Case 1/2006 – Eight-month-old Infant with Pulmonary Atresia and Ventricular Septal Defect and Increased Pulmonary Blood Flow

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Clinical findings

Eight-month-old, white, male infant presenting with cyanosis and shortness of breath during feedings since birth, with no progression. A heart murmur had been detected on auscultation on his first day of life. Weight gain was poor and mild dyspnea at rest remained unchanged. Physical examination revealed dyspnea with mild cyanosis; pulses were normal. Oxygen saturation was 88%, heart rate was 125 bpm, respiratory rate was 60 rpm and weight was 6,500 g. The aorta was not palpable in the suprasternal notch. In the precordium, mild systolic impulses were observed on the left sternal border and the apical impulse was palpable in the fourth left intercostal space, in the midclavicular line, slightly/more forceful, limited to two fingertips. A mild early systolic click was heard in the aortic area along with a blowing, grade 1-2/6, continuous murmur in the pulmonic and aortic areas and in the posterior chest, mainly on the right side. The liver was palpable 2 cm below the right costal margin. Hb: 13.9 g/dl, Hct: 42%. The electrocardiogram showed signs of right ventricular overload with an 18-mm R wave in V1 preceded by a Q wave, and RS morphology from V2 to V6. SÂP: + 30°, SÂQRS: +95°, SÂT: + 40°.

Radiographic imaging

The image shows enlargement of the cardiac silhouette due to a rounded long ventricular border with an upwardly displaced apex. The middle arch is excavated; the vascular pedicle is widened due to a dilated aorta, and the pulmonary vasculature is increased especially on the right side (Figure 1).

Diagnostic impression

This image suggests a heart disease with a morphology like that found in the tetralogy of Fallot, but with the presence of increased pulmonary blood flow, which suggests pulmonary atresia with ventricular septal defect with systemic-to-pulmonary collateral vessels.

Differential diagnosis

This anomaly should be distinguished from the tetralogy of Fallot itself, which presents with increased pulmonary flow because of moderate pulmonary stenosis. Tricuspid atresia and pulmonary atresia with an intact septum should also be considered given the leftward displacement of the ventricular border, also simulating left ventricular enlargement.

Diagnostic confirmation

Clinical findings strongly suggest the diagnosis of pulmonary atresia with ventricular septal defect because of the presence of an early systolic click in the aortic area and a continuous murmur in the posterior chest due to left-to-right flow through systemic-to-pulmonary collateral vessels, with increased pulmonary blood flow and mild cyanosis. The echocardiogram showed both pulmonary arteries and pulmonary trunk with a 3-mm diameter, in addition to pulmonary atresia and ventricular septal defect. Cardiac catheterization showed dilated systemic-to-pulmonary collateral vessels from the descending aorta to the right pulmonary arterial tree and to the left lower lobe. Pressure measurements were: RA: 8, RV: 70/8, LV: 70/8, Aorta: 70/45-53 mmHg.

Management

Ligation of the collateral to the right and Blalock-Taussig anastomosis were indicated and the arterial saturation changed to 80%, because of the resulting reduction in the pulmonary blood flow.

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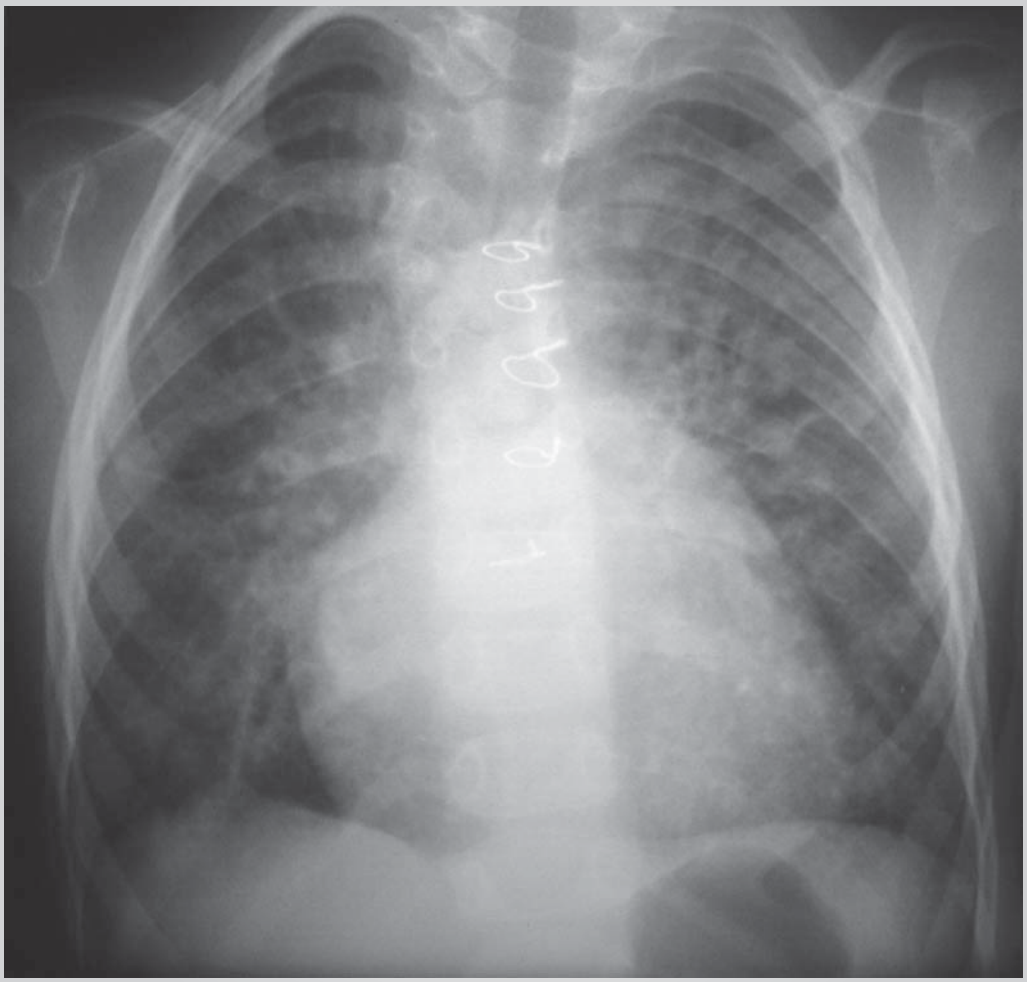

Fig. 1- *Chest radiograph shows a cardiac shape similar to that found in the tetralogy of Fallot. However, the increased pulmonary vasculature suggests pulmonary atresia with ventricular septal defect and large-diameter systemic-to-pulmonary collateral vessels*