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*Case Report*

# Prenatal diagnosis of aortopulmonary window associated with aberrant subclavian artery

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**Abstract** Aortopulmonary window is a rare cardiac developmental anomaly characterised by a communication between the ascending aorta and the pulmonary artery. Aortopulmonary window may be isolated or associated with cardiac defects such as ventricular septal defect, atrial septal defect, interrupted aortic arch, and tetralogy of Fallot. We report a case of aortopulmonary window associated with aberrant subclavian artery based on fetal two-dimensional echocardiogram. The mother was referred for fetal echocardiography because of multiple fetal anomalies. Prenatal echocardiography at 30 weeks of gestation revealed a defect between the main and right pulmonary arteries and the ascending aorta (type III). The patient was born at 38 weeks of gestation via caesarean delivery, and was admitted to the neonatal intensive care unit because of respiratory failure and multiple congenital anomalies. Postnatal echocardiogram and cardiac MRI confirmed the prenatal findings. In addition, this patient had severe Dandy–Walker malformation and renal anomalies with poor prognosis. The family decided to withdraw respiratory care support on day of life 4, and the neonate passed away shortly after.

Keywords: Aortopulmonary window; aortopulmonary septal defects; CHD

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## Case description

A 26-year-old woman – gravida 3, para 1, abortion 1 – with history notable for bipolar disorder and marijuana use underwent an anatomy scan at 28 weeks of gestation. The anatomy ultrasound revealed bilateral urinary tract dilation of low risk and severe bilateral ventriculomegaly. A fetal echocardiogram was performed at 30 weeks of gestation because of multiple fetal anomalies. The fetal heart showed situs solitus and levocardia, with normal venoatrial, atrioventricular, and ventriculoarterial connections. A 6 × 8-mm aortopulmonary window was visualised (Figs 1–3). Color and pulsed Doppler examinations confirmed unrestrictive flow across the defect. There was also evidence of an aberrant right subclavian artery on color flow mapping.

Delivery was by repeat caesarean delivery at 38 weeks of gestation due to severe hydrocephalus. A male infant was born with complications, weighing 5 kg with a length of 52.5 cm. Extracardiac malformations included severe hydrocephalus – Dandy–Walker malformation – vertebral anomalies, and bilateral urinary tract dilation of the kidneys. Postnatal chromosome analysis revealed a normal karyotype, but the microarray reported a pathogenic terminal gain in 7p and terminal loss in 6p. The postnatal echocardiogram and cardiac MRI confirmed prenatal diagnosis of type III aortopulmonary window and aberrant right subclavian artery.

## Discussion

Aortopulmonary window is one of the more rare forms of CHD comprising up to 0.2% of all live

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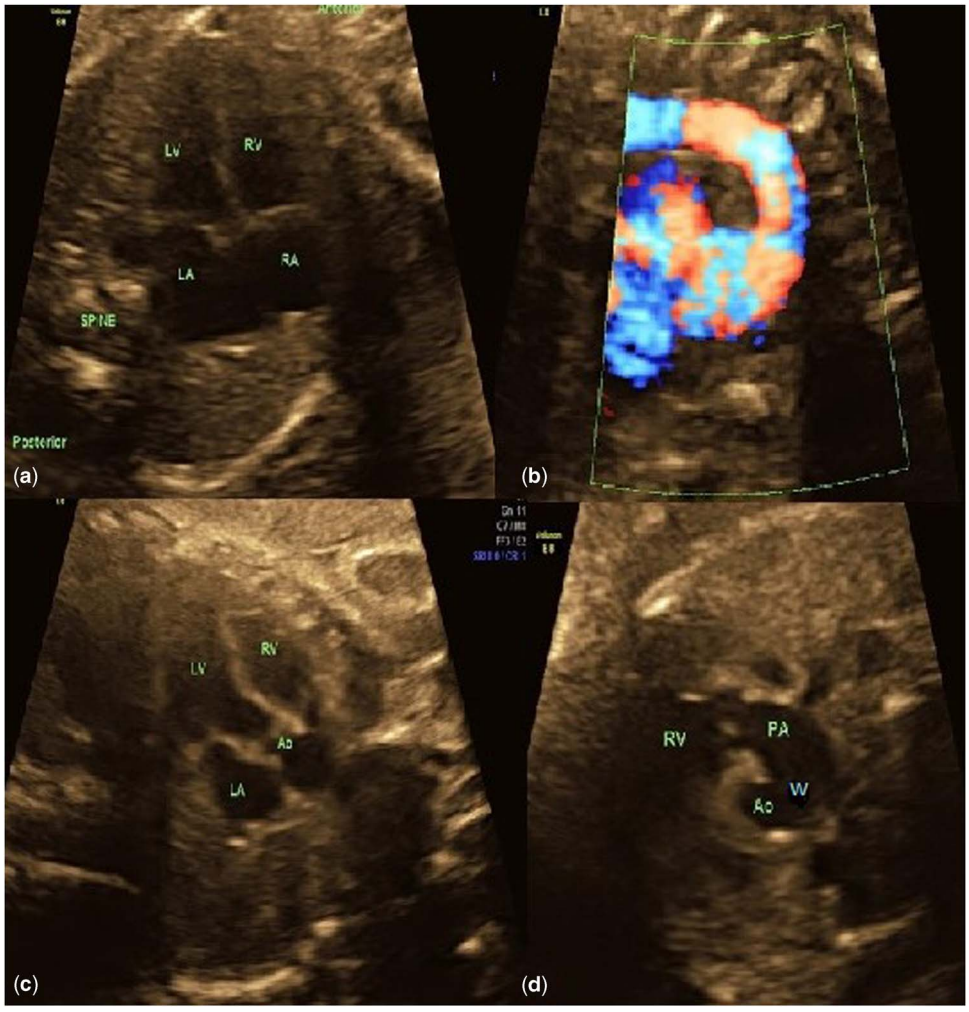


**Figure 1.**
*Fetal echocardiogram at the time of diagnosis at 30 weeks of gestation. (**a**) Normal four chamber. (**b**) Aortic arch. (**c**) Left ventricular outflow tract. (**d**) Right ventricular outflow tract in the short-axis view demonstrating the aortopulmonary window. AO = aorta; LA = left atrium; LV = left ventricle; PA = pulmonary artery; RA = right atrium; RV = right ventricle; W = window.*

birth cardiac defects.[1,2] Aortopulmonary window is defined as an opening or a communication between the ascending aorta and the pulmonary artery in the presence of two anatomically separate semilunar – aortic and pulmonary – valves. The Society of Thoracic Surgeons Congenital Heart Surgery Database Committee for aortopulmonary window recommended a classification system for aortopulmonary window (Table 1).[3] With fetal echocardiography, early detection is possible; however, without heightened suspicion, an aortopulmonary window may be missed, especially in fetuses with complex cardiac malformations and ever increasing maternal obesity.[2] Aortopulmonary window is associated with concomitant CHD in 25–35% of the time.[1,2] The most common of these include tetralogy of Fallot, ventricular septal defects, and aortic arch abnormalities,[1] but atrial septal defects, patent ductus arteriosus, and coronary artery anomalies have also been noted.[4] Early fetal diagnosis and treatment is critical in order to improve neonatal outcomes. This helps avoid congestive heart failure, which can occur as early as the 1st week of neonatal life and development of irreversible pulmonary hypertension.[5] Surgical repair is recommended as soon as diagnosis of aortopulmonary window is established.[3] As with all CHD, survival rates depend on concomitant defects and genetics and overall are reported to be close to 90%.[1]

## Conclusion

Visualising the aortopulmonary septum during a fetal echocardiographic examination is essential to allow for diagnosis of a very rare but critical fetal

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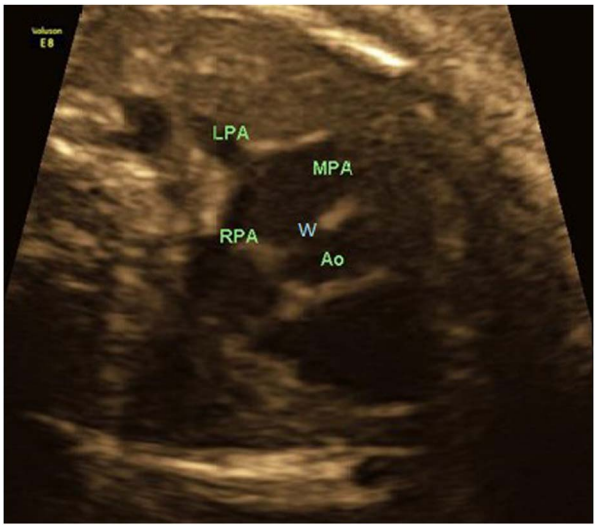


**Figure 2.**
*Fetal echocardiogram demonstrating the aortopulmonary window in the long axis. AO = aorta; LPA = left pulmonary artery; MPA = main pulmonary artery; RPA = right pulmonary artery; W = window.*

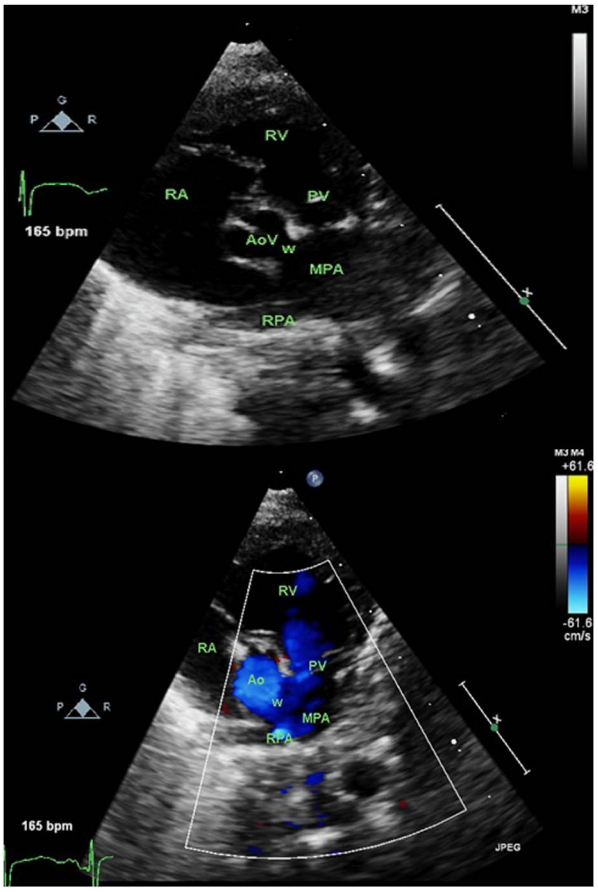


**Figure 3.**
*Postnatal echocardiogram demonstrating the aortopulmonary window in the short axis with and without color flow. AO = aorta; AoV = aortic valve; bpm = beats per minute; MPA = main pulmonary artery; PV = pulmonary vein; RA = right atrium; RPA = right pulmonary artery; RV = right ventricle; W = window.*

Table 1. Aortopulmonary window classification system*.

| | |
|---|---|
| Type I – proximal defect | Proximally located a few millimetres from the semilunar valves |
| Type II – distal defect | Defect located in the uppermost portion of the ascending aorta and overlies the portion of the right pulmonary artery |
| Type III – total defect | Defect involves the majority of the ascending aorta |
| Type IV – intermediate defect | Located at the middle of the ascending aorta, most suitable for device closure |

* Adapted from Backer and Mavroudis[3]

diagnosis. Prenatal diagnosis of aortopulmonary window is important because early corrective surgery in the neonatal period prevents congestive heart failure and pulmonary vascular obstructive disease.

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## Conflicts of Interest

None.

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