



Medical Coverage Policy

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Surgical Treatment of Chest Wall Deformities

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INSTRUCTIONS FOR USE

The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer’s particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer’s benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer’s benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Each coverage request should be reviewed on its own merits. Medical directors are expected to exercise clinical judgment where appropriate and have discretion in making individual coverage determinations. Where coverage for care or services does not depend on specific circumstances, reimbursement will only be provided if a requested service(s) is submitted in accordance with the relevant criteria outlined in the applicable Coverage Policy, including covered diagnosis and/or procedure code(s). Reimbursement is not allowed for services when billed for conditions or diagnoses that are not covered under this Coverage Policy (see "Coding Information" below). When billing, providers must use the most appropriate codes as of the effective date of the submission. Claims submitted

for services that are not accompanied by covered code(s) under the applicable Coverage Policy will be denied as not covered. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

Overview

This Coverage Policy addresses surgical repair of chest wall deformities in pectus excavatum, pectus carinatum, and Poland syndrome.

Coverage Policy

Coverage for surgical repair of a chest wall deformity varies across plans, may be subject to the provisions of a cosmetic and/or reconstructive surgery benefit and may be governed by state and/or federal mandates. Please refer to the customer's benefit plan document for coverage details.

If coverage for surgical repair of a chest wall deformity is available, the following conditions of coverage apply.

Pectus Excavatum

Surgical repair of pectus excavatum is considered medically necessary when imaging studies (e.g., computerized tomography [CT] scans, radiographs, magnetic resonance imaging [MRI]) confirm a pectus index (i.e., Haller index) greater than 3.25 and EITHER of the following criteria is met:

- pulmonary function studies demonstrate restrictive or obstructive lung disease
- cardiac imaging (e.g., echocardiography, stress echocardiography, MRI) demonstrates findings consistent with external cardiac compression

Pectus Carinatum

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.

Poland Syndrome

Surgical treatment of a chest wall deformity associated with Poland syndrome, including initial breast reconstruction*, is considered medically necessary when EITHER of the following criteria is met:

- preoperative frontal photographs demonstrate partial or complete absence of the pectoralis major muscle(s)
- preoperative imaging demonstrates absence or hypoplasia of upper costal cartilage and/or ribs

When the above criterion is met, coverage of initial breast reconstruction of the affected breast is limited to a one- or two-stage approach (e.g., direct-to-implant or tissue expander followed by permanent implant) to correct breast asymmetry.

Breast reconstruction of the contralateral breast to achieve symmetry in the absence of a severe physical deformity or disfigurement of the contralateral breast accompanied by functional deficit is considered cosmetic in nature and not medically necessary.

Breast reconstruction revision surgery of the affected breast following initial treatment of Poland syndrome is considered medically necessary when there is severe physical deformity or disfigurement accompanied by functional deficit.

Breast reconstruction revision surgery following initial treatment of Poland syndrome that is performed for the sole purpose of improving appearance is considered cosmetic in nature and not medically necessary.

***Note: Please reference the Cigna Medical Coverage policies "Breast Reconstruction Following Mastectomy or Lumpectomy" and "Breast Reduction" for conditions of coverage related specifically to breast reconstruction procedures.**

When performed for indications other than those listed above, surgical repair of a chest wall deformity is considered not medically necessary.

Magnetic Mini Mover Procedure (3MP) for the treatment of pectus excavatum is considered experimental, investigational or unproven.

General Background

The thorax (i.e., chest cavity) is a rigid structure that protects the thoracic organs and supports the upper extremities. Commonly reported chest wall deformities include pectus excavatum (PE), pectus carinatum (PC), and Poland syndrome. In many cases, primarily cosmetic complaints without functional impairment are associated with the abnormality. However, abnormalities of the chest wall can lead to restrictive pulmonary disease, impaired respiratory muscle strength, and decreased ventilatory performance in response to physical stress (Boas, 2020). Cardiac or respiratory impairment may result in functional limitations, such as activity intolerance. Other symptoms may include mild to moderate exercise limitation, respiratory infections, and asthmatic symptoms, as well as decreased stamina and endurance.

Pectus Excavatum

Pectus excavatum (PE), also referred to as a sunken chest or funnel chest, is the most common congenital chest wall deformity. The deformity may be deeper on the right side than on the left side and result in a rotation of the sternum. It is usually diagnosed within the first year of life, with wide variations in the degree of sternal depression. PE is three to four times more prevalent in males, and occurs most frequently in Caucasians (Mourisse and van der Heide, 2022; Gottlieb, et al.; 2018). Although most patients are asymptomatic, 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. Scoliosis, congenital heart disease, and functional heart murmurs can also be associated with PE.

Symptoms of PE 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. Pulmonary effects associated with PE generally include restrictive lung disease, atelectasis, and paradoxical respiration. Restricted lung disease is evident when the total lung capacity (TLC) (forced vital capacity plus residual capacity) is less than 80% of the predicted value for an individual (Mourisse and van der Heide, 2022; Boas, 2020; Rust and Westney; 2016).

Pectus Carinatum

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. The condition is more rare than PE, and is six times more prevalent in males than in females (Gottlieb, et al.; 2018). 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
- 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, Marfan's syndrome, scoliosis, mitral valve disease, and other cardiac changes.

Non-surgical treatment may include orthotic bracing. Bracing may be effective for correction of the chest wall deformity associated with PC, however this type of treatment is often aimed at improving cosmesis. Compression orthotic braces exert pressure on the anteroposterior direction and may be recommended for skeletally immature children (e.g., < age 18 years) with mild to moderate chest deformities. Prolonged use of the orthotic device is often required for total correction, and compliance is an important factor for successful remodeling. As a result candidates must be motivated to wear the brace (Gottlieb, et al.; 2018).

Poland Syndrome

Poland syndrome (i.e., Poland's anomaly, Poland's syndactyly), is a rare congenital disorder characterized by the absence or hypoplasia of the sternocostal head of the pectoralis major muscle, associated with a wide range of other malformations. Co-occurring anomalies include absence or hypoplasia of the pectoralis minor muscle, absence or underdevelopment the upper ribs, absence of costal cartilages, hypoplasia of the breast and subcutaneous tissue, and a variety of hand and upper-extremity anomalies. The right side of the body is affected twice as often as the left, and the condition occurs more frequently in men than in women. 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. 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

The severity of the chest wall abnormality is dependent upon the depth, symmetry and width of the deformity. Chest radiographs are commonly used to determine the degree of chest wall deformity. Cross-sectional imaging such as computed tomography (CT) scans and magnetic resonance imaging (MRI) may be used to evaluate the degree of cardiac compression (ventricular compression), pulmonary compression, and cardiac displacement. Plain anteroposterior and lateral radiographs, CT and MRIs are used to determine the Haller index (a measurement of chest diameter). 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 (Mourisse and van der Heide, 2022). The Correction Index (CI) is another, more recent thoracic index used to assess the severity of deformity in pectus excavatum. The CI corresponds to the depth of the anterior wall depression, reflected as a percentage of the sternal/costal cartilage depression to be corrected with surgery. There is no widely accepted CI cut-off point for surgical repair, though some authors have suggested surgical correction may be appropriate for symptomatic individuals with a CI \geq 28% (Zens, et al., 2022; Rodriguez-Granillo, et al., 2019; Sujka, et al., 2018; Poston, et al., 2014; St. Peter, et al., 2011). The Haller index remains the gold standard for estimating the degree of chest wall deformity in pectus excavatum.

Echocardiography and/or electrocardiography may also be used to evaluate cardiac status. Respiratory status can be determined with the use of pulmonary function studies. In some cases, pulmonary function studies may reveal a restrictive pattern (incomplete lung expansion) and a subsequent decrease in pulmonary volume and reserve. The forced expiratory volume (in one second) (FEV₁), forced vital capacity (FVC), and total lung capacity (TLC) are reduced while the ratio of FEV₁/FVC may be normal or increased in the presence of restrictive airway disease. TLC <80% predicted value signifies restrictive pulmonary disease (Rust and Westney; 2016).

The diagnosis of Poland syndrome is usually obtained by clinical examination. Chest wall abnormalities and determining the presence of latissimus dorsi muscles may require CT scans while chest radiographs may be utilized to evaluate rib formation.

Surgical Treatment

Indications for surgical correction 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.

Pectus Excavatum/Pectus Carinatum: While the optimal age for surgical repair is generally between the ages of 11 and 18 years, and may be performed in adults, each case must be reviewed individually for the presence of cardiopulmonary impairment. Criteria that may be used to demonstrate severe PE and the need for surgical repair include two or more of the following (Mourisse and van der Heide, 2022):

- a Haller CT index greater than 3.25
- pulmonary function studies that indicate restrictive or obstructive airway disease
- a cardiology evaluation, where the chest compression is causing murmurs, mitral valve prolapse, cardiac displacement, or conduction abnormalities on the echocardiogram or electrocardiogram (ECG)
- 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

Surgery for PE may be performed using any one 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, such as the Nuss procedure (i.e., a minimally invasive repair of pectus excavatum [MIRPE]), have been utilized. These procedures involve the insertion of a convex steel bar beneath the sternum through small thoracic incisions and do not require cartilage resection or osteotomy. It has been reported that the Nuss procedure has a shorter operative time and less operative blood loss compared to the Ravitch method (Mao, et al., 2017).

Surgical repair may also be recommended for the treatment of PC. Surgical repair of PC may involve 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

Poland Syndrome: Patients with Poland syndrome typically present for surgical reconstruction to correct deformity or defect of the pectoralis muscles and/or ribs, and to correct breast asymmetry. In cases of rib aplasia, 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. Surgical repair of the chest wall includes the reconstruction of the pectoral muscles and resection of deformed cartilages. This repair typically involves muscle transfers and/or flaps to match normal development of the unaffected side, reconstruction of the axillary line, and correction of infraclavicular flattening. Techniques for repair of large defects include the use of rib grafts, other bony allo- or autografts, mesh, or a combination of these techniques (Gottlieb, et al., 2018; Fokin and Robicsek; 2002).

Surgical treatment of Poland syndrome often consists of reconstruction of the breast and nipple on the affected side by a plastic surgeon, in addition to surgical repair of the chest wall muscles and hypoplastic bone. Surgery may be performed early (approximately age 13) in males, however, in females, reconstructive surgery is often deferred until breast development is complete. If there are rib abnormalities and paradoxical motion, the rib grafts or other chest wall stabilization may occur before breast development is complete. Generally, reconstruction of the breast involves tissue expansion, placement of permanent breast implants and may involve myocutaneous or latissimus dorsi flaps if there is an associated anomaly of the pectoral muscle. Nipple-areolar reconstruction is generally performed at a later stage. Consequently, for patients with Poland syndrome, treatment provided before complete breast development may involve the use of tissue expanders in the affected side which can be inflated periodically to match development of the unaffected breast. Expanders allow for tissue expansion and accommodation of a permanent implant and latissimus muscle upon completion of breast development. Once breast development is complete, the expander is removed, a permanent prosthesis is inserted, and breast reconstruction is performed.

For information on breast prostheses and products used in breast reconstructive procedures, please see Cigna Medical Coverage policy "Breast Reconstruction Following Mastectomy or Lumpectomy".

U.S. Food and Drug Administration (FDA)

Reconstructive repair of chest wall deformities may involve the placement of metal bars, struts, and stabilizers to correct the defect. These appliances are categorized as Class II medical devices by the FDA, and numerous manufacturers have received 510(k) FDA clearance for bone fixation

devices for repair of PE and other anterior chest wall deformities. Examples include Park's Pectus System (TDM Co. Ltd., Houston, TX; 2020) and the Lorenz Pectus Support Bar System (Walter Lorenz Surgical, Inc., Jacksonville, FL; 2006).

Literature Review

Published evidence evaluating surgical repair of chest wall deformities consists of meta-analyses, case series, cross-comparison studies, and prospective trials. The reported clinical outcomes are mixed; differences among outcomes may be related to patient selection criteria, the degree of severity of the deformity, the surgical technique utilized, and growth effects. Many authors evaluated and reported on the methods of surgical repair and improved cosmetic outcomes while some evaluated the impact of PE or PC on cardiopulmonary function. There is little consensus regarding the degree of cardiopulmonary impairment, if any, that is associated with these anomalies. The effects of surgery on exercise tolerance are not clearly established, however, data suggesting improvement in cardiovascular and/or pulmonary function and activity tolerance after surgical repair have been reported (Sonaglioni, et al., 2023; Kelly, et al., 2013; Jaroszewski and Fonkalsrud, 2007; Kubiak, et al., 2007; Lawson, et al., 2005; Bawazir, et al., 2005; Fonkalsrud and Anselmo, 2004; Haller and Loughlin, 2000; Fonkalsrud, et al., 1994). Outcome measures of these studies generally included total lung capacity (TLC), functional residual capacity (FRC), vital capacity,(VC), expiratory flow rate (EFR), and maximum expiratory flow rate (MEFR), exercise tolerance and endurance typically measured prior to surgery, immediately following surgery and three to six months postoperatively. Improvement was generally seen only with increased periods of exercise and not with routine pulmonary function testing at rest. The results of some meta-analyses and other published clinical studies in the medical literature are also mixed, some results tend to support improvement in cardiopulmonary function following surgery (Johnson, et al., 2008; Malek, et al., 2006a, Malek, et al., 2006b) while others do not (Zganjer and Zganjer, 2010; Castellani, et al, 2010; Guntheroth and Spiers, 2007).

Magnetic Mini-Mover Procedure (3MP)

A procedure that is currently under investigation for the treatment of PE is the Magnetic Mini-Mover Procedure (3MP) which utilizes a magnetically coupled implant to pull the sternum forward and remodel the deformed costal cartilage. The internal magnetic implant is surgically placed on the sternum and an external magnet is applied using a custom-fitted chest wall orthotic device. The Magnetic Mini-Mover device was the subject of a United States Food and Drug Administration (FDA)-sponsored Phase III Investigational Device Exemption (IDE) clinical trial. IDE approval allows the device to be used in a clinical study for the collection of safety and effectiveness data. Current evidence evaluating the magnetic mini mover procedure in the published peer-reviewed scientific literature is primarily in the form of pilot studies and small case series (n=15). Reported adverse events have included postoperative pneumothorax, surgical site infection, and device breakage occurred in approximately half of the cases (Graves, et al., 2017; Harrison, et al., 2012; Harrison, et al., 2007).

Professional Societies/Organizations

American Pediatric Surgical Association (APSA): In 2012, the APSA published its guideline on the evaluation and management of pectus carinatum. The guideline recommendations were based upon a systematic review of medical literature and peer-reviewed journals in or after 1973. Recommendations pertaining to surgical intervention included:

- Operative therapy plans should be individualized and age-appropriate, with surgery typically being deferred until pubertal growth is nearly complete.
- Open surgical techniques are acceptable for repair when performed by experienced pediatric surgeons.

- Thoracoscopic and other minimally invasive techniques are acceptable in some children, when performed by pediatric surgeons with advanced skills and experience in the technique.
- Surgery in children ages 5 years and younger is discouraged due to the risk of disruption of normal chest wall growth and chest wall restriction.
- The pediatric surgeon should be involved in extended postoperative follow up.

Medicare Coverage Determinations

| | Contractor | Determination Name/Number | Revision Effective Date |
|-----|------------|---------------------------|-------------------------|
| NCD | National | No Determination found | |
| LCD | | No Determination found | |

Note: Please review the current Medicare Policy for the most up-to-date information.
(NCD = National Coverage Determination; LCD = Local Coverage Determination)

Coding Information

Notes:

1. This list of codes may not be all-inclusive since the American Medical Association (AMA) and Centers for Medicare & Medicaid Services (CMS) code updates may occur more frequently than policy updates.
2. Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Pectus Carinatum/Pectus Excavatum

Considered Medically Necessary when criteria in the applicable policy statements listed above are met:

| CPT®* 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 |

[†]Note: Considered Experimental/Investigational/Unproven when used to report Magnetic Mini Mover (3MP) procedure.

Poland Syndrome

Considered Medically Necessary when used to report surgical repair of a chest deformity associated with Poland syndrome:

| CPT®* Codes | Description |
|-------------|---|
| 11970 | Replacement of tissue expander with permanent implant |
| 15734 | Muscle, myocutaneous, or fasciocutaneous flap; trunk |
| 15756 | Free muscle or myocutaneous flap with microvascular anastomosis |

| CPT®* Codes | Description |
|--------------------|--|
| 15777 | Implantation of biologic implant (eg, acellular dermal matrix) for soft tissue reinforcement (ie, breast, trunk) (List separately in addition to code for primary procedure) |
| 19325 | Breast augmentation with implant |
| 19357 | Tissue expander placement in breast reconstruction, including subsequent expansion(s) |
| 19361 | Breast reconstruction; with latissimus dorsi flap |
| 19380 | Revision of reconstructed breast (eg, significant removal of tissue, re-advancement and/or re-inset of flaps in autologous reconstruction or significant capsular revision combined with soft tissue excision in implant-based reconstruction) |
| 20900 | Bone graft, any donor area; minor or small (eg, dowel or button) |
| 20902 | Bone graft, any donor area; major or large |

***Current Procedural Terminology (CPT®) ©2023 American Medical Association: Chicago, IL.**

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Revision Details

| Type of Revision | Summary of Changes | Date |
|------------------|---|-----------|
| Annual review | <ul style="list-style-type: none"> • No clinical policy statement changes. | 4/15/2024 |

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