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Nuss procedure in the adult population for correction of pectus excavatum

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ABSTRACT

Minimally invasive repair of pectus excavatum has been successfully modified for use in adult patients. Many patients present in adulthood with progression of symptoms as the chest wall becomes less compliant with age. A thorough workup is completed including echocardiogram and chest CT to evaluate for anatomic abnormalities. Cardiopulmonary exercise testing is done to quantify the physiologic impact. Modifications of the original Nuss procedure required to allow for successful adult repair include the use of forced sternal elevation, the use of multiple bars, medial bar fixation, and interspace support to prevent bar rotation and migration. Occasionally, fractures may occur that require an open procedure and osteotomy or cartilage resection and hybrid approach incorporating the principals of intrathoracic support and osteotomy with bar stabilization.

Introduction

Pediatric surgeons have established that children are not just small adults and require specialized training and techniques for treatment. Likewise, successful repair of adults with pectus excavatum (PE) requires modifications of Nuss' procedure that has been universally adopted as the standard of care for repair of children and adolescents.[1] The optimal surgical treatment of adult patients with PE has been controversial with some surgeons warning against a minimally invasive repair of pectus excavatum repair (MIRPE) in patients beyond adolescence.[2,3] Repair of adults is more difficult and has greater complications reported.[4–7] Successful repair can however be obtained with an MIRPE approach.[4,6,8–10] We present our technical modifications of MIRPE that have enabled safe, cosmetically and physiologically successful and durable repair in adults up to age 72 years.

Adult patient presentation

Pectus excavatum is reported in the pediatric literature to be four times more common in men than women.[11,12] Similarly, surgical series of adults also report a higher frequency of men versus women undergoing surgical repair.[6,8,13] There may be, however, a large number of underdiagnosed or misdiagnosed females as breast tissue or breast implant augmentation obscures the defect.[11,12] Additionally, some PE patients may not experience symptoms until they enter adulthood.[14,13] Adult patients may present with notable progression of symptoms with aging as the chest wall becomes less compliant and compensatory mechanisms decrease[13,15–17] (Fig. 1 a and b). In nearly half of adults studied by Kratgen et al. development of symptoms did not occur until patients were in their 30's- to 40's.[15] They also noted symptoms improved significantly after surgical repair.[15]

There are a number of studies evaluating the physiologic impact of PE in children during exercise,[18–20] but there are few reports that exclusively examine adult patients.[21–29] The inward projection of the chest wall results in a negative cardiopulmonary impact due to varying degrees of cardiac compression.[23,26] With significant compression of the right heart chambers, right heart filling and flows are reduced leading to decreased stroke volume and cardiac output[17,22,23,26] (Fig. 1 c and d). Surgical repair of the PE alleviates compression, allowing for an increase in right heart chamber size, flow velocities, and improved cardiac output[6,15,23,26,25] (Fig. 1 e–g). In 101 PE patients ≥30 years, a mean increase in right ventricular output by 65% was seen after MIRPE by one center utilizing intraoperative transesophageal echocardiogram.[6] Krueger et al. also noted significant improvement in both right and left ventricular outputs after open repair.[25] Speckle tracking strain has also been used to evaluate myocardial contractile function of the right and left ventricle in PE patients before and after repair.[23] In 165 adults, significant improvements in all except LV radial strain rate were demonstrated after MIRPE repair.[23]

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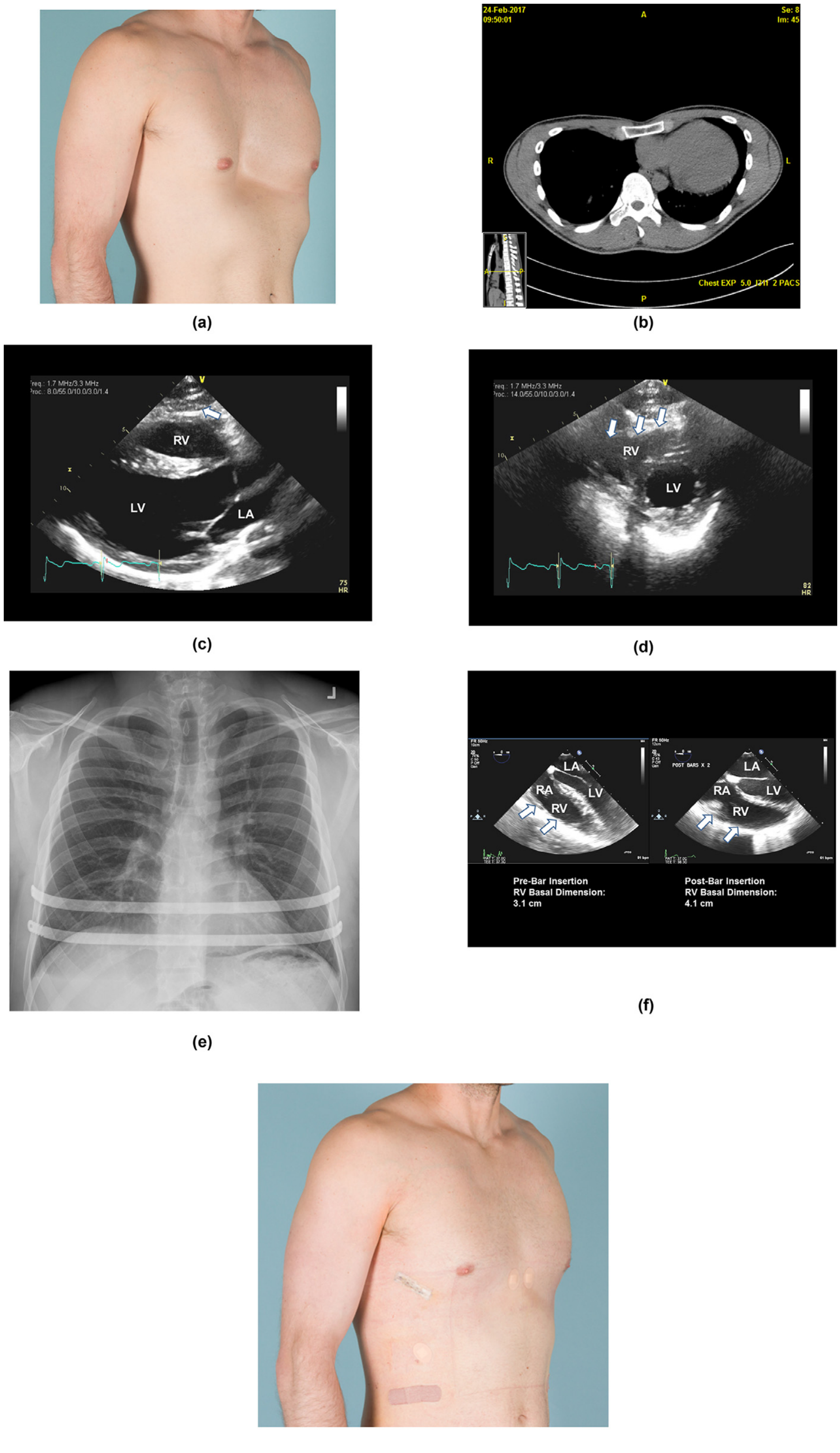


Fig. 1. a–g (a) Photograph of 30-year-old man with severe pectus excavatum which developed in his early teens. He noted significant progression of exertional dyspnea and tachycardia over the past 5 years. (b) Subsequent CT scan showed deformity causing cardiac compression and Haller index of 5. CPET testing showed significant decrease in exercise capacity with peak anaerobic VO_2 of 60% predicted. An echocardiogram was performed (c) Parasternal long-axis transthoracic view demonstrated basal to mid right ventricular external compression (white-arrow) due to pectus excavatum deformity (d) Parasternal short-axis transthoracic view demonstrating significant right ventricular external compression (white-arrows) due to pectus excavatum deformity. (e) An MIRPE with 2 bars was performed for PE correction (f) Four-chamber transesophageal echocardiogram imaging was performed during bar insertion. The pre-bar insertion images (left) demonstrated basal to mid right ventricular external compression due to pectus excavatum deformity (white-arrows) with basal right ventricular dimension measuring 3.1 cm. (Right) Following 2 bar insertion the right ventricular compression improved (white-arrows) and the basal right ventricular basal dimension increased to 4.1 cm. (g) Excellent cosmetic results and complete resolution of symptoms was obtained postoperative.

Improvements in right ventricular preload, stroke volume and cardiac output may be related to an intrinsic improvement in the RV strain and strain rate seen after relief of cardiac compression.

Maximal exercise capacity is directly correlated with oxygen delivery and oxygen delivery is an indirect measure of cardiac output.[22] Normalization of decreased cardiopulmonary function has been shown in teenagers three years following MIRPE[30] Neviere et al. reported significant improvement seen in his adult patients after 1 year post modified Ravitch.[21] Udholm, however, in contrast to their findings in children and teens, did not find significant improvement in the cardiopulmonary function of 11 adults tested at 1-year post MIRPE.[31,32] However, the series was very small, and there was a trend toward improvement which may be become significant with additional time, or when examined in a larger cohort.

Adult patient evaluation

When an adult patient presents with PE, an evaluation should be completed to understand the severity of the condition and to rule out other cardiopulmonary disorders. A thorough workup is indicated in patients with symptoms. Initial evaluation should include imaging, specifically magnetic resonance imaging or noncontrast chest computed tomography (CT), which allows for visualization of the deformity, evidence for cardiac compression or displacement, and potential intrathoracic pathology.[16,33] The scan should be performed with both inspiratory and expiratory phases as the severity of the defect may substantially worsen when a patient exhales (Fig. 2a and b).[17] In adults, changes between inspiratory and expiratory imaging may also be used as a measure of chest wall flexibility. Imaging is used to calculate the index of severity at the lowest level of the pectus deformity and can be assessed using the Haller index (normal 2.5–2.7) or the correction index. The PE is severe if the Haller index is greater than 3.2 or the correction index score is more than 10%.[34–36] Next, a 12-lead electrocardiogram is completed, and if abnormalities worrisome for ischemia are reported, a stress test and cardiology consultation should be initiated. Additional laboratory analyses based on patient comorbidities should be obtained at the physician's discretion.

In order to rule out anatomic cardiac abnormalities, an echocardiogram should be completed. Echocardiogram is particularly critical for patients who are suspected of having connective tissue disorders to assess aortic dimensions and valve function.[37] Measurement of ejection fraction should also be performed. Some centers also evaluate myocardial strain.

Cardiopulmonary exercise testing (CPET) is used to help quantify the degree PE affects a patient's ability to exercise and consume oxygen. Cardiac limitations due to cardiac compression by the PE are demonstrated by an abnormally low peak maximal anaerobic VO_2 during exercise testing.[21,32] Pulmonary function may not be markedly abnormal in patients with PE; however, in pediatric patients, static studies have reported a median percent predicted value lower than normal for FVC, FEV1 and $FEF_{25\text{-}75}$.[38] Symptoms of significant dyspnea in the adult warrant ruling out a pulmonary pathology.

Indications for surgery

As in pediatric patients, surgery is recommended for symptomatic adult patients with severe deformities having a Haller Index score of more than 3.2 or a correction index of more than 10%.[35] Evidence for cardiopulmonary dysfunction includes decreased peak anaerobic VO_2, restrictive pulmonary disease, or right-sided cardiac compression seen by echocardiogram or radiographic imaging. In addition, a significant decrease in cardiac output from supine to sitting position on echocardiogram may be used as medical evidence for repair.[39]

Basic principles of repair

Minimally invasive repair of pectus excavatum (MIRPE), or the "Nuss" procedure, is the standard of care for surgical repair of PE in children.[40,41] Initially, attempts at MIRPE for adults resulted in higher complication rates as compared to the open Ravitch repair.[3,42] With age, the chest wall becomes more rigid; therefore, elevating the sternum and supporting the repair with substernal bars is often more difficult.[2,43] Excellent results, however, have been shown with an MIRPE approach in adults; even in patients over 30 years.[3,6,8,9,44–51]

Multiple modifications of the technique Nuss first introduced for pediatrics in 1998[1] have allowed for successful extension of MIRPE into the repair of adults.[4–7,44,46,52,53] These include the following:

1) *Use of forced sternal elevation* - The force needed to elevate the sternum to normal position in adults is up to 250 N in male adults and is much larger than in pediatric patients.[8,43] Lifting the sternum anteriorly, before inserting the dissector and placing bars, decreases the force required to insert and rotate bars in the less flexible adult chest wall. This minimizes trauma to and lateral stripping of the intercostal muscles.[5,9,54–57] A variety of methods to obtain sternal elevation have been reported with the additional advantage of improved visibility and safety during mediastinal dissection and bar passage.[5,9,54–58]
2) *Use of multiple bars to support defect* – In the original description by Nuss, a single bar was recommended to be placed at the lowest point of the deformity. The median patient age in the original series was 4 years of age. In the adult chest wall, the weight and rigidity of the chest significantly increase the pressure applied to the intrathoracic bars. Two or more Nuss bars distribute the pressure and should be considered in adult patients.[4,6,8,59–62] The use of multiple bars also helps to decrease the risk of bar rotation and malposition.[4,59,60] Single-bar repairs have been shown to require reoperation for bar rotation or incomplete correction more frequently than those who had a double-bar repair.[60] In our own practice, we utilized two or more bars in 99% of patients over 18 years.[6] Additionally, bars that are shorter may reduce the risk of bar rotation.[63,64] Bars that are too long bars have been considered a risk factor for procedure failure.[65]
3) *Medial fixation and support of interspace holding pectus bar* – One of the causes of failure in the adult MIRPE is inability for the intercostal space to support the bar.[54,61,66,67] Nuss bars must remain anterior to apply force to the sternum. With lateral striping of the intercostal muscles, the bar migrates posterior-lateral and the bar falls lower into the thorax. This migration of the bar can be evident with lateral chest roentgenograms or CT imaging (Fig. 3a–d). Techniques for medial fixation include medially placed stabilizers,[7] FiberWire or sternal wire interspace reinforcement[6,9,67] a hinge reinforcement plate,[68] and sternal fixation.[5]

Hybrid procedure

Older patients have a higher risk of fracture and need of osteotomy or cartilage resection.[6,69–71] While greater than 88% of the patients in our cohort ≥ 30 years were successfully repaired with MIRPE, some did require an open procedure for successful repair.[6] Our procedure of choice is a hybrid approach that incorporates the

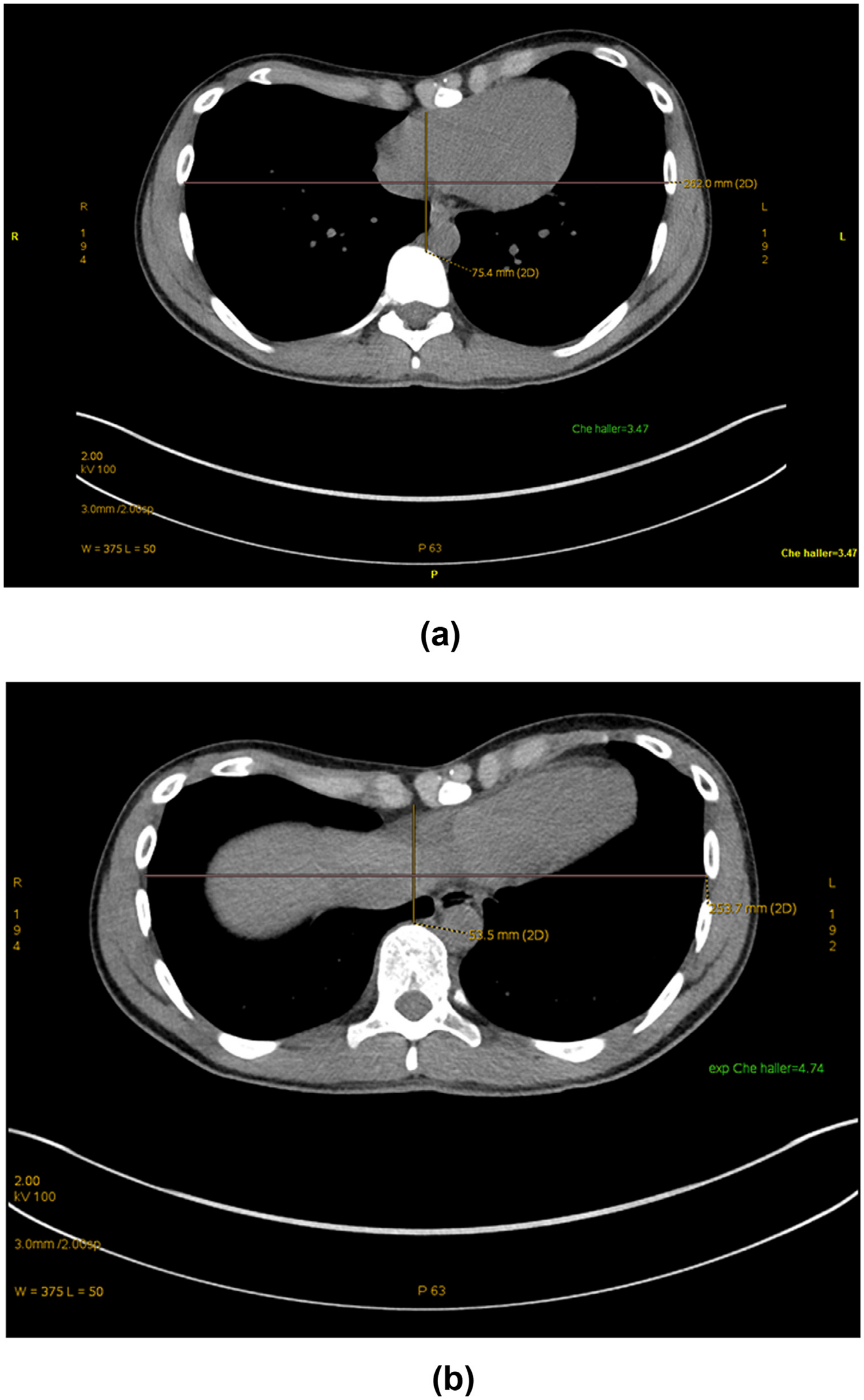


Fig. 2. Computed tomography imaging of a 21-year old male being evaluated for pectus excavatum deformity. Imaging was obtained in both (a) inspiratory and (b) expiratory views. Note the significant worsening of the cardiac compression and Haller index from 3.4 during inspiration to 4.7 in expiration.

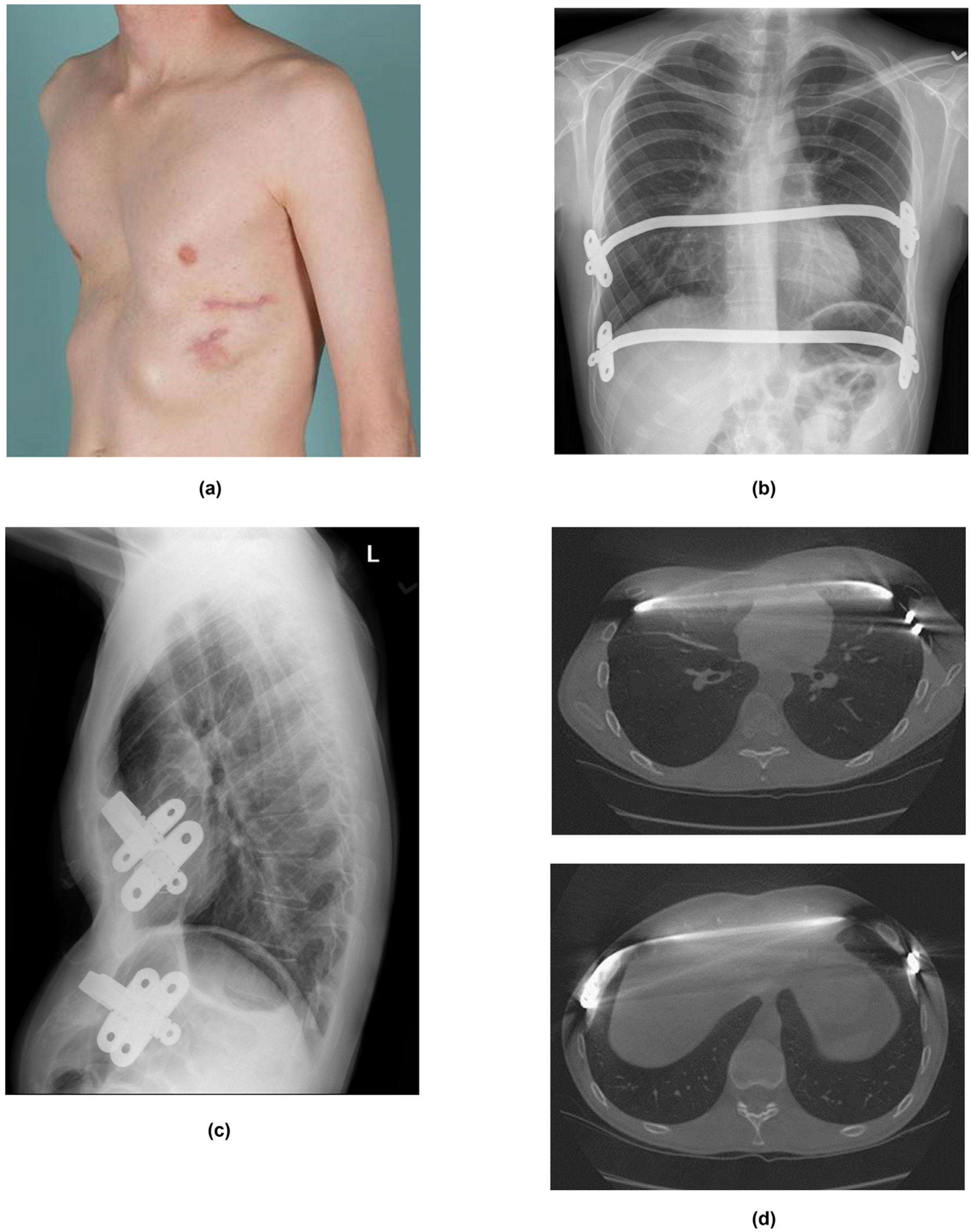


Fig. 3. (a) A 35-year old male presented for evaluation after undergoing MIRPE at an outside institution. On evaluation he had residual pectus deformity despite 2 indwelling support bars. His chest imaging confirmed failed MIRPE due to posterior-lateral migration of pectus support bars. (b) PA chest x-ray shows 2 Nuss bars with bilateral stabilizers (c) lateral imaging is critical to reveal rotation of the bars with residual pectus defect (di, ii) CT images confirm both pectus support bars with lateral intercostal exit and failure to contact the sternum for anterior elevate y.

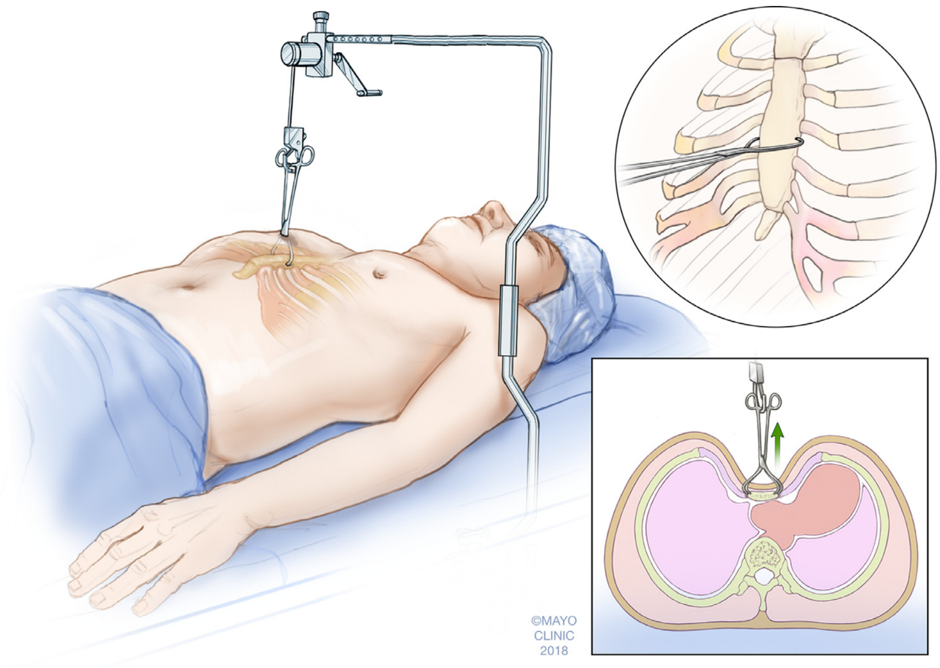
principals of intrathoracic support bars keeping the defect anterior and osteotomies with stabilization of the sternum and ribs.[6,66]

Procedure at our institution[5,6,52,67,66]

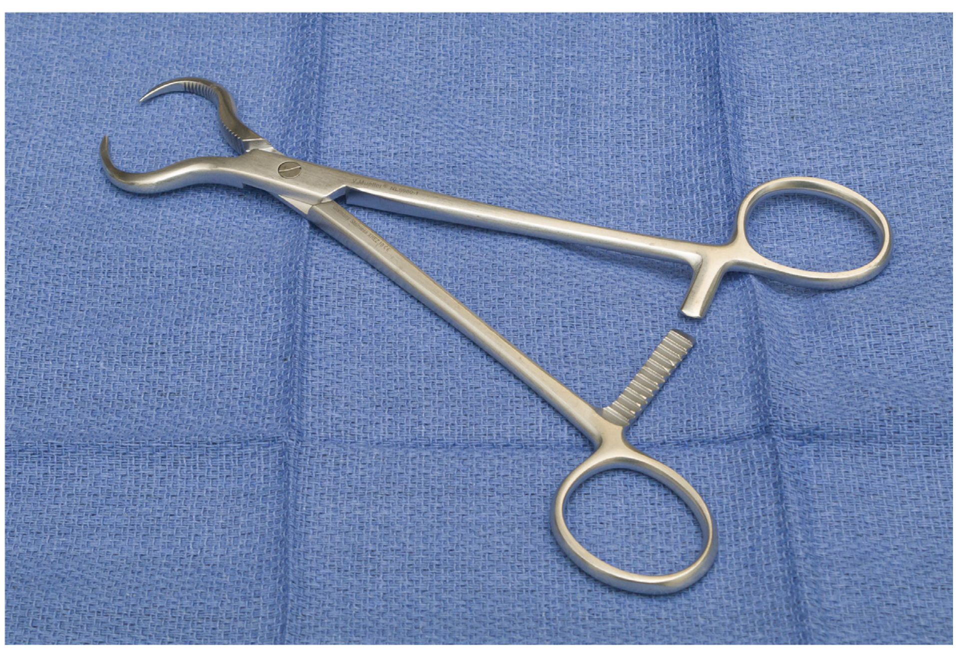
MIRPE technique

After induction of general anesthesia and placement of a double-lumen endotracheal tube for single lung ventilation, the patient is positioned supine with longitudinal gel rolls posterior and parallel to spine with the arms tucked. Patients receive antibiotic prophylaxis with intravenous cefazolin prior to the procedure.

At the lateral pectoral borders, 3-cm incisions are made bilaterally, and submuscular pockets are developed laterally and under the pectoralis muscles. Through the right incision, a thoracoscopic port is placed and carbon dioxide insufflation initiated at a pressure of 8 mmHg. Under thoracoscopic visualization, another 5-mm port is inserted on the right side superior to diaphragm. Forced sternal elevation is attempted. The Rultract retractor (Rultract Inc, Cleveland, OH) is attached to the bed on the patient's left. At the



(a)



(b)

Fig. 4. (a) Forced sternal elevation can be achieved with the use of a Rultract retractor positioned on the patient's left and attached by a clamp to the sternum. (b) A Lewin bone clamp is preferred at our institution as it allows a firm grip behind the anterior table of the sternum without having to introduce the prongs into the thorax.

center of the pectus defect, a penetrating bone clamp (Lewin Perforating Forceps [V. Mueller NL6960], CareFusion, Inc, San Diego, California) (Fig. 4a) is placed into the anterior table of the sternum. The bone clamp is attached to the Rultract retractor and the sternum elevated (Fig. 4b).[5] There are a variety of attachments available for the Rultract retractor to attach to the sternum at the defect. We prefer the basic bone clamp as the tips enter the sternum behind the anterior table and do not enter the thoracic cavity minimizing the risk of damage to the internal mammary artery.

With the cardiac compression relieved by sternal elevation, a thoracoscopic dissection is performed. Using a combination of a blunt tipped instrument, such as a laparoscopic endokittner, and electrocautery, dissection across the mediastinum is completed. The Lorenz dissector (Zimmer® Biomet®, Jacksonville, FL) is introduced into the superior interspace of defect through the right interspace and brought out through the contralateral interspace. A #5 FiberWire® (Arthrex, Inc, Naples, FL) is attached to the end of the dissector and used to guide the bar into place. The Fiberwires

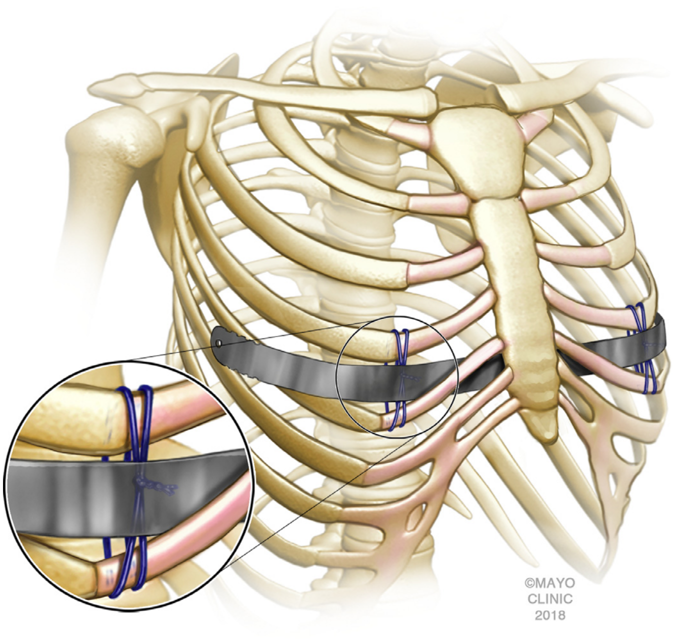


Fig. 5. Figure-of-eight FiberWire incorporating the rib above and below the interspace to reinforce the interspace medial and prevent displacement of the bar.

are temporarily crossed anterior to the sternum to keep them out of the way. Figure-of-8 FiberWire incorporating the rib above and below the interspace is used to reinforce the interspaces just lateral to the dissector entrance and exit sites to prevent later displacement of the bar (Fig. 5). Bars are then custom bent and sized to extend 2–3 cm beyond the anterior axillary line. A second bar is placed one or two interspaces below the superior bar. After placement of the second bar, if an inferior residual defect remains, a third bar may be required and is placed as above. The bars are rotated into position with the sternum still elevated to minimize intercostal rotational forces.

FiberWire® is additionally used for thoracoscopic bilateral circumferential fixation of the bars[52] in multiple sites (Fig. 5), including the central sternum. To fix the bar medially at the sternum, the Rultract® retractor and perforating bone clamp are removed. Under thoracoscopic view, a 1/8th inch drill bit with depth guard is used to make a hole in the sternum above and below the bar. A fascial suture passer is placed through the hole to retrieve the FiberWire, from inside the chest and brought out encircling both the sternum and bar and then tied on the anterior chest wall under the skin.

At conclusion of the procedure, an intercostal block is performed bilateral with bupivacaine. Through the inferior port incision, a 16-French chest tube is placed which is planned for removal the following morning. Variable flow subcutaneous continuous flow catheters are tunneled along the lateral chest wall (7.5 in. On-Q® Pain Relief System with a Select-A-Flow® Variable Rate Controller, Halyard® Health, Inc, Irvine, CA). We recommend that bars remain for a minimum of three years in adults.

Postoperative pain control in adults

Pain control is an integral part of the postoperative recovery process and are covered fully by Singhal and Jermain in this issue (pages xx-xx). Successful analgesic regimens to achieve adequate pain control after surgery have included the use of thoracic epidurals, intravenous on-demand patient-controlled anesthesia, subcutaneous continuous flow catheters and local paravertebral blocks.[72–76] Tunneled, anesthetic-infiltrating catheters are our preferred approach. They are being used increasingly after thoracic procedures in lieu of thoracic epidural analgesia (TEA) with studies showing earlier hospital discharge.[73,74,77] In our experience, subcutaneous continuous flow catheters with a standardized multimodal anesthetic protocol (including intraoperative methadone, NSAIDs, gabapentin & narcotics) achieved good pain control.[6,72,78,79]

Discussion

Repair of adults with PE is indicated when symptomatic and anatomically severe. In adults, symptoms can be resolved or improved with surgical correction.[15] Significant improvement in self-esteem, social functioning and improved levels in quality of life can also be expected with repair.[15,45,80]

A modified MIRPE technique can be utilized to safely and successfully repair most adult patients with PE. In our own series of adult patients, age did play a significant role in the difficulty of repair.[6] Older patients (≥30 years, 11%) were more likely to require open osteotomy or chondroplasty versus younger (18–29 years, 3%). Failure to lift with forced sternal elevation due to fixation of the sternum or costochondral attachments and sternal fracture with forcing elevation were common causes for requiring the addition of an open procedure in our cohort. Looking back at adult revision cases performed for failed MIRPE has also been helpful to develop modifications that may contribute to a successful repair. In a review of 47 adults that underwent revision from prior failed MIRPE, 98% were due to inadequate correction after initial repair as opposed to regression.[67] The majority of patients (66%) still had bars in place when evaluated at our institution. Successful MIRPE revision utilizing the technical modifications presented in this manuscript allowed MIRPE alone in 83% of these patients.[67] In our experience, failure of MIRPE in adults has been predominantly caused by intercostal stripping of the interspace supporting the bar which results in posterior and lateral bar migration. The weight and rigidity of the adult chest wall exacerbates this problem. Reinforcement of the interspace or medially placed stabilizers help prevent stripping of the interspace and posterior-lateral migration of the bars. In our experience, this and sternal elevation were the most critical technical modifications necessary to facilitate success in adults.

Conclusions

Adult patients may present with new-onset or progressive symptoms from pectus excavation. Repair should be recommended if cardiopulmonary compromise is identified in the setting of an anatomically severe PE. With the addition of technical modifications, an MIRPE approach can be used to achieve excellent correction with low complication rates in adults. Sternal elevation, use of multiple bars, medial stabilization, and reinforcement of the interspaces improve success in the rigid adult chest wall and should be considered to decrease the risk of bar migration and failed support. Overall, adult patients can expect improvement in symptoms and high satisfaction following repair of their PE.

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