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BRIEF COMMUNICATION

Aorta to Right Atrial Tunnel with Postnatal Transcatheter Treatment

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Abstract An antenatal referral for fetal echocardiography at 20 weeks of gestation with the suspicion of a cyst in the right atrium led to the diagnosis of an aorta to right atrial communication in a fetus. It was considered to be right coronary to right atrial fistula by fetal echocardiography. There was no other fetal anomaly and the cardiac hemodynamics remained stable throughout pregnancy. Postnatally, there was a progressive enlargement of the tunnel that led to a decision for intervention. Aortography showed an anterior aorta to right atrial tunnel, which was occluded in the catheterization laboratory. We describe in this report, the antenatal findings noted in ultrasound imaging along with the postnatal presentation and management of this rare congenital anomaly.

Keywords Aorta to right atrial tunnel · Fetal echocardiography · Transcatheter occlusion · Coronary artery-right atrial fistula

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Introduction

Aorta to right atrial tunnel, a very rare congenital anomaly first reported in 1980, is grouped into commoner posterior tunnels arising from left sinus and rarer anterior tunnels arising from the right sinus [1]. Only two cases of prenatal detection have been reported till date to the best of our knowledge. Progression into fetal heart failure was reported in one and the other fetus had an uneventful in utero course similar to the case described below [2, 3].

Report of Case

A primigravida mother with no antenatal risk factors was referred at 20 weeks of gestation for fetal echocardiography with the suspicion of a cyst in the right atrium of the fetal heart. Fetal imaging showed normal situs and segmental anatomy. Right atrium looked larger than usual with normal vena caval and tricuspid flow. A cystic structure in the right atrial cavity measuring 6.3 × 6 mm in size was seen in the four-chamber view located close to the interatrial septum superior to fossa ovalis (Fig. 1a). In the left ventricular outflow view a tubular structure was seen at the right atrioventricular groove, which seemed to end in the right atrial cystic cavity. Further imaging with color Doppler showed blood flow from the ascending aorta into the tubular structure which ended in the right atrial cavity (Fig. 1b). The end on view of this channel before emptying into the atrium had simulated a cyst in B-mode imaging. The channel had constrictions with flow turbulence at multiple sites. Due to the location near the right atrioventricular groove, it was considered to be right coronary artery to right

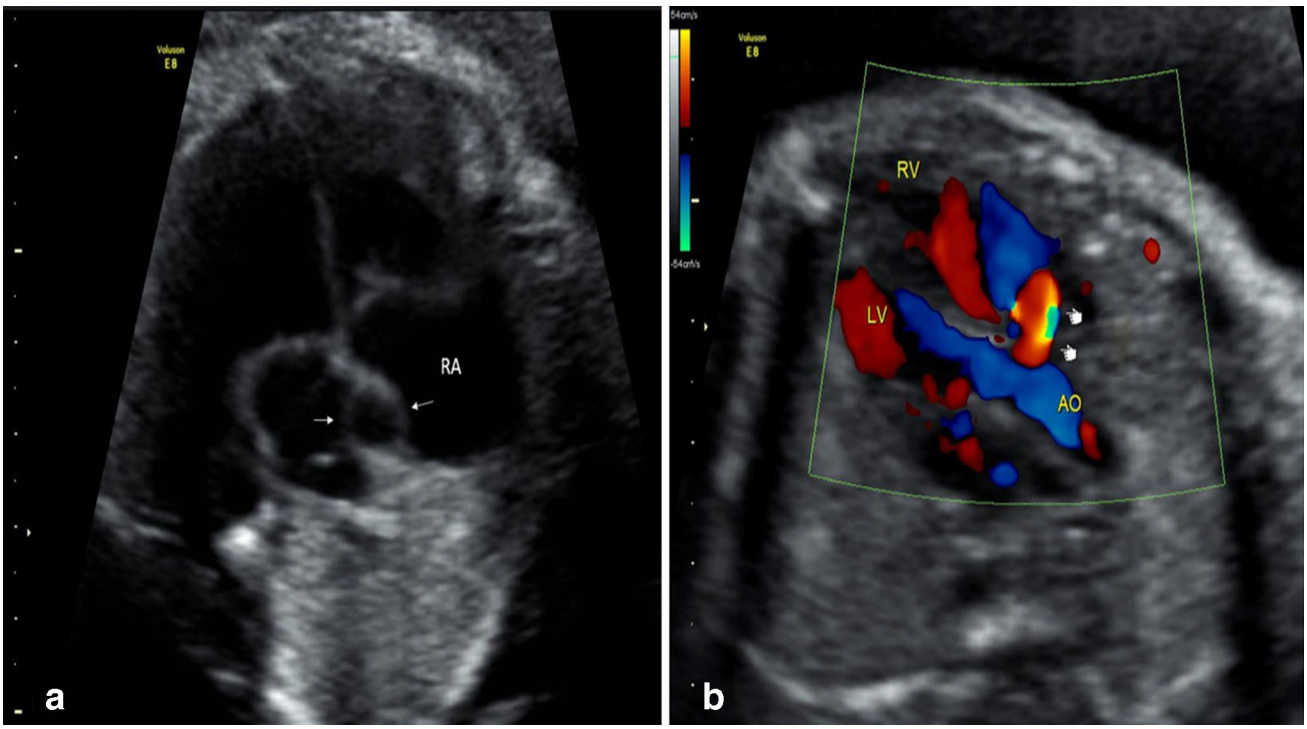

Fig. 1 **a** Apical four-chamber view in fetal echocardiography showing circular cystic structure (*arrows*) close to the interatrial septum in the right atrial cavity. **b** Color Doppler in left ventricular outflow view of fetal heart shows tubular structure from the ascending aorta (*hand arrows*) with flow away from aorta. *RA* right atrium, *AO* aorta, *RV* right ventricle, *LV* left ventricle

atrial fistula and family was counseled accordingly. Regular in utero monitoring was done which did not show any worsening in hemodynamics till delivery. Postnatal evaluation by transthoracic echocardiography was reported as right coronary to right atrial fistula and the child was on regular follow-up.

At 1½ years of age, the child developed symptoms and progressive enlargement of the tunnel to more than 2 cm on echocardiography. In view of the increasing size, angiography was performed for delineating anatomy. Aortography showed normal right and left coronary arteries and a large tunnel was seen arising from the anterior right aortic sinus opening in the right atrial cavity with two levels of mild constrictions, one near the aorta and the other at the right atrial end (Fig. 2a). Closure of the tunnel at one end alone would result in a thrombus laden tunnel sac with attendant risk of clot migration. So it was occluded at both ends using two ADO I occluder devices (St. Jude Medical, Plymouth, MN) that trapped the clot between the devices (Fig. 2b). The proximal device was carefully positioned not to encroach on the right coronary ostium which arose close to the aortic origin of the tunnel. On a two-year follow-up, there is complete closure with normal heart size and function.

Discussion

Embryogenesis of this defect remains unclear with a proposed abnormality in the development of outflow cushions or the inter-leaflet triangle at the aortic root [4, 5]. Limited numbers of cases have been reported after birth and surgical or transcatheter occlusion of the shunt seems possible with reasonable outcome [6–9]. From imaging point of view, it can be challenging to differentiate aorta to right atrial tunnel from coronary artery to right atrial fistula in fetal ultrasound as both abnormalities are seen in the right atrioventricular junction and coronary artery imaging is usually difficult. The challenge in echocardiographic imaging might persist even after birth due to the tunnel masking the smaller coronary artery. Ruptured sinus of Valsalva aneurysm can also look similar in ultrasound but not reported in fetal life. Confirmation in postnatal life is usually by angiographic methods. Hence, it may be better

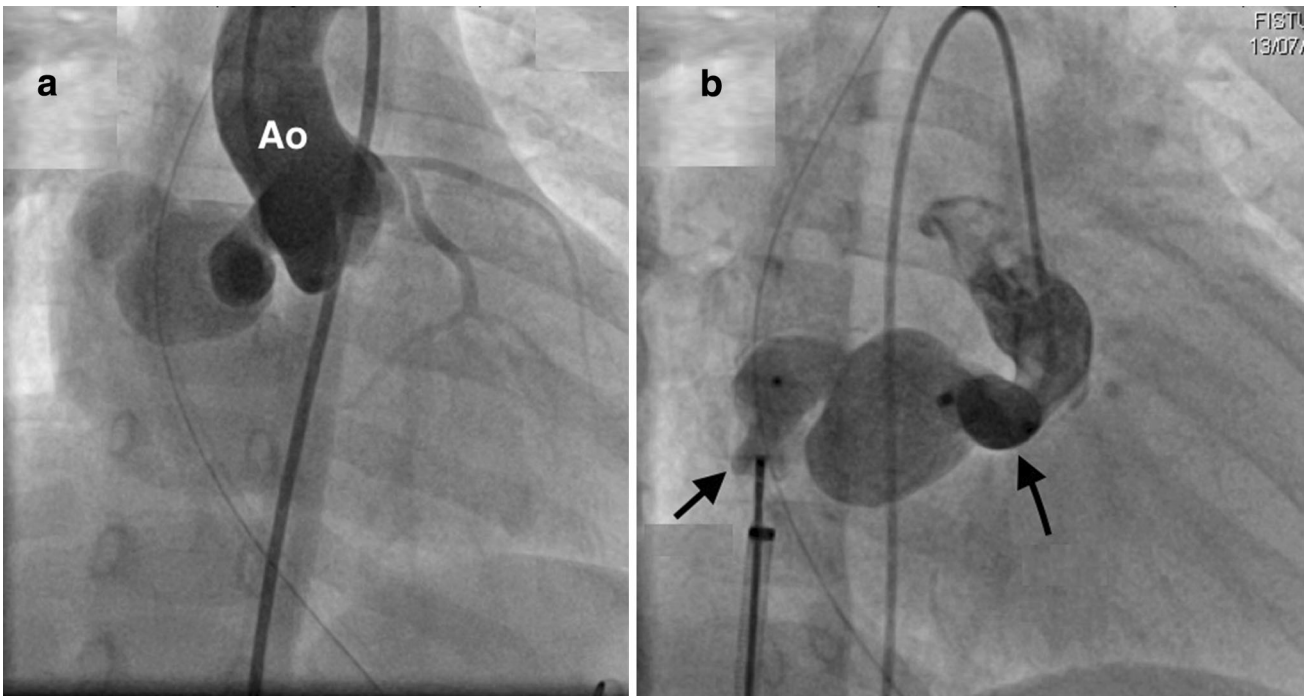

Fig. 2 Postnatal angiography images during transcatheter closure of the tunnel. **a** Aortography showing a large tunnel arising from the anterior right aortic sinus opening into the right atrial cavity. **b** Two occluder devices (*arrows*) deployed one at right atrial opening and the other at the proximal constriction. *Ao* ascending aorta

to consider this anomaly as a differential diagnosis for coronary artery to right atrial fistula in fetal life and in utero monitoring is warranted for optimal postnatal management.

Compliance with Ethical Standards

Conflict of interest None.

Summary Statement Parents of the child described in this case have given informed written consent for publication.

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