

POLICY NUMBER	LAST REVIEW DATE	APPROVED BY
MG.MM.SU.67C6	03/10/2023	MPC (Medical Policy Committee)

IMPORTANT NOTE ABOUT THIS MEDICAL POLICY:

Property of ConnectiCare, Inc. All rights reserved. The treating physician or primary care provider must submit to ConnectiCare, Inc. the clinical evidence that the patient meets the criteria for the treatment or surgical procedure. Without this documentation and information, ConnectiCare will not be able to properly review the request for prior authorization. This clinical policy is not intended to pre-empt the judgment of the reviewing medical director or dictate to health care providers how to practice medicine. Health care providers are expected to exercise their medical judgment in rendering appropriate care. The clinical review criteria expressed below reflects how ConnectiCare determines whether certain services or supplies are medically necessary. ConnectiCare established the clinical review criteria based upon a review of currently available clinical information (including clinical outcome studies in the peer-reviewed published medical literature, regulatory status of the technology, evidence-based guidelines of public health and health research agencies, evidence-based guidelines and positions of leading national health professional organizations, views of physicians practicing in relevant clinical areas, and other relevant factors). ConnectiCare, Inc. expressly reserves the right to revise these conclusions as clinical information changes, and welcomes further relevant information. Identification of selected brand names of devices, tests and procedures in a medical coverage policy is for reference only and is not an endorsement of any one device, test or procedure over another. Each benefit plan defines which services are covered. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered and/or paid for by ConnectiCare, as some plans exclude coverage for services or supplies that ConnectiCare considers medically necessary. If there is a discrepancy between this guideline and a member's benefits plan, the benefits plan will govern. In addition, coverage may be mandated by applicable legal requirements of the State of CT and/or the Federal Government. Coverage may also differ for our Medicare members based on any applicable Centers for Medicare & Medicaid Services (CMS) coverage statements including including National Coverage Determinations (NCD), Local Coverage Determinations (LCD) and/or Local Medical Review Policies(LMRP). All coding and web site links are accurate at time of publication.

Definitions

Vertical expandable prosthetic titanium rib (VEPTR)	Curved rod placed horizontally in the chest to shape the thoracic cavity for the treatment of spinal and thoracic deformities. In 2014, the FDA Center for Devices and Radiological Health (CDRH) cleared the VEPTR®- VEPTR II™ device for use in skeletally immature patients with severe, progressive spinal deformities and/or three dimensional deformity of the thorax associated with, or at risk of, Thoracic Insufficiency Syndrome (TIS)">Thoracic Insufficiency Syndrome (TIS) .
Cobb angle	Measurement of the degree of spinal curvature; the Cobb angle is considered the standard measurement to quantify a scoliosis for the purpose of measuring curve progression over time. A curve is considered to be scoliosis at a Cobb angle of ≥ 10°. Any increase ≥ 5° is regarded as a significant change; indicative of curvature progression with scoliosis considered mild at 10°-24°, moderate at 25°-50° and severe at > 50° in skeletally mature individuals. Cobb angles > 45° are considered severe in skeletally immature persons.
Ellis-van Creveld syndrome	Autosomal recessive genetic disorder characterized by skeletal dysplasia.



Hypoplastic thorax syndrome	Examples of the syndrome include achondroplasia, Ellis van Creveld syndrome, Jarcho- Levin syndrome and Jeune's syndrome.
Jarcho-Levin syndrome	Heritable axial skeleton growth disorder associated with malformation of the vertebral column and ribs.
Jeune syndrome	Congenital dwarfism associated with asphyxiating thoracic dystrophy.
Scoliosis	Musculoskeletal condition characterized by an abnormal lateral curvature of the spine. There are several different types of scoliosis that affect children and adolescents. The most common type is considered idiopathic but additional types of scoliosis include congenital, neuromuscular and syndromic scoliosis.
Thoracic Insufficiency Syndrome (TIS)	Rare condition defined as, "The inability of the thorax to support normal respiration or lung growth. This would include patients with progressive congenital, neuromuscular, idiopathic, or syndromic scoliosis" (FDA, 2014). TIS may include flail chest syndrome, hypoplastic thorax syndrome, as well as rib fusion and scoliosis.

Related Medical Guideline

Surgical Correction of Chest Wall Deformities

Guideline

The VEPTR 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.

Rib/chest wall defects may be secondary to any of the following scoliosis conditions:

- 1. Congenital scoliosis
- 2. Neuromuscular scoliosis
- 3. Infantile and juvenile idiopathic scoliosis
- 4. Syndromic scoliosis

Exclusions and Limitations

- Use of VEPTR for any condition other than those listed above (including Poland Syndrome) is not considered medically necessary due to insufficient evidence of therapeutic value.
- 2. Use of VEPTR as a scoliosis treatment in the absence of TIS (or risk for TIS) is not considered medically necessary.



Applicable Procedure Codes

20999	Unlisted procedure, musculoskeletal system, general
21899	Unlisted procedure, neck or thorax

Applicable Diagnosis Codes

M41.00	Infantile idiopathic scoliosis, site unspecified
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
M41.40	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
M41.46	Neuromuscular scoliosis, lumbar region
M41.47	Neuromuscular scoliosis, lumbosacral region
Q67.5	Congenital scoliosis NOS
Q76.3	Congenital scoliosis due to congenital bony malformation

References

American Academy of Orthopaedic Surgeons. Idiopathic Scoliosis in Children and Adolescents. Your Orthopaedic Connection. March 17, 2022. http://orthoinfo.aaos.org/topic.cfm?topic=A00353. Accessed March 20, 2023.



Campbell RM Jr, Smith MD, Mayes TC, et al. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. J Bone Joint Surg Am. 2003; 85-A(3):399-408.

Campbell RM Jr, 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. 2004; 86-A(8):1659-1674.

Campbell RM Jr. VEPTR: past experience and the future of VEPTR principles. Eur Spine J. 2013; 22 Suppl 2:S106-S117.

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. 2005; 30(17 Suppl):S58-68.

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). 2014; 39(22):1868-1874.

Flynn JM, Emans JB, Smith JT, et al. VEPTR to treat nonsyndromic congenital scoliosis: a multicenter, midterm follow-up study. J Pediatr Orthop. 2013; 33(7):679-684.

Gadepalli SK, Hirschl RB, Tsai WC, et al. Vertical expandable prosthetic titanium rib device insertion: does it improve pulmonary function? J Pediatr Surg. 2011; 46(1):77-80.

Hell AK, Campbell RM, Hefti F. The vertical expandable prosthetic titanium rib implant for the treatment of thoracic insufficiency syndrome associated with congenital and neuromuscular scoliosis in young children. J Pediatr Orthop B. 2005; 14(4):287-293.

Karlin JG, Roth MK, Patil V, et al. Management of thoracic insufficiency syndrome in patients with Jarcho-Levin syndrome using VEPTRs (vertical expandable prosthetic titanium ribs). J Bone Joint Surg Am. 2014; 96(21):e181.

Maruyama T, Takeshita K. Surgical treatment of scoliosis: a review of techniques currently applied. Scoliosis. 2008; 3:6.

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. 2006; 31(3):284-290.

National Institute of Arthritis and Musculoskeletal and Skin Diseases. Questions and Answers about Scoliosis in Children and Adolescents. © 2017. https://www.niams.nih.gov/Health Info/Scoliosis/default.asp. Accessed March 20, 2023.

Ortop Traumatol Rehabil. 2007 Sep-Oct;9(5):459-66. The vertical expandable prosthetic titanium rib (VEPTR) in the treatment of scoliosis and thoracic deformities. Preliminary report. Latalski M1, Fatyga M, Gregosiewicz A.

Pediatr Radiol. 2015 Apr;45(4):606-16. doi: 10.1007/s00247-014-3128-4. Epub 2014 Sep 21. Vertical expandable prosthetic titanium rib (VEPTR): a review of indications, normal radiographic appearance and complications. Parnell SE1, Effmann EL, Song K, Swanson JO, Bompadre V, Phillips GS.

Scoliosis Research Society. Information about Thoracic Insufficiency Syndrome Treatment. https://www.srs.org/patients-- and-families/conditions-and-treatments/parents/scoliosis/thoracic-insufficiency-syndrome. 2019. Accessed March 20, 2023.

Specially matched clinical peer review.

Surg Technol Int. 2009 Apr; 18:223-9. Vertical expandable prosthetic titanium rib (VEPTR): indications,



technique, and management review. Shah SC1, Birknes JK, Sagoo S, Thome S, Samdani AF.

U.S. Food and Drug Administration. 510(k) Premarket Notification Database. Vertical Expandable Prosthetic Titanium Rib 510 (k) Advisory Committee Summary. November 2014. https://www.fda.gov/downloads/advisorycommittees/committeesmeetingmaterials/pediatricadvisorycommittee/ucm4-38974.pdf. Accessed March 20, 2023.

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. 2007; 42(1):76-80.

White KK, Song KM, Frost N, et al. VEPTR growing rods for early-onset neuromuscular scoliosis: feasible and effective. Clin Orthop Relat Res. 2011; 469(5):1335-1341.

Revision history

DATE	REVISION
02/2021	ConnectiCare, Inc. has adopted the clinical criteria of its parent
	corporation, EmblemHealth