Evaluation and Surgical Treatment of Adult Pectus Excavatum Patients

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Abstract

Pectus excavatum is one of the most common congenital chest wall deformities, in which the sternum and the adjacent costal cartilages are displaced posteriorly. Physiological limitations and psychological impacts of the deformity may occur as the patient ages and the chest wall becomes more rigid. Adult patients can undergo surgical correction of their deformity with excellent results. A minimally invasive approach to the repair of adult patients can be successfully utilized.

Introduction

The two most common congenital chest wall deformities are pectus excavatum (PE) and pectus carinatum [1-3]. Although in the past physicians considered PE primarily an aesthetic problem, studies continue to document physiologic limitations caused by the deformity [4-8]. Significant improvement in cardiopulmonary function and exercise ability can be achieved with surgical correction [4,6,7,9-13]. In most patients the development of PE tends to be more prominent during periods of rapid skeletal growth with stabilization after reaching skeletal maturity [14]. Most patients begin experiencing symptoms in their adolescence, however, some may not develop symptoms until adulthood as the chest wall becomes more rigid [7,15-16]. Both open and minimally invasive techniques have been described for repair of adult PE with the modified Nuss procedure becoming a more common repair method with experience and technique modifications [7,15,17-26] (Figure 1).

Key Points

• Comprehensive history, physical examination and investigations are necessary for evaluation of PE repair.
• Symptomatic adult patients with evidence of cardiac compression are candidate for surgical repair.
• Modified Nuss procedure has been performed in adult patients with successful results.
• Some form of open repair is required only for a small percentage of more complex and recurrent PE cases.
• Female patients can have their deformity repaired along with breast implants during the same surgical procedure.

Presentation of Adult Pectus Patients

The age of presentation of PE varies. Although it can be present at birth, most patients present during their childhood period or teenage years [7,27]. PE can be a cosmetic problem with minor symptoms however, it can also present with compromise in cardiopulmonary function [4,6]. Symptoms such as shortness of breath, palpitations, chest pain and fatigue may become more prominent with advancing age due to the decrease in the chest wall compliance with subsequent increase in cardiac compression [7,16,28-29]. Kraagten et al. [16] reported the majority of his adult patients did not have symptoms occur until the 4th and 5th decades. Most commonly reported symptoms include progressive shortness of breath, tachycardia/palpitations, exercise intolerance and chest pain [16,27-30]. There is an association in some patients between connective tissue disorders such as Marfan, Ehlers-Danlos, and Noonan syndromes and PE [30-31].

Cardio-pulmonary Implications of Pectus on Adults

Studies have proven that severe PE can cause significant compression to the right heart chambers [4,29,32]. Displacement of the heart into the left chest may occur to varying degrees and significant...
Significantly narrower at the aortic root level and right ventricle end-diastolic and end-systolic areas were larger in controls. The study also showed significant morphological variations in the RV of PE patients compared to the control group, there was a higher incidence of RV with rounded apex, trabecular hypertrophy, and changes in the structure of moderator band were seen as well, as RV wall sacculation [34]. In a recent retrospective study conducted for adult patients who underwent surgical repair for their PE deformity using a modified Nuss procedure from 2011 to 2014, Chao et al. [4], reported significant increase in the RV outflow tract (16.2%), cardiac output (38.3%) and in the RV stroke volume (33.6%) in comparison to the preoperative values.

Cardiopulmonary exercise testing (CPET) is used to assess the exercise capacity and evaluate the physiological impairments which can result as a consequence of the pectus deformity [35-36]. Limitations in cardiopulmonary functions have been reported in PE patients with abnormally low peak oxygen uptake (VO2) [L/min] and O2 pulse (mL/beat) during exercise testing [9,12]. Fick equation is used to calculate the VO2: VO2 = cardiac output (CO)x difference between the oxygen content in the arterial and the venous blood ([a-v]O2), while the O2 pulse is used to determine the stroke volume during exercise testing [36]. In PE patients, one or more of these parameters can be affected which then lead to decline in the exercise endurance [36]. The patient’s standard exercise history must be documented during assessment. Cardiac output, which is the product of the stroke volume (SV) and the heart rate (HR), can decrease with inactivity [37]. Therefore it is important to take into consideration the patient’s exercise history as patient’s activity may decrease after surgery, which then can affect the measurements of the VO2 [36]. The effects of surgical correction have been debated with variable results published [6,9,10,38]. Neviere et al. [9] reported a significant increase in exercise ability and in the VO2 and O2 pulse at 1 year follow up in 70 patients who underwent surgical repair of PE compared to their preoperative measurements. Sigalet et al. [6] also found significant cardiopulmonary function improvement at >3 months following the removal of the pectus bars. There were improvements in VO2 max, O2 Pulse and respiratory quotient in comparison to the preoperative measurements. No long-term results have been published and a more recent mid-term evaluation by Udholm et al. [38] did not show any significant improvement in the cardiac output or in the VO2 max in adult patients 1 year after repair, however they previously reported normalization of the cardiopulmonary function in teenagers patients at three years following surgical repair [10].

There have been several Meta-analyses performed on cardiopulmonary changes after surgery with conflicting results. Malek et al. [39] included eight studies in their literature review and they reported significant improvement in cardiovascular function which increased by more than a half standard deviation after surgical repair of PE. Several parameters were used for assessing the cardiovascular function in these studies, such as the cardiac output, heart rate, stroke volume, ejection fraction, VO2 max and O2 Pulse. No evidence suggested publication bias or missing small studies with negative results in their Meta-analysis. Maagaard et al. [40] reported in their recent literature review that the majority of the articles that studied the exercise capacity showed an increase in the exercise capacity as shown either in the increased peak oxygen uptake or the oxygen pulse with only one study which showed a significant decrease. This could be explained by the short 3 month post-operative follow up period with decreased physical activity following repair. The wide variability in the included studies in terms of the surgical techniques, number/
age of the patients, the measured endpoints, the follow up period and the absence of matched group in most of the studies should be taken into consideration [40]. All of these analyses supported the need for surgical repair of PE deformity and contradict what has been thought previously that PE is merely a cosmetic problem [41].

**Indications for Surgical Intervention**

A comprehensive history, physical examination and investigational studies should be done to rule out other potential causes that might contribute to the patients’ symptoms, and to evaluate the necessity of surgical repair. More common symptoms of exercise intolerance, dyspnea with exertion and chest pain in adult patient with PE can be caused by a number of serious medical conditions that must be ruled out in the differential. In adults of advanced age, investigation for coronary disease may be prudent. Diagnostic studies include echocardiography, electrocardiography, cardiopulmonary exercise testing (CPET) and computerized tomography (CT) scan or magnetic resonance imaging (MRI) [28,42]. The severity index or Haller index is measured by dividing the maximum internal width of the chest by the distance between the sternum posteriorly and vertebral body anteriorly [43]. It is recommended to assess the deformity both in the inspiratory and the expiratory views, as when patient exhales the chest becomes more compressed and the deformity becomes more evident [44]. Another measurement has been used to assess the severity of pectus deformity, the correction index which estimates the percentage of correction necessary to normalize the chest wall and may be a more valid assessment in patients whose chest is barrel in shape and their Haller Index subsequently is falsely low [45,46].

**Table 1:** A summary of some published results of Adult PE patients with minimally invasive repair.

<table>
<thead>
<tr>
<th>Author</th>
<th>Numbers of adult patients</th>
<th>Median age (y)</th>
<th>Male (%)</th>
<th>Median operative time (min)</th>
<th>Numbers of bars (%)</th>
<th>Median postoperative stay (days)</th>
<th>Complications n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pileegard [17]</td>
<td>151</td>
<td>Mean: 40.4</td>
<td>108 (71.5)</td>
<td>Mean*: 121</td>
<td>2 (55), 3 (44.4), 4 (0.7), 1 (0.9), 2 (56.5), 3 (42.6)</td>
<td>Mean*: 2010-2012: 5 2013-2015: 3.3</td>
<td>Deep infection after re-operation caused by lateral bar dislocation 1 (2)</td>
</tr>
<tr>
<td>Jaroszewski et al. [15]</td>
<td>115</td>
<td>Mean: 23.7</td>
<td>88 (76.5)</td>
<td>Mean*: 111</td>
<td></td>
<td>Mean*: 2010-2012: 4 2013-2015: 3.1</td>
<td>*Bar rotation 2 (1.7), infection 1 (0.9), pleural effusion required thoracostomy3 (2.6), pneumothorax required chest tube insertion 1 (0.9), reoperation for bleeding 2 (1.7)</td>
</tr>
<tr>
<td>Esteves et al. [20]</td>
<td>19</td>
<td>Mean: 22.5</td>
<td>12 (63)</td>
<td>Mean: 96.5</td>
<td>1 (95), 2 (5)</td>
<td>Mean: 6.1</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Yoon et al. [21]</td>
<td>44</td>
<td>Mean: 20</td>
<td>38 (86.4)</td>
<td>90</td>
<td>1 (63.8), 2 (36.2)</td>
<td>3</td>
<td>Reoperation to correct bar rotation 2 (4.5), and incomplete repair 1 (2.2), Pneumothorax required thoracostomy 2 (4.5), wound infection required surgical revision 2 (4.5), Pneumopericarditis and pericardial effusion 1 (2.2)</td>
</tr>
<tr>
<td>Olbrecht et al. [22]</td>
<td>52</td>
<td>Mean: 23</td>
<td>42 (80.8)</td>
<td>82</td>
<td>1 (94.2), 2 (5.8)</td>
<td>3</td>
<td>Pneumothorax required tube thoracostomy 2 (3.9), superficial site infection 1 (21.6%), 3 of them (5.9) required surgical revision and 2 (3.9) required early bar removal, bar displacement required repair 4 (7.7), upper sternal depression required 2nd bar insertion 2 (3.9), development of pectus carinatum required repair 1 (1.9), recurrence of PE occur in 2 (3.9) patients none of them required repair</td>
</tr>
<tr>
<td>Schalmon et al. [23]</td>
<td>43</td>
<td>Mean: 22</td>
<td>39 (91)</td>
<td>Mean: 70</td>
<td>1 (81), 2 (19)</td>
<td>Mean: 9.3</td>
<td>Puncture of a pleural effusion 5 (12), pneumothorax required drainage 2 (5), bar displacement with no required repair 1 (2)</td>
</tr>
<tr>
<td>Hebra et al. [24]</td>
<td>30</td>
<td>Mean: 23</td>
<td>NR (75)</td>
<td>60% operative time: 1 to 2 hours</td>
<td>2 (16)</td>
<td>Mean: 6</td>
<td>Seroma (10%), bar displacement (6%), pneumothorax requiring tube thoracostomy (6%), superficial wound infection (3%), and stabilizer bar fracture (3%). Two patients required conversion to modified Ravitch repair.</td>
</tr>
<tr>
<td>Aronson et al. [25]</td>
<td>35</td>
<td>Mean: 23</td>
<td>28 (80)</td>
<td>65</td>
<td>2 (3)</td>
<td>7</td>
<td>Pneumothorax 9 (26), wound abscess in 3 (9), and bar displacement 5 (14).</td>
</tr>
<tr>
<td>Cheng et al. [26]</td>
<td>96</td>
<td>Mean: 24.5</td>
<td>80 (83)</td>
<td>80</td>
<td>2 (22.9)</td>
<td>Mean: 7.2</td>
<td>Pleural effusion 1 (1) required needle thoracostomy Pericardial effusion 1 (1) required pericardiocentesis and steroid 2 months postoperative, surgical revision for bar displacement 2 (2.6)</td>
</tr>
</tbody>
</table>

*151 patients, 30-72 years of age.  
*115 patients, 18-29 years of age.  
*Information on patients underwent minimally invasive repair of pectus excavatum.  
NR: not reported
is measured by calculating the distance between the spine anteriorly and most anterior internal rib (b) at its maximum point from the least distance between the spine anteriorly and the sternum posteriorly (a). The used equation is \((b-a)/b \times 100\) [45,46]. Typical surgical candidates are symptomatic with evidence of cardiac compression on echocardiography or CT scan/MRI studies, have abnormal CPET and Haller index >3.2 or correction index >10% [43,47].

### Surgical Therapy

The Ravitch is an invasive procedure that has been used for PE repair for many decades. The procedure involves open resection of varying degrees to the deformed costal cartilages [48]. A number of modifications have been introduced to the Ravitch technique which included decreasing the amount of cartilages being resected, preserving the perichondrium, and the use of a metal strut or mesh posterior to the sternum for anterior support [18,49]. A minimally invasive repair approach to PE was introduced for children and adolescents in 1998 [50]. The Nuss procedure involved temporary placement of substernal support bar to elevate the deformity for a minimum of 2 years and removal of the bar as an outpatient procedure without the morbidity and long term consequences of cartilage resection [17,28].

### MIRPE for Adult Patients

Although initially thought to be applicable only to young patients, a minimally invasive repair of pectus excavatum (MIRPE) is being increasingly used for adult patients [13,15,17,19-26]. An MIRPE can be performed in the majority of adults with excellent results and success [15,17] (Table 1). Modifications to the Nuss procedure have been introduced to minimize the higher risks of complications that were initially published in adults [24,51,52]. These modifications included the use of forced sternal elevation to facilitate the introduction of the pectus support bars with less rotational force and allowance of better visualization across the mediastinum [53], use of multiple support bars to distribute the force needed for the chest elevation [15,52] and improved methods to secure the bars to prevent rotation [15,19,54,55]. Pilegaard [55] recommended the use of a shorter bar and amulti-point fixation technique has also been used by Park et al. [52]. In our clinical practice, the use of multipoint Fiber Wire fixation of the bars bilaterally and reinforcement of the interspaces to avoid intercostal muscle stripping has greatly reduced the incidence of bar rotations [15,54].

Others have noted that multiple bars help balance the increased pressure of supporting the adult chest wall [52,56,57]. Several studies have shown increased need for multiple bars in older patients, 70% in patients over 30 years required 2 or more bars as reported by Pilegaard [17]. In our own practice, we utilized 2 or more bars in over 99% of patients over age 18 [15]. Some surgeons have advocated the Ravitch in older patients [18,49]. In our experience, over 88% of the patients over 30 years were successfully repaired with MIRPE however some required a hybrid procedure (described to follow) in which addition to pectus support bars, an open resection for fracture or osteotomy was needed to achieve adequate repair [15]. The length of hospital stay has been reasonable with most series reporting 3-9 days of hospitalization and a low percentage of complications (Table 1) [15,17]. Achieving adequate postoperative analgesia is one of the concerns after Nuss procedure especially in adult patients. Thoracic epidural, intravenous on demand patient administrated narcotics, and subcutaneous continuous flow catheters and local blocks have all been reported [58-62]. For our patients, a standardized polypharmacy protocol is followed with good results [15,58] and includes gabapentin, non-steroidal, local subcutaneous catheter delivery and narcotics [58].

### Open Hybrid Repair for Complex, Difficult Adult Patients

Asymmetric, calcified and complex deformities may require an open procedure for cartilage resection, osteotomy or repair of fractures with attempted elevation. A hybrid procedure incorporating both principals of osteotomy cuts and pectus support bars may also be considered and is our procedure of choice for these difficult deformities [15] (Figure 3). With age, the chest wall becomes less compliant and the force needed to elevate the deformity is increased [63]. Failure to elevate the sternum or its fracture during attempt elevation usually necessitates the use of an open procedure to correct the defect adequately [15,64]. Patients with severe recurrent deformities, extensive ossification and/or chest wall rigidity or defect severity that prevented elevation underwent a hybrid repair at our institution [65,66]. Extensive and more complex repair is associated with an increase in complications and length of hospital stay [15]. A recently published meta-analysis compared between the Ravitch and the Nuss procedures though it reported a higher complication rate, mostly early complications, in the adult patients underwent Nuss repair, only 90 adult patients were included in the analysis and they emphasized the importance of the availability of long term data to better compare the two procedures [67].

Patient should be counseled about the different surgical approaches for pectus repair. Age increases the difficulty of the repair and surgeon experience with complex cases and older patients is critical to successful outcomes [51,68-70].

### Adult Female Patients and Implants

PE is generally thought to be more common in men than women [71]. There are few publications that focused primarily on the PE repair in female patients [72-74]. Breast development can often conceal the PE deformity additionally some women have implants placed to mask their deformity. Symptoms from limitation of cardiopulmonary function may develop in later years. Augmentation together with correction of the chest wall deformity can be performed without significant complications and should be considered in patients who desire so [72,75].

### Conclusion

Adult patients may present with new onset or worsening...
symptoms from their pectus excavatum. Complete preoperative investigation should be performed to rule out other causes and surgery is offered if medically indicated. Surgical repair has been shown to relieve symptoms and improve cardiopulmonary function in adult patients after repair. Both open and minimally invasive techniques are options. Both patient choice as well as surgeon expertise should be considered in determining the best option for every individual patient repair.

References


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