7.01.110	Vertical Expandable Prosthetic Titanium Rib		
Original Policy Date:	February 1, 2016	**Effective Date:**	June 1, 2023
Section:	7.0 Surgery	**Page:**	Page 1 of 12

Policy Statement

I. 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 and children between 6 months of age and skeletal maturity.

II. Use of the vertical expandable prosthetic titanium rib for all other conditions, including but not limited to the treatment of scoliosis in individuals without thoracic insufficiency, is considered **investigational**.

NOTE: Refer to Appendix A to see the policy statement changes (if any) from the previous version.

Policy Guidelines

Due to complexity of thoracoplasty and the young age of the individuals undergoing such a procedure, implantation of the vertical expandable prosthetic titanium rib 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 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 through a humanitarian device exemption for the treatment of TIS in skeletally immature patients.[3,] In 2014, the VEPTR® was cleared for marketing by the Food and Drug Administration 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

Food and Drug Administration 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.[1,] 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.[2,] 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

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 1 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 (RCT) 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.

Promotion of greater diversity and inclusion in clinical research of historically marginalized groups (e.g., People of Color [African-American, Asian, Black, Latino and Native American]; LGBTQIA (Lesbian, Gay, Bisexual, Transgender, Queer, Intersex, Asexual); Women; and People with Disabilities [Physical and Invisible]) allows policy populations to be more reflective of and findings more applicable to our diverse members. While we also strive to use inclusive language related to these groups in our policies, use of gender-specific nouns (e.g., women, men, sisters, etc.) will continue when reflective of language used in publications describing study populations.

Progressive Thoracic Insufficiency Syndrome

Clinical Context and Therapy Purpose

Thoracic insufficiency syndrome is the inability of the thorax to support normal respiration or lung growth.[2,]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 thoracic insufficiency syndrome 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.

The purpose of the vertical expandable prosthetic titanium rib in individuals who have progressive thoracic insufficiency syndrome is to provide a treatment option that is an alternative to or an improvement on existing therapies.

The following PICO was used to select literature to inform this review.

Populations

The relevant population of interest is children who have progressive thoracic insufficiency syndrome.

Interventions

The therapy being considered is the vertical expandable prosthetic titanium rib. The vertical expandable prosthetic titanium rib 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 vertical expandable prosthetic titanium rib 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.

Comparators
Relevant comparators include respiratory supportive care.

Outcomes
The general outcomes of interest are symptoms, morbid events, functional outcomes, treatment-related mortality, and treatment-related morbidity. Based on existing literature, follow-up of 2 to 5 years is recommended.

Study Selection Criteria
Methodologically credible studies were selected using the following principles:

1. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs.
2. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.
3. To assess long-term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.
4. Studies with duplicative or overlapping populations were excluded.

Review of Evidence
Uncontrolled Studies
Thoracic insufficiency occurs in a limited patient population, and the literature on the use of the vertical expandable prosthetic titanium rib consists mostly of case series from single institutions (some series are from specialized pediatric centers); no comparative trials have been identified. Data submitted to the U.S. Food and Drug Administration (FDA) on thoracic insufficiency syndrome include an initial feasibility study involving 33 patients and a subsequent prospective study of 224 patients (214 with baseline data) at 7 study sites.[1,] Of these, 94 patients 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. 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 vertical expandable prosthetic titanium rib. Because standard pulmonary function testing was not possible for most of this population, an assisted ventilatory rating was used to assess impact on respiratory status. The assisted ventilatory rating ranged from 0 (unassisted breathing on room air) to 4 (full-time ventilatory support). In the multicenter prospective study, the assisted ventilatory rating 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 vertical expandable prosthetic titanium rib, and colleagues reported on 27 patients who had surgery for thoracic insufficiency syndrome and at least 2 years of follow-up data. This series was based on 41 patients treated between 1990 and the study reporting.[4,] Entry criteria for the study were acceptance by a pediatric general surgeon, pediatric pulmonologist, and pediatric orthopedist; age 6 months to skeletal maturity; progressive thoracic insufficiency syndrome; 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). Before surgery, the mean annual rate of progression was 15° per year (range, 2°-50°). 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 another needed supplemental oxygen for ventilatory support at final follow-up.

Flynn et al (2013) reported an average 40.7 month follow-up (range, 25-78) in 24 children with nonsyndromic congenital scoliosis.[5,] 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 mean Cobb angle improved by 8.9° and mean thoracic height improved by 3.41 cm. Eight (33%) patients had a total of 16 adverse events, all of which required surgery. Gadepalli et al (2011) examined growth and pulmonary function in 26 children who received a vertical expandable prosthetic titanium rib between 2006 and 2010.[6,] In this case series, the children underwent 29 insertions and 57 expansions, with an average of 3 surgeries per child. Each procedure required an average of 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 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). 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 for patients with thoracic insufficiency syndrome who underwent the procedure at a single children's hospital from 1999 to 2005.[7,] Thirty-one patients with fused ribs and thoracic insufficiency syndrome 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). A 3-member team selected patients for surgery, and cardiac function was evaluated preoperatively. Lengthening of the vertical expandable prosthetic titanium rib 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 computed tomography scan) in the operated lung increased from 157 cm^3 preoperatively to 326 cm^3 at the final follow-up visit.

Motoyama et al (2006) reported on 10 patients with thoracic insufficiency syndrome.[8,] Using a special portable pulmonary function test device, they evaluated lung function in 10 children who had a vertical expandable prosthetic titanium rib. The median age was 4.3 years (range, 1.8-9.8) at first test, and patients were followed for an average of 22 months (range, 7-33). At baseline, FVC showed moderate-to-severe decrease (69% of predicted), indicating the presence of significant restrictive lung defect. Forced vital capacity 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.

Waldhausen at al (2007) published a series of 22 patients.[9,] Seven (19%) of the 36 vertical expandable prosthetic titanium rib 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 vertical expandable prosthetic titanium rib in thoracic insufficiency syndrome[10,] or early changes in pulmonary function.[11,]

Section Summary: Progressive Thoracic Insufficiency Syndrome

The evidence evaluating use of the vertical expandable prosthetic titanium rib thoracoplasty to treat children with progressive thoracic insufficiency syndrome due to rib and/or chest wall defects consists of case series. Results from the case series reported by different specialty centers have

demonstrated improvement and/or stabilization in key measures with use of the vertical expandable prosthetic titanium rib in progressive thoracic insufficiency syndrome. 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 thoracic insufficiency syndrome, 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 vertical expandable prosthetic titanium rib technology.

Early-onset Scoliosis Without Thoracic Insufficiency Syndrome

Clinical Context and Therapy Purpose

The purpose of the vertical expandable prosthetic titanium rib in individuals who have early-onset scoliosis without thoracic insufficiency syndrome is to provide a treatment option that is an alternative to or an improvement on existing therapies.

The following PICO was used to select literature to inform this review.

Populations

The relevant population of interest is young children with early-onset scoliosis without thoracic insufficiency syndrome.

Interventions

The therapy being considered is the vertical expandable prosthetic titanium rib. The vertical expandable prosthetic titanium rib 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 vertical expandable prosthetic titanium rib 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.

Comparators

Relevant comparators include spinal fusion and bracing.

Outcomes

The general outcomes of interest are symptoms, morbid events, functional outcomes, treatment-related mortality, and treatment-related morbidity. Based on the limited literature available on the vertical expandable prosthetic titanium rib for early-onset scoliosis without thoracic insufficiency syndrome, follow-up of at least 4 years is recommended.

Study Selection Criteria

Methodologically credible studies were selected using the following principles:

1. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs.
2. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.
3. To assess long-term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.
4. Studies with duplicative or overlapping populations were excluded.

Review of Evidence

Non-randomized Controlled Studies

Farley et al (2014) used data from a prospective registry to compare treatment of congenital scoliosis using the vertical expandable prosthetic titanium rib (n=22) with treatment using spinal fusion (n=27)

and observation (n=184).[12,] Function, pain, and mental health status were measured with the 22-item Scoliosis Research Society questionnaire. Compared with the observation group, the vertical expandable prosthetic titanium rib 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.

Uncontrolled Studies

An uncontrolled cohort study conducted by El-Hawary et al (2017) enrolled 63 children (mean age, 6.1 years) with early-onset scoliosis measuring more than 45 degrees (mean, 72 degrees) and no rib abnormalities or thoracic dysplasia.[13,] Outcomes of interest were change in major and secondary scoliosis curves and spinal growth, based on change in coronal spine height and sagittal spine length. After 2.2 years follow-up, the mean major scoliosis curve was reduced from 72 to 57 degrees (p<.0001), while the secondary scoliosis curve was reduced from 42.8 to 39.6 degrees (p=.009). Results were similar for the change from baseline in coronal spine height (p<.0001) and sagittal spine length (p<.0001). Seventy-nine percent (42 of 65) of patients were deemed to have treatment success, based on a composite outcome that included controlling the major scoliosis curve and improving the coronal spine height. Nearly half of the patients (49%; 31 of 65) had an adverse event associated with vertical expandable prosthetic titanium rib surgery, including 15 instances of device migration.

Longer follow-up of the cohort was subsequently reported by El-Hawary et al (2020).[14,] Data were available for 59 patients (mean age, 6.1 years) at a mean follow-up of 6.9 years. At follow-up, the vertical expandable prosthetic titanium rib was in place in 24 patients. Among the other patients, 3 had the prosthetic rib removed, 11 converted to other devices, and 13 had undergone definitive fusion. Two patients had died and 6 were lost to follow-up. At final follow-up, the mean major scoliosis curve was 61 degrees (p<.001 vs. baseline), while secondary scoliosis curve regressed to nearly baseline (42 degrees; p=.54 vs. baseline). Coronal spine height (p<.001) and sagittal spine length (p<.001) remained significantly improved from baseline. Results were similar in a subset of 29 patients that had the vertical expandable prosthetic titanium rib in place for over 5 years. At 5-years follow-up, there were 24 instances of device migration and 1 occurrence of a device-related Grade 3 adverse event ; 2 deaths were deemed not treatment-related.

Case Series

A case series conducted by White et al (2011) reported on the off-label use of spine-to-spine vertical expandable prosthetic titanium rib to treat spinal deformity in 14 children without chest wall abnormalities.[15,] 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-vertical expandable prosthetic titanium rib 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 the 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."

Section Summary: Early-onset Scoliosis Without Thoracic Insufficiency Syndrome

The evidence evaluating use of the vertical expandable prosthetic titanium rib thoracoplasty to treat young children with early-onset scoliosis without thoracic insufficiency syndrome consists of a non-randomized controlled study, an uncontrolled cohort study, and a case series. The vertical expandable prosthetic titanium rib is being evaluated for curves greater than 45° in infants and juveniles without thoracic insufficiency. Similar to thoracic insufficiency syndrome, limited data are available on the use of the vertical expandable prosthetic titanium rib 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 trade-off of the vertical expandable prosthetic titanium rib surgery.

Adverse Events

Complications that occur with vertical expandable prosthetic titanium rib need to be considered by practitioners and families when discussing this procedure. The FDA review and the articles by Campbell et al (2004) and Emans et al (2005) have informed the summary on complications arising from vertical expandable prosthetic titanium rib.[1,4,7,] 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. Waldhausen et al (2016), in a single-center series, reported on device-related complications in 22 of 65 patients treated for thoracic insufficiency syndrome over a 13-year period.[16,]

Supplemental Information

The purpose of the following information is to provide reference material. Inclusion does not imply endorsement or alignment with the evidence review conclusions.

Practice Guidelines and Position Statements

Guidelines or position statements will be considered for inclusion in 'Supplemental Information' if they were issued by, or jointly by, a US professional society, an international society with US representation, or National Institute for Health and Care Excellence (NICE). Priority will be given to guidelines that are informed by a systematic review, include strength of evidence ratings, and include a description of management of conflict of interest.

No relevant 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 ongoing and unpublished trials that might influence this review are listed in Table 1.

Table 1. Summary of Key Trials

NCT No.	Trial Name	Planned Enrollment	Completion Date
Ongoing			
NCT01672749	Evaluation of a Growth Guiding Construct vs. Standard Dual Growing Rods and vertical expandable prosthetic titanium rib (VEPTR) for the Treatment of Early Onset Scoliosis Patients: A Prospective Multi-center Cohort Study With a Matched Historical Control	51	Apr 2027
Unpublished			
NCT02241954	Vertical Expandable Prosthetic Titanium Rib (VEPTR) for Thoracic Insufficiency Syndrome	7 (actual)	Feb 2020 (completed)

NCT: national clinical trial.

References

1. Food and Drug Administration. Vertical Expandable Prosthetic Titanium Rib (VEPTR). 2004; https://www.accessdata.fda.gov/cdrh_docs/pdf14/k142587.pdf. Accessed February 14, 2023.
2. Campbell RM, Smith MD, Mayes TC, et al. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. J Bone Joint Surg Am. Mar 2003; 85(3): 399-408. PMID 12637423
3. Campbell RM. VEPTR: past experience and the future of VEPTR principles. Eur Spine J. Mar 2013; 22 Suppl 2(Suppl 2): S106-17. PMID 23354777
4. Campbell RM, 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. Aug 2004; 86(8): 1659-74. PMID 15292413
5. Flynn JM, Emans JB, Smith JT, et al. VEPTR to treat nonsyndromic congenital scoliosis: a multicenter, mid-term follow-up study. J Pediatr Orthop. 2013; 33(7): 679-84. PMID 23812154
6. Gadepalli SK, Hirschl RB, Tsai WC, et al. Vertical expandable prosthetic titanium rib device insertion: does it improve pulmonary function?. J Pediatr Surg. Jan 2011; 46(1): 77-80. PMID 21238644
7. 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 expandable prosthetic titanium rib: growth of thoracic spine and improvement of lung volumes. Spine (Phila Pa 1976). Sep 01 2005; 30(17 Suppl): S58-68. PMID 16138067
8. 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). Feb 01 2006; 31(3): 284-90. PMID 16449900
9. 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. Jan 2007; 42(1): 76-80. PMID 17208544
10. 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). Nov 01 2009; 34(23): 2530-3. PMID 19927103
11. 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. PMID 19098643
12. Farley FA, Li Y, Jong N, et al. Congenital scoliosis SRS-22 outcomes in children treated with observation, surgery, and VEPTR. Spine (Phila Pa 1976). Oct 15 2014; 39(22): 1868-74. PMID 25099323
13. El-Hawary R, Kadhim M, Vitale M, et al. VEPTR Implantation to Treat Children With Early-Onset Scoliosis Without Rib Abnormalities: Early Results From a Prospective Multicenter Study. J Pediatr Orthop. Dec 2017; 37(8): e599-e605. PMID 28141685
14. El-Hawary R, Morash K, Kadhim M, et al. VEPTR Treatment of Early Onset Scoliosis in Children Without Rib Abnormalities: Long-term Results of a Prospective, Multicenter Study. J Pediatr Orthop. Jul 2020; 40(6): e406-e412. PMID 32501900
15. White KK, Song KM, Frost N, et al. VEPTR™ growing rods for early-onset neuromuscular scoliosis: feasible and effective. Clin Orthop Relat Res. May 2011; 469(5): 1335-41. PMID 21213088
16. Waldhausen JH, Redding G, White K, et al. Complications in using the vertical expandable prosthetic titanium rib (VEPTR) in children. J Pediatr Surg. Nov 2016; 51(11): 1747-1750. PMID 27397045

Documentation for Clinical Review

Please provide the following documentation:

- 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 /device,
  - 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,) as applicable
- Laboratory results if applicable
- Other pertinent multidisciplinary notes/reports: (e.g., psychological or psychiatric evaluation, physical therapy, multidisciplinary pain management) when applicable

Post Service (in addition to the above, please include the following):

- 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.

The following codes are included below for informational purposes. Inclusion or exclusion of a code(s) does not constitute or imply member coverage or provider reimbursement policy. Policy Statements are intended to provide member coverage information and may include the use of some codes for clarity. The Policy Guidelines section may also provide additional information for how to interpret the Policy Statements and to provide coding guidance in some cases.

Type	Code	Description
CPT®	22899	Unlisted procedure, spine
HCPCS	None	

Policy History

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

Effective Date	Action	Reason
02/01/2016	BCBSA Medical Policy Adoption	Medical Policy Committee
02/01/2017	Policy revision without position change	Medical Policy Committee
10/01/2017	Policy revision without position change	Medical Policy Committee
06/01/2018	Policy revision without position change	Medical Policy Committee
06/01/2019	Policy revision without position change	Medical Policy Committee
06/01/2023	Policy reactivated. Previously archived from 06/01/2020 to 05/31/2023.	Medical Policy Committee

Definitions of Decision Determinations

Medically Necessary: Services that are Medically Necessary include only those which have been established as safe and effective, are furnished under generally accepted professional standards to treat illness, injury or medical condition, and which, as determined by Blue Shield, are: (a) consistent with Blue Shield medical policy; (b) consistent with the symptoms or diagnosis; (c) not furnished primarily for the convenience of the patient, the attending Physician or other provider; (d) furnished at the most appropriate level which can be provided safely and effectively to the patient; and (e) 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 the Member's illness, injury, or disease.

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 and Feedback (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 at (800) 541-6652, or the Transplant Case Management Department at (800) 637-2066 ext. 3507708 or visit the provider portal at www.blueshieldca.com/provider.

We are interested in receiving feedback relative to developing, adopting, and reviewing criteria for medical policy. Any licensed practitioner who is contracted with Blue Shield of California or Blue Shield of California Promise Health Plan is welcome to provide comments, suggestions, or concerns. Our internal policy committees will receive and take your comments into consideration.

For utilization and medical policy feedback, please send comments to: MedPolicy@blueshieldca.com

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.

Appendix A

POLICY STATEMENT	
BEFORE	**AFTER** **Blue font: Verbiage Changes/Additions**
Reactivated Policy **Policy Statement:** N/A	**Vertical Expandable Prosthetic Titanium Rib 7.01.110** **Policy Statement:** I. 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 and children between 6 months of age and skeletal maturity. II. Use of the vertical expandable prosthetic titanium rib for all other conditions, including but not limited to the treatment of scoliosis in individuals without thoracic insufficiency, is considered **investigational**.