Surgery

Latest Review Date: June 2013
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 (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.

The vertical expandable prosthetic titanium rib (VEPTR, Synthes Spine Co) is a curved rod placed ~~horizontally~~ 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 four to six months as growth occurs and also to be replaced if necessary. Some patients require multiple devices.

A VEPTR has received FDA approval under a humanitarian device exemption (HDE). The data submitted to the FDA as part of the approval process consisted of a case series of 147 children ranging from ages six months to 15 years. The study showed the device was safe and showed probable benefit by enabling some patients to breathe unassisted. The labeled indications state that the device is not intended to correct conditions other than chest wall instability. It also should not be used before six months of age or beyond the age of skeletal maturity (14 years for girls, 16 for boys).

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 infants/children with thoracic insufficiency 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 members' 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:

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 seven study sites. 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 five-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. 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 four 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 two 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 six months to skeletal maturity; progressive TIS; more than 10% reduction in height of the concave hemithorax; and three or more anomalous vertebrae, with three 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 children who could complete pulmonary function tests (PFTs). Preoperatively, one patient required continuous positive airway pressure (CPAP), and one needed supplemental oxygen for ventilatory support at final follow-up.

Emans et al from Children's Hospital, Boston, report results on 31 patients with Thoracic Insufficiency Syndrome. Thirty-one patients with fused ribs and TIS were treated; four patients had prior spinal arthrodesis with continued progression of deformity. In 30 patients, the spinal deformity was controlled and growth continued in the thoracic spine during treatment at rates similar to normals. Increased volume of the constricted hemithorax obtained during expansion thoracostomy was maintained at follow-up. Complications included device migration (n=8), deep infection (n=2), and brachial plexus injury/thoracic outlet syndrome (n=2).

Another paper reported on European experience. In this case series, 15 patients (mean age 6 years, age range 11 months to 12 years) were treated from 2002 to 2005 for scoliosis and thoracic insufficiency. In all cases, patients and families reported improvement. Three complications were reported: skin breakage, lumbar hook displacement, and rib fracture.

Motoyama et al from Children's Hospital in Pittsburgh (PA) reported on follow-up of 10 patients with thoracic insufficiency with follow-up as long as 33 months. At baseline, FVC showed a moderate-to-severe decrease (69% of predicted values), indicating the presence of significant restrictive lung defect. FVC increased significantly over time, with an average rate of 26.8% per year, the rate of increase 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, indicating that in most children studied, lung growth kept up with body growth.

January 2009 Update

No articles were identified that would lead to a change in the policy statement. One additional series of 22 patients from another Children's Hospital has been published. Waldhausen et al (2007) published a retrospective chart review of 22 children who had 36 vertical expandable prosthetic titanium rib (VEPTR) devices placed. Most patients had carbon dioxide retention, pulmonary restrictive disease, or respiratory failure. Seven VEPTR devices required revision for erosion through the bone or dislodgement and three were removed. Five were outgrown and removed or replaced. One eroded soft tissue causing superficial infection that resolved with operative revision. Postoperative ventilation/perfusion scans improved most in younger children.

2010 – 2011 Update

No articles were identified that would lead to a change in the policy statement. Skaggs et al (2009) reported on a retrospective review that evaluated the nutritional status of children with TIS to determine if treatment with VEPTR leads to improvements in weight percentile. There were 76 patients (mean age 3.7 years, range 8 months – 14 years) who had placement of a VEPTR device. The average follow-up was 3.3 years (range 2 – 6 years). The results showed a significant increase in weight percentiles after VEPTR surgery. Of 76 patients, 60 (79%) were < or + 5% for weight before surgery. After surgery, 24 of 60 (40%) had an increased weight percentile. The authors concluded there was a significant improvement in nutritional status of children after VEPTR surgery.

Mayer et al (2009) reported on postoperative change in lung function after VEPTR insertion. They reported that although there is a clinically and radiographically apparent expansion of the thorax after VEPTR insertion, there is no similar improvement in lung volume. There was a decrease in forced vital capacity, an increase in residual volume, and no change in total lung

capacity at the first postoperative visit (7.7 ± 4.8 months). They noted that this needs further study.

October 2011 Update

Thoracic Insufficiency Syndrome

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 three 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 CT scans were similar when corrected for age. Pulmonary function tests were performed every six 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 one second (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, four for chest tube placement (pneumothorax), one for seroma drainage, six for hardware removal (for infection) and three for hardware repositioning (for dislodgement). Another 22 complications were treated nonoperatively.

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. 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 three rod fractures in two patients, three superficial infections, and one case of prominent hardware that threatened skin integrity. The authors concluded that 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.”

September 2012 Update

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. 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 four patients (15%) in the study published by Campbell.

May 2013 Update

There are no new articles in the peer-reviewed literature on this topic. The policy statement remains as stated.

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.

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 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
- Ellis van Creveld syndrome

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

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

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: FEP does not consider investigational if FDA approved. Will be reviewed for medical necessity.

Pre-certification requirements: Not applicable

Coding:

CPT Codes: **There is no specific CPT code for this procedure.**

References:

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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

This medical policy is not an authorization, certification, explanation of benefits, or a contract. Eligibility and benefits are determined on a case-by-case basis according to the terms of the member's plan in effect as of the date services are rendered. All medical policies are based on (i) research of current medical literature and (ii) review of common medical practices in the treatment and diagnosis of disease as of the date hereof. Physicians and other providers are solely responsible for all aspects of medical care and treatment, including the type, quality, and levels of care and treatment.

This policy is intended to be used for adjudication of claims (including pre-admission certification, pre-determinations, and pre-procedure review) in Blue Cross and Blue Shield's administration of plan contracts.