Pectus Excavatum: More Than a Matter of Aesthetics

Fizan Abdullah, MD, PhD; and Jamie Harris, MD

ABSTRACT

Pectus excavatum (PE) is the most common congenital chest abnormality, and affects males 5 times more frequently than females. PE results from improper fusion of the ribs with the sternum during embryologic development. The cardinal presenting sign is chest depression. Evaluation includes serial measurement of the chest deformity defect. Additional evaluation of cardiopulmonary function, including arrhythmias and pulmonary function tests, should be done as well. Computed tomography scans are used to determine the Haller index, a measure of deformity severity, with a measurement of greater than 3.2 deemed severe. The main indication of repair is decreased cardiopulmonary capacity, not cosmetic. Surgical repair should be timed such that it occurs after the pediatric growth spurt. Generally, the Nuss procedure, which is minimally invasive, is the first-line surgical repair. Ravitch, or open repair, is used for more complex or asymmetric deformities.

Pectus excavatum (PE), or funnel chest, is the most common congenital abnormality of the chest wall, with an estimated incidence of 1 in 400 births and prevalence of 2.6% in children age 7 to 14 years. PE accounts for 90% of all chest wall deformities and affects 5 times more males than females.1 A chest depression is the cardinal sign of PE. It is typically detectable at birth but milder cases may not manifest themselves until the adolescent growth spurt. Other anatomic hallmarks of PE include sloped ribs, pot belly, and rounded shoulders, all of which become more prominent as the child grows older.

Considerations for treatment go well beyond aesthetics, as PE can affect a child’s self-esteem and lead to social isolation, and in extreme cases adolescents may develop suicidal ideation.2 Furthermore, PE can cause serious physiologic sequelae. Depending on severity, patients may report anterior and left parasternal pain, dyspnea with exertion, and reduced cardiopulmonary endurance, all of which can be corrected with surgery.

Cardiac evaluation is important for several reasons, including the fact that a subset of patients may have an abnormally positioned and/or partly compressed heart due to the deformity. Some patients with PE may also have mitral valve prolapse.

Although PE occurs most commonly as a stand-alone deformity, up to 20% of patients may have other skeletal anomalies, most notably scoliosis.3 In rare cases, PE can signal the presence of an underlying connective tissue disorder, such as Marfan syndrome or Loeys-Dietz syndrome.5 Recognizing PE as a sign of these serious pathologies is critical because they put patients at risk for serious complications, such as aortic aneurysms, aortic dissection, or both. A small portion of children with PE may have Poland syndrome, marked by an underdeveloped or absent pectoralis muscle, syndactyly, and a malformed rib cage.

ETIOLOGY

PE is a disorder of embryonic development. The mechanism is rooted in failure of the ribs to fuse properly with the sternum, a process that begins around the 35th day of gestation, continues throughout gestation, and is completed by final ossification during adolescence.

Even though the condition exhibits a pattern of inheritance (about one-third of PE cases have another affected family member), there is no known genetic...
aberration directly responsible for this abnormality.6

PRESENTATION AND CLINICAL EVALUATION
Most cases of PE are detected by visual inspection of the chest wall. PE is a spectrum disorder, so although many cases are diagnosed at or shortly after birth, milder cases may not become pronounced until late childhood or early adolescence.

It is important that cardiac and pulmonary examinations be performed at the time of diagnosis or shortly thereafter. Depending on the severity of the condition and the extent of cardiac involvement, PE may cause cardiac arrhythmias, dyspnea, reduced cardiopulmonary fitness, compromised exercise capacity, and fatigue.

Auscultation may reveal a characteristic murmur or click, signaling mitral valve prolapse. Additionally, the clinician may hear a systolic click stemming from the proximity of the pulmonary artery and sternum.

A thorough patient history is essential not only in evaluating the severity of the condition, but also to rule out the presence of certain associated syndromes. When PE occurs as part of a syndrome (eg, Marfan), additional symptoms that should trigger suspicion include presence of scoliosis, joint hypermobility/laxity, flat feet, arm span 1.05 times greater than total height, high-arched palate, and certain craniofacial features such as elongated skull, downward slanted eyes, and receding jaw.5,6 Additionally, a family history of cardiac aneurysms can signal the presence of a connective tissue disorder.

DIAGNOSIS
Physical Examination
The severity of PE can be estimated by caliper measurement of the distance from the deepest depression of the sternum to the top of the rib cage. Serial physical examination measurements of the depression can help track the progression of the defect over time.

Imaging
A computed tomography (CT) scan is currently the preferred modality used to define the degree of severity of PE, and it can also detect any cardiac displacement. The Haller index is the gold standard measurement of severity and can be easily calculated from the axial sections of the CT image. The index is based on the ratio of the interior transverse chest diameter (horizontal distance between the two sides of the rib cage) to the anterio-posterior diameter (the distance between vertebrae and deepest point of the sternal depression).

A normal Haller index value is approximately 2.5. The degree of deformity is directly proportional to the Haller index value. A ratio greater than 3.2 is deemed severe and often requires surgical correction, although some patients with lower indices may also be symptomatic and warrant intervention. Although lateral and anterio-posterior chest radiographs can also be helpful in calculating the Haller index, CT scan is the preferred imaging modality because of its superior visualization and the additional clues it may provide with regard to cardiac involvement.7

Cardiac Evaluation
Cardiac evaluation with electrocardiogram and echocardiography can help identify any heart anomalies, such as mitral valve prolapse or right atrial and right ventricular compression.

Pulmonary Evaluation
Because PE can impinge on the lungs, children may be initially misdiagnosed with exertional asthma. Pulmonary function testing can help determine the presence and degree of lung involvement. Patients with PE can have decreased forced expiratory volumes and decreased vital capacity. A restrictive pattern of lung disease could provide an additional indication for repair or for an earlier intervention.

BASICS OF SURGICAL MANAGEMENT
The main indications for PE repair are noncosmetic. There is evidence to support that PE can alter physiologic function and decrease cardiopulmonary capacity, both at rest and with exertion.5 Research shows that surgical correction leads to both subjective and objective improvements in pulmonary function and exercise capacity.9,10 Many patients with severe defects report rapid symptomatic relief after the procedure. Surgical correction also leads to dramatic cosmetic improvement in anatomy of the upper torso.11

The timing of surgery is important. To minimize risk for postoperative complications, optimize outcomes, and reduce the likelihood of recurrence, surgery is best deferred until after the pediatric growth spurt, generally between ages 10 and 15 years.12 The emerging consensus among pediatric surgeons in the US is that older is better, with the mean age at surgery now approximately age 13.5 years.13 However, patients with severe PE and reduced cardiopulmonary function may warrant earlier correction. That decision should be made on a case-by-case basis, factoring in defect severity and cardiopul-
monary involvement, overall health, comorbidities, and growth status, and it is best determined by consulting a pediatric surgeon specializing in chest wall deformities.

**Nuss Procedure**

The Nuss procedure is a minimally invasive technique that involves the insertion of a custom-made steel or titanium bar under the sternum and affixing it laterally to the ribs. Most cases can be successfully corrected with a single bar, although more severe cases may require the use of a second bar.

Patients undergoing the Nuss procedure should be screened for allergy to metal. Those with suspected personal or family history benefit from skin testing. A titanium bar can be used in patients with allergy to metal.\(^{14}\)

On average, the postoperative length of stay is less than 1 week.\(^{15}\) Postoperative recovery involves pain management with intravenous (first 48 hours) and oral narcotics, and nonsteroidal anti-inflammatory drugs or acetaminophen thereafter, with tapering of the dose over a few weeks. Patients generally can resume normal daily activities within 1 week of surgery; however, this is patient specific and usually limited by postoperative pain.\(^{16}\) Patients are advised to abstain from heavy physical exertion for about 6 weeks. Return to aerobic activity, including soccer and basketball, is typically permitted around 6 weeks after surgery, but participation in heavy contact sports such as football or hockey should not be resumed until after bar removal. The bar is removed after a minimum of 2 years, once the desired sternum shape is achieved. Bar extraction is generally performed under general anesthesia on an outpatient basis.

The Nuss procedure results in less scarring, and evidence points to lower infection and bleeding risk compared to the open, Ravitch procedure. During the past 2 decades, the Nuss procedure has become the preferred option for most patients; however, technique choice can be predicated on a variety of factors, such as individual patient anatomy and the severity of the condition. Recurrent cases may be candidates for the Ravitch procedure, particularly patients with more complex anatomy.\(^{17}\)

**Ravitch Procedure**

The Ravitch procedure is an open repair and is done by removing part of the anterior cartilage, followed by placement of steel struts affixed to the ribs on the side to support and elevate the breast bone. The struts are removed once an appropriate level of correction is achieved.

**Follow Up**

Patients require regular follow up with the surgeon for at least 2 years after repair, at intervals between 3 and 6 months.

**SURGICAL OUTCOMES**

Patients who undergo repair at surgical centers with experience in treating PE have excellent outcomes. Overall recurrence rates vary among studies, but have been quoted between 2% and 10% in both open and Nuss procedures.\(^{16}\) Some studies have shown nearly 95% of patients report satisfactory functional and cosmetic results.\(^{18}\) Complication rates range from 15% to 20%, including both serious and less serious sequelae.\(^{19}\)

Early post procedural complications (<1 month) include pneumothorax, pleural effusion, pneumonia, hemothorax, pericarditis, and surgical site infections. Late-onset complications (1 month or more after the procedure) include prolonged chest pain and, rarely, overcorrection into pectus carinatum deformity, bar displacement or rib fracture, and hypertrophic scarring.

Choice of proper technique, surgical experience, and optimal timing are critical in reducing complication rates. A recent multicenter study found that bar or strut displacement was the most frequent late-onset complication, affecting 7% of 182 patients, with all but 1 of the 13 cases requiring reoperation.\(^{9}\) The same study revealed that 2.2% of patients developed surgical site infections, and 1.1% had rib erosion or fracture.\(^{9}\) These rates vary from institution to institution, depending on patient volume and staff experience, highlighting the importance of referring patients to centers with skilled surgical teams and high volumes of pediatric patients with chest wall malformations. One single-center study has shown a bar displacement rate of 2.9% and a surgical infection rate of 0.7% with the routine use of chlorhexidine scrubbing.\(^{20}\)

The most serious complication of the Nuss technique is direct cardiac injury. According to one report, 13 such incidents have occurred since the technique was introduced in the late 1980s.\(^{21}\) The risk for this complication may have decreased over the years due to increased experience at large-volume centers performing the Nuss procedure.

Patients with persistent postoperative chest pain should be evaluated via echocardiogram, chest radiography, complete blood count, and erythrocyte sedimentation rate to rule out pericardial or pleural effusion, bar displacement, and infection.
The recurrence rate of PE is approximately 5%, but a single-center study has reported a recurrence rate as low as 1.7%. Recurrence risk varies depending on patient age at the time of the original correction, presence of connective tissue disorders, and the type of repair. Most recurrent cases can be successfully corrected using the minimally invasive Nuss approach, regardless of initial repair technique. The Ravitch procedure may be more appropriate for patients with recurrent PE who have severe abnormalities of the sternocostal junction and cartilage regrowth under the sternum.

CONCLUSION

PE is the most common congenital abnormality of the chest wall. The degree and severity of PE vary widely, and additional skeletal anomalies are present in up to 20% of patients. PE can be syndromic and signal the presence of connective tissue defects. Depending on its severity, the condition can affect cardiopulmonary function. Thorough evaluation, including assessment for the presence of associated syndromes and cardiac and pulmonary function testing, is essential in the assessment. Considerations for correction of PE go beyond just aesthetic appearance. Corrective surgery has a high success rate but the timing and surgical approach should be determined on a patient-by-patient basis and performed by an experienced pediatric surgeon at a center that treats a high volume of patients with chest wall deformity.

REFERENCES