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Contemporary management of recurrent pectus excavatum

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ABSTRACT

Background: Optimal management of recurrent pectus excavatum (PE) has not been established. Here, we review our institutional experience in managing recurrent PE to evaluate long-term outcomes and propose an anatomic classification of recurrences, and a decision-making algorithm.
Methods: Clinical records of patients undergoing repair of recurrent PE (1996–2011) were reviewed. Univariate and multivariate logistic regression analyses were employed to examine patient characteristics as potential predictors for re-recurrence.
Results: Eighty-five patients with recurrent PE were identified during the study period. The initial operation was a Ravitch procedure in 85% of cases. Revision procedures were most frequently Nuss repairs ($N = 73$, 86%), with remaining cases managed via open approach. Overall cosmetic and functional results were satisfactory in 67 patients (91.8%) managed with Nuss and in 7 (58%) patients managed with other techniques. Seven (8%) patients required additional surgical revision. Multivariate analysis identified no statistically significant patient or procedural factors predictive of re-recurrence.
Conclusion: This study demonstrates that the Nuss procedure can be an effective intervention for recurrent pectus excavatum, regardless of the initial repair technique. However, open repair remains valuable when managing severe cases with abnormalities of the sternocostal junction and cartilage regrowth under the sternum.

Recurrent PE can occur irrespective of the surgical approach used at the time of initial repair. The chance of recurrence varies with the type of anomaly originally corrected, the presence of connective tissue disorders, and the age of the patient at the time of correction. The severity of recurrent PE ranges from mild cosmetic deformities to severe asphyxiating thoracic dystrophy, also known as "acquired Jeune's syndrome" [1–3]. Therefore, because of this wide spectrum of acquired deformities, their surgical repair may be technically challenging. There are no current guidelines regarding the management of recurrent PE; thus, it is controversial whether open or minimally invasive approaches are optimal for these conditions. In the era of the minimal invasive repair of PE (MIRPE), also known as Nuss procedure, surgeons performing PE repair have become less familiar with the open "Ravitch" techniques. Familiarity with MIRPE has also led to its adoption to repair recurrences.

Previous investigations have proven the effectiveness of the MIRPE in repairing recurrences after failed MIRPE [4–6]. However, there is a paucity of literature investigating the safety and effectiveness of the Nuss technique in patients with recurrence after failed open repair [6–10]. A small number of retrospective series suffer from relatively small simple sizes, lack of anatomic detail on the type of recurrences, and a short term follow-up [6–10] to draw generalized conclusions. It thus remains to be clarified which patients may benefit more from MIRPE and if this procedure provides durable functional and cosmetic results.

The aim of this study was to review a single large institution experience in managing recurrent PE and propose a decision-making algorithm to assist surgeons in selecting the appropriate operative approach for recurrent chest wall deformities.

1. Methods

Following institutional review board approval, a retrospective chart review was conducted of all patients who underwent surgical repair of recurrent PE at Johns Hopkins Hospital from January 1996 to June 2011. Medical records, including electronic patient records, hospital charts, and operative reports and outpatient visit notes, were used to obtain patient characteristics, perioperative data, and outcomes. Included in the analysis were patients requiring surgical revision of recurrent PE who had their initial repair during childhood or adolescence (aged 1 to 18 years). Excluded were patients with PE occurring after median

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sternotomy as well as severe growth restriction known as acquired Jeune's syndrome, which we have previously described [11].

1.1. Preoperative workup

Preoperative workup for patients with recurrent PE does not differ from that of patients presenting for primary repair [12–14]; it includes: chest CT scan, cardiopulmonary evaluation, including pulmonary function tests (PFT) and echocardiogram.

1.2. Surgical procedures

Surgical procedures used to repair recurrent deformities were selected according to the type of recurrent defect. The radiologic classification of the common "anatomic issues" observed in recurrent PE is provided in Table 1, and the decision-making algorithm used to select the surgical approach is displayed in Fig. 1. Axial computed tomography and three-dimensional reconstructions depicting common findings of recurrent PE are displayed in Figs. 2–5.

1.2.1. Open procedure

The prior longitudinal or transverse skin incision is reopened and myocutaneous flaps are bilaterally elevated to provide a complete exposure of the thoracic cage. The fibrous tissue scar surrounding the sternum is removed using electrocautery and the pleural cavity opened to remove the remaining scar tissue and complete the release of the sternum. At this point the sternum is freely mobile and can be easily elevated into a neutral position. Sternal rotation is further corrected by performing an oblique transverse osteotomy at the level of the anterior table. Three or four of the deformed cartilages are then bilaterally excised, preserving the perichondrium.

A more complex reconstruction is performed in cases in which the cartilages are partially or completely absent (floating sternum) or the entire sternocostal junction is replaced by fibrous tissue scar. After removing the fibrous band at the level of sternocostal margin, the residual lower cartilages are advanced and attached to the edge of the sternum with stainless steel wire. For patients in whom cartilages are completely absent, autologous rib or cadaveric bone grafts (fibular or femoral allografts) are used to bridge the gap between the ribs and the sternum. The sternum is further stabilized in all open procedures by placement of a retrosternal support. At our institution, the Adkins strut was utilized until 1997, after which the Lorenz bar was used exclusively.

1.2.2. Modified Nuss procedure

In the absence of severe anatomic deformity, recurrent PE was treated with a modified Nuss procedure. Technical modifications introduced at our institution have been previously described in detail [14]. Briefly, no thoracoscopic guidance is used, the left-to-right approach is used to create the retrosternal dissection and introduce the bar, and the four-point fixation technique, employing two lateral stabilizers and two medial wire sutures, stabilizes the bar. The retrosternal support is left in place for 2–4 years if a Lorenz bar was used and for 6–12 months if the support was an Adkins strut.

Table 1
Radiologic classification of anatomic issues in recurrent PE after failed Ravitch repair.

Anomalies of the sternum	Overgrowth of the manubrium
	Short and hypotrophic body
	Twisted sternum
	Malunion of the xiphoid process with associate epigastric hernia
Anomalies of the ribs and the sternocostal junction	Fused/deformed ribs
	Absence of cartilages or fibrous union between ribs and sternum (lung herniation/floating sternum)
	Evidence of cartilages regrowth under the sternum (dense tissue/calcifications)
	Complete replacement of the entire sternocostal junction by fibrous tissue
	Prominent cartilages in the left or right side of the thorax (pectus carinatum features)

1.3. Assessment of results after surgical revision of recurrent PE

Patients are followed for at least two years following bar removal. Final results, after bar removal, were defined as satisfactory if symptoms were completely resolved and the sternum remained in neutral position. Unsatisfactory results were defined as the presence at least one of the following conditions: persistence of thoracic pain, lack of improvement in aerobic activities, protrusion of the cartilages on either side, residual depression of the sternum.

The obstetrical caliper was used to objectively quantify the severity of residual defect after bar removal. The caliper index is the ratio between the thicknesses of the thorax at the level of the deepest part of the defect measured along the midclavicular lines (right and left) and the thickness of the thorax at the same level measured along the midsternal line [13]. With this index, a residual defect greater than 1.5 cm was considered mild recurrence, while a residual defect greater than 2.5 cm was considered severe recurrence with indication for further surgical revision.

1.4. Data analysis

Descriptive statistics are presented as a mean and standard deviation (SD), or median and range. Univariate and multivariate logistic regression analyses examined potential predictors for re-recurrence, including patient comorbidities, age at initial repair, and type and number of operations. P values <0.05 were considered significant. We hypothesized that patients with CTD, patients undergoing primary repair at a young age and patients that are status post multiple revisions for recurrent PE are likely to have markedly abnormal chest wall compliance; we further speculated that these patient factors, in combination with operative technique employed at surgical revision might serve as predictors for re-recurrence. We therefore included these factors in a logistic regression model to better assess the relative magnitudes of various contributing risk factors.

2. Results

During the study period, a total of 105 patients were evaluated for surgical repair of recurrent PE. Eighty-five patients met the inclusion criteria. Forty-five (52.9%) patients had their initial operation at our institution. Information regarding previous failed repair is displayed in Table 2. Overall the median age at initial operation was 8.4 years (range 2.5–18 years) and the mean age at the time of surgical evaluation for recurrent PE was 21.3 ± 8.5 years (Table 3). Seventy-one patients (83.5%) were male. Diagnosis of Marfan syndrome was genetically confirmed in 8.2% of cases ($N = 7$). One patient with structural congenital abnormalities of the spine and ribs, severe tracheomalacia and impairment of respiratory dynamic was suffering from Goldenhar syndrome. The mean depth of the defect measured with caliper was 3.5 ± 0.8 and the mean Haller index was 4.4 ± 1.2. Six patients had a floating sternum and in three patients the free floating sternum was associated with lung herniation.

PFT revealed a combined obstructive and restrictive pattern in the majority of patients. Predicted mean values of forced expiratory volume in 1 second (FEV1), forced vital capacity (FVC), and forced expiratory flow between 25% and 75% exhalation (FEF25-75%) were FEV1: 77.2 ± 13.8 FVC: 76.8 ± 14.4; and FEF25%–75%: 66.7 ± 20.

Revision procedures were performed by two pediatric surgeons experienced in chest wall reconstruction, with technique selected according to the type of recurrent deformity. Modified Nuss repair was performed in 73 cases, the Ravitch operation with retrosternal