Policy

*Please see amendment for Pennsylvania Medicaid at the end of this CPB.*

I. Aetna considers surgical repair of severe pectus excavatum deformities that cause functional deficit medically necessary when done for medical reasons in members who meet all of the following criteria:

A. Well-documented evidence of complications arising from the sternal deformity. Complications include but may not be limited to:

- Cardiac compression, displacement results in decreased cardiac output demonstrated by echocardiography; or
- Reduced lung capacity as demonstrated by a total lung capacity (TLC) less than or equal to 80% of predictive value per pulmonary function testing; or
- There is objective evidence of exercise intolerance due to reduced lung capacity as documented by exercise pulmonary function tests that are below the predicted values; and
B. An electrocardiogram or echocardiogram has been done if a heart murmur or known heart disease is present to define the relationship of the cardiac problem to the sternal deformity; and
C. A CT scan of the chest demonstrates a pectus index, derived from dividing the transverse diameter of the chest by the anterior-posterior diameter, greater than 3.25.

Aetna considers surgical repair of pectus excavatum cosmetic when criteria are not met.

II. Aetna considers surgical reconstruction of musculoskeletal chest wall deformities (congenital absence or hypoplasia of pectoralis major and minor muscles; congenital partial absence of the upper costal cartilage) associated with Poland's syndrome that cause functional impairment medically necessary (also see CPB 0185 - Breast Reconstructive Surgery (../100_199/0185.html)).

III. Aetna considers bracing and surgical procedures to correct pectus carinatum cosmetic because this deformity does not cause physiologic disturbances from compression of the heart or lungs.

IV. Aetna considers the following interventions for the treatment of pectus excavatum experimental and investigational because their effectiveness has not been established:

- The magnetic mini-mover procedure
- The vacuum bell
- Dynamic Compression System.

Background

Chest wall deformities result from abnormal growth of the rib cartilages which pushes the sternum either inward or outward, away from the plane of the chest. The deformities can range from mild, symmetric indentions or protrusions, to severe asymmetric deformities. The appearance of the deformity often changes dramatically around the time of adolescent growth. Chest wall deformities may be corrected using various techniques; most require surgical intervention.
Pectus excavatum (PE) is often a cosmetic defect, but it may have varied anatomic and symptomatic presentations. There is no conclusive evidence supporting the existence of a functional component whose physiological basis can be consistently defined.

Pectus excavatum (PE) surgery techniques include, but may not be limited to:

- **Nuss procedure**: Minimally invasive procedure in which an incision is made on each side of the chest wall. A concave bar is then inserted through one side of the chest under the sternum (breastbone) using a surgical clamp. Once the bar is pulled through, it is rotated, allowing the sternum to bend outward. Sutures are placed to temporarily attach the bar which eventually becomes held secure by muscle tissues growth occurring during recovery. The bar is left in place for several months or years.

- **Ravitch procedure**: Named for the surgeon that developed it, this technique involves removing the ends of the ribs in the area that is depressed at the sternum. The sternum is then straightened out at the point it turns downward by breaking it horizontally. Stitches and a metal bar are used to hold the sternum in place under the skin. After two to three years, when remolding has taken place, the bar may be removed.

Until recently, the indications for surgery in patients with PE were based solely on clinical judgment because the extensive literature on PE demonstrates that there is a discordance between patients' subjective assessment of shortness of breath and objective measures of cardiorespiratory function. In more recent years, the judgment of when to proceed with surgery has been made more objective by following the pectus index criteria advocated by Haller for surgical intervention. Computed tomography (CT) scans used in patients being evaluated for surgery document more clearly the severity of the fore-shortening of the antero-posterior diameter of the chest, the degree of cardiac compression and displacement, the degree of lung compression and other unexpected problems. It clarifies the need for operation by showing the dramatic internal morbidity of what is often portrayed as a "cosmetic" deformity.

The Haller index, also called the pectus index (PI) or pectus severity index (PSI), is the most commonly used scale for determining the severity of chest wall deformities. The index is defined as the width of the chest divided by the distance between the sternum and spine at the point of maximal depression. The normal
value is 2.54. In individuals with pectus carinatum, a lower PSI indicates a more severe deformity in contrast to individuals with excavatum, in which a higher PSI indicates a more severe deformity. An index greater than 3.25 is considered severe for pectus excavatum. Computerized tomography (CT) or magnetic resonance imaging (MRI) may be used to determine the index.

As originally described by Sir Alfred Poland, Poland's syndrome consists of absence or hypoplasia of the pectoralis major and minor muscles, hypoplasia or absence of nipple and breast, hypoplasia of subcutaneous fat, absence of axillary hair, and partial absence of the upper costal cartilages and portions of ribs, usually the 2nd, 3rd, and 4th. The absence of the sternal head of the pectoralis major muscle is considered the minimal expression of this syndrome (Wilhelmi and Cornette, 2002). Brachysyndactyly, ectrodactyly, and ectromelia are frequently described associations.

Poland syndrome surgery techniques include, but may not be limited to: augmentation with tissue from the opposite breast, musculocutaneous flap to fill hollow space on the exterior of the chest, prosthetic augmentation, and surgical repair of the chest wall.

In children with very severe deformity, staged procedures involving split rib grafts from the contralateral side combined with Teflon felt or Marlex mesh have been advocated. This results in a stable chest wall, abolition of paradoxical movement, and protection of the subjacent viscera. In the absence of the pectoralis major and with deficient breast and subcutaneous tissue, the chest is still visibly asymmetric. As soon as the asymmetry becomes a problem for the adolescent female patient, a round tissue expander can be placed beneath the pectoralis muscle and hypoplastic breast through a transaxillary incision, to avoid scars on the breast itself. The prosthesis is then inflated at appropriate intervals to maintain symmetry until development of the opposite breast stabilizes, at which time the expander can be replaced with a prosthetic mammary implant or an autologous soft-tissue transfer using pedicled myocutaneous flaps.

Pectus carinatum is a developmental deformity of the chest characterized by a protrusion of the sternum and ribs. It is extremely uncommon that pectus carinatum will cause a functional/physiological deficit. Pectus carinatum (PC) orthotic compression bracing uses a customized chest wall brace which applies direct, constant pressure to the protruding area of the chest with the goal of reshaping the
chest and sternum. The brace has front and back compression pads that are attached to aluminum bars which are bound together by a tightening mechanism. Regular monitoring and adjustment is generally required. PC surgery includes removing the affected cartilages to mobilize both the pectoralis (chest muscles) flaps and mobilizing the skin to straighten the sternum. These surgical techniques include, but may not be limited to: costal cartilage subperichondral resection, osteotomy, and wedge shaped osteotomy in the anterior sternal plate.

Schier et al (2005) described their experience in using a vacuum to pull the abnormal chest wall outward in patients with PE. A suction cup was used to create a vacuum at the chest wall. A patient-activated hand pump was used to reduce pressure up to 15 % below atmospheric pressure (atm). The device was used by 60 patients (56 males and 4 females), aged 6.1 to 34.9 years (median of 14.8 years), for a minimum of 30 mins, twice-daily, up to 5 hours per day (median of 90 mins). Patient progress was documented using photography, radiography, and plaster casts of the defect. In 14 children this method was used during the Nuss procedure to enlarge the retrosternal space for safer passage of the introducer. Follow-up occurred between 2 and 18 months (median of 10 months). Computed tomographic scans showed that the device lifted the sternum and ribs within 1 to 2 mins; this was confirmed thoracoscopically during the Nuss procedure. The suction cup enlarged the retrosternal space for safer passage of the introducer. Initially, the sternum sank back after few minutes. After 1 month, an elevation of 1 cm was noted in 85 % of the patients. After 5 months, the sternum was lifted to a normal level in 12 patients (20 %) when evaluated immediately after using the suction cup. All patients exhibited moderate subcutaneous hematoma, although the skin was not injured. One patient suffered from transient paresthesia in the right arm and leg; 2 patients experienced orthostatic disturbances during the first application of the suction cup. There were no other complications. In patients with PE, application of a vacuum effectively pulled the depressed anterior chest wall forward. The initial results proved dramatic, although it is not yet known how much time is required for long-term correction. The authors concluded that this vacuum method holds promise as a valuable adjunct treatment in both surgical and non-surgical correction of PE.

Haecker and Mayr (2006) examined the benefits of conservative treatment of patients with PE by means of the vacuum bell. A suction cup is used to create a vacuum at the anterior chest wall. A patient-activated hand pump is used to reduce the pressure up to 15 % below atm. Three different sizes of vacuum bell exist that
were selected according to the individual patient's age. When creating the vacuum, the lift of the sternum was obvious and remained for a different time period. The device should be used for a minimum of 30 mins (twice-daily), and may be used up to a maximum of several hours daily. Presently, a 12- to 15-month course of treatment is recommended. In addition, the device was used intra-operatively during the minimally invasive repair (MIRPE) procedure to enlarge the retrosternal space to ensure safer passage of the introducer in a few patients. A total of 34 patients (31 males and 3 females), aged 6 to 52 years (median of 17.8 years) used the vacuum bell for 1 to maximum 18 months (median of 10.4 months). Follow-up included photography and clinical examination every 3 months. Computed tomographic scans showed that the device lifted the sternum and ribs immediately. In addition, this was confirmed thoracoscopically during the MIRPE procedure. After 3 months, an elevation of more than 1.5 cm was documented in 27 patients (79 %). After 12 months, the sternum was lifted to a normal level in 5 patients (14.7 %). Relevant side effects were not noted. The authors concluded that the vacuum bell has proved to be an alternative therapeutic option in selected patients with PE. Moreover, they stated that while the initial results proved to be dramatic, long-term results are so far lacking, and further evaluation and follow-up studies are necessary.

Haecker (2011) provided additional data on the 2006 trial by Haecker and Mayr; but the conclusion remained unchanged. A total of 133 patients (110 males and 23 females) aged from 3 to 61 years (median of 16.21 years) used the vacuum bell for 1 to a maximum of 36 months. Computed tomographic scans showed that the device lifted the sternum and ribs immediately. In addition, this was confirmed thoracoscopically during the MIRPE procedure. A total of 105 patients showed a permanent lift of the sternum for more than 1 cm after 3 months of daily application; 13 patients stopped the application and underwent MIRPE. Relevant side effects were not noted. The authors concluded that the vacuum bell has proved to be an alternative therapeutic option in selected patients suffering from PE. The initial results proved to be dramatic, but long-term results are so far lacking, and further evaluation and follow-up studies are necessary.

Harrison et al (2007) noted that correction of PE results in measurable improvement in lung capacity and cardiac performance as well as improved appearance and self-image. The Nuss and modified Ravitch approaches attempt to correct the chest wall deformity by forcing the sternum forward in 1-step and holding it in place using a metal strut. The initial operation requires extensive
Manipulation under general anesthesia and results in post-operative pain, requiring hospitalization and regional anesthesia. Pain and disability may last for weeks. Both procedures are expensive. A better principle would be a gradual bit-by-bit repair via small increments of pressure applied over many months. These researchers developed the magnetic mini-mover procedure (3MP) and applied this strategy to correct PE. The procedure uses magnetic force to pull the sternum forward. An internal magnet implanted on the sternum and an external magnet in a non-obtrusive custom-fitted anterior chest wall orthosis produce an adjustable outward force on the sternum. Outward force is maintained until the abnormal costal cartilages are remodeled and the pectus deformity is corrected. These investigators implanted a magnet in human skeletons and measured the force applied to the sternum when the distance between the internal and external magnets was varied in increments. With the 2 magnets 1 cm apart, the outward force was adequate to move the sternum at least 1 cm. They also mapped the magnetic field in the 2-magnet configuration and found that maximum field strengths at the surface of the heart and at the outer surface of the orthosis were at safe levels. The authors concluded that the 3MP allows correction of PE by applying magnetic force over a period of months. Crucial questions raised during the design, re-design, and simulation testing have been satisfactorily answered, and the authors have received a Food and Drug Administration (FDA) Investigation Device Exemption (G050196/A002) to proceed with a phase I to II clinical trial.

Harrison et al (2012) performed a pilot study of safety, probable efficacy, and cost-effectiveness of 3MP. A total of 10 otherwise healthy patients, aged 8 to 14 years, with severe pectus excavatum (pectus severity index [PSI] greater than 3.5) underwent 3MP treatment (mean of 18.8 +/- 2.5 months). Safety was assessed by post-implant and post-explant electrocardiograms and monthly chest x-rays. Efficacy was assessed by change in pectus severity index as measured using pre-treatment and post-treatment computed tomographic scan. Cost of 3MP was compared with that of standard procedures. The 3MP device had no detectable ill effect. Device weld failure or mal-positioning required revision in 5 patients. Average wear time was 16 hrs/day. Pectus severity index improved in patients in the early or mid-puberty but not in patients with non-compliant chest walls. Average cost for 3MP was $46,859, compared with $81,206 and $81,022 for Nuss and Ravitch, respectively. The authors concluded that the 3MP is a safe, cost-effective, outpatient alternative treatment for pectus excavatum that achieves good results for patients in early and mid-puberty stages.
Ji and Luan (2012) reviewed the current development in therapy of congenital funnel chest. The main therapies for congenital funnel chest are thoracoplasty (Ravitch sternum elevation procedure and minimal invasive Nuss procedure) and prosthesis implantation. The magnetic mini-mover procedure and the vacuum bell are still in the research phase.

An UpToDate review on “Pectus excavatum: Treatment” (Mayer, 2013) states that “Currently, surgical correction for PE is done with either the modified Ravitch procedure (open resection of the subperichondrial cartilage and sternal osteotomy, with placement of an internal stabilizing device), or the Nuss procedure (minimally invasive technique in which a curved bar is inserted to lift the sternum; the bar is removed about two years later)”.

An UpToDate review on “Pectus carinatum” (Nuchtern and Mayer, 2014) states that “In more than 90 percent of patients, pectus carinatum deformity is first noted during early adolescence, and it often worsens dramatically during the adolescent growth spurt. The defect does not resolve spontaneously. The vast majority of patients have no physiologic symptoms, and cosmetic appearance is the primary concern … The decision of whether to treat depends on the severity of the defect, and the patient and family's level of concern”.

Johnson et al (2014) compared outcome measures of current PE treatments, namely the Nuss and Ravitch procedures, in pediatric and adult patients. Original investigations that stratified PE patients based on current treatment and age (pediatric = 0 to 21 years; adult 17 to 99 years) were considered for inclusion. Outcome measures were: operation duration, analgesia duration, blood loss, length of stay (LOS), outcome ratings, complications, and percentage requiring reoperations. Adult implant patients (18.8 %) had higher re-operation rates than adult Nuss or Ravitch patients (5.3 % and 3.3 %, respectively). Adult Nuss patients had longer LOS (7.3 days), more strut/bar displacement (6.1 %), and more epidural analgesia (3 days) than adult Ravitch patients (2.9 days, 0 %, 0 days). Excluding pectus bar and strut displacements, pediatric and adult Nuss patients tended to have higher complication rates (pediatric -- 38 %; adult -- 21 %) compared to pediatric and adult Ravitch patients (12.5 %; 8 %). Pediatric Ravitch patients clearly had more strut displacements than adult Ravitch patients (0 % and 6.4 %, respectively). These results suggested significantly better results in common PE surgical repair techniques (i.e., Nuss and Ravitch) than uncommon techniques (i.e., Implants and Robicsek). The authors concluded that these results suggested
slightly better outcomes in pediatric Nuss procedure patients as compared with all other groups. They recommended that symptomatic pediatric patients with uncomplicated PE receive the Nuss procedure. They suggested that adult patients receive the Nuss or Ravitch procedure, even though the long-term complication rates of the adult Nuss procedure require more investigation.

In a Cochrane review, de Oliveira Carvalho (2014) evaluated the safety and effectiveness of the conventional surgery compared with minimally invasive surgery for treating people with PE. With the aim of increasing the sensitivity of the search strategy, these researchers used only terms related to the individual's condition (pectus excavatum); terms related to the interventions, outcomes and types of studies were not included. They searched the Cochrane Central Register of Controlled Trials (CENTRAL), PubMed, Embase, LILACS, and ICTPR. Additionally they searched yet reference lists of articles and conference proceedings. All searches were done without language restriction. Date of the most recent searches was January 14, 2014. These investigators considered randomized or quasi-randomized controlled trials that compared traditional surgery with minimally invasive surgery for treating PE. Two review authors independently assessed the eligibility of the trials identified and agreed trial eligibility after a consensus meeting. The authors also assessed the risk of bias of the eligible trials. Initially the authors located 4,111 trials from the electronic searches and 2 further trials from other resources. All trials were added into reference management software and the duplicates were excluded, leaving 2,517 studies. The titles and abstracts of these 2,517 studies were independently analyzed by 2 authors and finally 8 trials were selected for full text analysis, after which they were all excluded, as they did not fulfill the inclusion criteria. The authors concluded that there is no evidence from randomized controlled trials to conclude what is the best surgical option to treat people with PE.

CPT Codes / HCPCS Codes / ICD-10 Codes

Information in the [brackets] below has been added for clarification purposes. Codes requiring a 7th character are represented by "+":

<table>
<thead>
<tr>
<th>Code</th>
<th>Code Description</th>
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<tbody>
<tr>
<td>Pectus excavatum:</td>
<td></td>
</tr>
<tr>
<td>CPT codes covered if selection criteria are met:</td>
<td></td>
</tr>
<tr>
<td>Code</td>
<td>Code Description</td>
</tr>
<tr>
<td>--------</td>
<td>----------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>21740</td>
<td>Reconstructive repair of pectus excavatum or carinatum; open</td>
</tr>
<tr>
<td>21742</td>
<td>minimally invasive approach (Nuss procedure), without thoracoscopy</td>
</tr>
<tr>
<td>21743</td>
<td>minimally invasive approach (Nuss procedure), with thoracoscopy</td>
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</tbody>
</table>

Other experimental and investigational interventions:

Dynamic Compression System, Vacuum bell:

No specific code

ICD-10 codes covered if selection criteria are met:

- **J98.4**: Other disorders of lung [Covered for compression of lung as demonstrated by a total lung capacity (TLC) less than or equal to 80% of predictive value per pulmonary function testing]
- **Q67.6**: Pectus excavatum [that causes functional deficit]
- **R94.2**: Abnormal results of pulmonary function studies [covered for exercise pulmonary function tests that are below the predicted values and show restrictive lung disease]

ICD-10 codes not covered for indications listed in the CPB:

- **Q67.7**: Pectus carinatum

Poland's syndrome:

CPT codes covered if selection criteria are met:

- **11960**: Insertion of tissue expander(s) for other than breast, including subsequent expansion
- **11970**: Replacement of tissue expander with permanent prosthesis
- **11971**: Removal of tissue expander(s) without insertion of prosthesis
- **19340**: Immediate insertion of breast prosthesis following mastopexy, mastectomy or in reconstruction
- **19342**: Delayed insertion of breast prosthesis following mastopexy, mastectomy or in reconstruction
- **19357**: Breast reconstruction, immediate or delayed, with tissue expander, including subsequent expansion
- **19361**: Breast reconstruction with latissimus dorsi flap, without prosthetic implant
- **19364**: Breast reconstruction with free flap
- **19366**: Breast reconstruction with other technique
ICD-10 codes covered if selection criteria are met:

<table>
<thead>
<tr>
<th>Code</th>
<th>Code Description</th>
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<tbody>
<tr>
<td>19367</td>
<td>Breast reconstruction with transverse rectus abdominis myocutaneous flap (TRAM), single pedicle, including closure of donor site</td>
</tr>
<tr>
<td>19368</td>
<td>with microvascular anastomosis (supercharging)</td>
</tr>
<tr>
<td>19369</td>
<td>Breast reconstruction with transverse rectus abdominis myocutaneous flap (TRAM), double pedicle, including closure of donor site</td>
</tr>
<tr>
<td>20900</td>
<td>Bone graft, any donor area; minor or small (e.g., dowel or button)</td>
</tr>
<tr>
<td>20902</td>
<td>major or large</td>
</tr>
</tbody>
</table>

The above policy is based on the following references:

Pectus Excavatum


42. Mayer OH. Pectus excavatum: Treatment. UpToDate [online serial]. Waltham, MA: UpToDate; reviewed February 2013.


Poland's Syndrome


**Pectus Carinatum**


Amendment to
Aetna Clinical Policy Bulletin Number: 0272 Pectus Excavatum and Poland's Syndrome Surgical Correction

There are no amendments for Medicaid.