Cardiol Young 2004; 14: 203–205

ISSN 1047-9511

Brief Report

An unusual case of aorto-left ventricular tunnel

Vladimiro L. Vida, Tomaso Bottio, Giovanni Stellin

Department of Cardiovascular Surgery, Paediatric Cardiac Unit, University of Padova Medical School, Padova, Italy

Abstract An asymptomatic 4-year-old child was referred to our Institution with a provisional diagnosis of severe aortic valvar regurgitation in association with a ventricular septal defect. Intraoperatively, the aortic valve appeared bicuspid, and a tunnel of 7 mm diameter extending between the aorta and the left ventricle was found dividing the anterior commissure into two hemi-commissures. Repair was achieved by reconstructing the anterior commissure by direct reapproximation, and simultaneous obliteration of the aortic side of the tunnel. The aortic valvar leaflet was resuspended onto the arterial wall. This combined approach will hopefully reinforce the poorly supported aortic sinus, and may maintain aortic valvar competence over the long term. Our intraoperative diagnosis of aorto-left ventricular tunnel suggests consideration of this diagnosis when evaluating any patient with suspected aortic valvar incompetence.

Keywords: Aorto-left ventricular tunnel; bicuspid aortic valve; misdiagnosis

THE AORTO-LEFT VENTRICULAR TUNNEL IS A RARE congenital malformation, taking the form of an abnormal channel that begins in the left ventricle, bypasses the aortic valve, and terminates in the ascending aorta. Due to the rarity of this cardiac malformation, cases of misdiagnosis often occur. We describe an unusual case of aorto-left ventricular tunnel in a patient with a bicuspid aortic valve.

Case report

In May 2003, an asymptomatic 4-year-old child was referred to our Institution with provisional diagnosis of severe aortic valvar regurgitation in association with a ventricular septal defect. A systolic and diastolic murmur was heard at the left upper sternal border. Chest X-rays revealed cardiomegaly, the cardiothoracic index being 0.57. Echocardiography, with cross sectional imaging showed a moderate dilated left ventricle with good systolic function, the ejection fraction being measured at 84%. The preoperative echocardiographic diagnosis was a large malalignment ventricular septal defect with a fibrous aneurysmal component on the right ventricular side (Fig. 1). There was presumed aortic valvar incompetence due the presence of a distorted leaflet tetherd within the ventricular septal defect. The child was scheduled for

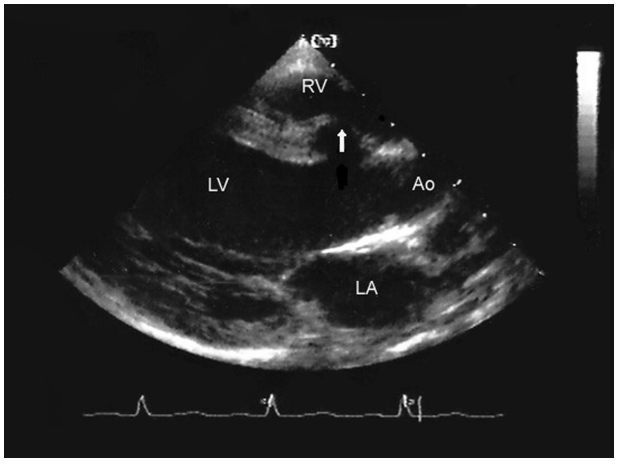

Figure 1.
Cross sectional echocardiographic imaging in the parasternal long-axis view showing prolapse of the anterior sinus into the right ventricle outflow tract (arrow), mimicking an aneurysm of the membranous septum. LV: left ventricle; LA: left atrium; RV: right ventricle; Ao: aorta.

Correspondence to: Vladimiro Vida MD, Department of Cardiovascular Surgery, Paediatric Cardiac Unit, University of Padova Medical School, via Giustiniani 2, 35128 Padova, Italy. Tel: +39 049 821 2410; Fax: +39 049 821 2409; E-mail: vladimirovida@interfree.it

Accepted for publication 6 October 2003

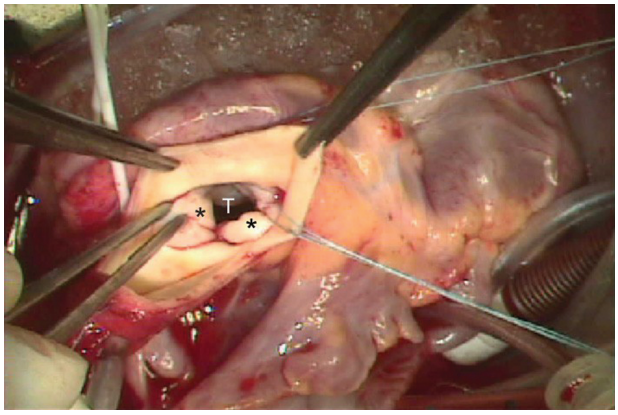

Figure 2.
Intraoperative view showing the aorto-left ventricular tunnel bypassing the anterior commissure of the bicuspid aortic valve. Note the dysplastic aortic leaflets (*), and the tunnel (T) dividing the anterior commissure in two hemi-commissures.

surgery with the plan to attempt reconstruction of the aortic valve and closure of the ventricular septal defect.

With the aid of moderate hypothermic cardiopulmonary bypass at 28°C, cross-clamping of the aorta and cold blood retrograde cardioplegic arrest, an oblique aortotomy was performed above the sinutubular junction. The aortic valve appeared bicuspid with thickened leaflets. An aorto-left ventricular tunnel of 7 mm diameter was found dividing the anterior commissure into two hemi-commissures (Fig. 2). No ventricular septal defect was detected in the region of the outflow tract. The orifice of the right coronary artery was very close to the tunnel at the level of the anterior leaflet.

Repair was achieved by reconstructing the anterior commissure by direct reapproximation of the two hemi-commissures, and simultaneous obliteration of the aortic side of the tunnel with interrupted mattress sutures. The reconstructed commissure was than resuspended onto the aortic wall to avoid prolapse of the leaflet. Intraoperative transoesophageal echocardiography showed only minimal residual aortic regurgitation through the centre of the valve. The child was easily weaned from cardiopulmonary by-pass in sinus rhythm and with stable haemodynamics. The postoperative course was uneventful.

Discussion

Aorto-left ventricular tunnel can be considered a defect of the aortic root affecting the interleaflet triangle, being found most often between the left and right coronary aortic valvar leaflets.[1] The essence of the lesion is the divorce of the attachment of the right or left coronary aortic leaflet from its normal location within the aortic root.[2] This removes the support for either the right or left coronary aortic leaflet, and invariably results in progressive aortic regurgitation, often necessitating valvar repair or replacement.

The tunnel appears as an extracardiac tubular communication between the aorta and the left ventricle, bypassing the aortic valvar apparatus, and producing massive regurgitation through the tunnel.[3] The tunnel courses between the posteriorly situated aortic sinuses and the anteriorly positioned subpulmonary infundibulum. This last part can bulge into the right ventricular outflow tract, causing the so-called septal aneurysm that, in our case, mimicked the ventricular septal defect with a presumed fibrous aneurysmal component on its right ventricular side.[1] Diagnostic echocardiographic criterions include an enlargement and hypertrophy of the left ventricle, an enlargement of the aortic root, and free regurgitation at the level of the aortic valve.[4] The most common diagnostic errors from echocardiography have been in confusing the ventricular end of the tunnel with a ventricular septal defect, while flow of blood through the tunnel has also been misinterpreted as valvar aortic regurgitation.[2] Resonance imaging and cardiac catheterization are often necessary to establish the correct diagnosis, and to characterize abnormal patterns of flow in the aortic root.[2] In our patient the presence of a septal aneurysm was judged to be the cause of distortion of the leaflets leading to aortic regurgitation.

The location of the aorto-left ventricular tunnel within the anterior commissure of a bicuspid, thickened and dysplastic aortic valve, dividing the anterior aortic valvar commissure into two hemi-commissures, mimicked echocardiographically aortic valvar incompetence. Surgery was accomplished with direct suture of the arterial side, with care to avoid distortion of both the aortic valve and the orifice of the right coronary artery. Closure of the tunnel, together with resuspension of the anterior aortic commissure, successfully eliminated any the aortic regurgitation. As reported by others,[5] the localized dilation of the aortic root at the site of the tunnel is a weakness that remains after the tunnel is closed, leaving a poorly supported aortic valve and a weak root. We believe, in our case, that resuspension of the aortic anterior commissure in association with closure of the tunnel may have reinforced the poorly supported aortic sinus and may maintain aortic competence over the long term.

Although rare, the aorto-left ventricular tunnel is the foremost cause during infancy of regurgitant flow of blood from the aorta to one or the other of the ventricles.[2]

The intraoperative diagnosis of this rare congenital cardiac malformation advises us to rule out the presence of aorto-left ventricular tunnel in evaluating any patient with "suspected" aortic valvar incompetence.

https://doi.org/10.1017/S1047951104002161 Published online by Cambridge University Press

References

1. Ho SY, Muriago M, Cook AC, Thiene G, Anderson RH. Surgical anatomy of aorto-left ventricular tunnel. Ann Thorac Surg 1995; 65: 509–514.
2. McKay R, Anderson RH, Cook AC. The aorto-left ventricular tunnels. Cardiol Young 2002; 12: 563–580.
3. Horvath P, Balaji S, Skovranek S, Hucin B, de Leval MR, Stark J. Surgical treatment of aortico-left ventricular tunnel. Eur J Cardiothoracic Surg 1991; 5: 443–444.
4. Cook AC, Fagg NL, Ho SY, Groves AM, Sharland GK, Anderson RH, Allan LD. Echocardiographic anatomical correlations in aorto-left ventricular tunnel. Br Heart J 1995; 74: 443–448.
5. Serino W, Andrade JL, Ross D, de Leval M, Somerville J. Aorto-left ventricular communication after closure: late postoperative problems. Br Heart J 1983; 5: 501–506.