



CIGNA MEDICAL COVERAGE POLICY

The following Coverage Policy applies to all plans administered by CIGNA Companies including plans administered by Great-West Healthcare, which is now a part of CIGNA.

**Subject Titanium Rib Implants –
Vertical Expandable Prosthetic
Titanium Rib (VEPTR™)**

**Effective Date 12/15/2010
Next Review Date.....12/15/2011
Coverage Policy Number 0259**

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Coverage Policies are intended to provide guidance in interpreting certain **standard** CIGNA HealthCare benefit plans as well as benefit plans formerly administered by Great-West Healthcare. Please note, the terms of a participant's particular benefit plan document [Group Service Agreement (GSA), Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a participant's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a participant's benefit plan document **always supercedes** the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable group benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. Proprietary information of CIGNA. Copyright ©2010 CIGNA

Coverage Policy

CIGNA covers the implantation of the Vertical Expandable Prosthetic Titanium Rib (VEPTR™) device as medically necessary when used in accordance with the Humanitarian Device Exemption (HDE) specifications of the U.S. Food and Drug Administration (FDA) for children with thoracic insufficiency syndrome (TIS).

CIGNA does not cover the implantation of the VEPTR device for ANY other indication (e.g., scoliosis without TIS) because it is considered experimental, investigational or unproven.

General Background

Thoracic insufficiency syndrome (TIS) is defined as the inability of the thorax to support normal respiration and lung growth (Campbell, 2004). For children with TIS, treatment is aimed at stabilizing their clinical symptoms while preserving ongoing thoracic spinal growth. Treatment modalities include ventilation support, bracing, implantation of autograft or allograft rib sections, splitting of fused ribs, and insertion of spacers. TIS with progressive scoliosis may be treated by fusing the spine and inserting supportive instrumentation (Schneerson, 2003; Prakash, 2004).

While standard surgical interventions assist in providing spinal support via fusion and/or instrumentation, these interventions do not typically result in added expansion of the anterior chest wall. The Vertical Expandable Prosthetic Titanium Rib (VEPTR™) is an alternative to spinal fusion for children with TIS. Construct options include rib-to-rib, rib to ilium, or rib-to-lumber attachment. The latter two options include a lumbar extension rod. The device can be lengthened manually to indirectly support the curvature of a young child's spine while allowing the thoracic cavity to expand, thus decreasing restriction of the lungs by the rib cage and spine. During surgery when maximum expansion has been achieved, the VEPTR is locked into position. Depending on the type and severity of the spinal deformity, more than one device may be inserted. The device length is surgically adjusted approximately every four to six months as needed to accommodate new spinal growth. Complete replacement of the device, to allow for ongoing growth of the child's spine and to maintain maximum support of the chest cavity may be indicated. Once the child reaches skeletal maturity the device may remain in place or may be removed. The advantages of this surgical technique are its indirect treatment of the anatomical causes of TIS, while not interfering with subsequent spinal procedures that may be needed later in life (Synthes, 2009; Freeman, 2003).

U.S. Food and Drug Administration (FDA)

The Vertical Expandable Prosthetic Titanium Rib (VEPTR) (SYNTHES® Spine Co., West Chester, PA) was approved under the FDA Humanitarian Device Exemption (HDE) program in August 2004. The device is indicated for "the treatment of thoracic insufficiency syndrome (TIS) in skeletally immature patients. The FDA stated that for the purpose of identifying potential TIS patients, the categories in which TIS patients often fall include: flail chest syndrome; rib fusion and scoliosis; and hypoplastic thorax syndrome (e.g., Jeune's syndrome, achondroplasia, Jarcho-Levin syndrome and Ellis van Creveld syndrome) (FDA, 2004).

FDA approval was based on a 14-year multicenter study (n=224) conducted by SYNTHES Spine. The children, age six months or older, were divided into four groups by diagnosis, including flail chest, rib fusion, hypoplastic thoracic syndrome, and progressive scoliosis. Follow-up visits occurred at 4, 8, 12, 16, 20 and 24 months, and then annually thereafter. Twelve patients died and two withdrew, leaving 210 patients to complete the study. Of the remaining patients, 93.4% showed improved assisted ventilatory rating outcomes. Thoracic dimensions demonstrated that patients with flail chest improved 80%; those with rib fusion improved 91.5%; those with hypoplastic thoracic syndrome improved 84.8%; and those with progressive scoliosis improved 77.4%. The complications, experienced by 119 patients, included: device migration (16%), device failure (2%), spinal abscess (4%), and spinal or surgical site infection (5%) (FDA, 2004).

Specific contraindications to the use of the titanium rib include:

- insufficient bone strength in the ribs or spine where the device would attach
- absence of proximal and distal ribs for attachment of the device
- absence of diaphragmatic function
- insufficient tissue to cover the VEPTR
- age less than six months
- skeletal maturity has been reached (i.e., age 14 for girls and age 16 for boys)
- known allergy to any device materials
- infection near or at the surgical site

Literature Review

Titanium rib implants are an accepted treatment option for children with thoracic insufficiency syndrome. Evidence in the published peer-reviewed scientific literature in the form of prospective case series (Campbell, et al., Sep 2007; Emans, et al., 2005; Hell, et al., 2005; Campbell, et al., 2004) and retrospective reviews (Flynn, et al., 2010; Motoyama, et al., 2006; Gollogly, et al., 2004) have reported significant improvements in respiratory function, lung growth, and lung volume. Up to 12-year follow-ups have been reported. The number of studies comparing the titanium rib to other treatment modalities is limited.

Other Indications: VPTR has been proposed for the treatment of various idiopathic, neuromuscular, or syndromic spinal deformities in children without TIS, including early-onset scoliosis, also called idiopathic infantile scoliosis (Hasler, et al., 2010; Ramiz, et al., 2009; Smith, 2007). VEPTR is not FDA approved for the treatment of spinal deformities without thoracic insufficiency syndrome because its use has not clearly been shown to provide clinical benefit. Studies investigating the use of VEPTR for the treatment of spinal deformities

and scoliosis in children without TIS are primarily in the form of retrospective reviews (Hasler, et al., 2010; Samdani, et al., 2009; Smith, et al., 2009; Ramirez, et al., 2009) and case series (Redding, et al., 2008) with small heterogeneous patient populations (n=10-31) and a mean follow-up of 32 months or less.

Professional Societies/Organizations

Professional society opinion on this device is lacking. Neither the American Academy of Pediatrics, the Pediatric Orthopedic Society of North America, nor the American Academy of Orthopedic Surgeons has published guidelines on the titanium rib.

Summary

Vertical Expandable Prosthetic Titanium Rib (VEPTR) is an accepted treatment option for children with thoracic insufficiency syndrome (TIS). While the evidence is not robust, studies in the published peer-reviewed scientific literature have reported that the VEPTR has been successfully used as an alternative to ventilator dependence or spinal fusion. There is insufficient evidence to support VEPTR for any other condition, including scoliosis without TIS.

Coding/Billing Information

Note: This list of codes may not be all-inclusive.

Covered when medically necessary when used to report the implantation of the Vertical Expandable Prosthetic Titanium Rib (VEPTR) device:

CPT ^{®*} Codes	Description
20999	Unlisted procedure, musculoskeletal system, general
21899	Unlisted procedure, neck or thorax

ICD-9-CM Diagnosis Codes	Description
518.82	Other pulmonary insufficiency, not elsewhere classified
756.3	Other congenital anomaly of ribs and sternum
756.4	Chondrodystrophy,
756.55	Chondroectodermal dysplasia
786.09	Other dyspnea and respiratory abnormalities
807.4	Flail chest

*Current Procedural Terminology (CPT[®]) © 2010 American Medical Association: Chicago, IL.

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Policy History

Pre-Merger Organizations	Last Review Date	Policy Number	Title
CIGNA HealthCare	12/15/2007	0259	Titanium Rib Implants – Vertical Expandable Prosthetic Titanium Rib (VEPTR)
Great-West Healthcare	10/26/2006	04.261.02	Titanium Rib Implant

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