LETTERS TO THE EDITOR

Pulmonary Atresia With Intact Ventricular Septum

Allan et al. (1) reported five cases of pulmonary atresia with intact ventricular septum in which both the right atrial and ventricular cavities were enlarged. They confirmed at autopsy that this degree of enlargement caused lung compression during fetal life. One fetus died during intrauterine life and the rest of the patients died in the early neonatal period. We recently observed similar findings in a full-term male neonate with critical pulmonary valve stenosis and intact ventricular septum. Cyanosis and a continuous murmur in the pulmonary area were noted at birth. The chest radiograph revealed massive cardiomegaly and decreased pulmonary vascularity. Two-dimensional echocardiogram documented critical pulmonary valve stenosis, intact ventricular septum, a dilated right atrial cavity (Fig. 1) and a small-sized right ventricular cavity and tricuspid valve. An interatrial communication of adequate size was also seen. The ratio of the right ventricular to left ventricular dimensions was 0.78 as assessed by M-mode echocardiogram. The patient was started on prostaglandin E infusion with adequate response. Cardiac catheterization and angiography were performed mainly to assess the size of the brachiocephalic vessels and pulmonary arteries in preparation for the construction of a modified Blalock-Taussig shunt. An aortogram showed opacification of small pulmonary arteries across the ductus arteriosus. The right ventricular cavity could not be entered. This cavity failed to opacify during an inferior vena cava angiogram. At surgery the pinhole pulmonary valve orifice was enlarged and the Blalock-Taussig shunt constructed. The ductus arteriosus was ligated. After completion of the shunt, ventricular fibrillation developed during irrigation of the pericardium and the infant failed to survive resuscitation attempts.

Autopsy was performed; and the heart and lungs were perfused and examined after fixation. The heart was enlarged. The right atrium was tremendously dilated with dimensions of 3 × 4 × 1.8 cm (Fig. 2). The tricuspid valve orifice was small and measured 1.6 cm in diameter. The valve was composed of two cusps with shortened chordae tendineae. The right ventricular cavity was hypoplastic and measured 0.3 × 0.6 × 1.0 cm. The free wall of this ventricle was 1.7 cm thick. The main pulmonary artery was normal in size and the surgical enlargement of the valve orifice was seen. However, the right and left pulmonary arteries were markedly hypoplastic and the more distal portions constricted. On microscopic examination, the arterioles were constricted with narrowed lumens and hypertrophied media. Both lungs were hypoplastic. The Blalock-Taussig shunt was patent. The left atrial and

Figure 1. Two-dimensional echocardiogram in the apical four chamber view shows dilation of the right atrium (Ra), a hypoplastic right ventricle (Rv) and a normal-sized left atrial (La) cavity. Only a section of the left ventricle (Lv) is seen in this view. Ias = interatrial septum.

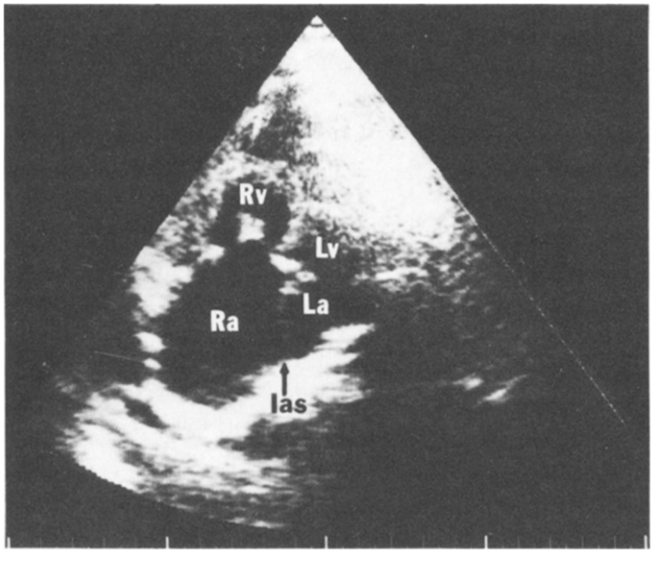
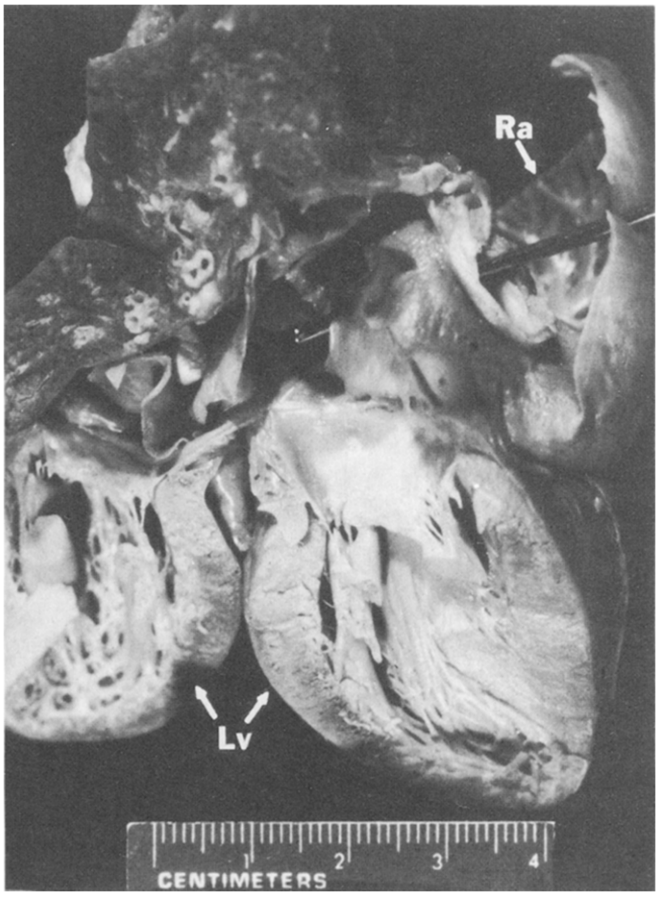


Figure 2. At postmorten examination the right atrial cavity was enlarged. A probe is seen across an adequately sized interatrial communication. Abbreviations as in Figure 1.



ventricular cavities were of normal dimensions. The free wall of the left ventricle was 0.7 cm thick. The coronary arteries had normal anatomy and distribution.

Enlargement of the right heart cavities in neonates with pulmonary atresia/critical pulmonary valve stenosis and intact ventricular septum should cause suspicion of hypoplastic lungs. The left to right shunt across a Blalock-Taussig shunt in this setting may be impaired by the increased pulmonary vascular resistance as suggested by lung microscopy in the patient described.

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Reference

1. Allan LD, Crawford DC, Tynan MJ. Pulmonary atresia in prenatal life. J Am Coll Cardiol 1986;8:1131–6.

Correction

We wish to correct an inadvertent transposition of numbers which occurred in our recently published study (Kudenchuk PJ, Kron J, Walance CG, et al. Reproducibility of arrhythmia induction with intracardiac electrophysiologic testing: patients with clinical sustained ventricular tachyarrhythmias. J Am Coll Cardiol 1986;7: 819–28). The text (the last paragraph of the right-hand column on page 823) corresponding to Figure 5 incorrectly read that 74 patients (65%) were taking a class I antiarrhythmic agent at the time of their initial sustained event, and that 40 patients were not taking such an antiarrhythmic agent. The text should read: Forty patients (35%) were taking a class I antiarrhythmic agent, and 74 patients were not taking such an antiarrhythmic agent at the time of their initial sustained arrhythmia event. The figure corresponding to this text was appropriately labeled, and designated the two groups correctly.

We apologize for the confusion this discrepancy between text and figure may have caused.

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