Aorto-left ventricular tunnel

Clinical features and surgical management

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Three patients aged 5 to 7 years with aorto-left ventricular tunnel had successful surgical closure, and a fourth patient aged 16 years with bad postoperative aortic regurgitation is described. The diagnosis should be considered in a child presenting with the features suggestive of severe aortic regurgitation. Early operation is recommended before the aortic annulus becomes too dilated.

Aorto-left ventricular tunnel is an uncommon condition which presents as apparent gross aortic regurgitation in childhood (Levy *et al.*, 1963). Severe aortic regurgitation is well tolerated in the young, and with the current uncertainty about the long-term function of any valve replacement one is reluctant to replace the aortic valve prematurely in a child or adolescent. However, the aorto-left ventricular tunnel may be 'cured' without replacement of the aortic valve providing surgery is performed before the valve cusps and annulus have become distorted (Roberts and Morrow, 1965). It is, therefore, important to distinguish patients with a tunnel from those with primary valvar regurgitation in order that appropriate management can be decided. The present report is concerned with the clinical and surgical management of three patients with proven aorto-left ventricular tunnel seen in the National Heart Hospital in the past four years, and the clinical data on a fourth patient operated on elsewhere are mentioned.

Findings

Presentation

There were two boys and one girl. Heart murmurs were recognized early in all, on day one by the general practitioner who delivered the child and at 3 weeks and 2 years in the others. The features suggesting aortic regurgitation were obvious in the first two by the age of 1 year, and the child whose heart abnormality was recognized at the age of 2 was diagnosed initially as a persistent duct.

Two children had recurrent bronchitis in the first three years and two had increasing effort dyspnoea by the age of 6 years. Consultation was sought in two be-

cause the mothers noted the vigorous chest pulsation. No patient had angina.

Clinical examination

The three children were of normal build, and were referred to the National Heart Hospital at 5, 6, and 7 years. All were referred for surgical treatment for supposed severe aortic valve disease.

Visible vigorous carotid pulses and a hyperdynamic enlarged left ventricle were constant features. The blood pressure varied from 140/50 to 150/10 mmHg. A diastolic thrill in the second and third left interspace was noted in two patients and was also evident to the right of the sternum in one and in the suprasternal notch in another. One patient had an intense systolic thrill while the others had a short murmur in systole only. All had a loud ejection sound at the apex and along the left sternal border. Aortic valve closure was always clear and the diastolic notch was obvious on the carotid trace in all three patients (Fig. 1). Pulsation to the right of the sternum was visible and palpable in all three.

Chest radiography

There was obvious dilatation of the ascending aorta in all patients, and this had been present since infancy in two (Fig. 2a, b). The left ventricle was enlarged in all, and the 7-year-old boy, who was breathless, had prominent upper lobe pulmonary veins suggesting pulmonary venous congestion. Two patients had a visible bulge in the region of the infundibulum below the main pulmonary artery shadow which was not obvious on all films but could be seen on screening to be pulsating vigorously particularly when the patient was turned into the right anterior oblique.

Electrocardiograms

All had significant increase in left ventricular voltage such as would be expected in long-standing aortic regurgitation.

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FIG 1 Phonocardiogram from 8-year-old boy with aorto-left ventricular tunnel showing an intense immediate diastolic murmur (Edm) and a short ejection systolic murmur (Esm) at the left sternal edge. The dicrotic notch (dn) is obvious on the carotid tracing. An a wave (a) has been recorded on the apex cardiogram taken from the left ventricle.

FIG 2 Chest radiographs from a patient with aorto-left ventricular tunnel. (a) Age 4 weeks showing cardiac enlargement and prominence of the ascending aorta which could be confused with thymus gland. (b) Age 6 years showing unusual prominence of the ascending aorta and a bulge below the pulmonary area on the left border due to the aneurysmal supracardiac part of the tunnel.

FIG 3 Biplane Elema aortograms from 7-year-old girl with aorto-left ventricular tunnel. (a) Anteroposterior view showing free regurgitation into the left ventricle mainly through the right side of the aortic valve. The aortic valve ring is dilated and there is some central regurgitation as well. (b) Lateral view. The right anterior aortic sinus is grossly dilated and distorted and the tunnel contributes to the abnormal appearance. Central regurgitation through the aortic valve ring is present.

FIG 4 The appearance of the heart and aortic root at operation for aorto-left ventricular tunnel as viewed through a median sternotomy. The dilated and rotated right aortic sinus. The right coronary artery was 'absent'. The ascending aorta is also dilated.

Investigation

Right heart catheterization was normal in all. Gradients of 5 mmHg and 18 mmHg were recorded across the aortic valve in two and there was no gradient in the other.

The striking features were seen on aortography. Two had aortograms using Elema still films and cineangiograms were available in two. One patient had both investigations. There was dilatation of the right aortic sinus which looked abnormal in both planes (Fig. 3). The aortic valve ring was large in all and aortic regurgitation was free. As the reflux of contrast medium was severe it was difficult to localize its origin, but in two there was the appearance of a short channel, seen on the lateral view, which appeared to originate from the abnormal right aortic sinus of Valsalva.

Surgical technique

In each patient a median sternotomy and normothermic bypass were used. The external appearance showed unusual dilatation of the ascending aorta and aortic root with a particular localized large bulge anteriorly in the region of the right aortic sinus which extended over the right ventricular outflow tract or base of the main pulmonary artery (Fig. 4). There was an intense 'to and fro' purring in this, and firm pressure on it abolished the thrill and caused the systolic pressure to rise.

On opening the aorta, the aortic valve was seen to be of tricuspid configuration in all and rotated anticlockwise as viewed from above so that the commissure between the right and non-coronary cusps was in the sagittal plane. There was thickening of the right and non-coronary cusps, and in one patient there were lacunae at one commissural edge of a deep left coronary cusp. One boy had a thick abnormal valve of tricuspid configuration with minor fusion of the points of commissural attachment only. One patient had an absent right coronary artery and in the others both orifices were normal and separate from the tunnel enabling coronary perfusion to be established. Two patients had no coronary perfusion.

In each patient there was an opening at the base of the right aortic sinus (Fig. 5) separate from and adjacent to the right coronary artery orifice when present. This defect, about 1 cm in diameter, led into an endothelium-lined tunnel which merged into a dilated ampulla beyond. This in turn opened by a slit-like orifice into the left ventricular outflow. A sound was placed in the right coronary artery to protect it and in one patient the ampulla was filled with gel foam and the orifice in the right sinus was patched with pericardium; the left ventricular opening was closed with multiple interrupted 000 sutures. In the other two patients repair consisted of obliteration of the tunnel and closure of the aortic and ventricular openings by direct suture. The bypass time was 31 to 50 minutes and the heart in each patient came off bypass without difficulty.

Postoperative course

In one boy, aged 8, an immediate diastolic murmur was noted on the second postoperative day and though he left hospital with a normal pulse and blood pressure he had signs of a moderate aortic regurgitation 18 months later.

Reinvestigation confirmed this to be occurring through the centre of the valve. It was decided not to reoperate because the heart was smaller than before operation, the patient was asymptomatic, and valve replacement might have been necessary.

The two other children, now aged 8 and 9 years, remain well, with normal sized hearts and regression of the electrocardiographic evidence of left ventricular hypertrophy. One has a soft immediate diastolic murmur noted two years later.

Discussion

Aorto-left ventricular tunnel is a rare anomaly and until now only 14 patients have been reported since Levy and his colleagues (1963) drew attention to the condition. As well as these three patients, another girl aged 16 with florid aortic regurgitation was diagnosed by Dr. Paul Wood as Marfan's syndrome affecting the right aortic sinus (Fig. 6) and operated upon elsewhere by Mr. W. Cleland in 1962. It was clear that she had a tunnel which was successfully closed, but she died 8 years later and after 2 further operations for severe central aortic regurgitation due to annular dilatation. Our first patient, in spite of his short thick set stature, was thought to have a Marfan aortic pathology because of the severe dilatation. Histology of the ascending aorta taken at the time of operation was normal and his tunnel was successfully closed. The next two patients were correctly diagnosed before operation.

The diagnosis of a tunnel should be considered in any infant or young child presenting with signs suggesting gross aortic regurgitation; the earlier the age at which the signs are found the more likely is this to be the diagnosis, particularly if the murmur is noted to be loud in the first few months of life. A diastolic murmur louder than the systolic, and the presence of a diastolic thrill of maximum intensity at right or left sternal edge are additional features suggesting the diagnosis; loud aortic valve closure is evidence of good cusp tissue. Congenital valvar lesions are rarely the cause of free aortic regurgitation unless complicated by infection or rupture and these events are unusual in early childhood.

Another clinical feature that supports the diagnosis of an aorto-left ventricular tunnel is a distinct dirotic notch in the presence of apparent free aortic regurgitation, as this is not present in regurgitation of valvar origin (personal observation, Dr. Richard Sutton).

Routine chest radiography shows not only remarkable dilatation of the ascending aorta which is seen with other forms of aortic regurgitation but also disproportionate dilatation of the right aortic sinus which was on occasions visible on the straight chest radiograph. The bulge was not constant and its ap-

FIG 5 The surgeon's view of the aortic valve (AV) and aortic ostium of the tunnel (T) to the left as seen from above through the opened ascending aorta. The right coronary orifice was absent in the sinus (RS). The aortic valve had three large cusps with slightly rolled edges.

pearance presumably depends on the phase of the cardiac cycle and slight rotation of patient. For this reason it is recommended that any child with a suspected tunnel should be carefully screened and turned to see if this vigorous pulsating bulge can be seen just below the pulmonary artery shadow. The dilatation of the ascending aorta may be present in the neonate and has been confirmed at necropsy (Morton, Murtagh, and O'Hara, 1969) and a personally observed (J.S.) 10-day-old child operated on in another unit. This may be confused with an enlarged thymus which is commonly seen at this age.

Another condition which may present in infancy with a loud diastolic or 'to-and-fro' murmur is Fallot's tetralogy with 'absent' pulmonary valve (Osman, Meng, and Girdany, 1969). The right ventricular hypertrophy, normal pulse pressure, and absence of dilatation of the ascending aorta should distinguish the conditions. There may also be difficulty in differentiating a ruptured sinus of Valsalva aneurysm, and even a ventricular septal defect with

FIG 6 Aortogram (AP view) from a 16-year-old girl with a proven aorto-left ventricular tunnel. The huge dilatation of the aortic sinus is shown on the left cardiac border.

pronounced aortic regurgitation. However, in our experience, patients with a ruptured sinus aneurysm do not present so early unless there is associated bacterial endocarditis; this, however, does not occur in infancy and the regurgitation is of acute onset later in life. It is likely that the 8-hour-old infant reported by Morton *et al.* (1969) as having an aneurysm of the ventricular septum had in fact an aorto-left ventricular tunnel with aortic valve stenosis. The true anatomical diagnosis of ruptured sinus into left ventricle must be extremely rare.

Aortography is necessary to confirm the diagnosis. The upper part of the right aortic sinus is constantly large, eccentrically deformed, and projects anteriorly. This should suggest the true anatomical diagnosis. It is more pronounced but similar to the appearance seen in ventricular septal defect with aortic regurgitation. The intracardiac part of the tunnel itself may be difficult to define, but, in two, a short localized jet was visible emerging from the abnormal right anterior sinus. There was obvious associated central aortic valve regurgitation in Case 1, caused by ring dilatation.

Apart from minor variations, the anatomy has been strikingly similar in each instance. The tunnel

arises from an aortic ostium high within the right sinus and about level with the right coronary ostium. Dilatation of the right aortic sinus is always present. Levy and his associates (1963) described the origin of the tunnel above the right coronary orifice as one of the anatomical features which distinguished a tunnel from a right coronary sinus aneurysm rupturing into left ventricle. However, Bove and Schwartz (1967) observed that the relation between the openings of the tunnel and the coronary artery were not constant, and agreed with Roberts and Morrow (1965) that the value of this criterion for distinguishing the two conditions was limited. When the right coronary ostium is absent, as in one of our patients, this adds to the difficulty.

Inspection beyond the opening shows there is a true tunnel leading from it, and this supracardiac part of the tunnel may be aneurysmally dilated. From this, it passes as a wide ampulla-like structure through that part of the ventricular septum which forms the posterior wall of the right ventricular infundibulum. In so doing it bypasses the aortic valve and enters the aortic vestibule beneath the intercoronary commissure.

Levy *et al.* (1963) also suggested that aneurysmal dilatation of the tunnel might be an acquired phenomenon. However, there does not appear to be any correlation between the presence of such dilatation and the age of the patient. It was seen in a 16-month-old boy (Cooley, Harris, and Rodin, 1965) and is present in the newborn, so it is likely to be part of the developmental anomaly or that the aorta dilated in fetal life.

In 4 of the 14 patients previously reported, gradients have been demonstrated across the out-flow tract of the right ventricle. It is presumed that the gradient results from compression of the posterolateral wall of the infundibulum by the tunnel as it passes through the ventricular septum. This area is well cephalad to the conduction pathways, and dysrhythmias have not been a feature of the condition which is unlike ruptured aneurysm of the sinus of Valsalva. It is of interest that two patients have been reported as having ruptured aortic sinus into left ventricle (Scott, Collins, and Sinclair-Smith, 1964; Morton *et al.*, 1969) and clearly in retrospect had aorto-left ventricular tunnels with the same anatomical features as the other described cases. It is likely that Tasaka *et al.* (1960) and Warthen (1949) also were describing a tunnel and not a ruptured sinus aneurysm.

Levy *et al.* (1963) presented angiographic evidence that blood flows through the tunnel in systole as well as during diastole, which is confirmed by cine-angiography in our two patients. This may contribute to the coarse systolic bruits noted in some patients.

In our cases digital compression of the tunnel at operation regularly obliterated the continuous thrill palpable over it. At the same time, the systolic pressure rose and the heart rate slowed as with other fistulous communications.

Associated aortic valve abnormalities have been found. These may be acquired from free aortic regurgitation stretching the cusps and valve ring as in two of our patients where the cusps were rolled and thickened, or congenital such as a bicuspid aortic valve (Roberts and Morrow, 1965) or aortic valve stenosis which caused the patient's death after closure of the tunnel (Cooley *et al.*, 1965). Because of these abnormalities the patients should be protected from bacterial endocarditis throughout life even in the absence of any haemodynamic dysfunction after successful operation.

Another feature noted is the anticlockwise rotation of the aortic valve when viewed from above, so the right coronary sinus is directed anteriorly and to the left. This is similar to the appearance of the aortic root in patients with ventricular septal defect and aortic regurgitation (Somerville, Brandao, and Ross, 1970) and suggests a possible common developmental origin. The angiographic appearance of the aortic root in the lateral view is also similar in the two conditions, but the sinus dilatation is greater in the patients with a tunnel. Ventricular septal defect with aortic regurgitation probably results from a disturbance in the incorporation of the proximal bulbus cordis into the ventricle. The bulbus forms not only part of the distal right and left ventricle (infundibulum and vestibule) and the membranous septum but also the semilunar valves and roots of the great vessels and sinuses. The tissue in the sinuses of Valsalva is relatively free of elastic tissue but distal to the bulbar root is the divided truncus and its thick elastic walls. We suggest that the tunnel arises at the junction of the distal bulbus where it joins the dividing truncus and results from a defect in that area. The re-entry into the left ventricle is difficult to explain. It may be as a result of ischaemic compression necrosis in the fetus, but the smooth appearance of the endothelialized ampulla is against this.

It is of interest that when associated abnormalities have been described with a tunnel they are ones that arise from defective bulbus cordis development, such as pulmonary and aortic valve stenosis. Levy *et al.* (1963) suggested that the tunnel represented an anomalous coronary artery and that its subsequent dilatation was an acquired phenomenon. Morgan and Mazur (1963) considered that the aorto-left ventricular communication might result from primary separation of the aortic annulus from the

fibrous skeleton of the heart, while Cooley *et al.* (1965) believed, on the basis of histological evidence, that the condition was related to Marfan's disease. However, none of these theories accounts for all the patients described and no other case had histological changes of Marfan's disease. We think that a disturbance in development and incorporation of distal bulbus cordis is more likely though it is not clear how the actual tunnel is formed.

Only five successful operations for this condition have previously been reported (Levy *et al.*, 1963; Bove and Schwartz, 1967; Bernhard, Plauth, and Fyler, 1970; Fishbone, DeLeuchtenberg, and Stansel, 1971). These authors' patients were aged 2 to 11 years. In each instance the only procedure performed was closure of the aortic ostium of the tunnel with or without obliteration of the tunnel. As in congenital aortic valve stenosis, aorto-left ventricular tunnel has a strong male preponderance. Only two female cases have previously been described and now a further two are added.

The importance of distinguishing aorto-left ventricular tunnel from congenital valvar regurgitation lies in the different indications for operation that exist in the two conditions. In children with valvar regurgitation, valve replacement is nearly always necessary to correct the lesion and operation should in our opinion be deferred for as long as is reasonably possible. Conversely, in patients with aorto-left ventricular tunnel, closure of the tunnel in early childhood is advisable before severe dilatation of the aortic annulus has caused secondary changes in the basically normal aortic valve. Ideally this should be before 5 years. This is suggested by our patient aged 8 who had persistent central aortic regurgitation after the tunnel was closed which might not have occurred if it had been corrected earlier. The lesson learnt from the 16-year-old girl, cited earlier, supports this view. In the other two cases operated on earlier, good aortic valve function was evident after closure of the tunnel and confirmed by postoperative aortography.

This lesion may cause heart failure in infancy and demand emergency correction at this time. More probably it will present in an asymptomatic child over 1 year with cardiomegaly and severe aortic regurgitation. Tunnel should be the first diagnosis in such a patient and should be confirmed by aortography. Early closure of the tunnel should obviate the need for aortic valve replacement which may arise if the child is allowed to wait until seriously symptomatic or until the second decade.

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Addendum

Since this paper was submitted a further patient, a man aged 26 years, has been operated on for this condition. After the aorta was opened to confirm the diagnosis, the ampulla was opened; this was lined by endothelium and was structurally similar to a dilated aortic sinus of Valsalva. There were two openings from this, one above the right coronary cusp opening into the ascending aorta and the lower one obliquely below the right coronary cusp

in the left ventricle. Thus, there was a deficiency above and below the base of the cusp enclosed by the dilated ampullary structure. In addition to this and opposite the lower opening was a jet lesion in the non-coronary cusp with a perforation; there was no history to suggest bacterial endocarditis, though six months earlier he had ex-

perienced sudden palpitation and his friends commented on the purring in his chest probably related to the small perforation of the cusp. In addition, the anterior cusp of the mitral valve was a thin, billowing, sail-like structure with a web of tissue between the chordae. The cusp was plicated.