

Prenatal findings of aortico-left ventricular tunnel: the good and the ugly

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Objective

To describe variable prenatal findings, management and outcome of aortico-ventricular tunnels.

Methods

Retrospective study focusing on prenatal presentation, follow-up, postnatal management and clinical outcome of two fetal cases with aortico-left ventricular tunnel.

Results

Two cases of fetuses with an aorta-to-left ventricular tunnel were identified after review of our databases. Both fetuses were male and were referred for fetal echocardiograms due to ventricular size discrepancy and for ascending aorta dilation. The first one was seen at 32⁺² weeks gestational age (GA) and carried the diagnosis of critical aortic stenosis and aortic insufficiency with mild to moderate hypoplastic left ventricle (LV), functional mitral valve stenosis/atresia and LV endocardial fibroelastosis and severe LV systolic dysfunction. There was also a thinned out membranous ventricular septum and moderate tricuspid regurgitation, a mildly dilated ascending aorta and retrograde flow in the transverse arch. Delivery was carried out to term and the definite diagnosis was made postnatally when an aorta-to-LV tunnel was identified. The newborn underwent bilateral pulmonary artery banding at 8 days of age, followed by closure of the tunnel, aortic valvotomy and placement of a ductal stent at two weeks of age. Due to worsening right ventricular dysfunction and non-compensated heart failure he finally underwent a heart transplantation at 6 months of age and continues to be alive almost five years out. The second case was referred at 23⁺³ weeks GA due to LV dilation, dilation of the ascending aorta and possible aortic insufficiency. The diagnosis of an aorta-to-LV tunnel was made, with moderate LV dilation and borderline LV systolic function as well as retrograde flow in the ascending aorta; the fetus underwent regular follow-up and was delivered at term with no signs of hydrops. The diagnosis was confirmed postnatally with the addition of the presence of LV non-compaction. Medical management was initially instituted with good response and the infant underwent successful, elective surgical repair and closure of the tunnel at 5 months of age. Follow-up at 4.5 years of age reveals an asymptomatic toddler with mild aortic insufficiency and mild residual LV dilation with mild systolic dysfunction.

Conclusion

Congenital aortico-ventricular tunnels pose a challenge when it comes to their prenatal diagnosis. Common presentation is ventricular discrepancy and ascending aorta dilation, usually associated with retrograde flow in the aorta and a competent aortic valve; associated myocardial dysfunction and non-compaction of the affected ventricle is not rare in that setting. Long-term outcome depends on the associated defects and the ventricular function, with closure of the tunnel being most of the times feasible.