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Brief Report

Percutaneous device closure of aortico-left ventricular tunnel using Amplatzer vascular plug III

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Abstract Aortico-left ventricular tunnel is a rare congenital heart defect. Reports on successful device closure of these defects are scarce. We are reporting the first case of device closure of aortico-left ventricular tunnel using an Amplatzer vascular plug III.

Keywords: Aortico-left ventricular tunnel; Amplatzer vascular plug III; device

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Aortico-left ventricular tunnel is a rare defect and accounts for 0.001% of all congenital cardiac defects.[1] Martins et al[2] described this defect among 0.05% of the 17,381 patients who underwent cardiac catheterisation over 35 years at the Boston Children's Hospital. It is an abnormal channel that begins in the ascending aorta, bypasses the aortic valve, and leads into the left ventricular cavity. These tunnels bypass the normal ventriculoarterial junction but do not penetrate the septal musculature.[3] Reports on device closure of aortico-left ventricular tunnel are scarce. This is the first report of device closure of aortico-left ventricular tunnel using an Amplatzer vascular plug III.

Case report

A 4.5-year-old boy was referred to the paediatric cardiology clinic for cardiac murmur noted as an incidental finding at the peripheral health centre. This child remained completely asymptomatic since birth. His clinical examination revealed regular high-volume radial pulses at a rate of 110/min, right arm supine blood pressure of 101/56 mmHg, grade 3/6 ejection systolic murmur, and grade 3/4 early diastolic murmur audible best at the aortic areas.

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Transthoracic echocardiogram revealed a sigmoid tract arising superior to the sinotubular junction and traversing antero-lateral to the right coronary cusp assuming an oval shape in the short-axis enface view venting inferiorly into the left ventricular ouflow tract. This aortico-left ventricular tunnel measured about 8.5 mm in length, and the width measured 4.5 mm, 6 mm, and 3.6 mm at the superior ostium at the aortic end, mid-portion, and inferior ostium at the left ventricular end, respectively. There were no associated aortopulmonary anomalies. Coronary arteries and both the ventricular outflow tracts were normal. We could demonstrate pan-diastolic flow reversal in the descending aorta on colour and pulse wave Doppler interrogations. There was mild left ventricular dilatation, with the left ventricular end-diastolic dimension measuring 41 mm (Z-score = +2.29).

This child underwent successful device closure of the aortico-left ventricular tunnel under trans-oesophageal and biplanar fluoroscopic guidance. Immediate pre-procedural trans-oesophageal echocardiogram assessment demonstrated the tunnel to be of 4.5 mm width and about 8 mm length (Fig 1). A minor aneurysmal bulge of the intracardiac portion of the tunnel into the right ventricular outflow tract was also noted with no obstruction. After securing a 5 F sheath in the right femoral artery, an aortic root angiogram using a 4 F pigtail catheter demonstrated the dye filling the left ventricle via the tunnel in diastole. Coronary arteries were normal. We positioned a 014 extra-stiff cross-it 300 coronary wire into the left ventricle via the tunnel

through a 5 F right Judkin's catheter to aid the threading of Tyshak II (B. Braun International Systems Inc., Bethlehem, PA, United States of America), 8 mm/4 cm balloon. This was inflated slowly to conform to the course and shape of the tunnel. The dimensions further measured with the balloon inflated in situ revealed the tunnel length to be 5.3 mm and diameter 4.3 mm. We then exchanged the coronary wire for 035 Amplatzer extra-stiff wire over which a 7 F Mullin's sheath was positioned in the left ventricle through which we advanced 10 mm/5 mm Amplatzer Vascular Plug III (St. Jude Medical, Inc., St. Paul, Minnesota, United States of America). Under trans-oesophageal and fluoroscopic guidance, we initially positioned the device to be seated entirely in the tunnel with inferior lobe optimally placed at the left ventricular end and the waist sitting in the body of the tunnel and the superior lobe closing the aortic end. We repeated aortic and left ventricular angiograms with a 4 F pigtail catheter advanced through a 4 F sheath secured in the left femoral artery, with the device fully deployed and not released. These angiograms and the trans-oesophageal echocardiogram did not reveal any antegrade or retrograde flow through the tunnel. The coronaries' filling was normal and there was no demonstrable aortic regurgitation. These findings were further confirmed on repeating the angiographic study after the release of the device (Fig 2). We administered intravenous bolus dose of unfractionated heparin, 100 units/kg at the beginning of the procedure. The child received aspirin at 5 mg/kg/day starting from 1 day before the procedure for the following 6 months duration.

Electrocardiogram monitoring during the entire procedure did not reveal any ischaemic changes.

We performed a transthoracic echocardiogram recently at 4 months after the device closure. This showed the device positioned well with no residual shunt and no aortic regurgitation (Fig 3).

Discussion

Ever since the first description by Levy et al in 1963,[4] there have been many reports of aortico-left

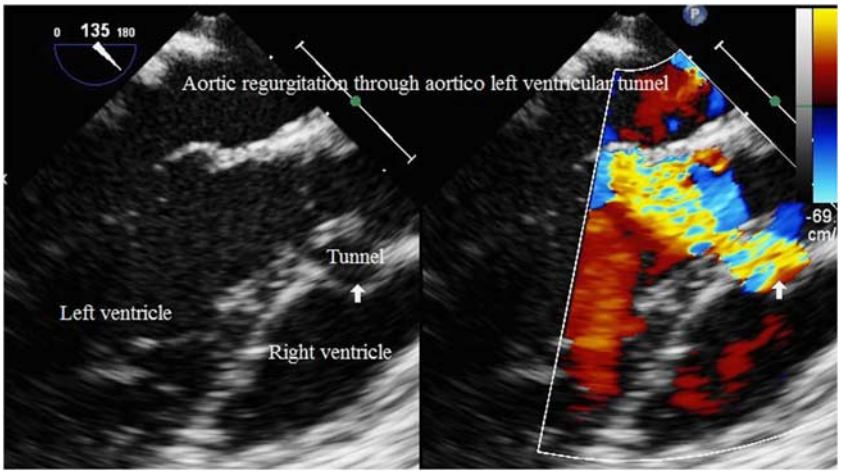

Figure 1.
Mid oesophageal long-axis view (trans-oesophageal echocardiogram) demonstrating aortic regurgitation through the aortico-left ventricular tunnel adjacent to the right coronary cusp

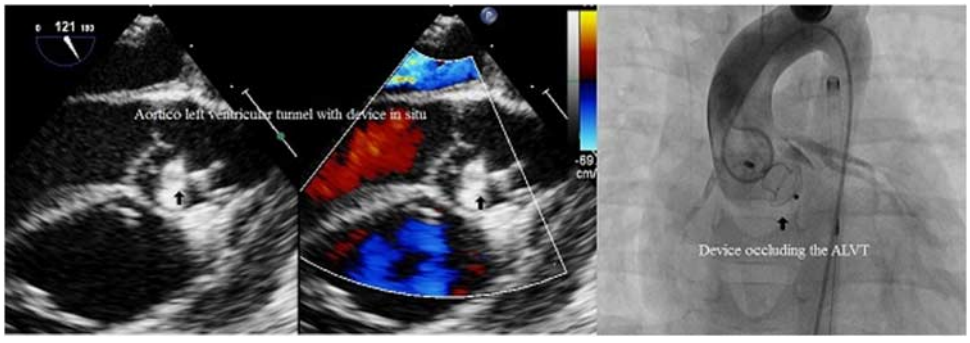

Figure 2.
Mid-oesophageal long-axis view (trans-oesophageal echocardiogram) and aortic root angiogram showing the device well placed in the aortico-left ventricular tunnel (ALVT) with no residual regurgitation and aortic valve integrity well preserved.

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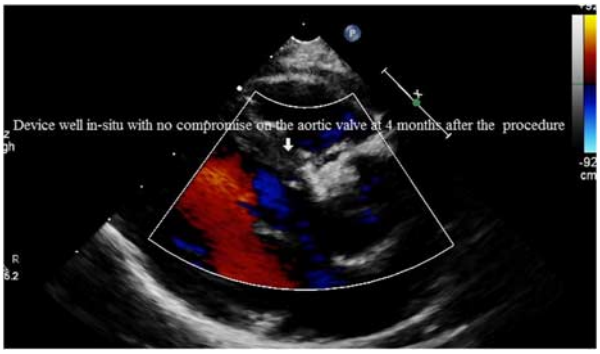

Figure 3.
Follow-up transthoracic echocardiogram after 4 months, revealing the device effectively closing the aortico-left ventricular tunnel with no evidence of aortic valve compromise.

ventricular tunnel focussing mainly on its surgical management. The exact aetiology and embryology of aortico-left ventricular tunnel still remain largely speculative. Suggested explanations include an anomalous coronary artery, possibly the conal vessel, opening in the left ventricle,[4] rupture of a sinus of Valsalva aneurysm,[5] an anterior aortic wall abnormality with communication into the left ventricle,[3] defective incorporation of the distal end of the bulbus cordis,[6] persistence of embryonic crests of the fifth aortic arch,[7] and early aortic dissection similar to Marfan's syndrome.[8] Hovaguimian et al in 1988 attempted to describe four types of aortico-left ventricular tunnel with their attendant implications on the surgical approach: type 1, a simple tunnel with a slit-like opening at the aortic end and no aortic valve distortion; type II, a large extracardiac aortic wall aneurysm of the tunnel with an oval opening at the aortic end with or without valvular distortion; type III, intracardiac aneurysm of the septal portion of the tunnel, with or without right ventricular outflow tract obstruction; and type IV, types II and III combined.[9] In our patient, the tunnel had an oval opening at the aortic end with no aortic wall aneurysm or any aortic valve distortion. There is in addition a minor aneurysmal bulge of the intracardiac septal portion of the tunnel, thus categorising this into type III.

Generally, aortico-left ventricular tunnel presents in childhood or infancy, and the natural course depends on the degree of aortic regurgitation. Recognition of aortic incompetence in a neonate or young infant should suggest aortico-left ventricular tunnel as a possible diagnosis. Other lesions that must be differentiated are ruptured aneurysm of the sinus of Valsalva, coronary cameral fistula, congenital isolated aortic incompetence, and aortic incompetence with ventricular septal defect.

Transthoracic echocardiography can identify the aortico-left ventricular tunnel and associated lesions. Associated cardiac anomalies are present in about 45% of these patients.[2] Coronary artery anomalies are commonly encountered in the form of complete absence of right or left coronary arteries, ostium of right coronary artery lying within the tunnel and right coronary artery to distal tunnel fistula. Critical aortic stenosis, bicommissural aortic valve, unicommissural aortic valve, sub-aortic stenosis, aortic incompetence, leaflet perforation of the aortic valve from haemodynamic trauma of the unsupported right coronary cusp, pulmonary valvar stenosis, right ventricular outflow tract obstruction and left ventricular non-compaction, ventricular septal defect, atrial septal defect, and patent ductus arteriosus are among the other reported associations.[10–12] Our patient did not have any other significant associated cardiac lesion.

Catheterisation should be reserved for patients with unclear non-invasive findings or for transcatheter closure. Other imaging modalities like transoesophageal echocardiography, foetal echocardiography, and magnetic resonance imaging are also helpful in defining the details in various clinical settings.[13,14]

There are very few reports of successful percutaneous closure of aortico-left ventricular tunnel.[15,16] The varied nature of the anatomy, close proximity of the tunnel to the coronary arteries, the necessity to improvise devices that are designed for other defects, and the potential deformation complications of the aortic valve annulus resulting in aortic regurgitation can all dissuade the interventionist. Transcatheter closure of the aortico-left ventricular tunnel may be attempted in all the four types when there is no associated lesion amenable only to surgical repair and when the device itself is not going to compromise the coronary arteries. Significant right ventricular and left ventricular outflow obstructions, more than mild aortic valve regurgitation, other aortic and pulmonary valve abnormalities and coronary artery abnormalities requiring surgery should be considered as contraindications for device closure.

Almost all the reports of device closures in the literature used Amplatzer duct occluder devices. These nitinol-based devices owing to their flexible nature conform to the shape of the aortico-left ventricular tunnel plugging the defect effectively. However, their retention skirt protruding into the left ventricular outflow tract can exert shearing stress on the red blood cells leading to haemolytic anaemia and hyperbilirubinaemia.[16] In this context, Amplatzer vascular plugs, which come in more suitable lengths and diameters, prove to be the better option. We used Amplatzer vascular plug III in our patient. The Amplatzer family of Vascular Plugs have four different types with a flexibility to select the correct vascular plug for the procedure

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based on vessel type, blood flow, and the available landing zone. The Amplatzer Vascular Plug III, with oblong cross-sectional shape, multiple nitinol mesh layers, and extended rims, provides the fastest occlusion of all Amplatzer Vascular Plugs and is ideal for high-flow situations. There is a small platinum marker on the distal rim centred in the long axis that further aids to improve visualisation for device orientation. In addition, 155 cm-long delivery wire accommodates a greater range of procedures for easy device delivery.

We feel that percutaneous device closure of aortico-left ventricular tunnel has the inherent advantage of plugging both ends of the tunnel, obviating the potential aneurysm formation noted in the earlier surgical methods of closing only the aortic end. It is also unlikely for aortic regurgitation to occur in future if no such problem is evident during the procedure itself. This presumption is nevertheless unsubstantiated as there are no long-term follow-up reports in these patients with device closure. On the contrary, regardless of the surgical technique used, valvar aortic regurgitation is a major concern in these patients, and despite successful surgical repair the incidence of aortic regurgitation ranges from 16% to 60%.[10,17]

References

1. Okoroma EO, Perry LW, Scott LP, McClenathan JE. Aortico-left ventricular tunnel: clinical profile, diagnostic features and surgical considerations. J Thorac Cardiovasc Surg 1976; 71: 238–244.
2. Martins JD, Sherwood MC, Mayer JE, Keane JF. Aortico–left ventricular tunnel: 35-year experience. J Am Coll Cardiol 2004; 44: 446–450.
3. Ho SY, Muriago M, Cook AC, Thiene G, Anderson RH. Surgical anatomy of aorto–left ventricular tunnel. Ann Thorac Surg 1998; 65: 509–514.
4. Levy MJ, Lillehei CW, Anderson RC, Amplatz K, Edwards JE. Aortico-left ventricular tunnel. Circulation 1963; 27: 841–853.
5. Turley K, Silverman NH, Teitel D, Mavroudis C, Snider R, Rudolph A. Repair of aortico-left ventricular tunnel in the neonate: surgical, anatomic and echocardiographic considerations. Circulation 1982; 5: 1015–1020.
6. Somerville J, English T, Ross DN. Aorto-left ventricular tunnel: clinical features and surgical management. Br Heart J 1974; 36: 321–328.
7. Palacio J, Peralta A, Sanchez B, Alperovich M. Intrapericardial congenital supravalvular aortic aneurysm communicating with the outflow tract of the left ventricle. J Cardiovasc Surg 1964; 5: 401–448.
8. Cooley RN, Harris LC, Rodin AE. Abnormal communication between the aorta and left ventricle: aortico-left ventricular tunnel. Circulation 1965; 31: 564–571.
9. Hovaguimian H, Cobanoglu A, Starr A. Aortic-left ventricular tunnel: a clinical review and new surgical classification. Ann Thorac Surg 1988; 45: 10–121.
10. Horvath P, Balaji S, Skovranek S, Hucin B, de Leval MR, Stark J. Surgical treatment of aortico-left ventricular tunnel. Eur J Cardiothorac Surg 1991; 5: 113–117.
11. Chehab G, Hayek J, Saliba Z, El-Rassi I. Images in cardiovascular medicine: angiography of an aneurysmal aorto-left ventricular tunnel. Circulation 2007; 116: 356–358.
12. McKay R. Aorto-ventricular tunnel. Orphanet J Rare Dis 2007; 2: 41.
13. Singh A, Reinhardt Z, Desai T, Rasiah SV. Case series of antenatally diagnosed aortico-left ventricular tunnel. Pediatr Cardiol 2010; 32: 822–825.
14. Kakadekar AP, Sandor GG, Patterson MW, LeBlanc JG. Role of transesophageal echocardiography in the management of aortic-left ventricular tunnel. Pediatr Cardiol 1995; 16: 137–140.
15. Chessa M, Chaudhari M, De Giovanni JV. Aorto-left ventricular tunnel: transcatheter closure using an Amplatzer duct occluder device. Am J Cardiol 2000; 86: 53–54.
16. Vijayalakshmi IB, Chitra N, Prabhu Deva AN. Use of an Amplatzer duct occluder for closing an aortico-left ventricular tunnel in a case of noncompaction of the left ventricle. Pediatr Cardiol 2004; 25: 77–79.
17. Sreeram N, Franks R, Arnord R, Walsh K. Aorticoventricular tunnel: long-term outcome after surgical repair. J Am Coll Cardiol 1991; 17: 950–955.