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Brief Report

Transcatheter closure of a residual aorto-left ventricular tunnel: report of a case with a 6-year follow-up

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Abstract Aorto-left ventricular tunnel is an exceedingly rare congenital cardiac defect. Early surgical closure is the treatment of choice. Residual or recurrent tunnel and aortic valve insufficiency are well-recognised complications after surgical repair. In this article, we report on successful transcatheter closure of a residual aorto-left ventricular tunnel using an Amplatzer duct occluder in a 7-year-old boy. The outcome after 6 years of follow-up is encouraging.

Keywords: Aorto-left ventricular tunnel; aortic regurgitation; cardiac catheterisation; Amplatzer duct occluder; child

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Aorto-left ventricular tunnel is an extremely rare congenital communication between the ascending aorta and the left ventricular cavity, bypassing the aortic valve.[1,2] It allows retrograde diastolic blood flow from the aorta to the left ventricle, eventually leading to congestive heart failure. Therefore, early surgical closure of the tunnel is currently the treatment of choice;[2–4] however, residual or recurrent tunnel and valvular aortic regurgitation are fairly common complications after surgical repair.[2,3]

Case report

A 7-year-old boy was referred to us for management of a residual aorto-left ventricular tunnel and aortic valve insufficiency. He had previously undergone surgical repair for aorto-left ventricular tunnel at the age of 2 months. At follow-up, the residual flow through the tunnel and aortic valve regurgitation were noticed. There were also signs of progressive left ventricular volume overload. The child was free of symptoms.

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On physical examination, peripheral pulses were full and symmetrical, and his blood pressure was 95/60 mmHg. There was a grade 2 diastolic murmur, which was best heard in the second interspace at the right sternal border. Electrocardiographic findings were unremarkable. Transthoracic echocardiography revealed a residual aorto-left ventricular tunnel arising above the right sinus of Valsalva and terminating in the left ventricle just below the aortic valve. The aortic annulus and the ascending aorta measured 21 mm (z score +3.25) and 24 mm (z score +3.35) in diameter, respectively. The aortic valvar leaflets appeared mildly dysplastic. There were two jets of aortic regurgitation, the larger one emerging from the residual tunnel (color Doppler jet width of 3–4 mm) and the smaller central one emerging through the valve. The left ventricle was slightly dilated with preserved systolic function. There were no anomalies in the origin of the coronary arteries.

Given the sufficient distance between the aortic opening of the tunnel and both the aortic valve leaflets and the right coronary artery, we elected to perform transcatheter closure of the residual tunnel.

Intravenous heparin (100 IU/kg) and ceftriaxone (50 mg/kg) were administered before the procedure.

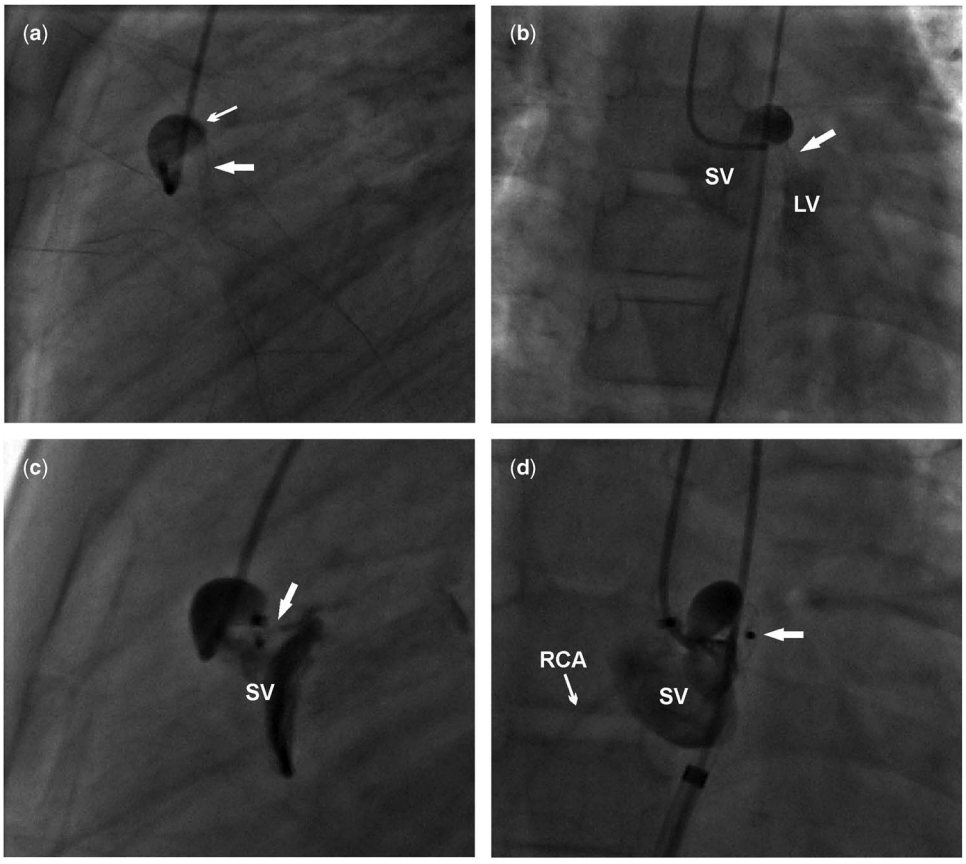

Figure 1.
*(**a**) A selective aortogram in the lateral projection reveals the aorto-left ventricular tunnel (large arrow) with an aneurysmal aortic end (small arrow). (**b**) A selective aortogram in antero-posterior projection shows the aorto-left ventricular tunnel (arrow). (**c**) A selective aortogram in the lateral projection after transcatheter closure of the tunnel with an Amplatzer duct occluder shows a trace residual shunt through the occluder (arrow). (**d**) A selective aortogram in the antero-posterior projection demonstrating complete closure of the tunnel with the Amplatzer duct occluder (large arrow). The right coronary artery flow is unobstructed (small arrow). SV = right sinus of Valsalva; LV = left ventricle; RCA = right coronary artery.*

Left-sided cardiac catheterisation was undertaken after accessing the right femoral artery. Angiography in several projections confirmed backflow of blood into the left ventricle through the residual tunnel and the aortic valve. Selective angiography showed a windsock appearance of the residual aorto-left ventricular tunnel – that is, type II according to the classification by Hovaguimian et al[4] (Figs 1a and b). It originated >4 mm distal to the right coronary ostium. Considering the morphology of the tunnel and its minimum diameter of 3 mm, it was decided to attempt closure using a 6/4-mm Amplatzer duct occluder (St. Jude Medical, Inc., St. Paul, Minnesota, United States of America). A long sheath was passed across the tunnel, and the device was successfully deployed under fluoroscopic and transthoracic echocardiographic guidance (Figs 1c and d). Repeat aortography demonstrated good position of the device completely occluding the tunnel with no impingement on the right coronary artery (Supplementary video 1). The degree of aortic valve regurgitation remained unchanged. There were no electrocardiographic features of myocardial ischaemia during or after the procedure. Following the intervention, the patient was started on continuous heparin infusion at a dose of 20 IU/kg/hour for the next 48 hours. He was then discharged home on aspirin therapy 75 mg once daily for 6 months.

The patient has been doing well during the 6-year follow-up period. His left ventricular size has returned to normal as demonstrated by serial echocardiograms. There has been no increase in aortic regurgitation or the presence of residual flow through the tunnel (Fig 2). Aortic annulus and ascending aortic diameter at the latest follow-up were 25 (z score +2.80) and 29 mm (z score +3.02), respectively. There have been no signs of device-related aortic valve distortion or injury to the right coronary artery as well. Repeat 24-hour Holter monitoring revealed no arrhythmias.

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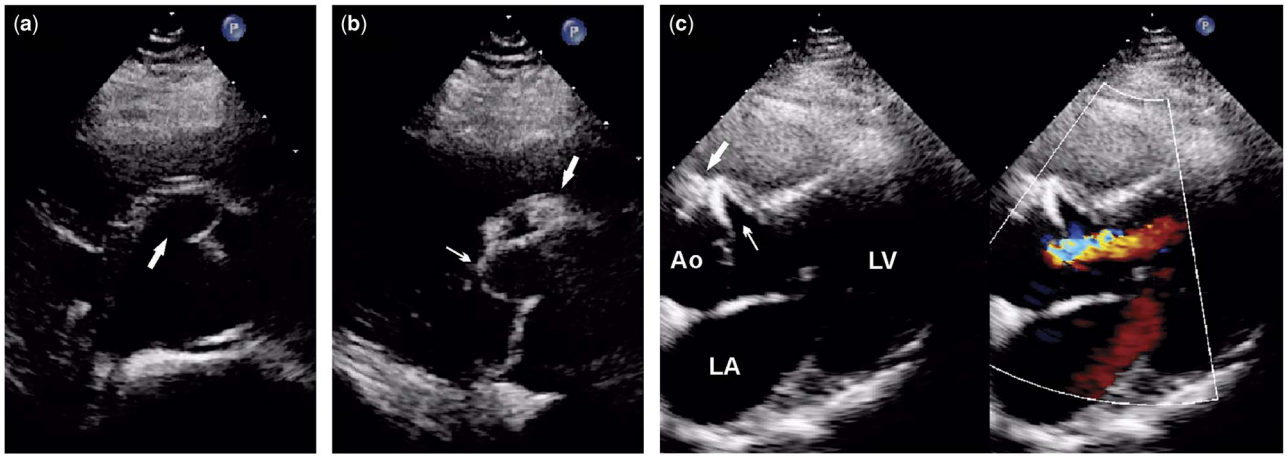

Figure 2.
*(**a**) The echocardiographic image obtained from a modified, parasternal long-axis view in systole shows the left ventricular end of the aorto-left ventricular tunnel (arrow). (**b**) This image taken from a modified, parasternal long-axis view in diastole shows the device occluding the tunnel (large arrow). The small arrow points to the unsupported right coronary cusp. (**c**). This is the apical three-chamber view. The left panel demonstrates the device placed into the tunnel (large arrow) and the left ventricular opening of the tunnel (small arrow). The right panel shows a central jet of aortic regurgitation. There is no residual flow through the tunnel. Ao = aorta; LA = left atrium; LV = left ventricle.*

Discussion

Haemodynamically significant aorto-left ventricular tunnel should be managed soon after establishing the diagnosis to prevent the development of heart failure, progressive aortic root and ascending aortic dilatation, and damage to the aortic valve leaflets.[2,4] Surgery is the preferred treatment modality, especially in the presence of associated lesions such as significant aortic valve abnormalities and anomalies of the coronary arteries.[3,5] The results of surgical repair are good, although postoperative complications including residual or recurrent aorto-left ventricular tunnel and aortic valve insufficiency can occur.[2,3] Transcatheter closure of the aorto-left ventricular tunnel is another possibility, but there is little experience with this approach.[3,5–10] Furthermore, current information on the management and outcome of postoperative residual or recurrent aorto-left ventricular tunnel is scarce. Treatment options for dealing with residual or recurrent tunnels comprise surgical and transcatheter repair.[3,8,9] To our knowledge, however, there are only two descriptions of successful transcatheter closure of postoperative residual or recurrent aorto-left ventricular tunnel.[8,9]

Although the transcatheter approach seems more appealing, several factors limit its use, including close relation of the tunnel to the coronary arteries, the risk of aortic valve damage or distortion by the device and consequent aortic regurgitation, variable tunnel morphology, distensibility of the tunnel, and the presence of coexisting anomalies.[3,5,6,9,10] The residual shunting through the occluder can sometimes cause significant haemolysis[5,9]. There is also a lack of devices specifically designed for this indication.[5,7] The choice of the appropriate device should be decided on an individual basis, depending on the underlying anatomy of the aorto-left ventricular tunnel and its relationship to the surrounding structures. Finally, data are lacking with respect to long-term results after transcatheter closure.

In conclusion, transcatheter closure of postoperative residual aorto-left ventricular tunnel can be a safe and effective alternative to re-operation in carefully selected cases. Successful transcatheter occlusion of residual aorto-left ventricular tunnel seems to slow the aortic disease process and at least delays the need for re-operation, as demonstrated by the results of a 6-year follow-up in our patient; nevertheless, close follow-up for the presence of aortic valve impairment, progressive ortic root and ascending aortic dilatation, and left ventricular dysfunction is warranted.

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Conflicts of Interest

None.

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Ethical Standards

The authors assert that all procedures contributing to this study comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the Ethics Committee of the University Children's Hospital in Belgrade, Serbia.

Supplementary material

To view supplementary material for this article, please visit https:/doi.org/10.1017/S1047951117000701

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