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Operative Techniques in
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Operative Strategies for Thoracic Insufficiency Syndrome by Vertical Expandable Prosthetic Titanium Rib Expansion Thoracoplasty

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The 3-dimensional thoracic deformity responsible for thoracic insufficiency syndrome in children can be addressed by correcting the respective volume depletion deformity through vertical expandable prosthetic titanium rib (VEPTR) expansion thoracoplasty. These deformities include absent ribs, fused ribs and scoliosis, windswept deformity of the thorax in early onset scoliosis, and hypoplastic thorax. The core principle of this surgical technique is the early restoration of maximum volume and symmetry of the thorax with indirect correction of the scoliosis without the growth inhibition effects of spine surgery. Operative strategies of VEPTR expansion thoracoplasty include stabilization of flail chest, expansion of constricted chest walls, and control of spinal deformity. This approach, especially when performed in infancy, is postulated to provide stimulus for lung growth by making available additional thoracic volume for these patients. Most of these patients also have exotic scoliosis, a rare and possibly lethal subset of spinal deformity, which requires special care and treatment. Although VEPTR expansion thoracoplasty is a straightforward surgical technique, the preoperative selection of patients for VEPTR surgery is complex, and meticulous attention to postoperative management is critical for success. The long-term goal of VEPTR treatment is to have the largest and most functional thorax possible by skeletal maturity to provide optimal respiratory function for adulthood.

KEYWORDS thoracic insufficiency syndrome, expansion thoracoplasty, opening wedge thoracostomy, exotic scoliosis, VEPTR, secondary thoracic insufficiency syndrome, volume depletion deformity, lateral flexion contracture of the thorax

Vertical expandable prosthetic titanium rib (VEPTR) expansion thoracoplasty treats thoracic insufficiency syndrome, which is the inability of the thorax to support normal respiration or lung growth,[1] with the presence of either enabling the diagnosis of thoracic insufficiency syndrome (TIS). Normal respiration depends on the thorax having a normal, stable volume and an ability to change that volume.[1] The thorax is the respiratory pump,[2] providing lung expansion through a positive thoracic volume change by contraction of the diaphragm with the rib cage providing passive circumferential support for the lungs. Additional positive thoracic volume change is provided by secondary breathing[1] with intercostal muscle contraction elevating the ribs to provide anterior lateral expansion of the chest wall. Lung growth and thoracic growth are interdependant, so symmetrical enlargement by growth of the surrounding thorax is necessary in order for lung growth to proceed normally.

Exotic scoliosis[2] refers to those early-onset cases of spinal curvature that are much more complex in nature, commonly associated with a thorax distorted by curve rotation and lordosis, causing volume depletion deformity[2] and thoracic growth inhibition with adverse effect on lung growth. Additional thoracic disability is observed if there is primary rib cage disease from absent or fused ribs. Spine fusion, commonly used for treatment of standard scoliosis[2] caused by idiopathic adolescent scoliosis, may not be the optimal ap-

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Table 1 Thoracic Volume Depletion Deformities

Type of Volume Depletion Deformity	Thoracic Deficit	Mechanism of Lung Volume Loss	Examples
I. Absent ribs and exotic scoliosis	Unilateral thoracic hypoplasia	Lung prolapses into the chest with volume loss.	VATER, absent ribs and congenital scoliosis
II. Fused ribs and exotic scoliosis	Unilateral thoracic hypoplasia	Constriction of lung due to fused ribs shortening hemithorax	VATER, fused ribs and congenital scoliosis, thoracogenic scoliosis from prior thoracotomy
III_A. Foreshortened thorax	Global thoracic hypoplasia	Bilateral longitudinal constriction of lungs from loss of thoracic height	Jarcho-Levin syndrome
III_B. Transverse constricted thorax	Global thoracic hypoplasia	Lateral constriction of lungs from rib deformity	Jeune's asphyxiating thoracic dystrophy, windswept deformity of the thorax in scoliosis

proach for exotic scoliosis because of issues of spine and thoracic growth, bone stock being too weak or too small for fixation, or inability of spine fusion to address three dimensional thoracic deformity. For most cases of exotic scoliosis, the most serious comorbidity is thoracic insufficiency syndrome.

The thoracic mechanism of respiration may be compromised early in life through congenital malformation of the ribs and the spine with reduction in thoracic volume available for the lungs. There also may be loss of the ability to change thoracic volume: the action of the diaphragm may be intrinsically compromised by abnormal proximal insertion proximally or through intrinsic defects such as congenital diaphragmatic hernia. The contribution of chest wall expansion to respiration may be compromised by chest wall absence caused by congenital flail chest or fused ribs as well as distortion of the convex hemithorax ribs caused by rib hump in early onset scoliosis. Mild thoracic deformities may result in subclinical thoracic insufficiency syndrome, whereas more severe thoracic deformity may result in thoracic insufficiency syndrome severe enough to cause respiratory insufficiency with the need for respiratory assistance such as nasal oxygen or ventilator support.

For surgical planning, the patient's thoracic abnormality should be classified as a defined volume depletion deformity[2] of the thorax (Table 1). Some complex patients may have one type of volume depletion deformity affecting one hemithorax whereas the contralateral hemithorax may have another type of volume depletion deformity. In these cases, appropriate surgical strategy is used for each respective volume depletion deformity and addressed in a staged fashion. The indications and contraindications for VEPTR treatment are considered preoperatively (Table 2).

Preoperative evaluation includes standard orthopedic history, respiratory history, physical examination (including assessment of chest wall deformity, thumb excursion test),[1] and

Table 2 VEPTR Indications

FDA Indications for VEPTR Expansion Thoracoplasty	Contraindications
Thoracic insufficiency in skeletally immature patients associated with 1. Flail chest syndrome 2. Constrictive chest wall syndrome, including fused ribs and scoliosis 3. Hypoplastic thorax 4. Progressive scoliosis of congenital or neurogenic origin without rib anomaly Specific indications for fused ribs and scoliosis volume depletion deformity (type II): 1. Progressive scoliosis with three or more fused ribs at the level of the vertebra at the apex of the concave hemithorax 2. Greater than 10% reduction in height of the hemithorax on the concave side compared to the convex side (space available for lung)[1] 3. Progressive thoracic insufficiency syndrome 4. An age of 6 months up to skeletal maturity 5. Concurrent approval of indications by a pediatric orthopaedist, a pediatric general surgeon and a pediatric pulmonologist	1. Stiff thoracic kyphosis greater than 50 degrees 2. Spinal cord syrinx, tethered spinal cord or other spinal cord abnormality 3. Inadequate soft tissue coverage for the VEPTR (usually body weight less than 25 percentile) 4. Absence of proximal ribs for attachment of the VEPTR 5. Inability to tolerate repetitive episodes of general anesthesia (usually due to pulmonary, cardiac disease, or other medical conditions) 6. Absence of diaphragm function 7. Active pulmonary infection