

Images in cardiovascular medicine

Ebstein's anomaly associated with ventricular septal defect and pulmonary stenosis

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(Ital Heart J 2000; 1 (10): 705)

This paper was supported by the Programma Operativo del Piano CCCN-5 B006 of the Ministero dell'Università e Ricerca Scientifica (MURST) and the European Community (no. 711/1998).

Received September 4, 2000; accepted September 14, 2000.

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A 15-day-old infant was referred to our hospital due to deep cyanosis and systolic murmur at the mid-sternal level. Chest X-ray showed a moderate cardiac enlargement with poor pulmonary vascular markings. At ECG, P wave anomaly suggestive of right atrial enlargement was found. Echocardiography showed a moderate-sized muscular ventricular septal defect, as well as a severe downward displacement of both septal and posterior tricuspid valve leaflets and a severe

stenosis of the pulmonary valve (Fig. 1). The Ebstein's anomaly of the tricuspid valve resulted in severe regurgitation and a marked reduction in size of the functional right ventricular chamber. At cardiac catheterization, any attempt of crossing the pulmonary valve to perform balloon dilation failed, so the baby underwent surgery. However, despite successful surgical pulmonary valvulotomy, it was impossible to wean the baby off the cardiopulmonary bypass.

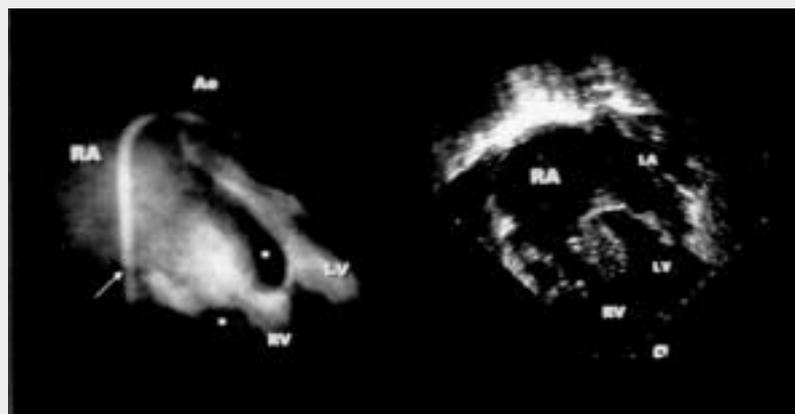


Figure 1. Angiographic (left panel) and echocardiographic (right panel) imaging of the Ebstein's anomaly of the tricuspid valve. Left ventricular angiography was performed in the 4-chamber view, in order to image the mid-muscular ventricular septal defect. Note the dysplasia and redundancy of both mural and posterior leaflet of the tricuspid valve (asterisks) that appear severely downward displaced with respect to the valvular annulus (white arrow). It results in severe tricuspid valve regurgitation and marked hypoplasia of the functional right ventricular chamber. Ao = aorta; LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.