CONGENITAL DEFORMITIES OF THE ANTERIOR CHEST WALL
A 10 Years Surgical Experience
Arterior göğüs duvarının doğumsal deformiteleri

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Summary: *We reviewed fifteen patients with chest wall deformity in our department in last ten years. All these patients were treated surgically for cosmetic improvement of thoracic wall deformity. Thoracic deformities were found to be as the following: pectus excavatum(53%), pectus carinatum (33 %), and atypical costal anomalies (14%). Scoliosis, Poland's syndrome, and mitral valve proplapse were associated with pectus excavatum and carinatum. The complications of pectus excavatum and carinatum repair were pneumothorax in which six patients required chest tube placement, wound infection, and local tissue necrosis. There was no major recurrence.*

Key Word: *Chest wall deformity*

Özet: *Anabilim Dalımızda son on yıldaki göğüs duvarı deformiteli onbeş hastayı gözden geçirdik. Tüm bu hastalar toraks duvarı deformitesinin kozmetik iyileşmesi için cerrahi olarak tedavi edildi. Gözlenen toraks deformitelerinin % 53'ü pektus ekskavatum, %33'ü pektus karinatum ve %14'ü atipik kostal anomalilerdi. Skolyoz, Poland sendromu ve mitral valv prolapsusu pectus ekskavatum ve karinatuma eşlik eden anomalilerdi. Pectus ekskavatum ve karinatum onarımının komplikasyonları, altı hastaya göğüs tüpü takılması gerektiren pnömotoraks , yara infeksiyonu ve lokal doku nekrozuydu. Majör rekürrens yoktu.*

Anahtar Kelime: *Göğüs duvarı deformitesi*

Costal and sternal deformities are sufficiently grotesque and obvious that one would expect them to have been recorded from ancient times. However, apart from descriptions of ectopia cordis, the congenital deformities received scant attention until the last half of the nineteenth century. The operative correction of chest wall deformities has been largely a development of thoracic surgery since World War II. Of the congenital deformities of the sternum, pectus excavatum(i.e.,funnel chest, koilosternia, trichterbrust)is the most common. Operative treatment offers a great deal to these patients and it can be performed effectively and safely with the expectation of encouraging results (14,15).

The deformities of the sternum are of three principal types:depression deformities (i.e.,pectus excavatum or funnel chest), protrusion deformities(i.e., pectus carinatum or pigeon or chicken breast), and sternal cleft (14). A ten year experience of the surgical management of these conditions at the Department of Thoracic and Cardiovascular Surgery, Gevher Nesibe Hospital, Kayseri, from1982 to1992 was presented.

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METHODS

Between the years 1982 to 1992, fifteen patients underwent operative correction of the anterior chest wall deformity. The mean age was 13.4 years (range,6 to 19 years). There were three girls and twelve boys. Eight (53%)patients had pectus excavatum, five(33%) pectus carinatum, and two (14%) atypical costal anomalies.

The following studies were made routinely in all patients:plain chest roentgenographies with/or without barium in posteroanterior and lateral plane electrocardiography; echocardiography; pulmonary function tests and examination of blood gases before and after effort; abdominal ultrasonography; pre-and post-operative chest photographs.

Chest roentgenograms were taken postoperatively to exclude a pneumothorax or effusion and, to check the position of the bar if placed. The width

and depth of deformity for pectus excavatum were determined by posteroanterior and lateral chest roentgenograms with barium that was filled in the 'sunk' (Fig.1).

Depression ratio and deformity grade were estimated to calculate "Welch index" for determining the severity of the anterior chest wall deformity on a scale of 1-10, as defined by Welch (20)(Fig.1). For this reason, each patient had a standing chest roentgenogram to asses the cardiothoracic ratio (CTR), 5th rib angle (i.e., caudal declination of the fifth rib), and prevertebral space of T3 and T9. (Fig.1)

Scoliosis, was defined as lateral curvature of the thoracic spinal column when viewed in the sagittal plane, was determined from the posteroanterior chest roentgenogram. The patients who had a positive clinical examination that included an Adams forward flexion screen for scoliosis and a lateral curvature of the spine >5o by radiographic masurement were accepted as one with scoliosis.

The echocardiographic study is and electrocardiography were undertaken to determine the incidence of mitral valve prolapse (MVP) and some dysrhytmias such as Wolff-Parkinson- White (WPW), Lown-Ganong-Levine(LGL) syndromes in our patients with pectus excavatum.

Pulmonary function studies were performed to determine vital capacity(VC), maximum breathing capacity(MBC),maximum voluntary ventilation, mean total lung capacity.

Pre-and post-oferative chest photographs were taken for comparision before and after surgical correction.

Surgery was recommended for patients whose anterior chest wall deformity was greater than 5 on a scale of 1 to 10. The incision in patients with pectus excavatum and pectus carinatum was usually a midline and vertical from the second costal cartilage downwards, extending over the xiphoid to the epigastrium, but in some patients, atransverse submammary incision was used. In operation for pectus excavatum, costal cartilages were exposed, deformed costal cartilages were resected, the xiphoid process was divided, the sternum was freed, sternal osteotomy was carried out, a retrosternal vacuum drain was placed behind the sternum and the wound was closed. In operation for pectus carinatum , muscles were reflected laterally off the costal cartilages, the deformed costal cartilages were resected subperichondrilly, the sternum was corrected and the wound was closed. The chest drainage tubes were normally removed in 24 hours.

RESULTS

Scoliosis(13.3%), Poland's syndrome(7.7%), and MVP(26.6%) was associated with pectus excavatum and carinatum.

Pain in the area of the deformed cartilages and a history of recurrent pneumonia were the most common symptoms(81%) .

Thirteen patients had no signifigant lateral curvature(<5°) by frontal roentgenograms, but two patients had a positive Adam's forward flexion screening and lateral curvature of the spine >5o by roentgenographic measurement.

A systolic ejection murmur of grade II-III/IV was indentified in five (33.3%) patients. Electrocardiographic abnormalities were not common. One patient had a LGL, one had a WPW,three (20%) had an intraventricular conduction defect (Rr waves in III and aVF) and one had a right bundle branch block. Four patients had a MVP.

Vital capacity was mean 9l percent of predicted VC in patients with pectus excavatum and carinatum. Maximum breathing capacity was also diminished. It was l2 percent less than predicted MBC. The differences among values of VC, MBC, FEV, FVC, pO2, and PCO2 of the patients before and after effort were not significant statistically.

Abdominal ultrasonographies did not demostrate any congenital defect. Routine laboratory tests of all patients were normal.
CTR was greater than 50% in five patients with

pectus excavatum. Three of the patients with pectus carinatum had an asymmetric deformity. All of the patients with pectus excavatum, expect one, had a Welch index greater than 5 on the scale of 1-10 (Fig.1).

The operation was performed in most cases(80%) for cosmetic reasons. Incisions were midline in nine(60%) patients (Fig. 2&3). In six patients, a transvers submammary incision was used (Fig. 4&5).

Complications of surgical correction in thirteen patients with pectus excavatum and carinatum were relatively unimportant. They were summarized in table1.

The results of surgical correction of our patients with anterior chest wall deformity were as the following:satisfactory correction in 73.3 percent of the patients , major recurrence in 6.6 percent, and mild recurrence in 13.3 percent.

$$WI = (1-D_1/D_2) \times 10 + \begin{cases} 0.5 \text{ if } \theta > 25^\circ \text{ and/or} \\ 0.5 \text{ if CTR} > 50^\circ \text{ and/or} \end{cases}$$

WI, Welch index; D_1/D_2=DR, Depression Ratio; (1-DR) XIO=DG, Deformity Grade; θ, Rib angle; CIR, Cardiothoracic Ratio

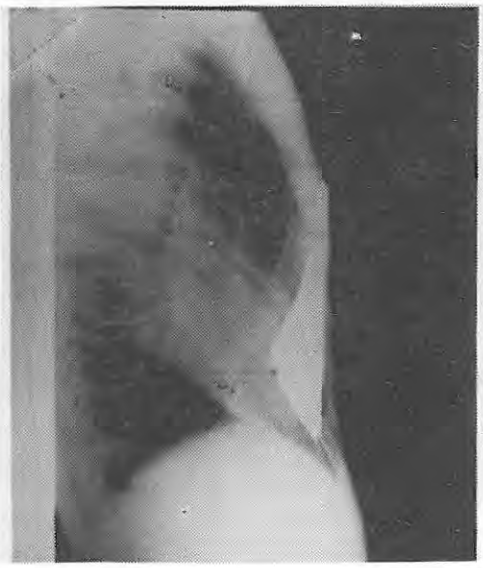

Figure 1. The calculation of depression rate, deformity grade, and welch index

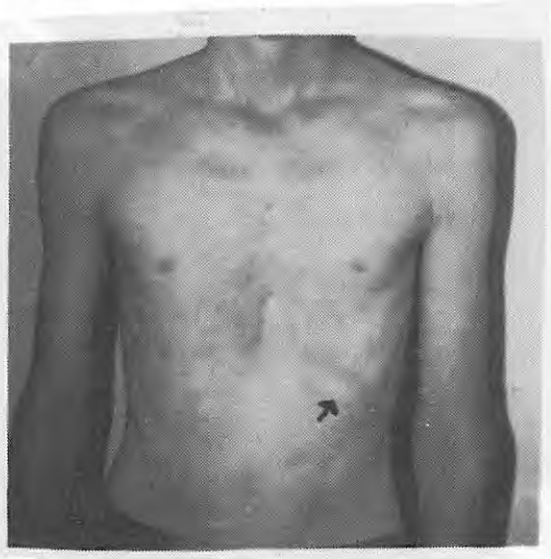

Figure 2. Pectus excavatum repair:preoperative photograph of a 13-year-old boy. Note the atypical thoracic deformity on left arcus costae

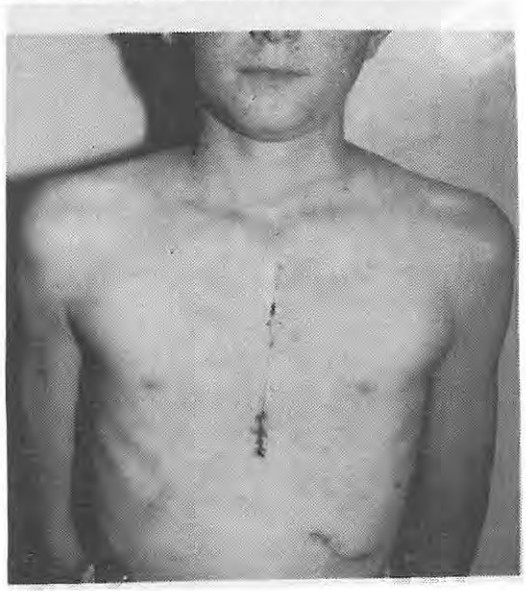

Figure 3. Pectus excavatum repair : postoperative photograph of the same patient.after seven days

Figure 4. Preoperative photograph of a 15-year-old boy with symmetric pectus carinatum

Figure 5. Postoperative photograph of the same patient shows operative correction thorough a transverse submammary incision (After ten days)

DISCUSSION

Since the late 15th century, chest wall deformities have been described, but no corrective surgical procedures were developed until 1911(6).These congenital chest wall deformities include bifid or cleft sternum which results from incomplete fusion of the halves of the developing sternum; Cantrell's pentalogy or thoracoabdominal ectopia cordis which consists of a split lower sternum, upper abdominal wall defect, partially absent pericardium, diaphragmatic hernia, and a major congenital defect such as tetralogy of Fallot;Poland's syndrome which consists of an absence of the lower one-half of the pectoralis mojor muscle, absence of the pectoralis minor muscle, and absence of parts of the underlying ribs, the second to fourth ribs, with or without an associated deformity of the arm and hand such as brachydactyly, syndactyly, or ectromelia; pectus carinatum or the pigeon breast which results from an overgrowth of the costal cartilages rather than a primary sternal deformity; and pectus excavatum or funnel chest which results from a depression of the sternum and the lower costal cartilages(6,20).

There are several theories about the etiology of pectus excavatum: (a)a functional abnormality of the anterior diaphragm, (b) a short subxiphoid tendon that holds the sternum posteriorly, (c)a short central tendon of the diaphragm, (d) failure of osteogenesis or chondogenesis, (e) an excessive misdirected growth of the lowermost costal cartilages, resulting in a concave deformity of varied proportions and inwardly forced lower sternal segment. In pectus excavatum, the superior manubrium, the first and second costa, and the corresponding costal cartilages are abnormally curved, resulting in a sharp posterior concavity of the gladiolus(4,6).

Pectus carinatum or pigeon chest which is described as anterior thoracic protrusion deformity is seen much less frequent than depression deformities-16.7 percent-. The pigeon breast abnormality that was defined as an overgrowth of the costal cartilages rather than a primary sternal deformity is divided into four types: (I) symmetric chondrogladiolar type, anterior displacement of the

sternum with symmetric concavity of the costal cartilages; (II) asymmetric chondrogladiolar type, anterior displacement of costal cartilages on one side and normally positioned or oblique sternum and cartilages on the contralateral side, (III) mixed carinatum/excavatum type, a carinate deformity on one side and depression or excavatum deformity on the contralateral side, often with sternal obliquity; (IV) chondromanubrial or "pouter pigeon"type, protrusion of the manubrium and second and third costal cartilages and relative depression of the gladiolus. The most frequent from is type I, -57.4%-, and the most unusual deformity is type IV, -2%-(6,21). There were three (60%) type I. pectus carinatum among our patients. The remaining two deformities were type II pectus carinatum.

Pectus excavatum was occured in much than one-half of our cases as compared with others (6,l4,l5,20), but there was no sternal cleft and Cantrel syndrome. Funnel chest is commonly present from birth, but occasional de novo cases appear at adolescence. There is an increased familial incidence of pectus deformities(20). Family history is present in 37 percent of cases with funnel chest (2l). In our cases with pectus excavatum, anterior thoracic deformity was present from birth.

Anterior chest wall deformites are associated with such disease as Marfan's syndrome or arachnodactyly,Poland's syndrome,Pierre-Robin syndrome,and newborn's asphyxiating thoracic dystrophy or Jeune's disease (1,20).Funnel chest occurs in almost 67 percent of patients with the Marfan syndrome. In a series (19), scoliosis that is frequently described as a major component of Marfan syndrome have been identified in 21.5 percent of patient with funnel chest . In our cases, the rate of scoliosis was 13.3 percent. In a study(10), a significant association between morphological abnormalities of the thoracic skeleton such as pectus excavatum or other nonclassic forms of anterior-posterior narrowing and scolisosis and pulmonary infection due to Mycobacterium avium complex have been observed.

Musculoskeletal abnormalities identified in patients with pectus excavatum include scoliosis(10,19,21), kyphosis, prune belly syndrome or abdominal musculature deficiency syndrome(21), Werdnig-Hofmann paralysis and other myopathies, chromosomal defects such as Turner's syndrome, Poland's syndrome, Marfan's syndrome, Coffin-Lowry syndrome, Noonan syndrome, Pierre-Robin syndrome, neurofibromatosis, cerebral palsy, tuberosis sclerosis, and congenital diaphragmatic hernia (1,21).

Congenital heart diseases (CHDs) such as transposition of great arteries, ventricular septal defect, tetralogy of Fallot, ASD either primum or secundum, complete atrioventricular canal, dextrocardia etc. are identified in 1.5 percent of cases of funnel chest (21). In our cases, CHDs were not identified.

Asthma is frequently identified in 5 percent patients with pectus deformity (21). There was no patient with asthma in our cases. Infants with funnel chest tolerate well its own conditions. There is a history of common recurrent pneumonia. Pain in the area of the deformed cartilages or precordial pain after sustained effort is a symptom of older children with sunken chest. A few patients complain of palpitations or syncope.

Posteroanterior and lateral thorax roentgenography, computed tomography scan, Moire photography, measurement of sternovertebral distance, pulmonary scintigraphy, static and dynamic lung function tests, multiple gated acquisition scans, electrocardiography, and echocardiography have been used to assess anterior chest wall deformities (2,4,5,11,12,14,15,17,20,21). We did not use the techniques such as CT scan, Moire photography, lung scan,and multiple gated acquisition scan.

A systolic ejection murmur which is attributed to the close proximity of or contact between the posterior sternal cortex and the pulmonary artery, and is magnified with a short interval of effort, is usually identified in 57 to 100 percent of patients with funnel chest (21).

Electrocardiographic abnormalities which are

attributed to the abnormal configuration of the thoracic wall and the displacement and rotation of the heart into the left thoracic cavity are common.Cardiac dysrhytmias which have been attributed to cardiac impingement by a depressed sternum, particularly paroxysmal supraventricular tachycardia(PST) and atrial fibrilation, have occured in patients with pectus deformity (11,12). It has been demonstrated that incidence of WPW syndrome was 4% in the patients with pectus excavatum, and that patients with WPW syndrome had a 27% incidence of pectus deformity (12) . In this study their findings indicate that WPW syndrome is the most important underlying mechanism for PST in patient with pectus deformity. In conclusion, pectus excavatum deformity is easily recognizable and should alert the physician to consider the possibility of WPW syndrome which develop a lifethreatening dysrhytmia,as an underlying condition when a cardiac dysrhytmia is present (12).

It has been showed that vital capacity and maximum voluntary ventilation in men with pectus excavatum were significantly decreased compared to control population (21). In an other study (3), the authors evaluated that the mean total lung capacity as a percentage of predicted in the excavatum patient who were symptomatic with exercise was 79 percent.

The pre-and post-operative studies of the patients with pectus deformity demonstrated that no abnormalities were demonstrated in the patients with pectus carinatum and low-normal vital capacity, a small improvement in total lung capacity and a significant improvement in maximal voluntary ventilation were seen en the pectus excavatum patients after surgery (2).

In adolescents with funnel chest, cardiac output and arterial blood gas values that become normal after repair of deformity are significantly abnormal under strenuous exercise conditions (6). Our study showed that arterial blood gas values unchanged importantly before and after effort in the pectus patients.

In adult patients with pectus excavatum, cardiopulmonary impairment that can only be demonstrated by exercise have been reported (7). In a study that was designed to identify if there is cardiopulmonary impairment in children with funnel chest and to evaluate if this impairment is related to the age of the child, it has been found that the pectus patients had normal working capacity, oxygen transport, and cardiac output in response to exercise,and showed a shortening in the preejection period and the preejection period to left ventricular ejection time ratio, additionally, the diastolic BP at maximal exercise and postexercise were significantly elevated in the pectus patients who were in the ages ll years or older, when compared with the normal patients (5). Unfortunately, we did not study cardiac performance in our patients with pectus patients.

Several authors identified in rates of 18-65 percent MVP in the pectus excavatum patients (1,12,21). It has been proposed that the cause of MVP in the pectus patients is anterior compression of the heart aganist the spine by the depressed sternum with resulting deformity of the ventricular chamber or the mitral annulus.

For surgical indication in the patients with pectus deformity, Ravitch said, ".. operation clearly indicated in all obviously symptomatic patients and in children with deep or progressive deformities, which we can expect to produce physiological derangement in time, whether symptoms are recognized as such or not. " (13). Ochsner-DeBakey's primary indication for repair has been to relieve pressure on the heart and lungs by elevating the sternum (6).The presence of a pectus excavatum is an indication for surgical correction. Some surgeon (20,21) prefer not to operate on the pectus excavatum patients who are younger than 3 or 4 years old, whereas some surgeons' patients have been infants in the ages of 1 to 2 years. Correction can be done both safely and effectively if a significant deformity is present at 1 or 2 old year. Operation should be done before a child reaches 5 years of age, however, the best results are obtained when operating on a child of 2 or 3 old year (13-15).

Sauerbruch's operation which consists of the rib

ends along the funnel sternum, and elevation of the sternum by traction sutures through the skin and suspended above the bed of the patient may be the first practice of external traction combined with resection (6).We applied traction to a 13-year-old boy with Ravitch-Poland Syndrome and a teenager with funnel chest.

Ravitch's operation has been modifications or extensions of all subsequent operations that have devised for pectus excavatum, with one exception, i.e., Wada's sternal turnover operation, without external or internal artifical support for the repositioned sternum (6,13).We operated on seven patients with funnel chest using modified Ravitch's technique. A midline incision offers better operative exposure and reduced blood loss in males and in all adults (4,13-16). In girls, a transvers submammary incision that yields a superior cosmetic results but involves more dissection may be used but in this incision, there is some risk to the vascular supply of the edge of the upper flap and also some loss of time and blood (13-15). We prefers the midline incision in the majority of our patients with pectus deformity.

The old-fashioned operative techiques for management of pectus excavatum include resection and discard of the plastron (Meyers, 1911), traction (Alexander, 1931; Lester, 1946), bone struts (Dailey, 1959), internal "hammock" support (Daniel,1958), sternal turnover (Nissen, 1944; Judet & Judet, 1954;Scheer, 1957;Judet &Valentin, 1964; Wada, 1968), metallic internal fixation (Ravitch, 1949; Brandt, 1953;Holmes, 1957), transvers metallic support (Grob, 1957;Rehbein, 1958), and non-structural cosmetic implants (Mason, 1970; Stanford , 1972)(6,13-16,20,21). We used a transvers metal strut for fixation of sternum in one patient, and a traction on the sternum, through a wire passed around it and through the skin in another patient.

Funnel chest chest costoplasty for deep, unilateral and asymmetric pectus deformity has been developed by Wada and Ikeda (16). Some surgeons have used sternal eversion, either with or without a vasculer pedicle attached to the sternum. Hawking et al(8) the vascular supply to the sternal graft has maintained by internal vascular pedicle in their sternal eversion technique for repair of pectus excavatum, and good results obtained in 81% of the patients. Hayashi and Maruyama (9), a vascularized costal strut based on the anterior intercostal branch of the internal thoracic artery has applied to provide rigid internal fixation of the thoracic wall after correction of funnel chest.

Even if the others (18) are still performing this, we did not performed a non-structural repair using Silastic implants for purely cosmetic reconstruction because we believe that important structural and physiological problems are associated with severe pectus deformities, and the implant would not be appropriate for a growing child.

Complications of pectus deformity repair are few and relatively unimportant, except for major recurrence of the abnormality years after the orginal correction. The complications of surgical correction for pectus deformity include pneumothorax (l.5%), wound infection that is the most serious complication in especially operative technique popularized by Koop, wound hematoma (0.4%), wound dehiscence (0.7%), pneumonia, trasnsient postoperative fever that is the most complication, seroma, hemoptysis, and hemopericardium (4,21). Major recurrence in our cases was fifteenth as compared with 2.7 percent of the others (21).

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