

Figure 3. Transthoracic echocardiography (apical 5-chamber view) demonstrates an elongated anterior mitral chordae tendineae protruding into the left ventricular outflow tract (arrow)

Video 1. Transthoracic echocardiography (apical 5-chamber view) demonstrates an elongated anterior mitral chordae tendineae protruding into the left ventricular outflow tract

Video 2. Transthoracic echocardiography (parasternal long axis) demonstrates an elongated anterior mitral chordae tendineae protruding into the left ventricular outflow tract

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Combination of tetralogy of Fallot with absent pulmonary valve and left pulmonary artery originating from patent ductus arteriosus: A rare association in an infant

A 6-month-old female infant was transferred because of respiratory distress. A chest radiograph demonstrated a well delineated, huge

mass on the upper and middle part of the right lung (Fig. 1). Two-dimensional echocardiography showed dilatation of the right ventricle, main pulmonary artery (20 mm, z score +4.4) and right pulmonary artery (22 mm, z score +7.8) but dilatation of the left pulmonary artery was not observed (Fig. 2a, b). There was no identifiable pulmonic valve tissue in the area of the right ventricular outflow tract. Color Doppler echocardiogram showed turbulent flow across the right ventricular outflow tract with systolic right ventricle pulmonary artery gradient of 70 mm Hg. There was a wide jet of pulmonic regurgitant flow essentially filling the right ventricular outflow tract (Fig. 2c-d). Computed tomographic angiography (256 Slices, Somatom Definition; Siemens Medical Solutions, Germany) showed non-confluent pulmonary arteries with dilated right and left pulmonary arteries connected to the patent ductus arteriosus (Fig. 3a, b). During the 7th month, she underwent a total corrective operation [ventricular septal defect (VSD) closure, right ventricular outflow tract (RVOT) reconstruction, and right ventricle-pulmonary artery conduit implantation with 19 mm pulmonary homograft, plication of the right pulmonary artery and unifocalization of the left pulmonary artery]. The infant has no significant residual symptoms after more than 2 years post successful surgery.

Tetralogy of Fallot with absent pulmonary valve syndrome is a rare variant of tetralogy of Fallot. It may clinically be present with airway compression from dilated pulmonary arteries or congestive heart fail-



Figure 1. Chest radiography shows well delineated huge mass on the upper and middle part of the right lung due to enlarged right pulmonary artery



Figure 2. a-d. Two-dimensional echocardiography shows malalignment ventricular septal defect, dextroposition of the aorta, dilatation of main pulmonary artery and right pulmonary artery but not of the left pulmonary artery (a, b). Color Doppler echocardiography shows turbulent flow across the right ventricular outflow tract and large regurgitation belonging to pulmonary insufficiency (c, d)

Ao - aorta; asterisk - indicates dextroposition of the aorta; LV - left ventricle; MPA - main pulmonary artery; PR - pulmonary regurgitation; RPA - right pulmonary artery; RV - right ventricle; and RVOT - right ventricular outflow tract

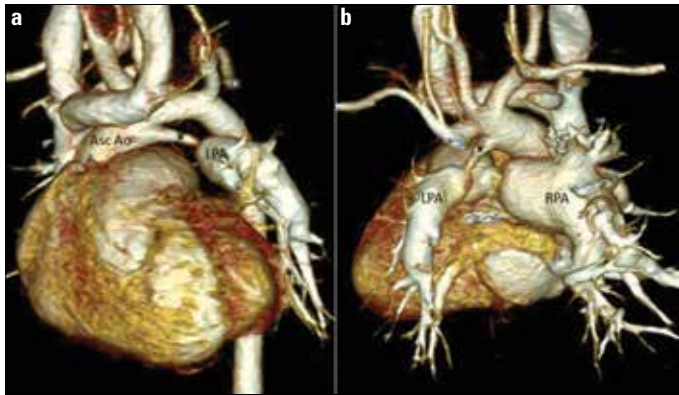


Figure 3. a, b. Computerized tomographic angiography shows non-confluent pulmonary arteries with dilated right pulmonary artery and left pulmonary artery connected to patent ductus arteriosus

Asc Ao - indicates ascending aorta; asterisk - indicates patent ductus arteriosus; LPA - left pulmonary artery; and RPA - right pulmonary artery

ure in early infantile period. Although there have been several cases of anomalous left pulmonary artery origin from the ascending aorta in

tetralogy of Fallot with absent pulmonary valve syndrome, the combination of tetralogy of Fallot with absent pulmonary valve syndrome and ductal origin of the left pulmonary artery is extremely rare.

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