Repair of Pectus Chest Deformities in 320 Adult Patients: 21 Year Experience

Dawn E. Jaroszewski, MD, and Eric W. Fonkalsrud, MD

Department of Surgery and Division of Cardiothoracic Surgery, David Geffen School of Medicine at University of California–Los Angeles, Los Angeles, California

Background. **Severe pectus chest deformities are common, often causing physiologic impairment. Patients who do not undergo repair during childhood often experience progressive worsening of symptoms during adulthood. There are few published reports regarding pectus repair in adults.**

Methods. **A retrospective review from January 1986 through January 2007 was performed on patients age 19 years and older, who underwent surgical correction of pectus excavatum (PE) or carinatum (PC) at one hospital.**

Results. **Adult patients, including 268 PE (84%), 41 PC (13%), and 11 with combined deformities (3%) underwent open repair with minimal cartilage resection and a temporary internal support strut. Ages ranged from 19 to 67 years (mean, 27). Patients experienced dyspnea, decreased endurance and tachypnea with mild exertion (99%), tachycardia (94%), and chest pain (69%). All patients reported worsening of symptoms during adolescence, which became more severe during adulthood. The mean severity score for PC and PE was 1.8 and 5.8, respectively (normal = 2.5). All patients experienced improvement in symptoms within four months after repair. Mean hospitalization was 2.9 days. Complications included pleural effusion (n = 8), pneumothorax (n = 4), and pericarditis (n = 2). Small localized protrusions persisted in eight patients. Four patients underwent repair of mild recurrent deformities. There were no deaths. Ninety-eight percent reported considerable improvement in exercise tolerance and indicated postoperative results as very good or excellent.**

Conclusions. **Uncorrected pectus deformities persist after childhood and often cause worsening symptoms with increasing age. Repair can be performed in adults with low morbidity, short hospital stay, and considerable improvement in physiologic function.**

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Pectus chest deformities are among the most common major congenital anomalies, occurring in approximately 1:400 Caucasian male births [1]. Males are afflicted approximately five times more often than females [2, 3]. Pectus excavatum (PE) with sternal depression is approximately six times more common than pectus carinatum (PC) with sternal protrusion. Pectus deformities are believed to often have a genetic origin, with an accelerated growth of the costal cartilages compared with the remainder of the bony skeleton. Pectus excavatum is frequently recognized during early childhood, is often of mild severity, and it produces few symptoms. During rapid adolescent growth, most patients experience a considerable increase in the severity of the depression until full skeletal maturity is achieved. In contrast, pectus carinatum deformities are often unrecognized until adolescent skeletal growth occurs [4, 5]. Pectus carinatum consists of a spectrum of deformities, most frequently with anterior displacement of the mid and lower sternum and adjacent costal cartilages.

Severe PE and PC can both cause symptoms and physiologic limitations [6, 7]. With PC, the thorax is held in a partially expanded position with increased residual air and reduced vital capacity, somewhat analogous to that which occurs with emphysema. Pectus carinatum prevents complete expiration of air from the lungs and thus restricts gas exchange considerably, causing dyspnea, tachypnea, and reduced endurance with exercise. The symptoms are often much more severe than the external appearance would suggest.

The heart in PE patients is displaced into the left chest to varying degrees by the depressed sternum. Cardiac compression reduces the stroke volume and cardiac output in severe deformities, causing accelerated fatigue and compensatory tachycardia [8, 9]. Sternal compression commonly decreases the thoracic volume, which reduces the tidal volume and vital capacity, causing dyspnea and a decrease in endurance with compensatory tachypnea during exercise. Many experience discomfort in the lower anterior chest [10].

Although pectus deformities are recognized to cause both physiologic as well as social impairment [11, 12], many symptomatic patients do not undergo repair during childhood. Many of these patients experience progressive worsening of cardiopulmonary function with increasing age. Publications regarding the surgical management of adult patients with symptomatic pectus deformities have been sparse [13]. The present study reviews the experience from one hospital with

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Address correspondence to Dr Fonkalsrud, Department of Surgery, David Geffen School of Medicine at UCLA, Dept. of Surgery, 10833 Le Conte Avenue, Los Angeles, CA 90095-1749; e-mail: efonkalsrud@mednet.ucla.edu.

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Table 1. Age of Pectus Patients Undergoing Repair

Age Range	Number of Patients	% of Patients
19–24	149	47
25–30	78	24
31–39	47	15
40–49	29	9
Over 50	17	5
	320	100

320 adult pectus patients who have undergone surgical repair.

Patients and Methods

The medical records of all patients 19 years of age and older who underwent correction of PE and PC deformities at the UCLA Medical Center from January 1986 to January 2007 were retrospectively reviewed. Consent for the study was granted by the UCLA Medical Center Institutional Review Board; individual consent for this specific study was waived. Since the initial pectus repair in 1969, 1,120 patients have undergone repair of pectus chest deformities in our hospital by one surgeon (977 since 1986). Since 1986, 320 adults (33%) underwent surgical correction. No adults underwent pectus repair prior to 1986. There were 243 males (76%) and 77 females (24%). The ages ranged from 19 to 67 years (mean, 27 years) (Table 1). There were 268 with PE (84%), 41 with PC (13%), and 11 (3%) with a combination of PC and PE. Twenty-five patients were referred for repair of symptomatic recurrent deformities [14], two with a previous Nuss repair [15]. The pectus severity index (inner width of the chest divided by the distance between the sternum and the spine), as determined by computed tomographic (CT) scan or chest X-ray, indicates the severity of the deformity (normal chest = 2.54) [16]. For PE patients in the present study the index ranged from 3.3 to 20.2 (mean, 5.8). For PC patients the index ranged from 1.3 to 2.1 (mean, 1.8). Seventy-three percent of the adult patients were referred from states or countries outside California.

The most frequent symptoms were dyspnea with mild exercise in both PE and PC patients, with progressive loss of endurance during exercise in 99%. Moderate compression type discomfort in the lower anterior chest with activity was noted by 69% of PE patients. Many PC patients experienced pain when pressure was applied to the anterior chest. Tachycardia with occasional palpitations during exercise was present in 94% of all patients. Exercise-induced wheezing was reported by 19% of all patients. Sixteen percent of PC patients had asthmatic symptoms requiring bronchodilator medications; 9% of PE patients had asthmatic symptoms. Eighteen percent of all patients had an increased frequency of bronchitis compared with other persons their age. Many patients who experienced increasing respiratory symptoms during adulthood had a history of asthma, pneumonia, or other pulmonary disorders during early childhood. Mild to moderate dysphagia was reported by 14 PE patients.

Associated major congenital disorders were present in less than 4% of patients and included Marfan's syndrome (n = 2) and congenital heart defects (n = 3). Sixteen percent of PE patients had a functional heart murmur. Mild to moderate scoliosis of the thoracic spine was present on 72% of PE patients. Varying degrees of kyphosis was present on 68% of PC patients. Only five patients had other major medical disorders.

All patients had a preoperative chest radiograph or CT scan. Most PE patients had a narrow anterior to posterior diameter of the chest with varying degrees of displacement of the heart into the left chest (Fig 1). An increase in the anterior to posterior diameter of the chest with narrow cardiac silhouette was observed in PC patients. Almost all patients over the age of 25 years had preoperative cardiac evaluation including electrocardiogram and echocardiography, pulmonary function tests, and spirometry. Right ventricular strain was noted on 61% of the PE patients studied. Fourteen percent had documented mitral valve prolapse. Sixteen patients had a diastolic filling pattern suggesting constrictive dynamics. Fifty-four percent of the tested patients showed mild to moderate restrictive pulmonary function.

The decision to repair the pectus deformity was based on the severity of symptoms and the severity of the deformity, verified by the pectus severity index. The major indication for operation was for physiologic improvement. All patients underwent variations of a highly modified Ravitch repair, with progressive reduction in the amount of cartilage resected over the 21-year period [5, 17]. The evolution of the operative technique used for both PE and PC during the past 21 years has been published in detail previously [5, 17, 18] and is here only briefly described as it is currently performed. During the past three years, we have resected only short segments of costal cartilages medially and laterally and then reattached the cartilages to the sternum after it had been elevated or lowered to the desired position. For the first 140 PE patients the right pleural space was routinely

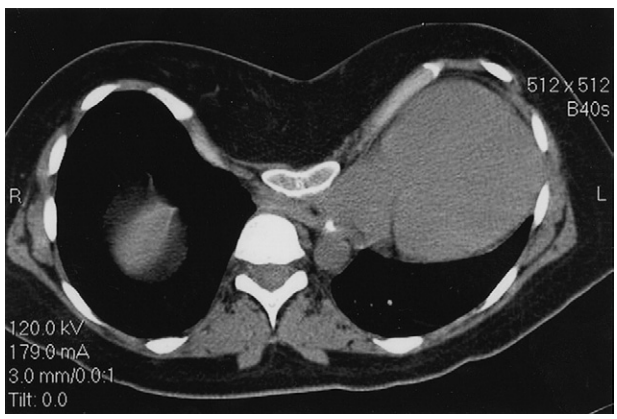

Fig 1. Computed tomographic scan from 51-year-old female with very severe pectus excavatum (severity index = 20.2). Note severe displacement of heart into left chest with compression of left lung.

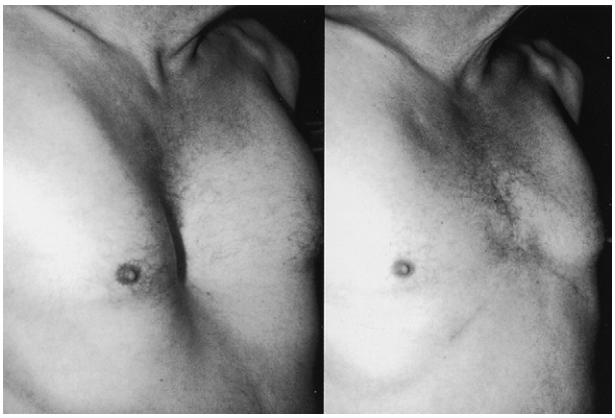

Fig 2. (A) A 59-year-old male with severe pectus excavatum (severity index = 5.2). (B) Appearance of chest 26 months after repair.

opened and the support strut was placed posterior to the sternum and the cartilage repair. All subsequent PE patients and all PC patients, as well as 22 of the 25 with recurrent deformities, had an anterior support strut with a large wire holding the sternum to the strut and did not require opening of either pleural space.

Under general endotrachael anesthesia, a transverse chevron incision was made with a short midline extension superiorly in almost all patients depending on the number of deformed cartilages. Short skin flaps were elevated; the pectoralis muscles were reflected laterally and the abdominal muscles were mobilized sufficient to expose the deformed lower costal cartilages. Subperiosteal resection of short segments of cartilage was performed adjacent to the sternum and laterally near or beyond the costochondrial junction where the chest wall was at the highest level. Five of the seven costal cartilages attached to the sternum on each side were reconstructed in 282 patients, six in 31 patients, and four in seven patients. The retrosternal space for both PE and PC patients was mobilized over a short distance. An attempt was made to avoid entering either pleural space in the last 180 patients. The pericardium was not entered in any patients. A transverse wedge osteotomy was made across the anterior table of the sternum at the level where the sternum depressed posteriorly for PE patients. The posterior table of the sternum was gently fractured at the osteotomy without detachment and the lower sternum was elevated, and twisted when asymmetric, to the desired level. For PC patients, a simple transverse osteotomy was made across the anterior sternum at the appropriate level and the distal sternum was lowered to the desired position. The costal cartilages were shortened sufficient to permit reattachment to the sternum medially and to the ribs laterally without pressure on either end. A thin stainless steel Adkins strut (Baxter Healthcare Corp, McGraw Park, IL) with a slight convex curvature was placed across the lower anterior chest and secured to the sternum with a heavy wire for both PE and PC patients to stabilize the chest. The pectoralis and abdominal muscles were reconstructed to completely cover the cartilaginous repair. A hemovac drain was placed between the cartilage and muscle repairs. The subcutaneous tissues and skin were closed with absorbable sutures.

Postoperative care followed a clinical pathway program that includes three days of intravenous cephazolin given perioperatively, and oral cephalexin for an additional four days. Pain was mild to moderate for almost all patients using patient-controlled analgesic with morphine during the first 48 hours. Subsequently, oral nonnarcotic medications were adequate. None of the patients received an epidural analgesic. The mean duration of the operation was 191 minutes (±26); however, it was slightly longer in patients undergoing repair of recurrent deformities. Only three patients were hospitalized for more than three days, with a mean hospitalization of 2.9 days. The sternal support strut was removed a mean of seven months after repair on an outpatient basis under light general anesthesia and rarely took more than 20 minutes. There were no complications after strut removal.

Results

Follow-up on all patients undergoing pectus repair in the present study was performed by office visit, telephone, or email questionnaire from one month to 21 years after sternal strut removal (mean, 26 months). Reporting of symptoms before and improvements after repair was subjective and reliant on patients' response to questions.

Postoperative complications included transient pleural effusion (n = 8) and pneumothorax (n = 4), one of which required aspiration, all in the first 143 patients who had a retrosternal support strut. Transient pericarditis of undetermined etiology (n = 2) resolved in both with a short course of indomethicin. One patient experienced intraoperative transient ventricular fibrillation of undetermined etiology, which reverted to normal sinus rhythm with one external shock. Four patients (three with PE and one with PC) underwent repair of mild recurrent deformities within three years after the primary repair, all performed before the minimal cartilage resection technique was used. Two of the six patients repaired after the age of 60 years experienced a moderate recurrence, with the severity index showing a mean of only 62% improvement compared with preoperative values. Small localized

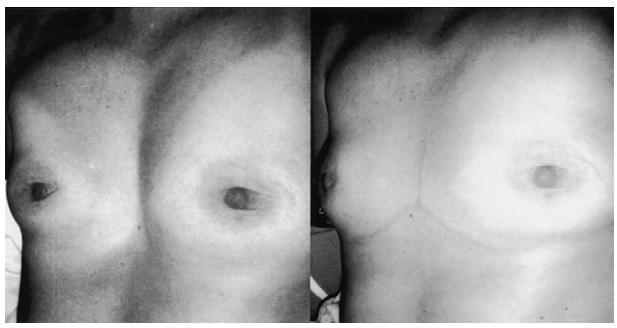

Fig 3. (A) A 46-year-old female with severe pectus excavatum (severity index = 7.2). (B) Appearance nine months after repair and three months after bar removal.

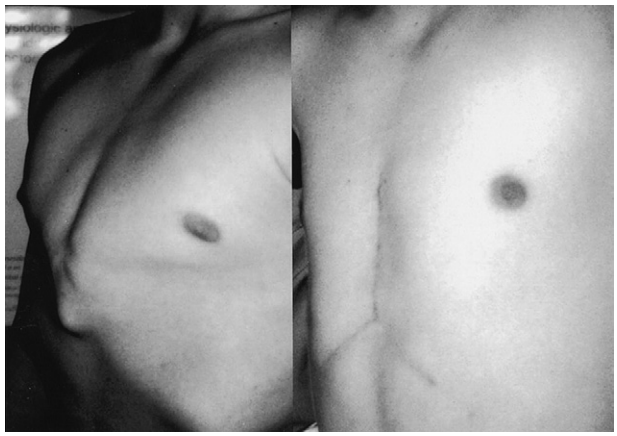

Fig 4. (A) A 23-year-old male with severe pectus carinatum (severity index = 1.72). (B) Appearance two months after removal of sternal support bar.

protrusions, usually of the lowest, or the second costal cartilage, were corrected at the time of outpatient sternal bar removal for eight patients. Mild to moderate hypertrophy of the cutaneous scar occurred in 22 patients. There were no perioperative deaths. Ninety-two percent of patients returned to work within two weeks and gradually increased both anaerobic and aerobic exercises over the ensuing weeks. All patients returned to unrestricted physical activities after removal of the sternal strut.

Although objective measurements of physiologic improvement after operation are not available for all patients, all but three of the PE patients, each with repair of recurrent deformities, showed shifting of the heart from the left chest toward the mid mediastinum on chest radiographs within the first three months. For 38 consecutive PE patients who underwent preoperative and postoperative measurement of vital capacity with an incentive spirometer, a mean improvement of 10.6% was noted within two months after sternal strut removal. Within four months after PE repair, functional heart murmurs were no longer audible in 29 of 42 patients with preoperative murmurs. Slightly over 98% of patients considered the result after repair to be very good or excellent and that they would highly recommend pectus repair to other patients (Figs 2; 3; 4). Moderate to marked improvement in exercise tolerance with much less dyspnea and increased endurance was reported within four months after PE or PC repair by all but six patients, three of whom had repair of recurrent deformities. Preoperative chest pain was resolved, or considerably improved, in all but two of the 221 patients after repair.

Comment

Increasing evidence indicates that both severe PE and PC deformities cause physiologic limitations in exercise tolerance, and are not merely cosmetic disorders [6, 8, 9, 19–21]. The extent of cardiopulmonary impairment and the amount of physiologic improvement achieved after surgical repair is currently the subject of several studies [6, 21, 22]. Symptomatic pectus patients frequently use wider diaphragmatic excursions to compensate for diminished chest wall expansion and contraction during respiration. Symptoms from pectus deformities are uncommon during early childhood, and repair in preadolescent children is associated with a higher recurrence rate and may adversely affect costal cartilage growth [23]. During the adolescent years of rapid growth, the majority of PE and PC deformities become increasingly severe and symptomatic until full skeletal growth has been achieved. Few further thoracic skeletal changes occur throughout adult life. Although the anatomic deformities rarely become more severe during adult years, symptoms from both PE and PC often gradually worsen with the aging of both the heart and lungs, which often makes compensatory mechanisms to alleviate physical limitations less successful. Dyspnea with mild exercise and decreased endurance are experienced by some adult patients with only moderately severe deformities, with PE severity scores occasionally as low as 3.3. Some patients describe a compression type discomfort in the anterior chest that on rare occasions may simulate angina.

Reports of favorable clinical experience with the minimally invasive repair of PE (MIRPE) have included primarily young children [15]. During recent years the MIRPE has been extended to a few older patients including adults, although the great force necessary to elevate the sternum and the complications and postoperative pain are higher in adults [24, 25]. Repair of PE and PC using the open technique with minimal cartilage resection (as described in the present report) is slightly more difficult technically than the MIRPE, particularly because the costal cartilages in adults are often brittle and partially ossified; however, this technique produces more consistently good results, with lower morbidity, less pain, shorter hospitalization, and more rapid return to full activity [24]. Resection of the entire deformed costal cartilages, as in the Ravitch repair, may damage the periosteum, often results in ossified rigid regenerated cartilage, and is unnecessary to achieve optimal results.

Pulmonary function studies to accurately document the severity of physical limitations caused by PE and PC should be performed during exercise, which was not the case for more than 60% of patients in the present study. Dynamic magnetic resonance imaging studies of the heart, chest wall, diaphragms, and lungs may be more definitive in identifying physiologic changes in dynamics caused by pectus deformities, as well as improvement after repair. An extensive three-year study of approximately 40 PE patients using these techniques before and after repair is near completion in our hospital. The significance of cardiac compression and displacement on older patients is speculative, although stroke volume and cardiac output have been shown to be reduced [26–28]. Chest radiographs and CT scans show that cardiac compression is nearly always reduced considerably by PE repair. Preoperative mitral valve prolapse in 16% of PE patients persisted in less than half after repair [17]. Most symptoms caused by PE and PC can be improved con-

siderably after surgical repair. The technique for repair using minimal cartilage resection and temporary support with a temporary internal support strut has provided better results than when more extensive cartilage resection was used over 15 years ago. Data from the present clinical experience indicate that many patients who do not undergo repair of severe PE and PC deformities in childhood will experience increasing symptoms throughout adult life.

There are sparse published data regarding indications for, and results after, operations for symptomatic adult patients with PE or PC. Data from the present clinical experience indicate that many adults with pectus deformities will experience a progression in severity of symptoms as they age. For those adults repair can be performed with low morbidity, low cost, short limitation of activity, and a high frequency of symptomatic improvement. Although the optimal age for correction of pectus deformities is during adolescent years when the operation is technically easier, repair of persistent deformities in symptomatic adult patients should be considered as a recommended treatment option.

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