



CIGNA MEDICAL COVERAGE POLICY

The following Coverage Policy applies to all plans administered by CIGNA Companies including plans administered by Great-West Healthcare, which is now a part of CIGNA.

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

Effective Date 3/15/2009
Next Review Date 3/15/2010
Coverage Policy Number 0309

Table of Contents

Coverage Policy	1
General Background	2
Coding/Billing Information	6
References	7
Policy History.....	11

Hyperlink to Related Coverage Policies

Breast Reconstruction Following
Mastectomy or Lumpectomy
Cardiopulmonary Exercise Testing (CPET)

INSTRUCTIONS FOR USE

Coverage Policies are intended to provide guidance in interpreting certain **standard** CIGNA HealthCare benefit plans as well as benefit plans formerly administered by Great-West Healthcare. Please note, the terms of a participant's particular benefit plan document [Group Service Agreement (GSA), 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 participant'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 participant's benefit plan document **always supercedes** 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 group 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. Proprietary information of CIGNA. Copyright ©2009 CIGNA

Coverage Policy

Coverage for surgical repair of chest wall deformities is dependent upon benefit plan language and 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 chest wall deformities 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. Please refer to the applicable benefit plan document to determine benefit availability and the terms, conditions and limitations of coverage.

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

CIGNA covers surgical repair of severe pectus excavatum as medically necessary when imaging studies (e.g., computerized tomography [CT] scans, radiographs) confirm a pectus index (i.e., Haller index) greater than 3.25 and EITHER of the following criteria is met:

- Pulmonary function studies demonstrate at least a moderately severe restrictive lung defect.
- Cardiac imaging (e.g., echocardiography, stress echocardiography, magnetic resonance imaging [MRI]) demonstrates findings consistent with external 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.

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 because they are 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. 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, 2004). In many cases however, cosmetic complaints are associated with chest wall abnormalities. Generally, severe chest wall deformities result in physiologic impairment and associated functional limitations, such as activity intolerance related to cardiac or respiratory impairment. Symptoms resulting from the abnormality may include mild to moderate exercise limitation, respiratory infections, and asthmatic symptoms. Moreover, the deformity may place physiological restrictions on the patient and result in decreased stamina and endurance.

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. 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. 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 (i.e., Poland's anomaly, Poland's syndactyly), a rare congenital disorder, is 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

There is controversy regarding whether there is 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). Cardiac effects associated with PE generally include decreased cardiac output, mitral valve prolapse and arrhythmias; pulmonary effects associated with PE generally include restrictive lung disease, atelectasis, and paradoxical respiration. Patients with PC are usually asymptomatic; however the deformity may be associated with other conditions such as 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. Chest radiographs are commonly used to determine the degree of chest wall deformity. Plain anteroposterior and lateral radiographs may be used to determine the Haller index. In addition, cross-sectional imaging such as computerized tomography (CT) scans and magnetic resonance imaging (MRI) are used to evaluate the degree of cardiac 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). 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.

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; 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: If patients with severe deformities do not undergo surgical repair in childhood, their symptoms will likely 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, 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. Another method of correction currently being investigated involves placing a magnet on the sternum (breastbone) and then applying an external magnetic force that will pull the sternum outward gradually. This method has been referred to as Magnetic Mini-Mover Procedure. Theoretically, this method applies constant outward force on the deformed cartilage with the use of magnetic forces in order to produce biologic reformation of cartilage and correction of the chest wall deformity. A magnet is implanted on the sternum in an outpatient procedure and is pulled outward by way of an external device molded to the patient's anterior chest wall (National Institutes of Health, [NIH], NCT00466206).

Goretsky et al. (2004) reported on their experience with surgical correction of chest wall deformities and identified criteria used to demonstrate severe PE and the need for surgical repair, which requires 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

For correction or improvement of PC, authors recommend bracing to exert pressure on the anteroposterior direction. More specifically, bracing may be utilized 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

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

Several studies have been published in the peer-reviewed scientific literature evaluating surgical repair of chest wall deformities. Many studies evaluate and report on the methods of surgical repair, improved cosmetic outcome, and the impact of PE or PC on cardiopulmonary function. Evidence primarily consists of meta-analyses, retrospective reviews, case series, cross-comparison studies and prospective trials. Data suggesting improvement in cardiovascular and/or pulmonary function and activity tolerance after surgical repair has been reported in some of the studies (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.

Recently, Johnson and colleagues (2008) conducted a meta-analysis to determine the effect surgical correction of pectus excavatum had on pulmonary function, cardiac output and exercise data. The authors analyzed 19 studies and concluded there was substantial evidence to support total lung capacity decreases after the Ravitch procedure; there was evidence supporting a modest increase in forced expiratory volume after bar removal (Nuss procedure); and there was evidence to suggest stroke volume increased after the Ravitch procedure (although it was not conclusive). There was no evidence repair of PE improved exercise capacity. A meta-analysis published by Guntheroth and Spiers (2007) assessed whether or not thoracic surgery for PE significantly improved cardiovascular function and reported that the studies they reviewed were flawed by inadequate methods and failed to show improvement in cardiac function after thoracic surgery for PE. Nonetheless, Malek et al. (August 2006) published the results of a meta-analysis (some of the studies overlapped with the Guntheroth publication) suggesting surgical repair of pectus excavatum significantly improved cardiovascular function. Additionally, this same group of authors (Malek, et al., 2006b) reported the results of a meta-analysis assessing the efficacy of pectus excavatum repair on pulmonary function, using similar methods, and concluded surgical repair does not significantly improve pulmonary function. Overall, the results of the meta-analysis lend some support that surgical correction improves cardiovascular function.

There is no consensus among authors regarding the degree of cardiopulmonary impairment, if any, that is associated with these anomalies. Although the effects of surgery on exercise tolerance are not clearly established—some of the published results are variable and may be considered controversial—authors have reported improvement in cardiopulmonary functioning postoperatively for treatment of PE and PC. Improvement is generally 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. Overall, the reported outcomes may be considered 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.

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.

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. 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 focused on 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 and is therefore considered a viable treatment option for selected candidates with severe deformity and functional impairment.

Coding/Billing Information

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

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

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

Not Medically Necessary/Cosmetic/Not Covered:

CPT ^{®*} Codes	Description
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

	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

[†]Note: Cosmetic in nature and not medically necessary when performed in association with surgical repair of chest wall deformity for Poland syndrome.

ICD-9-CM Diagnosis Codes	Description
	Multiple/Varied

*Current Procedural Terminology (CPT®) © 2008 American Medical Association: Chicago, IL.

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Policy History

<u>Pre-Merger Organizations</u>	<u>Last Review Date</u>	<u>Policy Number</u>	<u>Title</u>
CIGNA HealthCare	3/15/2008	0309	Surgical Treatment for Chest Wall Deformities (Pectus Excavatum/Carinatum and Poland Syndrome)

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