Medical Policy
Vertical Expandable Prosthetic Titanium Rib

Table of Contents
- Policy: Commercial
- Coding Information
- Information Pertaining to All Policies
- Policy: Medicare
- Description
- References
- Authorization Information
- Policy History

Policy Number: 305
New Policy Number: 7.01.110
NCD/LCD: N/A

Related Policies
- Orthotics for Progressive Scoliosis, #550

Policy
Commercial Members: Managed Care (HMO and POS), PPO, and Indemnity Medicare HMO BlueSM and Medicare PPO BlueSM Members

Vertical expandable prosthetic titanium rib in the treatment of progressive thoracic insufficiency syndrome due to rib and/or chest wall abnormalities in infants/children between 6 months of age and skeletal maturity (about age 14 for girls and age 16 for boys) may be considered MEDICALLY NECESSARY.

Notes:
- Implantation of this device should be performed in specialized centers, given the complexity of these procedures and patients.
- 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.

Vertical expandable prosthetic titanium rib for all other conditions, including but not limited to the treatment of scoliosis in patients without thoracic insufficiency, is considered INVESTIGATIONAL.

Prior Authorization Information
Inpatient
- For services described in this policy, precertification/preauthorization IS REQUIRED for all products if the procedure is performed inpatient.

Outpatient
- For services described in this policy, see below for products where prior authorization might be required if the procedure is performed outpatient.
CPT Codes / HCPCS Codes / ICD Codes
Inclusion or exclusion of a code does not constitute or imply member coverage or provider reimbursement. Please refer to the member’s contract benefits in effect at the time of service to determine coverage or non-coverage as it applies to an individual member.

Providers should report all services using the most up-to-date industry-standard procedure, revenue, and diagnosis codes, including modifiers where applicable.

CPT Codes
There is no specific CPT code for this service.

Description
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 (eg, severe scoliosis with rib absence or rib fusion) and various hypoplastic thorax syndromes (eg, Jeune syndrome, Jarcho-Levin syndrome). Spine, chest, and lung growth are interdependent.

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.

Summary
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.

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, a 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.

**Policy History**

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<tr>
<th>Date</th>
<th>Action</th>
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<tbody>
<tr>
<td>12/2016</td>
<td>New references added from BCBSA National medical policy.</td>
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<tr>
<td>7/2015</td>
<td>New references added from BCBSA National medical policy.</td>
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<tr>
<td>1/2014</td>
<td>Removed ICD-9 procedure code 78.51 as it does not meet the intent of the policy.</td>
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<tr>
<td>6/2013</td>
<td>New references from BCBSA National medical policy.</td>
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**Information Pertaining to All Blue Cross Blue Shield Medical Policies**

Click on any of the following terms to access the relevant information:

- Medical Policy Terms of Use
- Managed Care Guidelines
- Indemnity/PPO Guidelines
- Clinical Exception Process
- Medical Technology Assessment Guidelines

**References**