Subject: Surgical Repair of Pectus Carinatum & Pectus Excavatum Chest Wall Deformity		Original Effective Date: 10/12/15
Policy Number: MCP-254	**Revision Date(s):** 12/19/18	
Review Date: 12/16/15, 12/14/16, 6/22/17, 9/18/19		
MCPC Approval Date: 12/19/18, 9/18/19		

DISCLAIMER

This Molina Clinical Policy (MCP) is intended to facilitate the Utilization Management process. It expresses Molina's determination as to whether certain services or supplies are medically necessary, experimental, investigational, or cosmetic for purposes of determining appropriateness of payment. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered (i.e., will be paid for by Molina) for a particular member. The member's benefit plan determines coverage. Each benefit plan defines which services are covered, which are excluded, and which are subject to dollar caps or other limits. Members and their providers will need to consult the member's benefit plan to determine if there are any exclusion(s) or other benefit limitations applicable to this service or supply. If there is a discrepancy between this policy and a member's plan of benefits, the benefits plan will govern. In addition, coverage may be mandated by applicable legal requirements of a State, the Federal government or CMS for Medicare and Medicaid members. CMS's Coverage Database can be found on the CMS website. The coverage directive(s) and criteria from an existing National Coverage Determination (NCD) or Local Coverage Determination (LCD) will supersede the contents of this Molina Clinical Policy (MCP) document and provide the directive for all Medicare members.[1]

DESCRIPTION OF PROCEDURE/SERVICE/PHARMACEUTICAL [29-33]

Pectus deformities are the most frequently seen congenital thoracic wall anomalies. The cause of these conditions is thought to be abnormal elongation of the rib cartilages. Pectus carinatum (PC), or “chicken breast”, “pigeon chest”, “pyramidal chest”, “thorax cuneiform”, or “sternal kyphosis” is a protrusion of the sternum and associated convex deformity of the adjacent costal cartilages of the anterior chest wall. In most patients, pectus carinatum deformity is diagnosed when the defect worsens during the adolescent growth spurt. Cosmetic appearance is the primary concern but symptoms associated with pectus carinatum may include exertional dyspnea, exercise limitation, frequent respiratory infections, and asthma. Pectus excavatum (PE), or "funnel chest", “cobbler’s chest”, “sunken chest”, “and hallowed breast” is a deformity of the chest wall characterized by a sternal depression typically beginning over the midportion of the manubrium and progressing inward through the xiphoid process. A small number of patients are born with pectus excavatum but most are diagnosed when the defect worsens during the adolescent growth spurt. Symptoms include exertional intolerance, dyspnea, chest pain, and poor endurance. PE is frequently associated with scoliosis, Marfan syndrome, and congenital heart disease.

A mild PE deformity may be treated with posture and exercise, while mild degrees of PC may be treated with bracing or casting. One of the standard surgical treatments for PE or PC is using the open Ravtich approach, placing a sternal bar behind the sternum after mobilizing the deformed cartilages around the sternum; a second operation follows to remove the bar approximately 12 months after the initial repair. A minimally invasive approach called the Nuss Procedure involves the placement of a large curved bar through small incisions on the chest wall; the bar is rotated into position and kept in place for several months post-operatively. The severity of the chest wall deformity is determined by the pectus severity index (PSI), also known as the Haller index which

is the width of the chest divided by the distance between the sternum and spine. The normal value is 2.54. Surgical repair is usually recommended for patients who are symptomatic, have a functional impairment, and who demonstrate a pectus severity index greater than 3.25 for pectus excavatum and less than 2 for pectus carinatum calculated from the chest measurements of a CT scan of the area of the chest with the greatest depression.

RECOMMENDATION [2-20 29-35]

Molina State Health Plans may limit coverage for cosmetic procedures and may provide coverage for reconstructive procedures. Please review all applicable State Health Plan benefits and definitions of cosmetic and reconstructive surgery first before applying the below criteria.

- ❑ Surgical treatment of pectus excavatum or pectus carinatum, may be considered medically necessary and may be authorized when all of the following criteria are met:
  - Age > 5 years; [7 18 23] AND
  - Failure of conservative treatment that includes orthotic bracing if applicable; [23] AND
  - Documented functional impairment: [23 25-27] [ALL]
    - ➢ Abnormal cardiac stress test and/or pulmonary function during exercise; and
    - ➢ Clinical signs or symptoms that impair activities of daily living (i.e. shortness of breath, chest or pectoral rib pain, asthma, or frequent infections of the lower respiratory tract); and
    - ➢ Clinical signs of cardiopulmonary impairment that include: [ONE]
      - ❖ arrhythmias; or
      - ❖ mitral valve prolapse; or
      - ❖ signs of cardiac or pulmonary compression; AND
  - The surgical procedure is expected to correct the functional impairment; AND
  - Imaging documents a clinical chest wall deformity: [12] [ONE]
    - ➢ Pectus Excavatum: Haller Index is $\geq$ 3.2, OR
    - ➢ Pectus Carinatum: Haller Index is $\leq$ 2.0, OR
    - ➢ Poland syndrome: when rib formation is absent

- ❑ If NO functional impairment is present, surgical treatment of pectus carinatum and pectus excavatum is considered cosmetic and not medically necessary.

EXCLUSIONS [21-28]

The following non-surgical treatments are considered experimental and investigational because their clinical effectiveness has not been proven in peer reviewed literature:

- ❑ Magnetic mini-mover procedure (3Mp): sternal magnet
- ❑ Vacuum bell: sternal suction
- ❑ Dynamic Compression System: brace compression

SUMMARY OF MEDICAL EVIDENCE [2-20]

There is a large body of evidence in the published peer-reviewed literature that supports the open or minimally invasive surgical approach as a treatment for pectus excavatum. There is a smaller body of evidence supporting surgery as a treatment option for pectus carnivatum. Surgical repair is usually recommended for patients who are symptomatic, have a functional impairment, and who demonstrate a pectus severity index greater than 3.25 for pectus excavatum and less than 2 for pectus carinatum.[12] Studies have reported that operative repair of pectus chest wall deformities are to be discouraged in children ages 5 years and younger due to the risk of disruption of normal chest wall growth with resultant chest wall restriction. [7 18] Study sizes included 60-1200 participants and follow-up was up to 12 years. Studies included systematic reviews, meta-analysis and prospective and retrospective reviews that showed chest wall deformities can be repaired with a low rate of complications, a short hospital stay, and excellent long-term physiologic and cosmetic results. [2-20]

One of the largest single institution prospective reviews reported the results of 21 years with minimally invasive repair of pectus excavatum by the Nuss procedure in 1215 patients. The mean Haller CT index was 5.15 ± 2.32 (mean ± SD). Pulmonary function studies performed in 739 patients showed that FVC, FEV1, and FEF25-75 values were decreased by a mean of 15% below predicted value. Mitral valve prolapse was present in 18% (216) of 1215 patients and arrhythmias in 16% (194). Of patients who underwent surgery, 2.8% (35 patients) had genetically confirmed Marfan syndrome and an additional 17.8% (232 patients) had physical features suggestive of Marfan syndrome. Scoliosis was noted in 28% (340). At primary operation, 1 bar was placed in 69% (775 patients), 2 bars in 30% (338), and 3 bars in 0.4% (4). Complications decreased markedly over 21 years. In primary operation patients, the bar displacement rate requiring surgical repositioning decreased from 12% in the first decade to 1% in the second decade. Allergy to nickel was identified in 2.8% (35 patients) of whom 22 identified preoperatively received a titanium bar, 10 patients were treated successfully with prednisone and 3 required bar removal: 2 were switched to a titanium bar, and 1 required no further treatment. Wound infection occurred in 1.4% (17 patients), of whom 4 required surgical drainage (0.4% of the total). Hemothorax occurred in 0.6% (8 patients); 4 during the postoperative period and four occurred late. Postoperative pulmonary function testing has shown significant improvement. A good or excellent anatomic surgical outcome was achieved in 95.8% of patients at the time of bar removal. A fair result occurred in 1.4%, poor in 0.8%, and recurrence of sufficient severity to require reoperation occurred in 11 primary surgical patients (1.4%). Five patients (0.6%) had their bars removed elsewhere. In the 752 patients, more than 1 year post bar removal, the mean time from initial operation to last follow up was 1341 ± 28 days (SEM), and time from bar removal to last follow-up is 854 ± 51 days. Age at operation has shifted from a median age of 6 years (range 1-15) in the original report to 14 years (range 1-31). The minimally invasive procedure has been successfully performed in 253 adult patients aged 18 to 31 years of age. 95.8% good to excellent anatomic result in were achieved. [9]

There is insufficient published evidence to support the efficacy and safety of the following proposed minimally invasive treatments for chest wall deformities: The magnetic mini-mover procedure, the vacuum bell and the Dynamic Compression System. There are no randomized controlled trials on these treatments for chest wall deformities. Small prospective and retrospective reviews have reported improvement but long term results are needed to evaluate safety and effectiveness. [21-28]

CODING INFORMATION THE CODES LISTED IN THIS POLICY ARE FOR REFERENCE PURPOSES ONLY. LISTING OF A SERVICE OR DEVICE CODE IN THIS POLICY DOES NOT IMPLY THAT THE SERVICE DESCRIBED BY THIS CODE IS COVERED OR NON-COVERED. COVERAGE IS DETERMINED BY THE BENEFIT DOCUMENT. THIS LIST OF CODES MAY NOT BE ALL INCLUSIVE.

CPT	Description
21740	Reconstructive repair of pectus excavatum or carinatum; open
21742	Reconstructive repair of pectus excavatum or carinatum; minimally invasive approach (Nuss procedure), without thoracoscopy
21743	Reconstructive repair of pectus excavatum or carinatum; minimally invasive approach (Nuss procedure), with thoracoscopy

HCPCS	Description
	N/A

ICD-10	Description: [For dates of service on or after 10/01/2015]
Q67.6	Pectus excavatum
Q67.7	Pectus Carinatum
Q79.8	Other congenital malformations of musculoskeletal system , Poland Syndrome

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Review/Revision History:

10/12/15: New Policy
12/16/15, 12/14/16 & 6/22/17: Policy reviewed, no changes
12/19/18: Policy reviewed, added Poland syndrome to clinical criteria. Added the following exclusions for non-surgical treatments that are considered investigational: Magnetic mini-mover procedure, vacuum bell and the dynamic compression system. Updated summary of medical evidence, coding and references.
9/18/19: policy reviewed, no changes