



Cigna Medical Coverage Policy

Subject 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. Coverage Policies are intended to provide guidance in interpreting certain **standard** Cigna benefit plans. 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. 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. Proprietary information of Cigna. Copyright ©2014 Cigna

Coverage Policy

Coverage for surgical repair of a chest wall deformity is dependent upon benefit plan language, may be subject to the provisions of a cosmetic and/or reconstructive surgery benefit and may be governed by state and/or federal mandates. Under many benefit plans, surgery for a chest wall deformity is not covered when performed solely for the purpose of improving or altering appearance or self-esteem or to treat psychological symptomatology or psychosocial complaints related to one's appearance.

Under many benefit plans formerly administered by Great-West Healthcare reconstructive services and surgery are covered when the reconstruction services are being performed for one of the following primary purposes: 1) to relieve severe physical pain caused by an abnormal body structure; 2) to treat a functional impairment caused by an abnormal body structure or to restore an individual's normal appearance, regardless of whether a functional impairment exists, when the abnormality results from a documented illness that occurred within the preceding 12 months.

Please refer to the applicable benefit plan language to determine the terms, conditions and limitations of coverage.

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

Cigna covers surgical repair of pectus excavatum as 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.

Cigna covers surgical repair of pectus carinatum as 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.

Cigna covers the surgical repair of a chest deformity associated with Poland syndrome as medically necessary when rib formation is absent.

Not Covered

Cigna does not cover the Magnetic Mini Mover Procedure (3MP) for the treatment of pectus excavatum because it is considered experimental, investigational or unproven.

Cigna does not cover surgical repair of a chest wall deformity performed solely to improve or alter appearance or self-esteem or to treat psychological symptomatology or psychosocial complaints because it is considered cosmetic and not medically necessary.

Under many benefit plans, Cigna does not cover breast reconstruction procedures performed in association with surgical repair of a chest wall deformity for Poland syndrome, pectus excavatum, or pectus carinatum, because each is considered cosmetic in nature and not medically necessary. Such reconstruction procedures include, but are not limited to the following:

- breast reconstruction with latissimus dorsi flap or other technique
- mastopexy
- mammoplasty with or without prosthetic implant
- nipple/areolar reconstruction
- breast reconstruction with tissue expander
- revision of reconstructed breast
- insertion of breast prosthesis
- reconstructive surgery to produce a symmetrical appearance

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 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, 2011). 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

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 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. 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 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 indicative when the total lung capacity (TLC) (forced vital capacity plus residual capacity) is less than 80% of the predicted value for an individual (Rakel, 2011; Johnson and Brunetta, 2005).

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. 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, mitral valve disease, Marfan's syndrome, scoliosis and other cardiac changes.

Poland Syndrome

Poland syndrome (i.e., Poland's anomaly, Poland's syndactyly), is a rare congenital disorder associated with lateral depression of the ribs on one 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). Although the anomaly is associated with a wide range of malformations, 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

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. Plain anteroposterior and lateral radiographs are used to determine the Haller index (a measurement of chest diameter). Cross-sectional imaging such as computerized 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. 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). Another method being investigated to determine the severity of chest wall deformity is the correction index which involves measuring the minimum distance between posterior sternum and anterior spine and the maximum distance between anterior spine most anterior portion of the chest. The difference between the two is then divided by the latter ($\times 100$) to give the percentage of chest depth representing the defect (St. Peter, et al., 2011). The correction index is a newer method proposed to define the pectus deformity; however, in comparison to the Haller Index, the correction index has no clearly established threshold for determining the degree of chest wall deformity.

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) (FEV_1), forced vital capacity (FVC), and total lung capacity (TLC) are reduced while the ratio of FEV_1/FVC may be normal or increased in the presence of restrictive airway disease. $TLC < 80\%$ predicted value signifies restrictive pulmonary disease.

The diagnosis of Poland syndrome is usually obtained by clinical exam. 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 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.

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 impaired cardiopulmonary symptoms. Criteria that may be used to demonstrate severe PE and the need for surgical repair include two or more of the following (Goretsky, et al., 2004):

- 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

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 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. A procedure that is currently under investigation for the treatment of PE is the Magnetic Mini Mover Procedure (3M) 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 is currently under investigation in a United States Food and Drug Administration (FDA) sponsored Phase III Investigational Device Exemption (IDE) clinical trial (NCT01327274); 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 limited to pilot studies (Harrison, et al., 2007, Harrison, et al 2012).

Surgical repair is also often recommended for the treatment of PC. 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

While orthotic bracing may be effective for correction of the chest wall deformity associated with PC, 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 (Goretsky, et al., 2004; Obermeyer, Goretsky, 2012).

Poland Syndrome: Patients with Poland syndrome typically present for surgical reconstruction to improve physical appearance and correct breast asymmetry. Surgical procedures involving the breast and muscles to achieve symmetry are considered cosmetic since there is no significant impairment being corrected. Patients who present with absent ribs are also 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.

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 is 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 and a permanent prosthesis is inserted and breast reconstruction is performed.

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. If necessary, reconstruction of the rib cage may be performed at this time with autologous rib grafts.

Literature Review

Published evidence evaluating surgical repair of chest wall deformities consists of meta-analyses, retrospective reviews, 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 evaluate and report on the methods of surgical repair and improved cosmetic outcome while some evaluate 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 has been reported (Kelly, et al., 2013; Jaroszewski, 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 include 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 is 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).

Professional Societies/Organizations

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.

Use Outside of the US: No relevant information.

Summary

Congenital chest wall deformities may result in functional limitations such as activity intolerance related to cardiac or respiratory impairment. Some patients 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 cosmetic and aimed at improving appearance. Some of the evidence in the published, peer-reviewed scientific literature indicates that surgical repair for PE or PC does improve postoperative cardiopulmonary functioning and exercise tolerance, surgery therefore is considered a viable treatment option for selected candidates with severe deformity and functional impairment. Although surgical repair of Poland Syndrome is frequently cosmetic, with the absence of rib formation there is often functional impairment and surgical repair for this subset of patients may be considered medically necessary.

Coding/Billing Information

Note: 1) This list of codes may not be all-inclusive.

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

Covered when medically necessary:

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

Experimental, Investigational, Unproven and Not Covered when used to report Magnetic Mini Mover (3M) procedure:

CPT [®] * Codes	Description
21743	Reconstructive repair of pectus excavatum or carinatum; minimally invasive approach (Nuss procedure), with thoracoscopy

Poland Syndrome

Covered when medically necessary when used to report surgical repair of a chest deformity associated with Poland syndrome:

CPT [®] * Codes	Description
15734	Muscle, myocutaneous, or fasciocutaneous flap; trunk
15756	Free muscle or myocutaneous flap with microvascular anastomosis
20900	Bone graft, any donor area; minor or small (e.g., dowel or button)
20902	Bone graft, any donor area; major or large

Not Covered

Cosmetic/ Not medically necessary/Not covered when performed in association with surgical repair of chest wall deformity for Poland syndrome, pectus excavatum, or pectus carinatum:

CPT [®] * Codes	Description
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
19316	Mastopexy
19324	Mammoplasty, augmentation; without prosthetic implant
19325	Mammoplasty, augmentation; with prosthetic implant
19340	Immediate insertion of breast prosthesis following mastopexy, mastectomy or in reconstruction
19342	Delayed insertion of breast prosthesis following mastopexy, mastectomy or in reconstruction
19350	Nipple/areola 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
19367	Breast reconstruction with transverse rectus abdominis myocutaneous flap (TRAM), single pedicle, including closure of donor site;
19368	Breast reconstruction with transverse rectus abdominis myocutaneous flap (TRAM), single pedicle, including closure of donor site; with microvascular anastomosis (supercharging)
19369	Breast reconstruction with transverse rectus abdominis myocutaneous flap (TRAM), double pedicle, including closure of donor site
19380	Revision of reconstructed breast

HCPCS Codes	Description
L8600	Implantable breast prosthesis, silicone or equal
S2066	Breast reconstruction with gluteal artery perforator (GAP) flap, including harvesting of the flap, microvascular transfer, closure of donor site and shaping the flap into a breast, unilateral
S2067	Breast reconstruction of a single breast with "stacked" deep inferior epigastric perforator (DIEP) flap(s) and/or gluteal artery perforator (GAP) flap(s), including harvesting of the flap(s), microvascular transfer, closure of donor site(s) and shaping the flap into a breast, unilateral
S2068	Breast reconstruction with deep inferior epigastric perforator (DIEP) flap or superficial inferior epigastric artery (SIEA) flap, including harvesting of the flap, microvascular transfer, closure of donor site and shaping the flap into a breast, unilateral

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