

Supracardiac Type Total Anomalous Pulmonary Venous Return with Obstruction, a Rare Combination: Case Report

Obstrüksiyonla Birlikte Suprakardiyak Tip Total Anormal Pulmoner Venöz Dönüş, Nadir Bir Kombinasyon

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ABSTRACT Total anomalous pulmonary venous return (TAPVR) defines the anomaly in which the pulmonary veins have no connection with the left atrium. TAPVR is a rare congenital cardiac anomaly. TAPVR is divided into four groups according to the site of connection. Pathophysiologically, these four types are subclassified according to whether the pulmonary venous return is obstructed or nonobstructed. Early diagnosis and surgery are life saving, especially in newborns with pulmonary venous obstruction, which is most commonly seen in infracardiac type. A 3-day-old male infant was referred to our unit for cyanosis, who was diagnosed with supracardiac type TAPVR with obstructed pulmonary venous return and moderate coarctation of aorta. In this report a newborn with obstructed supracardiac type TAPVR is presented with echocardiography and catheterization findings, because of it's a rare disease.

Key Words: Pulmonary veins; infant, newborn; cyanosis

ÖZET Total anormal pulmoner venöz dönüş (TAPVD), pulmoner venlerin sol atriyum ile birleşememe anomalisi olarak tanımlanır. TAPVD nadir bir konjenital kardiyak anomalidir ve birleşme yerine göre dört gruba bölünür. Patofizyolojik olarak bu dört tip pulmoner venöz dönüşün obstrükte olup olmamasına göre alt gruplara ayrılır. Özellikle pulmoner venöz obstrüksiyonlu yenidoğanlarda, ki bunlarda obstrüksiyon daha çok infrakardiyak tipte görülür, erken tanı ve cerrahi tedavi hayat kurtarıcıdır. Siyanoz ile başvuran üç günlük erkek hasta suprakardiyak tip TAPVD ile birlikte pulmoner venöz obstrüksiyon ve orta derecede aort koarktasyonu tanısı aldı. Bu yazıda, nadir görülen bir hastalık olmasından dolayı obstrükte suprakardiyak tip TAPVD ekokardiyografi ve kateterizasyon bulguları ile sunuldu.

Anahtar Kelimeler: Akciğer ven; bebek, yenidoğan; siyanoz

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Total anomalous pulmonary venous return (TAPVR) defines the anomaly in which the pulmonary veins have no connection with the left atrium. It accounts for only 1-2.6% of congenital heart diseases.¹ TAPVR is divided into four groups according to the site of connection, as follows: Type I) anomalous connection at the supracardiac level (50%), type II) anomalous connection at the cardiac level (to the coronary sinus) (30%), type III) anomalous connection at the infracardiac level (15%), type IV) anomalous connection at two or more of the above levels (5%). Pathophysiologically, these four types are subclassified according to whether the pulmonary venous return is obstructed or nonobstructed. Although obstruction

may occur with any anatomic type of TAPVR, the highest incidence is encountered with the infracardiac type. Obstructed TAPVR is rarely seen with supracardiac type.² We report a case of a newborn with obstructed supracardiac type TAPVR, because of his rarely anomaly. The diagnosis was suspected on echocardiography. Cardiac catheterization and angiography was done for definitive diagnosis.

CASE REPORT

A 3-day-old male infant was referred to our unit for evaluation of cyanosis and respiratory distress. He was born via cesarean section at term. Prenatal and natal history were uncomplicated. His weight was 3330 g and height was 50 cm. On physical examination the baby had central cyanosis. Oxygen saturation was 54% on pulse oxymetry. He had a heart rate of 144/min and respiratory rate of 70/min. On auscultation, he had no cardiac murmur. His breath sounds were normal. Electrocardiogram showed normal sinus rhythm, normal axis, incomplete right bundle branch block. Telecardiography was normal. Echocardiography revealed high settled atrial septal defect (ASD), muscular trabecular ventricular septal defect (VSD), ductus with a right-to-left shunt, tricuspid insufficiency (4.2 m/s) and moderate aortic coarctation. Pulmonary veins were draining into left innominate vein via a vertical vein (Figure 1). Cardiac catheterization and angiography were performed for confirming the diagnosis and to describe the site of pulmonary venous obstruction. Cardiac catheterisation data is shown in Table 1. Pulmonary artery injection in AP view demonstrated pulmonary venous drainage into innominate vein and superior vena cava (SVC) through vertical vein. The obstruction between pulmonary venous chamber and vertical vein was observed in vertical vein injection (Figure 2 A, B). These findings suggested obstructed supracardiac type of TAPVC, ASD, VSD, pulmonary hypertension (PH), patent ductus arteriosus (PDA) and moderate coarctation of the aorta (CoA). Therefore, the patient was referred to another clinic for surgery. He has been followed up in outpatient clinic after a successful operation.

DISCUSSION

TAPVR is an anomaly that results from failure of the common pulmonary venous chamber to become incorporated into the left atrium during embryogenesis; there is persistence of communications between the pulmonary portion of the foregut plexus and the cardinal or umbilicovitelline system of veins, resulting in the connection of all the pulmonary veins either to the right atrium directly or to the systemic veins and their tributaries. There is no connection between the pulmonary veins and the left atrium. Although pulmonary ve-

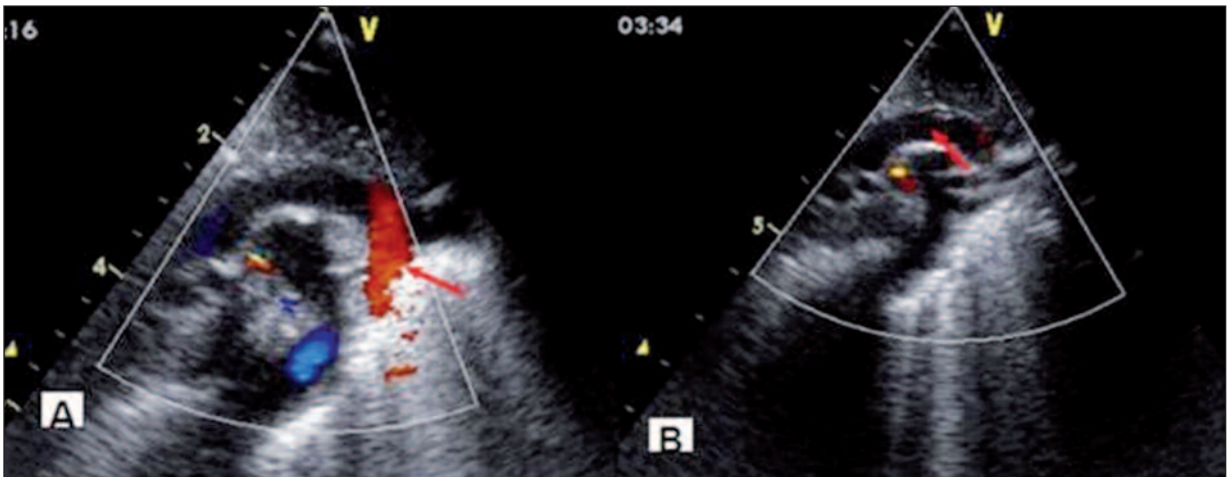


FIGURE 1A, B: Vertical and innominate veins are seen from suprasternal aspect on colour Doppler images (red arrows).

(See for colored from <http://cardiovascular.turkiyeklinikleri.com/>)

TABLE 1: Cardiac catheterization data.

Site	Pressure (mmHg)	Oxygen sat. %
SVC	-	75
IVC	-	60
RA	4	79
RV	99/5	79
PA	100/55/72	79
LV	-	69
aAo	105/60/82	70
dAo	74/51/58	-
LA	5	74

dAo: descending aorta, IVC:inferior vena cava, LA:left atrium, LV:left ventricle, PA:pulmonary artery, RA:right atrium, RV:right ventricle, SVC:superior vena cava, LIV:left innominate vein, aAo:ascending aorta

nous obstruction may occur with any anatomic type of TAPVR, the highest incidence is encountered with the infracardiac type. Obstructed TAPVR is rarely seen with supracardiac type.³

Clinical symptoms and signs depend on the patency of the interatrial communication and on the presence of an obstruction of the pulmonary venous return (PVO). The presence of intrinsic or extrinsic narrowing in the connecting vein also produces pulmonary venous obstruction. Patients with severe pulmonary venous obstruction present

in the first or second week of life with obvious cyanosis, dyspnea, difficulties with feeding even sudden death in the first 2 months. Patients without PVO mostly present within the first week of life with signs of heart failure as tachypnea, dyspnea, hepatomegaly and discrete but increasing cyanosis.⁴ Our patient presented with respiratory distress, cyanosis and hypoxia on third day of life because of pulmonary venous obstruction.

Patients with any one of the following criteria were considered having PVO: more than 50% reduction in the diameter of anomalous pulmonary vein in angiography or MRI, pressure gradient greater than 10 mm Hg across the narrowing site, high blood flow velocity detected within the pulmonary venous turbulent exceeding 1.5 m/sec by Doppler technique, the existence of pulmonary hypertension.¹⁻⁵ We observed supracardiac TAPVR on echocardiography, but we couldn't detect the site of obstruction. PVO associated with supracardiac type TAPVR was confirmed by cardiac catheterization and angiography (Figure 1).

Major cardiac malformations are seen in approximately 30% of the patients with TAPVR. These lesions include common atrium, atrial isomerism, single ventricle, truncus arteriosus, and anomalies

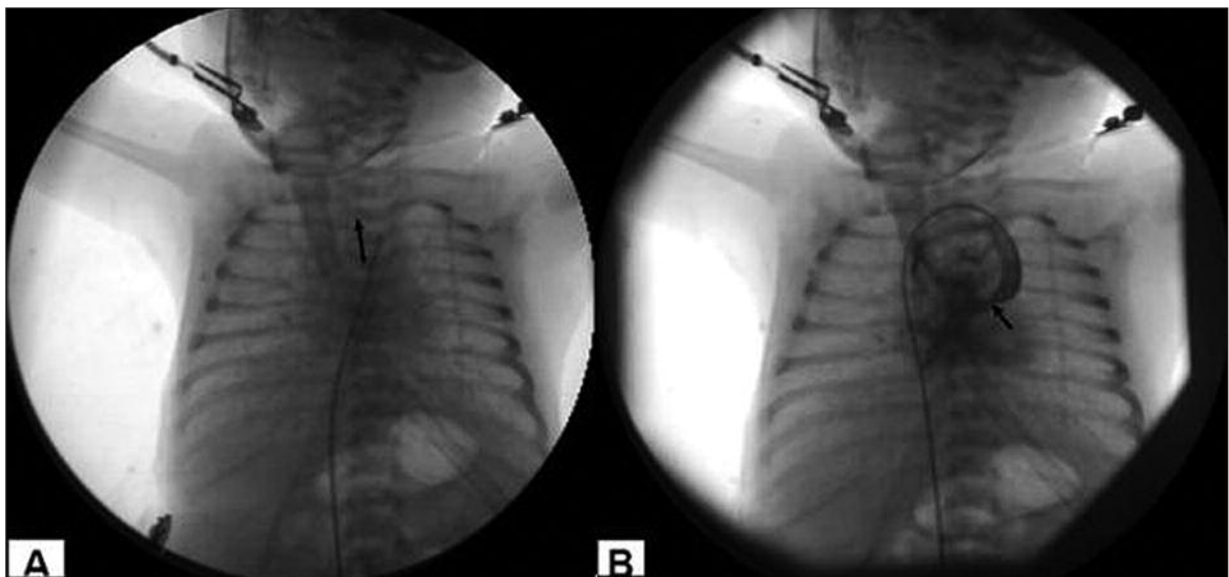


FIGURE 2A, B: Pulmonary artery injection in AP view demonstrating pulmonary venous drainage into innominate vein and SVC through vertical vein (black arrow). Vertical vein injection in AP view are showing the obstruction between pulmonary venous chamber and vertical vein (black arrow shows vertical vein).

of the systemic veins, multiple VSDs, coarctation of the aorta, vascular sling, tetralogy of Fallot.^{6,7} In our case, associated cardiac malformations were ASD, VSD, PDA and CoA. There is no known genetic predisposition in this lesion, but males are affected nearly twice as often as females.⁸ Our case was male.

Diagnosis of TAPVR is routinely made by echocardiography. Only rarely is cardiac catheterization and angiography now necessary; it is typically reserved for associated anomalies and to demonstrate the site of obstruction. Without proper surgery or treatment, the prognosis of these patients is grim in the first year of life. Although the results of operation for TAPVR can be useful, because the compliance of the left heart chambers is restricted, concluded that in a patient with preoperative pulmonary venous obstruction, the vertical vein and/or atrial septal defect should be left

open.⁹ The long-term outlook for patients with repaired TAPVR is very good, with an operative mortality rate of 10-15% (now closer to 10%) and long-term survival approaching 85%. Stenosis of the individual pulmonary veins or the pulmonary venous confluence is common postoperatively (around 15% after 1-2 years), especially in the infradiaphragmatic and miscellaneous types. However, with successful neonatal repair, the outlook for these patients is very good.¹⁰ Our patient has been followed up in outpatient clinic after a successful operation.

CONCLUSION

In conclusion, obstructive type is more rare in supracardiac group than infracardiac group of TAPVR. Prompt diagnosis and referral for surgical correction can provide successful results in specialized centers.

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