MEDICAL POLICY

POLICY TITLE	VERTICAL EXPANDABLE PROSTHETIC TITANIUM RIB
POLICY NUMBER	1.136

Original Issue Date (Created):	**3/1/2013**
Most Recent Review Date (Revised):	**8/16/2019**
Effective Date:	**10/1/2019**

I. POLICY

Use of the vertical expandable prosthetic titanium rib (VEPTR) is 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.

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** as there is insufficient evidence to support a conclusion concerning the health outcomes or benefits associated with this procedure.

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.

Cross-reference:

MP-1.120 - Interventions for Progressive Scoliosis

II. PRODUCT VARIATIONS

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This policy is only applicable to certain programs and products administered by Capital BlueCross and subject to benefit variations as discussed in Section VI. Please see additional information below.

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FEP PPO: Refer to FEP Medical Policy Manual MP-7.01.110, Vertical Expandable Prosthetic Titanium Rib. The FEP Medical Policy Manual can be found at: www.fepblue.org.

III. DESCRIPTION/BACKGROUND

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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, such as severe scoliosis with rib absence or rib fusion, and various hypoplastic thorax syndromes, such as Jeune syndrome and Jarcho-Levin syndrome. Spine, chest, and lung growth are interdependent. 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 also 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 also to be replaced if necessary. Some patients require multiple devices.

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. 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. For the purpose of identifying potential TIS patients, the categories in which TIS patients fall are as follows:

- Flail chest syndrome
- Rib fusion and scoliosis
- Hypoplastic thorax syndrome, including,

  o Jeune syndrome

  o Achondroplasia

  o Jarcho-Levin syndrome

  o Ellis-van Creveld syndrome

FDA product code: MDI.

IV. RATIONALE

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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 (eg, 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.

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V. DEFINITIONS TOP

NA

VI. BENEFIT VARIATIONS TOP

The existence of this medical policy does not mean that this service is a covered benefit under the member's health benefit plan. Benefit determinations should be based in all cases on the applicable health benefit plan language. Medical policies do not constitute a description of benefits. A member's health benefit plan governs which services are covered, which are excluded, which are subject to benefit limits and which require preauthorization. There are different benefit plan designs in each product administered by Capital BlueCross. Members and providers should consult the member's health benefit plan for information or contact Capital BlueCross for benefit information.

VII. DISCLAIMER TOP

Capital BlueCross's medical policies are developed to assist in administering a member's benefits, do not constitute medical advice and are subject to change. Treating providers are solely responsible for medical advice and treatment of members. Members should discuss any medical policy related to their coverage or condition with their provider and consult their benefit information to determine if the service is covered. If there is a discrepancy between this medical policy and a member's benefit information, the benefit information will govern. If a provider or a member has a question concerning the application of this medical policy to a specific member's plan of benefits, please contact Capital BlueCross' Provider Services or Member Services. Capital BlueCross considers the information contained in this medical policy to be proprietary and it may only be disseminated as permitted by law.

VIII. CODING INFORMATION TOP

Note: This list of codes may not be all-inclusive, and codes are subject to change at any time. The identification of a code in this section does not denote coverage as coverage is determined by the terms of member benefit information. In addition, not all covered services are eligible for separate reimbursement.

Covered when medically necessary:

CPT Codes®								
20999	21899	22899						

ICD-10-CM Diagnosis Codes	Description
J80	Acute respiratory distress syndrome
M41.00	Infantile idiopathic scoliosis, site unspecified

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ICD-10-CM Diagnosis Codes	Description
M41.02	Infantile idiopathic scoliosis, cervical region
M41.03	Infantile idiopathic scoliosis, cervicothoracic region
M41.04	Infantile idiopathic scoliosis, thoracic region
M41.05	Infantile idiopathic scoliosis, thoracolumbar region
M41.06	Infantile idiopathic scoliosis, lumbar region
M41.07	Infantile idiopathic scoliosis, lumbosacral region
M41.08	Infantile idiopathic scoliosis, sacral and sacrococcygeal region
M41.11	Juvenile idiopathic scoliosis, cervical region
M41.112	Juvenile idiopathic scoliosis, cervicothoracic region
M41.113	Juvenile idiopathic scoliosis, cervicothoracic region
M41.114	Juvenile idiopathic scoliosis, thoracic region
M41.115	Juvenile idiopathic scoliosis, thoracolumbar region
M41.116	Juvenile idiopathic scoliosis, lumbar region
M41.117	Juvenile idiopathic scoliosis, lumbosacral region
M41.119	Juvenile idiopathic scoliosis, site unspecified
M41.122	Adolescent idiopathic scoliosis, cervical region
M41.123	Adolescent idiopathic scoliosis, cervicothoracic region
M41.124	Adolescent idiopathic scoliosis, thoracic region
M41.125	Adolescent idiopathic scoliosis, thoracolumbar region
M41.126	Adolescent idiopathic scoliosis, lumbar region
M41.127	Adolescent idiopathic scoliosis, lumbosacral region
M41.129	Adolescent idiopathic scoliosis, site unspecified
M41.20	Other idiopathic scoliosis, site unspecified
M41.22	Other idiopathic scoliosis, cervical region
M41.23	Other idiopathic scoliosis, cervicothoracic region
M41.24	Other idiopathic scoliosis, thoracic region
M41.25	Other idiopathic scoliosis, thoracolumbar region
M41.26	Other idiopathic scoliosis, lumbar region
M41.27	Other idiopathic scoliosis, lumbosacral region
M41.30	Thoracogenic scoliosis, site unspecified
M41.34	Thoracogenic scoliosis, thoracic region
M41.35	Thoracogenic scoliosis, thoracolumbar region
M41.40	Neuromuscular scoliosis, site unspecified
M41.41	Neuromuscular scoliosis, occipito-atlanto-axial region
M41.42	Neuromuscular scoliosis, cervical region
M41.43	Neuromuscular scoliosis, cervicothoracic region
M41.44	Neuromuscular scoliosis, thoracic region
M41.45	Neuromuscular scoliosis, thoracolumbar region

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ICD-10-CM Diagnosis Codes	Description
M41.46	Neuromuscular scoliosis, lumbar region
M41.47	Neuromuscular scoliosis, lumbosacral region
M41.50	Other secondary scoliosis, site unspecified
M41.52	Other secondary scoliosis, cervical region
M41.53	Other secondary scoliosis, cervicothoracic region
M41.54	Other secondary scoliosis, thoracic region
M41.55	Other secondary scoliosis, thoracolumbar region
M41.56	Other secondary scoliosis, lumbar region
M41.57	Other secondary scoliosis, lumbosacral region
M41.80	Other forms of scoliosis, site unspecified
M41.82	Other forms of scoliosis, cervical region
M41.83	Other forms of scoliosis, cervicothoracic region
M41.84	Other forms of scoliosis, thoracic region
M41.85	Other forms of scoliosis, thoracolumbar region
M41.86	Other forms of scoliosis, lumbar region
M41.87	Other forms of scoliosis, lumbosacral region
M41.9	Scoliosis, unspecified
M41.9	Scoliosis, unspecified
M96.5	Postradiation scoliosis
Q67.5	Congenital scoliosis NOS
Q76.2	Congenital spondylolisthesis
Q76.3	Congenital scoliosis due to congenital bony malformation
Q76.425	Congenital lordosis, thoracolumbar region
Q76.49	Other congenital malformations of spine, not associated with scoliosis
Q76.6	Other congenital malformations of ribs (includes accessory rib, congenital absence of rib, congenital fusion of ribs, and congenital malformation of rib NOS)
Q76.7	Congenital malformation of sternum
Q76.8	Other congenital malformations of bony thorax
Q76.9	Congenital malformation of bony thorax, unspecified
Q77.2	Short rib syndrome
Q77.4	Achondroplasia
Q77.6	Chondroectodermal dysplasia
Q78.9	Osteochondrodysplasia, unspecified
Q79.8	Other congenital malformations of musculoskeletal system
Q79.9	Congenital malformation of musculoskeletal system, unspecified
Q87.2	Congenital malformation syndromes predominantly involving limbs (includes VATER syndrome)
S22.5XX	Flail chest

REFERENCES

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15. Blue Cross Blue Shield Association Medical Policy Reference Manual. 7.01.110, Vertical Expandable Prosthetic Titanium Rib. April 2019.

X. POLICY HISTORY

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MP 1.136	**CAC 10/28/12** New policy adopting BCBSA. Information regarding vertical expandable prosthetic titanium rib (VEPTR) was extracted from MP 1.120 Interventions for Progressive Scoliosis. Added additional information on VEPTR and placed in this new policy to match BCBSA. Remains medically necessary with criteria. Codes reviewed 9/20/12
	04/05/13- Policy recoded.
	CAC 11/26/13 Consensus review. References updated but no changes to the policy statements. Rationale added.
	CAC 11/25/14 Consensus review. No change to policy statements. References and rationale updated. Coding reviewed. 11/13/2014
	CAC 11/24/15 Consensus review. No changes to the policy statements. Rationale and reference update. Coding Reviewed
	CAC 11/29/16 Consensus review. No change to policy statements. Variation reformatting completed. FEP policy number corrected. Description/Background, Regulatory Status and Reference sections updated. Coding reviewed/updated.
	CAC 12/19/17 Consensus review. Description/Background, Rationale and Reference sections updated.
	7/6/18 Admin update: Coding reviewed and updated.
	11/2/18 Consensus review. Background and references updated. Rationale condensed.
	8/16/2019 Consensus Review. Policy statement unchanged. Policy Guidelines and Rationale updated. References updated.

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