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# Pectus Excavatum and Pectus Carinatum: Associated Conditions, Family History, and Postoperative Patient Satisfaction

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Background: Pectus excavatum (PE) and pectus carinatum (PC) are the most common chest wall deformities. In this study, we aimed to characterize how patients obtained information about these deformities, as well as patients' family history, associated medical problems, and postoperative satisfaction after the Nuss and Abramson procedures. Methods: This cross-sectional retrospective study included patients who were operated by a single surgeon between 2006 and 2013. Follow-up calls were made after approval of our institution's ethics committee. We reached 207 of the 336 PE patients (61.6%) and 73 of the 96 PC patients (76%). Results: The majority of the patients were male (85% of the PE patients and 91.8% of the PC patients). The age of diagnosis of PE was 14.52±0.51 years and the age at the time of operation was 17.89±0.42 years; for PC patients, the corresponding ages were 15.23±0.55 years and 16.77±0.55 years, respectively. A total of 70% of the PE patients and 63.8% of the PC patients obtained information about pectus deformities through the Internet. In 27.1% of the PE patients with an associated anomaly, 57.1% (n=13) had scoliosis, while 41.1% of the PC patients with an associated anomaly had kyphosis (n=5). Postoperative satisfaction, as evaluated on a scale from 0 to 10, was 8.17±0.15 for PE patients and 8.37±0.26 for PC patients. The postoperative pain duration was 51.93±5.18 days for PE patients and 38.5±6.88 days for PC patients. Conclusion: In this study, we found that most patients with pectus deformities were male. The Internet was an important resource for patients to learn about their deformities. Family history and associated anomalies were identified as important aspects for consideration in the clinical setting. The patients reported high levels of postoperative satisfaction, and pain management was found to be one of the most important elements of postoperative care.

Key words: 1. Funnel chest

2. Pectus carinatum

### INTRODUCTION

Pectus excavatum (PE) or 'funnel chest' is the most common (90%) chest wall deformity, with a 5-9:1 male predominance [1]. It is characterized by the inward displacement of

the sternum or costal cartilage. Although some families contain more than one affected person, PE is a sporadic disease [2]. It occasionally occurs concomitantly with other anomalies, such as Marfan syndrome or scoliosis [2]. PE is generally asymptomatic, but in some cases, chest pain, dyspnea,

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diminished exercise tolerance, syncope, poor self-image, and avoidance of social interactions may occur [3,4]. PE patients often suffer from difficulties adapting to daily social life, and aesthetic concerns negatively influence the self-esteem of patients who require a medical operation [5]. The deformity has a range of clinical presentations, and asymmetry is frequently noted on the right side of the chest wall, which may even result in rotation of the sternum [3]. Moreover, scoliosis is present in 5% of PE patients, while congenital cardiac anomalies and asthma are found less frequently [3,4]. The classical method of correcting RE, the Ravitch procedure, includes sub-perichondrial resection and sternal turnover [6]. The Nuss procedure, which is more commonly performed than the Ravitch procedure, is a minimally invasive technique in which a metal bar is used to elevate the depressed sternum and costal cartilage [6].

Pectus carinatum (PC) or 'pigeon chest' is the second most commonly seen chest wall deformity [1]. PC is also sporadic and is less common than PE. It also affects males more than females, and is characterized by anterior angulation of the sternum and protrusion of the costal cartilage [4]. Children with PC may be less self-confident and more self-conscious [7]. These problems result in an abnormal posture, which exacerbates PC [7]. A minimally invasive method of surgical correction, known as the Abramson procedure, was developed on the basis of the Nuss procedure. The Abramson procedure involves a presternally placed metal bar that compresses the sternum and is attached to both sides of the chest wall [8].

Minimally invasive repair of PE [9] and PC [10] (the Nuss and Abramson procedures) is the currently preferred treatment. In the present study, we aimed to characterize how patients obtained information regarding their condition, their family history, associated medical problems, and postoperative satisfaction and pain levels after the Nuss and Abramson procedures.

# **METHODS**

This cross-sectional retrospective study included patients with PE and PC who were operated by the same surgeon (MY) between January 2006 and June 2013. The operations were performed under general anesthesia. A curved bar was

inserted under the sternum, and lateral incisions were made on both sides of the chest. The bar (Tasarimmed Medical Devices Manufacturing and Marketing Inc., Istanbul, Turkey) was individually curved for each patient and used to elevate the depression of the chest wall in cases of PE or to depress the protrusion of the chest wall in cases of PC. The bar was fixed to the ribs on both sides, and the incisions were closed. A steel plate was used to stabilize and fix the bar to the ribs.

Follow-up telephone calls were made after approval of the Marmara University School of Medicine Local Ethics Committee. The study was explained to the patients and informed consent was obtained from each subject. We reached 207 of 336 PE patients (61.6%) and 73 of 96 PC patients (76.0%). The patients were asked questions about their demographics and clinical characteristics. The data were analyzed using SPSS ver. 15.0 (SPSS Inc., Chicago, IL, USA). Continuous variables were expressed as mean±standard deviation. Nominal variables were expressed as numbers and percentages. The chi-square test or the Student t-test was performed to compare two different groups. All p-values <0.05 were considered to indicate statistical significance.

### **RESULTS**

Pectus deformities were generally recognized by the patients' families (53.6% for PE and 55.8% for PC). The demographic characteristics of the patients with PE and PC are presented in Table 1. The patients were most often voluntarily admitted to the thoracic surgery outpatient clinic (82.6% for PE and 80.8% for PC). The diagnoses were most commonly made at the Marmara University Hospital Department of Thoracic Surgery (43.2% for PE and 45.2% for PC). Of the patients with PE, 70.0% obtained information about pectus deformities through the Internet, compared to 63.8% of patients with PC. In contrast, 46.4% of the patients with PE and 37.9% of the patients with PC also reported obtaining information from their doctors.

27.1% of the patients with PE had an associated anomaly (Table 2), and 57.1% (n=13) of that patients had scoliosis. Additionally, 18.4% had an associated cardiac anomaly, 12.24% had kyphosis, and 8.7% had another accompanying disease. The body mass index of the patients with PE was  $20.08\pm0.21$ 

Table 1. Demographic characteristics of the patients with PE and PC

Characteristic	PE	PC 73	
No. of patients	206		
Mean age (yr)	20.53±0.44 (range, 8-47)	18.89±0.63 (range, 9-35)	
Gender (male:female)	176:31 (85%:15%)	67:6 (91.8%:8.2%)	
Haller index	4.01±0.13 (range, 1.87–10.5)		
Mean recognition age (yr)	10.05±0.44 (range, 0-30)	11.66±0.44 (range, 1–17)	
Mean diagnosis age (yr)	14.52±0.51 (range, 0-37)	15.23±0.55 (range, 3-32)	
Mean operation age (yr)	17.89±0.42 (range, 4–46)	16.77±0.55 (range, 10-33)	
Previously vs. currently have knowledge about disease	31% vs. 81%	32% vs. 79%	

PE, pectus excavatum; PC, pectus carinatum.

Table 2. Medical characteristics of the patients with PE and PC

Characteristic	PE	PC	
Family history	47% (n=97)	41% (n=30)	
Associated anomaly	27.1% (n=56)	17.8% (n=13)	
Postoperative satisfaction (0-10 point interval)	8.17±0.15	$8.37 \pm 0.26$	
Postoperative pain duration (day)	51.93±5.18 (range, 0-500)	38.5±6.88 (range, 0-300)	
Postoperative pain killer usage (day)	37.89±5.66 (range, 0-500)	23.97±3.63 (range, 1-210)	
Returned their normal life after the operation (day)	74.69±8.07 (range, 0–450)	45.0±5.05 (range, 3–180)	

PE, pectus excavatum; PC, pectus carinatum.

kg/m² (range, 11.52–28.37 kg/m²), with 32.5% of patients being underweight, 61.9% normal, and 5.6% overweight. 17.8% of patients with PC had an associated anomaly: 41.1% (n=5) had kyphosis, 16.7% had scoliosis, 8.3% had Marfan syndrome, 8.3% had a foot anomaly, 8.3% had flair chest, 16.7% had other problems, and 11% had an accompanying disease. The body mass index of the patients with PC was  $20.52\pm0.38$  kg/m² (range, 12.50–35.56 kg/m²), with 26.4% of the patients being underweight, 68.1% normal, and 5.6% overweight and obese.

Approximately half of the patients with PE (53.4%) or PC (41.1%) had at least one person in their first degree relatives affected by a similar deformity. Among the patients with PE, 59.1% had a distant relative affected by a similar deformity, 30.9% reported an affected cousin, 21.8% reported an affected sibling, 21.8% reported an affected aunt our uncle, 22.7% reported an affected father, and 7.28% reported an affected mother. The mean number of the affected relatives of patients with PE was 0.94±0.08 (range, 0-5). The corresponding figures for patients with PC were 70% for distant relatives, 23.3% for siblings, 10% of aunt or uncles, 23.3% of

fathers, and 6.67% of mothers. The mean number of affected relatives of patients with PC was  $0.59\pm0.1$  (range, 0–4). Consanguineous marriages between the parents of the patients were found in 19.7% of the PE patients and 32.3% of the PC patients.

In the postoperative follow-up period, 65.2% of the patients with PE and 64.8% of the patients with PC reported bar discomfort. In the patients with PE, the conditions that were associated with bar discomfort were lying on the side in 38.4% of patients, heavy lifting in 20.7%, cold weather in 21.7%, and hot weather in 3%. In the patients with PC, bar discomfort was associated with lying on the side in 45.1% of patients, heavy lifting in 26.8%, cold weather in 23.9%, and hot weather in 8.5%. The satisfaction with the Nuss procedure among the PE patients was as follows: 3.9%, very dissatisfied; 3.4%, somewhat dissatisfied; 10.7%, neither satisfied nor dissatisfied; 23.3%, somewhat satisfied; and 58.7%, very satisfied. The satisfaction with the Abramson procedure among the PC patients was as follows: 2.8%, very dissatisfied; 2.8%, somewhat dissatisfied; 8.3%, neither satisfied nor dissatisfied; 20.8%, somewhat satisfied; 65.3%, very satisfied. In the post-

Table 3. Characteristics of pain and satisfaction from operation according to age groups in patients with pectus excavatum

Characteristic	Operation age (yr)	No.	Mean±standard deviation	p-value
Postoperative satisfaction from body image	≥20	65	7.74±2.35	0.02
-	< 20	142	$8.45 \pm 1.87$	
Satisfaction from operation	≥20	65	4.15±1.17	0.09
	< 20	142	$4.41\pm0.94$	
Postoperative duration of pain	≥20	65	76.50±95.81	0.001
	< 20	142	40.45±58.35	
Postoperative pain killer usage	≥20	65	64.03±99.71	0.000
	< 20	142	25.90±35.55	
Back to normal life	≥20	65	93.95±98.07	0.05
	< 20	142	68.07±78.47	

operative period, three patients with PE and two patients with PC developed contact dermatitis, four patients with PE and two patients with PC developed cellulitis at the surgical site, three patients with PE and three patients with PC developed pneumothorax, and one patient with PE developed pericarditis. All complications were treated promptly.

When outcomes were analyzed according to the age of the patient at the time of the operation, a significant negative relationship was observed between age and postoperative satisfaction relating to body image, satisfaction with the Nuss procedure, the postoperative duration of pain, the duration of painkiller usage, and return to normal life in patients with PE (Table 3). The definition of returning to normal life is the ability to perform daily activities without any restrictions caused by pain related with the operation. However, no such associations were found in the patients with PC (data not shown).

#### DISCUSSION

In the present study, PE and PC were predominant in males (with a male-to-female ratio of approximately 9:1), in agreement with previous studies [1]. The mean age at diagnosis was between 14 and 16 years; it has been proposed that the growth spurt in adolescence makes the deformity more prominent [4]. An interval of approximately four years was identified between recognition of the deformity and the diagnosis. This emphasizes the importance of early diagnosis. Associated anomalies were found in one of every four patients with PE and one of every five patients with PC.

Moreover, approximately half of the patients with PE and PC had a positive family history, confirming the importance of gathering appropriate information from a patient's medical history and performing a comprehensive physical examination. The short duration of the operation time, with minimal scarring, explains the good postoperative satisfaction scores reported by the patients with PE and PC. Additionally, several studies have shown improvements in the quality of life of patients with PE and PC after correction of the deformity [11-14].

The patients most commonly obtained knowledge about their condition from the Internet. In this digital era, medical knowledge found on the Internet plays an increasingly important role [15]. We also found that a lower than expected percentage of patients with pectus deformities stated that they obtained knowledge from their doctor. This may have been due to the crowded nature of outpatient clinics and the tremendous workload of thoracic surgery residents. More time should be spent with patients explaining their conditions and the outcomes of the procedures. For this purpose, after this study we prepared a patient booklet about anterior chest wall deformities. The role of the Internet in providing patients with medical knowledge should be considered further, since many young adults prefer medical consultation via the Internet [16]. Patients with pectus deformities who seek medical care are mostly young adolescents and adults, so Internet-based healthcare information is especially important in this context.

The prevalence of a family history may be very high, potentially reaching up to 40% in patients with PE [17]. In a

study from Brazil, a family history of pectus deformities was found in 65% of patients [18]. In the present study, nearly half of the patients had a positive family history. However, in our study, we found fewer patients with scoliosis than has been reported in previous studies (16%–51%) [18]. Other anomalies are well known to be associated with these deformities (e.g., kyphosis, cardiac anomalies, and foot anomalies) [2]. We also observed that patients with pectus deformities were mostly slim and tall. The patterns involved in the genetic inheritance of pectus deformities are not well known. Further research is needed in this area, in order to explain the occurrence of associated anomalies, family history patterns, and the presence of a relatively low body mass index in patients with pectus deformities.

Postoperative satisfaction relating to body image was high, as expected based on previous studies [19]. The postoperative duration of pain and usage of painkillers were longer than expected. However, similarly to our findings, Nakanii et al. [20] also reported long-term pain after the Nuss procedure. Returning to normal daily life also takes time, and we informed our patients about these life-affecting considerations. Moreover, the early diagnosis and treatment of PE and PC are important. Especially for family physicians and pediatricians, the diagnosis of these deformities and associated conditions, in combination with prompt referrals to thoracic surgeons, would increase the success of the treatment and improve patient satisfaction.

In conclusion, in the present study, we found that most patients with pectus deformities were male. The Internet was an important resource for patients with pectus deformities to obtain information about their deformities. We prepared a booklet about chest wall deformities and shared this booklet with patients to improve the doctor-patient relationship and to answer the most frequently asked questions about their deformity. Family history and associated anomalies were important aspects to consider in the clinical setting. Patients reported high levels of postoperative satisfaction relating to their body image. Pain management and bar discomfort were identified as some of the most important elements of postoperative patient care.

# CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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