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Brief Report

Prenatal diagnosis of anomalous origin of the right pulmonary artery from the ascending aorta

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Abstract We report a case of anomalous origin of the right pulmonary artery from the ascending aorta that was diagnosed by fetal ultrasound at 21 weeks of gestation. The clue to the diagnosis was present in the three-vessel view, this being one of the views that we use for fetal cardiac screening. The anomaly was corrected surgically at 11 days of age. We discuss the importance of prenatal diagnosis in the management of this rare anomaly.

Keywords: Hemitruncus; fetal echocardiography; three-vessel view; neonatal correction

ANOMALOUS ORIGIN OF THE RIGHT PULMONARY artery from the ascending aorta is a rare congenital malformation that is usually associated with high rate of mortality related to rapid development of obstructive pulmonary vascular disease.[1–4] Early recognition of this rare anomaly, and early surgical intervention, is therefore very important. There is no doubt that management would be much better if the diagnosis could be made at fetal examination. We herein describe an example of this lesion that was diagnosed by fetal ultrasound, permitting timely postnatal surgical treatment.

Case report

A 28-year-old woman was referred to us at 21 weeks of gestation for fetal echocardiography because her previous baby died of untreated hypoplastic left heart syndrome at one month of age. The three-vessel view, one of our primary views used for fetal cardiac screening,[5] demonstrated that the right pulmonary artery arose anomalously from the posterior wall of the ascending aorta (Fig. 1). The left pulmonary artery arose normally from the pulmonary trunk and the arterial duct was patent and left-sided. The heart was normal in size, and showed no other structural or functional abnormalities. On follow-up examination at 34 weeks of gestation, the heart was mildly enlarged. A baby girl was delivered spontaneously at full-term, weighing 3.4 kg. On physical examination, she was well developed and nourished. She was not cyanotic, but mildly tachypneic with a respiration rate of 50 breaths per minute. A systolic murmur graded at two in a scale of six was audible along the left sternal border. Echocardiography confirmed the fetal diagnosis, and demonstrated a widely patent left-sided arterial duct. On follow-up, she became more tachypneic with the respiration increasing to 60 breaths per minute. The chest radiograms showed a moderately enlarged cardiac silhouette, and increased pulmonary vascularity on both sides. She was treated with furosemide. On the 11th day of age, she underwent an elective operation. The right pulmonary artery was anastomosed end-to-side to the pulmonary trunk, the opening in the ascending aorta was closed, and the patent arterial duct was clipped. The postoperative course was uneventful, but follow-up echocardiography revealed stenosis at the site of anastomosis of the right pulmonary artery to the pulmonary trunk with a peak pressure gradient of 45 mmHg. The stenosis was successfully dilated with a balloon at 4 months of age. There is now no residual pressure gradient.

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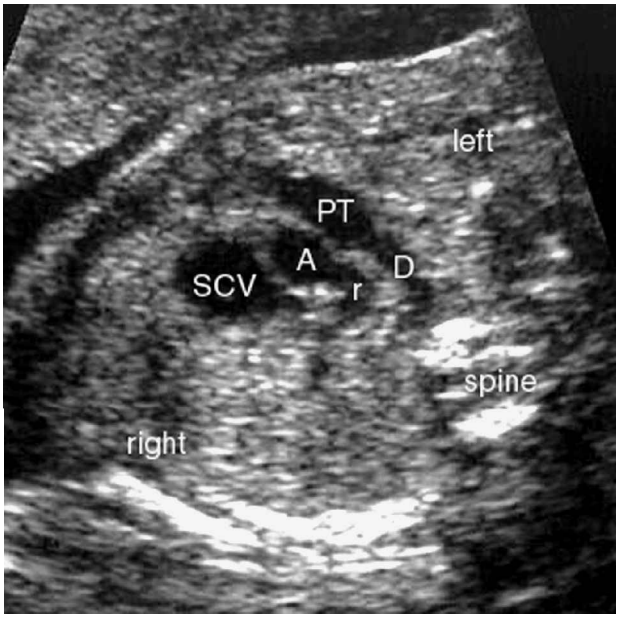

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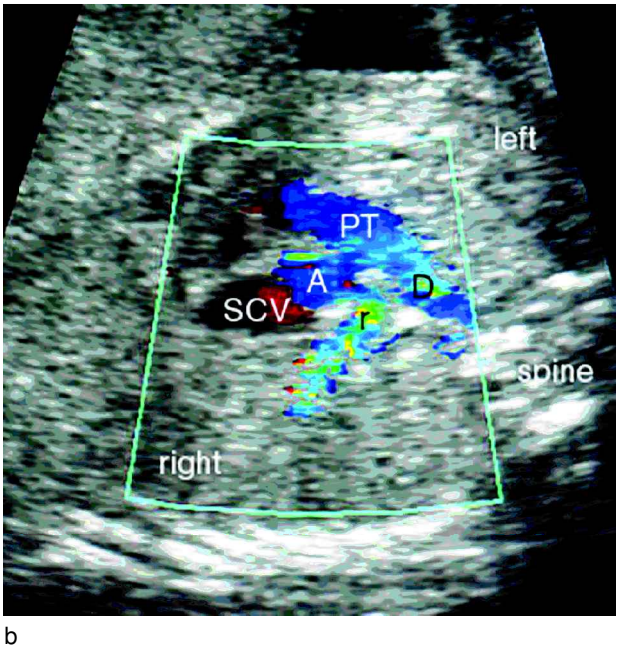

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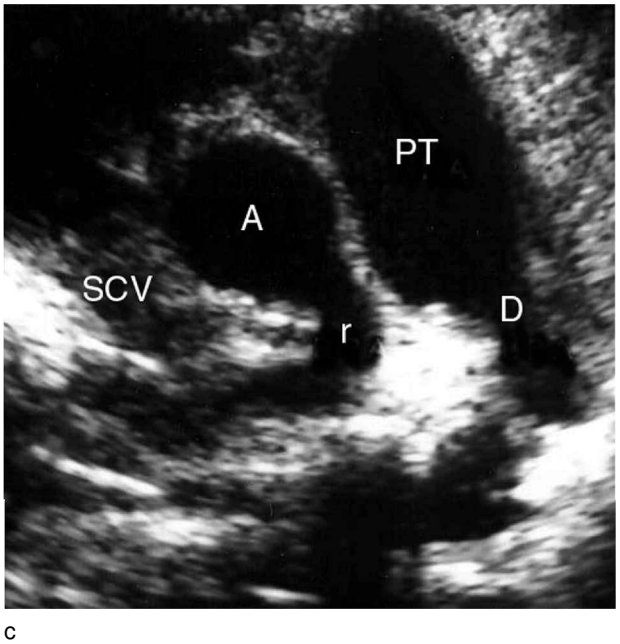

c

Figure 1.
Fetal (a & b) and postnatal (c) echocardiograms in three-vessel plane showing anomalous origin of the right pulmonary artery (r) from the ascending aorta (A). The arterial duct (D) is patent between the pulmonary trunk (PT) and the descending aorta in both fetal and postnatal studies. SCV = superior caval vein.

Discussion

Origin of the right pulmonary artery from the ascending aorta is a rare congenital malformation. It is commonly associated with patency of the arterial duct, aortopulmonary window, interruption of the aortic arch, or tetralogy of Fallot. As severe obstructive pulmonary vascular disease develops as early as 1 month of age not only in the lung fed by the anomalous artery but also in the lung fed from the pulmonary trunk, early surgical intervention is necessary, which mandates early diagnosis.[1–4] Although the postnatal diagnosis can be made without difficulty, the patients can escape diagnosis because of predominance of the symptoms and signs of pulmonary hypertension.[6,7] The ideal sequence would be the diagnosis at routine obstetric ultrasound, followed by referral of the baby to a tertiary cardiac center immediately after or before birth. This, of course, necessitates accurate fetal diagnosis.

In this respect, the three-vessel view, which Yoo et al.[5,8] introduced as one of the basic views for fetal cardiac screening, is useful. It is an orthogonal transverse view of the fetal mediastinum where the oblique section of the pulmonary trunk, and the

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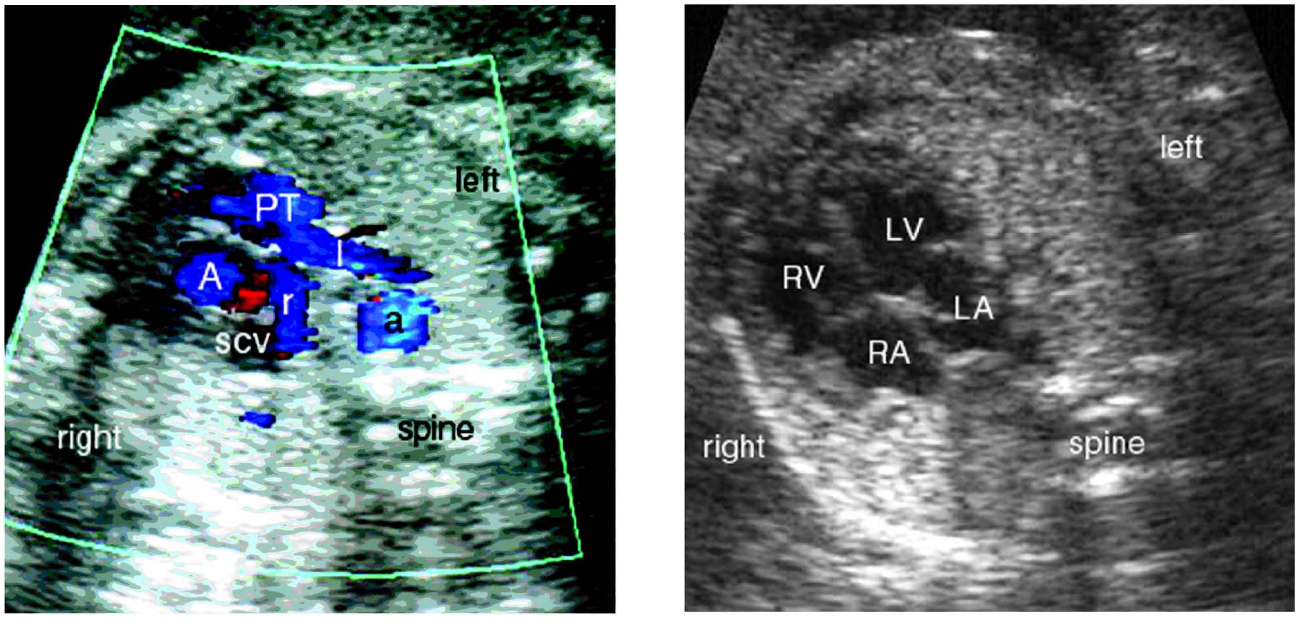

a b

Figure 2.
Fetal echocardiograms from a fetus with stenosis of the right pulmonary artery. On three-vessel view (a), the right pulmonary artery (r) was wrongly considered to arise from the ascending aorta (A). The postnatal examination revealed that the right pulmonary artery had a normal origin from the pulmonary trunk (PT) but was severely stenotic at its origin. The four-chamber view (b) shows that the heart is displaced to the right because of hypoplasia of the right lung. l = left pulmonary artery, SCV = superior caval vein, LA = left atrium, LV = left ventricle, RA = right atrium, RV = right ventricle.

cross sections of the ascending aorta and superior caval vein, are aligned in an oblique straight line from left and anterior to right and posterior. It provides important clues to the diagnosis of the malformations involving ventricular outflow tracts and great vessels. In the present case, the definitive diagnosis was made from a three-vessel view where the origin of the right pulmonary artery from the ascending aorta could clearly be defined. After we experienced the case described herein, we encountered another fetus having a similar fetal echocardiographic finding (Fig. 2). In this second case, postnatal echocardiography and angiography proved that the right pulmonary artery arose from the pulmonary trunk but was very stenotic. The misinterpretation at fetal examination of this case was due to the fact that the lumen of the stenotic segment was not visualized, while there was a false signal dropout of the walls of the right pulmonary artery and the adjacent ascending aorta. Despite this false positive diagnosis, our cases emphasize the importance of the three-vessel view as one of the basic cardiac screening views at fetal ultrasound, and demonstrate the impact on prenatal diagnosis in subsequent management of prenatal diagnosis.

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