



Pectus carinatum

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Purpose of review

Pectus carinatum has been termed the undertreated chest wall deformity. Recent advances in patient evaluation and management, including the development of nonoperative bracing protocols, have improved the care of children with this condition.

Recent findings

Recent evidence confirms that children with pectus carinatum have a disturbed body image and a reduced quality of life. Treatment has been shown to improve the psychosocial outcome of these patients.

Summary

Patients with pectus carinatum are at risk for a disturbed body image and reduced quality of life. Until recently, treatment required surgical reconstruction. A growing body of literature, however, now supports the use of orthotic bracing as a nonoperative alternative in select patients. This article reviews the current literature and describes the evaluation and management of children with pectus carinatum deformity.

Keywords

chondrogladiolar, chondromanubrial, orthotic bracing, pectus carinatum

INTRODUCTION

Pectus carinatum is the second most common chest wall deformity observed in children. Whereas the more common pectus excavatum deformity has received a great deal of recent attention in the literature due to the associated cardiac and pulmonary dysfunction and alternative surgical options, recent evidence affirms the long held belief that pectus carinatum can lead to significant psychological distress and thus warrants an equally aggressive management approach. Numerous studies have demonstrated the efficacy of both operative repair and nonoperative bracing for correction of pectus carinatum. The authors' current approach to evaluation and management of children with pectus carinatum will be reviewed.

DEFINITIONS

Pectus carinatum is a term used to characterize a range of chest wall deformities defined by anterior protrusion of the sternum and adjacent costal cartilages, and is in distinction to the more common pectus excavatum, which describes chest wall depression deformities. Pectus carinatum deformities can be subclassified into two distinct entities depending on the component of the sternum involved. The chondrogladiolar variant describes protrusion of the gladiolus, or body of the sternum

(Figs 1a and 2a). This deformity has also been referred to as 'keel chest'. The chondromanubrial variant describes protrusion of the manubrium, or superior component of the sternum, and has been termed the 'pouter pigeon breast', 'Currarino–Silverman syndrome', 'horseshoe chest', and 'horns of steer'. This deformity can present as either an isolated manubrial protrusion or 'mixed' defect with manubrial protrusion and gladiolar depression (Fig. 3a).

EPIDEMIOLOGY

Pectus carinatum is a relatively common chest wall deformity occurring with a male to female ratio of approximately 4 : 1 [1,2[•]]. Two groups have investigated the overall prevalence of both pectus deformities in children. Westphal *et al.* [3], in a cohort of 1332 children aged 11–14 years of age,

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KEY POINTS

- Correction of pectus carinatum is indicated for children with a significant deformity and associated pain or disturbed body image.
- Cardiac and pulmonary dysfunction is uncommon with pectus carinatum.
- Preoperative work-up of pectus carinatum typically requires only plain chest radiographs.
- Orthotic bracing is the preferred initial management technique in the majority of children with pectus carinatum requiring correction.
- The optimal timing of treatment of pectus carinatum is between the ages of 10 and 15 years to allow bracing protocols and to minimize recurrence risk.

reported a pectus carinatum prevalence of 0.675% and pectus excavatum prevalence of 1.275%. Coskun *et al.* [4], in their cohort of 1342 children aged 7–14 years of age, reported a similar pectus carinatum prevalence of 0.6% but reported a pectus excavatum prevalence of 2.6%. The chondrogladiolar variant is by far the most common, found in 92.3 to 95% of children with pectus carinatum deformity [5–7].

CAUSE AND ASSOCIATED CONDITIONS

Similarly to pectus excavatum deformities, the exact pathogenesis of pectus carinatum remains unknown. For the chondrogladiolar subtype, it is generally believed that abnormal costal cartilage

growth is the underlying defect [5]. In contrast, there is evidence to suggest that the chondromammary pectus carinatum subtype is due to sternal ossification abnormalities and is therefore a distinct entity [8,9]. The finding of concomitant thoracolumbar scoliosis in 12–34% of patients suggests that pectus carinatum may actually be the result of a ubiquitous connective tissue disorder [1,10,11]. A high degree of genetic predisposition is likely as a family history of chest wall deformities is found in approximately one-fourth of children with pectus carinatum [1,3,10]. Additionally, numerous genetic abnormalities may manifest pectus carinatum. Two relatively common genetic conditions that have associated pectus carinatum are Marfan and Noonan syndrome [12]. A pectus deformity is reported in 70–95% of children with Noonan syndrome and is a component of the diagnostic criteria for both Marfan syndrome and Noonan syndrome [13,14]. Finally, and as a unique cause, cases of ‘reactive’ or ‘iatrogenic’ pectus carinatum have also been reported to develop as a complication of surgical pectus excavatum repair [15].

CLINICAL PRESENTATION

Children with pectus carinatum may present at any age, but typically are brought to medical attention at the time of puberty, as the deformity becomes more prominent during the adolescent growth spurt. At the time of presentation, patient complaints typically consist of cosmetic concerns leading them to avoid activities such as swimming or changing in front of others [5,6]. As such, it is not surprising that patients with pectus carinatum have been shown to

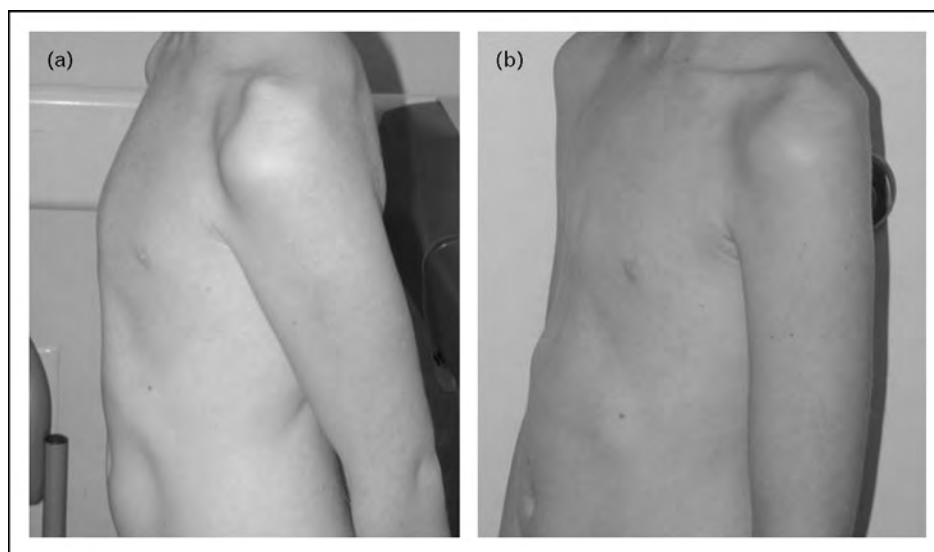


FIGURE 1. Prebracing (a) and postbracing (b) photographs of a patient with chondrogladiolar pectus carinatum.

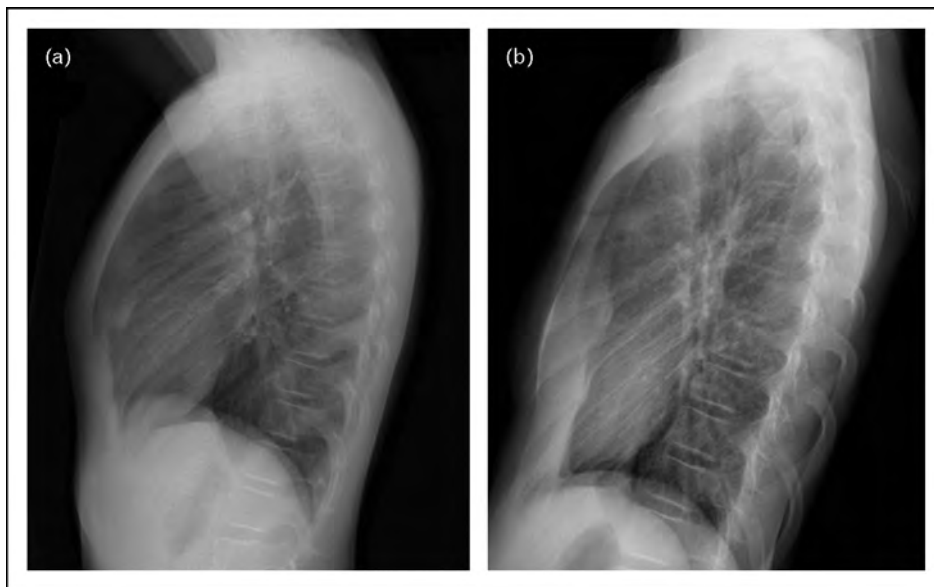


FIGURE 2. Prebracing (a) and postbracing (b) lateral X-rays of the patient with chondrogladiolar pectus carinatum pictured in Fig. 1.

have a disturbed body image and reduced quality of life when compared with controls [16[■]]. Additionally, patients with a pectus deformity often believe their physical appearance is more impaired than do adult raters without the deformity, suggesting an exaggerated impairment of self-image [17]. Children seeking surgical correction of pectus carinatum thus overwhelmingly report cosmetic reasons as the primary indication for correction [16[■],18[■]].

Physical symptoms may also be present with pectus carinatum and include musculoskeletal chest wall discomfort particularly while lying prone, respiratory symptoms, or palpitations [5,6,19–21]. Whereas some authors have reported respiratory symptoms to be present in only a minority of patients [5,6,21], others have described virtually all children having compensatory tachypnea, dyspnea, exercise intolerance, or wheezing [19,20]. Our experience suggests that respiratory symptoms are

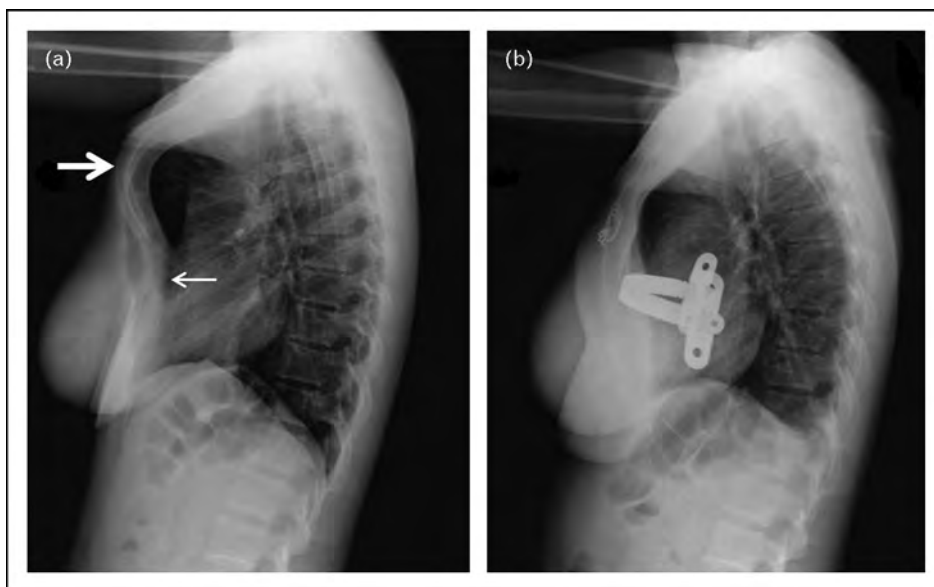


FIGURE 3. Preoperative (a) and 1-month postoperative (b) lateral X-rays of a patient with mixed-type chondromanubrial pectus carinatum who underwent reconstruction. Mixed type has both a carinatum (bold arrow) and an excavatum component (thin arrow).

only occasionally present and typically minor in nature. Finally, female patients in their late teens may also report an inability to wear a bra and body image concerns over breast symmetry.

EVALUATION

A number of authors have advocated pretreatment computed tomography (CT) scan to document the degree of sternal and cartilaginous elevation [22] and to predict success of treatment [23]. It is our experience, however, that the majority of children with pectus carinatum only necessitate a posterior–anterior and lateral chest X-ray. These images are typically adequate to document the sternal abnormality, assure no alternative pathology, and follow treatment progression. More advanced imaging (CT or MRI) may be appropriate for children with ‘mixed’ deformities to evaluate the degree of sternal depression and cardiac compression.

Select patients with pectus carinatum may require additional preoperative testing. Children who report dyspnea, palpitations, or other cardiopulmonary complaints require pulmonary function tests (PFTs) and cardiology evaluation. Although less frequent than in pectus excavatum, cardiac abnormalities have been reported in up to 6% of children with pectus carinatum [10]. An echocardiogram is particularly important in those patients with a ‘mixed’ deformity to document any cardiac dysfunction secondary to compression by the depressed component of the sternum. In those children with dysmorphic features suggestive of a genetic syndrome (Noonan or Marfan), genetics consultation is warranted to allow long-term multispecialty follow-up.

OPERATIVE TREATMENT

Operative repair of pectus carinatum, as first described by Ravitch [23], has been the mainstay of treatment for over 50 years. Since the first description, a number of large series have reported the technique and outcomes for this initial operative repair of pectus carinatum [1,5,11]. The surgical steps include an anterior midline or transverse incision, elevation of the pectoralis muscles off the sternum and thoracic cage, resection of the involved costal cartilages, and typically at least one sternal osteotomy to allow repositioning of the sternum. For additional sternal support, mesh [5] or a stainless steel strut [10] has been used to secure the sternum at the desired height.

Recent larger series have focused on further modifications to the initial technique. These augmentations include reduced cartilage resection

[20], the use of less extensive muscle dissection coupled with bioabsorbable sternal plating and postoperative bracing [7], and reduced cartilage resection coupled with endoscopic dissection [19].

As is the case in the repair of pectus excavatum, some authors have tried to come up with an even less invasive technique to repair pectus carinatum. Abramson *et al.* [24] describe repair through placement of a long curved bar anterior to the sternum via lateral chest wall incisions. The bar is fixed to the ribs after the chest deformity is manually depressed to the desired level. No cartilage is resected and the bar is not removed for at least 12 months. A nearly identical technique has also been reported by Yüksel *et al.* [25]. Schaarschmidt *et al.* [26] report using a similar bar with endoscopic dissection and limited cartilage resection. Whereas the more extensive Ravitch-based technique may be utilized for all subtypes of pectus carinatum, the bar-based technique, which acts like an externally applied brace, is not useful for treating chondromanubrial pectus carinatum due to the superior location of the sternal protrusion and more limited chest wall compliance found in these patients.

In general, the outcomes following repair of pectus carinatum are good. Regardless of the operative technique, the vast majority of patients report being satisfied with the results of their procedure [1,5,7,10,19,24,26]. Bostanci *et al.* [18[■]], utilizing a standardized questionnaire, showed a significant improvement in both the patient and parent rating of the psychosocial and physical function of patients between the preoperative and 6-month postoperative assessment. While subjective appraisal has its limitation, this is the way in which the success of a repair is determined. A recent survey of Canadian Pediatric Surgeons reported that ‘surgeon appraisal of appearance’ and ‘patient opinion’ are used by the overwhelming majority of clinicians [27[■]]. In addition to appearance, Fonkalsrud [20] reported extensive improvement in respiratory complaints following surgical repair. Cahill *et al.* [28], however, demonstrated no changes in preoperative and postoperative PFTs and progressive work exercise in their small series.

Complications following operative repair of pectus carinatum are rare and include pneumothorax, wound infection, and local skin necrosis, and seem to be unrelated to operative technique [1,7,20,24]. Numerous other procedure-specific complications have been described. Authors using techniques to minimize open dissection have reported a higher incidence of postoperative seroma formation [19,24]. Those utilizing a less invasive bar-based technique report cases of skin adherence to the bar, fracture of the wire used to secure the bar,

and overcorrection necessitating early bar removal [24,25]. Although not a complication of the procedure, recurrence has been shown to occur in up to 5.5% of patients treated with operation [7,20]. Surgical repair after completion of the pubertal growth spurt has been recommended to reduce the rate of recurrence [29].

Although typically a disease treated by pediatric surgeons, there is virtually no upper age limit for surgical correction of patients with pectus carinatum and series have included patients up to 49 years of age [7]. With regard to a lower age range, caution has been suggested for operating on children less than 4 years of age due to the fear of causing long-term growth problems and the development of iatrogenic thoracic dystrophy, which has been described with early repair of children with pectus excavatum [30]. The recurrence risk following early surgical repair also appears to be much higher and supports the decision to correct the condition as patients go through puberty.

BRACING TREATMENT

Evidence to support the use of bracing for pectus carinatum has existed since 1992 [31]. While this treatment option has only recently garnered more attention in the English-speaking surgical literature, a survey of pediatric surgeons shows that this technique is widely used, and has supplanted operative repair as the initial treatment of choice in select patients [27].

The basic principle of the bracing protocol is the application of continuous pressure over a long-term period to the protruding deformity to allow remodeling of the abnormal costal cartilage. Pectus carinatum corrective braces surround the thorax and have at a minimum two points of contact, one anterior contact point providing direct pressure to the deformity and a second contact point providing counter pressure, typically to the patient's back (Fig. 4). It is generally believed that a compliant

chest wall is a requisite for any bracing procedure to work and that the chest wall becomes less compliant after puberty [2^o,21,32,33]. As such, children aged 10–15 years are the best candidates for this technique. Martinez-Ferro *et al.* [2^o] have developed a bracing system that allows measurement of the pressure necessary for initial correction. Due to changes in chest wall compliance with age, they found that older children required a greater pressure and duration of bracing to achieve satisfactory correction.

The current literature suggests a great deal of variation in bracing protocols across centers. Duration of bracing is variable and has been recommended to continue until the patient and physician are content with the result [2^o,34], for a defined period of 6 months [31], or until the completion of linear growth [21,32,35]. The recommended time for wearing the brace per day ranges from 14 [32] to 24 h per day [33]. Other protocols have recommended wear for up to 23 h per day until initial resolution of the deformity. This is then followed by a maintenance phase lasting until the completion of linear growth, during which the patient is only asked to wear the brace overnight [21,35].

The most common cause of bracing failure is due to patient compliance. Recent series have reported patient compliance with bracing to range from 78 to 90% [2^o,7,32,34,35]. Regardless of the specific bracing protocol, it seems that the vast majority of compliant patients can achieve positive results from bracing [2^o,7,31–35]. While current studies indicate that bracing can lead to excellent patient satisfaction, the long-term efficacy of this treatment modality, however, is unknown. Martinez-Ferro *et al.* [2^o] in a series without a maintenance phase of bracing and mean follow-up of 3.3 years reported a recurrence rate of 15%, noting that all recurrences achieved complete resolution with additional bracing.

Complications with bracing are generally minor. The most common complaint is rash or skin



FIGURE 4. Demonstration of the orthotic bracing device used at our institution.

discoloration over the point of brace contact [2³,31,33]. In a series of 112 patients, back pain (8), hematoma (1), and skin ulceration (5), were reported [2³]. Finally, Haje and Haje [36] report 15 cases of overcorrection with bracing leading to an iatrogenic pectus excavatum. Frequent follow-up is therefore important to assure appropriate fit and correction.

OUR TREATMENT APPROACH

Individuals should be seen in the surgery clinic as soon as the child, parent, or primary provider expresses any concern about the pectus deformity. Early referral provides an opportunity to reassure all individuals involved that pectus carinatum is not life-threatening and that it can be effectively treated with all management options. After consultation and reassurance a number of children and parents opt to not undergo intervention. These patients are typically put at ease with an explanation of the deformity and its natural history. In patients who have completed their pubertal growth spurt we inform them that there is unlikely to be any further significant change. As others have previously described [31,34], we often suggest exercises to develop anterior chest wall musculature to minimize the visibility of the deformity. If the child has not completed their pubertal growth spurt, we explain that with the rapid growth of puberty the deformity may progress. Follow-up for progression of the deformity is important in these patients. Symptomatic children and those with body image concerns typically warrant interventions.

In appropriately aged children with a chondrogladiolar pectus carinatum, bracing should always be the first option for intervention. Chest wall compliance determines the upper age of bracing and can be evaluated by a manual compression test [31,33]. Anyone who can achieve a near-normal chest shape during anterior–posterior compression on the most prominent portion of the sternum may undergo a bracing trial. Our bracing protocol consists of brace wear for 14–16 h per day over a 16-month period. We warn patients to look for skin irritation and see them in follow-up every 3 months. Chest X-rays and digital images are obtained at the onset and completion of the bracing protocol to document progress (Figs 1 and 2).

In children with chondrogladiolar pectus carinatum who are not candidates for bracing or fail a bracing trial we would consider surgical intervention. For those patients with chondromanubrial pectus carinatum we do not feel that there is convincing evidence for success with bracing and do not offer this as a management option. Our surgical

repair typically utilizes a modified Ravitch technique. We have on occasion used wire to close the sternal osteotomy and implanted a bar, used more commonly for minimally invasive pectus excavatum repair, as a support strut. The bar's thickness, strength, and resistance to deforming stresses allows it to maintain its shape against the less compliant chest wall found in older children (Fig. 3). Consensus guidelines for the management of pectus carinatum have recently been published by the American Pediatric Surgical Association [37³].

CONCLUSION

Pectus carinatum is a relatively common chest wall abnormality with an unknown cause. While the impact of treatment on quality of life is well documented, the effect on physiologic dysfunction is less well established. Treatment should begin with a consideration of orthotic bracing in the vast majority of patients. Due to differences in chest wall compliance required for successful bracing, early referral is indicated. Overall, success rates with both bracing and operative management are high.

Acknowledgements

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Conflicts of interest

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- of outstanding interest

Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 434).

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