

Current Opinion

Pectus Carinatum: The Undertreated Chest Malformation

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Introduction

Pectus carinatum (PC) consists of a spectrum of congenital protrusion deformities of the chest, the most frequent being anterior displacement of the mid and lower sternum and adjacent costal cartilages, which is usually first recognized during early adolescence. Asymmetrical deformities with a tilted sternum are common, occasionally with localized depression of one or both sides of the lower anterolateral chest. Protrusion of the upper sternum (chondromanubrial) with occasional depression of the lower sternum occurs in approximately 12% of cases.¹ PC is approximately seven times less frequent than pectus excavatum (PE), and occurs more than six times more often in males.² More than 30% of PC patients have a familial occurrence of pectus deformities.¹ Less than 5% have congenital heart disease.¹ More than 60% have mild to moderate scoliosis.¹

Symptoms from PC are infrequent during early childhood, except for those few patients who develop chondromanubrial prominence at an early age (10%).¹ Although many adolescent children with PC deformities indicate to parents and physicians that they have increasingly severe shortness of breath, often with only mild exercise, and reduced stamina and endurance with activity, as well as exercise-induced wheezing, most physicians are likely to indicate to the family that there is no correlation between PC and cardiorespiratory dysfunction, and that the deformity is primarily a cosmetic problem.³ Patients are often advised that PC is difficult to correct surgically and that the operation is very painful with many complications and is minimally effective in producing physiological benefit. Each of these views are incorrect based on present knowledge of PC deformities and current techniques for surgical repair.¹ Several surgeons have observed decreased

respiratory excursions of the thorax with increased anteroposterior chest diameter, resulting in reduced chest wall movement with respiration.⁴ Few publications during the past two decades have indicated the long-term results following repair of PC deformities.

A paucity of physiological studies exists regarding the cardiorespiratory effects of severe PC deformities.⁵ Objective measurements to document the severity of physical limitations caused by PC while at rest have often been imprecise and misleading. Pulmonary function studies should be performed during exercise. The symptoms expressed by the patient often appear far out of proportion to the apparently mild protrusion of the chest. With PC, the thorax is held in a partially expanded position with increased residual air volume and reduced vital capacity. Some very competitive patients have been able to compensate for PC-induced limitations in chest expansion in short-duration athletic activities by wider diaphragmatic excursions, at a cost of greater energy expenditure.

A comprehensive review of many published reports indicating changes in cardiac or respiratory function after PC repair was provided by Shamberger and Welch.⁶ The severity of the pectus protrusion can easily be measured by calculating the pectus severity index from chest radiographs or computed tomography (CT) scans.^{2,7} Because of the high cost and often invasive nature of the procedures used for physiological evaluation before and after pectus repair, we do not favour the routine use of such tests.

Asthmatic symptoms are more frequent in patients with PC than in those with PE. Several patients have complained of pain from pressure on the protruding sternum and/or cartilages. The symptomatic child often pleads for the parents to seek help in correcting the deformity. During the past

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several years, an increasing number of patients over the age of 18 years have sought correction of PC deformities because of worsening symptoms and persistent limitation of activity.

Surgical repair

Although a wide variety of PC configurations have been described, there are two basic types. Young children occasionally develop severe protrusion of the manubrium and upper sternum, which may cause shortness of breath, frequent respiratory infections, and wheezing with exercise compared to other children.⁸ Repair may require more extensive reconstruction in these young children because five or six of the cartilages on each side may require reconstruction and the sternum may need two or more transverse osteotomies. External compression harnesses have had minimal effect in correcting PC and are cumbersome and painful.

Repair of the more common mid and lower sternal protrusion in adolescents and adults often requires resection of segments of four to six lower cartilages on each side. For those patients with asymmetrical deformities and prominent unilateral cartilaginous protrusion, resection of short segments of the abnormal cartilages on each side will often allow the sternum to depress to the desired level. A transverse osteotomy through the anterior table of the sternum is often helpful in lowering the sternum and correcting the tilt. Placement of a sternal support bar (Adkins strut)⁹ across the chest anterior to the sternum, and securing the bar to the sternum with absorbable sutures for 4 to 6 months, provides stability, minimizes pain, and prevents depression or recurrent sternal protrusion for almost all patients.¹ The sternal support bar eliminates postoperative flail chest and paradoxical respirations, which reduces pain, permitting early ambulation and deeper respiratory excursions, thus reducing the length and cost of hospitalization. The bar is easily removed and ensures good long-term results. For patients with bilateral severe costal and sternal protrusion after subperiosteal resection of medial and lateral segments of the deformed cartilages, a wedge of cartilage may be inserted into the anterior sternal osteotomy to depress the sternum to the desired position.

Costal cartilage regeneration from the perichondrial sheaths is remarkably rapid in children and adults, with the chest becoming very stable within 4 to 6 weeks. We prefer surgical repair in the mid adolescent years for those patients who are experiencing symptoms, and who have a low pectus severity index; the operation is technically easier in this age group than in adults.

Clinical experience

During the past 33 years, we have performed surgical correction of carinatum deformities in 115 patients (99 males and 16 females). Minimal deformity was observed in 102 of the 115 patients (89%) before the age of 10 years. In 11 patients, sternal protrusion was noted during the first 5 years of life; 10 of them had chondromanubrial deformities whereas only four of 102 patients with later onset of PC had this type of deformity. Protrusion of the mid and lower sternum was first noted after the age of 10 years in 97 patients. Fifty-six patients had asymmetrical protrusion deformities, with the left anterior chest usually being more prominent. The age at the time of operation ranged from 3 to 37 years (mean, 17.9 years). Chest radiographs from all patients showed an increase in the anteroposterior diameter of the chest, somewhat emphysematous lungs, and a narrow cardiac silhouette. The pectus severity index, determined by dividing the inner width of the chest at its widest point by the distance between the posterior surface of the sternum and anterior surface of the spine at the widest point, ranged from 1.32 to 1.98 (mean, 1.76). The mean pectus severity index in normal persons is 2.54.^{7,10}

The operative technique used for repair of the 115 PC patients was a modification of that described by Ravitch¹¹ and similar to that detailed in previous reports of PE.¹² The repair has varied, depending on the location of the sternal protrusion, the degree of asymmetry, and the severity of any associated depression, but includes the following essential features, detailed below.

Transverse curvilinear incision is made across the lower anterior chest midway between the nipples and the costal margins. Patients with more than four deformed costal cartilages benefit from a short cephalad extension of the incision in the midline (Figure 1).

Limited skin flaps are elevated over the pectoralis muscles using needlepoint electrocautery.

The pectoralis major muscles are reflected laterally just sufficient to expose the deformed costal cartilages.

The lowermost deformed costal cartilages are exposed by mobilizing the attached abdominal muscles.

Short cautery incisions (1.0–1.5 cm) are made through the perichondrium of deformed costal cartilages adjacent to the sternum and laterally near or beyond the costochondral junction, where the chest wall is at a near normal elevation.

Short segments of cartilage (1.0 cm) are resected medially and laterally from each of the deformed ribs using Freer elevators, carefully preserving the perichondrium.

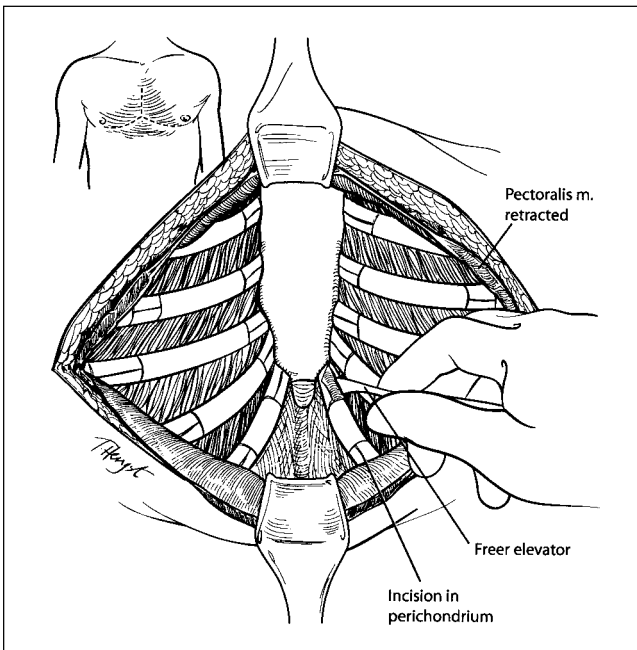


Figure 1. Pectoralis muscles are reflected laterally over a short distance, and abdominal muscles are mobilized inferiorly just sufficient to expose the deformed cartilages. Short incisions are made through the perichondrium on the medial and lateral ends of the deformed cartilages, and short segments are removed.

The xiphoid is detached from the lower sternum if it extends anteriorly or posteriorly.

A transverse osteotomy is made through the anterior table of the sternum at the level where it begins to protrude outward from the normal chest contour in patients with severe protrusion. The posterior table of the sternum is gently fractured but not displaced, lowering the sternum to the desired level. In eight patients with chondromanubrial prominence, it was necessary to make a second or third osteotomy across the anterior table of the sternum to flatten the localized protrusion.

For 25 patients with severe symmetrical protrusion, a small triangular wedge of costal cartilage was placed into the anterior sternal osteotomy and secured to the sternum with non-absorbable sutures to provide stability and haemostasis.

A thin stainless steel Adkins strut⁹ was placed across the lower anterior chest and secured to the appropriate rib on each side with fine wire for 83 of the 115 patients (Figure 2). The sternum was secured to the strut with absorbable sutures to prevent sternal depression and provide stability. For 14 patients with sternal twisting or combined upper protrusion and lower depression defects, the strut was placed posterior to the lower tip of the sternum, similar to that used for PE.

The xiphoid is reattached to the lower sternum.

Finely minced fragments of the patient's resected cartilage are placed into the perichondrial sheaths to enhance cartilage regeneration.

The pectoralis muscles are sutured together in the midline and the abdominal muscles are sutured to the pectoralis muscles across the lower anterior chest (Figure 3).

A small polyethylene suction catheter is placed into the space between the muscles and cartilaginous repair.

The skin is closed with subcuticular absorbable sutures and steristrips.

Thorough haemostasis was achieved with electrocautery and the wounds were copiously irrigated with antibiotic solution (cefazolin; Ancef, GlaxoSmithKline, Philadelphia, PA, USA) throughout the operation. Hemovac drains were removed within 4 days. Intravenous antibiotic (cefazolin; Ancef), was given for 3 days, and an oral antibiotic (cephalexin; Keflex, Eli Lilly and Company, Indianapolis, IN, USA) was given for a further 4 days. Postoperative pain was remarkably mild for all patients and was controlled using intravenous analgesics for the first 48 hours and oral non-narcotic medications thereafter. Analgesic medications were rarely used after 1 week. Epidural analgesia was not used for any of the patients.

The duration of the operation averaged 144 minutes (less than for PE deformities); the total period of hospitalization

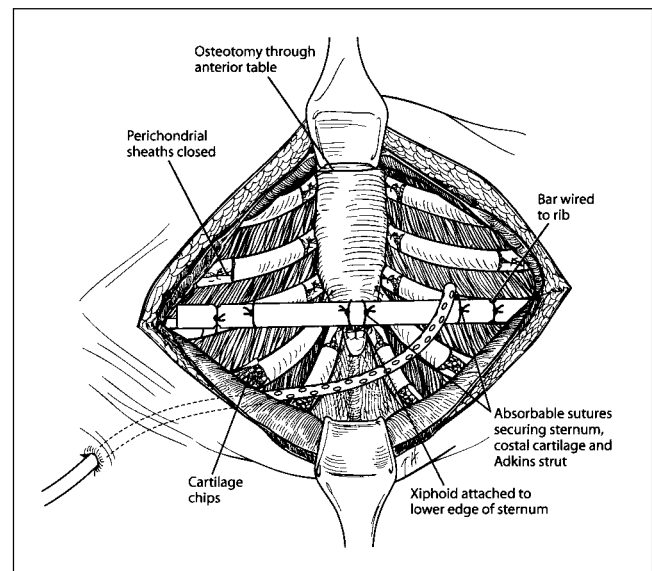


Figure 2. Finely minced fragments of autologous costal cartilage are placed into the open perichondrial sheaths. The perichondrial sheaths are closed loosely with absorbable sutures. An Adkins strut is placed across the lower anterior chest anterior to the sternum and attached to the appropriate rib on each side with fine wire. The sternum is secured to the strut with absorbable sutures to prevent sternal depression. The xiphoid is sutured back to the lower sternum. A Hemovac catheter is placed across the lower anterior chest.

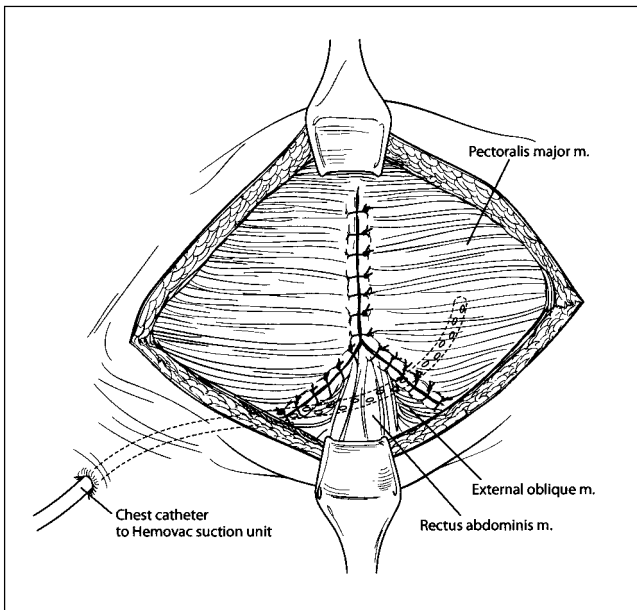


Figure 3. The pectoralis muscles are reapproximated in the midline with absorbable sutures. The abdominal muscles are attached to the pectoralis muscles across the lower anterior chest. The Hemovac catheter is positioned between the muscle closure and cartilaginous repair.

rarely exceeded 3 days (mean, 2.7 days). Mean blood loss was 78 mL and no patients received a blood transfusion. Full physical activity, except for body contact sports, was resumed by almost all patients within 8 weeks. The sternal struts were removed on an outpatient basis 5 to 6 months after repair through a 1.5 cm incision, with the patient under light general anaesthesia. Removal of the bar rarely took more than 15 minutes.

Results and comments

Each patient with preoperative severe shortness of breath or limitation in stamina and endurance experienced marked improvement within 3 to 4 months after surgery. Patients with asthmatic symptoms experienced fewer episodes of wheezing and a decreased need for medications. All patients with chest discomfort noted considerable improvement within 3 months. Each of 29 patients who underwent preoperative and postoperative measurement of vital capacity with an incentive spirometer experienced improvement within 6 months (mean improvement, 9%). There were no deaths within the first 5 years after surgery. Only three patients did not achieve a very

good or excellent long-term result during a mean follow-up of 9.2 years; two of these patients had other major anomalies, and the third had undergone attempted previous reconstruction. Postoperative complications included wound seroma (5), pleural effusion (4), unintentional pneumothorax (3), and recurrent mild protrusion of one or two lower or upper costal cartilages (6). Mild to moderate hypertrophy of the cutaneous scar occurred in 11 patients.

The described operative technique has provided excellent results in more than 97% of patients, with short hospitalization, a low rate of complications, and low cost. The high frequency of improvement in respiratory symptoms, exercise tolerance and endurance, as well as cosmetic appearance, supports the view that symptomatic patients with PC at all ages will benefit from surgical repair.

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