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Cardiac anomalies in pediatric patients with pectus excavatum

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SUMMARY

OBJECTIVE: Pectus excavatum is the most prevalently encountered deformity of the thoracic wall. It can be accompanied by congenital anomalies.

METHODS: The cardiac findings of 36 children who were diagnosed at the Thoracic surgery outpatient clinic of our university between 10 February 2021 and 1 October 2021 and 57 healthy children in a similar age group were analyzed.

RESULTS: We determined that the pectus excavatum patients in our study had a higher risk of having mitral insufficiency, mitral valve prolapse, tricuspid valve prolapse, cardiac malposition, and congenital heart disease.

CONCLUSION: Our study showed that the prevalence of cardiac pathologies was higher in pediatric pectus excavatum patients than in healthy children in the control group. Thus, we recommend clinicians to refer pediatric pectus excavatum patients to pediatric cardiology outpatient clinics for the early diagnosis of potential cardiac pathologies.

KEYWORDS: Pectus excavatum. Mitral valve prolapse. Tricuspid valve prolapse.

INTRODUCTION

Pectus excavatum (PE) is the most prevalently encountered deformity of the sternum and constitutes 90% of all congenital thoracic wall deformities[1]. In this deformity that is seen in one in every 300–400 live births, the female-male ratio is 1:4, where the prevalence of the condition is higher in males[2]. As the person ages, especially during the rapid development in adolescence, the deformity becomes more pronounced.

PE starts in childhood, and the deformity increases progressively. These patients, who have exercise intolerance at the beginning, start to have reduced cardiopulmonary function levels as the deformity increases[3]. The complaints of patients include fatigue, malaise, lower exercise capacity, palpitations, and chest pain. Through aging, these complaints increase.

Congenital cardiac diseases are observed in 2% of pediatric PE patients[4]. Patients with Marfan, prune belly, and Turner syndromes have an increased probability of having PE[5,6].

Varying degrees of systolic ejection murmurs can be heard in the physical examinations of individuals with PE deformity. This situation is more noticeable, especially following exercise. The closure of the distance between the pulmonary artery and the posterior sternal cortex or contact between these structures is shown as the cause of these murmurs.

In this study, we aimed to examine the electrocardiography (ECG) and echocardiography (ECHO) findings of pediatric patients with PE deformity comparatively with a control group without a thoracic deformity and discuss the results with the literature.

METHODS

This study was carried out in compliance with the Declaration of Helsinki. Before the inclusion of the patients, ethical approval was obtained from the Non-Interventional Clinical Studies Ethics Committee of our university on February 8, 2021 (meeting no: 2021/5 and decision no: 2). Additionally, the families of the patients provided informed consent.

Study design

The cardiac findings of 36 children diagnosed with PE at the Thoracic Surgery outpatient clinic of our university between February 10, 2021 and October 1, 2021 and 57 healthy control group patients in the same age group were analyzed. Routine posteroanterior and lateral chest radiographs of all patients were taken. Each patient was screened for cardiac pathologies by a pediatric cardiology specialist with ECG and ECHO.

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The demographic characteristics, complaints at the time of presentation, and ECG and ECHO results of all patients were recorded.

ECG: The patients were examined by 12-lead ECG. ECG examinations were made using a Gehealthcare Mac 2000 device. The electrocardiographs were examined in detail in terms of rhythm, heart rate, P-wave, ST-segment, QRS interval and amplitude, QTc interval, and T-wave values.

ECHO: All patients were examined by a pediatric cardiology specialist. All examinations were made with a Vivid 7 Pro ECHO device (GE Healthcare Vingmed Ultrasound AS) using the appropriate cardiac sector probe based on the patient's age and weight and in the left lateral decubitus position. The patients were examined with conventional ECHO methods based on the pediatric ECHO guidelines of the American Society of Echocardiography[7]. Patients who were followed up for congenital heart diseases (CHDs) and were diagnosed before the start of the study were excluded.

Statistical analyses

The statistical package for the Social Sciences for Windows 22 software was used for the statistical analyses. Descriptive variables are presented as frequency (n)–percentage (%) and mean±standard deviation values. The normal distribution of the variables was tested using the Kolmogorov-Smirnov test. The normally distributed parameters were analyzed by one-way analysis of variance (ANOVA) or Student's t-test. Kruskal-Wallis test or Mann-Whitney U test was used for the numerical variables that did not show normal distribution. Student's t-test, Mann-Whitney U test, and χ^2 test were used to find statistical significance. The risk factors were evaluated with univariate and multivariate logistic regression models. The variables found to be significant in the univariate analyses were included in the logistic regression analysis. $p<0.05$ was considered statistically significant.

RESULTS

Among the children who were included in the study, 24 were females, and 69 were males. The mean age of the patients was 11.10±4.04 (4–17.9) years. There was no statistically significant difference between the PE patients (PE group) and the healthy group (control group) in terms of sex or age (p=0.265 and p=0.506, respectively) (Tables 1 and 2).

In the comparison of the cardiological findings of the PE and control groups, no significant difference was found between the aortic valve pathology rates of the groups (p=0.740). The cardiac pathology, cardiac malposition, mitral valve prolapse (MVP), mitral insufficiency (MI), tricuspid valve prolapse (TVP), and CHD rates of the PE group were significantly higher than those of the control group (p<0.001, p<0.001, p<0.001, p<0.001, p<0.001, and p=0.027, respectively) (Table 1).

In the comparison of the ECHO findings, no significant difference was found between the PE and control groups in terms of their age, right ventricle end-diastole Z-score (RVED ZS), left ventricle end-diastole internal diameter Z-score (LVIDd ZS), end-diastolic left ventricle posterior wall thickness Z-score (LVPWd ZS), left ventricle end-systole internal diameter Z-score (LVIDs ZS), ejection fraction (EF), shortening fraction (SF), aortic sinus Z-score (Aort ZS), main pulmonary artery diameter Z-score (MPA ZS), Mitral annulus Z-score (Mannulus ZS), or pulmonary circulation values (p=0.506, p=0.093, p=0.146, p=0.195, p=0.238, p=0.656, p=0.900, p=0.736, p=0.921,

Table 1. Echocardiographic findings in patients with pectus excavatum and healthy control.

	Healthy control (57) n (%)	Patients with pectus excavatum (36) n (%)	p-value
Sex			
Female	17 (29.8)	7 (19.4)	0.265
Male	40 (70.2)	29 (80.6)	
Cardiac pathology	3 (5.3)	23 (63.9)	**<0.001**
Cardiac malposition	0 (0)	8 (22.2)	**<0.001**
MVP	4 (7)	20 (55.6)	**<0.001**
Mitral insufficiency	0 (0)	9 (25)	**<0.001**
TVP	1 (1.7)	9 (25)	**<0.001**
CHD	0 (0)	3 (3.2)	0.027
Aortic valve pathology	1 (1.8)	1 (2.8)	0.740

Statistics: Crosstabs, chi-squared tests. MVP: mitral valve prolapse; TVP: tricuspid valve prolapse; CHD: congenital heart disease. Statistically significant values are indicated in bold.

Table 2. Echocardiographic morphometric measurements in patients with pectus excavatum and healthy control.

	Healthy control (57) x±SD	Patients with pectus excavatum (36) x±SD	p-value
Age (years)	10.87±3.89	11.46±4.31	0.506
RVDD ZS	0.953±0.395	0.811±0.376	0.093
IVSd ZS	0.423±0.492	0.190±0.586	**0.042**
LVIDd ZS	-0.391±0.683	-0.622±0.823	0.146
LVPWd ZS	0.349±0.471	0.180±0.675	0.195
LVIDS ZS	-0.744±0.704	-0.912±0.602	0.238
EF	72.378±3.701	72.722±3.746	0.656
SF	41.386±3.211	41.472±3.193	0.900
Aort ZS	-0.555±0.785	-0.613±0.794	0.736
MPA ZS	-0.226±0.710	-0.242±0.807	0.921
Mannulus ZS	-0.447±0.461	-0.637±0.581	0.089
Tannulus ZS	-0.458±0.511	-0.722±0.518	**0.020**
LA ZS	0.700±0.585	0.109±0.833	**<0.001**
TAPSE ZS	-0.322±0.829	-0.809±0.784	**0.007**
Ascending Aorta ZS	-0.371±0.768	0.140±0.796	**0.003**
Pulmonary flow	1.045±0.059	1.044±0.090	0.941

Statistics: Student's t-test. RVDD ZS: end-diastole right ventricle diameter Z-score; IVSd ZS: diastolic interventricular septum diameter Z-score; LVIDd ZS: end-diastole left ventricle diameter Z-score; LVIDs ZS: end-systole left ventricle diameter Z-score; EF: ejection fraction; SF: shortening fraction; MPA ZS: main pulmonary artery diameter Z-score; LA ZS: left atrium diameter Z-score; TAPSE ZS: tricuspid annular plane systolic excursion Z-score. Statistically significant values are indicated in bold.

p=0.089, and p=0.941, respectively). The diastolic interventricular septum thickness Z-score (IVSd ZS), tricuspid annulus Z-score (Tannulus ZS), left atrium diameter Z-score (LAD ZS), and tricuspid annular plane systolic excursion Z-score (TAPSE ZS) values of the PE group were significantly lower than those of the control group (p=0.042, p=0.020, p<0.001, and p=0.007, respectively). The ascending aortic sinus Z-score (aorta ZS) of the PE group was significantly higher than that of the control group (p=0.003) (Table 2).

According to the results of the risk analysis that we carried out with the logistic regression analysis method for MVP and TVP development in the PE patients, MVP development increased 16.56-fold, and TVP development increased 18.66-fold in the PE patients (p<0.001 and p<0.001, respectively) (Table 3).

DISCUSSION

PE constitutes 90% of congenital thoracic cage deformities, and it is seen frequently in men[8]. In our study, similar to the literature, 80% of the PE deformity cases consisted of male children. In this deformity, while the first and second costal cartilages and the manubrium are usually in their normal positions, the lower costal cartilages adhering to the sternum and the body of the sternum are concave. Although the affected cartilages are curbed inward, the costae at the lateral of the costochondral junction remain unaffected. In approximately half of all cases, the sternum forms a curvature, mostly to the right on the frontal[9].

It can lead to reduced cardiopulmonary functions and physical capacity by causing a lower thoracic volume and compression in the heart. While its symptoms are rarely seen in early childhood, an increase can be observed in symptoms as the person gets older[9].

As the thoracic wall is flexible in young patients, the heart moves to the left, and this situation allows the compression on the heart to decrease to some extent. However, throughout the aging process, the flexibility of the thoracic wall decreasesand it becomes harder, the leftward deviation of the heart decreases, and compression on the heart and symptoms increase[10]. In the early adolescence period, complaints of early exhaustion are seen after started doing exercise. Moreover, exercise dyspnea, reduced stamina, chest pain, palpitations, exercise-triggered wheezing, and frequent upper respiratory tract infections can be seen[11]. While most of our patients did not have any complaints, the most common complaint in patients with symptoms was shortness of breath. Similar to the cases in the literature, in

Table 3. Regression values of mitral valve prolapse and tricuspid valve prolapse.

	OR	95%CI	p-value
MVP	16.56	4,937–55,563	<0.001
TVP	18.66	2,249–154,956	<0.001

Statistics: Logistic regression analysis. MVP: mitral valve prolapse; TVP: tricuspid valve prolapse.

our clinical study, it was found that the number of complaints increased with increasing age.

Cardiovascular functions can become more complicated with the displacement of the heart and rotation of the large blood vessels as the compression created by the sternum increases[12]. In our study, 8 (22%) patients had malposition on standard ECHO examination. This deformity, which is characterized by the depression of the sternum, may result in a significant decrease in the posterior-anterior diameter of the thoracic cage. This decrease may lead to a reduction in the stroke volume during systole as a result of the insufficient expansion of the heart during diastole[12]. This can lead to a failure in meeting the increased metabolic need, especially during exercise, and result in a lower LAD ZS value, as found in our study.

CHDs that can accompany PE are a vascular ring, pulmonary stenosis, atrial septal defect (ASD) primum, idiopathic hypertrophic subaortic stenosis, ASD secundum, total anomalous pulmonary venous return, transposition of the great arteries, tetralogy of Fallot, complete atrioventricular canal, tricuspid atresia, dextrocardia, truncus arteriosus, Ebstein anomaly, ventricular septal defects, aortic regurgitation, and patent ductus arteriosus. In our study, we found CHD in 3 (3.2%) patients: small secundum ASD that did not require intervention in one patient, and secundum ASD that required transcatheter closure in one patient and patent ductus arteriosus (PDA) in another patient.

As this deformity is in a concave form, it reduces venous return and myocardial perfusion by exerting compression on both the lungs and the ventricle. The compression of the right atrium and the right ventricle with an increased sternal depression index value in severe PE cases can be easily detected in echocardiographic tests[13]. TAPSE ZS is a value that shows the systolic function of the right ventricle[14]. In our study, the PE group had significantly lower tricuspid annulus diameter, tricuspid valve diameter, and TAPSE ZS values, which could be secondary to the increased sternal depression mentioned above. Furthermore, in the ECHO examination of 8 (22%) patients, malposition secondary to this sternal compression was detected.

In agreement with the information in the literature, in our study, the PE group had significantly higher rates of cardiac pathology, cardiac malposition, MVP, MI, TVP, and CHD ($p<0.001$, $p<0.001$, $p<0.001$, $p<0.001$, $p<0.001$, and $p=0.027$, respectively) (Table 1). Besides, when we conducted a risk analysis regarding MVP and TVP development in the PE patients using logistic regression analysis, we determined that the PE patients had a 16.56-fold increased risk of developing MVP and an 18.66-fold increased risk of developing TVP ($p<0.001$ and $p<0.001$, respectively) (Table 3).

In advanced-stage PE patients, as a result of the reduced volume based on the different positions of the heart, it is possible to observe tachycardia, right axis deviation in ECG, ST-segment depression (68% of patients), functional systolic murmurs originating from the compression of the left ventricular outflow tract (18% of patients), MVP (7–20% of patients), and conduction defects (branch blocks) (16% of patients)[15]. In our study, none of the patients showed significant ECG findings. The ECG findings that have been reported in the literature may be developing most probably due to the chronic effects of long-lasting sternal compression at older ages.

The fact that our hospital is a tertiary health center may have led to the inclusion of patients with severe thoracic deformities in the sample. Despite these limitations, the fact that this study is one of the very few studies in the literature that made a cardiac assessment of pediatric PE patients makes this study valuable.

Consequently, our study demonstrated that the prevalence of cardiac pathologies in pediatric PE patients was higher than the healthy control group. Accordingly, we recommend that pediatric PE patients are referred to pediatric cardiology outpatient clinics for the early diagnosis of cardiac pathologies.

AUTHORS' CONTRIBUTIONS

AA: Conceptualization, Data curation, Formal Analysis, Writing – original draft, Writing – review & editing. **UUG:** Data curation, Formal Analysis, Writing – original draft. **ŞG:** Conceptualization, Writing – review & editing.

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