Aorto-left ventricular communication after closure

Late postoperative problems

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SUMMARY The long-term follow-up of six patients operated on for aorto-left ventricular communication has been reviewed in detail. All had residual aortic regurgitation after the initial repair of the defect. It was severe in four and required repeated reoperation in three with ultimate aortic valve replacement.

The failure of early repair to solve the haemodynamic problem has provoked a reconsideration of the basic anatomy, of the surgical approach, and of the postoperative physiology of this anomaly. The so called “tunnel” is not a tunnel with length but should be considered as a localised breach at the insertion of the right coronary cusp. The localised aortic root dilatation at the site is a weakness that remains after closure of the tunnel leaving a poorly supported aortic valve and a weak root. Thus, the initial repair of the aorto-left ventricular communication must not only close the communication but reinforce, strengthen, and support the right aortic sinus in order to maintain cusp competence.

A congenital aorto-left ventricular communication, known as a “tunnel”, presents as severe aortic regurgitation in the neonate, infant, and child. Theoretically, closure of the communication should solve the haemodynamic problem, but reports show that severe aortic regurgitation may persist. Unfortunately, later aortic valve repair has failed in our hands so that ultimately aortic valve replacement has been necessary in some cases.

It was hoped that early closure of the defect would prevent secondary effects on the aortic root and cusps, but our recent experience suggests that this is not so.

This has prompted us to review the late results of previously reported patients, add new experience, and reconsider the anatomy of this congenital defect.

Subjects and methods

The clinical status of six patients operated on for aorto-left ventricular communication (tunnel), seen by one of us (JS), from 1970 to 1982, has been reviewed in detail. Patients were observed for one to 12 years (mean 8.5 years) after operation and no patient has been lost to follow-up.

Clinical details, and operative and postoperative findings are summarised in the Table.

All the patients have been reviewed regularly at annual intervals and particular attention has been directed to the presence and degree of aortic regurgitation. At each attendance an electrocardiogram, chest x-ray, and M-mode echocardiogram were done. Real time echocardiography was available only during the last year and so in only one patient was there a record before operation.

The severity of left ventricular hypertrophy on the electrocardiogram has been graded 1 to 4 according to previous communications.

Postoperative cardiac catheterisation with aortography and left ventriculography was performed two to seven years later in three patients. The degree of aortic regurgitation was assessed according to Sellers et al., and graded from 1 to 4.

M-mode echocardiography was performed in three patients preoperatively and in all after the operation, using an Ekoline 20 A echocardiograph, positioning the probe in the third left intercostal space. Real time echocardiography was performed before operation in one patient and after operation in all, using an ATL

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wide angle mechanical sector scanner, following a routine approach to long axis and short axis of the main chambers, from the parasternal, apical, and subcostal position.

Results

PREOPERATIVE STATE
Before the first operation all patients presented with signs of severe aortic regurgitation, bounding peripheral pulses, wide blood pulse pressure, but no evidence of heart failure. The electrocardiogram showed grade 1 left ventricular hypertrophy in four patients, grade 3 in one, and grade 4 in one. The chest x-ray film showed mild enlargement of the heart size in two (CTR 0.6 to 0.55), moderate in three (CTR 0.73 in all), and severe in one (CTR 0.73). There was enlargement of the aortic root in all, as previously noted.11

SURGICAL FINDINGS
The aortic root and the ascending aorta were grossly dilated in all patients, with pronounced bulging of the right aortic sinus over the right ventricular outflow tract.12-14 This looked like a “cherry” at the base of the sinus or a small “plum” in the older patients. On opening the aorta, a defect was seen between the outer border of the right coronary cusp and the left ventricle (Fig. 1) within the “cherry”. The diameter of the communication ranged from 2 mm to 1.25 cm. The orifice of the right coronary artery was seen to be separate from the defect. The coronary arteries were normal in all but one in whom the right coronary artery was displaced superiorly by the dilated sinus (case 2).

The aortic valve was tricuspid in all patients. There was anticlockwise rotation of the aortic ring in one patient (case 2) so that the commissure between right and non-coronary cusp lay in the sagittal plane; in one patient (case 4) there was a slight fusion of the posterior commissure, an association reported also by others.15

The communication between the right aortic sinus and left ventricle was closed by mattress sutures in five patients and by a pericardial patch in one (case 2). In case 6, the right coronary cusp looked distorted after the closure.

The bypass time was 30 to 60 minutes and no perioperative arrhythmia was noted.

Fig. 1 Diagrammatic demonstration of the anatomy of aorto-left ventricular communication. The arrows indicate the regurgitation through the communication and the turbulence against the aortic wall. There is no real “tunnel” present. Ao, aortic root; LV, left ventricle; LA, left atrium.
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Fig. 2 Two dimensional echo taken from case 6, before operation. (A) The parasternal long axis view of the left ventricle shows the delineation of the communication (c) and its opening in the left ventricle, beneath the right coronary cusp. (B) The parasternal short axis view of the aortic valve shows the large bulging of the communication (c) over the right ventricular outflow tract (RVOT). The aortic ring, valvular commissures, and origin of the left coronary artery are visualised. Ao, aortic root; LA, left atrium; LV, left ventricle; RV, right ventricle.

ECHOCARDIOGRAPHY

M-mode

In three patients, a double contour of the anterior wall of the aorta with systolic obliteration was seen before operation.7,16 The postoperative echo showed persistent left ventricular enlargement and dilated aortic root, with thickened right coronary cusp in all six patients, as well as fluttering of the mitral valve in five, but no double contour was seen over the root.

Real time

In the parasternal long axis views of the left ventricle...
there was a persistent drop-out at the anterosuperior part of the interventricular septum, extending to the right coronary cusp, so that a free communication between left ventricle and aorta was seen (Fig. 2A). The same abnormal drop-out was seen in parasternal short axis view of the aortic valve, as a "crescent" shaped structure wrapping around anteriorly the right coronary cusp, clearly distinct from the aortic root. The size of the aortic root was increased (Fig 2B).

After the repair, the two dimensional echocardiogram showed continuity between interventricular septum and the anterior wall of the aorta. No drop-out was seen in any part. The aortic valve presented with floppy and prolapsing cusps. Left ventricular enlargement and impairment of the contraction were also present (Fig. 3).

FOLLOW-UP
The immediate postoperative course was uneventful in five patients; one aged 19 months had chronic left ventricular failure from severe aortic regurgitation, requiring inotropic drugs and mechanical ventilation for four days.

An aortic diastolic murmur was obvious in all patients soon after the operation. In two patients (cases 1 and 3) the aortic regurgitation has remained mild or moderate for nine years, with decrease of heart and aortic size on chest x-ray film and improvement in the electrocardiogram.

The other four patients showed progressive worsening of aortic regurgitation, with persistent or increased cardiomegaly on the chest x-ray film and signs of increased left ventricular hypertrophy on the electrocardiogram. Three had further cardiac catheterisation, showing pronounced aortic regurgitation, with thickened aortic cusps, and dilated left ventricle with reduced contractility. One patient (case 6) now aged 2½ years has serious aortic regurgitation, and cardiomegaly is controlled by medical treatment in the hope of delaying valve replacement.

Reoperation
Three patients were reoperated on four to five years after the first operation. In one (case 2) the aortic ring was grossly dilated, without central coaptation of the cusps. In two (cases 4 and 5), also with considerable ring dilatation, there were new holes in the cusps.

Aortic valve repair was attempted in all. After this, the three patients had persistent and increasing aortic regurgitation, so that eventually the aortic valve was replaced four to seven years later. At reoperation a dilated aortic ring, with thickened rolling edges of the cusps, was found, leading to loss of central coaptation. In one patient (case 5) a residual communication between the aorta and left ventricle was found at the site of previous repair by mattress sutures.

The aortic valve was replaced by Carpentier-Edwards xenograft valve in two cases and by a Dacron conduit with a Starr mechanical valve in one.

All patients are alive and leading a normal life. Three with replaced aortic valve aged 12 to 21 years have no aortic regurgitation. Two patients had mild
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aortic regurgitation for nine to 10 years and one awaits reoperation.

Discussion

The word "tunnel" was first used by Levy et al. in 1963,\textsuperscript{1} to describe what was thought to be an ampulla-like or a vessel-like structure with two distinct openings, one proximal in the right coronary sinus and the other distal, in the left ventricle, just below the aortic valve.\textsuperscript{2} This term has been maintained over the years to classify the 28 cases already reported.\textsuperscript{11,15} Tunnel, however, means "an elongated passageway, usually open at both ends".* If this was the true morphology of the lesion, closure or obliteration should solve the problem. Our experience shows this is not so. The pathological problem, in fact, lies in the peculiar susceptibility of the right coronary sinus to congenital anatomical defects like aneurysms and ruptured sinus of Valsalva, aorto-left ventricular tunnel, and ventricular septal defect with aortic regurgitation.\textsuperscript{18,19} All relate to this thinned out anterior wall of the left ventricular outflow where the right aortic sinus meets the membranous septum.\textsuperscript{20}

At this site, any intrauterine injury or noxious effect on cardiac development can produce a breach between aortic sinus and left ventricle, and the regurgitation through the communication determines high flow turbulence against the right coronary sinus wall, causing its progressive dilatation (Fig. 1). Closing such a defect by direct suture, which seems the obvious thing to do, may distort the cusps, pulling them towards the weak aortic wall which remains unsupported within the dilated aortic sinus. Because of the distortion, it is not surprising that aortic regurgitation may persist and progress even if repaired in infancy.\textsuperscript{21}

Even after a patch was placed, the cusp was seen to be distorted and pulled out the thin dilated aortic wall of the right sinus, a situation noted by others.\textsuperscript{4,7,21}

It is interesting to note that two patients aged 34 and 19 had stable mild aortic regurgitation only; no significant difference could be found in their anatomical and clinical features or in the surgical management, compared with the other cases, to justify such unusual behaviour.

From our experience, it appears that the early operation has not determined a good haemodynamic result as previously suggested.\textsuperscript{3,6} Despite this, we believe that the defect must be repaired early, but modification in technique and understanding is mandatory. The surgeon is not only required to close the communication, distorting the cusps as little as possible, but also to reduce the size of the aneurysmal sinus and aortic root and strengthen the aortic wall in the same region. This could reduce the subsequent turbulence and avoid progressive distortion of the cusps and later regurgitation with the need for subsequent valve replacement.

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