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Clinical, Demographic and Echocardiographic Characteristics of Pediatric Chest Deformities

Pediatrik Göğüs Deformitelerinin Klinik, Demografik ve Ekokardiyografik Özellikleri

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Abstract

Aim: We aimed to retrospectively evaluate the clinical, demographic and echocardiographic findings of children diagnosed with chest deformity in the pediatric cardiology clinic.

Material and Method: This study enrolled children under the age of 18 years who were referred with chest deformity to our pediatric cardiology unit, over a period of six years (January 2017-December 2022).

Results: The mean age of the patients was 9.9 ± 5.2 years, median 11 years (0-18 years old). 89 patients with abnormal echocardiographic findings: 42 (9.56%) mitral valve prolapse, 18 (4.1%) atrial septal defect, 9 (2%) ventricular septal defect, 8 (1.82%) bicuspid without aortic valve stenosis aortic valve, 5 (1.1%) patent ductus arteriosus, 2 (0.45%) pulmonary stenosis, 2 (0.45%) great artery transposition, 2 (0.45%) hypertrophic cardiomyopathy, subaortic ridge 1 (% 0.27). Cardiac compression was present in 13.4% of the cases with pectus excavatum. 13(%3) patients were operated by a thoracic surgeon. Marfan Syndrome was diagnosed in 7 patients and Noonan Syndrome was diagnosed in 2 patients who applied to our clinic with chest deformity.

Conclusion: We suggest that echocardiographic examination in patients with chest deformity is important in the diagnosis of congenital heart diseases, early diagnosis and treatment of heart compression finding.

Keywords: Children, chest deformity, echocardiography

Öz

Amaç: Çocuk kardiyoloji kliniğinde göğüs deformitesi tanısı alan çocukların klinik, demografik, klinik ve ekokardiyografik bulgularını retrospektif olarak değerlendirmeyi amaçladık.

Gereç ve Yöntem: Bu çalışmaya altı yıl boyunca (Ocak 2017-Aralık 2022) pediatrik kardiyoloji birimimize göğüs deformitesi ile başvuran 18 yaş altı çocuklar dahil edildi.

Bulgular: Hastaların yaş ortalaması 9,9 ± 5,2 median:11 yaş (0-18 yaş) idi. Anormal ekokardiyografik bulguları olan 89 hasta: 42 (%9,56) mitral kapak prolapsusu, 18 (%4,1) atriyal septal defekt, 9 (%2) ventriküler septal defekt, 8 (%1,82) aort kapak darlığı olmayan biküspid aort kapak, 5 (%1,1) patent duktus arteriozus, 2 (%0,45) pulmoner darlık, 2 (%0,45) büyük arter transpozisyonu, 2 (%0,45) hipertrofik kardiyomiyopati, subaortik ridge 1 (%0,27) idi. Pektus ekskavatumlu olguların %13.4'ünde kardiyak bası vardı. 13(%3) hasta göğüs cerrahisi tarafından opere edildi. Göğüs deformitesi ile kliniğimize başvuran 7 hastada Marfan Sendromu, 2 hastada Noonan Sendromu teşhis edildi.

Sonuç: Göğüs deformitesi olan hastalarda ekokardiyografik incelemenin doğumsal kalp hastalıklarının tanısında, kalp sıkışması bulgusunun erken tanı ve tedavisinde önemli olduğunu düşünüyoruz.

Anahtar Kelimeler: Çocuklar, göğüs deformitesi, ekokardiyografi

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INTRODUCTION

Congenital chest wall deformities can be classified as pectus excavatum, pectus carinatum, Poland syndrome, and sternal defects.[1] The most common deformation of the anterior chest wall is pectus excavatum.[1] It constitutes 90% of all chest wall deformities.[2] Pectus excavatum is seen in approximately one in 400 live births.[3] The rates are four times higher in boys according to girls.[3]

Chest wall deformities are accompanied by other malformations of the skeletal system, cardiovascular, gastrointestinal and genitourinary anomalies.[4] Pectus excavatum and carinatum are important clinical findings in Marfan syndrome.[4]

The degree of recession in the pectus excavatum determines the clinical findings. When there is moderate and severe inward collapse, pressure on the heart and lungs occurs. With exertion, chest pain and shortness of breath develop. In patients with pectus excavatum, a decrease in cardiac output occurs in the supine position and during exertion. Patients tend to avoid movement. Mitral valve prolapse is the most common cardiac anomaly in patients with pectus excavatum.[4] Mitral valve prolapse as a result of anterior compression of the heart thought to be developing. Atrial septal defect, and ventricular septal defect and aortic coarctation have also been reported in patients with pectus excavatum.[4]

The second most common chest wall deformity is pectus carinatum. It constitutes approximately 16% of all chest deformities.[6] It is more common in men than women. Approximately 25% of patients with pectus carinatum have a positive family history.[7]

There is protrusion of the sternum and chondrocostal joints in the pectus carinatum. Protrusion in pectus carinatum can be symmetrical or asymmetrical. Although it is more common in pre-adolescence and adolescence, it can also be seen in infancy. There is an increase in the severity of pectus carinatum during periods.

Approximately 18% of cases with pectus carinatum have a congenital heart disease.[8] The most important reasons for patients to apply to the clinic are chest pain and exertional dyspnea. Echocardiographic examinations of the patients not only evaluate the mitral valve, but also provide detailed information about other congenital heart diseases. Pectus carinatum may be accompanied by Marfan or Noonan syndromes.

We aimed to retrospectively evaluate the clinical, demographic, clinical and echocardiographic findings of children diagnosed with chest deformity in the pediatric cardiology clinic.

MATERIAL AND METHOD

We retrospectively analysed the demographic, clinical and echocardiographic data of patients with chest deformities.

Study Population

This study enrolled children under the age of 18 years who were admitted with chest deformity to our paediatric cardiology unit, over a period of six years (January 2017-December 2022). Pectus deformities were identified by physical examination of the chest.

Echocardiographic Examination

Echocardiographic examination was performed with the Philips EPIQ 7C (USA) device, by taking multiple orthogonal parasternal, apical and subcostal images of the patients lying in the left lateral decubitus position by the same pediatric cardiologist. Traditional echocardiographic evaluation includes measurements of left ventricular end-diastolic and end-systolic diameter, septal and left ventricular posterior wall thicknesses in diastole and systole, left ventricular ejection fraction (EF) and left ventricular fractional shortening (FS) from the parasternal long-axis view. In four-chamber imaging, it was evaluated whether there was any external pressure on the heart.

Statistical Analysis

Descriptive statistics were applied to the obtained data, and their distribution by age and gender was examined. SPSS 21.0 Software Program was used for statistical analysis in the study.

Ethics Committee Approval

The study was approved by Local Ethic Committee (Decision No: 2022/533). All procedures were carried out in accordance with the ethical rules and the principles of the Declaration of Helsinki.

RESULTS

During the study period, 439 patients presented or referral to our unit with chief complaint of chest wall deformity. There were 337 (76.8%) boys and 102 (23.2%) girls. The mean age of the patients was 9.9 ± 5.2 years with a median of 11 years and a range of 0-18 years. The patients with chest wall deformities were included, 189 (43%) pectus excavatum, 132 (30%) pectus carinatum and 118 (27%) were both, pectus excavatum and carinatum.

Echocardiograpy were performed in all patients. 350 (79.72%) patients with pectus deformity revealed normal echocardiography. Echocardiographic diagnoses included 42 (9.56%) mitral valve prolapsus, 18 (4.1%) atrial septal defect, 9 (2%) ventricular septal defect, 8 (1.82%) bicuspid aortic valve without aortic valve stenosis, 5 (1.1%) patent ductus arteriozus, 2 (0.45%) pulmonary stenosis, 2 (0.45%) transposition of the great arteries, 2 (0.45%) hypertrophic cardiomyopathy, subaortic ridge 1 (0.27%). **Table 1** summarizes echocardiographic findings of patients with chest deformity. **Figure 1** and **Figure 2** show the appearance and echocardiographic evidence of right ventricular compression of our patient diagnosed with pectus excavatum as cardiac compression finding.

13.4% of the cases with pectus excavatum had cardiac compression. 13(%3) patients were operated by a thoracic surgeon.

Marfan Syndrome was diagnosed in 7 patients and Noonan Syndrome was diagnosed in 2 patients who applied to our clinic with chest deformity. Pectus excavatum deformity was accompanying in patients with Marfan syndrome and Noonan syndrome.

Table 1. Echocardiographic findings of patients with chest deformity

	n	%
Mitral Valve Prolapse	42	9.56
Atrial Septal Defect	18	4.1
Ventricular Septal Defect	9	2
Bicuspid Aortic Valve	8	1.82
Patent Ductus Arteriozus	5	1.1
Pulmonary Stenosis	2	0.45
Transposition of Great Arteries	2	0.45
Hypertrophic Cardiomyopathy	2	0.45
Subaortic Ridge	1	0.27

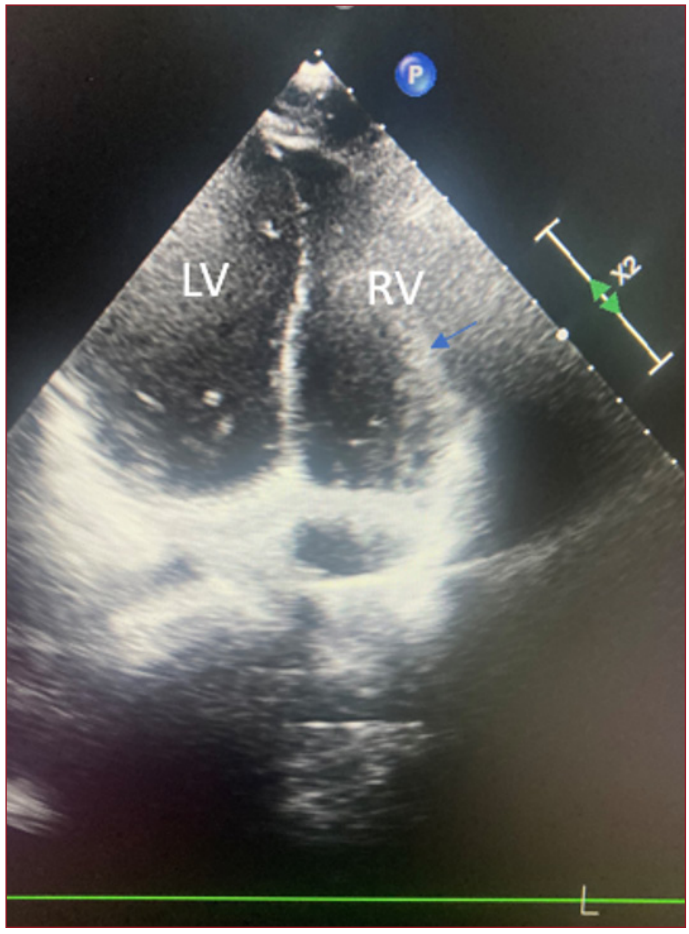

Figure 1: Arrow indicates right ventricular compression. RV: Right ventricul, LV: Left ventricul

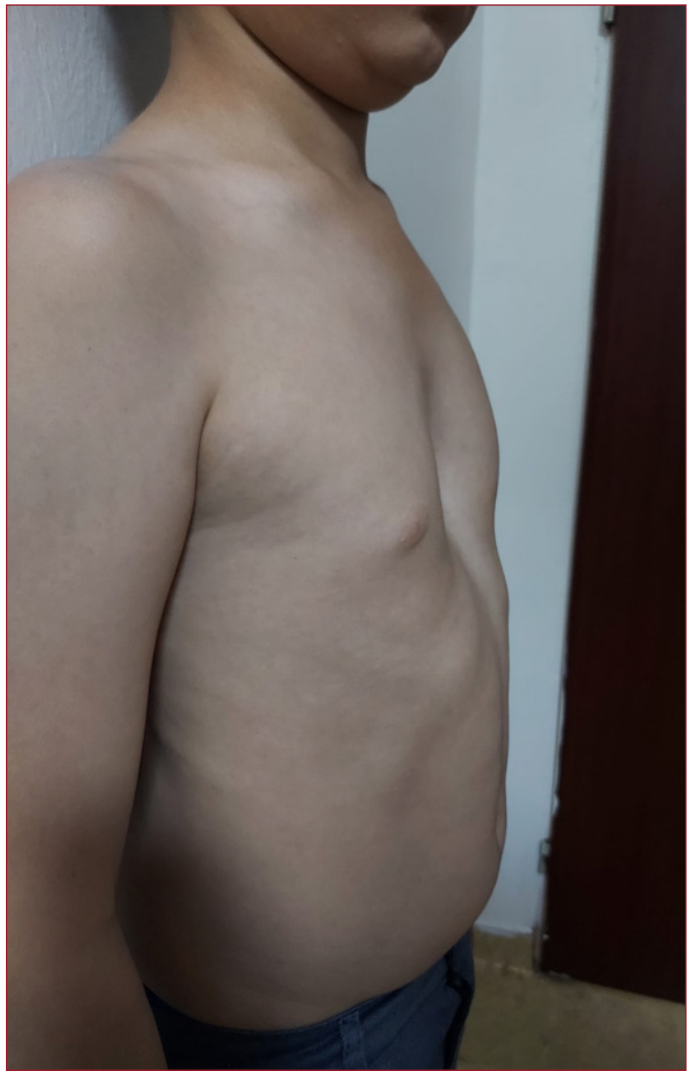

Figure 2: Appearance of the patient with pectus excavatum

DISCUSSION

Chest deformities are 4 times more common in boys than in girls.[9] In our study, similar to the literature, 76.2% of the patients with chest deformity were boys.

Congenital heart defects with pectus deformity is relatively common. Birth prevalence of congenital heart disease is estimated to be 8 cases per 1000 live births.[9] We found 20.8% congenital heart disease in the patients with pectus deformities.

The most common cardiac pathologies accompanying chest deformities are mitral valve prolapse and atrial septal defect. Park et al. found the incidence of mitral valve prolapse in the patients with pectus excavatum was 23%.[11] We found mitral valve prolapse (9.56%) as the most common cardiac pathology accompanying chest deformity. Mitral valve prolapse is thought to develop as a result of chest deformity pressing on the heart. Coln et al. found that in 123 patients, while 54 patients had preoperative mitral valve prolapse,

only seven patients had mitral valve prolapse after pectus correction surgery.[12] The improvement in a significant proportion of patients with mitral valve prolapse after pectus surgery supports the hypothesis that mitral valve prolapse develops due to cardiac compression.

Atrial septal defect is more common in children with chest deformity than in the normal population. The rate of atrial septal defect in patients with chest deformities was reported 2.1%, 2.8% and 15%, by Simsek et al, Akcalı et al and Sanchez et al respesctively.[13-15] We found the frequency of atrial septal defect in patients with chest deformity to be 4.1%. In patients with chest deformity, the evaluation of the atrial septum with transthoracic echocardiography can be difficult. We think that in selected cases transesophageal echocardiography can be performed.

Cyanotic congenital heart dissease is uncommon with pectus deformities. Kikuchi et al. reported pectus excavatum in a patient with tetralogy of Fallot.[16] We found transposition of great arteries in 2 patients with pectus deformity.

In chest deformities, cosmetic reasons, psychological reasons, lung and heart compression are surgical indications. Patients with a diagnosis of pectus excavatum should be carefully evaluated in terms of cardiac compression during echocardiographic examination. Zhao et al found that exercise capacity was limited as a result of reduced filling of he right heart by the compressive effects of pectus excavatum. [17] Cahill et al found an increase in cardiac stroke volume after surgical correction in patients with chest deformities. [18] Kowalewski et al found statistically significant increases in right ventricular volume indices after surgery.[19]

In our study, 13.4% of the cases with pectus excavatum had cardiac compression. Patients with cardiac compression were consulted with the thoracic surgery department. 13 (3%) patients were operated by a thoracic surgeon. After surgery, echocardiographic examination should be performed in terms of pressure on the heart. All patients were asymptomatic after surgery.

Chest deformities may occur as part of various syndromes. [20] Andrescu et al found that pectus excavatum and pectus carinatum were associated with Marfan and Noonan syndrome.[21]

Marfan syndrome involves the skeletal, ocular, and cardiovascular systems. Patients with Marfan syndrome present with tall stature, ectopia lentis, and a positive family history. Skeletal manifestations are the primary signs of Marfan syndrome. Pectus carinatum and pectus excavatum can be seen frequently in Marfan syndrome. Mitral valve prolapse, enlargement of the aortic root, rupture of sinus valsalva may be seen in patients with Marfan syndrome.[22]

Noonan syndrome is characterized by characteristic facies, short stature, congenital heart defect, pectus carinatum or pectus ekskavatum and developmental delay of variable degree.[23] We diagnosed Marfan syndrome in 7, Noonan Syndrome in 2 of the patients who applied to our clinic with chest deformity.

Since chest wall deformities may be associated with a genetic disorder or syndrome, a thorough family history taking, along with appropriate pediatric and genetic investigations, can provide valuable information.

CONCLUSION

Chest deformities can have a significant cardiac impact depending on the type and severity of the deformity. Cardiac compression, especially in pectus excavatum, which is the most common chest deformity, is an indication for surgery. Echocardiographic examination in patients with chest deformity is important in the diagnosis and treatment of the congenital heart diseases and heart compression. Echocardiographic evaluation should be performed in these patients, and they should be investigated for cardiac compression and congenital heart diseases. The patient's follow-up plan should be made with the thoracic surgery department by sharing the examinations of cardiac involvement.

ETHICAL DECLARATIONS

Ethics Committee Approval: Approval for this study was obtained from the Local Ethics Committee of Selçuk Medical Faculty Ethics Committee, 2022/533.

Informed Consent: Because the study was designed retrospectively, no written informed consent form was obtained from patients.

Referee Evaluation Process: Externally peer-reviewed.

Conflict of Interest Statement: The authors have no conflicts of interest to declare.

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