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Pectus Deformities in Children: A Nine-Year Experience

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From January 1988 through to September 1996, 114 children with pectus excavatum and 15 with pectus carinatum were seen in our department. Both conditions occurred more frequently in boys (105 cases) than girls (24 cases). Three children with pectus excavatum had a definite history of trauma through playing basketball. Associated anomalies were identified in 39 children with pectus excavatum and in four with pectus carinatum. A familial history of chest wall defects was present in five cases with excavatum and in two with carinatum defects. Scoliosis was observed in four with pectus excavatum and in two with pectus carinatum. Pectus excavatum deformity was noted much earlier (mean age 6.3 years) than pectus carinatum (mean age 11.6 years). The deformities were severe in 36%, moderate in 46%, and mild in 2% of children on clinical judgement. Cardiopulmonary symptoms, attributed to the excavatum deformities, were exercise intolerance, recurrent respiratory tract infections, and asthma and were recorded in 66 (51%) children before surgery. Symptoms ^{were} relieved in the majority of cases following surgery. Surgical repair was performed in 123 patients (108 pectus excavatum and 15 carinatum) and the average age at operation was 7.7 years and 13.4 years in pectus excavatum ^{and} pectus carinatum, respectively. Our operative procedure is a modification of the Welch technique and consists of bilateral resection of deformed costal cartilages with preservation of perichondrial sheaths and anterior wedge osteotomy of sternum with no internal or external strutting. All repairs were completed without a need of blood transfusion and with a low complication rate (5.6%) with an average hospital stay of 4.6 days. Satisfactory long-term results were achieved in 95% of cases (excellent 49%, very good 53%) with follow-up ranging from two weeks to five months (mean 17.5 months). One residual deformity (in pectus excavatum) and three major recurrences (2 pectus carinatum and 1 pectus excavatum) were noted and led to revision in three cases. It was believed that children with moderate to severe pectus deformities require surgical correction for both physiological as well as cosmetic reasons. Early repair of pectus excavatum deformity is efficacious in providing suitable body contour and to allow normal growth of the lungs once structural compression of the chest wall is relieved. Surgery is safe, does not require blood transfusion, complications are minimal, and the long-term results are very satisfactory in both types of defects. (*Asian J. Surgery* 1999;22(3):0-0)

Pectus excavatum and carinatum are common congenital anterior chest wall deformities in children.^{1,2}
³ Pectus excavatum (funnel chest) is more common and characterized by prominent posterior curvature of the body of the sternum, usually involving its lower half or two thirds, with its deepest point just above the junction with xiphoid. This is often noted at birth, but becomes

more noticeable in later infancy. Pectus carinatum (pigeon chest) is 10 times less frequent than excavatum defects, is almost imperceptible in early childhood, and becomes prominent during the rapid growth in early adolescence. Each condition is not a single entity but a spectrum of abnormal thoracic development varying from symmetric concavity or convexity of the costal cartilages on one side to an asymmetric one on the other side. Both

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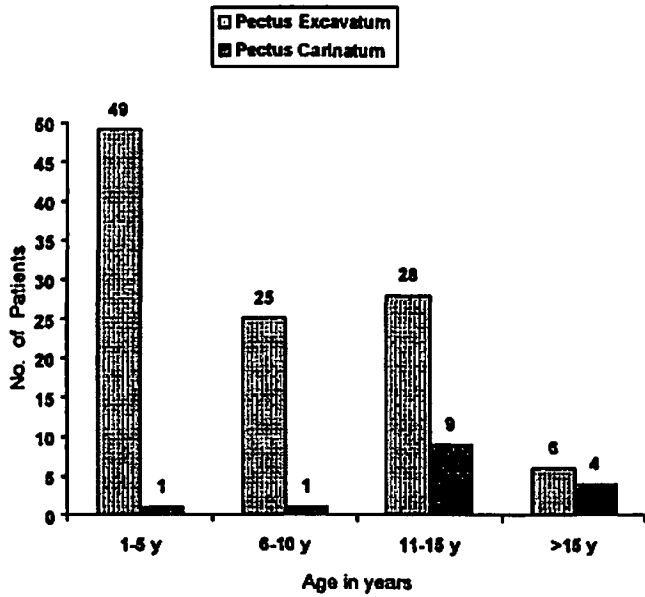


Figure 1. Pectus deformities; age of patients at operation.

deformities may give rise to physical and cosmetic complications.

Controversies involving the indications, optimal age, and method of operation still exist due to conflicting conceptions of the pathogenesis and results. The present study is an overview of our nine-year experience with these deformities with an analysis of indications, age at operation, and long-term results of operative correction using a single surgical technique.

PATIENTS AND METHODS

From January 1988 through to September 1996, 129 children and teenagers with congenital and acquired pectus deformities were referred to our department for evaluation and surgical correction. The medical records and office charts of these patients were reviewed to document type, gender, age at diagnosis and operation, family history of chest wall deformities, scoliosis, associated malformations, severity of defect, symptoms, cardiopulmonary studies, surgical technique, and long-term results on follow-up. Of the 129 deformities, 114 (88%) children had pectus excavatum and 15 (12%) had pectus carinatum deformity. Three children with pectus excavatum had a definite history of basketball trauma before they noticed their defect. The timing of surgical repair for both varieties is illustrated in Figure 1.

Surgical technique

Our surgical technique is a modification of the Welch procedure, which does not utilize internal or external strutting.⁴ A submammary incision within the nipple lines is preferred. The vertical incision is only used when the deformity extends to the second cartilage. In girls, to avoid breast deformity, as reported by some authors,⁵ we pay particular attention to place the incision within the projected inframammary crease. In both the procedures, the initial incision is carried through the pectoral muscles to the level of the sternum and the costal cartilages. Using the standard Bovie electrocautery or Argon beam cautery, the myocutaneous flaps are developed superiorly, laterally, and inferiorly to expose the sternum and all the deformed costal cartilages on both sides. Beginning superiorly, bilateral subperichondrial resection of costal cartilages is performed with the help of a blunt "whale tail" periosteal elevator. This usually involves the third through to the eighth costal cartilages. The lower costal cartilages often have bridges, which require a perichondrial incision to facilitate the complete removal of each deformed costal cartilage. The anterior wedge osteotomy is done in the space above the most superior cartilage excision. The posterior table is broken. The xiphoid is removed and the fascia of linea alba is sutured to the xiphoid base with permanent sutures to hold the sternum in a forward position. Reefing of perichondrium is done with permanent interrupted 2-0 silk sutures placed in a figure-of-eight manner, alternating from one side to the other, to avoid deflection of the sternum towards one side. The wound is closed leaving one or two chest tubes beneath the myocutaneous flaps to avoid postoperative fluid collection. Drains are usually removed after 48 hours. An antibiotic, usually Cefazolin (Ancef®), is utilized starting peri-operatively and is continued for two days. Postoperatively, all patients wear a clavicular splint continuously (except when bathing) for six weeks to three months. They are also advised to protect themselves from "roughhouse" activities during the initial six weeks and from upper body exercise for three months. Our redo operations were also done in a similar manner.

RESULTS

Males were affected more in both conditions yielding a ration of 4.4:1 (Figure 2). A family history of chest wall deformity was present in seven (5.4%) patients (5 relatives

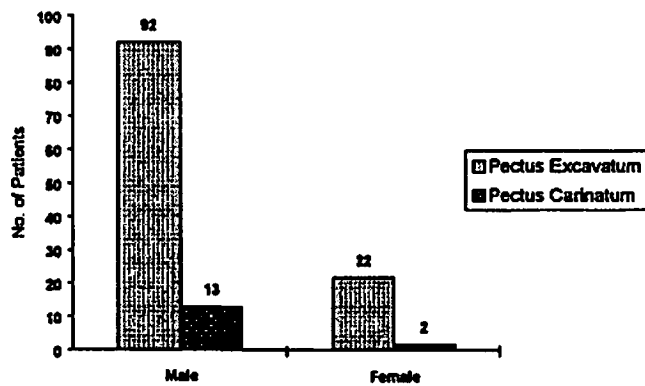


Figure 2. Gender distribution in 129 cases of pectus deformities.

had pectus excavatum while 2 had pectus carinatum) and six (4.6%) had a positive family history of scoliosis. Forty-four (34%) children had associated malformations of other systems (Table 1). There were six patients who had multiple and lethal abnormalities which precluded surgical treatment. Symptoms related to their defects were noted in 66 patients (Table 2). The mean age of diagnosis was 6.3 years for pectus excavatum and 11.6 years for pectus carinatum. The deformity, as estimated clinically, was severe in 58 cases (35%), moderate in 46 cases (45%), mild in two cases (1%), and not recorded in 23 patients (Table 3). Fifteen (11.6%) patients of the pectus excavatum variety had cardiopulmonary studies done before surgery, because of their physiological problems or associated malformations (Table 4), and these were abnormal in nine patients. The surgical procedures done in the 123 patients were completed with no mortalities. There was a 5.7% complication rate with two cases each of wound infection (one patient with associated Ehler-Danlos Syndrome) and hypertrophic scarring, and one case each of recurrent respiratory tract infection (a patient with BPD & GER required a longer hospital stay and antibiotics), brachial plexus neuropraxia (secondary to clavicular splint), and chest pain with dyspnea (Table 5). The mean hospital stay was 4.6 days and none of the patients required a blood transfusion. One hundred and nineteen (97%) patients were available for follow-up; the remaining four could not be located because of migration. Satisfactory long-term results were achieved in 112 patients (101/106 pectus excavatum and 11/13 pectus carinatum) with follow-up ranging from two weeks to 85 months (mean 17.5 months) although a mild recurrence was noted in three pectus

Table 1. Associated malformations in pectus deformities

Associated malformations	No. (%)
Cardiovascular System	10 (22%)
PDA	3
ASD	2
Mitral Valve Prolapse	2
VSD	1
Regurgitation of tricuspid valve	2
Coarctation of aorta	1
Murmur, unspecified	2
(3 patients had more than 1 lesion)	
Musculoskeletal System	22 (50%)
Bilateral inguinal hernia	9
Scoliosis	3
Nevus	3
Club foot	2
Umbilical hernia	2
Congenital diaphragmatic hernia	2
Undescended testis	1
Bilateral hallus valgus	1
Epigastric hernias	1
(5 patients had more than 1 lesion)	
Other Systems:	12 (27%)
Central nervous system	
Hydrocephalus	1
Gastrointestinal tract	
GI perforation	1
Renal	
Post brethral valves ← Posterior urethral valves	2
Respiratory	
Laryngomalacia	1
RDS	2
BPD	2
Eye	
Strabismus	1
Trisomy 21	2
Total	45 (34%)

excavatum patients. Unsatisfactory results were observed in four (3.2%) patients, where three had major recurrences (2 pectus carinatum and 1 pectus excavatum) and in one patient with pectus excavatum and a residual deformity. Revision surgery was done in three (2.4%) children and the cosmetic results after redo were acceptable in two patients and good in one case six months after repeat

Table 2. Symptoms in patients with pectus deformities

System Involved	Pectus Excavatum	Pectus Carinatum	Total	(%)
Respiratory				
Asthma	16	0	16	12.4%
Recurrent respiratory tract infection	24	2	26	20.1%
Chronic cough	1	0	1	0.7%
Cardiovascular				
Exercise intolerance	28	2	30	23.5%
Pericardial pain	6	0	6	4.6%
Murmur, compression	3	0	3	2.3%
Miscellaneous				
Tenderness over the protrusion	0	2	2	1.6%
Dysphagia	1	0	1	0.7%

19 patients had multiple symptoms

Table 3. Severity of deformity in 129 patients as estimated clinically

Severity	Pectus Excavatum	Pectus Carinatum	Total	(%)
Severe	51	7	58	(36%)
Moderate	40	6	46	(45%)
Mild	2	0	2	(1%)
Not recorded	21	2	23	(18%)
Total	114	15		

surgery. Overall satisfactory results were obtained in 95% (113) patients (Table 6).

DISCUSSION

Pectus excavatum and carinatum are common anterior chest wall deformities in children. Both occur more frequently in boys and are most likely polygenic or multifactoral.^{2,3,6,7} We also had a predominance of male patients with a ratio of 4.4:1 with a familial occurrence of 3.8% in pectus excavatum and 1.5% in pectus carinatum defects. Some authors⁷ have reported 14% familial incidence for pectus excavatum. Congenital heart and musculoskeletal anomalies are common associated malformations reported in these children.⁸⁻¹¹ Sometimes

Table 4. Cardio-pulmonary studies in 15 patients with pectus excavatum done before surgery

Test	No.	Indications
Pulmonary function tests	7	Severity of defect (4) Exercise intolerance (1) Broncho pulmonary dysplasia (1) Recurrent respiratory tract infection (1)
ECG	7	Multiple malformations (2) Murmur (2) Exercise intolerance (1) Mitral valve prolapse (1) Multiple cardiac anomalies (1)
Echo	6	Multiple malformations (2) Mitral valve prolapse (1) Severity of defect (1) Murmur (1) Multiple cardiac anomalies (1)
ETT	3	Exercise intolerance (1) Murmur (1) Recurrent respiratory tract infection (1)

this association is multiple or lethal, and may preclude the surgical intervention, as occurred in six cases (all pectus excavatum) of all studied. The incidence of associated congenital abnormalities seen in the present study is higher than those reported earlier in the literature.⁸⁻¹¹

Table 5. Post-surgical complications in pectus deformity patients

Complication	No.	%
Wound infection	2	1.6
Hypertrophic scar	2	1.6
Recurrent respiratory tract infection	1	0.8
Brachial plexus neuropraxia	1	0.8
Chest pain and dyspnea	1	0.8
Total	7	5.6

Table 6. Cosmetic results in 117 pectus cases based on postoperative follow up

Cosmetic Results	Pectus Excavatum	Pectus Carinatum	Total	(%)
Excellent	47	10	57	(49%)
Very Good	50	3	53	(45%)
Acceptable	3	0	3	(2.5%)
Unsatisfactory	4	0	4	(3.4%)
Total	104	13		

It is now generally accepted that pectus deformities influence the patient both physically as well as psychologically. In pectus excavatum, there is a decrease in the antero-posterior (AP) diameter of the chest, which interferes with optimum filling of the heart, producing a decrease in stroke volume and cardiac output. This diminished intra-thoracic space may influence the future growth of lungs in early years and may also cause cardiorespiratory symptoms. The toddlers and teenagers with pectus deformities are shy, withdrawn, and often refuse to participate in swimming, gymnastics, or other activities that disclose their deformity. Some authors and most primary physicians believe that these defects do not produce any cardiovascular or pulmonary impairment.¹² This opinion is in contrast to our study, which shows that 66 (55%) children had different cardiopulmonary symptoms (Table 2). Many patients required regular medications for their persistent asthma and recurrent respiratory tract infections before surgery, which were relieved in a vast majority following surgery. The exercise

tolerance was also increased, as assessed subjectively. Various physiological and radiological methods have been attempted to evaluate these children before and after surgery.^{2,13-20} These techniques are sophisticated, invasive, expensive, often require skill, and usually have involved small groups of patients. These methods also do not have any significant advantage in the decision for surgical management. Therefore, the basis for operation was usually clinical judgement.

Different opinions exist regarding the optimum age for surgical repair for pectus excavatum deformity, as there is no universally accepted age. Most surgeons advocate early operation,²¹⁻²⁷ in contrast to others who recommend operation after completion of growth because they found their results to be less satisfactory when surgical correction was done at a younger age.²⁸⁻²⁹ Recurrence was more commonly reported by some when they operated on younger children.^{30,32} However, satisfactory anatomical and cosmetic results are reported in children who were operated upon under 36 months of age on one- to 12-year follow-up.²⁵ We prefer and agree for an early operation for pectus excavatum, as evident in our study (mean age at operation 7.7 years). The repair of pectus carinatum is done once they have gone through their growth spurt to minimize recurrence.

Since the first repair performed by Meyer and Sauerbruch in 1911 and 1913,^{33,34} an enormous variety of surgical techniques for pectus deformities have evolved with time.^{3,35-43} Many series claim good results with a specific technique, but no method has been universally accepted as the optimal method. All have some risks of recurrence, which plagues the management of this problem. Today, the major area of controversy centers on the use of metallic struts or wires for stabilization of the sternum in the early or late recovery period. Various authors have reported satisfactory results with and without struts.^{38,39,44} We have performed all our surgical repairs with a single surgical technique, which does not require internal or external struts. It is safe and effective in achieving the two basic objectives of repair: the correction of deformity and the maintenance of correction. There is minimal blood loss and low complication rates as evident from our study. The meticulous preservation of perichondrial sheaths, in our technique, provide the basis for satisfactory regeneration of costal cartilages in long-term correction of the defects. We agree with those who feel that internal support provides no additional benefit,

causes complications on occasions, and requires a second operation.^{8,40,43,44}

The results of our surgical repair have been gratifying (48% excellent and 45% very good) as judged by patient satisfaction and linear follow-up ranging from two weeks to 85 months (mean 17.5 months). Unsatisfactory results, in the form of major recurrence or residual deformity, have occurred in four (3.4%) patients (2 pectus excavatum and 2 pectus carinatum) while three (2.5%) others, two with excavatum and one with carinatum, have had acceptable results. One patient with major recurrence had a mixed or asymmetrical deformity of the excavatum and carinatum variety associated with Poland Syndrome and an absent third rib. One case of pectus carinatum required revision surgery twice and another patient with residual deformity (pectus excavatum) had a minor revision surgery. An asymmetric deformity often results in a less satisfactory repair. In a female patient with pectus excavatum, a revision of her postoperative hypertrophic scar was also done a year after her pectus repair. Overall satisfactory results in our study were achieved in 113 (95%) patients, with a low morbidity and no mortalities. Our long-term follow-up results compare with those reported by others.^{8,9,45-47}

In conclusion, we believe that in children with pectus deformities, satisfying long-term results can be obtained with a technique that avoids struts. The optimal age for surgical repair of pectus excavatum is 305 years, when they may benefit from more space for their lungs to grow and have not suffered psychologically from peers due to cosmetic concerns. Physical as well as psychological (cosmetic) factors are positively influenced by the operation and may serve as indications for surgical intervention. Finally, surgery for pectus deformities is safe, does not require blood transfusion, complications are minimal, and long-term results are very pleasing.

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