Policy Statement

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

Use of the VEPTR® for all other conditions, including but not limited to the treatment of scoliosis in patients without thoracic insufficiency, is considered investigational.

Policy Guidelines

Due to complexity of thoracoplasty and the young age of the patient population undergoing such a procedure, implantation of the vertical expandable prosthetic titanium rib (VEPTR®) should be performed in specialized centers. Preoperative evaluation should require input from a pediatric orthopedist, a pulmonologist, and a thoracic surgeon. In addition, preoperative evaluation should require (when possible) a test for positive nutritional, cardiac, and pulmonary function.

Coding

There is no specific CPT code for this procedure. The procedure would most likely be reported with the following code:

- 22899: Unlisted procedure, spine

Description

The vertical expandable prosthetic titanium rib (VEPTR®) is a curved rod placed vertically in the chest to help shape the thoracic cavity. It is being evaluated in skeletally immature patients with thoracic insufficiency syndrome (TIS) to support thorax and lung development and in pediatric patients with scoliosis without TIS to slow or correct curve progression.

Related Policies

- Interventions for Progressive Scoliosis

Benefit Application

Benefit determinations should be based in all cases on the applicable contract language. To the extent there are any conflicts between these guidelines and the contract language, the contract language will control. Please refer to the member’s contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

Some state or federal mandates (e.g., Federal Employee Program (FEP)) prohibits plans from denying Food and Drug Administration (FDA)-approved technologies as investigational. In these instances, plans may have to consider the coverage eligibility of the FDA-approved technologies on the basis of medical necessity alone.

Regulatory Status

The VEPTR® (Depuy Synthes Spine, Raynham, MA) was initially cleared for marketing by the U.S. Food and Drug Administration (FDA) through a humanitarian device exemption for the
treatment of TIS in skeletally immature patients. In 2014, the VEPTR® was cleared for marketing by the FDA through the 510(k) process. The VEPTR® and VEPTR II™ devices are 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.

To identify potential TIS patients, the following categories are used:

- Flail chest syndrome
- Rib fusion and scoliosis
- Hypoplastic thorax syndrome, including
  - Jeune syndrome
  - Achondroplasia
  - Jarcho-Levin syndrome
  - Ellis–van Creveld syndrome.

FDA product code: MDI.

Rationale

Background

Thoracic Insufficiency Syndrome

Thoracic insufficiency syndrome (TIS) is the inability of the thorax to support normal respiration or lung growth. The condition results from serious defects affecting the ribs or chest wall (e.g., severe scoliosis with rib absence or rib fusion) and various hypoplastic thorax syndromes (e.g., Jeune syndrome, Jarcho-Levin syndrome). Spine, chest, and lung growth are interdependent.3

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.

Treatment

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

The vertical expandable prosthetic titanium rib (VEPTR) device 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. The VEPTR may be described as “rib-based” growth-sparing instrumentation, which is compared with “spine-based” growing rods for Cobb angle correction. The VEPTR device is designed to be expanded every 4 to 6 months as growth occurs and to be replaced if necessary. Some patients require multiple devices.

Literature Review

Thoracic insufficiency occurs in a limited patient population, and the literature on the use of the vertical expandable prosthetic titanium rib (VEPTR) consists mostly 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) on thoracic insufficiency syndrome (TIS) include an initial feasibility study involving 33 patients and a subsequent prospective study of 224 patients (214 with baseline data) at 7 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 TIS. Three- and 5-year follow-up rates for the multicenter study were approximately 95%. Of the 247 patients enrolled in either study, 12 (4.8%) patients died, and 2 withdrew. None of the deaths, as determined by investigators, were related to the VEPTR. 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 (unassisted breathing on room air) to 4 (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 dependent on a ventilator.

Campbell (2004), who developed the VEPTR, and colleagues reported on 27 patients who had surgery for TIS and at least 2 years of follow-up data; this series was based on 41 patients treated between 1990 and the study reporting. Entry criteria for this study were acceptance by pediatric general surgeon, pediatric pulmonologist, and a 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 for an average of 3.2 years (range, 2-12 years). Before surgery, the mean annual rate of progression was 15° per year (range, 2°-50° per year). Following surgery, the Cobb angle (of scoliosis) improved from 74° to a final value of 49°. Spine growth was at a 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. Preoperatively, 1 patient required continuous positive airway pressure, and one needed supplemental oxygen for ventilatory support at final follow-up. Another publication (2013) 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 years). With a mean of 5 expansion surgeries per patient (range, 1-10), the Cobb angle improved by a mean of 8.9° and thoracic height improved by a mean of 3.41 cm. Eight (33%) patients had a total of 16 adverse events, all of which required surgery.

In another series, Gadepalli et al (2011) examined growth and pulmonary function in 26 children who received a VEPTR between 2006 and 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 (54.6 L vs 51.8 L), FVC (58.1 L vs 55.9 L), or residual volume (145.3 L vs 105.6 L), 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 (2005) reported results on patients with TIS who underwent the procedure at a single children’s hospital from 1999 to 2005. 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, 0.5-5.4 years). A 3-member team selected patients for surgery, and cardiac function was evaluated preoperatively. Lengthening of the VEPTR 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, 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, final FVC was 73.5% of predicted levels. Prior to the procedure, 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 the operated lung increased from 157 cm³ preoperatively to 326 cm³ at the final follow-up visit.

Motoyama et al (2006) from a children’s hospital reported on 10 patients with TIS. Using a special portable pulmonary function test device, they reported on lung function in 10 children who had a VEPTR. Median age was 4.3 years (range, 1.8-9.8 years) at first test, and patients were followed 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 pace with body growth.

A series of 22 patients from another children’s hospital was published in 2007. Seven (19%) of the 36 the VEPTR units placed required revision and 10 of 22 children reported better activity levels while 2 of 22 children reported better respiratory function.

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

Scoliosis without Thoracic Insufficiency Syndrome
In 2011, White et al reported on 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 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 (43%) of 14 patients and included 3 rod fractures in 2 patients, 3 superficial infections, and 1 case of prominent hardware that threatened skin integrity. As noted by authors, while results were 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 the 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) and 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 at the second and third visits and higher function scores at the third and fourth visits. Interpretation of this study is limited due to confounding factors, including age at treatment, unknown comorbidities, and the rationale for treatment selection.

Adverse Events
Complications that occur with VEPTR need to be considered by practitioners and families when 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 4 (15%) patients in the study by Campbell. In a single-center series (2016) reporting on complications for 65 patients treated for TIS over a 13-year period, device-related complications occurred in 22 patients.

Summary of Evidence
For individuals who have progressive TIS due to rib and/or chest wall defects in childhood who receive VEPTR thoracoplasty, the evidence includes a few case series. Relevant outcomes are symptoms, morbid events, functional outcomes, and treatment-related mortality and morbidity.
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 case series reported at 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 (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 difficult to track in patients suffering with TIS, 1 study has demonstrated an age-specific increase in forced vital capacity; further still, that same study reported a final forced vital capacity 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. 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 a few case series. Relevant outcomes are symptoms, morbid events, functional outcomes, and treatment-related mortality and morbidity. The VEPTR is being evaluated for curves greater than 45° in infants and juveniles without thoracic insufficiency. Similar to TIS, very limited data are available on the use of the VEPTR for early-onset scoliosis without thoracic insufficiency; additionally, little is known about the disease progression of early-onset scoliosis, and therefore little is known regarding the risk-benefit tradeoff of the VEPTR surgery. The evidence is insufficient to determine the effects of the technology on health outcomes.

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

U.S. Preventive Services Task Force Recommendations
Not applicable.

Medicare National Coverage
There is no national coverage determination. In the absence of a national coverage determination, coverage decisions are left to the discretion of local Medicare carriers.

Ongoing and Unpublished Clinical Trials
Some currently unpublished trials that might influence this review are listed in Table 1.

Table 1. Summary of Key Trials

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<th>Trial Name</th>
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NCT: National Clinical Trial.

References


Documentation for Clinical Review

Please provide the following documentation (if/when requested):

- History and physical and/or consultation notes including:
  - Clinical findings (i.e., pertinent symptoms and duration)
  - Comorbidities
  - Activity and functional limitations
  - Family history if applicable
  - Reason for procedure/test/device, when applicable
  - Pertinent past procedural and surgical history
  - Past and present diagnostic testing and results
  - Prior conservative treatments, duration, and response
  - Treatment plan (i.e., surgical intervention)
- Consultation and medical clearance report(s), when applicable
- Radiology report(s) and interpretation (i.e., MRI, CT, discogram)
• Laboratory results
• Other pertinent multidisciplinary notes/reports: (e.g., psychological or psychiatric evaluation, physical therapy, multidisciplinary pain management) when applicable

Post Service
• Results/reports of tests performed
• Procedure report(s)

Coding

This Policy relates only to the services or supplies described herein. Benefits may vary according to product design; therefore, contract language should be reviewed before applying the terms of the Policy. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement.

MN/IE
The following services may be considered medically necessary in certain instances and investigational in others. Services may be considered medically necessary when policy criteria are met. Services may be considered investigational when the policy criteria are not met or when the code describes application of a product in the position statement that is investigational.

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<th>Type</th>
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<td>0PR14JZ</td>
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<td>0PR24JZ</td>
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Policy History

This section provides a chronological history of the activities, updates and changes that have occurred with this Medical Policy.

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Definitions of Decision Determinations

Medically Necessary: A treatment, procedure, or drug is medically necessary only when it has been established as safe and effective for the particular symptoms or diagnosis, is not investigational or experimental, is not being provided primarily for the convenience of the patient or the provider, and is provided at the most appropriate level to treat the condition.
Investigational/Experimental: A treatment, procedure, or drug is investigational when it has not been recognized as safe and effective for use in treating the particular condition in accordance with generally accepted professional medical standards. This includes services where approval by the federal or state governmental is required prior to use, but has not yet been granted.

Split Evaluation: Blue Shield of California/Blue Shield of California Life & Health Insurance Company (Blue Shield) policy review can result in a split evaluation, where a treatment, procedure, or drug will be considered to be investigational for certain indications or conditions, but will be deemed safe and effective for other indications or conditions, and therefore potentially medically necessary in those instances.

Prior Authorization Requirements (as applicable to your plan)

Within five days before the actual date of service, the provider must confirm with Blue Shield that the member’s health plan coverage is still in effect. Blue Shield reserves the right to revoke an authorization prior to services being rendered based on cancellation of the member's eligibility. Final determination of benefits will be made after review of the claim for limitations or exclusions.

Questions regarding the applicability of this policy should be directed to the Prior Authorization Department. Please call (800) 541-6652 or visit the provider portal at www.blueshieldca.com/provider.

Disclaimer: This medical policy is a guide in evaluating the medical necessity of a particular service or treatment. Blue Shield of California may consider published peer-reviewed scientific literature, national guidelines, and local standards of practice in developing its medical policy. Federal and state law, as well as contract language, including definitions and specific contract provisions/exclusions, take precedence over medical policy and must be considered first in determining covered services. Member contracts may differ in their benefits. Blue Shield reserves the right to review and update policies as appropriate.