The Synthes Spine Vertical Expandable Prosthetic Titanium Rib (VEPTR) allows new growth sparing surgical procedures for treatment of spine deformity in early childhood. The prime FDA indication for its use is the presence of thoracic insufficiency syndrome [1], which is the inability of the thorax to support normal respiration or lung growth. This syndrome is most often due to three-dimensional thorax deformity. A major shift in paradigm has occurred: spine deformity can no longer be considered an isolated deformity, but rather should be considered a component of the total thoracic deformity that adversely impacts thoracic volume, function, and growth. VEPTR is not a new "growing rod"; it is an instrumentation that stabilizes volume enhancing thoracic reconstructions. Specific VEPTR expansion thoracoplasties can address the different anatomic volume depletion deformities of the thorax [2] (Table I), indirectly correcting scoliosis without fusion, allowing the thoracic spine to grow and contribute to thoracic volume with probable benefit to the growth of the underlying lungs. Early spine fusion, a growth inhibition procedure, does not seem to address thoracic insufficiency syndrome (TIS) with recent reports [3, 4, 5] emphasizing that early spine fusion is associated with decreased vital capacity by maturity. VEPTR is a "buy time" procedure, correcting the thoracic deformity early in life, so rib cage and spinal growth can nurture lung development with definitive spine fusion postponed until adolescence when thoracic volume is optimal. At this time, unfortunately little is known about the normal interrelationship between spine, rib cage and lung growth, or how spine deformity distorts the rib cage with loss of thoracic volume for lung growth, or how biomechanically the spine deformity disables the thoracic ability to expand the lungs through rib cage motion. While much remains to be learned, some basic knowledge exists.

Lung growth is dependent on thoracic growth. The relationship between chest and lung growth was emphasized as early as 1947 by Eng [6]. In 1977, Roaf [7] emphasized that in scoliosis, movements of the chest wall did not increase the volume of the thorax with failure of development of the lungs. In 1979, Chopin [8], through CT scan study, first analyzed the distortion of the rib cage in scoliosis. Two natural history models of thoracic insufficiency syndrome (TIS), Jarco-Levin Syndrome and Juene’s Asphyxiating Thoracic Dystrophy, have a high mortality rate from restrictive lung disease secondary to severe congenital constriction of the chest. Volume of the normal thorax depends on the rib cage providing width and depth and thoracic spine providing height, and the volume is a function of age. Demiglio and Bonnel [9] reported that the thorax is 6.7% adult volume at birth, enlarges to 30% adult size by age 5, becomes only 50% adult size by age 10, but doubles in size to adult volume by skeletal maturity. Lung growth parallels thoracic growth and the increase in lung size depends on two mechanisms: alveolar cell multiplication that is most rapid in the first two years of life and probably continues until at least age 8, then lung alveolar cell hypertrophy, an important but poorly understood aspect of lung growth, en-
larges the lung to adult size [10]. The normal thorax has two important characteristics: it must have a normal volume, and the ability to change that volume (thoracic function) through both the primary breathing of the diaphragm and the secondary breathing of rib cage expansion [1]. The thorax should be of optimal volume and function by skeletal maturity, because aging adversely affects pulmonary function. Normal vital capacity decreases with time [11]. Children with an abnormal thorax due to spine deformity and associated chest wall abnormality probably have additional losses of vital capacity with aging, with possible pulmonary morbidity and an adverse effect on long-term survival.

The first prototype VEPTR operation was done in 1987 at our institution, Christus Santa Rosa Children’s Hospital in San Antonio, Texas, USA. Vertical Steinmann pins were used to treat a potentially lethal congenital chest wall deficiency. Postoperatively the child was successfully weaned off his ventilator within five days of surgery and his scoliosis improved. We began to see more children with rare chest and spine syndromes associated with severe respiratory disease. A multi-specialty approach to evaluation was developed, including a pediatric orthopedist, a pediatric general surgeon and a pediatric pulmonologist. The classic diagnostic syndromes of these children we saw could not explain their severe thoracic and pulmonary disability, so the concept of thoracic insufficiency syndrome was developed by us to explain their findings. TIS is not the same as respiratory insufficiency, which is the failure of pulmonary function to provide adequate body oxygenation. In TIS a child may have abnormal respiration, such as having fused ribs of the chest wall with inability to expand the lung with rib motion, but he may compensate by increasing respiratory rate through diaphragmatic breathing. The child may look normal clinically, but has a severe underlying problem. The other component of TIS is a thorax that cannot grow. A marginally hypoplastic thorax in early childhood may be sufficient, but if the thorax fails to keep pace with skeletal growth, the young adult will have both insufficient thoracic size and lung capacity for pulmonary health. Severe, progressive TIS may eventually cause respiratory insufficiency with the need for oxygen or even ventilator support. In progressive TIS, early VEPTR intervention is best before respiratory insufficiency develops.

A VEPTR custom device was available by 1989 and an FDA sole site feasibility study was begun in San Antonio. Research was supported by grants from the National Organization of Rare Disorders (NORD) and by the FDA Office of Orphan Products Development. The refined Synthes VEPTR was approved for use in an FDA multi-center trial in 1996. (Fig 1) Investigative sites included the Children’s Hospital of Pittsburgh, Boston Children’s Hospital, Seattle Children’s Hospital, Los Angeles Children’s Hospital, Primary Children’s Hospital in Salt Lake City, Shriner’s Hospital of Philadelphia, and the Children’s Hospital of Philadelphia. The Synthes VEPTR device was approved in Europe in 2003 and by the US FDA in 2004 under Humanitarian Device Exemption. FDA indications for VEPTR limit it to skeletally immature patients with thoracic insufficiency syndrome with a defined anatomic diagnosis (Table II).

Assessment techniques for TIS continue to be developed. The thumb excursion test [1] assesses thoracic function: the hands are placed around the base of the thorax with the thumbs equidistant from the spine; respiration through chest wall motion moves the thumbs away from the midline. Greater than 1.0 cm movement is normal. The rib hump in scoliosis disables thoracic function unilaterally, and the thumb excursion test often shows no motion on the affected side. Compromise of diaphragmatic breathing in secondary thoracic insufficiency syndrome is diagnosed by the marionette sign [12]: a bobbing of the head synchronous with respiration. The thorax has collapsed inferiorly towards the pelvis, usually from lumbar kyphosis in myelomeningocele, and the diaphragm is compromised by increased reactive abdominal pressure; literally it is doing a push-up against body weight. This can predispose a child to respiratory failure. Radiographically, the height of the thoracic spine shortened by congenital anomalies or early spine fusion can be compared to normative values for age [9] and a percent normal height derived. Emans et al [13] recently described the relationship between normal thorax width and the width of the pelvic inlet so a percent normal thoracic width can be measured. CT scan normative lung volumes have been recently published by Gollogly and Smith et al [14] so CT scan percent normal lung volumes can now be analyzed. Standard pulmonary function testing is difficult for patients younger than age 6, but research continues to develop.

Fig 1
The evolution of VEPTR (from left to right). The prototype Steinmann pin, the first custom VEPTR, the improved custom VEPTR, and the Synthes IDE VEPTR.
Despite these advances, the diagnosis of thoracic insufficiency syndrome remains complex, with many clinical and laboratory factors to be considered with multiple specialty input being optimal for patient selection.

VEPTR thoracic reconstruction procedures rely on three devices: a VEPTR rib-to-rib device of 200-mm radius, a VEPTR hybrid rib device that can span from rib to either the spine through a laminar hook or to the pelvis with a Dunn-McCarthy hook, and, for the hypoplastic thorax, a 70-mm radius device. Congenital scoliosis is treated with transverse osteotomy of the fused ribs with lengthening of the hemithorax and indirect correction of the scoliosis [12, 15] (Fig 2). For children under 18 months, a single rib to rib device is used. In older children with an adequate spinal canal width, a rib to spine hybrid VEPTR device is used, as well as a second lateral rib to rib device. Device expansion is performed every six months on schedule. Results of congenital scoliosis and fused ribs [12] note an average curve correction of 25° with an average thoracic spinal height growth of 0.71 cm/year, an average growth in width of the thorax of 0.51 cm/year. There was a highly statistically significant growth of both the concave side of the curves and unilateral unsegmented bars [16]. Space available for lung went from 63% preoperatively to 80% at follow-up. The children having surgery younger than age two, when lung growth is at its greatest, had an average vital capacity of 80% at follow-up. The children having surgery younger than age two, when lung growth is at its greatest, had an average vital capacity of 80% at follow-up. Those who had a prior history of spinal fusion and were older than age two at surgery had a vital capacity of only 36%.

These results suggest that with regard to pulmonary outcome, early intervention with VEPTR is preferable. The VEPTR complications are similar to those encountered in use of growing rods, with the most common being an asymptomatic proximal device migration into the rib over an average of 3.2 years, treated by reseating onto the reformed rib of attachment. Absent ribs and scoliosis are treated by multiple devices. Jarchco-Levin syndrome is treated by bilateral expansion thoracoplasty with VEPTR, and Jeune’s asphyxiating thoracic dystrophy is treated with a staged bilateral expansion thoracoplasty with 70-mm radius devices.

**Why did it take so long?** Why did it take over 14 years for VEPTR to gain FDA approval? The reasons are complex. VEPTR could be considered revolutionary (a long-term AO tradition). It is the first truly new concept in FDA-approved spinal instrumentation since the Harrington rod in 1962. It may be said that truly new medical devices need long-term clinical data comparing treatment outcome to controls from large patient populations to prove efficacy and safety to both practicing surgeons and the FDA, but pediatric spine disease is rare and controls are seldom practical. The Synthes VEPTR development eventually proved to be successful and was recently cited as an exemplary example of safe pediatric medical device development in a report to Congress by the US Institute of Medicine (Fig 3). The VEPTR experience, however, confirmed that there are numerous barriers to pediatric device development in the US. In response to inquiries by the US Congress, the FDA in 2004 conducted small group pediatric device task force committee meetings in Washington, DC and the committee identified general obstacles to pediatric device
development which included economic and regulatory issues, reimbursement problems with the humanitarian device exemption law, and the fact that there was no data on extent of unmet pediatric devices. The American Academy of Orthopaedic Surgeons (AAOS) in 2005 subsequently conducted the first pediatric device use survey of SRS and POSNA members and the results were presented to FDA and Congress. Many orthopedic device needs were identified for pediatric patients. Senate legislation to facilitate pediatric device development in the US is now pending.

VEPTR can only be considered the “first step” in the development of the ideal growth-sparing deformity treatment instrumentation for three dimensional thoracic deformity and dysfunction. Future tasks include development of a self-expanding VEPTR, expanded VEPTR indication studies, studies to determine the right time for VEPTR intervention, and an animal model to test the hypothesis of a “surgical trigger” for lung growth. There is also much more to learn about the natural history of TIS as well as outcomes of other spinal treatments, such as early spine fusion, growing rods, and others with regard to their ability to correct three dimensional thoracic deformity and dysfunction. The focus of spine deformity treatment for the growing child has definitely shifted from Cobb angle correction to a more comprehensive three-dimensional thoracic reconstruction. The future will be interesting for continuing AO efforts in these directions.

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<tr>
<th>Thoracic volume depletion deformities</th>
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<tr>
<td><strong>Type of volume depletion deformity</strong></td>
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<td>--------------------------------------</td>
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<tr>
<td>I. Absent ribs and scoliosis</td>
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<td>II. Fused ribs and scoliosis</td>
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<td>IIIa. Foreshortened thorax</td>
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<td>IIIb. Transverse constricted thorax</td>
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Table 1
### FDA HDE anatomic indications for VEPTR treatment

1. **Flail chest syndrome**

2. **Constrictive chest wall syndrome, including**
   - rib fusion and scoliosis

3. **Hypoplastic thorax syndrome, including**
   - Jeunes syndrome
   - Achondroplasia
   - Jarcho-Levin syndrome

4. **Progressive scoliosis of congenital or neurogenic origin without rib anomaly**

### Bibliography