

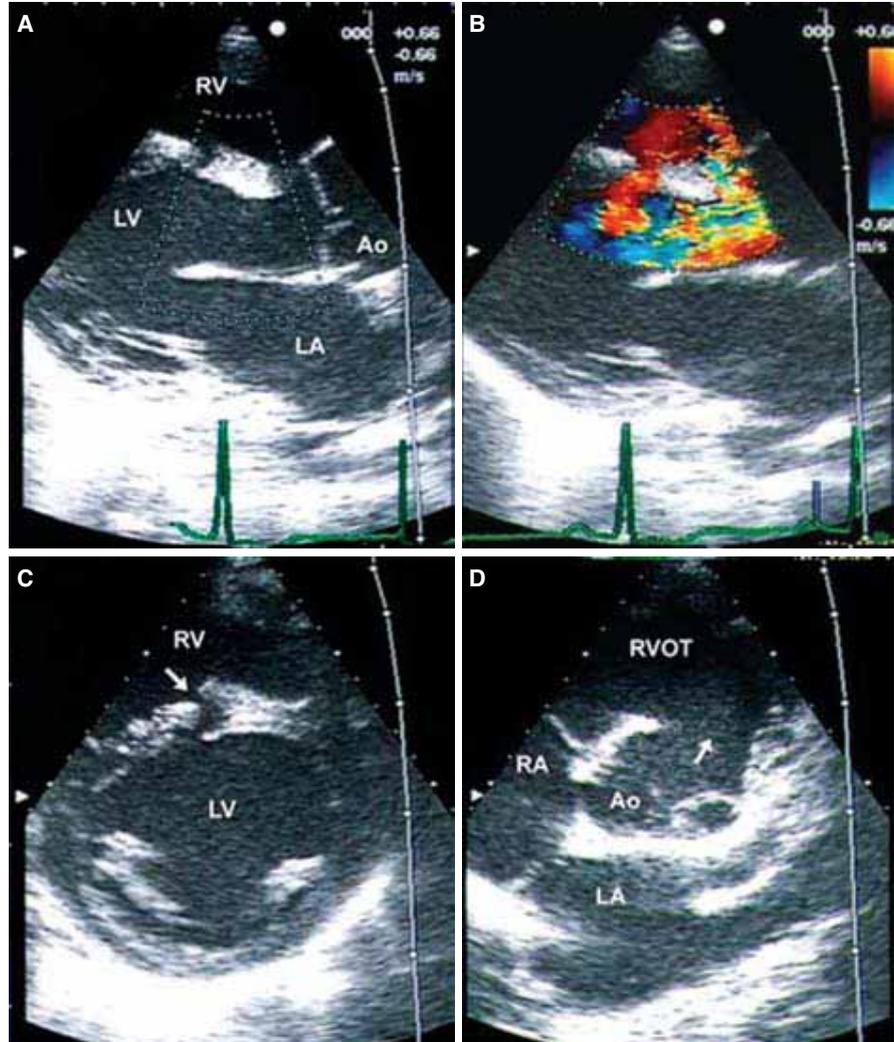
Pulmonary atresia with double ventricular septal defect

Çift ventriküler septal defektli pulmoner atrezi

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Pulmonary atresia with ventricular septal defect (VSD) is characterized by underdevelopment of the right ventricular outflow tract with atresia of the pulmonary valve, a large VSD, and overriding of the aorta, and is the ultimate expression of severity in Fallot's tetralogy. Additional muscular VSD occasionally coexist with malaligned VSD. A 16-year-old cyanotic boy was referred to our hospital for heart murmur. The electrocardiogram showed right ventricular hypertrophy with right axis deviation. The chest radiograph revealed a normal-sized, boot-shaped heart with a prominent right ventricular contour, absence of the pulmonary artery segment, and an upwardly displaced apex. Two-dimensional and color flow imaging in the parasternal long-axis views (Fig. A, B) showed malalignment and muscular VSD, and severe aortic regurgitation. The parasternal short-axis views also demonstrated double VSD and lack of



patency of the right ventricular outflow tract (Fig. C, D). The patient was not considered eligible for corrective surgery because of the absence of the central pulmonary arteries.

Figures. Two-dimensional and color flow imaging. The parasternal (A, B) long- and (C, D) short-axis views show combination of malalignment and muscular ventricular septal defect, and severe aortic regurgitation with pulmonary atresia. Ao: Aorta; RV: Right ventricle; RA: Right atrium; LV: Left ventricle; LA: Left atrium; RVOT: Right ventricular outflow tract.