



Evolution in the management of aorta to left ventricular tunnel in a national congenital cardiology centre

Brief Report

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Abstract

An aorto-ventricular tunnel is a rare congenital cardiac defect, where a channel connects the lumen of the ascending aorta to the left or right ventricle. Four patients presented with an aorto-left ventricular tunnel over two decades at a median age of 8 months (range 0.1–10 months). Two patients (50%) had associated cardiac anomalies including hypoplastic left heart syndrome and left ventricular noncompaction/hypertrophic cardiomyopathy with aortic/pulmonary valve dysplasia in one patient each. Although traditionally surgical treatment has addressed this problem, management has evolved to transcatheter closure with excellent outcomes in appropriately selected patients at our national centre.

Aorto-ventricular tunnel is a rare congenital cardiac defect with an incidence of less than 0.1% of all children born with congenital cardiac disease.¹ The tunnel originates in the ascending aorta and bypasses the aortic valve creating a direct connection between the ascending aorta and either the right or left ventricular cavity. It was originally reported by Levy et al. who coined the phrase aorto-left ventricular tunnel.² The majority of cases originate above the right coronary cusp and present in the first year of life.¹ In one series, the median age at presentation was 25 days with one-third of patients presenting with moderate to severe aortic valve regurgitation.¹

Methods

This is a retrospective case series of all patients who were diagnosed with an aorto-left ventricular tunnel from 2001 to 2021 at the National Paediatric Cardiology Centre at Children’s Health Ireland, Crumlin, Dublin. Information about their diagnosis and subsequent treatment plans were collated. Informed consent was obtained from patient one to use the images for publication. The case series was approved by the Institutional Review Board at Children’s Health Ireland, Crumlin, Dublin.

Results

Four patients presented during the study period with aorto-left ventricular tunnel at a median age of 8 months (range 0.1–10 months) (Table 1). Two patients (50%) had associated cardiac anomalies including hypoplastic left heart syndrome and left ventricular noncompaction/hypertrophic cardiomyopathy with aortic and pulmonary valve dysplasia in one patient each. Management included surgical closure in the patient with associated noncompaction cardiomyopathy and dysplastic aortic and pulmonary valves. Management evolved to transcatheter closure with excellent outcomes in three other patients.

Catheterisation procedure

The cases were performed under general anaesthetic in the hybrid cardiac catheterisation laboratory. Heparin 100 units/kg was given once access was obtained. Access was obtained via the right femoral artery and the tunnel was crossed with a 0.035” tapered Terumo wire (Terumo UK, Surrey, UK) and this was subsequently exchanged out for an Amplatz super stiff wire (Boston Scientific, Clonmel, Ireland). A destination sheath (Terumo UK, Surrey, UK) was placed in the left ventricle and the appropriate device (AVP II or Amplatzer ductal occluder I) as outlined above was positioned. A device which was 2 mm bigger than the minimum diameter of the tunnel was chosen. The reason a particular device was chosen was based on the likelihood of disk protrusion into the aorta and potential coronary origin interference. The device position was confirmed with both angiographic and transoesophageal imaging. In all cases, there was an

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Table 1. Profile of Aorto-left Ventricular Tunnel Patients

Patient	Age at diagnosis	Gender	Presenting complaint	Imaging	Type	Associated anomalies	Treatment
1	8 months	Male	Murmur	Echocardiography to make diagnosis and CT angiogram to evaluate the relationship to the coronary arteries	1	Nil	Closure with Amplatzer vascular plug AVP II in catheterisation laboratory
2	Birth	Male	Antenatal concern	Echocardiography to make diagnosis and CT angiogram to evaluate the relationship to the coronary arteries	2	Severe hypertrophic cardiomyopathy and left ventricular non compaction cardiomyopathy with dysplasia of the pulmonary and aortic valves	Surgical repair with ultimate cardiac transplant for his remaining underlying cardiac anomalies
3	10 Months	Female	New diastolic murmur	Echocardiography to make diagnosis and CT angiogram to evaluate the relationship to the coronary arteries	1	Hypoplastic left heart syndrome. Status post Norwood/Sano and Glenn operations.	Closure with AVP II in catheterisation laboratory
4	9 months	male	Murmur	Echocardiography and confirmed with angiography in catheterisation laboratory	2	Nil	Closure with Amplatzer duct occluder I in the catheterisation laboratory

optimum final device position. Patients received aspirin at a dose of 5 mg/kg (max 75 mg) for 6 months after the procedure.

Discussion

Aorto-left ventricular tunnel is a rare congenital cardiac defect which has an uncertain aetiology but is thought to evolve from a maldevelopment of the cushions which give rise to the aortic and pulmonary roots and abnormal separation of these structures.³

Hovaguimian et al. proposed a classification in 1988 as a way to guide the surgical strategy.⁴ Type 1 is a simple tunnel with a slit-like opening at the aortic end and no aortic valve distortion. Type 2 is a large extracardiac aortic wall aneurysm of the tunnel with an oval opening at the aortic end, with or without valvular distortion. Type 3 has an intracardiac aneurysm of the septal portion of the tunnel, with or without right ventricular outflow tract obstruction. Type 4 is a combination of types 2 and 3.⁴ In our series, two patients had a type 1 aorto-left ventricular tunnel (patient one and three) and two patients had a type 2 aorto-left ventricular tunnel (patient two and four). This classification has been used subsequently by some institutions in guiding which cases are best suited to transcatheter closure,⁵ with recommendations for only closing type 1 aorto-left ventricular tunnel using a transcatheter approach.

The clinical presentation is variable with previous reports having demonstrated the heterogeneity of the presentations from asymptomatic patients referred with a murmur to acutely unwell neonates with complex cardiac anatomy.⁵ This case series confirms the previous reports and also highlights the importance of the clinical examination prior to echocardiography. Patients with aorto-left ventricular tunnel will often demonstrate “to and fro” murmurs associated with thrills and bounding pulses. This can be difficult to distinguish from severe aortic regurgitation but in aorto-left ventricular tunnel the second heart sound will be normal.³ The classic clinical examination findings complement the imaging modalities. Interestingly, one of our patients presented with ventricular noncompaction/hypertrophic cardiomyopathy phenotype in combination with dysplastic aortic and pulmonary valves which has been previously described in this setting.⁶

Echocardiography is the primary imaging modality and in the majority of cases is sufficient in making the diagnosis of aorto-left ventricular tunnel.⁷ We demonstrate (Fig 1 and 2) how useful echocardiography can be in establishing the diagnosis but also how important multi-modality imaging can be when presented with rare cardiac lesions. Trans-thoracic parasternal long-axis and parasternal short-axis views best demonstrate the location, size, and effect of the tunnel on the aortic valve. The use of contrast-enhanced CT in three of the cases (Fig 3) was helpful in outlining the tunnel further and clarifying its relationship to the coronary arteries prior to intervention.

The mainstay of treatment is procedural with medical management reserved for the interim period prior to the tunnel being closed. Older reports focus primarily on surgical closure,⁷ with some more recent reports⁸ describing successful surgical closure but the importance of long-term follow-up for aortic valve disease in a five patient data series. However, in more recent times, transcatheter closure has been described as a safe and feasible alternative. Chessa et al. first reported on the transcatheter closure using an Amplatzer patent ductus arteriosus occluder device (AGA Medical Corporation, Golden Valley, Minnesota) in a 14-year-old boy with a good outcome and no follow-up complications.⁹

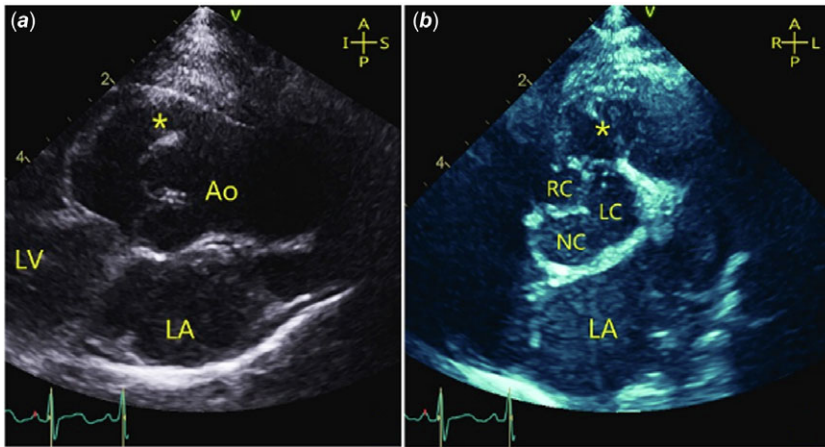


Figure 1. Transthoracic echocardiography with a parasternal long- (left image) and short-axis (right image) views demonstrating the length of the aorto-left ventricular tunnel (ALVT)(*). Ao: aorta, LV: left ventricle, LA: left atrium, RC: right coronary cusp, LC: left coronary cusp, NC: non-coronary cusp.

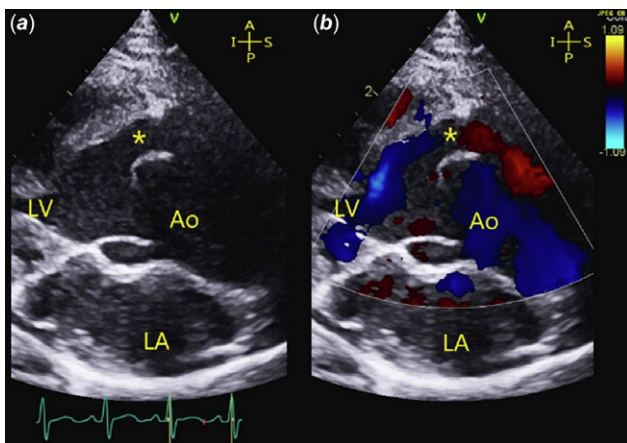


Figure 2. Colour doppler imaging of the aorto-left ventricular tunnel (ALVT)(*) in a parasternal long-axis view. Ao: aorta, LV: left ventricle, LA: left atrium.

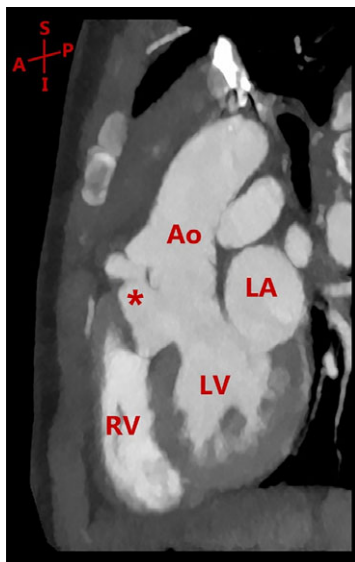


Figure 3. Computed tomography contrast image on aorto-left ventricular tunnel (ALVT) (*). Ao: aorta, LV: left ventricle, LA: left atrium, RV: right ventricle.

A subsequent report has demonstrated the use of transcatheter closure in a child with left ventricular non-compaction cardiomyopathy who would not tolerate surgical closure.⁶

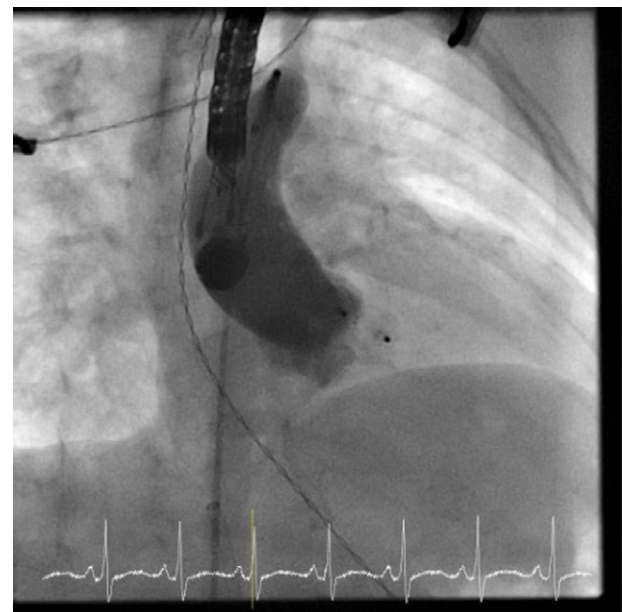


Figure 4. AP angiographic image demonstrating device position and occlusion of the aorto-left ventricular tunnel (ALVT).

In this series, three of the four children underwent a transcatheter approach as first-line treatment strategy (Fig 4). The fourth case had multiple anomalies which required surgical repair. In the three cases, there were no peri-procedure complications and for the two patients who were asymptomatic, they were discharged 24 hours after admission. There were no cases of coronary artery compression or distortion of the aortic valve.

Follow-up of the four patients revealed that patients one and four had mild aortic regurgitation before and after the procedure that has persisted at the most recent follow-up. Patient 2 had dysplastic aortic and pulmonary valves as well as the aorto-left ventricular tunnel and underwent a cardiac transplantation soon after surgical repair. Patient 3 had a complex background with hypoplastic left heart syndrome who had undergone a Norwood Sano operation and subsequently a bidirectional Glenn 2 years prior to the diagnosis. There was no aortic regurgitation before the aorto-left ventricle tunnel diagnosis or after its closure. They have subsequently undergone a cardiac transplantation for right ventricular failure.

Conclusion

Aorto-left ventricular tunnel is a rare anomaly which previously necessitated surgical repair. This case series highlights not only the importance of the clinical examination in its detection but also the usefulness of multi-modality imaging prior to definitive closure. Furthermore, we demonstrate the safety and feasibility of a transcatheter closure approach in appropriately selected patients among this heterogenous patient cohort.

Supplementary material. To view supplementary material for this article, please visit <https://doi.org/10.1017/S1047951123000586>

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Conflicts of interest. None.

Ethical standards. Approval of the above study was obtained from the Ethics Department at CHI Children's Health Ireland, Crumlin, Dublin, Ireland.

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