

## Medical Policy Manual

**Topic:** Vertical Expandable Prosthetic Titanium Rib

**Date of Origin:** June 2007

**Section:** Surgery

**Last Reviewed Date:** May 2014

**Policy No:** 159

**Effective Date:** August 1, 2014

### **IMPORTANT REMINDER**

Medical Policies are developed to provide guidance for members and providers regarding coverage in accordance with contract terms. Benefit determinations are based in all cases on the applicable contract language. To the extent there may be any conflict between the Medical Policy and contract language, the contract language takes precedence.

PLEASE NOTE: Contracts exclude from coverage, among other things, services or procedures that are considered investigational or cosmetic. Providers may bill members for services or procedures that are considered investigational or cosmetic. Providers are encouraged to inform members before rendering such services that the members are likely to be financially responsible for the cost of these services.

### **DESCRIPTION**

The vertical expandable prosthetic titanium rib (VEPTR) is a curved rod placed vertically in the chest that helps stabilize and shape the thoracic cavity. It is positioned either between the ribs or between the ribs and either the spine or pelvis. VEPTR may be described as a “rib-based” growth sparing instrumentation, which is compared with “spine-based” growing rods for Cobb angle correction. It is being evaluated for use in skeletally immature patients with thoracic insufficiency syndrome (TIS) and to slow or correct curve progression in pediatric scoliosis patients without TIS. 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.

TIS is the inability of the thorax to support normal respiration or lung growth.<sup>[1]</sup> It results from serious defects affecting the ribs or chest wall such as severe scoliosis, rib fusion (which may accompany scoliosis), and various hypoplastic thorax syndromes such as Jeune’s Syndrome and Jarcho-Levin syndrome. Spine, chest, and lung 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 the concave side occurs with worsening respiratory compromise. This

progression is often accompanied by a need for supplemental oxygen and can require mechanical ventilation. While spinal fusion is one approach to treatment, it may not be successful and may also limit growth (lengthening) of the spine.

Given the complexity of these procedures and patients, implantation of this device should be performed in specialized centers. Preoperative evaluation requires input from pediatric orthopaedists, pulmonologist, and thoracic surgeon. In addition, preoperative evaluation of nutritional, cardiac and pulmonary function (when possible) is required.

## Regulatory Status

The Vertical Expandable Prosthetic Titanium Rib (VEPTR) (DePuy Synthes) device received US Food and Drug Administration (FDA) approval under a humanitarian device exemption (HDE) for the treatment of TIS in skeletally immature patients.<sup>[2]</sup> The FDA review defined TIS as, “the inability of the thorax to support normal respiration or lung growth” and created the following categories to aid in the identification of potential TIS patients:

- Flail chest syndrome
- Rib fusion and scoliosis
- Hypoplastic thorax syndrome, including:
  - Jeune's syndrome
  - Achondroplasia
  - Jarcho-Levin syndrome
  - Ellis van Creveld syndrome

Because the FDA clearance of the VEPTR device is specific to pediatric patients with TIS, use of the device in patients with scoliosis, but without TIS, is an off-label indication. In addition, the FDA review noted that the device should not be used in patients less than 6 months of age. Skeletal maturity occurs at about age 14 for girls and age 16 for boys.

## MEDICAL POLICY CRITERIA

- I. Use of the Vertical Expandable Prosthetic Titanium Rib may be considered **medically necessary** in the treatment of progressive thoracic insufficiency syndrome due to rib and/or chest wall defects in infants/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 patients without thoracic insufficiency, is considered **investigational**.

## SCIENTIFIC EVIDENCE

The most clinically relevant outcomes include the following:

- The ability of the device to restore and maintain a more normal anatomical shape while allowing for the continued growth of the child until skeletal maturity
- Durability of any beneficial treatment effects
- Safety, including the rate of adverse effects and reoperations

The following is a summary of the current published evidence for the use of VEPTR in the treatment of thoracic insufficiency syndrome (TIS) and scoliosis without TIS.

## **Thoracic Insufficiency Syndrome**

### Literature Appraisal

The current evidence for the use of VEPTR in the treatment of TIS is limited to data from case series and retrospective reviews.<sup>[2-10]</sup> Results from these case series have consistently demonstrated improvement and/or stabilization in key measures with the use of VEPTR in children with TIS, with or without congenital scoliosis. This improvement was noted in measures related to thoracic structure, growth of the thoracic spine and lung volumes, and stable or improved ventilatory status.

In general, conclusions based on data from small case series are uncertain, largely due to the lack of an appropriate comparison group. However, since TIS is a rare disease with limited treatment options, it is unlikely that data from large randomized controlled trials will become available. In addition, TIS is a progressive disease with a natural history of worsening deformity, pulmonary function, and pulmonary insufficiency that is unlikely to improve in the absence of intervention. Therefore, the available case series evidence, which consistently reports improved health outcomes, is considered sufficient to conclude that VEPTR may provide a treatment benefit in select patients.

## **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.<sup>[11]</sup> 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 to 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 3 rod fractures in 2 patients, 3 superficial infections, and 1 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.” Additional trials with more patients and longer duration are needed before conclusions about the safety and effectiveness of the VEPTR device in pediatric patients with spinal deformity, but without TIS, can be made.

## **Adverse Effects**

Complications associated with this device need to be considered by practitioners and families. Data from the FDA review and the papers by Campbell and Emans<sup>[2-4]</sup> reported a 25% device migration rate (although no significant long-term consequences were associated with this complication), a 10% rate of infection-related complications, and a 1% to 7% rate of brachial plexus injury or thoracic outlet

syndrome.

Other adverse effects that have been reported include:<sup>[6,11-15]</sup>

- Implant breakage
- Rib fractures
- Hip joint destabilization
- Perforation of the iliac ala
- Seroma
- Spontaneous ossification (e.g., lumbar spine, rib, iliac crest)
- Tissue reaction to metal
- Esophageal rupture

### **Clinical Practice Guidelines**

No evidence-based clinical practice guidelines were identified which specifically recommend the use of the VEPTR for treatment of TIS or scoliosis in the absence of TIS.

### **Summary**

Current evidence for use of vertical expandable prosthetic titanium rib (VEPTR) in thoracic insufficiency syndrome (TIS) is limited to results from small studies which consistently suggest improvement and/or stabilization in key health outcomes. Given the rare occurrence of TIS, the progressive natural history of the disease, and the limited treatment alternatives, the current evidence is considered sufficient to conclude that VEPTR may provide a treatment benefit in select patients. Therefore, use of the VEPTR device may be considered medically necessary in skeletally immature children at least 6 months of age with progressive thoracic insufficiency syndrome due to rib and/or chest wall defects.

Current evidence is insufficient to permit conclusions on the safety and effectiveness of the use of the vertical expandable prosthetic titanium rib (VEPTR) for the treatment of scoliosis without thoracic insufficiency syndrome. Nor are there any clinical practice guidelines that recommend VEPTR for this condition. Therefore, use of VEPTR for the treatment of scoliosis is considered investigational.

### **REFERENCES**

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## CROSS REFERENCES

None

CODES	NUMBER	DESCRIPTION
CPT	20999	Unlisted procedure, musculoskeletal system, general
	21899	Unlisted procedure, neck or thorax
	22899	Unlisted procedure, spine

HCPCS	None	
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