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Çocukluk Yaş Grubunda İzole Pulmoner Kapak Yetersizliğinin Sağ Ventrikül Diyastolik Fonksiyonlarına Etkisi

Right Ventricular Diastolic Functions in Pediatric Patients with Isolated Pulmonary Valve Regurgitation

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ÖZ

GİRİŞ ve AMAÇ: *İzole pulmoner kapak yetersizliği (IPKY) nadir görülen bir klinik durumdur ve genellikle yıllarca iyi tolere edilir. IPKY'de sağ ventrikül diyastolik fonksiyonlarını değerlendirmek istedik.*

YÖNTEM ve GEREÇLER: *Çalışma grubu, kurumumuza yönlendirilen 3 kız ve 10 erkek çocuk hastadan oluşuyordu. Hastalar 5-16 yaş arasındaydı. Bu hastalar yaş ve cinsiyete göre benzer bir grup olan 27 sağlıklı çocukla karşılaştırıldı. Tüm hastalar için ayrıntılı fizik muayene, elektrokardiyogram, ekokardiyografi ve akciğer grafisi çekildi.*

BULGULAR: *Çalışmamızda grup (grup I) 13 IPKY hastasından oluşuyordu. Kontrol grubunda (grup II) 6-19 yaşları arasında 27 çocuk (15 kız, 12 erkek) vardı. Grup I ve II arasında pik erken diyastolik akım hızlarında (E) istatistiksel olarak anlamlı fark vardı. Gruplar arasında pik geç diyastolik akım hızı (A) ile istatistiksel olarak anlamlı fark yoktu. Grup I'de Grup II ile karşılaştırıldığında E: A oranı anlamlı olarak azdı. Grup II ile karşılaştırıldığında grup I'de izovolemik gevşeme süresi (IVRT) uzundu, ancak gruplar arasında istatistiksel olarak anlamlı fark yoktu. Gruplar arasında karşılaştırıldığında; grup I'de E hız zaman integrali (VTI E) az, A hız zaman integrali (VTI A) aynı ve VTI E / A azalmıştı.*

TARTIŞMA ve SONUÇ: *Her ne kadar IPVR'nin iyi huylu bir lezyon olduğu kabul edilmese de bu hastalarda sağ ventrikül diyastolik disfonksiyonunun olduğunu gördük. Bu arada, klinik semptomlar ortaya çıkmadan önce aritmi ve sağ taraflı yetmezliğin belirlenmesi önemlidir.*

Anahtar Kelimeler: *Pulmoner kapak, yetersizlik, diyastolik fonksiyon.*

ABSTRACT

INTRODUCTION: *Isolated pulmonary valve regurgitation (IPVR) is a rare clinical entity is usually well tolerated for many years. We wanted to evaluate right ventricular diastolic functions in IPVR.*

METHODS: *The study group was consisted of 3 girls and 10 boys who were referred to our instution. Patients were aged between 5-16 years. These patients compared with 27 age and sex matched healthy children. All patients detailed physical examination, electrocardiogram, echocardiography and chest X-ray were recorded.*

RESULTS: *In our study the group (group I) was consisted of 13 IPVR patients. In the control group (group II) there were 27 children (15 girls, 12 boys) aged between 6-19 years There was statistically significant difference in peak early diastolic flow velocity (E) between group I and II. There was no statistically significant difference in peak late diastolic flow velocity (A) between groups. The E: A ratio was significantly decreased in group I when compared with group II.Isovolemic relaxation time (IVRT) was prolonged in group I when compared with group II but it was not statistically significant different between groups. When compared between groups; in group I the E velocity time integral (VTI E) was decreased, A velocity time integral was (VTI A) was unchanged and VTI E/A was decreased.*

DISCUSSION and CONCLUSION: *Although it is consent that IPVR is a benign lesion we found that right ventricular diastolic dysfunction in this patients. Worthwhile it is important to determine the arrythmias and right sided failure before the clinical symptoms appear.*

Keywords: *Pulmonary valve, regurgitation, diastolic function.*

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INTRODUCTION

Isolated pulmonary valve regurgitation (IPVR) is a rare clinical entity first described in 1936 by Kissin (1). IPVR can occur either a congenital or acquired basis. Congenital IPVR is more common and may result because of absence or rudimentary development of the pulmonic valve (2-3), a supernumerary valve cusp (1), bicuspid pulmonary valve (4)and dilatation of the main pulmonary artery. The latter may occur as a primary lesion (idiopathic) or in association with other major systemic defects, eg, Marfan syndrome or Ehlers-Danlos syndrome (5). IPVR can also occur in patients with severe thoracic cage deformity. This situation distorts the right ventricular outflow tract (6). Acquired IPVR is relatively uncommon and can be encountered under various clinical situation (7). The most common reason is surgical pulmonary valvulotomy or balloon dilatation to correct pulmonary valvular stenosis in tetralogy of Fallot patients (8). The three pathogenetic factors common to most patients with IPVR are retrograde blood flow from the pulmonary artery into the right ventricular outflow tract, dilatation of the right ventricle (7). The regurgitant blood flow augments diastolic volume resulting in increased transvalvular flows. Clinical progression consists of right ventricle dilatation, prolonged QRS duration, tricuspid regurgitation, right ventricular systolic dysfunction (9). Pulmonary regurgitation is usually well tolerated for many years (9-10). Clinical manifestations comprise exercise intolerance, congestive heart failure, atrial and ventricular arrythmia and sudden cardiac death when there is a right ventricular dysfunction clinical heart failure sypmptoms occur (9). The treatment of pulmonary regurgitation depends on the underlying etiology.

In acquired pulmonary regurgitation, patients usually symptomatic and ventricle dysfunction occur in early period (8). In congenital pulmonary valvular regurgitation patients are free of symptoms and are asymptomatic for years. There are some studies in acquired pulmonary valve regurgitation for searching right ventricle diastolic functions. But we could not find any studies searching right ventricular diastolic functions in IPVR. Therefore we wanted to evaluate right ventricular diastolic functions in congenital pulmonary valve regurgitation patients and wanted to compare with healthy subjects.

MATERIALS and METHODS

Study population; The study group was consisted of 13 children (3 girls, 10 boys) who were referred to our instution because of heart murmur. Patients were aged between 5-16 years. Each were healthy and had a normal growth-up period. These patients compared with 27 (15 girls, 12 boys) age and sex matched healthy children. For all patients detailed physical examination, electrocardiogram and chest X-ray were performed. Heart rate, ax, rythm, PR, QRS, QT intervals were recorded.

Doppler examination; We used a commercial system (Toshiba) with a 3-5 MHz transducer to provide simultaneous cross sectional echocardiography and colour Doppler imaging. Examination was undertaken with subjects supine position. The same investigator performed the echocardiographic study to all patients. We used the high parasternal short axis view for the pulmonary valve. For the tricuspid valve we recorded traces in the right ventricular inflow view and parasternal and apical four chamber views. If the flow signal in colour Doppler flow mapping showed reversed flow away from the valve when the valve was closed regurgitation is was said to be present Signals of very short duration detected only at the time of valve closure were not regarded as true regurgitation. Studies with pulsed and continuous wave Doppler echocardiography were performed to confirm the presence of regurgitation and to measure the maximal velocity. Only patients with more than 1.5 m/sn pulmonary flow regurgitation were taken to the study. Tricuspid flow velocities peak initial velocity (E), velocity at time of atrial contraction (A), E:A, and the velocity time integral of peak initial velocity (VTI E), velocity time integral of atrial contraction (VTI A) and the ratio of integrals (VTI E:A) were measured.

RESULTS

In our study (group I) was consisted of 13 isolated pulmonary valve regurgitation patients. These 13 children were aged between 5-16 years (medium 10.6±3.4 years).There were 3 girls and 10 boys.

Echocardiographically pulmonary valve regurgitation was obtained only.

In the control group (group II) there were 27 children (15 girls, 12 boys) aged between 6-19 year. (medium 12.4±4.06 years). There was no statistical difference between groups according to ages and body surface area ($p<0.01$) (Table I).

Table 1. Demographic features of the groups

	Group1	Group2	**Total**
Number	13	27	**40**
Age	10.6±3.47	12.4±4.06	**11.85±3.93**
Male	10(%77)	12 (%44)	**22**
Female	3(%23)	15 (%56)	**18**
Body mass area *(m^2)	**1,16±0.31**	**1.32±0.32**	**1.26±0.32**

Pulmonary valve regurgitation flow rate was between 1.5-2.57 m/sn. In both groups tricuspid valve E, A, E:A, VTI E, VTIA, VTI E:A values measured. E value was decreased in group I. There was statistically significant difference in peak early diastolic flow velocity (E) between group I and II. There was no statistically significant difference in peak late diastolic flow velocity (A) between groups. The E:A ratio was significantly decreased in group I when compared with group II (Table II).

Table 2. **Tricuspid E, A, E / A values of groups**

	E (cm/second)	A (cm/second)	**E/A**
Group I	69.23±13.2	48.57±4.8	**1.42±0.26**
Group II	**79.29±12.5**	**49.88±8.5**	**1.60±0.24**

Isovolemic relaxation time (IVRT) was prolonged in group I when compared with group II but it was not statistically significant different between groups. When compared between groups; in group I the E velocity time integral (VTI E) was decreased, A velocity time integral was (VTI A) was unchanged and VTI E/A was decreased. There was no significant correlation between pulmonary valve regurgitation flow rate and right ventricular diastolic function parameters (Table III).

Table 3. **E, A, E / A speed-time integrals of groups**

	VTI E	VTIA	**VTI E/A**
Group I	8.64±2.27	4.90±0.76	**1.78±0.53**
Group II	**10.97±3.46**	**4.80±1.37**	**2.38±0.77**

DISCUSSION

Isolated pulmonary valve regurgitation is a rare clinical entity (7). While in the past the diagnosis of pulmonary valve regurgitation was confirmed only by cardiac catheterization. Abbott recorded eight cases of congenital pulmonary valve regurgitation in a study of 1000 autopsy cases (11).There are a few cases diagnosed by cardiac catheterization. More patients are now being described in whom the clinical diagnosis is confirmed by Doppler echocardiograpy. Today catheterization only used for severe cases. Doppler echocardiography is a sensitive and spesific technique for detecting valve regurgitation (12). We also used Doppler echocardiography to evaluate our patients.

Isolated pulmonary valve regurgitation is more commonly accompanies other cardiovascular diseases, especially pulmonary stenosis or pulmonary hypertension, its presence is an indication to search other cardiovascular diseases (13). We described 13 pulmonary valve regurgitation patients in this study and it was the only cardivascular defect.

Isolated pulmonary valve regurgitation is usually compatible with many decades of normal life. Patients often remain free of overt symptoms until marked right ventricle dilatation and systolic dysfunction ensue (9,10). When patients become symptomatic, right ventricle dysfunction is usually well established and may have become irreversible. In patients with isolated congenital pulmonary regurgitation and otherwise normal hearts, symptoms are rare before the age of 30. After the age of 40, however, patients manifest symptoms of right heart failure (9). In our study all patients were free of symptoms.

Other investigations are electrocardiogram and chest X-ray. Most patients with pulmonary regurgitation have sinus rythm, but arrhythmias can be present. QRS duration and QRS change have prognostic implications for malignant arrythmia and

sudden cardiac death in these patients (9,13,14). Patients with severe pulmonary regurgitation characteristically have dilatation of the pulmonary trunc and central pulmonary arteries. There is also right ventricle enlargement. We performed electrocardiogram and telecardiography for all patients. Electrocardiographic study and telecardiograms were all normal.

The filling characteristics of right side of the heart is shown by the tricuspid flow velocity curve. The diastolic velocities are slightly lower than those of the mitral valve. The normal tricuspid flow velocity curve, similar to the mitral flow velocity curve, consists of an early diastolic velocity that denotes rapid fillling, a rapid descent during middiastole, and an increase in velocity during atrial contraction The peak velocity in normal persons changes during the respiratory cycle, with an increase during inspiration and a decrease during expiration. Abnormalities of tricuspid flow velocity curves are similar to those of the mitral flow velocities. Patients with abnormal relaxation of the right ventricle may show a prolongation of the deceleration time and a decrease in the E:A ratio (15). By performing Doppler echocardiography we measured tricuspid flow velocities

Pulmonary regurgitation is an important problem in repair of tetralogy of Fallot. Its effects on right ventricular diastolic function in children are unknown. In a study 19 children with repair of tetralogy of Fallot and 12 healty subjects were evaluated. Tricuspid time to peak velocities and duration of pulmonary regurgitation were studied by Doppler echocardiography. Finally they found that impaired relaxation and restriction to filling affects right ventricular function in children with repair of tetralogy of Fallot and pulmonary regurgitation (16). In a similar study Cardoso et al compared 30 patients who had undergone to repair of tetralogy of Fallot with 30 healthy children. The velocity of the A wave and the E/A ratio for the tricuspid valve showed significant differences between the groups. Nineteen patients (63.3 %) patients had restrictive right ventricular physiology, which had a longer postoperative period, longer duration of the QRS complex, and a lower E/A ratio in inspiration These studies showed the right ventricle diastolic dysfunction but all these studies performed in acquired pulomary valve regurgitation patients. In our study we found that the velocity of the E wave and E/A ratio was decreased and the time integral of E wave and the time integral of E/A was decreased (17,18). In near past a study was performed on pigs whose pulmonary regurgitation created. Echocardiographic study revealed that decreased right ventricule longitudinal function and increased compensatory septal pumping (19).

Isolated pulmonary valve regurgitation is a rare condition. Although there is only case series in the literature and it is reported to be benign in childhood, a 21-year-old young woman with isolated pulmonary valve regurgitation had right ventricular diastolic dysfunction in hospital admission because of syncope. Therefore, close follow-up of patients with isolated pulmonary valve regurgitation has been reported to be beneficial (20).

In conclusion in our study we found significant differences between groups. Although it is consent that isolated pulmonary valve regurgitation is a benign lesion we found that right ventricular diastolic dysfunction in this patients. Worthwhile it is important to determine the arrythmias and right sided failure before the clinical symptoms appear. So that we thought that these patients must be folllowed by echocardiographic and electrocardiographic studies. Because we have limited patients in this study, it is needed much more studies in the future.

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