Case Reports

Functional Pulmonary Atresia in a Newborn with Normal Intracardiac Anatomy

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Abstract. Functional pulmonary atresia is a relatively rare clinical condition usually associated with
Ebstein’s malformation, tricuspid valve dysplasia, Uhl’s anomaly, or transient myocardial ischemia with
severe tricuspid regurgitation. The occurrence of functional pulmonary atresia associated with tran-
sient tricuspid regurgitation in a newborn with an anatomically normal heart is even more uncommon.
We describe a case in which color Doppler flow mapping played an essential role in diagnosis and
follow-up of this clinical condition in a newborn who had normal intracardiac anatomy.

Key words: Pulmonary atresia — Doppler echocardiography

Increased perinatal pulmonary vascular resistance may induce functional pulmonary atresia in the neo-
brate resulting in inadequate forward ejection of the right ventricle and severe tricuspid regurgitation [1].
Although this is a relatively uncommon clinical condition, usually occurring in structurally abnormal
hearts (Ebstein’s anomaly, Uhl’s anomaly, tricuspid valve dysplasia, and transient myocardial ischemia
with severe tricuspid regurgitation), it may occur even more rarely in anatomically normal hearts of new-
borns with transient tricuspid regurgitation. Functional pulmonary atresia must be differentiated from
anatomic pulmonary atresia with intact ventricular septum because of the specific management implica-
tions. Two-dimensional echocardiography and color Doppler flow mapping play an important role in the
differential diagnosis between these two clinical conditions [2–6]. The structural and physiologic charac-
teristics usually emphasized for Doppler echocardiographic diagnosis of functional pulmonary valve
atresia include (1) an anatomically normal pulmonary valve and right ventricular outflow tract and (2) pulmonary valve leaflets that do not open during systole but present diastolic regurgita-
tion [1–6].

Case Report

A 1.4-kg male infant born at 28 weeks gestation with Apgar scores of 7 and 9 at birth presented with cyanosis and respiratory distress. He was transferred to the intensive care unit, where he was intu-
bated and mechanically ventilated. Physical examination revealed, in addition to severe cyanosis, tachypnea and an early (grade 3/6) systolic murmur. Upon admission to the intensive care unit, arterial
oxygen saturation was 35%, pCO₂ was 84 mmHg, and metabolic acidosis was documented (pH = 7.16). The chest X-ray showed mild cardiomegaly and reduced pulmonary vascular markings. Doppler echocardiographic study was performed 3 hours after birth with a commercially available ultrasound system (Hewlett-Packard Model 2500) using a 5.0-MHz transducer. Although intracardiac anatomy was normal, the right ventricle was mildly
dilated, the right atria was moderately dilated, and severe tricuspid

Fig. 1. A systolic frame obtained in paraesternal short-axis view shows normal right ventricular anatomy and a pulmonary valve with normal echo density that do not open during systole (arrow). AO, aorta; PA, pulmonary artery; RA, right artery; RV, right ventricle.
regurgitation was documented (very large pansystolic turbulent jet). Based on peak tricuspid jet velocity (310 cm/sec), pulmonary systolic pressure was estimated (50 mmHg). The anatomy of the right ventricle infundibular region was normal and the pulmonary valve had normal echo density but the leaflets did not open during systole (Fig. 1). There was no antegrade flow across the pulmonary valve, but during diastole a mild pulmonary regurgitation was documented by color Doppler flow mapping (Figs. 2 and 3). The ductus arteriosus was not documented in this first study.

The atrial septum bulged to the left and a patent foramen ovale presenting right-to-left flow was seen. The tricuspid valve was inserted at normal level without displacement. Mechanical ventilation controls were then modified to produce hyperventilation. Two hours later cyanosis was no longer observed, arterial pH was 7.6, pCO₂ was 23 mmHg, and oxygen saturation increased to 97%. Doppler echocardiography was repeated 15 hours after birth and showed (1) an important reduction in the dimensions of jet area indicating a mild to moderate degree of tricuspid regurgitation, (2) a reduction of peak jet velocity (220 cm/sec, systolic pulmonary pressure 27 mmHg), and (3) that flow across the patent foramen ovale was left to right and a normal systolic opening of the pulmonary valve was observed. Color Doppler flow mapping documented a normal flow across the pulmonary valve and the ductus arteriosus was now patent (Figs. 3 and 4). The infant was weaned from mechanical ventilation by 5 days of age and was discharged home at the age of 10 days. Follow-up Doppler echocardiography performed at 5 months of age disclosed a completely normal examination, including the absence of tricuspid regurgitation, closure of the ductus arteriosus, the absence of flow across the foramen ovale, and normal antegrade flow in the pulmonary artery.

**Discussion**

In this case, Doppler echocardiography played an essential role in raising the suspicion of functional pulmonary atresia. As previously reported [2–6], this diagnosis was supported by the absence of antegrade flow across the pulmonary valve associated with the occurrence of pulmonary valve regurgitation documented by color Doppler flow mapping in a newborn who had normal anatomy of the right ventricle infundibular region, normal echo density of the pulmonary valve, and immobile pulmonary valve leaflets during the cardiac cycle.

The occurrence of functional pulmonary valve atresia in a neonate with normal intracardiac anatomy has rarely been reported. Although the prognosis of