Echocardiographic diagnosis of total anomalous pulmonary venous connection of the infracardiac type

Infrakardiyak total anormal pulmoner venöz bağlantının ekokardiyografik tanısı

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Introduction

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital anomaly, corresponding to approximately 2% of all congenital heart defects (1). A recognized classification divides TAPVC into four groups according to the site of connection (2). With type I, having a supracardiac connection (50%), the common pulmonary trunk joins the left vertical vein, the innominate vein, or the superior vena cava. With type II, having a cardiac connection (30%), the anomalous pulmonary venous drainage enters the coronary sinus or flows directly into the right atrium. With type III, the site being infracardiac (15%), connection occurs to the portal vein, venous duct, or inferior vena cava below the diaphragm. With type IV, a mixed site variety (5%), the anomalous venous return occurs at several levels (1). Pathophysiologically, these four types are subclassified according to whether the pulmonary venous return is obstructed or nonobstructed. The clinical presentation and prognosis are different for the two latter conditions, being poorer for the obstructed type. Although obstruction may occur with any anatomic type of TAPVC, the highest incidence is encountered with the infracardiac type. We report a case of a newborn with infracardiac total anomalous pulmonary venous connection. The diagnosis was suggested by echocardiography and confirmed by catheter-angiography, which allowed definition of the anatomy.

Case Report

A 25-day-old male baby was admitted to our department because of respiratory distress and cyanosis. He was 3120 gram in weight and his general condition was poor. Subcostal retractions and tachypnea were present. No cardiac murmur was noted. Chest roentgenograms showed pulmonary venous congestion with interstitial and alveolar pulmonary edema. Echocardiography revealed that the right atrium, right ventricle and pulmonary trunk were enlarged. Pulmonary veins connections to left atrium could not be identified. Coronary sinus was found to be normal in size. There was a 5 mm defect in interatrial septum and right–to-left shunt was seen through the defect. On long-axis view, an echo-free space lying behind the left atrium was observed and thought to represent the common pulmonary vein (Fig. 1). Together with subcostal study, it was demonstrated, an anomalous vertical venous vessel running in the midline, across the diaphragm, and connecting with the portal vein, and draining into inferior vena cava via the ductus venosus (Fig. 2). Marked dilatation of the portal vein was also observed (Fig. 3). A diagnosis of infracardiac type TAPVC was established, to confirm the diagnosis and to exclude the possibility of mixed type TAPVC, catheter-angiography was performed. After recording of pulmonary wedge pressure (20 mmHg), contrast medium was injected into the right and left pulmonary arteries. During venous return phase, it was observed that, left upper and lower pulmonary veins came together and then joined to vertical vein, on the other hand right upper and lower pulmonary veins drained separately into the vertical vein (Fig 4 A, B). Surgical intervention on the 30th day of life confirmed the anomalous pathway of pulmonary venous drainage and demonstrated a small atrial septal defect (ASD). Complete repair was performed during deep hypothermic circulatory arrest. Despite positive inotropic support, patient died during early postoperative period, due to insufficient cardiac output.

Discussion

Total anomalous pulmonary venous connection accounts for only 1%–2% (1) of all congenital cardiac anomalies. It is a developmental disorder, in which the pulmonary veins fail to connect with the left atrium (3); they drain the oxygenated blood directly or indirectly through systemic veins into the right atrium resulting in a left-to-right shunt. Survival depends on the presence of a right-to-left intracardiac shunt, which almost always occurs through a patent foramen ovale that is rarely restrictive.

Total anomalous pulmonary venous connection is one of the most important differential diagnoses in infants presenting with respiratory distress and cyanosis during the newborn period. For

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early recognition of the disease, clinic suspicion is necessary. Echocardiography with color Doppler is usually a reliable tool for detecting anomalous pulmonary venous return noninvasively (4), although diagnosis is occasionally difficult because of the poor far-field resolution and small field of view. In the various reported studies, TAPVC was detected with an 85–100% sensitivity and 99–100% specificity (5).

A common feature in all types of total anomalous pulmonary venous drainage is the impossibility of defining the connections of the pulmonary veins with the left atrium. Furthermore, patients have volume overload of the right heart, and atrial septal defects of various sizes. The site of anomalous drainage of the pulmonary veins may be assessed by means of multiple cuts from subcostal, precordial and suprasternal windows (6, 7). The site of the orifice of TAPVC in supracardiac TAPVC is best seen from a suprasternal approach, in intracardiac TAPVC in the four-chamber projection, in infracardiac TAPVC from a subcostal approach (8).

In our case, infracardiac type TAPVC diagnosis was established by echocardiographic examination. During echocardiographic study, the transducer was placed in the subcostal region in a way to acquire images of the descending aorta and the vertebral column. Apart from the aorta and the inferior vena cava a third vessel was identified, lying anterior to the aorta, the proximal part lying behind the left atrium, the distal part curving anterior in the liver, the distal end being distended. With pulse Doppler ultrasound, the characteristic normal flow signals in the aorta and inferior vena cava were obtained. The signal from the anomalous pulmonary vein was a continuous venous signal, the direction of flow being away from the heart. These findings are highly characteristic and diagnostic for total anomalous pulmonary venous connection of the infracardiac type (6-8). So, we accept patient as infracardiac type TAPVC with venous obstruction and started digoxin and furosemide treatment. Catheter angiographic examination was undertaken the following day and the diagnosis was

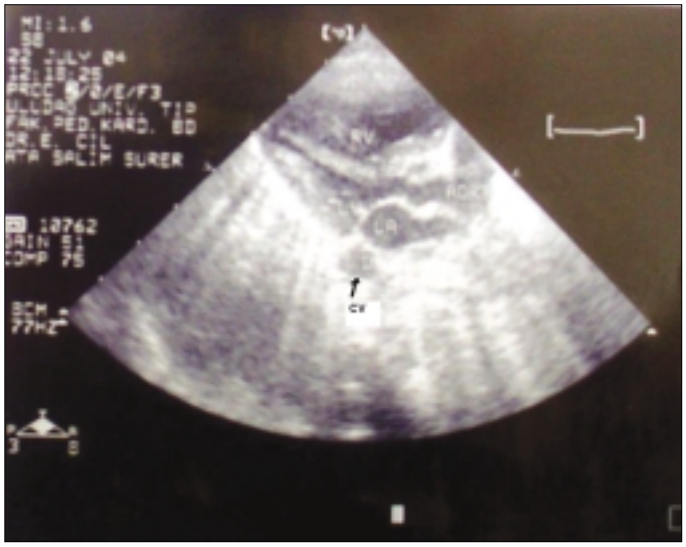

Figure 1. Long-axis view showing an echo free space behind the left atrium
CV- common pulmonary vein, LA- left atrium, LV- left ventricle, RV- right ventricle

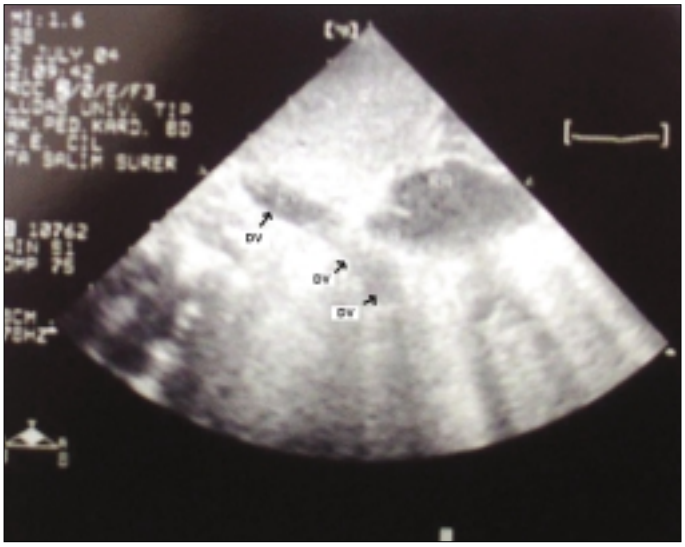

Figure 2. Subcostal view, demonstrating an anomalous vertical venous vessel
DV- descending vein

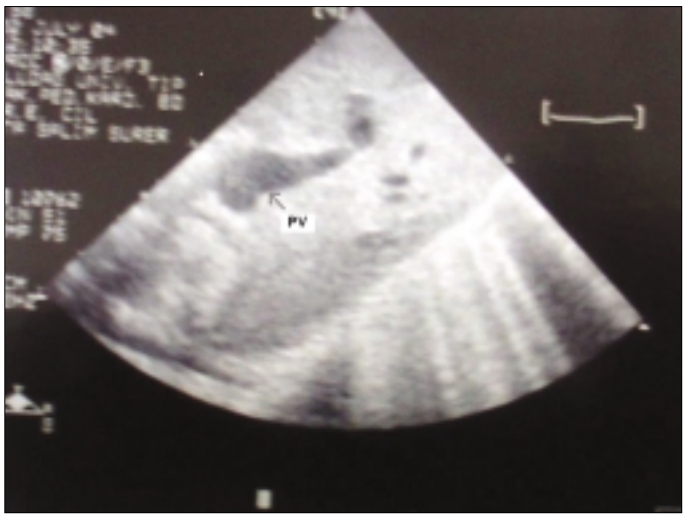

Figure 3. Subcostal view showing marked dilatation of the portal vein
PV- portal vein

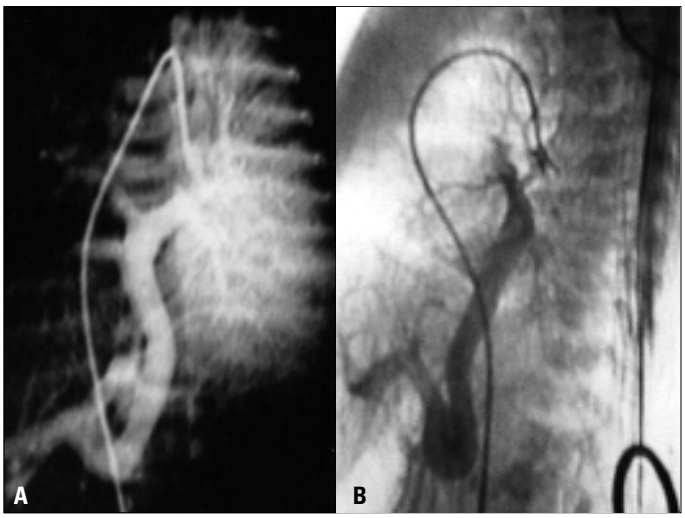

Figure 4. Angiographic images showing selective left pulmonary artery injection in anteroposterior (A) and left lateral (B) views, demonstrating vertical venous vessel and pulmonary veins

confirmed. Catheter angiography is accepted as diagnostic for TAPVC. But, in case of pulmonary hypertension due to pulmonary-venous obstruction, it carries an increased risk. In experienced centers, echocardiography has become the definite diagnostic method with a high level of sensitivity and specificity. Since infracardiac total anomalous pulmonary venous drainage associated with venous obstruction has been considered a surgical emergency (9), patient underwent surgery on an emergency basis. Because of a relatively small and non-compliant left atrium and left ventricle (10), low cardiac output persisted after repair and patient was lost at early postoperative period.

Total anomalous pulmonary venous connection is one of the most important differential diagnoses in infants presenting with respiratory distress and cyanosis during the newborn period. Echocardiography makes it possible to assess the diagnosis of TAPVC by a non-invasive approach, to find the site of the orifice and indicate operation without the use of invasive examination methods.

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