Double outlet right ventricle with intact ventricular septum

Ventrikül septumu normal çift çıkışlı sağ ventrikül

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Summary– Double outlet right ventricle is a cardiac malformation in which both the aorta and pulmonary artery arise from the right ventricle. A double outlet right ventricle with intact ventricular septum is extremely rare. A nine-day-old boy born at 38 weeks with a birth weight of 3200 g was referred due to cyanosis and murmur. The history of the patient was nonsignificant. On the physical examination, oxygen saturation of the patient on room air was 84%, and cardiac activity and a 2/6 systolic murmur at the right sternal border were found. On electrocardiography, deep q's were determined in D2, D3, and aVF. On echocardiography, left atrial isomerism, dextrocardia, wide, high venosum atrial septal defect, double outlet right ventricle with intact ventricular septum, malposition of the great arteries, and moderate pulmonary stenosis were observed. Catheter angiography was performed for diagnostic confirmation. Herein, a newborn with cyanosis and murmur diagnosed as double outlet right ventricle with intact ventricular septum is reported.

Özet– Çift çıkışlı sağ ventrikül hem aorta hem de pulmoner arterin sağ ventrikülden çıktığı bir kalp malformasyonudur. Ventrikül septumda defekt bulunmayan çift çıkışlı sağ ventrikül oldukça nadir görülür. Doğum ağırlığı 3200 gr olan ve 38 haftada doğan 9 günlük erkek bebek siyanoz ve üfürüm nedeni ile gönderildi. Hastanın öz geçmişinde özellik yoktu. Fizik muayenede sternumun sağ kenarında kalp aktivitesi ve 2/6 sistolik üfürüm bulunmaktaydı. Diğer system muayeneleri normaldi. Hastanın oda havasında oksijen yoğunluğu %84 idi. Elektrokardiyografisinde D2, D3 ve aVF'de derin q dalgaları mevcuttu. Ekokardiyografi incelemesinde sol atriyum izomerizmi, dekstrokardi, geniş yüksek venozum tipi atriyum septumu defekti, ventrikül septumunun sağlam oluşu ile birlikte çift çıkışlı sağ ventrikül, büyük arterlerin malpozisyonu ve pulmoner stenoz izlendi. Tanı doğrulanması açısından kateter-anjiyografi yapıldı. Bu yazıda, yendioğan döneminde siyanoz ve üfürümü olan, ventrikül septumu normal bulunan ve çift çıkışlı sağ ventrikül tanısı konan bir olgu sunuldu.

Double outlet right ventricle is a cardiac malformation in which both the aorta and pulmonary artery arise from the right ventricle.[1] A ventricular septal defect (VSD) is necessary for survival, since it represents the only outlet of the left ventricle.[2] Double outlet right ventricle with intact ventricular septum is extremely rare.[3] According to embryological development, VSD is expected in all conotruncal abnormalities. Displacement of the great arteries in conjunction with movement of the outlet septum results in the formation of VSD.[4] Thus far, only a limited number of double outlet right ventricle cases with intact ventricular septum have been reported. Some of these have pointed to previously existing VSD closure by various mechanisms. However, others have argued the ventricular septum was intact since birth.[4] The majority of patients in the neonatal period require urgent septostomy.[5] Any associated anomalies significantly worsen the course. Hypertensive left ventricle arrhythmogenic and ischemic potential should be considered during the selection of treatment.[6]

Abbreviations:

ASD *Atrial septal defect*
SVC *Superior vena cava*
VSD *Ventricular septal defect*

CASE REPORT

A nine-day-old boy born at 38 weeks with a birth weight of 3200 g was referred due to cyanosis and murmur. The history of the patient was nonsignificant. Family history revealed that the parents were third-degree relatives. Although the pregnancy was followed,

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she had not undergone fetal echocardiographic study. On the physical examination, oxygen saturation of the infant on room air was 84%, and cardiac activity and a 2/6 systolic murmur at the right sternal border were found. Other system examinations were normal. Teleradiography showed dextrocardia, cardiothoracic ratio of 0.70, and low perfusion in the lung fields. On electrocardiography, deep q's were determined in D2, D3, and aVF. On echocardiography, left atrial isomerism, left-handed topology, dextrocardia, wide, high venosum atrial septal defect (ASD), double outlet right ventricle with intact ventricular septum, malposition of the great arteries, and moderate pulmonary stenosis were observed (Figure 1). Additionally, fistulous lesions without any direct connection to the coronary artery, left ventricle hypertrophy, and 3rd degree mitral insufficiency were seen. Catheter angiography was performed for diagnostic confirmation. A 5 Fr sheath was inserted into the right femoral vein. It was inserted into the aorta and right ventricle, and 90/50/70 mmHg and 70 mmHg aorta and right ventricular pressure, respectively, were determined. At straight posterior-anterior position, it was seen that both great arteries completely originated from the right ventricle and the interventricular septum was intact. Because of the intact ventricular septum and mitral valve hypoplasia, it was impossible to enter the hypoplastic left ventricle. On angiography, the abnormal course of the aorta was revealed. The coronaries were observed to branch from different roots. There was no direct connection between the coronary artery and fistulous lesions seen on angiocardiography.

DISCUSSION

Ventricular septal defect is the single outlet of the left ventricle in double outlet right ventricle cases, and usually presents.[3] Double outlet right ventricle with intact ventricular septum is a very uncommon entity.[6] There are only a limited number of cases reported in the literature. The most important reason for the limited number is that the cases are lost immediately after birth. Double outlet right ventricle with intact ventricular septum was reported more in autopsy series than in living patients.[5]

Normally, in the development of double outlet right ventricle, VSD is inevitably expected since the transposition of the outlet septum is in question. Embryologically, there have been attempts to explain, by several mechanisms, why restrictive VSD or intact ventricular septum coexists with double outlet right ventricle. Explanations have included marked subaortic conal tissue, alignment of that conal tissue with a muscular interventricular septum, and the limitation of the interventricular passage by the muscle filaments, which are widened due to the hypertrophy of both ventricles.[1,2,7,8] Specifically, enlargement of the subaortic conus causes flow limitation even in the cases with wide VSD; thus, in another word, causing functional restriction.[2]

Spontaneous closure of previously shown VSDs has also been mentioned in the literature. The presence of tricuspid and mitral accessory tissues, progressive muscular tissue invasion, aneurysmal formation of

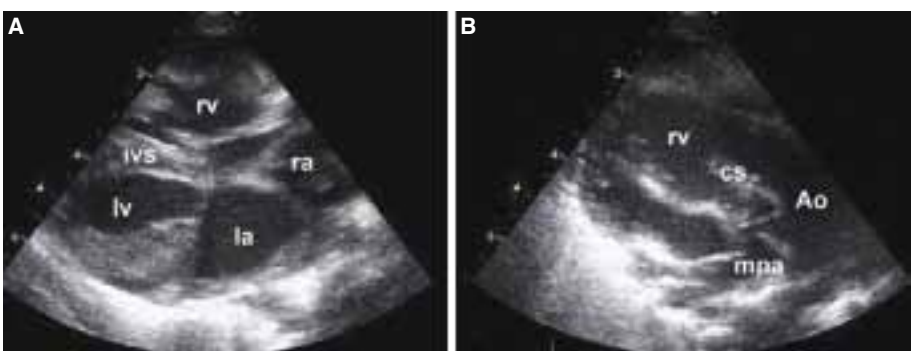

Figure 1. Double outlet right ventricle with intact ventricular septum is monitored on the 2D echocardiography in the apical four-chamber and parasternal long-axis study. **(A)** Intact ventricular septum and hypoplastic left ventricle with mitral valve are shown. **(B)** Both great arteries arise from only the right ventricle; the aorta is to the right and anterior to the pulmonary artery. la: Left atrium; ra: Right atrium; lv: Left ventricle; rv: Right ventricle; cs: Conus septum; ivs: Intact ventricular septum; Ao: Aorta; mpa: Main pulmonary artery.

the interventricular septum, endocardial proliferation and fibrosis, and occlusion by thrombus and infected vegetation are among VSD closure mechanisms.[1,9] According to Somerville et al.,[10] chances of spontaneous closure of VSDs that reach the inlet region, have subaortic or subpulmonic localization or resemble "swiss cheese" in shape are low. However, VSDs localized in the inferior portion of the tricuspid medial papillary muscle and which are limited or totally in the muscular region tend to close spontaneously.

During the closure of a previously existing VSD, changes such as fibrosis, etc. may also occur in the conduction system, which passes posteroinferiorly of the defect, and these changes may be reflected in ECG findings. According to reported cases, while the patients followed due to tetralogy of Fallot have right axis and right ventricle hypertrophy, left axis hypertrophy is observed due to the left hemi-block as the VSD is closing.[10]

In the double outlet right ventricle cases with intact ventricular septum, the pressure of the left cavities increases in utero due to the high-pressured and hypoplastic left ventricle. The patent foramen ovale flap moves with the effect of pressure in a way that limits the interatrial communication. Thus, intact ventricular septum as well as intact atrial septum or restrictive ASD are usually observed in these patients.

Patients are usually presented urgently in the neonatal term due to restrictive ASD. Unless balloon atrial septostomy or stent intervention to the interatrial septum, which maintains the mixture, is performed, patients may be lost. The clinical picture of our patient did not appear immediately in the neonatal period due to the wide, high venosum ASD; however, he was referred to the cardiology clinic following realization during the physical examination that the murmur and heart sounds were on the right. In the literature, there has been only one case in which alignment of the left atrial isomerism and wide, high venosum ASD was reported. In that case, reported by Anderson et al.,[11] left atrial isomerism and double superior vena cava (SVC) were present; continuity of the azygos was supplied by left SVC. In our case, continuity by the right SVC was maintained.

Development of the left ventricle and its structures is restrained following decrease in the blood volume passing to the left system. Another reason for the emergent and poor presentation of the patients is the accompanying anomalies belonging to the left system, such as mitral atresia, mitral stenosis, subaortic stenosis, and coarctation of the aorta. Levoatriocardinal vein presentation was reported in several cases secondary to the obstruction in front of the pulmonary vein flows.[3] Left SVC presence is not uncommon in these patients. The left SVC may be a reason for the reduction in the blood volume going to the left ventricle by limiting the mitral flow in the embryologic term. As seen in our case, the rare accompaniment of heterotaxy syndromes was reported in the literature.[11]

The high-pressured coronary fistula formation in the patients, on which the coronary circulation depends, is one of the crucial reasons for not being able to pursue biventricular repair in these patients. In treatment, VSD enlargement was considered in the patients with restrictive VSD in the first place. However, the success of this process is limited due to the proximity to the conduction system.[1,2,5,6]

Presence of subvalvular infundibular stenosis in the patients is important in terms of protection of the lung fields and affording the patient the opportunity for surgery in the long-term. However, if the subaortic conus is significant, tight band application should be avoided in those patients with concerns of systemic outlet obstruction in the long-term.[5] Since our patient had pulmonary stenosis, lung fields were already protected.

A second problem in the treatment is how to intervene with respect to the hypertensive and nonfunctional ventricle. While some authors accept a simple patch technique, leaving the left ventricle in its position, some prefer mitral avulsion with the concern that these ventricles might be resources for arrhythmia and fistula in the long-term.[5,6]

The large, high venosum ASD provided a chance of survival in our patient and thus did not require septostomy or septectomy. Absence of subaortic conus and moderate pulmonary stenosis made our patient suitable for bidirectional cavopulmonary shunt later. Many studies have implied that a residual hypertensive-hypoplastic left ventricle may cause long-term complications like small right ventricle in pulmonary atresia with intact ventricular septum.[5,6] Although presence of a large ASD and mitral insufficiency help in left ventricle decompression, long-term complications in our patient cannot be estimated. Furthermore, malposition of the great arteries is an important point

for planning treatment. Our patient was followed for four months, and bidirectional cavopulmonary shunt was planned.

In conclusion, double outlet right ventricle with intact ventricular septum is extremely rare. The degree of left ventricular and mitral hypoplasia, restrictive interatrial communication and additional abnormalities can influence the mortality and morbidity. Therefore, optimal treatment modalities should be selected according to the patient status.

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Anahtar sözcükler: Aort darlığı/komplikasyonlar; çocuk; koroner dolaşım; siyanoz/etiyoloji; kalp defekti, doğumsal/komplikasyonlar; kalp septal defektleri, ventriküler/etiyoloji; kalp kapak hastalıkları/komplikasyonlar; bebek, yenidoğan.