

Case 2/2008 – Three Year Old Female Patient with Subvalvular, Valvular, and Supravalvular Pulmonary Stenosis

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Clinical data

Cardiac murmur identified upon routine auscultation at 12 months of age which had presented no symptoms or clinical complications since birth.

Physical examination

Patient was eupneic, with normal skin color, and pulses were palpated on all 4 limbs. Weight: 15 Kg, Height: 100 cm, BP: 95/60 mm Hg, HR: 86 bpm. Aorta was not palpable at the supraasternal notch.

Precordium showed no deformities or impulsions. *Ictus cordis* was not palpated. Slight systolic thrill at supraasternal notch and

upper border of the sternum, with rough ejection systolic murmur +/++ in the same locations. Heart sounds were normal with consistent splitting of the second sound, and the pulmonary component was less pronounced than the aortic component.

Lungs and abdomen showed no abnormalities.

The electrocardiogram showed sinus rhythm and signs of right chamber overload. $\hat{A}P$: + 60°, $\hat{A}QRS$: indeterminate, $\hat{A}T$: +30°.

Radiographic image

Cardiac area with normal dimensions, rounded morphology, and diminished pulmonary vascular network (rete) (Fig. 1).

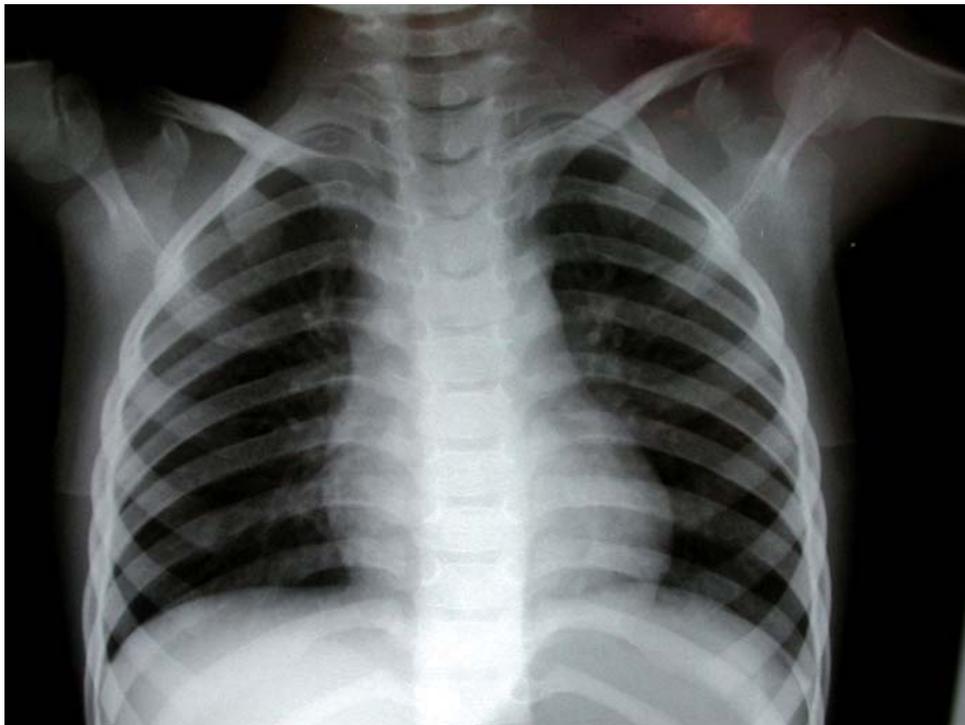


Fig. 1 - Chest X-ray shows rounded cardiac area, elevated tip, and normal limits.

Key words

Pulmonary stenosis, Congenital cardiopathy, surgery.

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Diagnostic impression

this image is consistent with the diagnosis of obstructive acyanogenic cardiopathy on the right, more likely infundibular-valvular pulmonary stenosis.

Differential diagnosis

absence of dilation of the pulmonary middle arch suggests the diagnosis of pulmonary stenosis with a preponderant infundibular component, as well as the possibility of supralvular pulmonary stenosis or right ventricular inlet stenosis.

Diagnostic confirmation

Clinical elements lead to the diagnosis of infundibular-valvular pulmonary stenosis confirmed by the echocardiogram with trivalvular commissural fusion, 70 mmHg pressure

gradient, and 10 mm diameter pulmonary ring. Cardiac catheterization, initially indicated to perform pulmonary valvuloplasty, evidenced supralvular pulmonary stenosis with a 41 mmHg pressure gradient and clear infundibular reaction. Pressure of the right ventricle was 55/7 mmHg and of the pulmonary trunk, 14/7 mm Hg. The pulmonary valve was deemed normal due to its ample valvular mobility (Fig. 2).

Treatment

In surgery, obstructive components were found at infundibular, valvular, and supralvular pulmonary levels. Resection of the muscle was performed in the right ventricle outlet, with valvular commissurotomy and widening of the pulmonary trunk with a bovine pericardium patch. Postoperative echocardiogram showed a residual 25 mmHg pressure gradient between the right ventricle and the pulmonary trunk.

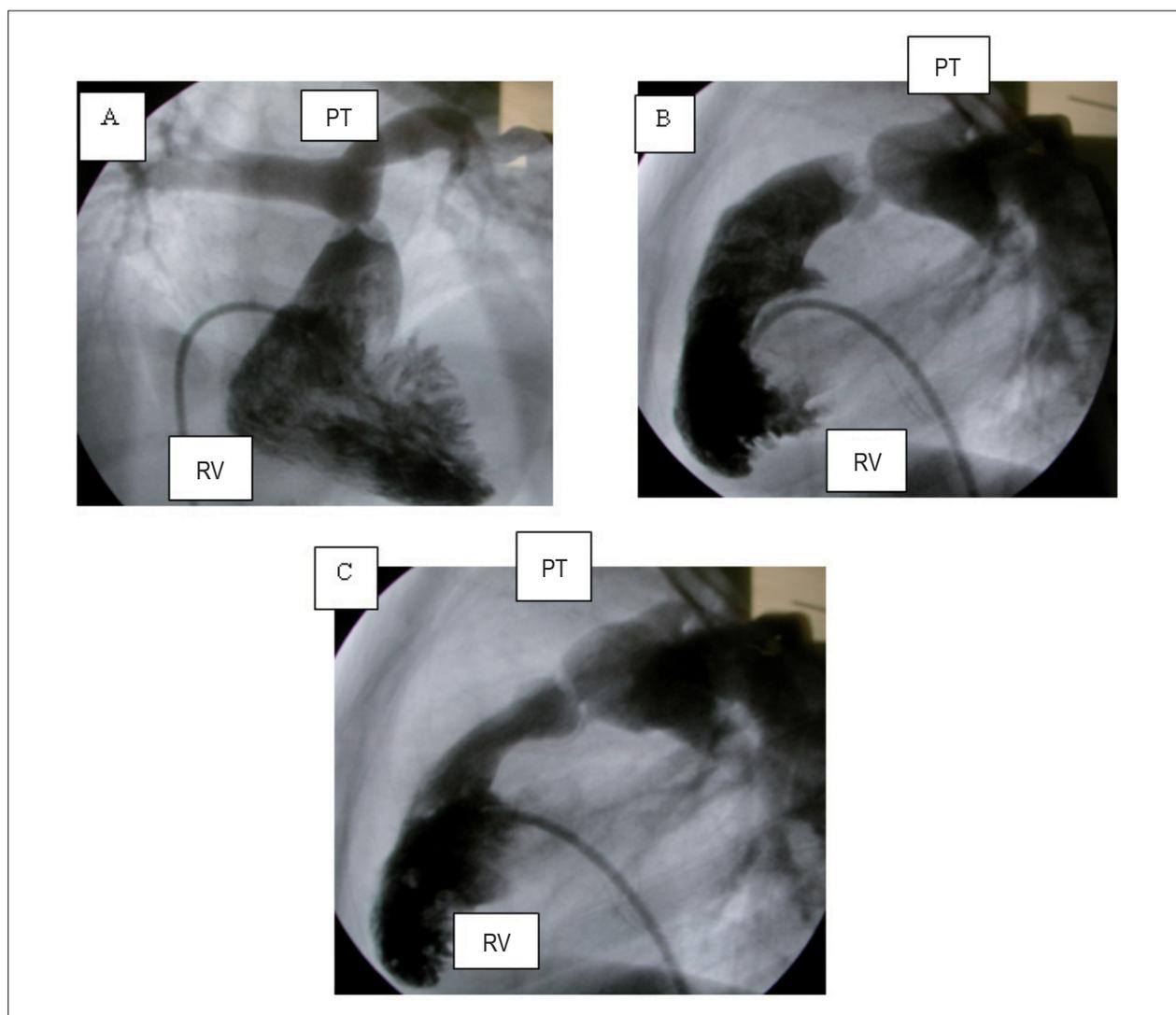


Fig. 2 - Angiogram of the right ventricle shows supralvular pulmonary stenosis in A (posteroanterior projection) and in B (left anterior oblique projection) with normal valvular motility, besides infundibular pulmonary stenosis in C (left anterior oblique projection). PT - pulmonary trunk, RV - right ventricle.