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Göğüs Duvarı Deformitesi Olan Çocukların Demografik, Klinik ve Ekokardiyografik Özellikleri

Demographic, Clinical and Echocardiographic Characteristics of Children with Chest Wall Deformities

Eyüp Aslan[1], Ahmet Sert[2], Fatih Şap[3], Ebru Aypar[4], Dursun Odabaş[3]

1Denizli Devlet Hastanesi, Çocuk Kardiyolojisi, Denizli
2Konya Eğitim ve Araştırma Hastanesi, Çocuk Kardiyolojisi, Konya
3Necmettin Erbakan Üniversitesi, Çocuk Kardiyolojisi, Konya
4Hacettepe Üniversitesi, Çocuk Kardiyolojisi, Ankara

Öz:

***Giriş ve Amaç*:** Göğüs duvarı deformitesi olan çocukların demografik, klinik ve ekokardiyografik özelliklerinin değerlendirilmesi amaçlandı.

Yöntem ve Gereçler: Çocuk kardiyoloji ünitesinde üç yıl süreyle göğüs duvarı deformitesi tanısı alan hastalar retrospektif olarak değerlendirildi.

***Bulgular**:* Ortanca yaşı 8 olan hastaların ortalama yaşı 7.6 ± 4.5 idi. Göğüs duvarı deformitesi olan yüz altmış dört (% 80) hastanın ekokardiyografik değerlendirmesi normaldi. Ekokardiyografik tanıları 10 (% 4.87) atriyal septal defekt, 8 (% 3.90) mitral kapak prolapsusu, 7 (% 3.41) hafif mitral yetersizliği, 5 (% 2.44) darlık olmayan biküspit aort kapağı, 4 (% 1.95) ventriküler septal defekt, 2 (% 0.98) hafif aort kapak yetersizliği, 2 (% 0.98) dekstrokardi, 2 (% 0.98) aort koarktasyonu ve 1 (% 0.49) kompleks kalp defekti (pulmoner atrezi ve ventriküler septal defekt) idi. Ekokardiyografi ile değerlendirilen sağ kalbe bası bulgusu 15 (% 7.3) idi.

***Tartişma ve Sonuç**:* Göğüs duvarı deformitelerinin doğuştan kalp hastalıkları ile ilişkili olması ve kalbe bası bulgusu oluşturabilmeleri sebebiyle ekokardiyografi ile değerlendirilmesi defektlerin en uygun şekilde yönetilmesini sağlayacaktır.

***Anahtar Kelimeler**:* Çocuk, ekokardiyografi, göğüs duvarı deformitesi

***Türkçe Kısa Başlık**:* Göğüs Duvarı Deformitesi Olan Çocukların Özellikleri

Sorumlu yazar yazışma adresi: Eyüp ASLAN. Denizli Devlet Hastanesi. Çocuk Kardiyoloji Servisi, Denizli
E-posta:

SUMMARY

Introduction*:* To determine the demographic, clinical and echocardiographic characteristics of children with chest wall deformities.

Methods*:* The patients diagnosed with chest wall deformities were investigated retrospectively in the pediatric cardiology unit over a period of three years. The study enrolled 205 children under the age of 18 years diagnosed with chest wall deformities.

Results**:** The mean age of the patients was 7.6 ± 4.5 years with a median of 8 years. One hundred and sixty four (80 %) patients with chest wall deformities were found to have normal echocardiography results. Incidental echocardiographic diagnoses included 10 (4.87%) atrial septal defects, 8 (3.90%) mitral valve prolapses, 7 (3.41%) mild mitral regurgitations, 5 (2.44%) bicuspid aortic valve without aortic valve stenoses, 4 (1.95%) ventricular septal defects, 2 (0.98%) mild aortic regurgitations, 2 (0.98%) dextrocardia, 2 (0.98%) coarctations of the aorta and 1 (0.49%) complex cardiac defect (pulmonary atresia and VSD). Compression in the right heart was evaluated by echocardiography 15 (7.3%).

Discussion and Conclusion*:* Because chest wall deformities are associated with congenital heart defects in children and may cause compression in the right heart, evaluation by echocardiography can ensure an optimal management of the defects.

Keywords: Child, echocardiography, chest wall deformity

Running Head: Characteristics of Children with Chest Wall

Introduction

Chest wall deformities occur when there is anomalous skeletal development and/or formation of the thoracic cavity [1]. Rib cage overgrowth leads to depression or protuberance of the sternum [pectus excavatum (PE) and pectus carinatum (PC)] and accounts for greater than 90% of congenital chest wall deformities. Pectus excavatum is accounting for approximately 85% of all the cases. It presents in approximately one of every 300 to 400 births. Pectus excavatum is most frequently occur during early childhood and becomes much more severe during adolescent growth years until full skeletal maturity is achieved [1]. Most patients with PE are asymptomatic, but in severe cases they have dyspnea with exertion, progressive loss of endurance, tachycardia, palpitations and chest discomfort [2]. The patients with PE should be evaluated by echocardiography. Mitral valve prolapse (MVP) and atrial septal defect (ASD) are the most in the patients with PE [3]. Although intracardiac anomalies are not often, if presents, the degree of compression of right ventricle, right atrium and the great vessels, arises from the heart, should be determined. Surgical repair of PE is indicated when the deformity is severe and when the defect has a significant impact on the patient's physicology. According to multiple studies, surgery can improve ejection fraction if cardiac compression is evident and improve psychosocial function [4].

Pectus carinatum (PC) is the second most common congenital abnormality of the chest wall. The PC is a disease that most often is a cosmetic problem, with no impact on a cardio-respiratory system [5].

Methods and Materials

Study population: This study enrolled children under the age of 18 years who were referred with chief complaint of chest deformity to our paediatric cardiology unit, over a period of three years (February 2010-June 2013). The study was approved by the local ethics committee and written informed consent was obtained from parents of patients. Pectus deformities were identified by physical examination of the chest.

Echocardiogram was obtained from reports generated at the time of the study. Diagnoses that are associated intracardiac anomalies with chest wall deformity included MVP and ASD or possible cardiac compression.

Statistical Analysis: Clinical symptoms were expressed as counts and percentages. Statistical analyses were performed using a computer software package (SPSS for Windows, version 15.0).

Results

During the study period, 205 patients presented or referral to our unit with chief complaint of chest wall deformity. There were 145 (70.7%) boys and 60 (29.3%) girls. The mean age of the patients was 7.6 ± 4.5 years with a median of 8 years and a range of 0-17 years. The patients with chest wall deformities were revealed, 142 (69.3%) pectus excavatum, 56 (27.3%) pectus carinatum and 7 (3.4%) were both, pectus excavatum and carinatum.

Echocardiograms were performed in all patients in order to diagnose a possible of an intracardiac anomalies and depression of the right heart. One hundred and sixty four (80 %) patients with pectus deformity revealed normal echocardiography. Incidental echocardiographic diagnoses included 10 (4.87%) atrial septal defect, 8 (3.90%) mitral valve prolapsus, 7 (3.41%) mild mitral regurgitation, 5 (2.44%) bicuspid aortic valve without aortic valve stenosis, 4 (1.95%) ventricular septal defect, 2 (0.98%) mild aortic regurgitation, 2 (0.98%) dextrocardia, 2 (0.98%) coarctation of the aorta and 1 (0.49%) complex cardiac defect (pulmonary atresia and VSD). Depression to the right heart was evaluated by echocardiography 15 (7.3%). All of patients were consulate with thoracic surgeon department. None were undergone to surgery. Follow-up cardiologic evaluations were recommended in these patients.

Discussion

While the prevalence of CHD is 0.8% live births in general population, CHD with pectus deformity is relatively common. We found 22.9% CHD in the patients with pectus deformities. Shamberger reported that, among 20,860 infants and children with CHD seen at their institution, 36 (0.17%) had associated anterior thoracic deformities, the rate of pectus deformities in the patients visited to our pediatric cardiology clinic among 12636 patients was 205 (1.62%).

Acyanotic congenital heart defects, MVP and ASD are the most common in PE. In our study, the most intracardiac defect was atrial septal defect (5,4%). The rate of this anomaly was reported 2.1% and 2.8% by Simsek et al [6] and Akcali et al, [7] respectively. The incidence of MVP in the patients with PE reported in the literature is from 2.9% to 23% [3,8] while the rate is 3-6% in the general population. In the present study MVP has found 4,4% with pectus deformity.

Pectus excavatum is present in approximately two thirds of patients with Marfan syndrome (MS) which is a connective tissue disorder. In MS progressive aortic dilatation, usually maximal at the sinus of valsalva, associated with aortic valve incompetence leads to aortic dissection or rupture and is the principal cause of mortality, but MVP with incompetence may be significant [9]. In our study there were only one case with MS who had mitral valve prolapsus and normal aortic root.

Cyanotic CHD is uncommon with pectus deformities. There were reported fallot tetralogy with PE [10]. Pulmonary atresia with VSD was the only CHD in the present study.

While, Coln et al. [11] demonstrated that 95% of 123 patients had cardiac compression with pectus excavatum before surgical correction, in Guntheroth's metaanalysis [12] no cardiac dysfunction was noticed in resting teenagers. There were 7.3% of cases with PE had cardiac compression in our study, but none of them necessitated surgery.

In conclusion, pectus deformities are associated some of cardiac anomalies and could be make cardiac compression to the heart. Evaluation by an echocardiography resulted in detected and management early of these anomalies.

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