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Absent Pulmonary Valve Syndrome with Left Lung Atresia

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Absent pulmonary valve syndrome (APVS) is a rare anomaly characterized by the absence of the pulmonary valve. The absence of the pulmonary valve, along with the hypoplasia of the pulmonary annulus, leads to aneurysmal dilatation of the pulmonary artery and its branches. Other consequences include pulmonary stenosis and pulmonary insufficiency (1).

The 3-day-old baby presented with cyanosis and tachypnea. He had to and fro murmurs at the upper left sternal region, as well as hepatomegaly. The electrocardiogram showed right ventricular hypertrophy. The posterior-anterior chest X-ray showed opaque left hemithorax with cardiac displacement (Figure 1). Upon echocardiographic examination, the following findings were noted: the right cardiac chambers were dilated and left ventricular hypoplasia was present (Video 1. See corresponding video/movie images at www.balkanmedicaljournal.org). A large subaortic ventricular septal defect and a main pulmonary artery aneurysm were seen (Video 2. See corresponding video/movie images at www.balkanmedicaljournal.org). Hypoplastic pulmonary annulus and absent pulmonary valve leaflets were present (Figure 2). Moderate pulmonary stenosis (peak systolic gradient was 45 mmHg) and mild pulmonary regurgitation were present (Video 3. See corresponding video/movie images at www.balkanmedicaljournal.org). The aortic arc was left sided and no patent ductus arteriosus was seen. In addition to the echocardiographic findings, atresia of the left pulmonary artery was observed with computed tomography (Figures 3, 4). The patient died due to respiratory failure on fourth day after birth.

Absent pulmonary valve syndrome is a rare anomaly, which is characterized by aneurysmal dilatation of the pulmonary artery, depending on atresia or dysplasia of the pulmonary valve leaflets. The disease is usually associated with tetralogy of Fallot. Due to the absence of pulmonary valve leaflets, pulmonary regurgitation and mild pulmonary stenosis were seen (2). Respiratory distress signs could be seen to varying degrees due to compression of the airway with the pulmonary artery. In the severe forms, due to complete obstruction of the trachea

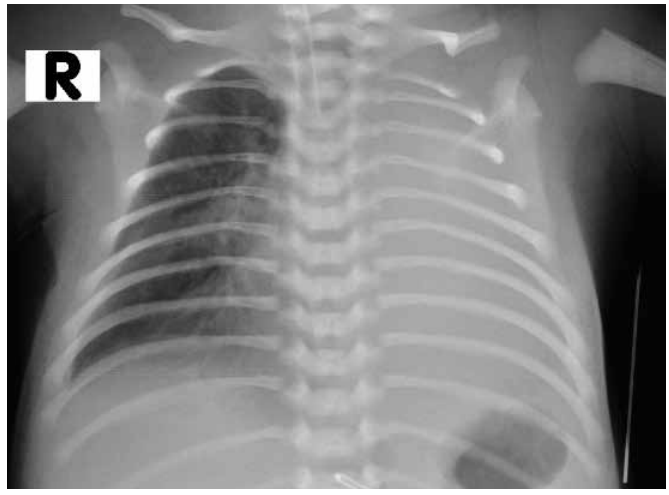

FIG. 1. Posterior-anterior chest X-ray showed opaque left hemithorax with cardiac displacement

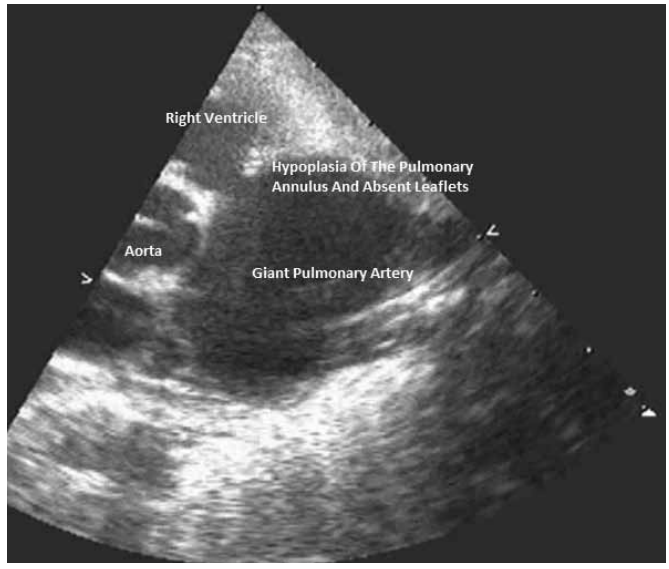

FIG. 2. Hypoplastic pulmonary annulus and absent pulmonary valve leaflets

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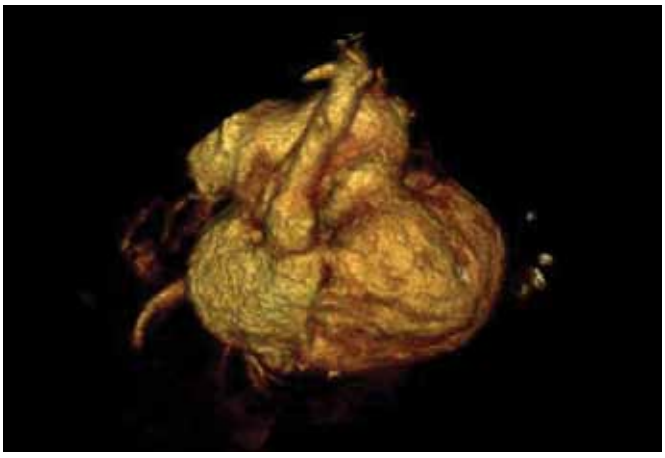

FIG. 3. CT 3D reconstruction of aneurysmal pulmonary arteries

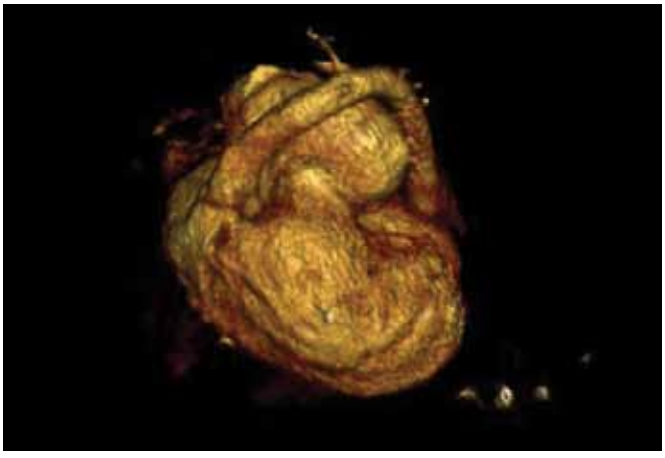

FIG. 4. Left pulmonary artery atresia

or bronchi, rapid progress can be seen in the neonatal period. More severe forms of the disease may result in death during the newborn period due to the pressure effect exerted by the pulmonary artery on the tracheobronchial structures.

The patient presented herein had an absence of the pulmonary valve, together with the non-restrictive ventricular septal defect. Also, mild pulmonary valve stenosis and insufficiency, widened pulmonary artery, and atresia of the left pulmonary artery were seen. Although catheterization could not be performed during the early stage, as the patient had severe respiratory dysfunction, computed tomography confirmed the presence of a widened pulmonary artery and atresia of the left pulmonary artery. Also, arterial branches from the abdominal or thoracic aorta supplying the left lung could not be detected with computed tomography.

The combination of APVS with atresia of the left pulmonary artery is extremely rare (3). Early diagnosis and supportive treatment in the newborn period are important for the survival of these severely affected infants.

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Video 1. Dilated right heart chambers.
Video 2. Large subaortic VSD.
Video 3. Hypoplastic pulmonary annulus and absent pulmonary valve leaflets.

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