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Brief Report

Aortico-left ventricular tunnel and left ventricular non-compaction: a case series

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Abstract Aortico-left ventricular tunnel and left ventricular non-compaction are rare congenital cardiac anomalies with varied clinical presentations and sparsely described co-existence in the medical literature. Owing to the limited information about these diagnoses in tandem, we present four cases of aortico-left ventricular tunnel and left ventricular non-compaction from our institution and discuss the clinical presentation, management, and follow-up.

Keywords: Aortico-left ventricular tunnel; left ventricular non-compaction; CHD; foetal echocardiography

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Aortico-left ventricular tunnel represents a rare congenital cardiac anomaly in which a connection exists between the aortic root, classically above the right coronary ostium, and the left ventricle, causing free aortic insufficiency with left ventricular volume overload.[1] Clinical presentation varies from acute neonatal heart failure or sudden death to symptomatic chronic compensated heart failure. Left ventricular non-compaction is an equally uncommon disorder characterised by failure of myocardial compaction resulting in hypertrabeculated myocardium, with non-compact/compact myocardium ratio greater than 2:1, and varied degrees of ventricular systolic performance.[2] The co-existence of aortico-left ventricular tunnel and left ventricular non-compaction is described in three isolated case reports.[3–5] Owing to the sparse information regarding an association between aortico-left ventricular tunnel and left ventricular non-compaction, we report four cases with varied presentation and outcomes.

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Case 1

Antenatal diagnosis of aortico-left ventricular tunnel with left and right ventricular dilation and moderately depressed left and right ventricular systolic function was made by foetal echocardiogram at 23 weeks of gestation, at which time maternal digoxin therapy was initiated. Repeat foetal echocardiogram at 27 weeks of gestation demonstrated significant left ventricular hypertrophy concerning for non-compaction and progression of left ventricular systolic dysfunction from moderate to severely depressed (Fig 1). Serial echocardiograms showed no change, and following term delivery the postnatal echocardiogram confirmed the diagnoses of aortico-left ventricular tunnel with the right coronary artery noted to arise from the tunnel and left ventricular non-compaction (non-compact/compact myocardium ratio 2.4:1) with mildly depressed systolic function (Fig 2). At 3 days of life, the patient underwent repair for the aortico-left ventricular tunnel by patching of the aortic portion of the tunnel. Owing to the fact that post-operative echocardiograms showed retrograde flow into the right coronary artery, cardiac catheterisation was performed demonstrating no residual aortico-left

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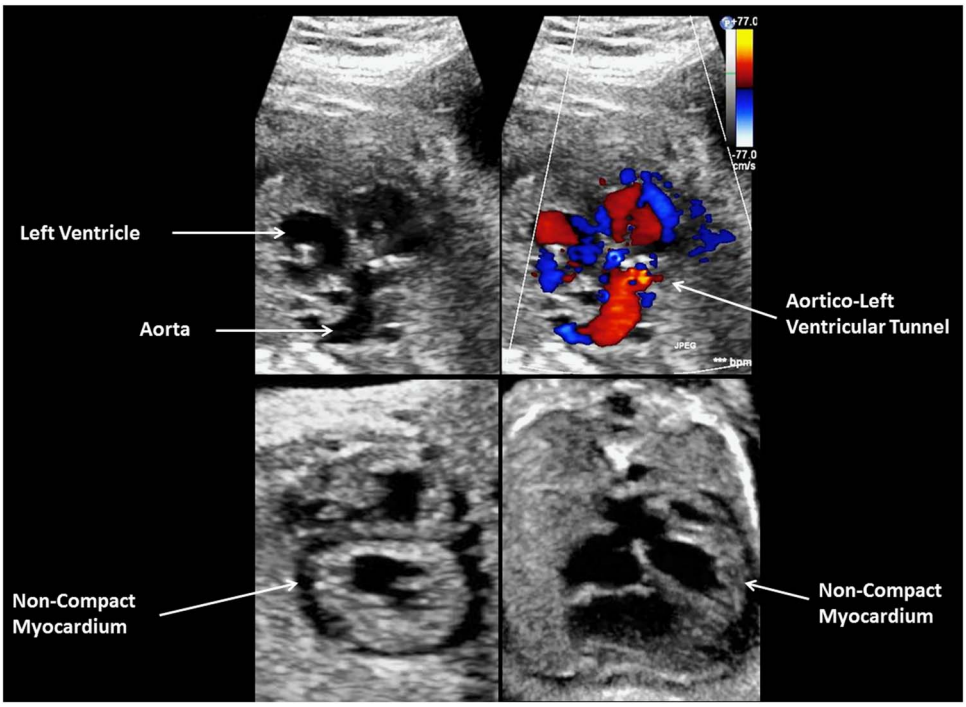

Figure 1.
Foetal echocardiogram. Prenatal echocardiogram demonstrating the aortico-left ventricular tunnel and increased left ventricular trabeculations consistent with non-compaction.

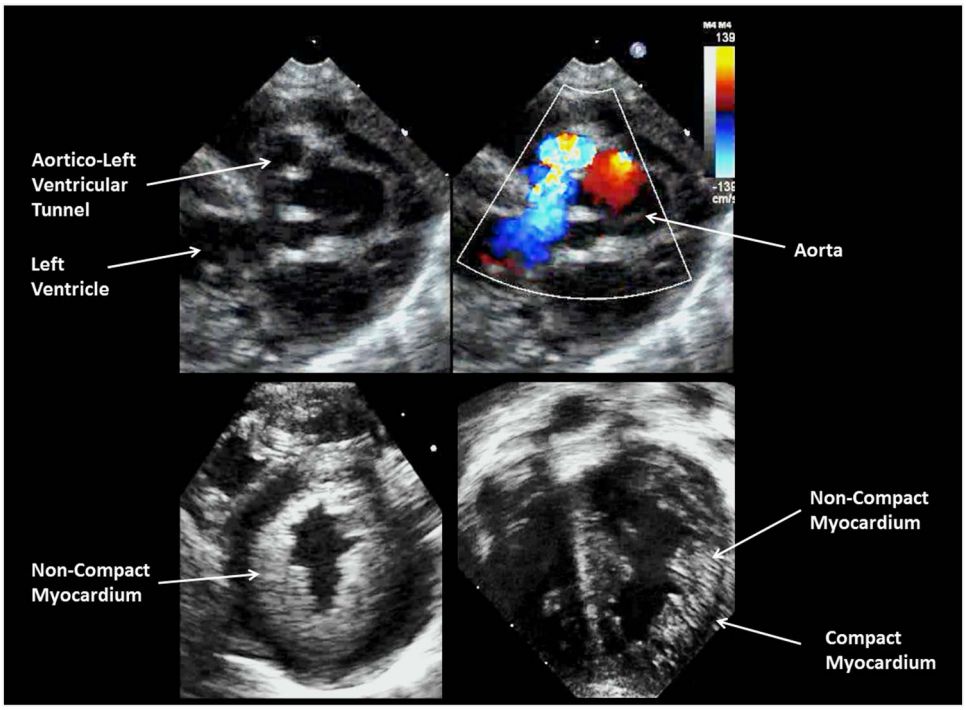

Figure 2.
Postnatal echocardiogram. Postnatal echocardiogram demonstrating the aortico-left ventricular tunnel and left ventriclar non-compaction.

ventricular tunnel, a patent right coronary artery arising from the left ventricular outflow tract portion of the tunnel (inferior to the patch), and retrograde filling of the right coronary artery from collateral vessels arising from the left anterior descending coronary artery. Discharge echocardiogram on post-operative

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day 11 showed mild-to-moderately depressed left ventricular systolic function with a regional wall motion abnormality of the anterior ventricular septum, no residual aortico-left ventricular tunnel, mild aortic insufficiency, and unchanged retrograde flow in the right coronary artery. The patient was discharged home on digoxin, captopril, and aspirin for thromboprophylaxis. At the 5-month follow-up, the patient has remained asymptomatic with stable ventricular function.

Case 2

A 14-year-old asymptomatic patient with a history of aortico-left ventricular tunnel status post neonatal repair and ascending aorta dilation was referred to our institution for aortic root replacement. Pre-operative echocardiogram showed left ventricular non-compaction (non-compact/compact myocardium ratio 2.6:1) with normal systolic function, mild aortic insufficiency, and severe dilation of the ascending aorta. The patient underwent valve sparing aortic root replacement, and the post-operative trans-oesophageal echocardiogram showed normal origins and colour flow into both coronary arteries and stable mild aortic insufficiency. The post-operative course was as expected with discharge home on post-operative day 5.

Case 3

Following antenatal diagnosis of aortico-ventricular tunnel with foetal congestive heart failure, patch closure of the aortico-ventricular tunnel was performed at 1 day of life. The post-operative course was complicated by persistent hypoxia requiring extra-corporeal membranous oxygenation for 4 days. The patient was discharged home at 26 days of age with an echocardiogram demonstrating no residual aortico-left ventricular tunnel, no aortic insufficiency, and left ventricular dilation and hypertrophy with severely depressed systolic function and septal dyskinesis. At follow-up after 2 decades, the patient developed chronic chest pain beginning at the age of 10 years. Serial echocardiograms demonstrated trace aortic insufficiency, stable mildly depressed left ventricular systolic function, and increasing trabeculations becoming consistent with left ventricular non-compaction (non-compact/compact myocardium ratio 2.8:1) at 19 years of age. Multiple Holter studies showed no dysrhythmia. Owing to ST segment depression on exercise testing, cardiac catheterisation was performed showing no residual aortico-left ventricular tunnel, elevated left ventricular end-diastolic pressure, and no evidence of right coronary artery patency. The patient has been medically managed with digoxin, enalapril, and aspirin for thromboprophylaxis and remains in NYHA class I with no limitations.

Case 4

A neonate with prenatal concern for congenital heart disease was transferred to our institution at 3 days of life with worsening congestive heart failure. Echocardiogram revealed aortico-left ventricular tunnel, severe aortic stenosis, and left ventricular non-compaction (non-compact/compact myocardium ratio 2.9:1) with severe systolic dysfunction. Following group discussion, the patient was determined a poor surgical candidate and listed for heart transplantation. Successful aortic balloon valvuloplasty was performed with resolution of recurrent metabolic acidosis and ventricular tachycardia; however, there was no improvement in ventricular function. The patient was maintained on continuous inotropic support and underwent successful orthotopic heart transplantation at 1 month of age.

Discussion

Aortico-left ventricular tunnel and left ventricular non-compaction are rare conditions with incidence of 0.1% and prevalence of 0.014%, respectively.[2,6] The mechanism of the relationship between these two conditions has not been well-delineated; however, their association is supported by the relationship of left ventricular non-compaction with congenital heart block and pregnancy, which cause ventricular volume overload.[2,7]

The four presented cases of aortico-left ventricular tunnel and left ventricular non-compaction include two foetal diagnoses, which was previously unreported, and provide previously undescribed follow-up of these diagnoses in tandem. The three previous isolated reports include the case of a neonate with resolution of left ventricular non-compaction following surgical closure of an aortico-left ventricular tunnel,[3] discussion of the diagnosis of these conditions by echocardiography in an adult,[4] and the description of transcatheter closure of an aortico-left ventricular tunnel in a child also with left ventricular non-compaction,[5] all with limited follow-up. This case series provides additional considerations for follow-up, including post-surgical right coronary artery flow, aortic root dilation, and observation for progressive ventricular dysfunction.

Conclusion

Aortico-left ventricular tunnel and left ventricular non-compaction are two uncommon cardiac conditions. The four cases presented above highlight an association,

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possibly related to ventricular volume overload, and demonstrate the concerns and complications requiring continued follow-up.

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Conflicts of Interest

None.

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