

#### UnitedHealthcare® West Medical Management Guideline

# **Pectus Deformity Repair**

**Guideline Number**: MMG101.R **Effective Date**: June 1, 2024

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# Related Medical Management Guideline • Cosmetic and Reconstructive Procedures Related Benefit Interpretation Policy

Cosmetic, Reconstructive, or Plastic Surgery

## **Coverage Rationale**

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Surgical repair of Pectus Excavatum is considered reconstructive and medically necessary when the following criteria has been met:

- Imaging studies confirm <u>Haller Index (HI)</u> > 3.25 or <u>Correction Index (CI)</u> ≥ 28%; and
- A Functional Impairment defined in physician office notes; and
  - For restrictive lung capacity the total lung capacity is documented in the physician office notes as < 80% of the predicted value; or
  - o There is cardiac compromise as demonstrated by decreased cardiac output on the echocardiogram; or
  - There is objective evidence of exercise intolerance as documented by cardiopulmonary exercise testing that is < 80% of the predicted value</li>

Surgical repair of Pectus Carinatum may be considered reconstructive and medically necessary in severe symptomatic disease that has failed first line treatment with corrective bracing or corrective bracing is not indicated for the member. Requests for coverage of repair of Pectus Carinatum will be reviewed by a UnitedHealthcare Medical Director on a case-by-case basis.

#### **Documentation Requirements**

Benefit coverage for health services is determined by the member specific benefit plan document and applicable laws that may require coverage for a specific service. The documentation requirements outlined below are used to assess whether the member meets the clinical criteria for coverage but do not guarantee coverage of the service requested.

#### **Required Clinical Information**

#### **Pectus Deformity Repair**

Medical notes documenting the following, when applicable:

- Diagnosis
- History of the medical condition(s) requiring treatment or surgical intervention
- Documentation of functional limitation/impairment
- Results of all recent imaging studies and applicable diagnostics, including results of:
  - CT scan including Haller Index or Correction Index calculation
  - Pulmonary function test total lung capacity

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#### **Required Clinical Information**

#### **Pectus Deformity Repair**

- Echocardiogram including ejection fraction
- o Stress test including cardiopulmonary function values
- Treatments tried, failed, or contraindicated; include the dates and reason for discontinuation
- Physician treatment plan

#### Definitions

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The following definitions may not apply to all plans. Refer to the member specific benefit plan document for applicable definitions.

**Correction Index (CI)**: This index corresponds to the depth of the anterior wall depression, reflected as a percentage of the sternal/costal cartilage depression that should be corrected with surgery. For its calculation, a horizontal line should be extrapolated through the anterior border of the spine, and two measurements should be obtained: the APmin, and the largest inner anterior-posterior distance between such horizontal line and the most anterior portion of the chest wall (APmax). Thus, the CI is calculated using the following formula: [(APmax-APmin) / APmax] x 100. (Rodríguez-Granillo et al, 2019)

**Functional or Physical or Physiological Impairment**: A Functional or Physical or Physiological Impairment causes deviation from the normal function of a tissue or organ. This results in a significantly limited, impaired, or delayed capacity to move, coordinate actions, or perform physical activities and is exhibited by difficulties in one or more of the following areas: physical and motor tasks; independent movement; performing basic life functions. (Medicare, 2023)

Haller Index (HI): The widest transverse diameter of the internal chest divided by the distance between the anterior spine and posterior sternum. (Sujka, 2018)

**Pectus Carinatum**: A protrusion of the sternum and costal cartilages caused by an abnormal growth of these structures. (Kelly & Martinez-Ferro, 2020)

Pectus Excavatum: Posterior depression of the sternum and adjacent costal cartilages. (Jaroszewski et al., 2010)

#### **Applicable Codes**

The following list(s) of procedure and/or diagnosis codes is provided for reference purposes only and may not be all inclusive. Listing of a code in this guideline does not imply that the service described by the code is a covered or non-covered health service. Benefit coverage for health services is determined by the member specific benefit plan document and applicable laws that may require coverage for a specific service. The inclusion of a code does not imply any right to reimbursement or guarantee claim payment. Other Policies and Guidelines may apply.

CPT Code	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

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#### **Description of Services**

Pectus deformities are anomalies of the thoracic cage caused by excessive growth of the cartilage in the chest.

Pectus Excavatum, also known as funnel chest or sunken chest, is characterized by a depression in the anterior chest wall. Cardiac and pulmonary compression can occur depending on the size of the deformity. Although the deformity may be identified in infancy, it can progress with growth. Surgical repair of Pectus Excavatum is usually reserved for severe deformities. Surgical repair typically occurs during adolescence while the chest is still soft and malleable.

Pectus Carinatum is characterized by a protrusion of the sternal and costal cartilages. Although cardiopulmonary disorders are rare with Pectus Carinatum, symptoms can include chest wall pain, back pain, scoliosis, kyphosis, dyspnea, and reduced endurance. Orthotic bracing is the first line treatment for thoracic reshaping, often yielding better results than surgical correction. (Kelly & Martinez-Ferro, 2020)

#### **Benefit Considerations**

Some states require benefit coverage for services that UnitedHealthcare considers cosmetic services and surgery, such as repair of external congenital defects in the absence of a Functional or Physical Impairment. Refer to the member specific benefit plan document.

- Cosmetic services and surgery are excluded from coverage. Procedures that correct an anatomical congenital defect without improving or restoring physiologic function are considered cosmetic services and surgery. Refer to the <u>Definitions</u> section.
- Procedures that do not meet the reconstructive criteria in the <u>Coverage Rationale</u> section are excluded from coverage.

#### **Clinical Evidence**

#### **Pectus Excavatum**

In a retrospective cohort study, Infante et al. (2023) assessed quality of life and adverse events in patients in whom bar duration was extended beyond 3 years, to ascertain whether this approach is safe and has any benefits. Two hundred and thirty-one patients were included in this study. The study focused on patients who underwent primary Nuss repair from 2007 to 2018 and had a follow-up of at least 24 months. Pectus bars had been left in place beyond 3 years in patients concerned over possible recurrence after bar removal. Structured interviews were held to assess pain, chest tightness, or other discomfort, and any adverse events related to pectus bars. Results were compared between patients in whom pectus bars were removed after 3 years (standard group) and those in whom bars were left in place longer (extended bar duration group). Bar duration was  $30.6 \pm 6.6$  mo in the standard group (51 patients) versus  $69.1 \pm 26.3$  mo in the extended group (180 patients). Some discomfort was reported by 81.6% in the standard group versus 62.9% in the standard versus 30.3% in the extended group. Quality of life improved in 92.6% of the standard group versus 94.7% of the extended group. No significant adverse events were reported in either group. In conclusion the authors suggest that an extended bar duration after the Nuss repair may not cause any adverse event nor negatively affect quality of life. The study had limitations which included its observational design and retrospective data collection.

Walsh et al. (2023) conducted a systematic review to assess the physiological and psychological outcomes on patients who had pectus excavatum (PEx) surgery. Fifty-one articles (34 physiological outcomes and 17 psychological outcomes). Physiological outcomes were divided into cardiopulmonary function changes [e.g., cardiopulmonary exercise testing (CPET), pulmonary function testing (PFTs), transthoracic echocardiography (TTE)]. Psychological outcomes investigated change in QoL scores, pre and postsurgical intervention and patient satisfaction. Twenty-one studies investigated pulmonary function at rest. There was no change in forced vital capacity or forced expiratory volume in 1 second following open repair and transient reductions followed closed repair. In the 11 studies investigating echocardiography, transthoracic rarely demonstrated cardiac compression; however, transoesophageal demonstrated intraoperative relief in cardiac compression in severe cases. Sixteen studies investigated exercise testing (cardiopulmonary exercise testing, CPET), 12 of which demonstrated significant improvement following surgery, both in maximal oxygen consumption and oxygen pulse. Seventeen studies investigated quality of life, all but one of which showed improvement following repair of PEx. All papers that reported on patient satisfaction following surgery found high rates, between 80% and 97%. The authors concluded that for many patients with PEx, there exists a cardiopulmonary limitation that while difficult to objectify, is likely to improve with surgical repair. Resting parameters offer little yield in aiding this except in the most severe cases. CPET therefore offers a better option for dynamic assessment of this limitation and improvements following repair. Surgery significantly improves psychological well-being and quality of life for patients with PEx.

In a cross-sectional multicenter study, Norlander et al. (2022) evaluated the health-related quality of life (HRQoL) in patients who have underwent the Nuss procedure for pectus excavatum (PE). The study identified patients (n = 420) who underwent the Nuss procedure in Sweden between 2000 and 2019. They were invited to answer the RAND-36 (generic HRQoL) and Nuss Questionnaire modified for Adults (NQ-mA), a disease-specific HRQoL. The results included a total of 236 patients with a response rate of 56.2%. Men scored significantly better on the modified Nuss Questionnaire total (p = 0.01) and psychosocial (p = 0.02) subscales. Patients younger than 20 years of age had significantly better scores on the same scales (p = 0.007 and 0.006, respectively) compared to patients aged 20–30 years at the time of surgery. Patients greater than 30 years of age had no significant difference in comparison. Patients who had their bar removed had

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significantly better values on both scales. The researchers concluded that the generic and disease specific HRQoL differs in different subgroups. Male gender, young age and bar removal seem to be associated with better HRQoL. Further studies are warranted to observe longitudinal data and confirm the study findings.

In a retrospective cohort study, Sollie et al. (2022) reported outcomes after performing Ravitch type repairs using a permanent titanium plate with screws to pectus deformities. A retrospective review of 61 PE and pectus carinatum (PC) cases from August 2013 to April 2021 was performed. Data were extracted from medical records and statistical analysis was reported. Fifty-four patients underwent PE repair, 6 underwent PC repair, and 1 underwent mixed repair. Median Haller index was 3.8. The postoperative bleeding was 30% in the first 10 patients and caused a change in protocol. Protocol changes including postponing chemical deep vein thrombosis prophylaxis, using intraoperative hemostatic agents, and using shorter implantation screws decreased this to 0% for the remaining cases. The most frequent complication was postoperative urinary retention in 13 cases (21.3%). Thirty- seven postoperative surveys were completed out of 50 patients. Seventy-five percent reported health improved, 65% reported exercise capacity improved, 75% reported breathing improved, and 59% reported chest pain improved. Of those that reported, 90% were satisfied with the overall outcome and 86% would have the operation again. The authors concluded Ravitch type repair with permanent titanium plate fixation is a safe and effective procedure for correction of PE and carinatum. Based on the postoperative survey results, the short terms findings were favorable, and patients were satisfied.

In a retrospective cohort study, Brungardt et al. (2021) studied the demographics and outcomes of adult patients who underwent surgical repair of PE via open and minimally invasive thoracoscopic methods. A retrospective analysis of the American College of Surgeons National Surgical Quality Improvement Program (ACS NSQIP) database from 2015 to 2018 was performed, capturing patients 18 years or older with PE as the postoperative diagnosis. Patients were placed into two groups of minimally invasive (Nuss) and open (Ravitch) repair procedure code. Baseline characteristics and postoperative outcomes were analyzed. A total of 168 adult patients were captured. Most of these patients were white (84.52%) male (69.64%) and 26 years old on average. Median operative time was longer in the open repair group [250 (IQR, 173-308) versus 122 (IQR, 94-160) minutes, p < 0.0001]. Median length of stay was five days (IQR, 4-6) in the open group and three days (IQR, 2-4) in the minimally invasive group (p = 0.2873). The authors concluded that complications after repair of PE occur at similar rates between open and minimally invasive repair. They further noted minimally invasive repair decreases operative time and may decrease length of stay, the decision of type of procedure depends upon clinical scenario and factors unique to the individual patient. The authors indicated Nuss repair, while the preferred method of repair in children, may be a less attractive option in adults undergoing repair of PE due to decreased pliability of the chest wall. The study was limited by database limitations, including lack of long term follow up and lack of complex case information.

In a retrospective cohort study, de Loos et al. (2021) examined the risk of complications after the Nuss procedure in adult patients compared with young patients with PE. This single-center retrospective cohort study evaluated all patients who underwent the Nuss procedure between 2006 and 2018. Patients were stratified by age as young ( $\leq$  24 years old) and adult (> 24 years old). The primary end point was the occurrence of perioperative or postoperative complications, subdivided into major (Clavien-Dindo class III or higher) and minor (less severe than Clavien-Dindo class III). Between-group differences were analyzed using the Mann-Whitney U and the  $\chi^2$  test with post hoc analysis. A total of 327 participants were included, 272 in the young group [median age, 16 years; interquartile range (IQR), 15 to 18 years; range, 11 to 24 years] and 55 in the adult group (median age, 32 years; IQR, 27 to 38 years; range, 25 to 47 years). The median HI was similar between groups (young, 3.7; IQR, 3.2 to 4.4 vs adult, 3.6; IQR, 3.0 to 4.3; p = .44). The median follow-up was 34 and 36 months, respectively. The incidence of major complications was comparable between young and adult participants (p = .43). Minor complications occurred more often among adults (young, 4% vs adult, 11%; p = .002). Chronic postoperative pain was the only minor complication with a significant difference in incidence (young, 1% vs adult, 7%; p = .008). The authors concluded the Nuss procedure is a safe surgical treatment for PE in both young and adult patients. They indicated the risk of major complications is comparable, however, adults more often have chronic pain. The study is limited by the difference in study group size.

In a retrospective case series, Ramadan et al. (2021) studied cardiopulmonary function (CPF) impairment, especially in PE and PC patients. The study goal was to determine any correlation between pectus malformations and cardiopulmonary symptoms and function based on systematic assessment of CPF and thoracic measurements, such as HI and sternal torsion angle (STA). Data from 76 adolescent patients with PE (n = 30) or PC (n = 46) were retrospectively collected referred between January 2015 and April 2018. CPF measurements and thoracic imaging were performed in all patients. HI and STA correction indexes were measured in all patients. Medical records from 76 patients (PE n = 30; PC n = 46) were analyzed. Patients were predominantly male (> 93.3%), and aged between 13 and 14½ old. PE was associated with airway obstruction, with a forced expiratory volume in 1 s value under the lower limit of normal in 13% of cases (p < 0.001). Restrictive syndrome was observed in 23% of cases (p < 0.001), with a Z score for total lung capacity under the lower limit of normal. In PC, pulmonary function was not affected. All patients showed slightly decreased values

of left and right ejection fraction and cardiac index at rest, although values were within normal range. There were no significant correlations between pulmonary and cardiac functions or between low CPF and thoracic measurements. The authors concluded the study results confirmed the modest impact of pectus malformations on CPF at rest, without correlation with anamnestic dyspnea on exertion, nor with chest pain or anatomical measurements (such as HI or STA). The authors noted that validation of new correction indexes could be helping characterize these malformations and choose optimal therapeutic management. The study was limited by a small number of patients, a lack of control group, and evaluation being performed at rest.

Sakamoto et al. (2021) analyzed changes in lung capacity and thoracic morphology based on computed tomography (CT) imaging in adults with PE before surgery, during bar insertion and after bar removal in a retrospective cohort study. Patients who underwent the Nuss procedure for PE after the age of 20 were included in this study. Chest CT scans of the included participants were taken before the Nuss procedure, immediately before removal of the pectus bar and 6 months after removal of the pectus bar. Lung capacity and thoracic morphology measurements were made from the CT scans. Six patients aged 24–43 years were included in this study. After the Nuss procedure, lung capacity was decreased in all patients. Although the pectus bar was removed, lung capacity had not significantly increased and was almost the same volume as before the Nuss procedure. After the Nuss procedure, the funnel chest shape had improved in all cases, patients' thoracic spine had also moved forward as the thorax moved forward and patients' stoop had improved. The authors concluded that while functional recovery is poor, the improvements in cosmetics such as decreased sternal depression and stoop are the main focus in the adult PE. The authors also concluded as changes in the morphology of the thoracic spine are present and have led to severe pain, surgical modification that reduces the stress of the thoracic spine should be considered. Sakamoto et al. (2021) noted further long-term observation seems necessary to determine the long-term effects of the Nuss procedure. This study was limited by a small sample size, no long term follow up, and lack of a comparison group.

In a prospective cohort study, Satur et al. (2021) explored categorization of exercise dysfunction in patients with PE. Cardiopulmonary exercise test data were delineated by maximal oxygen uptake values > 80%, which was tested to examine whether patterns of exercise physiology were distinguished. Seventy-two patients considered for surgical treatment underwent assessment of pulmonary function and exercise physiology with pulmonary function tests and cardiopulmonary exercise test between 2006 and 2019. Seventy who achieved a threshold respiratory gas exchange ratio of > 1.1 were delineated by maximal oxygen uptake > 80%, (group A, n = 33) and < 80% (group B, n = 37) and comparison of constituent physiological parameters performed. The cohort was 20.8 (±SD 6.6) years of age, 60 men, with a Haller's Index of 4.1 (±SD 1.4). Groups A and B exhibited similar demography, pulmonary function test results and Haller's index values. Exercise test parameters of group B were lower than group A; work 79.2% (±SD 11.3) versus 97.7 (±SD 10.1), anaerobic threshold 38.1% (±SD 7.8) versus 49.7% (±SD 9.1) and O2 pulse 77.4% (±SD 9.8) versus 101.8% ( $\pm$ SD 11.7), but breathing reserve was higher, 54.9% ( $\pm$ SD 13.1) versus 44.2% ( $\pm$ SD 10.8), p < 0.001 for each. Both groups exhibited similar incidences of carbon dioxide retention at peak exercise. A total of 65 (93%) exhibited abnormal values of at least one of four exercise test measures. The authors concluded that patients with PE exhibited multiple physiological characteristics of compromised exercise function. They noted this is the first study that defines differing patterns of exercise dysfunction and provides evidence that patients with symptomatic PE should be considered for surgical treatment.

Rodríguez-Granillo et al. (2019) provided expert opinion to attempt to establish recommendations about preoperative imaging in patient with PE. The authors noted chest CT, stress echocardiography (Echo), and cardiac magnetic resonance (CMR) allow the evaluation of specific information regarding structural and functional characteristics of vital importance to assess surgical candidacy and define surgical strategies. The authors provided recommendations for preoperative image acquisition and analysis, aimed at the assessment of the severity of the chest wall deformity (CT); the site of maximum cardiac compression, extent of increased interventricular dependence, and presence of pericardial effusion (CMR); and the effect of PE on the functional capacity and exercise-related systolic and/or diastolic function, and tricuspid annulus compression (Echo). The authors proposed that, the surgical decision in patients with PE is based upon the presence of at least two of the following criteria: 1) HI larger than 3.25, and/or a CI larger than 20%; 2) exercise-related symptoms; 3) pulmonary manifestations (atelectasis, bronchiectasis, repetitive pneumonia); 4) significant cardiac compression (type 1 or 2) and one or more of the following: systolic dysfunction, inspiratory septal flattening, moderate pericardial effusion, signs of diastolic dysfunction at stress, or elevated transtricuspid gradient at stress (mean gradient ≥ 6 mmHg)].

In a critical review of the literature, Obermeyer et al. (2018) evaluated contemporary literature to provide an understanding of the physiologic impact of repairing PE on pediatric and adult patients separately, noting the adverse physiologic effects of PE and subsequent resolution following correction have been a subject of controversy. Obermeyer et al. (2018) reviewed literature surrounding resting pulmonary function testing, resting cardiac function testing, and cardiopulmonary exercise testing in pediatric and adult patients. The authors concluded pediatric and adult patients experience subjective

clinical improvement in exercise tolerance after PE repair in the majority of cases. They indicated the benefits are likely multifactorial as suggested by studies demonstrating improved respiratory mechanics and increased stroke volume due to relief of right ventricular compression. The authors noted the culmination of these physiologic effects is difficult to assess objectively, but cardiopulmonary exercise testing (CPET) currently represents the best non-invasive method to evaluate exercise capacity. The authors indicated available CPET studies are limited but are beginning to demonstrate a physiologic explanation for perceived improved exercise tolerance after MIRPE. The authors recommended future studies should focus on CPET with consistent methodologies using control groups to provide a more objective evaluation of the physiologic effects after PE repair.

Sujka et al. (2018) provided commentary on the creation, calculation, and limitations of the methods guantifying pectus defects. Quantification of defect severity can be performed with multiple imaging modalities or external thoracic measures, but is most commonly quantified by the HI or Pectus Correction Index (PCI). These two measures provide a measure of the chest based on cross sectional imaging, most commonly CT scans, allowing for standard comparison and definitions of pectus defects. HI has been the clinical standard for the past few decades. This metric was defined as the widest transverse diameter of the internal chest divided by the distance between the anterior spine and posterior sternum. A width to depth ratio of 3.25 has been set as the discriminator to define patients with a significant enough PE defect to be a potential candidate for repair. The decision to operate for PE is also based on physical limitations secondary to the defect and psychosocial disposition, but it has been a generally held belief that surgical candidacy revolves around an HI greater than or equal to 3.25. The authors concluded that based on recent literature, a HI of 3.25 as a cut-off point for surgical intervention is no longer a good discriminator and bares no conclusive relationship with the aesthetic complaints observed. They further concluded that the limitation of the HI include variation with thoracic shape, age, gender. breathing, lack of consideration for asymmetry, and lack of consideration for cardiac compression. The PCI was developed in an effort to remove width from the calculation which has little relation to depth of the defect. The PCI measures the distance between the posterior sternum and the anterior spine and the inner margin of the most anterior portion of the chest. The difference between the two lines is the amount of defect the patient has in their chest. To generate the percentage of chest depth the patient is missing centrally the difference between the measurements is then divided by the maximum prominence of the chest (the longer measurement) and multiplied by 100. The PCI provides a number which represents the percentage of chest depth to be corrected by bar placement and regained by the patient. The authors note that any given PCI does not demand repair, only that a PCI of 10% is noticeable enough to reach diagnosis. The authors indicated PCI and HI had a strong correlation and the PCI that was equivalent to an HI of 3.25 was 28%, however, any specific anatomic threshold is probably inadequate as a stand-alone trigger for repair. The authors concluded HI was the initial measurement to quantify PE defects, but because of its dependence on width, the PCI is more descriptive and accurate in describing these defects. They also concluded that while other imaging studies have been suggested to obtain cross sectional measurements, CT is the standard method due to its accuracy, availability, and simplicity. Other imaging methods such as 3D scanners and external thoracic measurements using calipers are areas of ongoing investigation.

#### **Pectus Carinatum**

In a study conducted by de Beer et al. (2023), 740 patients were treated with the Dynamic Compression System (DCS), between 2013 and 2020. At time of analysis 203 patients were still being treated. The remaining patients either finished treatment successfully (n = 406), stopped treatment prematurely (n = 74) because of a lack of motivation, insufficient results, or were lost to follow up (n = 57). The lost to follow up group was excluded from analysis. The study included the effect of age, gender, pectus height, symmetry and pectus rigidity on treatment time and symptoms with linear multiple regression analyses. Carinatum height and high pressure of initial correction at the start of treatment duration increased with 1.9 months. An initial correction pressure of  $\geq$  7.6 pounds per square inch (psi), increased the treatment duration with 3.5 months compared to an initial correction pressure of  $\leq$  5.0 psi. A high initial pressure of correction of  $\geq$  7.6 psi increased the odds of having somatic symptoms with 1.19) and psychosocial symptoms with 1.13 compared to a low initial pressure of correction of  $\leq$  5.0 psi. An initial pressure of correction of 5.1–7.5 psi increased the odds of having somatic symptoms with 1.19) compared to an initial pressure of correction of  $\leq$  5.0 psi. An initial pressure of correction of  $\leq$  5.0 psi. Patients with asymmetric chests were more likely to abandon therapy. The authors concluded high carinatum height and high initial pressure of correction of  $\leq$  5.0 psi. Patients with prolonged bracing treatment and a higher failure rate.

In a retrospective cohort study (n = 738), van Braak et al. (2022) evaluated the results of both the surgical procedures (Ravitch or Abramson) and dynamic compression bracing (DCS) bracing for patients with pectus carinatum (PC). A total of 631 patients started with DCS bracing, 105 patients for Ravitch surgery and 2 patients for Abramson procedure. Of the 631 patients who underwent DCS bracing treatment, 553 finished treatment, and 78 patients are still under treatment. A total of 73.8% (n = 408) of these patients successfully completed treatment, 13.6% (n = 75) experienced treatment failure, and 12.7% (n = 70) were lost to follow-up. Ravitch surgery was performed in 105 patients, with a success rate of 92.4% finishing treatment. Complications of Ravitch surgery occurred in 32.4% of patients. No statistical relevant relations were

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found between osteotomy or sternal fixation and outcomes or complications. The Abramson procedure was successfully performed in 2 patients. The researchers concluded that DCS bracing should be the first treatment of choice in patients with PC. DCS bracing is noninvasiveness, has good results, and lower complication rate compared with surgery. Other factors to take into consideration are pressure of initial compression, compliance in wearing the brace, and patients with complex deformities. Surgery should be considered for those who have underlying syndromes such as Marfan or Poland. Limitations in the study include electing appropriate course of treatment, the quantification of initial deformities and outcomes, and loss of follow-up.

Geraedts et al. (2022) performed a systematic review of the outcomes after minimally invasive PC repair by the Abramson method. The PubMed and Embase databases were systematically searched. Data concerning subjective postoperative esthetic outcomes after initial surgery and bar removal were extracted. In addition, data on recurrence, complications, operative times, blood loss, post-operative pain, length of hospital stay, planned time to bar removal and reasons for early bar removal were extracted. The postoperative esthetic result, was selected as primary outcome since the primary indication for repair in PC is of cosmetic nature. Six cohort studies were included based on eligibility criteria, enrolling a total of 396 patients. Qualitative synthesis showed excellent to satisfactory esthetic results in nearly all patients after correctional bar placement (99.5%, n = 183/184). A high satisfaction rate of 91.0% (n = 190/209) was found in patients after bar removal. Recurrence rates were low with an incidence of 3.0% (n = 5/168). The cumulative postoperative complication rate was 26.5% (n = 105/396), of whom 25% required surgical re-intervention. There were no cases of mortality. The authors concluded minimally invasive repair of PC through the Abramson method is effective and safe. They further noted its efficacy is demonstrated by the excellent to satisfactory esthetic results in 99.5% and 91.0% of patients after respectively correctional bar placement and implant removal. The authors recommended future studies should aim to compare different treatment options for PC in order to elucidate the approach of choice for different patient groups. The review was limited by the small number of studies and lack of a comparison group.

Martinez-Ferro et al. (2019) explored the evolution of the diagnosis and treatment of PC up to its current management in a critical review. Management alternatives have shifted from open resective to minimally invasive strategies, and finally, to reshaping the chest using both surgical and non-surgical modalities. Open resective surgeries of the affected cartilages associated with sternal osteotomies were the first operations performed for this thoracic deformity. These surgeries, or variations of them, are still the first choice in cases of very deformed, rigid, non-articulated sternums with associated malformations, or after other approaches have failed. Minimally invasive resective techniques, such as the Nuss procedure, are also used. Non-resective techniques are more effective in flexible thoracic cages that are also amenable to non-operative bracing, so their use in PC is gradually decreasing. Bracing therapy has gained popularity as a non-operative alternative that has proven to be as good as operative strategies to correct PC in patients with flexible thoracic cages and high compliance. This is a good choice as the first line of treatment in almost all cases, and the only solution necessary in many cases with non-complex deformities, flexible thoracic cages and a high level of compliance. There is still a lack of consensus regarding the minimum number of hours patients should wear the brace, and whether bracing may play a role in adult patients.

In a systematic review, de Beer et al. (2018) reviewed studies on the treatment of PC with measured dynamic compression allows measurement and adjustments of the brace's pressure on the thoracic wall, leading to a controlled correction. The authors performed an electronic database search (PubMed and Cochrane) of the medical literature on measured dynamic compression. A total of 14 studies were found and eight studies between 2008 and 2018 were included. Study designs ranged from retrospective chart reviews to cross-sectional cohort studies. From the 8 studies, 1185 patients were included. The median age was 14 years (range 2–28) and 87% were male. The mean study follow up period was 16 months; 44% of patients were still under treatment, 29% of patients successfully completed treatment. 6% dropped out and 21% were lost to follow-up. Dropout was mainly caused social discomfort (7.2%) and failure of treatment (5.8%). Complications were infrequent. Mild chest discomfort or tightness was reported in 12% and skin lesions occurred in 5.1%. The overall recurrence rate was 2.6%. The authors concluded dynamic compression appears to be a safe, non-invasive and efficient treatment to correct PC in patients with a non-rigid thorax. The noted patients experience less discomfort, which in turn results in better compliance, however, accurate selection of patients based on age, pressure of initial correction and motivation is important and an objective scoring system to assess the esthetic and long-term physical and psychological results of the treatment is needed.

#### **Clinical Practice Guidelines**

#### American Pediatric Surgical Association (APSA)

In a clinical practice guideline, APSA (2012) indicates when a pectus carinatum chest wall deformity represents a significant deviation from normal and is associated with symptoms, nonoperative or operative corrective therapy is considered reconstructive as it restores function and alleviates symptoms. When it is performed for therapeutic purposes, the surgery for pectus carinatum falls under the definition of reconstructive surgery, and not cosmetic surgery, as defined

Pectus Deformity Repair UnitedHealthcare West Medical Management Guideline Proprietary Information of UnitedHealthcare. Copyright 2024 United HealthCare Services, Inc. by the American Medical Association and the Centers for Medicaid and Medicare as it is "performed to improve function, but may also be done to approximate a normal appearance."

APSA makes the following recommendations:

- For the child diagnosed with a pectus carinatum deformity physical evaluation for scoliosis should be performed. Dictated by the clinical presentation, an evaluation for congenital heart disease and Marfan's syndrome may also be performed.
- Symmetry of the pectus deformity, degree of sternal rotation, chest wall compliance, and the presence of a concomitant pectus excavatum deformity should be assessed.
- Although not required, chest computed tomography may assist in the surgical planning and play a role in determining the extent of the deformity in the child with a significant pectus carinatum.
- In prepubertal children, a period of observation to follow the progression of the pectus carinatum and to allow for discussion regarding the optimal method of therapy is appropriate. Without strong evidence for ideal timing of treatment, expert opinion suggests that the age for operative therapy must be individualized, but is typically deferred until pubertal growth in nearly complete.
- As reconstructive therapy for the compliant pectus deformity, nonoperative compressive orthotic bracing is usually an appropriate first line therapy as it does not preclude the operative option. For appropriate candidates, orthotic bracing of chest wall deformities can reasonably be expected to prevent worsening of the deformity and often result in a lasting correction of the deformity. Orthotic bracing is often successful in prepubertal children whose chest wall is compliant. Expert opinion suggests that the noncompliant chest wall deformity or significant asymmetry of the pectus carinatum deformity caused by a concomitant excavatum-type deformity may not respond to orthotic bracing.
- Open surgical reconstructive techniques are acceptable surgical options in the hands of experienced pediatric surgeons.
- Thoracoscopic reconstructive and other minimally invasive techniques are acceptable in some children, based on the advanced minimally invasive skills and experience of the pediatric surgeon.
- Unless there is some overwhelming indication for repair, operative repair of pectus chest wall deformities is 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.
- Expert opinion suggests ongoing evaluation through adolescence by a pediatric surgeon is appropriate in the child who has undergone nonoperative or operative chest wall reconstruction therapy. Due to rib, cartilage, and pubertal linear growth with resultant ongoing changes in the chest wall contour that may occur, the pediatric surgeon should be involved in the extended follow up of these children.

### U.S. Food and Drug Administration (FDA)

This section is to be used for informational purposes only. FDA approval alone is not a basis for coverage.

Pectus deformity repair is a procedure and, therefore, not subject to FDA regulation. However any medical devices, drugs, biologics, or tests used as a part of this procedure may be subject to FDA regulation. Refer to the following website for more information on devices used for pectus deformity repair (search by product code HRS): https://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfPMN/pmn.cfm. (Accessed February 23, 2024)

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# **Guideline History/Revision Information**

Date	Summary of Changes		
06/01/2024	Supporting Information		
	• Updated Clinical Evidence and References sections to reflect the most current information		
	<ul> <li>Archived previous policy version MMG101.Q</li> </ul>		

# **Instructions for Use**

This Medical Management Guideline provides assistance in interpreting UnitedHealthcare standard benefit plans. When deciding coverage, the member specific benefit plan document must be referenced as the terms of the member specific benefit plan may differ from the standard plan. In the event of a conflict, the member specific benefit plan document governs. Before using this guideline, please check the member specific benefit plan document and any applicable federal or state mandates. UnitedHealthcare reserves the right to modify its Policies and Guidelines as necessary. This Medical Management Guideline is provided for informational purposes. It does not constitute medical advice.

UnitedHealthcare may also use tools developed by third parties, such as the InterQual<sup>®</sup> criteria, to assist us in administering health benefits. UnitedHealthcare West Medical Management Guidelines are intended to be used in connection with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

Member benefit coverage and limitations may vary based on the member's benefit plan Health Plan coverage provided by or through UnitedHealthcare of California, UnitedHealthcare Benefits Plan of California, UnitedHealthcare of Oklahoma, Inc., UnitedHealthcare of Oregon, Inc., UnitedHealthcare Benefits of Texas, Inc., or UnitedHealthcare of Washington, Inc.