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Case report

Aortopulmonary window and right aortic arch in a fetus

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Abstract

We present the first description of a prenatal diagnosis of aortopulmonary window in association with right aortic arch and perimembranous ventricular septal defect. The diagnosis was made with fetal echocardiogram and subsequently confirmed using 3D motion-corrected fetal cardiac magnetic resonance imaging and postnatal echocardiogram. The genetic testing revealed normal array CGH, and the child underwent successful surgery in the neonatal period.

Key words: aortopulmonary window, congenital heart disease, right aortic arch, fetal heart.

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Aortopulmonary window (APW) is a rare congenital cardiac abnormality in which there is a failure of formation of the separate and adjacent walls of the intrapericardial arterial trunks, with persistence of the aortopulmonary foramen [1]. It occurs either in isolation or in combination with other cardiac anomalies such as transposition of the great arteries, tetralogy of Fallot, or interrupted aortic arch [2–4]. In this paper we focus on the first description of a prenatal diagnosis of APW in association with right aortic arch and perimembranous ventricular septal defect (Figure 1). Nuchal translucency was normal and risk for aneuploidies was low at the first trimester. There were no extracardiac anomalies found on subsequent ultrasound scans. An abnormal appearance of the three vessel and tracheal view was noted during a growth scan at 32 weeks of gestation. The diagnosis of APW was made on fetal echocardiography and confirmed on fetal cardiac magnetic resonance imaging (Figures 1C and 2), which is adjunct to our clinical practice [5].

The baby was born at term, no dysmorphic features were present, and the array CGH was normal. Postnatally the cardiac diagnosis was confirmed by transthoracic echocardiogram, and no further imaging was required. The neonate developed symptoms of congestive heart failure and was commenced on diuretics at two weeks of age. Successful cardiac surgery was accomplished at three weeks of age, comprising closure of the ventricular septal defect and the large, akin type II (distal) [6], aortopulmonary window (Figure 3) with Gore-Tex patches. Two years later the infant is thriving with no ongoing cardiac symptoms.

Aortopulmonary window is a challenging diagnosis to make on echocardiography pre- and postnatally. Our described case also has a right aortic arch, which is an unusual combination and has not been demonstrated prenatally. The three-vessel and tracheal view is a key view for prenatal ultrasound screening. However, care should be taken to ascertain that the heart and

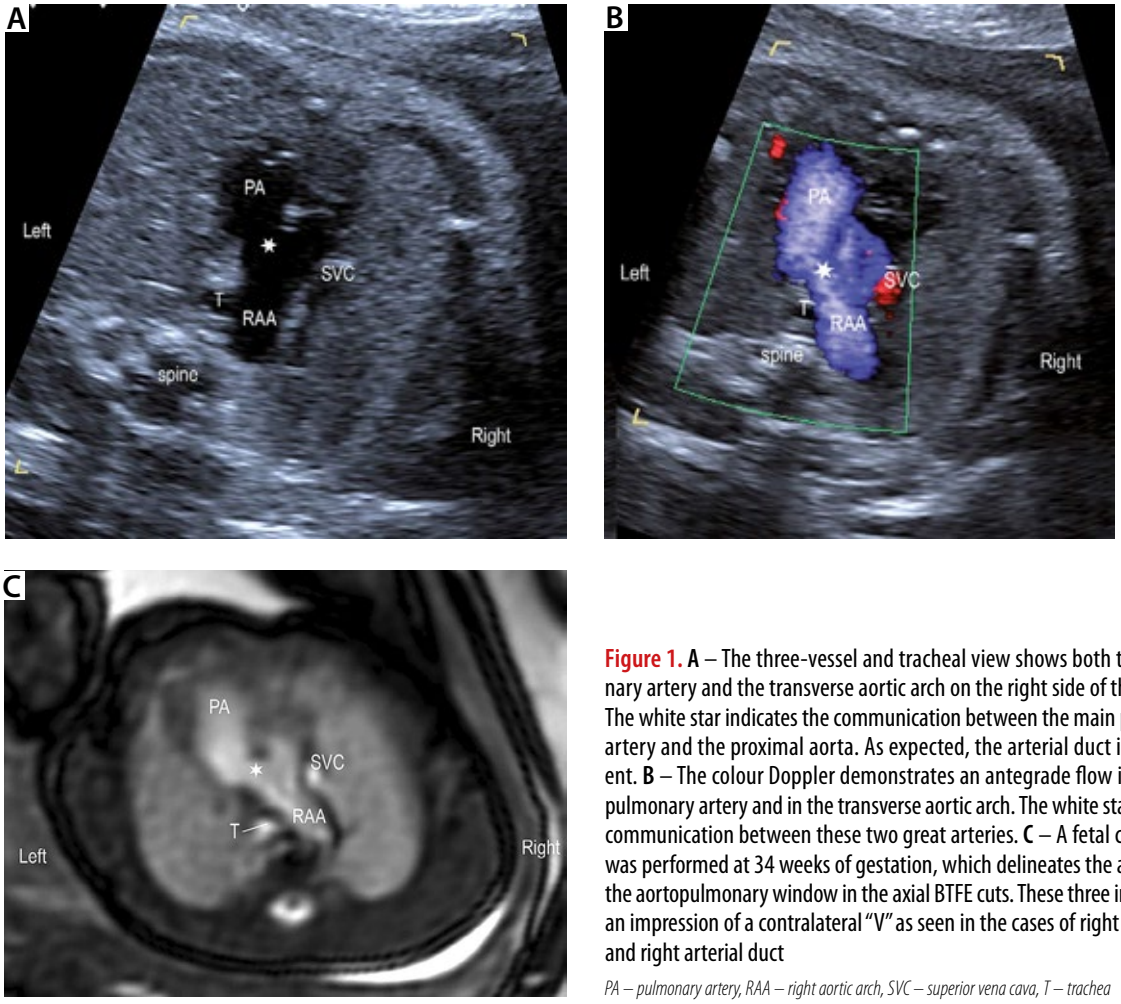

Figure 1. **A** – The three-vessel and tracheal view shows both the pulmonary artery and the transverse aortic arch on the right side of the trachea. The white star indicates the communication between the main pulmonary artery and the proximal aorta. As expected, the arterial duct is not present. **B** – The colour Doppler demonstrates an antegrade flow in both the pulmonary artery and in the transverse aortic arch. The white star indicates communication between these two great arteries. **C** – A fetal cardiac MRI was performed at 34 weeks of gestation, which delineates the anatomy of the aortopulmonary window in the axial BTFE cuts. These three images give an impression of a contralateral "V" as seen in the cases of right aortic arch and right arterial duct

PA – pulmonary artery, RAA – right aortic arch, SVC – superior vena cava, T – trachea

the "V" sign are on the ipsilateral side to avoid overlooking the combination of right aortic arch and right arterial duct.

In this case (Figure 1) the three-vessel and tracheal view shows both the pulmonary artery and the transverse aortic arch on the right side of the trachea. A large well-demarcated communication between the pulmonary artery trunk and the proximal aorta is visualised. There is no arterial duct seen on either the left or the right side joining the descending aorta.

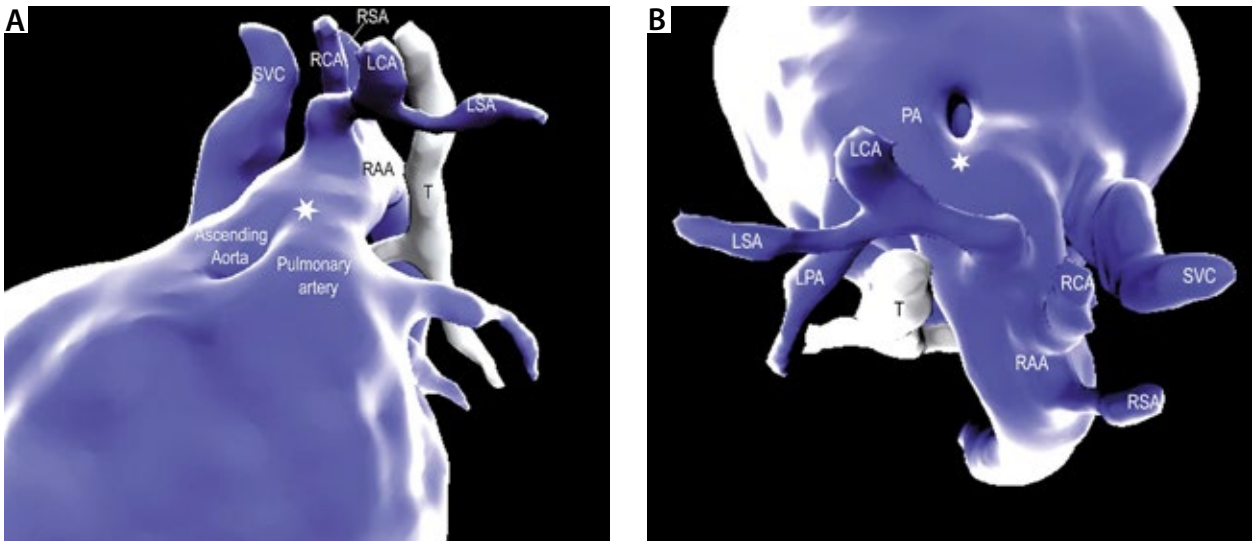

Figure 2. Fetal cardiac MRI demonstrates the anatomy in 3D using motion-corrected segmentation. The left lateral view (**A**) and the superior view (**B**) show the right-sided aortic arch with mirror image head and neck artery branching and a large communication between the main pulmonary artery and the proximal aorta. There is no arterial duct present

LCA – left carotid artery, LPA – left pulmonary artery, LSA – left subclavian artery, PA – pulmonary artery, RAA – right aortic arch, RCA – right carotid artery, RSA – right subclavian artery, SVC – superior vena cava, T – trachea

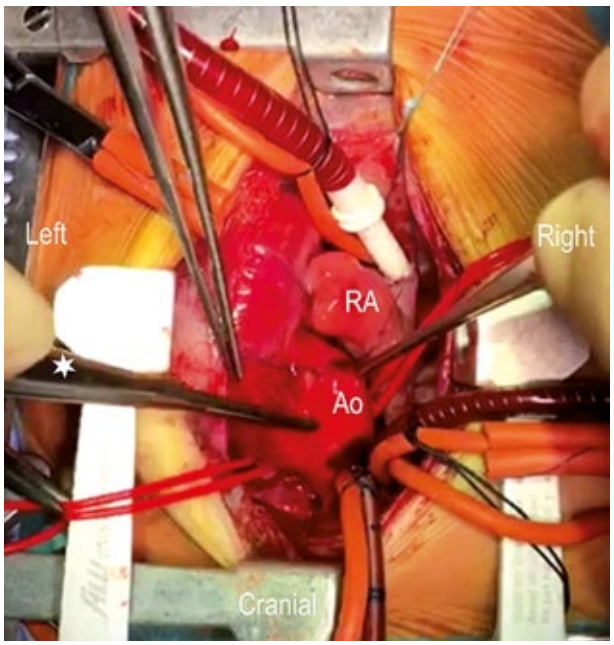

Figure 3. Surgical approach via a medium sternotomy with the patient on cardiopulmonary bypass is shown. The forceps on the left (asterisk) point to the large aortopulmonary window before repair

Ao – aorta, RA – right atrial appendage

Conversely, a right arterial duct would arise from the posterosuperior aspect of the junction of the pulmonary trunk and right pulmonary artery and course posteriorly, on the right of the trachea, joining the junction of the right aortic arch and descending aorta distal and opposite to the origin of the right subclavian artery.

Such a diagnosis is unlikely to have cardiorespiratory implications after birth. Contrary to this, an APW will result in congestive cardiac failure and may lead to irreversible pulmonary vascular disease if diagnosed late. Thus, prenatal diagnosis of APW would lead to timely postnatal management with potentially good long-term outcome.

Compliance with ethical standards

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Conflict of interest

The authors declare no conflict of interest.

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