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IMAGE

Anomalous origin of the pulmonary artery from the ascending aorta in a neonate, assessed by two-dimensional echocardiography

Shunt néonatal : artère pulmonaire naissant de l'aorte ascendante

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A full-term baby presented at 2 weeks of life with severe congestive heart failure. On clinical examination a 2/6 holosystolic murmur was found, bounding femoral pulses and hepatomegaly. Echocardiography found massive dilatation of both ventricles with severe atrioventricular regurgitation and pulmonary hypertension (Fig. 1). No intracardiac shunt was found but an abnormal right pulmonary artery arising from the ascending aorta was diagnosed (Figs. 2 et 3). On Doppler investigation of the aortic arch, diastolic reflux from the aorta to the right pulmonary artery was found (Fig. 4). The patient underwent immediate and successful complete surgical repair, consisting of detachment of the anomalous pulmonary artery from the ascending aorta with direct anastomosis to the main pulmonary artery.

Anomalous origin of the right pulmonary arteries arising from the ascending aorta, also called hemitruncus, is a very rare anomaly. This lesion with two normal semilunar valves should be distinguished from truncus arteriosus communis, in which there is only one semilunar valve. Associated defects such as tetralogy of Fallot or aortopulmonary window are present occasionally. The right lung receives blood directly from the aorta, with resulting massive volume and/or pressure overload of the left pulmonary branch. The right lung receives the entire right ventricular output, resulting in volume overload. Biventricular cardiomyopathy and pulmonary hypertension occur rapidly in neonates, leading to respiratory distress and congestive heart failure. Diagnosis is by two-dimensional echocardiography and surgery should be performed as soon as possible. Treatment consists of surgical division of the anomalously connected pulmonary artery branch and reanastomosis

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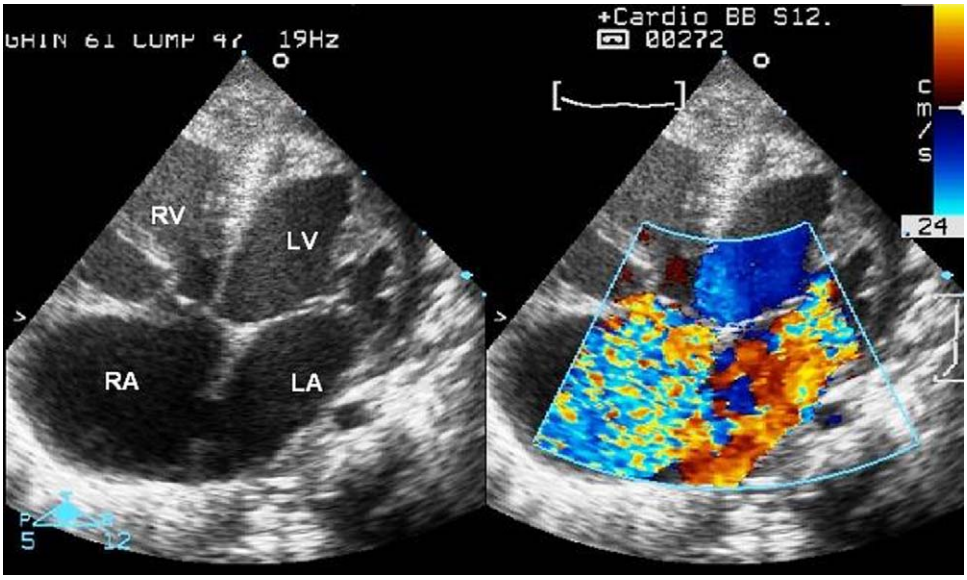

Figure 1. Apical four-chamber view without (left) and with colour Doppler (right). The right and left cavities are dilated massively. In systole, colour Doppler scans show severe tricuspid and mitral regurgitation. LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle (Supplementary data).

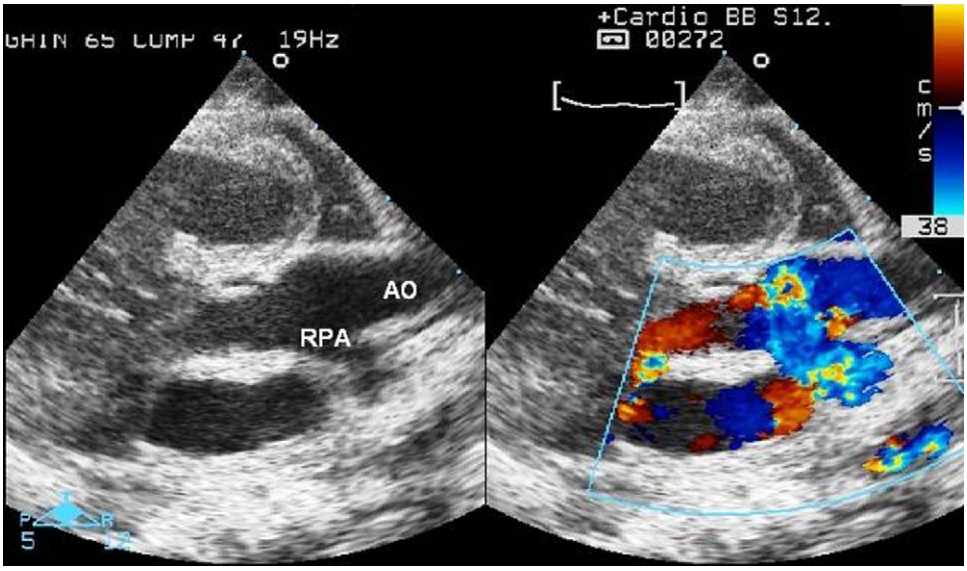

Figure 2. Parasternal long-axis view without (left) and with colour Doppler (right). The right pulmonary artery arises from the posterior wall of the aorta. The colour Doppler scan shows the massive shunt into the right pulmonary branch. Ao: aorta; RPA: right pulmonary artery(Supplementary data).

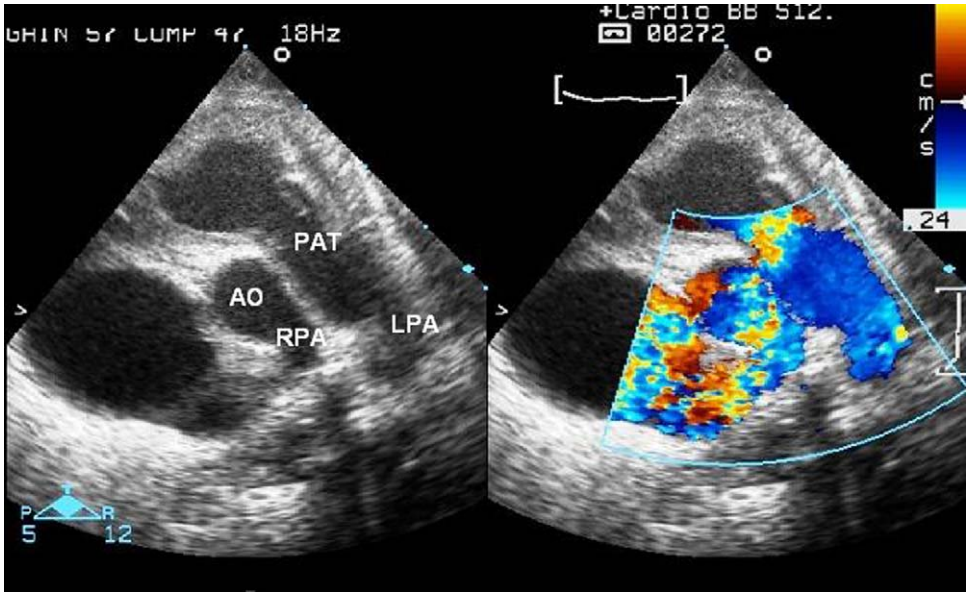

Figure 3. Parasternal short-axis view without (left) and with colour Doppler (right). The right pulmonary artery arises from the aorta while the left pulmonary artery arises from the pulmonary trunk. Ao: aorta; LPA: left pulmonary artery; PAT: pulmonary artery trunk; RPA: right pulmonary artery(Supplementary data).

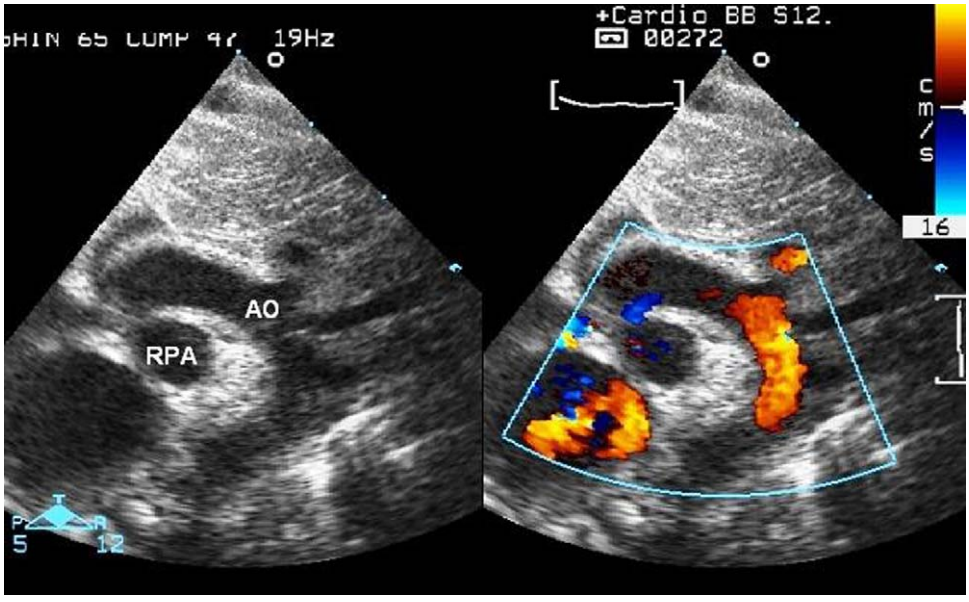

Figure 4. Suprasternal view without (left) and with colour Doppler (right). The right pulmonary artery arises from the posterior wall of the aorta. The colour Doppler scan shows a diastolic reflux from the aortic arch going to the right pulmonary artery. Ao: aorta; RPA: right pulmonary artery(Supplementary data).

either directly or with a graft to the main pulmonary artery. After few weeks of age, in infants who survive, shunt symptoms decrease because of pulmonary vascular obstructive disease, which contraindicates surgical correction.

Conflict of interest

None.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.acvd.2010.01.008.