Chest wall anomalies: pectus excavatum and pectus carinatum

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In 1996, Sydney S. Gellis, MD, editor of the *Pediatric Notes* wrote, "It has been evident for a long time that far too many children have had surgery for pectus excavatum" [1]. For many years, controversy has existed as to the surgical necessity for repair, because the perception was that pectus excavatum was purely a cosmetic defect. With recent modifications of the surgical procedure and a better understanding of the physical and psychological perceptions of this condition, however, surgery can have distinct and lifelong benefits.

Congenital chest wall anomalies fall into two groups: those with overgrowth of the rib cartilages causing either a depression or protuberance, and those with varying degrees of either aplasia or dysplasia. In the authors' patient population, pectus excavatum, a chest wall depression, accounts for more than 87% of the deformities and is by far the most common chest wall deformity (Table 1). As its name suggests, it presents with an "excavated, sunken or funnel chest." Pectus carinatum, a chest wall protuberance, constitutes approximately 5% of chest wall deformities. Combined excavatum/carinatum deformities constitute 6.1% of chest wall anomalies.

Chest wall deformities frequently are associated with systemic weakness of the connective tissues and with poor muscular development of the abdominal region, thorax, and spine. There is, therefore, a markedly increased association with Marfan's syndrome, Ehlers-Danlos syndrome, and scoliosis as well as with

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Table 1
Incidence and etiology

	Number of patients
Total evaluated	1124
Total with pectus excavatum only	982 (87.3%)
Total with mixed pectus excavatum/pectus carinatum	69 (6.1%)
Total with pectus carinatum only	51 (5.0%)
Total with Poland syndrome	8 (0.7%)
Total with other deformities	7 (0.6%)
Surgical patients	557
Total with pectus excavatum only	550 (99%)
Total with mixed pectus excavatum/pectus carinatum	7 (1.2%)
Total with Poland syndrome	3 (0.5%)
Total with other deformities	1 (0.2%)
Male to female ratio	4:1
Family history of pectus excavatum	45%
Incidence of scoliosis	29.3%
Incidence of Marfans-diagnosed	3.1%
Marfanoid (presumed Marfans)	19%
Patients with Ehlers-Danlos	2.1%

omphalocele in the case of bifid sternum, all of which complicate the management of these patients (see Table 1).

Surgical management has undergone major changes over the last 15 years. In the 1960s and 1970s, radical surgical operations were in vogue, even in very young children. It came to be realized, however, that pulmonary function actually decreased over time because of the scarring of the anterior chest wall, and some patients developed acquired asphyxiating chondrodystrophy from resections that were too extensive and performed at too early an age. As a result, surgeons stopped operating on prepubertal patients and reverted to modified resections of the deformed cartilages. Recently, a minimally invasive procedure with no resection, only internal bracing, has been introduced.

Pectus excavatum

Introduction

Pectus excavatum is a depression of the anterior chest wall of variable severity that may be mild, moderate, or severe. All variations of depth, symmetry, and breadth of the deformity may be seen. The deformities may be small in diameter and deep, cup-shaped or of large diameter and shallow, saucer-shaped, or eccentric. The depth and extent of the depression determine the degree of cardiac and pulmonary compression, which in turn determines the degree of incapacitation. The deformity frequently is noted at birth and progresses with growth.

Progression may be especially pronounced during puberty, a fact unknown to many pediatricians who mistakenly advise younger patients that the condition

will resolve spontaneously. The authors have seen many families that received this inaccurate advice, and whose children missed the opportunity to have the deformity repaired before puberty, when the chest was still soft and malleable, and before it interfered with physical performance. Approximately one third of patients have a deformity thought severe enough to require operative correction.

History

Pectus excavatum was recognized as early as the 16th century. In 1882, a report of five cases by Ebstein [2] covered the clinical spectrum of the condition. Treatment at that time was limited to "fresh air, breathing exercises, aerobic activities, and lateral pressure" [3,4].

Thoracic surgery remained forbidden territory until the early years of the 20th century. At that time, surgical correction consisted of several modifications of costal cartilage resection with sternal osteotomies, with or without the use of support bars. In the 1920s, Sauerbruch performed the first pectus repair that used the bilateral costal cartilage resection and sternal osteotomy technique [5] later popularized by Ravitch. He also advocated external traction to hold the sternum in its corrected position for 6 weeks postoperatively.

In 1958, Welch [6] advocated a less radical approach than Ravitch. He produced excellent results in 75 cases without cutting through all the intercostal bundles or through the rectus muscle attachments. Haller [7] drew attention to the risk of "acquired asphyxiating chondrodystrophy" in his paper entitled "Chest Wall Constriction After too Extensive and too Early Operations for Pectus Excavatum." As a result, most surgeons stopped performing open pectus repair in young children and waited until after puberty. They also decreased the amount of cartilage resected and wrote about a modified Ravitch procedure.

In 1997, Nuss et al [8,9] published their 10-year experience with a minimally invasive technique that required no cartilage incision, no resection, and no sternal osteotomy, but instead, relied on internal bracing made possible by the flexibility and malleability of the costal cartilages. The rationale for this technique was based on the following three observations:

1. Malleability of the chest. Children have a soft and malleable chest. In young children, the chest is so soft that even minor respiratory obstruction can cause severe sternal retraction. Trauma rarely causes rib fractures and flail chest in children because "the chest is so soft and malleable" [10–12]. The American Heart Association recommends "using only two fingers" when performing cardiac resuscitation in young children and "only one hand in older children" for fear of crushing the heart.
2. Chest reconfiguration. Even middle-aged and older adults develop a barrel-shaped chest configuration in response to chronic obstructive respiratory diseases such as emphysema. If older adults are able to reconfigure the chest wall, children and teenagers should be able to do the same, given the increased malleability of their anterior chest wall.

3. Bracing. The role of braces and serial casting in successfully correcting skeletal anomalies such as scoliosis, clubfoot, and maxillomandibular mal-occlusion by orthopedic and orthodontic surgeons is well established. The anterior chest wall, being even more malleable than the previously mentioned skeletal structures, is therefore ideal for this type of correction.

Incidence and etiology

Pectus excavatum occurs in approximately 1 in 1000 children and constitutes more than 87% of all the chest wall deformities (see Table 1). This is not the case in all countries, however. In Argentina, pectus carinatum is more common than is excavatum (M. Martinez-Fero, personal communication, 2001). Pectus excavatum is also very rare in Africa; to date, the authors have seen only 10 African-American patients out of more than 1000 patients with pectus excavatum. A family history of pectus excavatum was present in 45% of patients (see Table 1). The mode of inheritance, however, remains unclear. The authors have seen families with three siblings and cousins and other family members who have had a pectus deformity severe enough to require surgery. The male-to-female ratio is four to one in the authors' series of pectus excavatum patients; this is similar to that of other large series [13]. Males have an increased risk of this deformity, whereas females have an increased risk of associated scoliosis.

The association with connective tissue disorder is higher than in the normal population. A definitive diagnosis of Marfan's syndrome was present in 3.1% of the authors' patients, and an additional 19% had clinical features suggestive of Marfan's syndrome. Scoliosis was identified in 29% of the patients. Because severely asymmetric pectus excavatum tends to aggravate the postural abnormality of scoliosis, early correction of the pectus excavatum has improved mild scoliosis in some patients. Ehlers-Danlos syndrome was present in another 2.1% of patients. The vast majority of the authors' patients had an asthenic build and resembled basketball players; very few had an endomorphic or mesomorphic build.

Clinical features

Pectus excavatum most frequently is noted in infancy [13] and usually progresses slowly as the child grows. Most young children are asymptomatic, because young children have significant cardiac and pulmonary reserves. Additionally, a child's chest wall is very pliable. As children mature, however, the deformities become more severe, and the chest wall develops increased rigidity. They find that they have difficulty keeping up with their peers when playing aerobic sports. A vicious cycle may develop, because patients stop participating in aerobic activities secondary to inability to keep up. This results in a further decrease in their exercise capacity. Furthermore, these patients, already embarrassed by their deformity, will avoid situations where they have to take their shirt off in front of other children, again contributing to less participation in school and team activities.

Some patients may exhibit depression by withdrawing from activities with their peers and a declining quality of schoolwork. Most pectus patients have a typical geriatric or "pectus posture" that includes thoracic kyphosis, forward sloping shoulders, and a protuberant abdomen. A sedentary lifestyle may aggravate this posture, and the poor posture depresses the sternum even further. For this reason, the authors recommend an aggressive pectus posture exercise and breathing program.

Many patients have a relatively mild deformity during childhood. There is significant potential for marked progression of the deformity with growth, because the deformity rarely resolves spontaneously. Although the deformity may not always deepen, it is unlikely that it will resolve spontaneously. When the patients grow rapidly during puberty, the deformity often suddenly accelerates, and a mild deformity may become severe in as little as 6 to 12 months. These patients present with a history that "my chest suddenly caved in." It is the rapid progression that alarms parents and induces them to seek surgical consultation despite their pediatrician's reassurance. Patients with a rapid progression of their deformity exhibit the most pronounced symptom complex.

The earliest complaints include shortness of breath and lack of endurance with exercise. As the deformity progresses, chest pain and palpitations with activity may occur, giving rise to exercise intolerance. Other symptoms include frequent and prolonged respiratory tract infections, which may lead to the development of asthma (Table 2).

A recent study by Lawson [14] showed that patients suffer from poor body image, which has a major impact on self-worth. Therefore, it is important to correct the deformity before it affects their ability to function normally. One of the authors' patients, a 35-year-old lawyer, confided that he had not married, because he was too ashamed of his chest abnormality to have a serious relationship. A 16-year-old patient left a note for his parents before attempting suicide detailing the harassment and abuse that he had received from children

Table 2
Presenting symptoms of 557 surgical patients

Shortness of breath, lack of endurance, exercise intolerance	92.6% (516)
Chest pain, with or without exercise	71% (395)
Frequent respiratory infections	34.3% (191)
Asthma/asthma-like symptoms	34% (190)
Cardiology indicators	
Cardiac compression (by CT, echo)	85% (473/557)
Cardiac displacement (by CT, echo)	73.4% (409/557)
Murmur on exam	26% (142/543)
Mitral valve prolapse	15% (81/543)
Other anomalies (BBB, aortic insufficiency, regurgitation, hypertrophy, malformations)	16% (85/543)
Pulmonary indicators	
FVC below 80%	26.3% (116)
FEV1% below 80%	32.1% (142)
FEF$_{25-75}$% below 80%	52.1% (230)

at school. Just as one would not consider leaving a child with a cleft lip untreated, one should not leave a child with a severe pectus untreated. Both have a physiological and psychological impact on the patient.

Cardiac and pulmonary effects

Much has been written about cardiopulmonary function in patients with pectus excavatum [15]. Some authors have shown significant compromise of cardiac or pulmonary function [16,17], whereas others have been unable to demonstrate any change [18]. Several factors play a role when testing cardiopulmonary function. These include the severity of the deformity, the inherent physical fitness of the individual patient, the patient's age, associated conditions, whether the tests are done supine or erect, and whether they are done at rest or during exercise.

Cardiac effects fall into three categories: decreased cardiac output, mitral valve prolapse, and arrhythmias (see Table 2). Compression of the heart results in incomplete filling and decreased stroke volume, which in turn result in decreased cardiac output [16,17]. Secondly, the compression interferes with normal valve function. Mitral valve prolapse was present in 15% of the authors' patients and in up to 65% of other published series [19,20], compared with only 1% in the normal pediatric population [21]. The third effect on the heart is arrhythmia including first degree heart block, right bundle branch block, Wolff-Parkinson White syndrome [22], and so forth, which were present in 16% of the authors' patients.

Pulmonary effects also fall into three categories: restrictive lung disease caused by decreased intrathoracic capacity, atelectasis caused by cardiac displacement causing left lung compression, and paradoxical respiration in severe cases caused by disturbed mechanical forces. The result is restrictive lung disease, pulmonary atelectasis, frequent and prolonged respiratory infections, and even the development of asthma (see Table 2). Children with a severe deformity from birth tend to compensate by increasing the diaphragmatic component of their respiration. This partly compensates for their deformity and is seen in patients who, despite a severe deformity, are able to achieve low-to-normal pulmonary function studies at rest. They demonstrate a lack of endurance during exercise, however, because their compensation is finite. Stress testing has shown an increase in oxygen consumption for a given exercise when compared with normal patients [23]. These results indicate that the work of breathing is increased and explains why patients lack endurance.

Evaluation and indication for operation

A complete history is taken, and a physical examination is performed for all patients and includes documenting photographs. Patients who have a mild-to-moderate deformity are treated with a posture and exercise program in an attempt to halt the progression, and they are followed at 6-month intervals (Fig. 1).

Patients who have a severe deformity or who have a documented progression also are treated with the exercise and posture program. In addition, they undergo
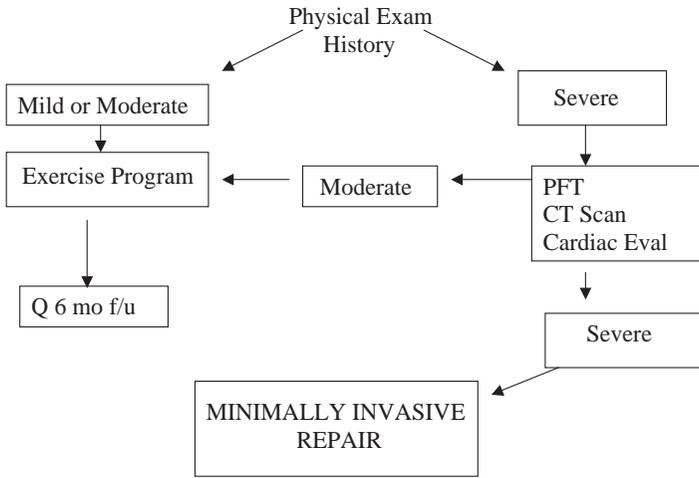
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                        Physical Exam
                           History

    Mild or Moderate                         Severe

    Exercise Program  ◄──  Moderate   ◄──   PFT
                                            CT Scan
                                            Cardiac Eval

       Q 6 mo f/u                              Severe

              MINIMALLY INVASIVE
                   REPAIR
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Fig. 1. Evaluation and treatment pathway.

objective studies to see whether their condition is severe enough to warrant surgical correction. These studies include a thoracic CT scan, pulmonary function tests, and a cardiac evaluation that includes an EKG and an echocardiogram (see Table 2, Fig. 1).

CT scans are very helpful, because they clearly show the degree of cardiac compression and displacement, the degree of pulmonary compression and atelectasis, asymmetry of the chest, sternal torsion, compensatory development of a barrel chest deformity in long-standing deformities, and ossification of the cartilages in patients with previous repairs. They also are used to calculate the Haller CT index, which gives an objective measurement for comparing the severity between different patients. The CT index is calculated by dividing the transverse diameter by the antero–posterior diameter [24] (Fig. 2).
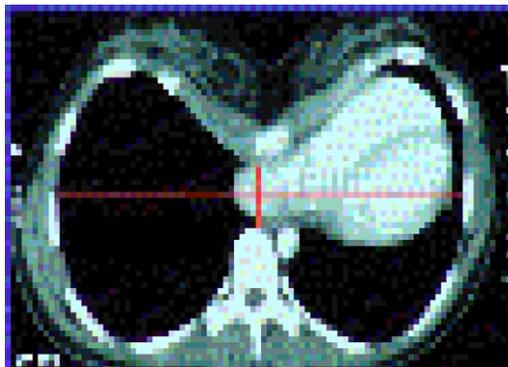

Fig. 2. Chest CT.

Pulmonary function tests are performed in all patients old enough to cooperate with testing. They are done best while exercising, but are also helpful in the resting state (see Table 2). The cardiology evaluation includes an EKG, an echocardiogram, and an examination by a pediatric cardiologist to determine the presence of cardiac compression, murmurs, mitral valve prolapse, conduction abnormalities, or other structural abnormalities (see Table 2).

Determination of a severe pectus excavatum and the need for repair include two or more of the following criteria:

- A Haller CT index greater than 3.25
- Pulmonary function studies that indicate restrictive or obstructive airway disease
- A cardiology evaluation where the compression is causing murmurs, mitral valve prolapse, cardiac displacement. or conduction abnormalities on the echocardiogram or EKG tracings
- Documentation of progression of the deformity with associated physical symptoms other than isolated concerns of body image
- A failed Ravitch procedure
- A failed minimally invasive procedure

When using these criteria, approximately 50% of patients are found to have a deformity severe enough to warrant surgery [8,9,25].

The age parameters for surgical correction depend on the type of procedure selected. Unlike the more invasive procedures (eg, Ravitch procedure or sternal turnover), there is no interference with growth plates when using the minimally invasive procedure [7,26]. Therefore, it can be done at any age, as evidenced by the fact that the authors have operated on patients from 21 months to 29 years of age successfully (Fig. 3). The concern with patients younger than 6 years,
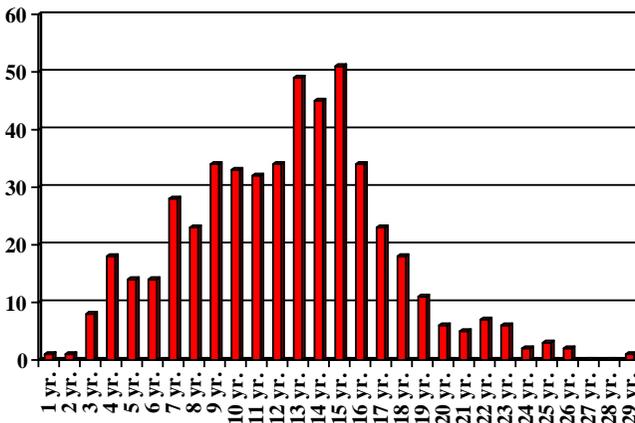
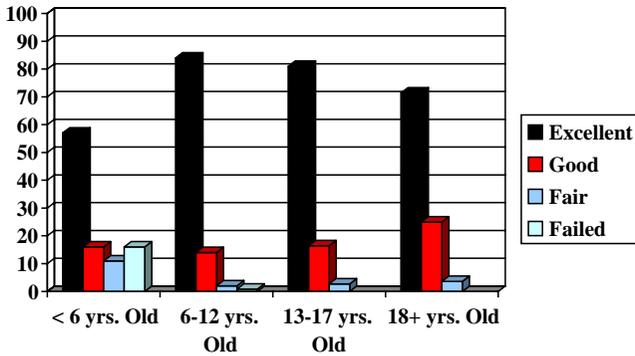

Fig. 3. Number of primary operations by age.

Fig. 4. Long-term results by age at time of surgery.

however, is that if the procedure is done at too young an age, there are many years of subsequent growth during which the pectus excavatum may recur.

The authors' experience has shown that the optimal age is 7 to 14 years, because, before puberty, the patients' chests are still soft and malleable; they show quick recovery, a rapid return to normal activities, and have excellent results (Fig. 4). After puberty, the flexibility of the chest wall is decreased, requiring the insertion of two bars, making the procedure more difficult. It also takes the patients longer to recover. All of the authors' patients over 20 years of age, however, have been extremely pleased with their results. Several other university centers have reported success with patients up to 44 years of age [27,28] (P. Colombani, MD, unpublished data, 2003) (see Fig. 4).

Results

Demographics

The minimally invasive technique for the repair of pectus excavatum received rapid acceptance by the surgical community, because the technique requires neither rib incision nor resection, no sternal osteotomy and has minimal blood loss, short operating time, and rapid return to regular activity [28–35].

Although the initial paper by Nuss et al presented a 10-year experience [8], the numbers were limited (42 patients). Additionally, the long-term results were affected by the early learning experience of using a support bar that was too soft, and in some patients, the bar was removed too soon. The authors' experience, as of June 30, 2003, encompassed 557 patients who had their primary operation at the authors' institution, with 298 patients postpectus bar removal (Table 3). Since the original presentation, numerous modifications have been made to the surgical technique (eg, routine use of thoracoscopy) and to the instruments to minimize the risks of the procedure and to facilitate insertion and stabilization of the substernal support bar. These have reduced the risks and complications markedly and have been documented in recent publications [25].

Table 3
Minimally invasive technique materials and methods

1124 patients were evaluated for chest wall deformity.		
608 patients had minimally invasive pectus repair.		
557 patients underwent primary operations.		
88 patients evaluated had prior pectus repairs.		
51 have had redo operations		
28 failed Ravitch procedures		
23 failed Nuss procedures[a]		
2 failed Leonard procedures[b]		
298 patients have had their bar(s) removed		
Minimally invasive technique operation and length of stay		
Single pectus bar:	84%	(466/557)
Double pectus bar:	16%	(90/557)
No stabilizers	20.1%	(112/557)
Stabilizers	80%	(445/557)
- Wired	83%	(369/445)
- Not wired	17%	(76/445)
Median age	13.3 years	(21 months–29years)
Median Haller CT index	4.8	(2.4–21)
Epidural:	3 days	(2–5)
Length of stay:	5 days	(3–10)

[a] 20 done elsewhere.
[b] 1 patient was a prior Ravitch; the other was a prior Nuss. Only counted once.

In the 16-year period from 1987 through June 2003, 1124 patients were evaluated for chest wall deformities, and 608 were judged to be severe enough to require surgery (see Table 3). Of these, 557 had their initial minimally invasive procedure done at the authors' facility, and 51 had redo operations.

Of these 557 patients, 550 (99%) had pectus excavatum, and seven (1.2%) had mixed pectus excavatum and carinatum. Three patients (0.5%) had associated Poland syndrome, and one (0.2%) had associated complex cardiac anomalies. Marfan's syndrome was confirmed or suspected in 123 patients (22%) and Ehlers-Danlos syndrome was noted in 12 patients (2.1%; see Table 1). The male-to-female ratio in patients undergoing repair was four to one (see Table 1). The median age was 13.3 years, with a range from 21 months to 29 years (see Table 3). Preoperative evaluation included CT scans in 516 patients with a median Haller CT index of 4.8 (range: 2.4 to 21). Cardiac compression was noted on echocardiography or CT scan in 473 of 557 patients (85%). Mitral valve prolapse was noted in 81 (15%) patients. Resting pulmonary function testing (PFT) was completed in 443 patients and demonstrated abnormalities as measured by FEF_{25-75} in 52% of the patients (see Table 2).

Operative procedure, analgesia, and length of stay

In 466 patients (84%), a single bar was inserted (see Table 3). Two bars were inserted in 90 patients (16%). Blood loss in most patients was minimal in the range of 10 cc, with the exception of one patient who developed a hemothorax.

Epidural analgesia was used for 3 days in most patients. The median hospital length of stay (LOS) was 5 days (range 3 to 10 days).

Complications

Early complications

As shown in Table 4, there were no deaths, nor were there any cardiac perforations during the 557 repairs. Postoperative pneumothoraces required chest tube drainage in 2.7% of the repairs and percutaneous aspiration in 0.5% of patients. Hemothorax requiring drainage occurred after one (0.2%) repair. Less than 1% of patients had pleural effusions treated by chest tube or aspiration.

Pericarditis requiring treatment with indomethacin occurred following six repairs (1.1%), with only one requiring pericardiocentesis. Pneumonia occurred after six repairs (1.1%), and medication reactions occurred following 24 repairs (4.3%). Wound infection occurred after four repairs (0.7%), resulting in bar infection and eventual early bar removal in three patients (0.5%). One hundred fifty-nine patients had a transient Horner's syndrome at varying times during the thoracic epidural administration.

Late complications

There were 66 (12%) bar displacements, and two-thirds of these patients required repositioning (see Table 4). Of these 47 displacements requiring revision, 15 occurred before stabilizers were available, a time period covering the

Table 4
Early postoperative complications

Deaths	0	
Cardiac perforations	0	
Pneumothorax	327	(59%)
Chest tube	15	(2.7%)
Aspiration	3	(0.5%)
Hemothorax	1	(0.2%)
Pleural effusion requiring drainage	4	(0.7%)
Pericarditis	6	(1.1%)
Wound/bar infection	4/3	(0.7)/(0.5%)
Pneumonia	6	(1.1%)
Medication reactions	24	(4.3%)
Transient Horner's	159	
Late complications		
Bar displacement	66/557	(12%)
Requiring revision	47/557	(8.4%)
Prior to stabilizer	15/112	(14.9%)
With stabilizer	32/445	(7.2%)
With wired stabilizer	10/445	(2.2%)
Hemothorax (post-traumatic)	2	(0.4%)
Bar allergy	3	(0.5%)
Overcorrection	27/557	(4.8%)
Skin erosion	1	(0.2%)

authors' first 112 repairs. After the introduction of stabilizers, the incidence of bar displacement dropped from 14.9% to 7.2% of patients. When the bar and stabilizers were wired together, the incidence of bar displacement dropped to 2.2% of patients.

Two patients developed late hemothorax secondary to trauma. Both underwent thoracoscopy with drainage of the hemothorax. No active bleeding was found, and the presumed etiology was injury to an intercostal vessel. Whether these patients would have developed hemothoraces as a result of their thoracic trauma if they had not had a pectus bar in situ is unknown. Several patients who were involved in major automobile accidents sustained head and musculoskeletal trauma but no chest injuries.

Three of 557 patients (0.5%) had unsuspected allergies to the metal in the bars. These presented as rashes in the area of the bar or stabilizer and required revision to custom-made bars of other alloys. Of 557 patients, 27 (4.8%) developed a mild overcorrection of their deformity, and four (0.7%) developed a true carinatum deformity. Of the patients who developed a true carinatum deformity, three had Marfan's syndrome or features consistent with Marfan's syndrome, and the fourth had symptoms of Ehlers-Danlos syndrome. No patient has developed thoracic chondrodystrophy.

Overall results and long-term follow-up

Patients were followed at 6 months postoperatively, then yearly. Long-term assessments classified the results into excellent, good, fair, or failed categories (Figs. 5, 6).

An excellent repair is achieved when the patient experiences total repair of the pectus and resolution of associated symptoms. A good repair is distinguished by a markedly improved but not totally normal chest wall appearance and resolution of associated symptoms. A fair result indicates a mild residual pectus excavatum without complete resolution of symptoms. A failed repair is marked by a
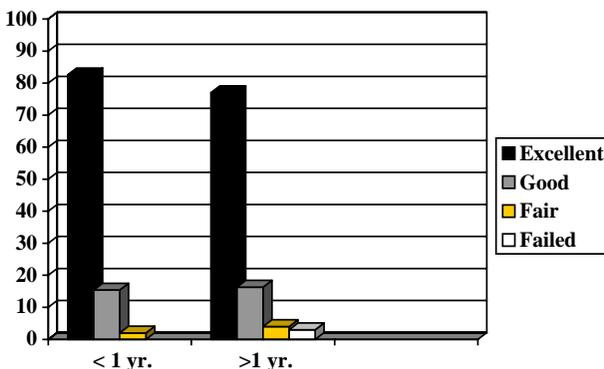

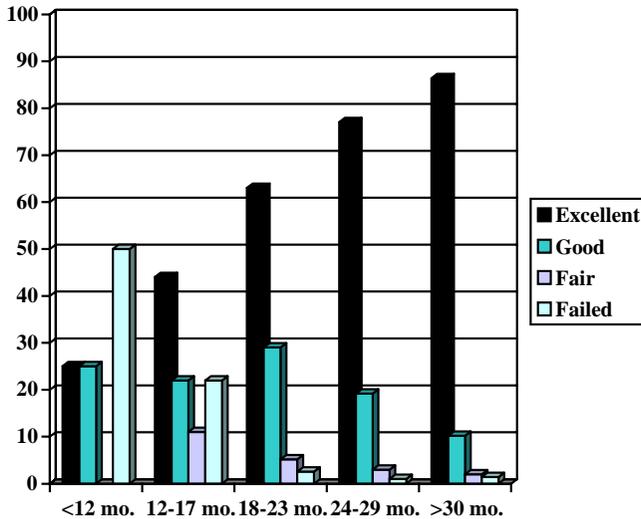Fig. 5. Long-term results by time since bar removed.

Fig. 6. Long-term results by length of time bar in situ.

recurrence of the pectus deformity and associated symptoms or need for additional surgery after final removal of the bar.

In addition, patients with EKG conduction abnormalities or mitral valve prolapse (MVP) had follow-up assessments. Patients old enough to have PFT studies were reassessed with repeat studies.

It has been noted that patients who are sedentary and who do not perform the pectus breathing exercises tend to develop mild recurrence over the long term. The authors therefore strongly emphasize the importance of aerobic activities and deep breathing exercises.

The initial cosmetic and functional results are excellent in 492 patients (88%), good in 63 patients (11%), fair in 1 patient (0.2%), and failed in 1 patient (0.2%) overall. The bars have been removed in 298 patients (54%), with 245 patients (82%) more than 1 year postbar removal and 53 patients (18%) less than 1 year postbar removal. In the group whose bar had been out for less than 1 year, the results were excellent in 83%, good in 15%, and fair in 2%. In the group in which patients bars had been removed more than 1 year ago, the results were excellent in 77%, good in 16.3%, fair in 4%, and failed in 3% (see Fig. 5). The long-term results were affected by the length of time the bar was left in place (see Fig. 6) and by the age of the patients at the time of surgery (see Fig. 4).

Bar removal

The authors advise that the pectus bar be left in place for 2 to 4 years. The authors evaluate patients on an annual basis and monitor their growth, activity level, and PFTs, and encourage them to do their pectus exercises and participate in aerobic sports. Patients between the ages of 6 to 10 often do not grow rapidly.

Therefore, they tolerate the bar well for 3 or even 4 years. On the other hand, the authors have had teenagers who have had a massive growth spurt, growing 14 cm a year. They completely outgrow the bar and require bar removal after only 2 years.

The authors consider the exercise programs to be just as important as the surgery. Many children and adults lead sedentary lifestyles and never perform aerobic activities. Therefore, their lungs never expand beyond the resting tidal volume, or approximately 10% of total lung capacity. Deep breathing with breath holding for 10 to 15 seconds and aerobic activities like running (such as in soccer or basketball) and swimming are encouraged. Long-term, the authors have seen mild recurrence of the pectus anomaly in patients who do not follow an exercise protocol.

Pectus excavatum can be corrected with excellent long-term results without necessitating costal cartilage incision or resection or sternal osteotomy. Many patients have been managed safely and effectively at long-term follow-up. Pectus excavatum repair without cartilage resection is a simpler operation with tolerable morbidity. As a result, it has received rapid acceptance by the surgical community.

Pectus carinatum

Introduction

Pectus carinatum, or protrusion deformity of the chest, occurs less frequently than pectus excavatum. It comprises about 15% of patients with chest wall deformities [36]. The prominence may be in the sternal manubrium [37], which is called a chondromanubrial deformity or "pigeon breast." The most common protrusion occurs in the lower or body of the sternum (the gladiolus), and is called chondrogladiolar or "chicken breast." The protrusion may be unilateral, bilateral, or mixed [38]. About 80% of patients are male. Although the etiology is unknown [39], a genetic component of causation is suggested by the approximately 25% of patients with a family history of chest wall defect [36]. Pectus carinatum has been reported to occur following treatment for pectus excavatum [40].

Pathophysiology

The natural history of the condition differs from pectus excavatum. Pectus carinatum is noted in childhood usually, especially around the time of a growth spurt, rather than at birth, as generally happens with pectus excavatum. Most patients are asymptomatic, and when symptoms occur, they are confined to tenderness at the site of the protrusion [38,41]. Associated mitral valve disease has been reported [42,43]. In patients without congenital heart disease, cardiopulmonary limitation caused by the condition has not been reported. Other

associations include Marfan's syndrome and scoliosis (in 15%) [36]. This re-inforces a possible etiology for pectus disorders as an abnormality of connective tissue development.

Therapy

There has been limited experience with orthotic bracing that has consisted of both dynamic chest compressors and body casting. Two reports have described correction or improvement in the condition by means of a brace analogous to that used for treatment of scoliosis, but exerting pressure in the anteroposterior direction [41]. The authors' group has had limited success with bracing. The ideal candidate is one who is around puberty and highly motivated to wear the brace. Those with mild deformities and a malleable skeleton will have the most potential benefit. If unsuccessful, orthotics do not preclude or complicate surgery.

Surgical treatment is by costochondral resection with sternotomy [38,44,45]. These studies emphasize the importance of performing a bilateral cartilage resection, even with unilateral deformity of the cartilages, to prevent recurrence. Overall, patients tolerate surgery very well with few postoperative problems. Recurrence is reported to be rare by centers with large experience. Deformities that recur tend to be in children who underwent correction at an early age or who had incomplete correction at the time of the original surgery.

Acknowledgments

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