Operative Management of Acquired Thoracic Dystrophy in Adults After Open Pectus Excavatum Repair

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**Background.** In young children, acquired thoracic dystrophy (ATD) is associated with extensive resection of cartilage, often during open pectus excavatum (PE) repair. Progressive dyspnea or exercise intolerance may develop in these patients secondary to cardiac compression or restrictive pulmonary function. Surgical treatment of ATD by attempting to increase the overall thoracic volume has been controversial. We describe our experience with adults presenting for surgical correction of ATD.

**Methods.** A retrospective medical record review was performed for all patients with ATD presenting for surgical evaluation from December 2010 through February 2013.

**Results.** Ten adult male patients were evaluated for treatment of ATD after an open Ravitch procedure for PE. Nine patients, whose mean age was 34 years (range, 21-42 years), elected to proceed with surgical treatment. The mean age of the initial repair was 3.7 years. Extensive reconstruction, chest wall expansion, and placement of stainless steel support bars and titanium plating were performed in all patients. Eight patients had minor complications, and major complications occurred in 3 patients. Respiratory failure with prolonged ventilator support occurred in 3 patients. There were no reoperations or deaths. At mean follow-up of 16 months (range, 6-31 months), all patients subjectively reported improvement in their ability to exercise and in their symptoms, including dyspnea with exertion.

**Conclusions.** ATD may be associated with early childhood Ravitch repair. Adults may present with disabling symptoms related to cardiac compression and restrictive pulmonary function. Reconstruction with sternal elevation and expansion of the anterior chest subjectively improves symptoms.

(Early aggressive open repair of pectus excavatum (PE) in young children (before the age of 7 years) using the Ravitch technique or other modified open repair technique has resulted in the development of a hypoplastic abnormally developed thorax in some patients (Fig 1) [1–3]. Acquired thoracic dystrophy (ATD) is thought to occur after extensive resection of rib cartilage in young children during PE repair [2, 3]. Damage to or resection of the cartilage growth plates subsequently prevents normal growth and maturity of the bony chest wall. Progressive dyspnea and exercise intolerance secondary to cardiac compression and restrictive pulmonary function may develop in these patients [1, 3–5] Surgical treatment of ATD, which attempts to increase the overall thoracic volume, has been controversial [6–12]. Reconstructive techniques have been described to treat this disorder in children and often involve staged procedures that allow for progressive chest expansion as the children grow [9, 10, 12]. ATD is rare in adults, and treatment is primarily aimed at decreasing symptoms by relieving cardiac compression and increasing thoracic volume. Long-term chest wall stability and healing are concerns. We describe our experience with adults presenting for surgical correction of ATD.

**Patients and Methods**

**Patients**

Patients presented to the Division of Cardiothoracic Surgery, Mayo Clinic in Arizona in conjunction with our chest wall reconstruction program. Mayo Clinic Institutional Review Board approval was obtained for a retrospective review of all patients presenting for treatment of ATD from December 2010 through February 2013. All patients were identified and diagnosed as having ATD by a history of a previous Ravitch-type open procedure for correction of PE at a young age, with subsequent abnormal chest wall growth and restriction. Operative records for the childhood operations were not available in

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the majority of patients. Thus precise details of surgical procedures and extent of cartilage resection previously performed could not be analyzed. All patients had undergone a medical evaluation, including at least 2 of the following: computed tomography or magnetic resonance imaging of the chest, cardiopulmonary exercise testing (CPET), echocardiography, pulmonary function testing, and cardiac catheterization. Cardiopulmonary exercise stress testing was performed using a standard bicycle ergometer with an incremental 25 W/min protocol. Patients who showed evidence of cardiac limitation underwent preoperative echocardiographic assessment. Intraoperative transesophageal echocardiography was performed in all patients, and images for 7 of the 9 patients were available for review and analysis by 2 separate cardiologists. Statistical analysis was performed for preoperative and postoperative echocardiographic variables using a paired $t$ test and sign test.

**Operative Procedure**

A cardiac-certified anesthesiologist and cardiac operating room team took part in all procedures. All patients had thoracic epidural anesthesia preoperatively and received 0.2% ropivacaine or 0.25% bupivacaine. Narcotic agents were used during the operation and for postoperative pain control. Intraoperative transesophageal echocardiography was performed throughout the procedure. Arterial blood pressure monitoring was performed in all patients, and central venous access was established if needed.

Patients were positioned supine with 2 rolls placed vertically along the paraspinal area. Arms were tucked at
the sides. Prophylactic cephazolin was administered intravenously before the skin incision. Chlorhexidine gluconate (ChloraPrep; CareFusion, Leawood, KS) was applied to the entire anterior thorax, anterior axillary space, and abdomen. Sterile draping was placed to allow for complete exposure of the thorax and axilla. An antimicrobial drape (3M Ioban 2 Antimicrobial Incise Drape; 3M, Minneapolis, MN) was applied to secure the position of the towels.

Incisions were made using the scar from the previous open PE repair and were extended when necessary. Dissection down to the bony chest wall and elevation of the pectoral muscles bilaterally were performed. The upper portion of the rectus abdominis muscle was also freed from the lower chest wall inferiorly. After the muscle was elevated from the chest wall, the extent of the deformity was evaluated.

The objectives for repair were both correction of the recurrent PE depression by anterior elevation of the chest wall defect and expansion of overall chest volume. Because of significant scar tissue and fixed bony deformities, multiple transverse rib osteotomies, both medially at the sternal attachments and laterally, were necessary. Once all abnormal ribs and sternocostal sites were cut, the sternum was elevated using a Lorenz bar passer followed by placement of stainless steel bars (Biomet MicroFixation, Jacksonville, FL). The bars were placed through the intercostal spaces in a fashion similar to that used in the Nuss procedure [13, 14], with the bar placed under the sternum and affixed laterally to the ribs. Additional fixation was performed to hold the chest wall in the anterior position. Braided composite suture (FiberWire No. 5; Arthrex, Naples, FL) was used for circumferential attachment of the bars to the sternum and lateral ribs at multiple sites bilaterally.

Bridging spaces between the medial sternum and the lateral rib/cartilage were created between osteotomy sites to increase the overall dimensions of the anterior chest in both the anteroposterior and lateral aspects. Titanium plating (Synthes, West Chester, PA) was then used to plate across, stabilize, and secure to adjacent chest wall and sternum (Fig 2A). The sternal osteotomy site of the anteriorly elevated sternum was also plated with titanium sternal plate. Polyglactin 910 woven mesh (Vicryl; Ethicon, Somerville, NJ) was attached posteriorly as vertical strips to bridge the gaps and allow a platform to contain graft. Bone graft paste (Grafton Demineralized Bone Matrix; BioHorizons USA, Birmingham, AL) was injected into the spaces (Fig 2B).

In 1 patient, there was a large area of absent bony chest wall with lung herniation that was felt to be significant

### Table 1. Preoperative Characteristics of Patients With Acquired Thoracic Dystrophy

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Mean</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at initial Ravitch procedure (y)</td>
<td>3.7</td>
<td>4–6</td>
</tr>
<tr>
<td>Age of ARD reconstruction procedure (y)</td>
<td>34</td>
<td>22–42</td>
</tr>
<tr>
<td>Preoperative pulmonary function test results (% of predicted normal value)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>FVC</td>
<td>50</td>
<td>22%–50%</td>
</tr>
<tr>
<td>FEV₁</td>
<td>51</td>
<td>22%–52%</td>
</tr>
<tr>
<td>MVV</td>
<td>53</td>
<td>21%–61%</td>
</tr>
<tr>
<td>Recurrent pectus excavatum index</td>
<td>4.26</td>
<td>2.8–7.0</td>
</tr>
<tr>
<td>Functional capacity</td>
<td>&lt; 75% predicted</td>
<td>&lt; 50% to &lt; 75% predicted</td>
</tr>
<tr>
<td>VO₂ at anaerobic threshold</td>
<td>&lt; 40% predicted</td>
<td>&lt; 30% to &lt; 50% predicted</td>
</tr>
</tbody>
</table>

ARD = acute respiratory distress syndrome; FEV₁ = forced expiratory volume in 1 second; FVC = forced vital capacity; MVV = maximum voluntary ventilation; VO₂ = oxygen consumption.
enough to require the defect be filled. To enhance stability, antibiotic-impregnated (1 g each of vancomycin, tobramycin, and cefazolin) bone cement (Palacos polymethyl methacrylate [PMMA] bone cement; Zimmer Surgical, Dover, OH) was mixed and allowed to harden for 2 minutes. While still malleable, the bone cement mixture was fashioned to fit into the chest wall defect with overlap of the bordering bony structures (“molding”). Titanium plating was compressed into the PMMA before it completely hardened. It was then removed and allowed to completely mature for another 20 minutes. While still malleable, the bone cement mixture was fashioned to fit into the chest wall defect with overlap of the bordering bony structures (“molding”).

Titanium plating was compressed into the PMMA before it completely hardened. It was then removed and allowed to completely mature for another 20 minutes. The hardened implant was the placed into the defect and attached to the sternum and lateral ribs. Titanium plating (Synthes) was then used to plate across, stabilize, and secure the implant to adjacent chest wall and sternum. Drill holes were made to allow screws of sufficient length to be placed, incorporating both the PMMA and underlying bony support structures (Fig 2C). Biological mesh sheeting was then placed across the chest wall in the majority of patients to cover the reconstruction completely and was secured to the bony structures with polyglactin 910 suture. The pectoral and rectus abdominis muscles were then brought back across the chest and secured to the midline and lower chest wall using nonabsorbable braided suture (Ethibond; Ethicon) (Fig 2D). Drains were left under the muscle flaps and removed when output was less than 30 mL/24 h. Chest tubes were placed bilaterally and the incisions were closed with layered absorbable suture.

Results
Ten adult male patients were diagnosed with and evaluated for ATD during this period. An extensive discussion regarding the risks of surgical intervention was undertaken with each patient. One patient elected not to proceed with an operation. Nine underwent surgical correction of their ATD. Patient characteristics are listed in Table 1. The mean age at which the initial Ravitch procedure was performed was 3.7 years (range, 4–6 years). At the time of surgical reconstruction for their ATD, the mean age was 34 years (range, 22–42 years), with a recurrent PE Haller severity index of 4.26 (range, 2.8–7.0). Because of the severe chest deformity and hypoplasia, there was not a clear correlation between the Haller index and severity of the deformity. All patients...
reported symptoms of dyspnea with even minimal exertion as well as exercise intolerance (often interfering with activities of daily living). One patient required oxygen supplementation (2-4 L/min) for ambulation. Pulmonary functions were generally consistent with severe restrictive disease. Appearance on physical examination was consistent with thoracic hypoplasia and included an abnormal thoracic inlet with prominent clavicle and manubrium (Fig 3). A small, deformed, and rigidly immobile anterior chest wall with primary diaphragmatic breathing was uniformly present. All patients had scars from previous Ravitch-type procedures for PE deformity performed in early childhood. One patient had undergone a second operative attempt at correction. Computed tomography or magnetic resonance imaging of the chest was consistent with small hypoplastic chest walls, narrow anteroposterior diameters, and compression or displacement (or both) of the heart into the left thorax (Fig 1). Some level of recurrent PE deformity was seen in all patients. Intraoperative transesophageal echocardiography was performed both after induction of anesthesia and at completion of surgical correction, with confirmation of relief of cardiac compression (Fig 4). Preoperative functional assessment with CPET confirmed extensive functional limitations related to both respiratory and cardiac aspects. All patients tested had exercise capacity less than 75% of that predicted for their age and sex, and 3 of 9 had functional capacity less than 50% of predicted. Preoperative and postoperative echocardiographic results are presented in Table 2.

Nine patients had surgical correction (Table 3). All patients underwent release of the sternum from the lateral scar tissue and calcified costosternal attachments. All but 1 patient had abnormal lateralized attachment of these muscles, and advancement flaps were necessary to allow closure and reattachment to the lower chest wall. Multiple vertical osteotomies of the anterolateral chest wall were made, and at least 2 stainless steel support bars (1 patient had 3 support bars) were placed behind the sternum through intercostal incisions. In 1 patient, a large portion of the anterior bony chest wall was absent. Reconstruction of the absent segment using MMR was performed as described in the Methods section. The mean bar length was 11.5 inches. All patients had platting of the anterior chest with titanium plating. Biological mesh sheeting (7 patients—XCM Biologic Tissue Matrix, Synthes; 1 patient—AlloDerm Tissue Matrix, LifeCell, Bridgewater, NJ) was then placed across the chest wall to cover the reconstruction completely and was secured to the bony structures with polyglactin 910 suture. Estimated blood loss was a median of 358 minutes (range, 246-526 minutes). Estimated blood loss was a median of 700 mL (range, 400-1,000 mL).

Serious complications included prolonged intubation (3 patients) with subsequent temporary tracheostomy (2 patients) and vasodilatory shock (2 patients) with right-sided heart failure (2 patients) requiring vasopressor and inotropic support. Table 4 summarizes major and minor complications. Two patients with prolonged respiratory failure had evidence of pneumonia that was treated with antibiotic therapy. There were no short- or long-term wound infections. No bar migrations, evidence of chest wall instability, or malunion has occurred. Hospitalization was a mean of 10.8 days (median, 8 days;
range, 6–24 days), with mean intensive care time of 6 days (median, 3 days; range, 2–18 days). No deaths occurred.

Mean follow-up was 16 months (median, 13.4 months; range, 6–31 months). All patients reported a subjective improvement in preoperative symptoms and ability to exercise. Intraoperative transesophageal echocardiography showed significant improvement in the right ventricular chamber size and ejection function in all patients (Table 2). Only 2 patients underwent postoperative testing at follow-up of more than 1 year after surgery. These included pulmonary function tests and CPET. The results did not show a notable improvement despite the patient’s subjective observance of symptomatic improvement. No patients had reoperation or removal of their stainless steel support bars. Bar removal is planned for 3 to 4 years from the date of operation.

### Table 3. Patient Operative and Hospitalization Information

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age (y)</th>
<th>Operative Time (min)</th>
<th>Estimated Blood Loss, mL</th>
<th>No. of Support Bars</th>
<th>Mean Length of Support Bars (inches)</th>
<th>Sternal and Anterior Expansion Plating</th>
<th>Mesh Coverage</th>
<th>Prosthetic Re Construction</th>
<th>ICU Time (d)</th>
<th>Length of Stay (d)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>34</td>
<td>246</td>
<td>600</td>
<td>3</td>
<td>11.5</td>
<td>Yes</td>
<td>AlloDerm a</td>
<td>None</td>
<td>2</td>
<td>7</td>
</tr>
<tr>
<td>2</td>
<td>42</td>
<td>438</td>
<td>600</td>
<td>2</td>
<td>10.0</td>
<td>Yes</td>
<td>XCM b</td>
<td>None</td>
<td>18</td>
<td>24</td>
</tr>
<tr>
<td>3</td>
<td>22</td>
<td>395</td>
<td>400</td>
<td>2</td>
<td>10.5</td>
<td>Yes</td>
<td>XCM</td>
<td>None</td>
<td>5</td>
<td>9</td>
</tr>
<tr>
<td>4</td>
<td>30</td>
<td>317</td>
<td>750</td>
<td>2</td>
<td>12.0</td>
<td>Yes</td>
<td>XCM</td>
<td>None</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>5</td>
<td>21</td>
<td>307</td>
<td>1,000</td>
<td>2</td>
<td>11.5</td>
<td>Yes</td>
<td>XCM</td>
<td>None</td>
<td>3</td>
<td>8</td>
</tr>
<tr>
<td>6</td>
<td>41</td>
<td>358</td>
<td>700</td>
<td>2</td>
<td>11.5</td>
<td>Yes</td>
<td>None</td>
<td>None</td>
<td>15</td>
<td>21</td>
</tr>
<tr>
<td>7</td>
<td>39</td>
<td>316</td>
<td>700</td>
<td>2</td>
<td>12.0</td>
<td>Yes</td>
<td>XCM</td>
<td>None</td>
<td>2</td>
<td>7</td>
</tr>
<tr>
<td>8</td>
<td>31</td>
<td>526</td>
<td>600</td>
<td>2</td>
<td>11.5</td>
<td>Yes</td>
<td>XCM</td>
<td>PMMA</td>
<td>6</td>
<td>9</td>
</tr>
<tr>
<td>9</td>
<td>41</td>
<td>403</td>
<td>700</td>
<td>2</td>
<td>11.5</td>
<td>Yes</td>
<td>XCM</td>
<td>None</td>
<td>2</td>
<td>6</td>
</tr>
</tbody>
</table>


ICU = intensive care unit; PMMA = polymethyl methacrylate.

### Table 4. Major and Minor Complications Occurring After Reconstructive Operations for Acquired Thoracic Dystrophy

<table>
<thead>
<tr>
<th>Complication</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Major</td>
<td></td>
</tr>
<tr>
<td>Prolonged respiratory failure requiring ventilator support</td>
<td>3</td>
</tr>
<tr>
<td>Tracheostomy for ventilator weaning</td>
<td>2</td>
</tr>
<tr>
<td>Ventricular fibrillation</td>
<td>1</td>
</tr>
<tr>
<td>Right-sided heart failure requiring isotropic support</td>
<td>2</td>
</tr>
<tr>
<td>Vasodilatory shock requiring vasopressor support</td>
<td>3</td>
</tr>
<tr>
<td>Minor</td>
<td></td>
</tr>
<tr>
<td>Anemia requiring transfusion</td>
<td>5</td>
</tr>
<tr>
<td>Pneumonia</td>
<td>4</td>
</tr>
<tr>
<td>Urinary retention</td>
<td>1</td>
</tr>
<tr>
<td>Ileus/severe constipation</td>
<td>2</td>
</tr>
<tr>
<td>Deep venous thrombosis without embolism</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary edema</td>
<td>8</td>
</tr>
<tr>
<td>Renal insufficiency</td>
<td>2</td>
</tr>
</tbody>
</table>

Comment

ATD occurs when thoracic chest growth is affected by an extensive early open PE repair [1–3]. A small percentage of these patients with hypoplastic abnormally developed thoraxes present as adults with progressive dyspnea and exercise intolerance secondary to cardiac compression and restrictive pulmonary function [1, 3–5]. Although surgical treatment of ATD has been reported in children, experience in the adult population is limited [6–12]. Our reconstruction efforts focused primarily on decreasing cardiac compression and secondarily on increasing overall thoracic volume. The technique of lateral thoracic expansion was described by Davis and colleagues [9] in Jeune’s syndrome, with healing documented. Despite anterior elevation of the chest wall, the percentage of increased overall thoracic space is limited unless expansion gaps are created. Long-term chest wall stability and healing of the separated sternum and multiple rib bridges in the adult are a concern. The addition of bone graft to these spaces may improve healing. The durability of the repair after removal of sternal support bars and the risk of PE recurrence are unknown.

In pediatric patients, improvement in pulmonary function after repair has been demonstrated [7, 12]. Our patients reported subjective improvement in their symptoms and ability to perform exercise. However, limited postoperative evaluations using pulmonary function testing and CPET have not objectively demonstrated improvement. Weber and Kurckhubasche [7] also noted some patients who had no improvement in pulmonary function at 24 months despite improved exercise tolerance. Radiographically and echocardiographically, overall chest volume improved (Fig 5) and cardiac compression was relieved, with improved ventricular size and function. We hypothesize that our patients benefited from surgical repair by increased cardiac output after relief of compression. All patients reported subjective improvement in their symptoms and ability to perform exercise. Although not cosmetically “normal,” patients were satisfied with their overall results and felt they were significantly
improved from preoperative levels. There were no long-term objective measures of patient improvement performed.

This series reviews our experience with adults presenting for surgical correction of ATD. Within the limitations of small numbers, retrospective evaluation, and limited follow-up, subjective improvement in clinical symptoms is suggested. Radiographically, cardiac compression appears to be relieved. Although these were complex procedures with numerous potential complications, the majority of patients were able to undergo successful repair without mortality or persistent complications. The long-term durability and results remain to be assessed.

In conclusion, although rare, ATD can occur after early Ravitch repair, and adult patients may present with disabling symptoms related to cardiac compression and restrictive pulmonary function. Reconstruction with anterior sternal elevation and expansion of the anterior chest can be safely performed. This improvement of overall thoracic space may improve symptoms and patient quality of life, although definitive evidence for physiologic improvement is lacking.

References


Fig 5. Computed tomographic axial images of a patient with acquired thoracic dystrophy (ATD) (A) before and (B) after reconstructive surgical intervention.