Management of aortic left ventricular tunnel

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SUMMARY A 22 year old man had a diagnosis of left ventricular aortic tunnel established during infancy. Surgical repair was deferred at that time because his severe aortic root deformity would also have required aortic valve replacement. His current lack of symptoms together with a normal exercise capacity could be used as an argument against routine early surgical repair of the tunnel, particularly when the possible need for additional aortic valve replacement is considered.

Aortic left ventricular tunnel is a rare congenital cardiac malformation. The physiology of this cardiac defect is similar to that of aortic valvar regurgitation. Surgical repair of the tunnel, even in the absence of heart failure, is the usual management.¹⁻³ Gross aortic valve deformity is, however, usually already present in the neonatal period, most probably as a result of the turbulence through the tunnel in utero. Despite successful surgical closure of these tunnels most patients are left with haemodynamically important aortic regurgitation and will need aortic valve replacement in the future.

We report a patient with severe left ventricular volume overload who was without symptoms. Structural change in architecture of the left ventricular muscle may have developed in utero in response to the early expansion of left ventricular volume. The continued stability of the patient's cardiac function and lack of symptoms are an argument against early surgical repair.

Case report

NEONATAL HISTORY

A 22 year old man was referred to our hospital at 6 weeks' old because of a cardiac murmur. Labour and delivery had been normal at 36 weeks' gestation (birthweight 2.85 kg). He was a healthy looking baby, without cyanosis or signs of heart failure. All peripheral pulses were present and collapsing in character. A pronounced apical and left parasternal heave was present. Auscultation detected a grade 3/4 ejection systolic murmur and a 3/4 decrescendo

diastolic murmur over the entire precordium.

A radiograph of the chest showed pronounced cardiomegaly with a grossly dilated aortic root and ascending aorta. The lung fields were normal. A clinical diagnosis of aortic regurgitation was made, and this was subsequently changed to aortic left ventricular tunnel when this was first described shortly afterwards.⁴ He was without symptoms, and subsequent growth and development were normal. The clinical diagnosis was confirmed by cardiac catheterisation at the age of 4 years. Measurements of right heart pressures were normal. Ascending aortic diastolic pressure was 40 mm Hg, and left ventricular end diastolic pressure 13 mm Hg. Right heart oximetry excluded a left to right shunt. Aortography showed gross dilatation of the aortic root, with pronounced expansion of the right and non-coronary sinuses. The tunnel was evident and arose at the level of the right coronary sinus. Immediate opacification of the left ventricle through both the tunnel and the aortic valve occurred. The aortic cusps seemed to be grossly deformed. Since surgical repair would probably have also entailed aortic valve replacement and especially as the patient was symptomless operation was deferred. Throughout childhood he remained completely symptomless and as an adult led a normal active life. At school he played competitive sport including soccer, without developing symptoms. At yearly review the results of his physical examinations were unchanged. Blood pressure remained at about 140/20 mm Hg and systolic ejection and early diastolic murmurs were repeatedly heard all over the precordium. Electrocardiograms always showed voltage criteria of left ventricular hypertrophy and no ST-T abnormality. By chest radiography the cardiothoracic ratio was unchanged

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Fig. 1 M mode echocardiogram showing the left ventricular aortic tunnel (T) in between the aortic root (AR) and the right ventricular outflow tract (RVOT). La, left atrium: PCG, phonocardiogram; ECG, electrocardiogram.

(59%), and was proportionately identical with that on the radiographs during infancy.

Serial M mode echocardiograms were repeated after the age of 6 years. The left ventricle was hypertrophied symmetrically, its volume had increased, and fractional shortening was 45–55% when corrected for body surface area. The anterior leaflet of the mitral valve had always shown a decreased excursion and fluttering throughout diastole. In all the serial studies a chamber was seen lying between the anterior wall of the aortic root and the right ventricular outflow tract (Fig. 1), below and at the level of the aortic annulus. This space had become continuous with the aortic root above the annulus.

Cross sectional echocardiography clearly showed the tunnel, which was best visualised in the parasternal long axis view (Figs. 2(a) and 2(b)). The tunnel entered the left ventricle at the upper part of the septum beneath the aortic root and was continuous with the right aortic sinus of Valsalva. The leaflets were considerably thickened and deformed. Symmetrical hypertrophy and increased left ventricular volume was seen by M mode echocardiography. From the aortic short axis (Figs 2(c) and 2(d)) three valve cusps could be seen; they were all severely

Fig. 2 (a) parasternal long axis recording of the left ventricle (LV) and (b) parasternal short axis recording of the aortic root with accompanying diagrams showing the appearance of the left ventricular aortic tunnel (T). Note the thickened tricuspid aortic valve (b). RV, right ventricle; AO, aortic root; LA, left atrium; RA, right atrium.

thickened and deformed. A rudimentary left coronary cusp and a large ballooning right coronary cusp were seen. Scans from the aortic root showed the tunnel extending from the aortic root at the level of the right coronary sinus of Valsalva to the left ventricle. The tricuspid and pulmonary valves looked normal, and the left atrium and right sided heart chambers were of normal size.

Discussion

Since Levy *et al* first described aortic left ventricular tunnel in 1963⁴ 33 cases have been reported. The aortic left ventricular tunnel is a vascular channel of congenital origin that connects the aortic root in the region of the right coronary sinus of Valsalva (above the aortic valve) with the left ventricle immediately below the aortic valve annulus. The aortic valve is often deformed,^{1 3-6} and other cardiac abnormalities may be associated.^{4 7-11} These findings and the subsequent hypertrophy of the left ventricle have been found at operation or necropsy^{12 13} from the neonatal period onwards.¹¹

Although there is evidence to support the congenital nature of left ventricular aortic tunnel, the embryology of the tunnel is controversial. Cooley *et al* postulated Marfan's syndrome based on the aneurysmal dilatation of the sinus of Valsalva with an excess deposition of mucopolysaccharide substances in the elastic media of the aortic wall and the adjacent portion of the tunnel.⁸ It was proposed that this could lead to cystic areas, which in turn could rupture into the left ventricle. These views, however, have not found support. Levy *et al* showed continuation of the elastic fibres of the aortic media into the tunnel wall and suggested that the tunnel was an anomalous coronary artery that communicated through an enlarged myocardial sinusoid with the left ventricle (coronary-cameral fistula).⁴ The tunnel wall in the ventricular septum represented histologically a sinusoid consisting mainly of connective tissue. Roberts and Morrow, however, concluded that the structure of the tunnel differed from that of the blood vessel.⁶

The diagnosis should be suspected in neonates presenting with signs of aortic regurgitation. The differential diagnosis is between absent aortic valve, endocarditis with destruction of the aortic valve, and tetralogy of Fallot with absent pulmonary valve.⁷ Rarely an aneurysm of the sinus of Valsalva may dissect into the left ventricle through the ventricular septum.¹⁴

Echocardiography is complementary to invasive studies, especially in detecting other associated cardiac abnormalities such as aortic and pulmonary stenosis³ or, less commonly, compression of the left

coronary artery by a deformed aortic valve.⁹ Cross sectional echocardiograms should detect progressive compression of the right ventricular outflow tract by the tunnel.¹⁰

Various method of surgical repair have been reported—for example, direct closure and external plication. Aortic regurgitation often persists post-operatively. Spooner *et al* reported a "triple repair" that was used successfully in one case.¹⁵ At the time of publication aortic regurgitation was not present. Similarly, in three cases where patch closure was used aortic regurgitation was avoided.^{7 10 16}

The risk of surgical repair was 10%, and even if the surgical repair is performed in the neonatal period or early infancy aortic valve deformities are already present.^{3 11 15 17 18} These and aortic root abnormalities can be explained by turbulent flow through the tunnel, beginning in utero.

The natural history of the left ventricular aortic tunnel is not known, and the oldest patient reported was a 25 year old man who was symptomless until he died of bacterial endocarditis.¹³

Three patients who had surgical repair and were symptomless were left with aortic regurgitation.¹⁻³ Two other patients with symptoms died without surgical intervention, but they had other major cardiac defects.^{6 19} In our patient the need for aortic valve replacement at an early age because of gross aortic valve deformity was the main reason for deferring surgery during childhood.

A valve defect occurring in utero may be better tolerated than a similar one occurring after birth. The earliest recognisable embryonic cardiac system is a straight heart tube. The visible contraction of this tube is similar to peristaltic movements propelling blood unidirectionally.²⁰ Growth of this primitive heart results in the formation of a cardiac loop and later the cardiac septa. Throughout this development the basic direction of the peristaltic wave is well preserved, and there are recognisable features of this in the adult heart.²¹ The formation of the cardiac valves is not complete until the ninth week, by which time the four chambers of the heart are fairly well formed. The primitive fetal heart is therefore adapted to pumping without the help of heart valves. Persistent incompetence of the valves beyond the ninth week would tend to exert developmental pressure on the fetal heart to adapt and to retain some of the peristaltic form of contraction and hence to preserve pumping efficacy into adulthood. This might lead to modification of the myocardial fibre direction and of the activation and deactivation sequence. At the molecular level there may be functional adaptation in the fetal contractile units, such as in terms of selection of the myosin types^{22 23} and hyperplasia of the myocardial cells.

An expectant attitude to the treatment of aortic left ventricular tunnel may be justified on account of favourable adaptation of the developing heart to this anomaly in utero. These considerations may explain the favourable prognosis of such patients compared with the less good outlook for most patients with severe aortic regurgitation acquired in childhood or adult life.

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