Coexistence functional pulmonary atresia with other congenital heart anomalies.

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Case Report

A 35 year-old woman presenting to our center at 21 weeks of gestation was initially diagnosed as suspect pulmonary atresia (PA) associated with small right ventricle, slow velocity of pulmonary artery flow and reverse PDA flow, minimal tricuspid diastolic flow with occasional very-tiny TR, persistent left superior vena cava, and mild pericardial effusion. A newborn was delivered by Cesarean delivery at 36 6/7 weeks of gestation with Apgar scores: 6->8.

After birth, PGE1 was administrated as a result of suspected PA. Postnatal echocardiography revealed mild hypoplasia of right ventricle with normal-echogenic pulmonary valve showing restricted mobility (compatible with functional PA), atrial septum bulging to the left, 8.6mm ASD with right to left shunt, 6mm PDA, and one apical muscular VSD (3.6mm) with left to right shunt. The ductus arteriosus was closed at the day 6 with no desaturation nor respiratory distress.

Subsequently, the newborn was hemodynamically stable and she was discharged on day 16. Final diagnosis of the newborn was congenital heart disease with functional PA with a muscular VSD and ASD.

Fig.A: mild pericardial effusion, a small RV and minimal tricuspid valve flow during diastolic phase that may be misdiagnosed as tricuspid atresia

Fig.B: reverse PDA flow (red color) and PLSVC (hollow arrow)

Fig.C: postnatal echocardiography showing functional PA with bidirectional pulmonary flow