JBJS ESSENTIAL Surgical Techniques

Expansion Thoracoplasty for Thoracic Insufficiency Syndrome Associated with Jarcho-Levin Syndrome

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Introduction

Although surgical treatment of spondylothoracic dysplasia (STD) is controversial, we have found that an expansion thoracoplasty using a Vertical Expandable Prosthetic Titanium Rib (VEPTR; DePuy Synthes Spine, Raynham, Massachusetts) results in favorable outcomes, including 100% survivability (at an average follow-up of 6.2 years), increased thoracic spinal length, and decreased requirements for ventilation support[1].

Jarcho-Levin syndrome includes two distinct anatomic variants that have severe shortening of the chest: spondylocostal dysostosis (SCD) and STD. SCD is usually associated with severe scoliosis and chaotic patterns of absent and/or fused ribs and can be treated with previously reported VEPTR techniques[2]. STD is a much more severe type-IIIA volume depletion deformity of the thorax[3], having mild scoliosis but extreme shortening of the thoracic spine, averaging 25% of the normal height[4] with almost all of the posterior ribs fused in a "crablike" pattern. STD has a high mortality rate, reported in the earlier literature as approaching 100% within the first year of life, because of severe extrinsic lung disease[5,6]. However, recent reports[4,7,8] have noted an improved survival rate of 55% to 58%, likely from more advanced intensive care in modern times, with most deaths occurring in the first year of life from respiratory failure. The adult survivors in the series described by Ramírez et al. survived even though the average vital capacity was 28% of the normal amount expected for this age, and it is unclear why they survived despite an extremely low vital capacity[4].The present article describes a VEPTR expansion thoracoplasty developed for the surgical treatment of STD using a unique v-osteotomy VEPTR expansion thoracoplasty of the posterior fused rib mass, which was first performed by Campbell and Smith in 1996[3]. Selecting appropriate patients for this surgery requires clinical judgment. The core indications for this procedure are a diagnosis of STD, a thoracic spine that is <50% of the predicted height, a crablike rib cage with extensive rib fusion, and clinical and/or radiographic signs of worsening thoracic insufficiency syndrome. The key parameters to assess clinically are an elevated respiratory rate; an increased rate of respiratory illness; a decrease in play activities; an increased need for respiratory assistance, such as oxygen or CPAP (continuous positive airway pressure) or BiPAP (bilevel positive airway pressure); or increased ventilator dependence. A worsening thoracic spine height over time, measured radiographically as a percentage of the expected height, as well as decreasing lung volumes on a computed tomography (CT) scan as a percentile for age, suggest surgical intervention should be considered. The patients who have improvement should probably be followed with reassessment clinically and radiographically every six months.

The treatment goals of the v-osteotomy expansion thoracoplasty include lengthening the fused posterior aspect of the thorax. This moves the insertion of the posterior diaphragm distally, thereby enlarging the overall space available for the lung. The procedure is staged; first, the right side is enlarged because there is more lung tissue on that side, and three to six months later, the left side is enlarged. Bilateral expansion thoracoplasty during the same surgical session is too much surgery for these fragile children to tolerate and therefore is not recommended. The devices, once implanted, are lengthened through the standard VEPTR technique of limited 3-cm incisions every four to six months under general anesthesia. We observe these patients overnight as outpatients after expansion surgery to minimize the risk of respiratory complications such as mucus plugs.

Step 1: Preoperative Preparation

Make anteroposterior and lateral radiographs of the spine.

- Make lateral radiographs of the cervical spine in flexion and extension to provide details of the cervical spine as well as to assess for any instability.
- Acquire a magnetic resonance imaging (MRI) scan of the entire spinal cord to assess the cord for any abnormality.

- Perform CT scans of the chest and lumbar spine at 5-mm intervals, unenhanced, to clearly define the anatomy of the chest and spine anomalies. Also, calculate lung volumes.
- Acquire a dynamic lung MRI scan, if possible, to assess baseline thoracic function.
- Upper airway anomalies are common, and an otolaryngology consultation is recommended. Cardiology consultation may be necessary if early cor pulmonale is suspected. Early consultation with the anesthesia staff is advised so that they can plan for possible fiber-optic-assisted intubation because of airway abnormalities and any other potential issues complicating general anesthesia.
- To complete the tri-specialty workup, pediatric general surgery and pediatric pulmonary consultation are also recommended.

Step 2: Position the Patient for the Procedure

The patient is placed in the prone position.

- The presence of a tracheostomy is common in these patients, and the skin underneath the tracheostomy collar is usually inflamed from friction. Carefully shield the tracheostomy from the operative field by draping (e.g., Steri-Drape 1010 adhesive barriers; 3M, Saint Paul, Minnesota). It is not necessary to sew in the tracheostomy tube for the procedure. Work closely with the anesthesia staff with regard to head and body positioning so that the tracheostomy tube is not in danger of kinking, and access to all intravenous lines is easily available for anesthesia.
- Use prophylactic intravenous Ancef (cefazolin) and gentamicin.
- Ensure that the patient has somatosensory and motor-evoked potential spinal cord monitoring during the procedure.

Step 3: The Incision

A curvilinear skin incision is made, starting proximally between the spine and the medial edge of the scapula (Figs. 1 and 2).

- Carry the incision distally and then gently curve it anteriorly in line with the tenth rib.
- Continue the dissection with cautery through the trapezius, rhomboid, and latissimus dorsi muscles.
- Next, use cautery to reflect the paraspinous muscles from lateral to medial, up to the transverse processes of the thoracic spine.
- Isolate the common insertion of the middle and posterior scalene muscles proximally.
- Note that the neurovascular bundle is anterior to the common middle and posterior scalene muscle insertion on the second rib.
- The medial portion of the rib cage is a very homogeneous broad mass of bone. Look for vestigial transverse grooves that laterally become a separation between the ribs.
- Identify the upper two ribs and then, using cautery, place a line between the second and third ribs. Carry the line medially until the spine is approached and then direct the line to end at the midportion of the fused mass of medial ribs just adjacent to the transverse processes of the spine.
- Place a similar line with cautery inferiorly, starting at the top of the eleventh rib and then drawing it medially to join the upper line at the edge of the spine to form the "v." If more bone stock thickness is needed for support of the rib cradles, then include the third rib in the superior section of ribs and the tenth rib in the inferior section, slightly narrowing the central v-shaped section of fused ribs.

Step 4: The Osteotomy

Perform the v-osteotomy (Figs. 3 and 4 and Video 1).

- Start the osteotomy superiorly at the lateral border of the fused ribs, usually at the mid-axillary line.
- Strip the periosteum anteriorly by inserting a number-4 Penfield elevator beneath the rib interval where the osteotomy is to be cut. The elevator protects the underlying lung.
- Then use a Kerrison rongeur to carefully cut along the line marked by the cautery. Continue this medially until the transverse processes of the spine are reached. Cut no further medially.
- Make a similar second cut inferiorly with the Kerrison rongeur to complete the v-osteotomy.
- Next, place two small lamina spreaders in the osteotomies, one proximally and one distally, and use them to gently distract the superior and inferior osteotomies apart, leaving the central v-shaped portion stationary.
- Utilizing a Freer elevator, gently strip down the pleura proximally, stripping it from the rib cage, so it lies loosely on the surface of the lung, and do the same thing distally through each osteotomy widened by the lamina spreaders to ease the lengthening of the hemithorax.
- Gradually widen the osteotomy using this maneuver with the lamina spreaders until it

is widened enough that a partially assembled number-4 VEPTR-I 220-mm-radius (superior rib cradle, rib sleeve, and inferior rib cradle) device will fit.

Step 5: Placement of the VEPTR Device

A number-4 VEPTR-I device is wedged in, starting laterally within the osteotomy sites, wedging the osteotomies apart, distracting the superior ribs proximally and the inferior ribs distally, lengthening the hemithorax, and stopping approximately at the posterior axillary line, when there is maximum stress on the superior and inferior ribs, to avoid fracture, and the lamina spreaders are then removed (Figs. 5 and 6).

- With the device held in place, insert a Freer elevator under the site of the fused mass of the first and second ribs to protect the lung. Then use a powered burr to cut a hole in the fused rib mass just underneath the first rib for the placement of the rib cradle end half of the superior rib cradle of the VEPTR.
- Once this is accomplished, carefully thread the cradle end halves into the hole in the rib mass and mate them to the rib cradles, proximally and distally, and lock them into place with the cradle locks.
- Lock the rib sleeve with a distraction lock, completing the implantation. As long as there is reasonable bone thickness of the first rib above the device, the risk of brachial plexus impingement is minimal.

Step 6: Wound Closure

Insert drains and local anesthetic catheters and close the wound.

- Insert number-7 and number-10 round Jackson-Pratt drains on the operative field, with the number-10 drain placed medially and the number-7 drain placed laterally.
- Place a local anesthetic catheter for assistance with pain control.
- Gently stretch the soft-tissue musculocutaneous flaps before closure. Reapproximate the paraspinal muscles in their anatomic position and suture them down with interrupted Vicryl suture (polyglactin; Ethicon). Close all remaining muscle layers with running Vicryl suture.
- Perform skin closure with 4-0 Monocryl (poliglecaprone; Ethicon), and use Dermabond (Ethicon) to seal the incision along with Steri-Strips (3M).
- Make intraoperative anteroposterior and lateral radiographs of the entire spine, including the chest, to check on device positioning.
- Leave the drains in place until the drainage is ≤20 mL per twenty-four hours.
- Leave the pain catheters in place for forty-eight hours.

Step 7: Expansion and Replacement Procedures

Lengthen the devices with the standard VEPTR technique of limited 3-cm incisions every four to six months.

- For expansion of the devices, make 3-cm linear incisions bilaterally with the patient under general anesthesia to expose the devices. The distraction locks are removed, the devices are expanded 0.5 to 1.0 cm, stopping when the reactive force becomes excessive, and then new distraction locks are inserted. Closure is in the usual fashion. No drains are used. The patient is observed overnight for any respiratory difficulties.
- For replacement of completely expanded devices, the rib sleeve and inferior rib cradles are exposed through the prior thoracotomy incisions. They are unlocked and removed and then longer devices are inserted. Number-7 round Jackson-Pratt drains are used bilaterally. Closure is in the usual fashion. The patient is observed overnight.

Results

VEPTR treatment in patients with STD is associated with increased thoracic spine height and reduced thoracic width-to-height ratio, suggesting a greater gain in height than in width[1] (Video 2). In addition, these patients display a decreased requirement for ventilation support, representing an important improvement in quality of life. Blood gas and hemoglobin levels remained stable, while respiratory rate improved by decreasing. Few preoperative pulmonary function tests were performed because of the young age of many of the patients; however, sequential postoperative pulmonary function tests that were done once the patients were old enough to be able to cooperate with standard spirometry, usually after the age of six years, showed that the raw vital capacity tended to increase over time but decreased as a percentage of the vital capacity predicted on the basis of arm span (Video 3). Despite this decrease, the percentage of the predicted value remained higher than those reported for the natural history[4,7,8]. Overall, the reductions in ventilation needs and age-appropriate reductions in respiratory rate suggest that these patients were improving clinically from a respiratory viewpoint. Following the VEPTR treatment, our patients with STD had stabilization of their condition and all were living at the time of writing.

These results are consistent with findings after VEPTR treatment of other spinal and chest wall deformi-

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ties, including SCD[1] and congenital scoliosis[9,10] as well as Jeune syndrome[11], myelomeningocele, and cerebrocostomandibular syndrome (all unpublished data). In these patients, thoracic growth, improved respiratory functioning, and improved survivability are also positive outcomes of treatment.

What to Watch For

Indications

- Spondylothoracic dysplasia with a thoracic spine that is <50% of normal height in patients six months or older.
- Severe widespread fusion of the posterior aspect of the ribs in a crablike type of configuration.
- Early respiratory insufficiency based on an elevated respiratory rate or decreased activity levels.
- More advanced clinical respiratory insufficiency based on the need for respiratory assistance such as O_2 by nasal prongs, a CPAP-BiPAP device, or need for a ventilator.

Contraindications

- Inability to survive a thoracotomy operation.
- Inability to tolerate multiple lengthening and/or device replacement operations for the VEPTR, usually performed every four to six months.
- Inadequate bone stock for insertion of a number-4 VEPTR device, which can span 4 cm. For this procedure, the approximate minimum height of the most laterally fused ribs on the radiograph should be $\geq$6 cm.

Pitfalls & Challenges

- Complications that may be encountered include a skin slough or wound infection.
- The proximal incision is at risk of infection because of the common presence of a tracheostomy collar and the contaminants beneath it.
- Wound problems are treated with debridement, antibiotics, vacuum-assisted closure, and eventual closure.
- These patients are extremely fragile medically and are prone to pneumonia, cor pulmonale, and upper airway management problems. Given the complexity of these patients, pediatric critical care management input is advisable to ensure the best possible outcomes.

Clinical Comments

- Some have argued that surgery for STD is never needed since the natural history of the disorder is favorable if the patient survives the first year of life[7]; however, this seems to apply primarily to the STD survivors who reside in the tropical climate of Puerto Rico where there may be a lower prevalence of pneumonia and other respiratory illnesses that can prove fatal to patients with extremely low vital capacity such as is found in STD. Our anecdotal experience in North America is much less favorable about the natural history of STD, with patients experiencing substantial mortality after infancy. Therefore, we argue that surgery for STD should be considered if the clinical course is becoming unfavorable.

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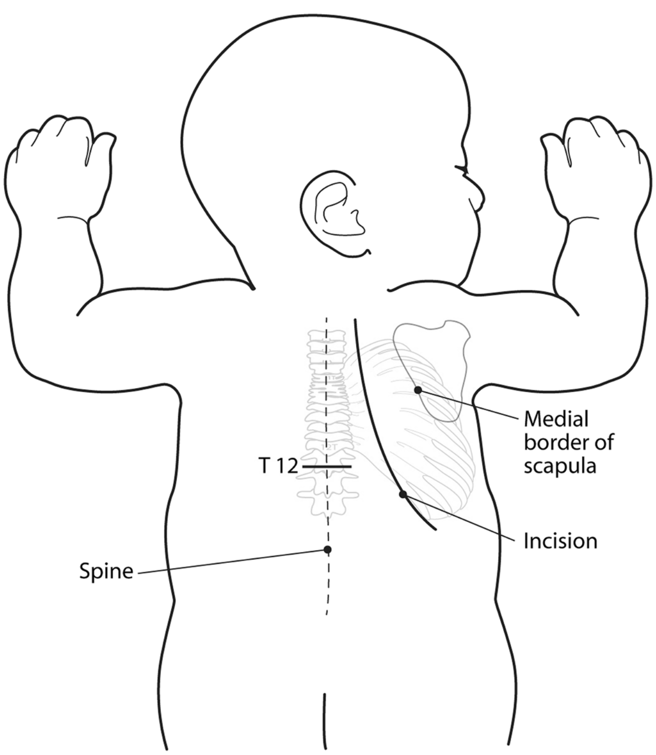

Fig. 1

The patient is placed in the prone position. Longitudinal chest rolls are placed under the chest. A transverse roll is placed under the pelvis with care taken to avoid pressure on the genitals. The arms and legs are padded with sheet foam. After sterile preparation, a curvilinear skin incision is made, beginning midway between the spine and the medial border of the scapula, and is carried distally toward the palpable edge of the fused rib mass.

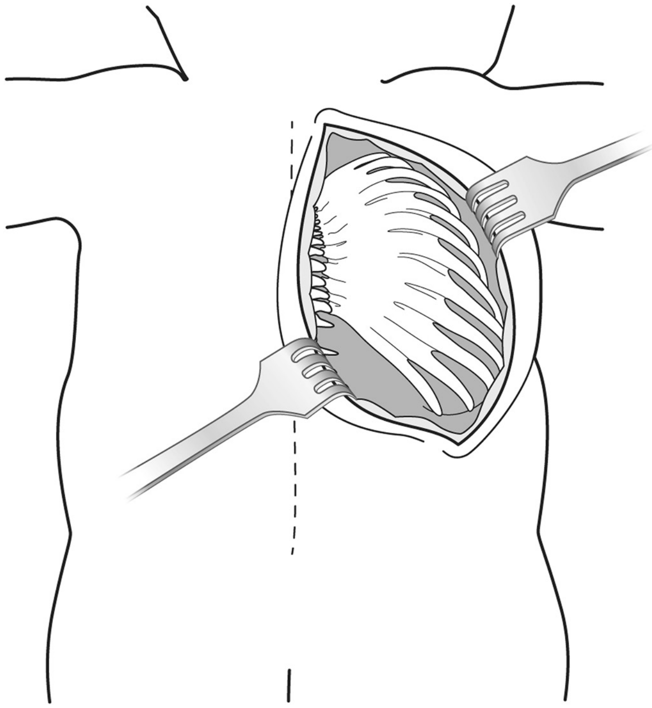

Fig. 2

Cautery is used to section the trapezius, rhomboid, and latissimus dorsi muscles in line with the skin incision. The paraspinous muscles are elevated by cautery from lateral to medial until the tips of the transverse processes of the spine are visualized.

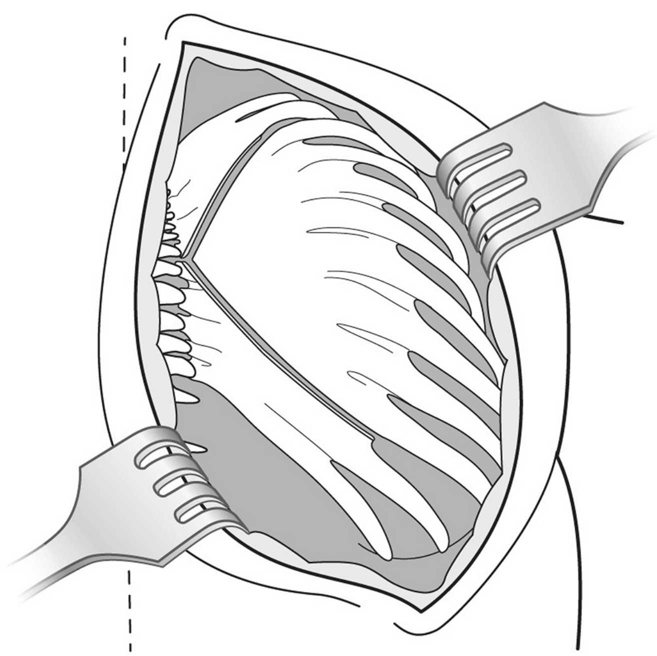

Fig. 3

The v-osteotomy is cut, beginning laterally and going medially, first starting superiorly. The ribs begin to separate laterally, and the first two ribs are identified. Cautery is used to mark between the second and the third rib, and the line of the osteotomy is brought down medially to the midportion of the fused rib cage. A second line is made by cautery on the top of the eleventh rib and is carried medially and upward to end at the point where the superior line was drawn to form the apex of the "v." This is the ideal placement of the lines of the osteotomies; however, if more bone stock for the support ribs is needed, then the lines are drawn to include the third rib in the upper portion and tenth rib in the lower portion of the fused rib mass. To begin the osteotomy, gently insert under the fused rib mass a number-4 Penfield elevator approximately 2 cm medially at the point where the second and third ribs begin to separate under the ribs and in line with the planned osteotomy. This strips down the periosteum and the pleura, protecting the lung underneath it. Next, use a Kerrison rongeur to cut the osteotomy from lateral to medial, gradually advancing the Penfield elevator as the osteotomy progresses. Stop the cut at the tips of the transverse processes of the spine. Use the same technique to cut the inferior osteotomy.

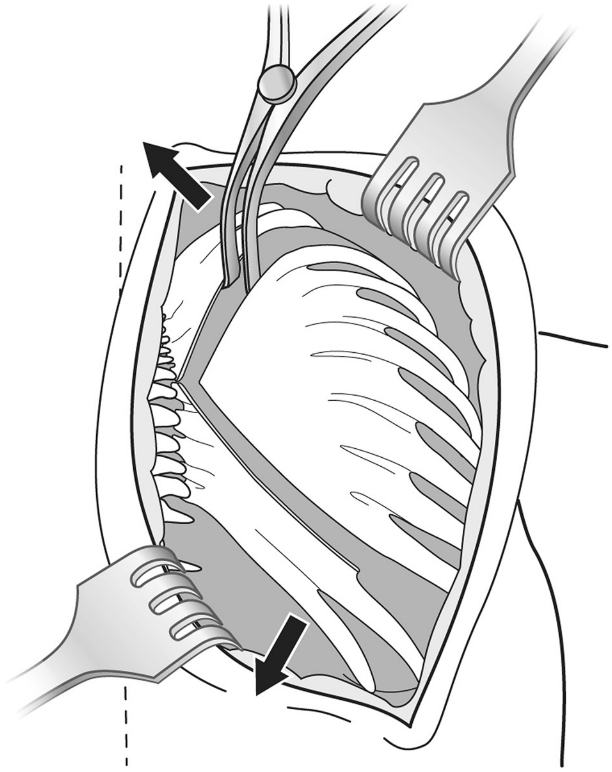

Fig. 4

Lamina spreaders are used to widen the osteotomies, leaving the central portion stationary. While the osteotomy should be widened as much as possible, care should be taken not to fracture the upper and lower ribs that are still attached to the spine. A Freer elevator is inserted through the osteotomies to strip down the pleura to aid in widening the osteotomies.

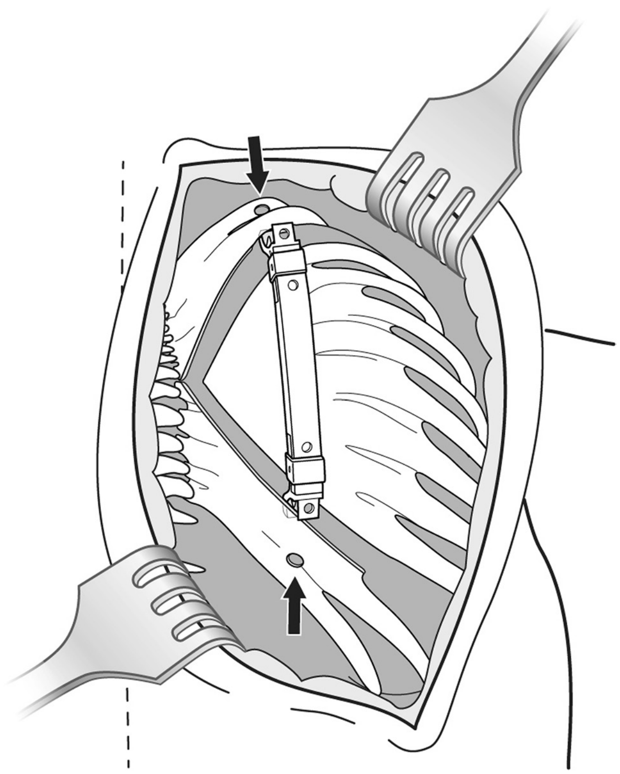

Fig. 5

Next, a number-4 (220-mm-radius) VEPTR-I device is wedged in, from lateral to medial, as far medial as possible without fracturing the superior and inferior ribs of attachment. A Freer elevator is inserted through the osteotomy site under the first and second fused ribs to protect the lung, and a powered burr is used to place holes (arrows) so that a rib cradle end half can be inserted to lock in the VEPTR.

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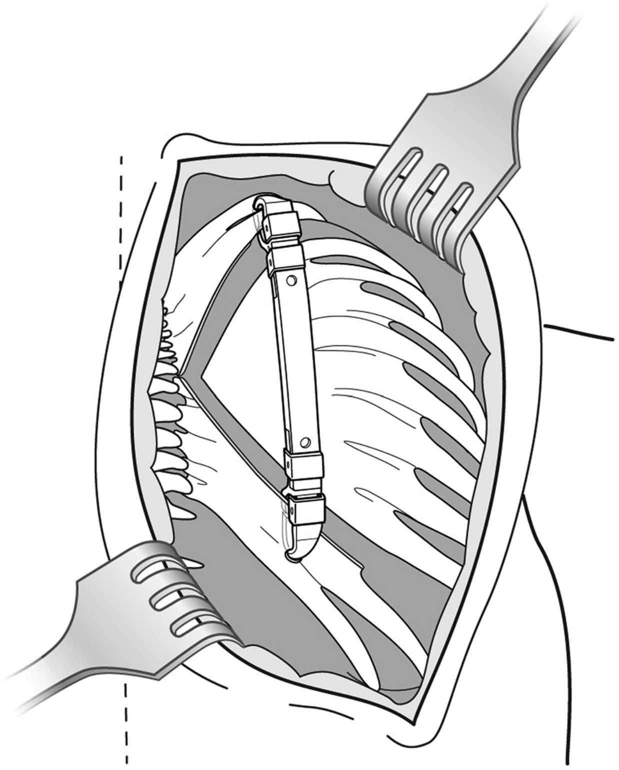

Fig. 6

The rib device is placed and locked with the cradle end half blue locks. Distraction locks are inserted into the rib sleeve of the device to complete assembly. Note that the upper portion of the device should never extend above the first rib to avoid brachial plexus compression.

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