

**Subject: Surgical Treatment for Chest Wall Deformities (Pectus Excavatum/Carinatum and Poland Syndrome)**  
**Number: 0309**

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**Surgical repair of severe pectus excavatum is considered medically necessary when the pectus index (i.e., Haller index) is greater than 3.25 and ANY ONE of the following criteria is met:**

- Pulmonary function studies demonstrate at least moderately severe restrictive lung defect.
- Cardiac testing, such as echocardiography or stress echocardiography, demonstrates findings consistent with external compression.

**Surgical repair of pectus carinatum is considered medically necessary when there is documented evidence of significant physical functional impairment (e.g., cardiac or respiratory insufficiency), and the procedure is expected to correct the impairment.**

**Surgical repair of a chest deformity associated with Poland syndrome is considered medically necessary when rib formation is absent.**

**The surgical treatment of chest wall deformities is considered cosmetic in nature and not medically necessary when performed solely to improve physical appearance or to treat psychological symptomatology or psychosocial complaints.**

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## **General Background**

The thorax is a rigid structure that protects the thoracic organs and supports the upper extremities. Abnormalities of the thorax often result in cosmetic complaints, but some chest wall deformities result in functional limitations, such as activity intolerance related to cardiac or respiratory impairment. Reported symptoms include mild to moderate exercise limitation, respiratory infections, and asthmatic conditions. Ultimately, the deformity may place physiological restrictions on the patient and result in decreased stamina and endurance. The most commonly reported chest wall deformities include pectus excavatum (PE), pectus carinatum (PC) and Poland syndrome. While pectus excavatum and carinatum may occur as isolated abnormalities, they may be associated with Marfan syndrome, congenital heart disease and scoliosis.

### **Pectus Excavatum**

PE, also referred to as a sunken chest or funnel chest, is the most common congenital chest wall deformity, occurring in approximately one in 400 births. It occurs in males more frequently than in females (i.e., 4:1). Although the etiology is not fully understood, it is hypothesized that PE occurs as a result of imbalanced or excessive growth of the lower costal cartilages that leads to a concave appearance of the anterior chest wall. The deformity may be deeper on the right side than on the left and result in a rotation of the sternum. It is typically diagnosed within the first year of life, with wide variations in the degree of sternal depression. During periods of rapid bone growth (e.g., puberty), the appearance of the chest may worsen and symptoms may develop. Moderate to severe deformities may displace the heart into the left chest, decreasing stroke volume and cardiac output. Chest deformities may also depress the sternal volume, adversely affecting the flow of air in and out of the lungs. Symptoms may include fatigue, dyspnea, chest discomfort and palpitations with mild exercise. The body generally compensates by increasing the heart rate with activity to overcome the decreased cardiac output and by more rapid, shallow breathing to compensate for the respiratory deficit. Scoliosis, congenital heart disease and functional heart murmurs can also be associated with PE.

### **Pectus Carinatum**

PC (i.e., pigeon breast or chicken breast) is a congenital chest deformity characterized by an anterior protrusion deformity of the sternum and costal cartilages. Although this condition also affects males more frequently than females (4:1), it occurs less frequently than PE. PC is typically not confirmed until after the growth spurts of early adolescence. This deformity produces a rigid chest and, while symptoms are uncommon, it may result in inefficient respiration as a result of the restrictive chest formation. Three types of PC-related defects have been identified in the literature:

- anterior displacement of the body of the sternum and symmetrical concavity of the costal cartilages
- lateral depression of the ribs on one or both sides of the sternum
- the pouter pigeon breast (the least common of the three): a defect that consists of an upper or chondromalacial prominence with protrusion of the manubrium and depression of the sternal body

The degree of physiological impairment is related to the degree of chest deformity. Patients with PC may develop symptoms as a result of restricted air exchange; complete expiration of air from the lungs may not occur. In addition, pain may result from the secondary pressures that develop from the overgrowth of cartilage. Other conditions that may be associated with PC include frequent respiratory infections, asthma, rickets and cardiac changes.

### **Poland Syndrome**

Poland syndrome (Poland's anomaly, Poland's syndactyly), a rare congenital disorder occurring in approximately one in 30,000 births, is associated with lateral depression of the ribs on one side or both sides of the sternum. The right side of the body is affected twice as often as the left. When the anomaly occurs on the left side of the body, the heart and lungs are vulnerable, because they may be covered only by skin, fascia and pleura (Rush, Ginsberg, 1999). Authors hypothesize that a circulatory defect in the subclavian artery during early pregnancy may be a related cause, but the primary cause is unknown. The condition is characterized by absence or hypoplasia of the pectoralis major muscle, absence or hypoplasia of the pectoralis minor muscle, absence of costal cartilages, hypoplasia of the breast and subcutaneous tissue, and a variety of hand and upper-extremity anomalies. In cases of severe cartilage deficiency, patients may develop lung hernia and paradoxical respiratory motion. In less severe cases, patients may develop a simple flattening of the anterior chest wall.

### **Diagnosis and Evaluation**

There is much controversy regarding abnormal cardiopulmonary function in patients with chest wall deformities, particularly PE. When testing, various factors may affect cardiopulmonary function including the severity of the deformity, the patient's age, and associated conditions, whether the tests are done supine or erect, and whether the tests are done at rest or during exercise (Goretsky, et al., 2004). According to this group of authors, cardiac effects associated with PE generally include decreased cardiac output, mitral valve prolapse and arrhythmias; pulmonary effects associated with PE generally include restricted lung disease, atelectasis, and paradoxical respiration. Regarding PC, patients are

usually asymptomatic; however the deformity may be associated with mitral valve disease, Marfan's syndrome, and scoliosis.

The severity of the chest wall abnormality is dependent upon the depth, symmetry and width of the deformity. Radiographs (i.e., X-ray and computed tomography [CT] scans) are commonly used to determine the degree of chest wall deformity. Children with chest wall deformities are often asymptomatic because in early childhood the chest wall is very pliable, and as children grow, the chest wall becomes more rigid, and the child with PE may become symptomatic. The degree of cardiac compression depends on the depth of the deformity and may result in compromise of the underlying structures. Authors have reported that the amount of pressure from the defect on the heart may affect normal cardiac function and lead to decreased cardiac output and stroke volume, in addition to interference with normal valve function. Typically, increased cardiac pressure results in decreased filling of the atria and a resultant decrease in the ejection fraction of the ventricle. The decrease in cardiac output and stroke volume may contribute to symptoms experienced by the patient such as fatigue and dyspnea, particularly with increased physical activity. CT scans are used to determine abnormalities which include but are not limited to the degree of cardiac compression, pulmonary compression, and cardiac displacement. CT scans are also used to calculate the Haller index, which is used to determine the degree of deformity. The degree of deformity can be determined by dividing the inner width of the chest at the widest point by the distance between the posterior surface of the sternum and the anterior surface of the spine. CT scans are better able to define the ratio of anterior-posterior (AP) borders to transverse diameters, also referred to as the pectus index or Haller Index. Diameters are taken at the deepest level of the sternal depression. CT scan ratios that reveal transverse to AP diameter of greater than 3.25 are considered significant for pectus excavatum. A normal chest has an index of 2.5 (Malek, et al., 2003; Fonkalsrud, 2004).

In most cases, cardiopulmonary function tests will be normal at rest. If the patient is subjected to intense exercise, cardiac output and respiratory function may decrease. It has been reported that patients with PE generally have lower lung volume than normal or that it is mildly reduced, and some authors have attributed respiratory impairment to decreased lung volume. According to textbook sources (Tzelepis and McCool, 2005), "If there is a restrictive impairment it is positively associated with the degree of sternal compression." Furthermore, the author states if there is an associated scoliosis it may be more severe. Patients with PC may develop a rigid chest wall resulting in less efficient respiratory efforts (i.e., poor chest wall expansion with inspiration) with a reduction in vital capacity and an increase in residual air. Respiratory status can be determined with the use of pulmonary function studies, and in some cases the results may reveal a restrictive pattern, including a decrease in pulmonary volumes and reserve.

Echocardiography and/or electrocardiography may be used to evaluate cardiac status. Echocardiograms may occasionally demonstrate mitral valve prolapse. According to a textbook source (Tzelepis and McCool, 2005), other cardiac anomalies that may be associated with compression, such as narrowing of the ventricular outflow tract, compression of the right ventricle and sacculations of the right ventricular wall, have been observed with echocardiography. Measurements of cardiac output and stroke volume have been demonstrated to be decreased in patients with increasing pectus severity index during periods of cardiopulmonary exercise (Malek, et al, 2003). Electrocardiographic abnormalities are common, consisting primarily of right-axis deviation and depressed ST segments, which occur as a result of the rotation of the heart. In approximately 18% of patients, a functional systolic cardiac murmur may be heard, due to compression of the right ventricle (Fonkalsrud, 2004).

Evidence in the peer-reviewed, published scientific literature and textbook sources indicates that in most cases, pulmonary function testing at rest in patients with PE is normal. Consequently, cardiopulmonary exercise testing has been utilized by some authors as a method of determining the degree of preoperative cardiopulmonary impairment and/or postoperative cardiopulmonary improvement in patients with PE. In many cases, despite subjective complaints of exercise intolerance, cardiopulmonary exercise testing is normal. However, patients with more severe deformities often demonstrate abnormal results with cardiopulmonary exercise testing. A textbook source (Boas, 2004) indicates, "lowered ventilatory reserves at peak exercise are common, although the clinical significance of these findings remains unclear." Nonetheless, according to Malek et al. (2006), who published a recent meta-analysis, "It is well established in the clinical exercise physiology literature that examination of pulmonary function during incremental cardiopulmonary exercise testing is as important in patient assessment as assessing cardiovascular function."

## **Surgical Treatment**

Patients with chest wall deformities often have poor body image and self-esteem. Some individuals attempt to cover the defect with clothing and avoid activities that may require exposure of the chest. Indications for surgical correction are controversial and vary widely. Surgical repair is offered primarily as a method of improving cosmesis and psychological factors but may be necessary to improve cardiopulmonary function in some patients, as the disfigurement may be accompanied by physiologic impairment.

The scientific literature is controversial as to whether PE and PC are primarily cosmetic disorders or whether they result in actual physiological impairment of function. Patients with mild PE deformity may be treated with posture and exercise, while patients with mild degrees of PC may be treated with bracing or casting. Most surgical corrections are performed for cosmetic reasons and, in some cases, to improve functional impairment. Authors agree that if patients with severe deformities do not undergo surgical repair in childhood, their symptoms worsen in adulthood.

If surgical repair is performed at an early age, it has been reported there is a high recurrence rate due to periods of rapid bone growth (Fonkalsrud, 2004). While the optimal age for surgical repair is generally between the ages of 11 and 18 years, each case must be reviewed individually for the presence of impaired cardiopulmonary symptoms. In some cases, surgery may be performed in adults to correct pectus deformities. Adults who have uncorrected PE deformity and experience symptoms of activity limitation may undergo surgical repair with low morbidity, short-term limitation of activities and improvement of symptoms (Fonkalsrud, 2003).

Surgery for PE may be performed using any of several techniques, including a sternal osteotomy (i.e., a modified osteotomy that involves supporting, removing and repositioning the sternum) or implantation of a Silastic mold in the subcutaneous space to fill the defect without altering the thoracic cage. Surgical correction often employs a metal bar behind the sternum; the bar may be removed in one to two years, after remodeling has occurred. The standard surgical procedure is the open Ravitch procedure, which involves extensive dissection, cartilage resection and sternal osteotomy. More recently, minimally invasive techniques, including the Nuss procedure (i.e., a minimally invasive repair of pectus excavatum [MIRPE]), have been utilized that involve the insertion of a convex steel bar beneath the sternum through small thoracic incisions. These recently developed minimally invasive methods do not require cartilage resection or osteotomy.

Some authors suggest the use of bracing to exert pressure on the anteroposterior direction to correct or improve PC. Bracing may be recommended for skeletally immature children with mild deformities; however, the candidate must be motivated to wear the brace (Goretsky, et al., 2004). If unsuccessful, bracing does not preclude surgery. The initial surgical repair for PC involves removing the affected cartilages and mobilizing the skin and pectoralis muscle flaps. To straighten the sternum, any one of the following surgeries may be performed:

- an osteotomy
- a subperichondrial resection of the involved costal cartilages
- a wedge-shaped osteotomy in the anterior sternal plate

## **Literature Review**

Several studies have been published in the peer-reviewed scientific literature evaluating surgical repair of chest wall deformities. Many of these studies evaluate and report on the methods of surgical repair and improved cosmetic outcome. Patients with chest deformities often report exercise intolerance and/or poor endurance, and recently some authors have reported on the impact of PE or PC on cardiopulmonary function. Additionally authors have reported on improvement in cardiovascular and/or pulmonary function postsurgical repair.

The results of a meta-analysis conducted by Malek et al. (August 2006) suggested that surgical repair of pectus excavatum significantly improves cardiovascular function. The authors reported the findings of each study in terms of an effect size; effect size is a statistically standardized measure of the study findings so that the numerical values are interpreted consistently across variables and measures in all

studies. The mean weighted effect size for cardiovascular function across all studies meeting inclusion criteria (i.e., eight studies involving 169 patients) was statistically significant. The authors reported an effect size of .59, indicating that the average patient improved by .59 standard deviations on the measures of cardiovascular functioning. In order to ensure there was no bias and that the effect size was not inflated, two post hoc analyses were conducted, and the results were consistent. In conclusion, the authors reported that their findings indicate surgical correction significantly improves cardiovascular function, and their results contradicted arguments that surgical repair is primarily cosmetic, resulting in minimal postoperative cardiovascular improvement.

Additionally, this same group of authors reported the results of a meta-analysis assessing the efficacy of pectus excavatum repair on pulmonary function using similar methods (i.e., effect size). After reviewing 12 studies involving 313 patients the authors concluded surgical repair does not significantly improve pulmonary function (i.e., effect size .08). However, the authors stated that their findings may be the result of evaluating studies that conducted pulmonary function testing under conditions in which pectus excavatum does not manifest itself. It has been reported that the dynamics of ventilation differ greatly when measured at rest rather than during exercise, and better measures of chest wall deformity would be obtained through incremental exercise testing. Measures of pulmonary function taken at rest were typical of the studies in the current meta-analysis (Malek, et al., October 2006).

Rowland and associates (2005) conducted a cross-sectional comparison of cardiac and ventilatory variables at rest and during maximal exercise in a study group that consisted of 12 boys with moderate to severe PE and a control group of 20 boys without any musculoskeletal deformity. The following outcomes were measured: endurance fitness, respiratory rate, tidal volume, minute ventilation, cardiac output, and stroke volume. Exercise was performed in an air-conditioned laboratory and consisted of upright cycling until an exhaustive effort was achieved. Comparisons were made at rest, at a given absolute submaximal workout and during maximal exercise. At rest, the stroke index was 13% higher in the control group, and resting tidal volume was significantly diminished in the PE group. Findings were similar during the absolute submaximal workout. At maximal exercise, the trend for lower stroke volume in the PE group continued, and tidal volume remained greater in the control group. The results of this study suggest patients with severe PE demonstrate lower endurance fitness than controls. Nevertheless, the authors reported, "There is a wide variability of exercise capacity in children with PE, and implicating a pectus defect as being responsible for claims of exercise intolerance in a given patient is difficult."

In 2005, Lawson and colleagues reported on the impact of PE on pulmonary function before and after repair with the Nuss procedure. The authors studied preoperative repair pulmonary function testing (PFT) from 408 patients approved for surgery. The patient population was primarily located out of state. In a subset of 45 patients, preoperative and post-bar removal PFT results were compared. At the time of publication, sufficient time had not elapsed for the bar to be removed in the remaining patients, or they did not return for post-bar removal PFTs (PFT studies were not available for 175 patients who were post-bar removal). The preoperative forced vital capacity and expiratory volumes were lower than normal by 13%, and the forced expiratory flow rate was lower than normal by 20%. The postoperative group had statistically significant improvement in all parameters; however, the volumes were still below normal. The authors concluded that the study did support a small but significant improvement in pulmonary function after bar removal. The authors did not assess cardiopulmonary function under exercise conditions.

Bawazir et al. (2005) conducted a prospective study of patients who underwent closed repair of PE. The authors studied the effects over time of PE on pulmonary function, cardiac function, progressive exercise tolerance and the patient's perception of appearance and subjective ability to exercise. The authors believe that, over time, the patient's pulmonary function and exercise tolerance would improve. The patients were evaluated preoperatively, at three to four months post-operation, at 21 to 24 months post-operation and again at three months post-bar removal. A total of 48 patients completed the initial preoperative assessment and then the three-month postoperative follow-up; 22 completed the 21-month follow-up; and 11 patients were followed to bar removal. Pulmonary function studies revealed a reduction in forced vital capacity and forced expiratory volume in one second at initial evaluation with normal total lung capacity and vital capacity. At three months post-operation, all values showed a remarkable decline. By the phase of final bar removal, the forced vital capacity and forced expiratory volume in one second had both improved significantly and were near initial preoperative values. According to the authors, pulmonary function remained below normative values for patients without PE of similar size and age.

Cardiac function was increased at three months and maintained thereafter. Initially, exercise tolerance declined but increased by the 21-month evaluation and after bar removal. The patients reported improvement in the ability to exercise immediately after bar insertion. The results of this study demonstrate that after closed repair of PE, there is an immediate subjective improvement in the ability to exercise, paralleled by an improvement in cardiac output. In addition, there was an early postoperative decline in pulmonary function, although this did improve over time. Further studies are needed to determine if these results are maintained or improve further over time.

Bohosiewicz et al (2004) conducted a prospective case series assessing outcomes after surgical treatment using the Nuss procedure for patients with pectus excavatum. The study population consisted of 66 children age one to 19 years. The pectus index ranged from 2.8 to 5.8. The main indications for surgery were chest deformities that were unacceptable for the children and parents. Some of the children had minor pulmonary and cardiac abnormalities, and only two were symptomatic (i.e., dyspnea, particularly with respiratory infections). Preoperative evaluation included chest x-ray, CT scan, pulmonology consultation with pulmonary function studies, and cardiology consultation. The surgeons used a modified technique; the supporting bar was inserted directly from the right side with no thoroscope, no sharp tools and no guiding tape, in contrast to the Nuss procedure, which uses thoracoscopy. Follow-up ranged from the time of operation to 56 months. Early cosmetic results were very good and similar to those reported by Dr. Nuss. The bar was removed in 24 children two years after the surgery. No postoperative complications have been reported after bar removal. One child did experience pectus excavatum recurrence six months after bar removal; deformation recurrence has been reported to be 7.8% after bar removal. The result of this clinical study supports good results in correcting chest deformity using a modified Nuss procedure for patients with pectus excavatum.

Goretsky et al. (2004) published a review and reported on their experience with surgical correction of chest wall deformities. In a 16-year period of time, 1124 patients were evaluated for chest wall deformity, and 608 were severe enough to require surgery. Of the 608 patients, 557 had their primary procedure, described as a minimally invasive technique, performed at the author's facility, and 51 had re-operations. Of the 557 patients, 550 had PE and seven had mixed PE/PC. According to the authors, patients with mild to moderate PE deformity are initially treated with a posture and exercise program and evaluated at six-month intervals with the goal of halting progression of disease. Patients with a severe deformity or with documented progression undergo posture and exercise programs in addition to objective studies (e.g., thoracic CT scans, pulmonary function tests and cardiac evaluation, including electrocardiogram (EKG) and Echo studies) to determine whether or not the condition is severe enough to warrant surgical correction. The authors suggest that when using the criteria below, 50% of the patients are found to have a deformity severe enough to warrant surgery. The criteria used by the authors to demonstrate severe PE and need for surgical repair include two or more of the following:

- a Haller CT index greater than 3.25
- pulmonary function studies that indicate restrictive or obstructive airway disease
- a cardiology evaluation, where the compression is causing murmurs, mitral valve prolapse, cardiac displacement, or conduction abnormalities on the echocardiogram or EKG
- documentation of progression of the deformity with associated physical symptoms other than isolated concerns of body image
- a failed Ravitch procedure
- a failed minimally invasive procedure

Fonkalsrud and Anselmo (2004) reported on 154 patients with symptomatic PC (mean severity index = 1.76) who underwent PC repair at the University of California/Los Angeles (UCLA) Medical Center using modifications of the Ravitch approach. Each of the 154 patients with reduced endurance or dyspnea with mild exercise demonstrated functional improvement within six months of surgery. The last 60 patients underwent less extensive open repair, and experienced less frequent complications, less severe pain, shorter hospitalization and improved postoperative results than did the patients who had undergone the more extensive repair used earlier. The authors' findings support improved exercise tolerance, endurance, alleviated respiratory symptoms and improved cosmetic appearance for PC repair.

Authors have suggested that cardiac filling is decreased in patients with PE, resulting in limited stroke volume, especially during exercise. Zhao et al (2000) evaluated whether limitation of venous return to the heart contributes to exercise intolerance in patients with PE. Thirteen patients with PE were enrolled in the study, along with a control group of 20 healthy patients. The authors assessed lung function, incremental exercise capacity and stroke volume in the sitting and supine positions. The study results demonstrated that patients with PE who were exercising in a sitting position had reduced oxygen uptake and stroke volume and, when exercising in a supine position, oxygen uptake and stroke volume values were similar to those demonstrated in the control group. The authors concluded that the exercise capacity in the upright position was affected by reduced filling of the heart.

Haller and Loughlin (2000) studied pulmonary and cardiac function with incremental exercise before and after surgery in 35 adolescents with PE and 10 age-matched healthy control subjects. Prior to surgery, the forced vital capacity (FVC) of the PE group was lower than that of the control group. There was no change in FVC after surgery. Prior to surgery, 58% of the PE patients had exercise limitations; 66% of the patients improved after surgery. PE subjects exercised at similar workload to the control group. After surgery, PE subjects exercised longer and had higher oxygen-pulse (i.e., measure of cardiac output) than before surgery. While some patients exhibited mild restrictive lung function, surgical repair did not influence this mild degree of restriction. The author's conclusions suggest that PE repair improves cardiopulmonary function.

Some authors have reported improvement in cardiopulmonary functioning postoperatively for treatment of PE and PC (Malek, Fonkalsrud, 2004; Sigalet, et al., 2003; Haller, Loughlin, 2000; Fonkalsrud et al., 1994). Authors have reported that improvement is seen only with increased periods of exercise and not with routine pulmonary function testing at rest. Patient selection criteria are dependent upon the degree of deformity and degree of activity intolerance demonstrated through cardiopulmonary testing. While reported outcomes have been controversial, differences among studies may be related to patient selection criteria, the degree of severity of the deformity, the surgical technique utilized, and future growth effects. A review of the published, peer-reviewed scientific literature indicates that surgical repair for PE or PC does improve postoperative cardiopulmonary functioning and exercise tolerance and is considered a viable treatment option for selected candidates with severe deformity and functional impairment.

Primarily, patients with Poland syndrome present for surgical reconstruction to improve physical appearance and correct breast asymmetry. Surgical treatment often consists of reconstruction of the chest wall muscles, breast, and nipple on the affected side by a plastic surgeon. Surgery is performed early (approximately age 13) in males. In females, however, surgery is often deferred until breast development is complete. Surgical treatment involves the use of tissue expanders and muscle transfers, to match normal development of the unaffected side and, ultimately, of breast reconstruction, with reconstruction of the axillary line and correction of infraclavicular flattening.

Patients who present with absent ribs are considered candidates for surgical repair (Townsend, 2004). In such cases, operative reconstruction may eliminate paradoxical motion, improving respiratory impairment. For more severe conditions, reconstructive surgery also provides protection of the underlying heart and lung structures. While there are a variety of surgical techniques to correct the deformity, a common approach is to use the latissimus dorsi muscle with autologous rib grafts to reconstruct the chest wall.

### **Professional Organizations/Societies**

A review of current professional society recommendations and policy statements from the American Thoracic Society and the American Academy of Pediatrics does not confirm existence of established guidelines for the treatment of congenital chest wall deformities. Regarding breast augmentation in teenagers, the American Society of Plastic Surgeons (ASPS) has a policy statement that supports breast augmentation for reconstructive purposes related to congenital defects (ASPS, 2004). Regarding cardiopulmonary exercise testing (CPET) with ventilatory gas analysis, the American College of Cardiology/American Heart Association (ACC/AHA) (Gibbons, et al., 2002), and the American Thoracic Society/American College of Chest Physicians (ATS/ACCP) (ATS/ACCP, 2002) have established indications and guidelines for exercise testing; however, these recommendations do not address the utility of CPET for chest deformities such as PE, PC or those associated with Poland syndrome.

### **Summary**

Congenital chest wall deformities may result in functional limitations such as activity intolerance related to cardiac or respiratory impairment. Patients often report symptoms which include mild to moderate exercise limitation, respiratory infections, and asthmatic conditions. In many cases, the deformity does not lead to a functional impairment, and treatment is considered to be solely cosmetic in nature. Authors have reported in the peer-reviewed, published scientific literature that surgical correction of the deformity can be an effective treatment and may reduce cardiopulmonary symptoms.

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## Coding/Billing Information

**Note:** This list of codes may not be all-inclusive.

**When medically necessary:**

CPT <sup>®</sup> * Codes	Description
21740	Reconstructive repair of pectus excavatum or carinatum; open
21742	Reconstructive repair of pectus excavatum or carinatum; minimally invasive approach (Nuss procedure), without thoracoscopy
21743	Reconstructive repair of pectus excavatum or carinatum; minimally invasive approach (Nuss procedure), with thoracoscopy

HCPCS Codes	Description
	No specific codes

ICD-9-CM Diagnosis Codes	Description
754.81	Pectus excavatum
754.82	Pectus carinatum
754.89	Other specified nonteratogenic anomalies
738.3	Acquired deformity of chest and rib

\*Current Procedural Terminology (CPT<sup>®</sup>) © 2006 American Medical Association: Chicago, IL.

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## References

1. American Society of Plastic Surgeons (ASPS). Breast augmentation in teenagers. Policy statement. Approved 2004 Dec. Accessed January 2007. Available at URL address: [http://www.plasticsurgery.org/medical\\_professionals/Policy\\_Statements/loader.cfm?url=/commonspot/security/getfile.cfm&PageID=15619](http://www.plasticsurgery.org/medical_professionals/Policy_Statements/loader.cfm?url=/commonspot/security/getfile.cfm&PageID=15619)
2. American Society of Plastic Surgeons (ASPS). Pediatric Plastic surgery, part II: congenital anomalies. Accessed January 2007. Available at URL address: [http://www.plasticsurgery.org/medical\\_professionals/publications/Physician-Counseling-Guides-Pediatric-Congenital-Anomalies.cfm](http://www.plasticsurgery.org/medical_professionals/publications/Physician-Counseling-Guides-Pediatric-Congenital-Anomalies.cfm)
3. American Thoracic Society/American College of Chest Physicians (ATS/ACCP) Joint Statement on Cardiopulmonary Exercise Testing (Corrected version). 2001. Am J Respir Crit Care Med. 2003; 167:211-277. (Erratum letter: Am J Respir Crit Care Med. 2003; 167:1451-52.) Available at URL address: <http://www.thoracic.org/sections/publications/statements/pages/pfet/cardioexercise.html>
4. Bawazir OA, Montgomery M, Harder J, Sigalet DL. Midterm evaluation of cardiopulmonary effects of closed repair of pectus excavatum. J Pediatr Surg. 2005 May;40(5):863-7.

5. Bento L, Martinez M, Conde J, Perez Martinez A, Esparza J, Gonzalez A. Early surgery in Poland syndrome. *Cir Pediatr*. 2002 Jul;15(3):91-3.
6. Boas S. Pectus Excavatum. *Skeletal Diseases Influencing Pulmonary Function*. In: Behrman RE, Kliegman RM, Jenson HB, editors; *Nelson Textbook of Pediatrics*, 17th ed. Copyright © 2004. Ch 409.
7. Bohosiewicz J, Kudela G, Koszutski T. Results of Nuss procedure for the correction of pectus excavatum. *Eur J Pediatr Surg*. 2005 Feb;15(1):6-10.
8. Borschel GH, Izenberg PH, Cederna PS. Endoscopically assisted reconstruction of male and female Poland syndrome. *Plast Reconstr Surg*. 2002 Apr;109(5):1536-43.
9. Cahill JL, Lees GM, Robertson HT. A summary of preoperative and postoperative cardiorespiratory performance in patients undergoing pectus excavatum and carinatum repair. *J Pediatr Surg*. 1984 Aug;19(4):430-3.
10. Coln E, Carrasco J, Coln D. Demonstrating relief of cardiac compression with the Nuss minimally invasive repair for pectus excavatum. *J Pediatr Surg*. 2006 Apr;41(4):683-6; discussion 683-6.
11. Colombani PM. Recurrent chest wall anomalies. *Semin Pediatr Surg*. 2003 May;12(2):94-9.
12. Davis JT, Weinstein S. Repair of the pectus deformity: results of the Ravitch approach in the current era. *Ann Thorac Surg*. 2004 Aug;78(2):421-6.
13. Fonkalsrud EW, Anselmo DM. Less extensive techniques for repair of pectus carinatum: the undertreated chest deformity. *J Am Coll Surg*. 2004 Jun;198(6):898-905.
14. Fonkalsrud EW, Beanes S. Surgical management of pectus carinatum: 30 years' experience. *World J Surg*. 2001 Jul;25(7):898-903.
15. Fonkalsrud EW, Bustorff-Silva J. Repair of pectus excavatum and carinatum in adults. *Am J Surg*. 1999 Feb;177(2):121-4.
16. Fonkalsrud EW. Current management of pectus excavatum. *World J Surg*. 2003 May;27(5):502-8. [Epub. 2003 Apr 28]
17. Fonkalsrud EW, DeUgarte D, Choi E. Repair of pectus excavatum and carinatum deformities in 116 adults. *Ann Surg*. 2002 Sep;236(3):304-12; discussion 312-4.
18. Fonkalsrud EW. Management of pectus deformities in female patients. *Am J Surg*. 2004 Feb;187(2):192-7.
19. Fonkalsrud EW, Mendoza J. Open repair of pectus excavatum and carinatum deformities with minimal cartilage resection. *Am J Surg*. 2006 Jun;191(6):779-84.
20. Fonkalsrud EW, Salman T, Guo W, Gregg JP. Repair of pectus deformities with sternal support. *J Thorac Cardiovasc Surg*. 1994;107:37-42.
21. Frey AS, Garcia VF, Brown RL, Inge TH, Ryckman FC, Cohen AP, et al. Nonoperative management of pectus carinatum. *J Pediatric Surg*. 2006 Jan;41(1):40-5;discussion 40-5.
22. Gibbons RJ, Balady GJ, Bricker JT, Chaitman BR, Fletcher GF, Froelicher VF, et al. ACC/AHA 2002 guideline update for exercise testing: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Committee on

Exercise Testing). 2002. American College of Cardiology Web site. Accessed January 29, 2007. Available at URL address: [www.acc.org/clinical\\_guidelines/exercise/dirIndex.htm](http://www.acc.org/clinical_guidelines/exercise/dirIndex.htm)

23. Glicenstein J. Corrective surgery of thoracic anomalies in Poland syndrome: general review of 20 patients. *Annales de Chirurgie Plastique et Esthetique*. 2001 Dec;46(6):640-51.
24. Goretsky MJ, Kelly RE, Croitoru D, Nuss D. Chest wall anomalies: pectus excavatum and pectus carinatum. *Adolesc Med Clin*. 2004 Oct;15(3):455-71.
25. Haecker FM, Bielek J, von Schweinitz D. Minimally invasive repair of pectus excavatum (MIRPE)-the Basal experience. *Swiss Surg*. 2003;9(6):289-95.
26. Haller JA Jr., Loughlin GM. Cardiorespiratory function is significantly improved following corrective surgery for severe pectus excavatum: proposed treatment guidelines. *J Cardiovasc Surg (Torino)*. 2000 Feb;41(1):125-30.
27. Inge TH, Owings E, Blewett CJ, Baldwin CE, Cain WS, Hardin W, Georgeson KE. Reduced hospitalization cost for patients with pectus excavatum treated using minimally invasive surgery. *Surg Endosc*. 2003 Oct;17(10):1609-13. Epub 2003 July 21.
28. Lawson ML, Mellins RB, Tabangin M, Kelly Jr RE, Croitoru DP, Goretsky MJ, Nuss D. Impact of pectus excavatum on pulmonary function before and after repair with the Nuss procedure. *J Pediatr Surg*. 2005 Jan;40(1):174-80;discussion 180.
29. Malek MH, Berger DE, Housh TJ, Marelich WD, Coburn JW, Beck TW. Cardiovascular function following surgical repair of pectus excavatum: a metaanalysis. *Chest*. 2006 Aug;130(2):506-16.
30. Malek MH, Berger DE, Marelich WD, Coburn JW, Beck TW, Housh TJ. Pulmonary function following surgical repair of pectus excavatum: a meta-analysis. *Eur J Cardiothorac Surg*. 2006 Oct;30(4):637-43. Epub 2006 Aug 8.
31. Malek MH, Fonkalsrud EW. Cardiorespiratory outcome after corrective surgery for pectus excavatum: a case study. *Med Sci Sports Exerc*. 2004 Feb;36(2):183-90.
32. Malek MH, Fonkalsrud EW, Cooper CB. Ventilatory and cardiovascular responses to exercise in patients with pectus excavatum. *Chest*. 2003 Sep;124(3):870-82.
33. Mansour KA, Thourani VH, Odessey FA, Durham MM, Miller JI Jr, Miller DI. Thirty-year experience with repair of pectus deformities in adults. *Ann Thorac Surg*. 2003 Aug;76(2):391-5;discussion 395.
34. Mcguigan RM, Azarow KS. Congenital chest wall defects. *Surg Clin N Am*. 2006 Aug;86(2):353-70,ix.
35. National Institute for Clinical Excellence (NICE). Interventional procedures: minimally invasive placement of pectus bar. Guidance. IP guidance number: IPG0003. Accessed January 2007. Available at URL address: <http://www.nice.org.uk/page.aspx?o=56898>
36. Nuss D, Kelly RE Jr., Croitoru DP, Katz ME. A 10-year review of a minimally invasive technique for the correction of pectus excavatum. *J Pediatr Surg*. 1998 Apr;33(4):545-52.
37. Park HJ, Lee SY, Lee CS, Youm W, Lee KR. The Nuss procedure for pectus excavatum: evolution of techniques and early results on 322 patients. *Ann Thorac Surg*. 2004 Jan;77(1):289-95.
38. Petersen C, Leonhardt J, Duderstadt M, Karck M, Ure BM. Minimally invasive repair of pectus excavatum - shifting the paradigm? *Eur J Pediatr Surg*. 2006 Apr;16(2):75-8.

39. Peterson RJ, Young WG Jr., Godwin JD, Sabiston DC Jr., Jones RH. Noninvasive assessment of exercise cardiac function before and after pectus excavatum repair. *J Thorac Cardiovasc Surg.* 1985 Aug;90(2):251-60.
40. Rowland T, Moriarty K, Banever G. Effect of pectus excavatum deformity on cardiorespiratory fitness in adolescent boys. *Arch Pediatr Adolesc Med.* 2005 Nov;159(11):1069-73.
41. Rush VW, Ginsberg RJ. Chest wall. In: Schwartz SI, Shires GT, Spencer FC, Daly JM, Fischer JE, Galloway AC, editors. *Principles of surgery.* Chapter 16: chest wall, pleura, lung, and mediastinum. The McGraw-Hill Companies, Inc.; 1999.
42. Sigalet DL, Montgomery M, Harder J. Cardiopulmonary effects of closed repair of pectus excavatum. *J Pediatr Surg.* 2003 Mar;38(3):380-5; discussion 380-5.
43. Townsend. Chest wall. In: Sabiston textbook of surgery, 17<sup>th</sup> ed. Elsevier; 2004. p. 1711-4.
44. Tzelepis GE, McCool FD. The Lings and Chset Wall Disease. Pectus excavatum. In: Mason RJ, Murray JF, Broaddus VC, Nadel JA, editors; In Mason: Murray & Nadel's Textbook of Respiratory Medicine. 4<sup>th</sup> ed. W. B. Saunders Company; Copyright © 2005 Ch 83.
45. Warner BW. What's new in pediatric surgery. *J Am Coll Surg.* 2004 Aug;199(2):273-85.
46. Wilhelmi BJ. Breast, Poland syndrome. eMedicine. Updated 2002 Aug. Accessed Jan, 2005. Available at URL address: <http://www.emedicine.com/plastic/topic132.htm>
47. Williams AM, Crabbe DC. Pectus deformities of the anterior chest wall. *Paediatr Respir Rev.* 2003 Sep;4(3):237-42.
48. Yalamanchili K, Summer W, Valentine V. Pectus excavatum with inspiratory inferior vena cava compression: A new presentation of pulsus paradoxus. *Am J Med Sci.* 2005 Jan;329(1):45-7.
49. Zhao L, Feinberg MS, Gaides M, Ben-Dov I. Why is exercise capacity reduced in subjects with pectus excavatum? *J Pediatr.* 2000 Feb;136(2):163-7.