Operative management of acquired Jeune's syndrome

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ABSTRACT

Background: Acquired Jeune's syndrome is a severe iatrogenic deformity of the thoracic wall following a premature and aggressive open pectus excavatum repair. We report herein our technique and experience with this rare condition.

Methods: From 1996 to 2011, nineteen patients with acquired Jeune's syndrome were retrospectively identified in a tertiary referral center. The technique used to expand and reconstruct the thoracic wall consisted of 1) release of the sternum from fibrous scar tissue, 2) multiple osteotomies along the lateral aspect of the ribs with anterior advancement of costal-cartilages to protect the heart, 3) stabilization of the thorax by placing a curved bar for retrosternal support and, 4) restoration of the sterno-costal junction by wiring the lower cartilages to the edge of the sternum.

Results: Major complications observed in this series were: bar displacement (seven cases), postoperative death from cardiac arrest following bronchoscopy (one case), late cardiac tamponade from migration of wire suture fragment (one case), and need for multiple reoperations (one case). Long-term cosmetic results and improvement in daily quality of life were reported as positive in the majority of cases.

Conclusions: Anterior chest wall reconstruction successfully treated our series of patients with acquired Jeune's syndrome. This multifaceted technique is an effective procedure that allows expansion of the thoracic cavity and improvement of aerobic activity.

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Acquired Jeune's syndrome, also known as acquired restrictive thoracic dystrophy (ARTD) [1], is the most severe iatrogenic deformity of the thoracic wall following open pectus excavatum (PE) repair performed at a preadolescent age [2]. The cause of this failure of growth of the chest wall is not completely clear, but is probably a consequence of an overzealous resection of the deformed costal cartilages at the time of primary PE repair and devascularization of the growth ossification center of the sternum due to an extensive anterior osteotomy [2,3]. In many cases, perichondrial sheets and costal cartilages have been completely removed. The damage is further aggravated in cases in which the inferior costal cartilages were sutured together under the sternum as additional posterior support. The cartilage regeneration and subsequent bone replacement of this tissue lead to the formation of a semicircular band underneath the sternum, which results in a frozen thorax [4].

Due to the limited experience at any single center and the variability of clinical presentation, the treatment of this condition is still a challenge for surgeons and current results have not suggested easily generalizable surgical principles. The aim of the present study is to report the experience at Johns Hopkins Hospital over 15 years in operative management of acquired Jeune's syndrome by anterior thoracic expansion using the Lorenz bar as retrosternal support, to analyze and discuss complications, and to evaluate long term outcomes and quality of life changes after surgical repair.

1. Patients and methods

Following Institutional Review Board approval (IRB #82036), a retrospective review was performed of all patients who underwent repair of recurrent PE at the Johns Hopkins Hospital from January 1996 through June 2011. Preoperative evaluation consisted of: physical examination, static and exercise cardiopulmonary function tests, echocardiogram, and computed tomography (CT) scans. In some cases three-dimensional CT scans were reconstructed in order to clarify the extent of the deformity, the shape and the rotation of the sternum, and the presence of fused, missing or bifid ribs. Included in this analysis are patients labeled as having acquired Jeune's syndrome, which we defined as patients with markedly abnormal chest wall growth after failed primary open repair of PE performed in preadolescent age, associated with severe restrictive pulmonary disease (FVC and TLC <60% of the predicted value on pulmonary function testing), physical and CT characteristics as shown in Figs. 1, 2 and 3, and significant issues with aerobic daily activity and social life. Exclusion criteria were other acquired deformities of the thoracic wall and congenital asphyxiating thoracic dystrophy.
Medical records, including electronic patient records, hospital charts, and surgical operative notes, were used to obtain information on demographics, preoperative, operative, and postoperative characteristics. To evaluate long-term quality-of-life improvement in daily activities after surgical repair, a daily activities questionnaire (Table 2) was administered to patients, via telephone, by a member of the research team. A disease severity classification scale (Table 3) was created to identify the extent of functional limitation on the basis of activities of daily living.

1.1. Surgical procedure

A transverse incision was made in the anterior aspect of the chest wall at the level of the previous PE incision. Thorough exposure of the thoracic cage was obtained by mobilizing a myocutaneous flap of both major and minor pectoralis muscles and separating the medial fibers of the rectus abdominis from the xiphoid.

The expansion of the chest wall was achieved using a reconstruction scheme detailed in Fig. 4. After placement of a Lorenz bar (Biomet Microfixation, Jacksonville, Fl; Fig. 5) for posterior sternal support, a transverse anterior osteotomy of the sternum at the sterno-manubrial junction was performed and the deformed costal cartilages were resected to allow further elevation of the sternum.

2. Results

From January 1996 to June 2011, a total of 105 patients were evaluated for recurrent pectus excavatum. Recurrent deformities were repaired with a modified Nuss procedure in seventy-five patients and Ravitch operation or complex anterior chest wall reconstruction in thirty patients. Of these, nineteen patients with acquired Jeune’s syndrome were identified. Fifteen were male. Preoperative characteristics are summarized in Table 1. One patient had a concomitant diagnosis of connective tissue disorder (Marfan syndrome). The mean age at prior primary PE operation was 4.63 years (range 1.6–10 years), which had been performed by Ravitch procedure in all cases. Only one patient had undergone primary PE repair at our institution. Mean preoperative PFT values (SD) were: FEV1 38.4% (11.2), FVC 38.7% (9.1) and TLC 45.6% (9.7) of predicted values for age, height and sex. The mean age at the time of chest wall repair for acquired Jeune’s syndrome was 21.7 years (range 11.2–36.7). All patients had experienced progressive dyspnea and fatigue with moderate exercise (such as walking up a flight of stairs, carrying a shopping bag, or walking without stopping a distance of about 300 m); three had dyspnea at rest and two of them required baseline oxygen supplementation. One patient with severe impairment of pulmonary function (FEV1 18.1% and FVC: 21.5% of predicted) had been evaluated for lung transplantation, but due to an extremely narrow thorax (Fig. 3), he could not be put on the transplant waiting list.

Seven patients complained of chest pain and palpitations, and four had recurrent episodes of pneumonia requiring hospitalization. Two patients suffered from dilated cardiomyopathy, and one had a pacemaker implantation. Severe fatigue and impairment of physical mobility resulted in social isolation for most of these patients.

In seventeen cases, the operative management was complete anterior chest wall reconstruction and the remaining two patients underwent a modified Ravitch procedure. Two surgeons experienced in pediatric chest wall reconstruction performed all procedures. A single Lorenz bar as retrosternal support was placed in fourteen cases, and two bars in four cases. Intraoperative findings are shown in Fig. 4. In two cases, costal cartilages from fifth to seventh ribs were absent and the costal margin was reconstructed, using autologous rib grafts in one case and femur allograft in the other. All patients were
extubated in the operating room, and nine were admitted to an intensive care unit (ICU) for close monitoring, of whom six were transferred to routine nursing-level care on the first postoperative day. One patient required noninvasive respiratory support with Bi-level Positive Airway Pressure (BiPAP) for 48 h and the other two patients had a postoperative course complicated with respiratory distress requiring reintubation (on the first and fifth postoperative day, respectively). In one of these two patients, who had a past history

![Computed tomography show a typical “bell shape thorax” in a 26-year-old male with severe impairment of cardiopulmonary function.](image)

**Fig. 3.** Computed tomography show a typical “bell shape thorax” in a 26-year-old male with severe impairment of cardiopulmonary function.

![Depiction of typical anatomic configuration and surgical principles.](image)

**Fig. 4.** (A) Depiction of typical anatomic configuration and surgical principles. The manubrium, clavicular head and 1st and 2nd cartilages and ribs are overgrown; the body of the sternum is atrophic, short and depressed; some ribs are fused; from V to VII ribs the cartilages are often absent and the ribs are attached to the sternum through fibrous scar tissue. A semicircular fibrous band, formed by cartilages regrown and calcified, lying beneath the sternum plate is often present and further restricts the thoracic cavity. Two longitudinal incisions are made at the level of the fibrous tissue scar that connected the deformed cartilages to the edge of the sternum. Once the fibrous scar tissue is removed and the cartilages are resected, the sternum is free. Lateral rib osteotomy is used as an adjunct for anterior release. (B) A transverse incision is performed at the level of the junction between the manubrium and the body of the sternum. The body is elevated up to the top edge of the manubrium and wired to it. The sternum can be further elevated if necessary with one or two wedge anterior osteotomies; the sternoplasty in some case is completed by inserting rib grafts in the bony defect of the anterior table of the sternum. (C) The cartilages are advanced and the lower cartilages are wired to edge of the sternum to restore the sternocostal junction and give more stability at the anterior chest wall. (D) The Lorenz bar is placed under the sternum. The ends of bar are fixed with two lateral stabilizing plates; the bar is also secured to adjacent rib with two suture wires. The sternum may also be directly wired to Lorenz bar.
including surgical repair of congenital diaphragmatic hernia and severe scoliosis requiring spinal fusion, the prolonged ICU stay was complicated with aspiration pneumonia. The patient developed sepsis and multiple organ failure requiring extracorporeal membrane oxygenation (ECMO). The patient died on postoperative day ten due to severe hypoxia causing pulseless electrical activity arrest. In the other patient, with dilated cardiomyopathy and severe obstructive and restrictive pulmonary disease requiring preoperative home oxygen supplementation, the postoperative course was complicated with a new onset of atrial fibrillation and subsequent cardiac failure.

In the immediate postoperative period, chest pain was managed with intravenous patient-controlled analgesia (PCA) using hydrocortisone in twelve patients and epidural with fentanyl and bupivacaine in seven. PCA therapy was provided for an average of 5.0 (SD: 4.5) days and epidural for 5.4 (SD: 4.8) days. Patients were then gradually transitioned to an oral narcotic regimen. The Average hospital length of stay was 6.9 (range 3–23) days.

Postoperatively, close monitoring with clinical evaluations every 6 months was offered until bar removal. Notable late postoperative complications included bar displacement requiring revision (seven cases), and upper sternal depression with bar in situ, required placement of a second bar (one case). One patient experienced late hemopericardium and chronic cardiac tamponade related to migration of a retained wire. Removal of the retained wire and drainage of hemopericardium allowed recovery. Two years after this event, the same patient underwent redo repair for recurrence, but the late postoperative course was complicated by right ventricle erosion, related to the instability of the bar. The bar was then removed and the cardiac injury repaired. The patient is now in good general condition, and the sternum is stable and in neutral position. He is satisfied with the cosmetic result, but still complains of dyspnea with moderate exercise.

Finally, one patient required multiple surgical procedures to obtain a satisfactory chest wall expansion. Anterior chest wall reconstruction was complicated by bar displacement, recurrence and worsening of clinical condition. At this point, a multistage procedure was performed. The lateral thoracic expansion was obtained by dividing and opposing in staggered fashion, the ribs 3rd to 7th allowing expansion of the superior part of the thorax. The lower part of the sternum remained depressed and was elevated, a second time, by performing the split of the sternum with interposition of two femoral bone grafts to support the sternum halves apart. This area was then covered with Marlex mesh, AlloDerm graft and skin flaps. The postoperative course was complicated with lower graft displacement and exposure, and partial collapse of the lower chest wall. The lower bone graft and the hardware were removed and stabilization and consolidation of the thoracic wall was finally obtained positioning two new fibular bone grafts between the split sternal halves. The anterior chest wall was then covered with an anterolateral microvascular free-flap from the right thigh. Now the thorax is stable, the flap has good perfusion and the wound has healed. The symptoms are improved, although oxygen supplementation is still necessary and the patient requires support when walking.

The bars were left in place for a mean period of 2.3 (SD: 0.9) years. Static PFTs showed an increase of pulmonary volumes with mean values (SD) of FEV1, FVC and TLC as, respectively: 54.8% (4.0), 52.8% (3.2), and 50.7% (8.2) of predicted. In two patients with severe combined obstructive and restrictive ventilatory defects, PFTs were not improved and both continued to require oxygen supplementation. One of these patients had a progressive deterioration of his cardiopulmonary function and died ten years after his reconstruction. Finally another patient with a history of multidrug abuse died several years after the operation following IV narcotic overdose. Sixteen patients are alive at a median follow-up of 11.9 years (range 3.7–15.6 years) after surgery, and were attempted contact by telephone to outline functional status and ability to perform activities of daily living. Nine patients were successfully contacted and agreed to administration of the quality-of-life questionnaire (Table 2): 75% of patients are in Class I or II, 12.5% are in Class III, and 12.5% are in Class IV of our functional classification system (Table 3).

### 3. Discussion

Acquired Jeune’s syndrome is a rare iatrogenic thoracic deformity following extensive and premature surgical repair of PE. Because few cases have been recognized, to date there is little generalizable experience in the operative management of this condition. The surgical principles described in the present study are meant to safely expand the anterior chest wall and increase the volume of the thoracic cavity. Similar to the management of congenital Jeune’s syndrome, the goal of surgical treatment is to expand the thoracic cage in order to release the entrapped lung, unload mechanical pressures on the heart, and improve diaphragmatic excursion. A number of techniques have been described to treat this rare disorder. The first thoracic wall expansion was performed in 1965 in a case of Jeune’s syndrome by Durand by performing multiple costochondral resections [5].

### Table 1

Preoperative characteristics.

<table>
<thead>
<tr>
<th>Demographics characteristics and comorbidities</th>
<th>Patients n = 19</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at operation, mean (range) years</td>
<td>21.7 (11–37)</td>
</tr>
<tr>
<td>Male (%)</td>
<td>15 (79)</td>
</tr>
<tr>
<td>Mean age at first operation, years (range)</td>
<td>4.6 (1.6–10)</td>
</tr>
<tr>
<td>Pulmonary function test results</td>
<td></td>
</tr>
<tr>
<td>FEV1, mean (SD) of predicted value</td>
<td>38.4 (11.2)</td>
</tr>
<tr>
<td>FVC, mean (SD) of predicted value</td>
<td>38.7 (9.1)</td>
</tr>
<tr>
<td>Preoperative symptoms</td>
<td></td>
</tr>
<tr>
<td>Dyspnea and fatigue with moderate exercise, N (%)</td>
<td>17 (90)</td>
</tr>
<tr>
<td>Dyspnea at rest, need for oxygen supplementation, N (%)</td>
<td>2 (11)</td>
</tr>
<tr>
<td>Chest pain and palpitations, N (%)</td>
<td>7 (37)</td>
</tr>
<tr>
<td>Recurrent pneumonia, N (%)</td>
<td>4 (21)</td>
</tr>
</tbody>
</table>

FEV1: Forced expiratory volume at 1 s; FVC: Forced vital capacity; SD: standard deviation.

### Table 2

Quality of daily life questionnaire.

<table>
<thead>
<tr>
<th>Question</th>
<th>Rating</th>
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<tbody>
<tr>
<td>Do you frequently get breathless at rest?</td>
<td></td>
</tr>
<tr>
<td>Can you bathe and get dressed without shortness of breath?</td>
<td></td>
</tr>
<tr>
<td>Do you get breathless walking on level ground?</td>
<td></td>
</tr>
<tr>
<td>Can you comfortably complete a grocery store trip?</td>
<td></td>
</tr>
<tr>
<td>After walking up one flight of stairs, could you still have a conversation?</td>
<td></td>
</tr>
<tr>
<td>Can you walk up two flights of stairs without stopping?</td>
<td></td>
</tr>
<tr>
<td>Are there social activities that you refuse to join because they are too strenuous?</td>
<td></td>
</tr>
</tbody>
</table>

### Table 3

Functional classification scale.

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>No activity limitations</td>
</tr>
<tr>
<td>II</td>
<td>Comfortable at rest, minor limitations of ordinary activity</td>
</tr>
<tr>
<td>III</td>
<td>Comfortable at rest, marked activity limitations but able to perform activities of daily living</td>
</tr>
<tr>
<td>IV</td>
<td>Severe limitations, unable to perform activities of daily living, need of oxygen supplementation</td>
</tr>
</tbody>
</table>

*Fei* Lorenz bar used to stabilize the thorax after reconstruction.
Subsequently, the majority of cases of congenital or acquired Jeune's syndrome reported in literature have been managed with an anterior approach. One early approach suggested that considerable increase in thoracic volume would be achieved by splitting the sternum [6]. The window obtained in the anterior part of the thorax by splitting the sternum was maintained opened with the interposition of synthetic materials, such as methyl methacrylate plate, or graft (autologous rib, femur or fibular allograft) [7–9]. Other approaches are a modified Ravitch procedure with Rehbein split as sternal support [2], Nuss procedure [10,11] and lateral thoracic expansion. This last technique allows increasing the thoracic wall by dividing and opposing the ribs in staggered fashion. The newly created elongated ribs are then stabilized, during the healing phase, by titanium miniplates [12].

Characteristically in our experience, these patients have a very low-lying diaphragm and a narrow thoracic cavity with immovable anterior chest wall due to the presence of fibrous tissue scar that has replaced the sterno-costal junction. The most important step of our procedure is the release and elevation of the sternum. Once this maneuver was performed intraoperatively, in fact, the tidal volumes tended to immediately increase. The second step is the reconstruction of the anterior chest wall. To protect the lungs and heart, the anterior gap between the sternum and the cartilages is bridged by cutting the ribs laterally. Finally, the stability of the anterior chest wall is obtained by placing a retrosternal bar and restoring the sterno-costal junction. This technique allows reconstructing the anatomic configuration of the anterior chest wall in a more physiologic way than other procedures. More invasive approaches, such as the split of the sternum or the lateral thoracic expansion, should be reserved for the most severe defects or in cases of recurrence of the condition after attempted repair with Ravitch procedure or anterior chest wall reconstruction.

The anterior chest wall reconstruction described in this study includes the placement of a retrosternal bar to stabilize and maintain the expansion of the thorax. The major limitation of this procedure is the instability of the bar; in this series the bar migration occurred in seven cases (37%). Although this high rate of bar displacement can be partially related to a lack of adequate fixation (in three cases no lateral stabilizers and wire sutures were used to fix the bar), other causes such as the overall rigidity of the thorax should be considered. There is prior report of an increased risk of this complication in patients who had a previous failed open PE repair [10]. An unstable retrosternal strut can erode into the sternum or the pericardium with dramatic and life-threatening consequences. Appropriate fixation of the bar is thus necessary to prevent this feared complication, and in some cases can be ensured by wiring the bar directly to the sternum. The use of two bars in select cases may increase the stability of the thoracic cage, but we did not achieve sufficient experience with that solution in this series.

Patients and their families should be informed about the limitations of the surgical procedure and the high risk of major complications and possible recurrence. These patients are at high risk of the following intraoperative and postoperative complications: 1) difficulty in controlling hemorrhage; 2) cardiac arrest; 3) cardiac and respiratory failure; and 4) sepsis. The release of the sternum from the scar tissue is a difficult and risky maneuver, during which injuries of the internal mammary artery or intercostal vessels can occur. In the majority of cases prompt control limits blood loss but should be done carefully to preserve the vascular supply of the sternum. Intraoperative cardiac arrest has been described in the literature in two cases of ARTD, due to a ventricular dysfunction as a consequence of severe pulmonary restrictive disease [10]. None of our patients had cardiac arrest, but the post-operative course was complicated in one case by progressive cardiac failure.

Other complications related to thoracic cavity expansion are: migration and exposure of grafts, wires and struts, or bar displacement. Although splitting the sternum is an attractive procedure and may appear simpler than our reconstruction, the presence of heart and lungs herniated through the anterior chest wall is at high risk of major injury. This vulnerability thus limits daily activity and social life. The sternal split could furthermore be complicated by displacement and migration of the rib grafts interposed between the two sternal halves. The sternum of patients with acquired Jeune’s syndrome is quite fragile and hypotrophic, and the interposed grafts are at high risk of displacement, migration and necrosis. In addition, sternal split can be complicated with skin erosion and exposure of grafts and wires if the coverage of the sternum is obtained only with skin flaps. Coverage of this area with a pectoralis muscle flap, Marlex mesh and Alloderm graft decreases the risk of non-healing and improves the cosmetic result [13].

In contrast to the comprehensive approach described herein, in our opinion, a lateral only approach as described by Davis et al. [12] does not allow adequate expansion of the chest wall because the middle part of the thorax remains depressed and immovable. Lateral thoracic expansion increases the thoracic volume, but the thorax remains frozen without any improvement of regional chest wall motion. In addition, loss of volume expansion can occur if fracture of the titanium miniplates occurs before healing of the newly created ribs.

The present study is limited by the retrospective nature of the study and the rarity of the presentation of this acquired deformity of the thoracic wall. The quality of life questionnaire and functional classification scale were created specifically for this study and were not previously validated, as there are no previously reported metrics specifically for restrictive lung disease reported in the literature to our knowledge. We plan on future validation of the tools with our patient population of chest wall deformities.

The overall finding in this series is that comprehensive anterior chest wall reconstruction with retrosternal support is an effective procedure that improves pulmonary function and quality of life for patients with acquired Jeune's syndrome. Individualized surgical planning is still required considering the variability of clinical presentations. In cases of very severe cardiopulmonary disease, the anterior chest wall reconstruction should be considered as the first step of a multistage procedure to expand the thoracic cavity, and in extreme cases the final attempt to give these patients a chance of lung transplantation.

References


Discussion

Discussant: Dr. David Notrica (Phoenix, AZ): Thank you for a nice paper. In some of these that we've done, once we've expanded the chest wall, there are clearly gaps in the chest after we've elevated the sternum and increased the total area. What do you do about the spaces in between the chest wall that you have created?

Response: Dr. Maria Grazia Sacco Casamassima: We have a gap in the anterior chest wall between the sternum and the cartilage. We try to advance the cartilage and wire the cartilage to the sternum if it is possible. Generally we wire the lower cartilage but in some cases also other cartilage in the upper part of the sternum depending on the expansion that we can obtain.

Dr. David Notrica: The other question is, have you used any of the plating systems to stabilize the sternum? There are a lot of commercially available plating systems for rib fixation and sternal fixation. Have you used any of those?

Response: Dr. Maria Grazia Sacco Casamassima: No, we use only the Lorenz bar that is fixed with wire to the rib and in some cases also we fix the bar to the sternum to improve stabilization.

Discussant: Dr. Alberto Pena (Cincinnati, OH): Over 30 years ago we presented a paper at the American Academy of Pediatrics warning the pediatric surgical community about the danger of removing, resecting completely the costal cartilages in treating pectus defects very early in life because those patients have a problem not only with fibrosis of the anterior chest wall but the entire chest did not grow. We reproduced the operation in animals and found that ribs and the entire chest simply doesn't grow because we believe that there are growth centers in the cartilages when they are resected completely. So taking care only of the anterior part of the chest perhaps may not improve completely the problem because – have you measured the entire volume of the chest in these patients? How do you deal with that? How do you expect to improve that?

Response: Dr. Maria Grazia Sacco Casamassima: We do not particularly measure the volume of the chest wall. I'm sorry. I don't have an answer for this question.

Discussant: Dr. Robert Shamberger (Boston, MA): You very nicely demonstrated the degree of pulmonary restriction on the patients prior to surgery and you concluded that the operation improved the pulmonary function but I didn't see any results – either total lung volumes or FEV1 – that demonstrated you had improved pulmonary function with this surgery.

Response: Dr. Maria Grazia Sacco Casamassima: We didn't show this data because 70% of patients had improvement of pulmonary function but this improvement was not really high. They passed from 30% of predicted value to 50%, but the most important thing is that the quality of life was really improved in these patients. These patients before the operation were unable to walk up a flight of stairs to go shopping or do ordinary activities like ride a bicycle or stay with peers. Afterward they had real improvement in quality of life and this is the most important result for us.