

D-transposition of the great arteries with bilateral peripheral pulmonary artery stenosis and arcus hypoplasia: A rare association

Periferik pulmoner stenoz, arkus hipoplazisi ve büyük arter transpozisyonu: Nadir görülen bir birliktelik

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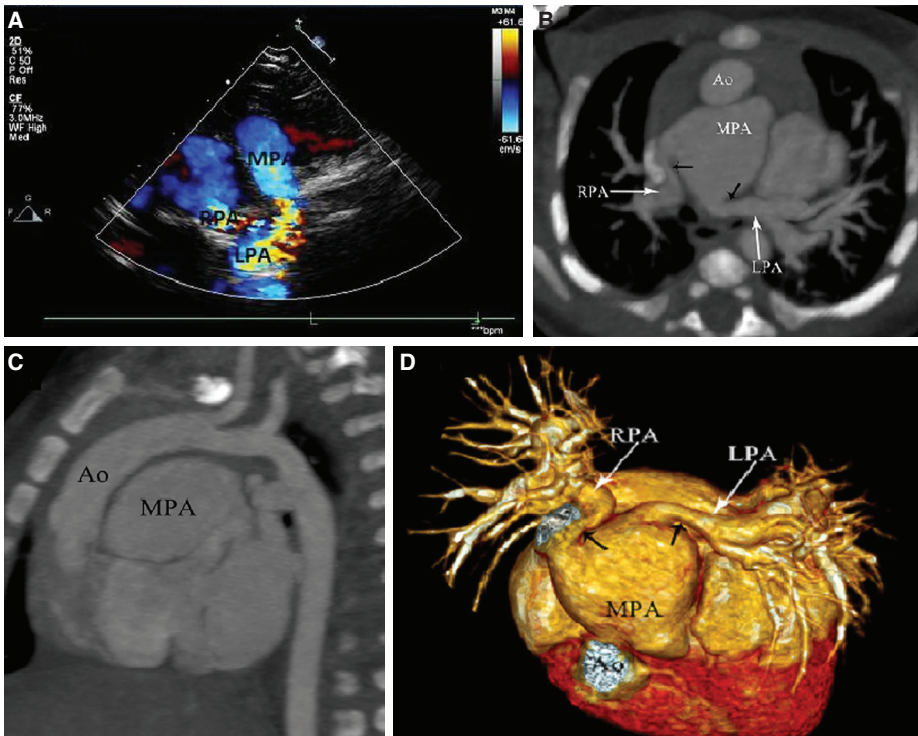
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A full-term 38-day-old female infant weighing 4 kg was admitted to our clinic with cardiac murmur. Oxygen saturation was 85%. Physical examination revealed a 2/6 systolic murmur along the left sternal border. Transthoracic echocardiography revealed a d-transposition of the great arteries. The two ventricles were balanced with a preserved left ventricle systolic function (LVEF = 70%). A 2 mm ventricular septal defect, a 6 mm secundum atrial septal defect, and a large patent ductus arteriosus were detected. In addition, there were peripheral pulmonary artery stenosis

(Figure A, Video 1*) with a continuous-wave Doppler gradient of 50 mmHg in both, and arcus hypoplasia. The main pulmonary artery was dilated (21 mm); right and left pulmonary arteries were small in size (3.5 mm and 3 mm respectively). The proximal transverse arch (5 mm, z-score: -3.41) and distal transverse arch (4.5 mm, z-score: -2.11) were hypoplastic. Isthmus was of normal size (5.5 mm, z-score: -0.41). Computerized tomography (CT) angiogram and 3D reconstruction confirmed severe right and left pulmonary artery stenosis and arcus aorta hypoplasia (Figure B-D). The patient was operated for arterial switch with bilateral pulmonary artery and aortic arch reconstruction. Transposition of the great arteries is a common congenital heart defect that causes cyanosis in neonates. It has been reported as occurring concomitantly with many congenital heart malformations. However, to the best of our knowledge, transposition of the great arteries associated with a peripheral pulmonary artery stenosis with arcus hypoplasia has not been reported. Careful echocardiographic assessment with CT confirmation may be the most important factor in developing the operative plan.



Figures– (A) Echocardiographic view of peripheral pulmonary artery stenosis. (B) Oblique axial thin maximum intensity projection multidetector computed tomographic image of peripheral pulmonary artery stenosis. (C) Volume rendering multidetector computed tomographic image of pulmonary arteries. (D) Oblique sagittal thin maximum intensity projection multidetector computed tomographic image. Ao: Aorta; MPA: Main pulmonary artery; LPA: Left pulmonary artery; RPA: Right pulmonary artery; Black Arrows: Stenosis.

*Supplementary video file associated with this presentation can be found in the online version of the journal.

