Chapter

49 Vertical Expandable Prosthetic Titanium Rib Insertion

Paulus Steyn

A two-year-old boy is scheduled for placement of a vertical expandable prosthetic titanium rib (VEPTR) system for progressive thoracic insufficiency syndrome in the setting of unilateral thoracic hypoplasia. His past medical history includes a staged repair of a tracheoesophageal fistula and a protracted hospital stay in the newborn period, including a tracheostomy that remained in place until about 1 year of age. His current medications include metoprolol and preoperative vancomycin. His current vital signs are: blood pressure 86/52 mmHg; heart rate 110/min and regular; respiratory rate 37/min; temperature 36.5°C; SpO_2 93% on room air. Physical exam reveals an alert and anxious child with normal facial structures. He has a tracheostomy scar. He is not flaring, but he is using his left-sided accessory muscles of breathing. He has right-sided neck torticollis but his airway exam is otherwise unremarkable.

Chest radiograph demonstrates scoliosis with a curve of 100 degrees in the thoracic spine and left sided lung volumes four times smaller than the right side (Figure 49.1).

What Is Thoracic Insufficiency Syndrome?

Thoracic insufficiency syndrome results from any condition that alters thoracic development and results in decreased lung growth. The normal thorax has two important functions: to maintain a constant and normal lung volume and to effortlessly change this volume. Lung volume depends on thoracic spine height and an equally broad and wide enough rib cage. Ventilation (lung volume exchange) depends on normal bilateral diaphragm excursion, rib orientation (recall the bucket handle mechanism) and functioning of the secondary muscles of respiration. The components of the normal thorax consist of the spine, sternum, ribs, secondary muscles of respiration, and diaphragm.

Lung development depends on normal thoracic growth, which entails a coordinated increase in thoracic spine height, and symmetrical enlargement of the rib cage that requires both rib growth and the correct orientation of the ribs. The thoracic spine lengthens by about 1.5 cm/year for the first five years of life, then slows down to about 0.5 cm/year until it experiences another growth spurt of 1.2 cm/year from 11–15 years of age. In the first two years of life, the rib cage is square shaped, the ribs are orientated horizontally and grow in length. Between two and 10 years of age it courses downward and forms an oval-shaped thoracic cross-section. There is a rapid increase in rib growth after the age of 10 years until the child reaches maturity; the final cross-section of the thorax is rectangle shaped.

The diagnosis of thoracic insufficiency is made when the patient has physiologic signs of respiratory impairment, a decrease in chest wall mobility, and worsening lung volumes on imaging. If obtained, pulmonary function tests demonstrate a restrictive pattern with a smaller than predicted vital capacity.

What Conditions Are Associated with Thoracic Insufficiency Syndrome?

Causes of thoracic insufficiency syndrome can be divided into disorders that affect stable lung volumes and disorders that affect normal ventilation.

Disorders that affect stable lung volume include scoliosis with fused or missing ribs, scoliosis with a windswept deformity, Jeune syndrome (asphyxiating thoracic dysplasia, a rare genetic disorder marked by a narrow/small thorax and progressive kidney failure), and spondylocostal dysplasia (previously known as Jarcho–Levin syndrome), a rare congenital disorder that presents with at least 10 consecutive vertebrae that are fused, missing, or partially formed. *Spondylocostal dysplasia* is also marked by ribs that are broadened, fused, growing in bizarre directions, or altogether missing.

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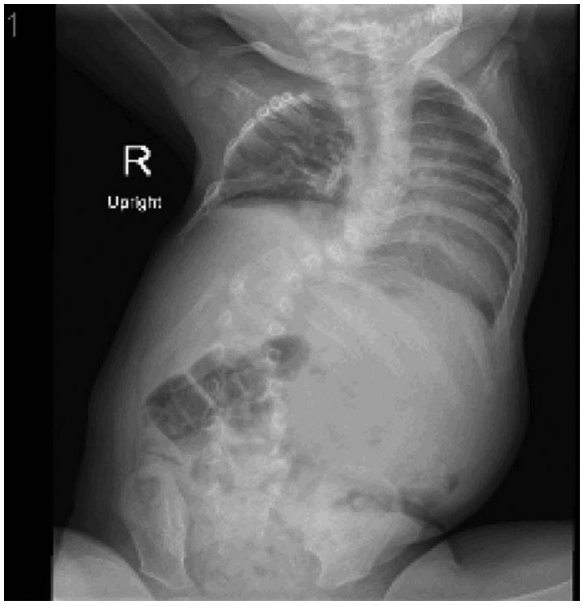

Figure 49.1 AP chest X-ray demonstrating poorly formed and concave vestigial ribs. Reproduced with permission from Springer Nature from Campbell, R.M., VEPTR: past experience and the future of VEPTR principles. *Eur Spine J* 2013;22 Suppl 2:S106–17

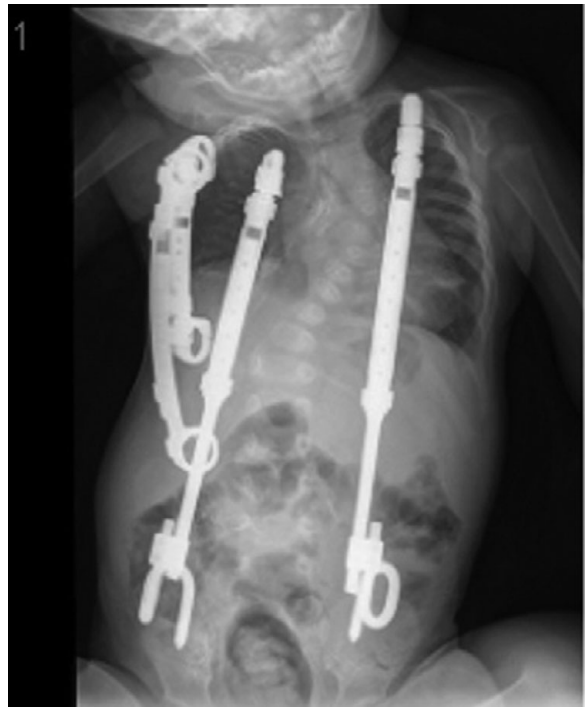

Figure 49.2 Postoperative AP chest X-ray demonstrating VEPTR placement. Reproduced with permission from Springer Nature from Campbell, R.M., VEPTR: past experience and the future of VEPTR principles. *Eur Spine J* 2013;22 Suppl 2:S106–17

Disorders that affect normal ventilation (movement of lung volume) include: congenital diaphragmatic hernia and hemidiaphragmatic paralysis (think phrenic nerve injury from lateral hyperextension of the neck at birth).

Note that standard scoliosis is not typically considered a cause of thoracic insufficiency syndrome.

What Is a VEPTR Procedure?

The Vertical Expandable Prosthetic Titanium Rib (VEPTR) is a device designed to address the complex disorders that cause progressive thoracic insufficiency syndrome in a three-dimensional manner. It is used to stabilize and progressively expand the thorax. Depending on the exact condition it is placed in conjunction with a wedge/expansion thoracotomy, but can also act as the sole device used to stabilize and expand a hemithorax such as in cases where absent ribs cause flail chest. The goals of a VEPTR system are to improve thoracic volume and function by correcting scoliosis and establishing symmetry of the thorax. The idea is to maintain these corrections as the patient grows using successive expansions every four to six months (Figure 49.2).

What Are the Indications for a VEPTR?

The VEPTR device is indicated to treat patients with conditions that cause thoracic insufficiency syndrome. They include conditions that cause progressive multidimensional defects of the thorax that are present from birth (congenital), such as scoliosis with absent ribs and flail chest, neurogenic scoliosis (rib abnormalities not necessarily present), and scoliosis with fused ribs, congenital conditions that cause hypoplastic chest wall defects such as Jeune Syndrome or spondylocostal dysplasia (Jarcho–Levin Syndrome), or acquired chest wall defects that may arise from trauma or tumor resections.

What Are Common Surgical Complications of the VEPTR Device?

Described complications include wound infection, skin breakdown, and brachial plexus injuries. The device can migrate or break, which will entail surgical repair.

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Is There Anything Else You Want to Know before Proceeding with This Case?

A thorough medical assessment will reveal associated cardiac and genitourinary conditions. Recall that patients with congenital defects of the vertebrae may also have anal atresia, tracheoesophageal fistulas, cardiac anomalies, radial atresia, and renal abnormalities (VACTERL syndromes). Echocardiography is important for assessing cardiac status. MRI can delineate spinal cord deformities and preexisting areas of spinal stenosis. The patient may have chronic hypoxia with compensatory metabolic alkalosis and polycythemia. A preoperative CT scan or MRI is helpful as a blueprint to map out chest wall deformities and assess lung volumes and the extent of thoracic insufficiency.

What Are the Important Anesthetic Implications?

Since the spinal cord will be manipulated and/or stressed, in our practice we routinely monitor somatosensory and motor evoked potentials, facilitated by a total intravenous general anesthetic with propofol and the anesthesiologist's opioid of choice. Intraoperative changes in neurophysiological signals warrant optimization of hemoglobin level and blood pressure, and immediate investigation of a surgical cause.

For the child that receives their first VEPTR device or presents for an expansion, we typically place one or two intravenous lines following inhalational induction using sevoflurane. Because of their underlying condition, and the relatively high frequency of their need for procedures, it is common for these children to be quite challenging to obtain IV access. Therefore, we usually employ ultrasound guidance, often with the aid of our radiology colleagues. Many of these children will already have had placement of an indwelling peripheral or central venous access. Unless dictated by the severity of their underlying condition, direct arterial monitoring is usually not necessary. Original insertion of the VEPTR will occasionally warrant red cell transfusion for intraoperative or preexisting anemia.

Intraoperative hypothermia is common, due to prolonged exposure during induction, IV placement, and surgical positioning. Therefore, all modalities should be taken to prevent hypothermia, including adjusting the OR temperature, use of a forced air warming blanket, and IV fluid warming device. A urinary catheter will only be necessary for procedures expected to last longer than three to four hours.

The surgical procedure is performed in the prone position. Therefore, adequate attention is paid to optimization of position and airway stabilization. All patients for VEPTR receive an endotracheal tube unless they have a preexisting tracheostomy. The anesthesia provider must have flexible endotracheal tube extensions, a soft bite block, and different sized prone pillows immediately available.

What Are Important Postoperative Concerns?

The most important postoperative concern is pain. Many of these children do not have appropriate age-related or cognitive development to use patient controlled analgesic devices; therefore, extra attention is warranted to optimize pain control with opioids and additional adjuvants. Ideally, the institution's pain team should be consulted on these children during the initial VEPTR placement.

Suggested Reading

Campbell RM Jr. VEPTR: past experience and the future of VEPTR principles. *Eur Spine J*. 2013;22 Suppl 2:S106–17. PMID: 23354777.

Harris JA, Mayer OH, Shah SA, et al. A comprehensive review of thoracic deformity parameters in scoliosis. *Eur Spine J*. 2014;23(12):2594–602. PMID: 25238798.

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