# CURRENT OPINION

# Pectus carinatum

*Thomas J. Desmarais[a,b] and Martin S. Keller[a]*

**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,

[a]Division of Pediatric Surgery, Washington University School of Medicine in Saint Louis, Saint Louis, Missouri and [b]Geisel School of Medicine at Dartmouth, Hanover, New Hampshire, USA

Correspondence to Martin S. Keller, MD, FACS, FAAP, Department of Pediatric Surgery, Saint Louis Children's Hospital, 1 Children's Place, Saint Louis, MO 63110, USA. Tel: +1 314 454 6022; e-mail: kellerm@wudosis.wustl.edu

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 chondromanubrial 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
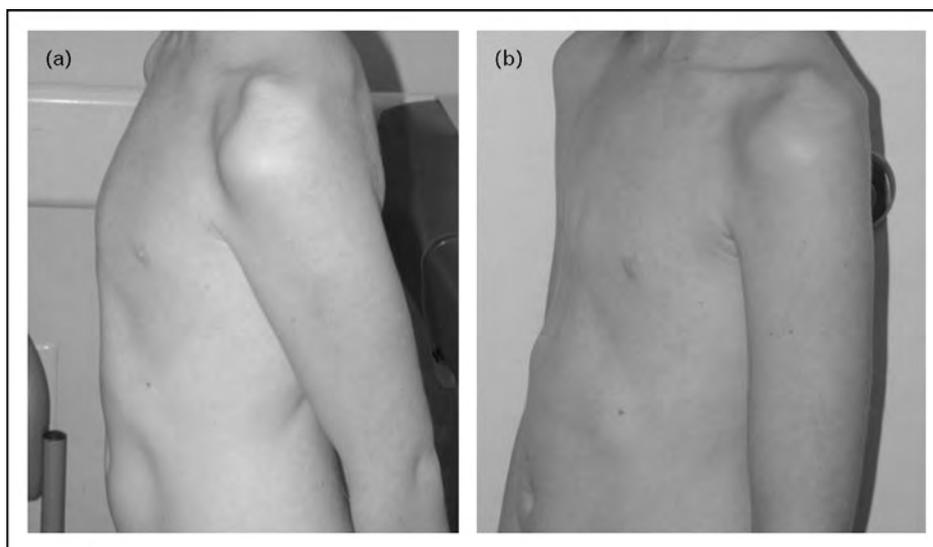


**FIGURE 1.** Prebracing (a) and postbracing (b) photographs of a patient with chondrogladiolar pectus carinatum.
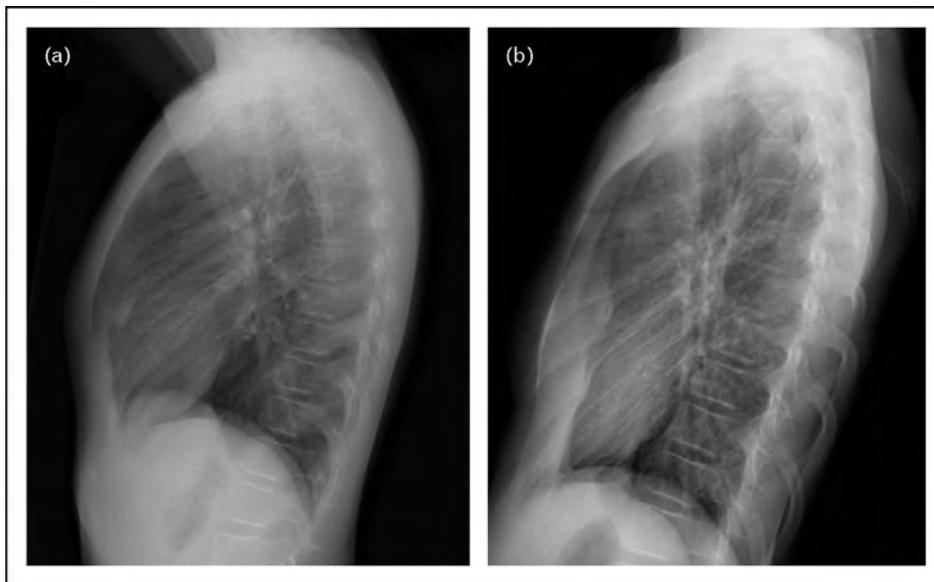
**FIGURE 2.** Prebracing (a) and postbracing (b) lateral X-rays of the patient with chondrogladiolar pectus carinatum pictured in Fig. 1.

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
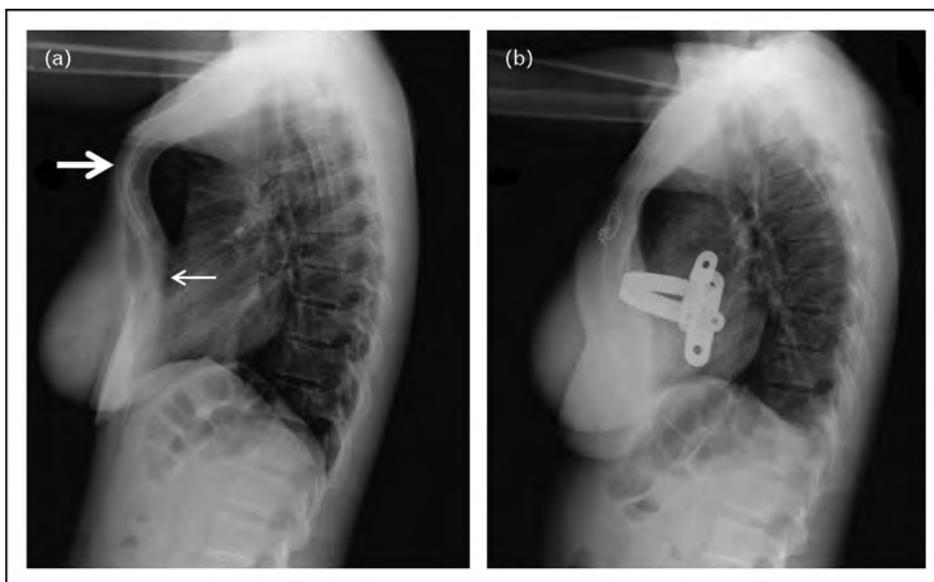


**FIGURE 3.** Preoperative (a) and 1-month postoperative (b) lateral X-rays of a patient with mixed-type chondromanubrial pectus carinatum who underwent reconstruction. Mixed type has both a carinatum (bold arrow) and an excavatum component (thin arrow).

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■,21,32,33]. As such, children aged 10–15 years are the best candidates for this technique. Martinez-Ferro et al. [2■] 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■,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■,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■,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■] 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 [2■,31,33]. In a series of 112 patients, back pain (8), hematoma (1), and skin ulceration (5), were reported [2■]. 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 [37■].

## 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.

## REFERENCES AND RECOMMENDED READING

Papers of particular interest, published within the annual period of review, have been highlighted as:
■  of special interest
■■  of outstanding interest
Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 434).

1. Shamberger RC, Welch KJ. Surgical correction of pectus carinatum. J Pediatr Surg 1987; 22:48–53.

2. Martinez-Ferro M, Fraire C, Bernard S. Dynamic compression system for the
■ correction of pectus carinatum. Semin Pediatr Surg 2008; 17:194–200.
This is one of the largest series reporting orthotic bracing as treatment for pectus carinatum and demonstrates a unique bracing system that allows real-time pressure measurement.

3. Westphal FL, Lima LC, Lima Neto JC, et al. Prevalence of pectus carinatum and pectus excavatum in students in the city of Manaus, Brazil. J Bras Pneumol 2009; 35:221–226.

4. Coskun ZK, Turgut HB, Demirsoy S, Cansu A. The prevalence and effects of pectus excavatum and pectus carinatum on the respiratory function in children between 7–14 years old. Indian J Pediatr 2010; 77:1017–1019.

5. Robicsek F, Cook JW, Daugherty HK, Selle JG. Pectus carinatum. J Thorac Cardiovasc Surg 1979; 78:52–61.

6. Welch KJ, Vos A. Surgical correction of pectus carinatum (pigeon breast). J Pediatr Surg 1973; 8:659–667.

7. Del Frari B, Schwabegger AH. Ten-year experience with the muscle split technique, bioabsorbable plates, and postoperative bracing for correction of pectus carinatum: the Innsbruck protocol. J Thorac Cardiovasc Surg 2011; 141:1403–1409.

8. Currarino G, Silverman FN. Premature obliteration of the sternal sutures and pigeon-breast deformity. Radiology 1958; 70:532–540.

9. Haje SA, Harcke HT, Bowen JR. Growth disturbance of the sternum and pectus deformities: imaging studies and clinical correlation. Pediatr Radiol 1999; 29:334–341.
10. Fonkalsrud EW, Beanes S. Surgical management of pectus carinatum: 30 years' experience. World J Surg 2001; 25:898–903.
11. Waters P, Welch K, Micheli LJ, et al. Scoliosis in children with pectus excavatum and pectus carinatum. J Pediatr Orthop 1989; 9:551–556.
12. Kotzot D, Schwabegger AH. Etiology of chest wall deformities: a genetic review for the treating physician. J Pediatr Surg 2009; 44:2004–2011.
13. Van der Burgt I. Noonan syndrome. Orphanet J Rare Dis 2007; 2:4.
14. Loeys BL, Dietz HC, Braverman AC, et al. The revised Ghent nosology for the Marfan syndrome. J Med Genet 2010; 47:476–485.
15. Swanson JW, Colombani PM. Reactive pectus carinatum in patients treated for pectus excavatum. J Pediatr Surg 2008; 43:1468–1473.
16. Steinmann C, Krille S, Mueller A, et al. Pectus excavatum and pectus
■  carinatum patients suffer from lower quality of life and impaired body image: a control group comparison of psychological characteristics prior to surgical correction. Eur J Cardiothorac Surg 2011; 40:1138–1145.
This is an in-depth demonstration of the quality of life and body image problems experienced by patients with uncorrected pectus deformities.
17. Krille S, Müller A, Steinmann C, et al. Self- and social perception of physical appearance in chest wall deformity. Body Image 2012; 9:246–252.
18. Bostanci K, Ozalper MH, Eldem B, et al. Quality of life of patients who have
■■  undergone the minimally invasive repair of pectus carinatum. Eur J Cardiothorac Surg 2013; 43:122–126.
This is the first study to document improved quality of life following correction of pectus carinatum. Improvements were shown for both the psychosocial and physical functioning components. This serves as a benchmark for future quality of life studies in pectus carinatum.
19. Schaarschmidt K, Kolberg-schwerdt A, Lempe M, Schlesinger F. New endoscopic minimal access pectus carinatum repair using subpectoral carbon dioxide. Ann Thorac Surg 2006; 81:1099–1103.
20. Fonkalsrud EW. Surgical correction of pectus carinatum: lessons learned from 260 patients. J Pediatr Surg 2008; 43:1235–1243.
21. Kravarusic D, Dicken BJ, Dewar R, et al. The Calgary protocol for bracing of pectus carinatum: a preliminary report. J Pediatr Surg 2006; 41:923–926.
22. Colombani PM. Preoperative assessment of chest wall deformities. Semin Thorac Cardiovasc Surg 2009; 21:58–63.
23. Ravitch MM. Unusual sternal deformity with cardiac symptoms operative correction. J Thorac Surg 1952; 23:138–144.
24. Abramson H, D'Agostino J, Wuscovi S. A 5-year experience with a minimally invasive technique for pectus carinatum repair. J Pediatr Surg 2009; 44:118–123.
25. Yüksel M, Bostanci K, Evman S. Minimally invasive repair of pectus carinatum using a newly designed bar and stabilizer: a single-institution experience. Eur J Cardiothorac Surg 2011; 40:339–342.
26. Schaarschmidt K, Lempe-Sellin M, Schlesinger F, et al. New Berlin-Buch 'reversed Nuss,' endoscopic pectus carinatum repair using eight-hole stabilizers, submuscular $CO_2$, and presternal Nuss bar compression: first results in 35 patients. J Laparoendosc Adv Surg Tech A 2011; 21:283–286.
27. Emil S, Laberge JM, Sigalet D, Baird R. Pectus carinatum treatment in
■  Canada: current practices. J Pediatr Surg 2012; 47:862–866.
This study is the first to report the current practice pattern of a large group of physicians treating pectus carinatum.
28. Cahill JL, Lees GM, Robertson HT. A summary of preoperative and postoperative cardiorespiratory performance in patients undergoing pectus excavatum and carinatum repair. J Pediatr Surg 1984; 19:430–433.
29. Colombani PM. Recurrent chest wall anomalies. Semin Pediatr Surg 2003; 12:94–99.
30. Haller JA, Colombani PM, Humphries CT, et al. Chest wall constriction after too extensive and too early operations for pectus excavatum. Ann Thorac Surg 1996; 61:1618–1624.
31. Haje SA, Bowen JR. Preliminary results of orthotic treatment of pectus deformities in children and adolescents. J Pediatr Orthop 1992; 12:795–800.
32. Frey AS, Garcia VF, Brown RL, et al. Nonoperative management of pectus carinatum. J Pediatr Surg 2006; 41:40–45.
33. Lee SY, Lee SJ, Jeon CW, et al. Effect of the compressive brace in pectus carinatum. Eur J Cardiothorac Surg 2008; 34:146–149.
34. Banever GT, Konefal SH, Gettens K, Moriarty KP. Nonoperative correction of pectus carinatum with orthotic bracing. J Laparoendosc Adv Surg Tech A 2006; 16:164–167.
35. Stephenson JT, Du Bois J. Compressive orthotic bracing in the treatment of pectus carinatum: the use of radiographic markers to predict success. J Pediatr Surg 2008; 43:1776–1780.
36. Haje SA, Haje DP. Overcorrection during treatment of pectus deformities with DCC orthoses: experience in 17 cases. Int Orthop 2006; 30:262–267.
37. APSA Practice Committee. Pectus carinatum guidelines. American Pediatric
■  Surgical Association; August 2012. Available from: http://www.eapsa.org// Surgeons/Content/NavigationMenu/AboutAPSA/StatementsGuidelines/ default.htm. [Accessed December 2012]
This is the first set of practice guidelines published for pectus carinatum.