Aortic Regurgitation with Ventricular Septal Defect

Surgical Management and Clinical Features

By JANE SOMERVILLE, M.D., M.R.C.P., A. BRANDAO, M.D., and DONALD N. ROSS, F.R.C.S.

SUMMARY
Twenty patients with ventricular septal defect and aortic regurgitation had surgical correction using cardiopulmonary bypass. Direct suture of the ventricular defect through the aorta and repair or replacement of the aortic valve were performed. Homograft replacement of the aortic valve was preferred to repair and was associated with less aortic regurgitation after operation. There were one operative death and one late death due to aortic regurgitation. Follow-up for 1 to 6 years showed that 17 of the 18 survivors have had a satisfactory result.

Additional Indexing Words:
Cardiac catheterization Aortic valve abnormality Angiocardiology
Homograft Electrocardiograms

THE ASSOCIATION of aortic regurgitation with ventricular septal defect is well known. Views about the pathology and surgical treatment of this combination of lesions have differed. Robinson and co-workers1 and Blumenthal and his colleagues2 believe that the prolapsed right coronary cusp causing aortic regurgitation is related to the site of the ventricular septal defect and that closure of the defect early in the natural history may prevent or cure aortic regurgitation. Ellis and associates3 found that the ventricular septal defect was often relatively small and that repair of the aortic valve as well as closure of the ventricular septal defect was necessary to improve the patient. In the current series, the aortic valve abnormality has been considered to be a related but an important separate congenital abnormality from the ventricular septal defect and repair or replacement of the aortic valve has been performed in all patients.

Group Studied
Twenty patients with ventricular septal defect and aortic regurgitation were operated on (by D.N.R.) using cardiopulmonary bypass between January 1963 and March 1969. The clinical details, electrocardiograms, and chest roentgenograms were studied before and after surgery.

Results
Clinical Features
The clinical data has been summarized in table 1. Patients were aged seven to 46 years at the time of surgery. Fifteen of the patients were under 20 years, and three of the 15 were below 10 years. There were 14 males and six females. A murmur was heard during infancy in 13 patients and detected in childhood in all the rest, except for patient 6 in whom it was noted for the first time at 36 years of age during bacterial endocarditis. It is presumed that the murmur noted in infancy was systolic since the clinical diagnosis of aortic regurgitation or the documentation of an immediate diastolic murmur was made after the age of 3 years in all except patient 2 in whom aortic regurgitation was noted in the first year. The pathologic abnormality in the

From the National Heart Hospital and Institute of Cardiology, University of London, London, England.

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### Table 1

**Clinical, Anatomic, and Surgical Data**

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*Perfect result until 16 months after operation when dyspnea and diastolic thrill appeared.

Abbreviations: Ang = angina; AF = atrial fibrillation; AR = aortic regurgitation; AS = subaortic stenosis; ASD = atrial septal defect (secundum); BP = blood pressure; CHF = congestive heart failure; CTR = cardiothoracic ratio; DS = direct suture; FU = follow-up; H = homograft; HD 64 = heart disease found; IDM = immediate diastolic murmur; LA + = left atrium enlarged; LVF = left ventricular failure; LC = left coronary cusp; LVH = left ventricular hypertrophy; NC = noncoronary cusp; OP = operation; P = patch; PND = paroxysmal nocturnal dyspnea; Pulm. pleth. pulmonary plethora; PS = organic subpulmonary stenosis; P. vein + = pulmonary veins prominent; R = repaired; RC = right coronary cusp; Resut. = resutured; RVH = right ventricular hypertrophy; S = Starr-Edwards prosthesis; SBE = subacute bacterial endocarditis; SM = systolic murmur; ST or T Abn. = S-T and T-wave segment abnormal in V1 and V6; VSD = ventricular septal defect.

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aortic valve in this patient was similar to the 
rest, but at the time of surgery, when the 
patient was 14 years of age, the heart was 
grossly enlarged and the wedge pressure at 
rest was elevated which suggested that the 
severe aortic regurgitation was long standing.

Recurrent respiratory infections and difficul-
ties in thriving occurred during the first year 
of life in three patients, and heart failure 
requiring digoxin was documented in two of 
them. The ventricular septal defect was 
subsequently found to be large, 2 cm or more 
in diameter. Seventeen patients had no 
thriving problems in infancy and only one of 
these (case 12) had a large defect over 2 cm 
in diameter.
Thirteen patients had mild symptoms, either effort dyspnea or palpitation at the time that aortic regurgitation was noted. Dyspnea was long standing and was associated with central cyanosis and severe infundibular stenosis in case 5. At the time of operation 17 patients had symptoms, four had left ventricular failure, three angina, and the rest effort dyspnea, palpitation, or subacute bacterial endocarditis. Those with attacks of left ventricular failure had serious aortic regurgitation; the ventricular septal defect was small in all except one in which it measured 2.5 cm by 1.5 cm which was only associated with a small left-to-right shunt. Effort angina started at 17, 21, and 43 years of age in three patients who had extreme left ventricular hypertrophy with S-T depression and T-wave inversion. Palpitation experienced by five was probably due to extrasystoles which were documented in three and were not precipitated by medication. Patient 17 appeared at the National Heart Hospital for the first time at age 14 in gross cardiac failure with large liver, ascites, edema, and extreme dyspnea, a difficult diagnostic problem.

Three patients had no symptoms at the time of operation which was performed because of severe left ventricular hypertrophy and cardiomegaly. Three patients had bacterial endocarditis due to the Streptococcus viridans. In two there was perforation of the noncoronary cusp but there was no evidence of infective damage in the third (case 3).

**Physical Examination**

Central cyanosis and clubbing was present in one patient with severe pulmonary infundibular stenosis and large ventricular septal defect. There was no central cyanosis in case 20 with equally severe pulmonary stenosis in which ventricular septal defect was small. All patients had a collapsing pulse and visible carotid pulsations. The resting diastolic systemic blood pressure was below 60 mm Hg in 18. Blood pressures were recorded from 200-100/70-0 mm Hg. Large A waves in the jugular venous pulse were seen in seven patients with severe aortic regurgitation. The left ventricle was clinically enlarged and overactive in all, but separate right ventricular pulsations were also identified clinically in five patients.

A pansystolic murmur with a thrill was present in every patient except the cyanosed patient who had a long ejection systolic murmur. An additional ejection systolic murmur was recorded in five patients all of whom had a pressure gradient across the outflow tract of the right ventricle. An immediate diastolic murmur was constantly present but was not associated with a thrill in any; delayed apical diastolic murmurs were noted in four patients with severe aortic regurgitation. In these patients the measured pulmonary blood flow was less than twice the systemic flow; thus it was assumed that this murmur was an Austin Flint type and related to the aortic regurgitation. Analysis of the second sound was difficult as aortic valve closure ($A_2$) was often drowned by the loud systolic murmur (fig. 1).

**Electrocardiogram**

Sinus rhythm was present in all patients. Widened P waves due to left atrial enlargement were seen in four, all of whom had important dyspnea. A QRS in the frontal plane was between $+20^\circ$ and $+90^\circ$ in 17 patients, and right axis deviation was present in six. Five patients with left axis deviation ($-30^\circ$) had gross aortic regurgitation and as the ventricular septal defect was in the usual

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**Figure 1**

Phonocardiogram from case 12 showing typical intense pansystolic murmur with midsystolic accentuation at pulmonary area (PA) and left sternal edge (LSE). Immediate diastolic murmur (DM) recorded at PA at high frequency (HF).
supracristal position, the left axis deviation was attributed to fibrosis in the severely hypertrophied left ventricle and not to the site of the defect. Important left ventricular hypertrophy was constantly present (fig. 2) with changes in S-T segment or T waves in 14. Associated right ventricular hypertrophy was present in three, two of whom had organic pulmonary stenosis with gradients of 92 mm Hg and 60 mm Hg and one with pulmonary artery pressure of 60/38. No electrocardiographic evidence of right ventricular hypertrophy was present in one patient whose right ventricular pressure was 60 mm Hg. The electrocardiogram in case 17 with a large pericardial effusion showed the high voltage of left ventricular hypertrophy which presumably would have been even greater in the absence of the effusion.

Chest Roentgenogram

Cardiac enlargement was present in all patients and was gross in 15 with cardiothoracic ratios of 60 to 80% (fig. 3). Left atrial enlargement was present in seven. Eleven of this group had right heart catheterization which confirmed a left-to-right shunt in 10. The aorta was prominent in its ascending portion or knuckle, or both, in 11 patients. The aortic arch was always left sided.

Thirteen patients were considered to have pulmonary plethora by clinicians, but independent assessment by radiologists diagnosed it in only eight, although the left-to-right

Figure 2

Electrocardiograms from case 12 before operation and 14 months after closure of the ventricular septal defect and homograft replacement of the aortic valve showing regression of left ventricular hypertrophy.
Figure 3
Chest radiographs from case 13. (A) Before and (B) 15 months after operation.

Figure 4
Chest radiographs. (A) Before and (B) 6 months after closure of the ventricular septal defect, homograft replacement of the aortic valve, and treatment of a possible myocarditis in case 17.
shunt was confirmed in 10 of the 13 and the other three were not catheterized. The lung fields appeared normal in six and a small left-to-right shunt was demonstrated in two. Underfilled lungs were seen in the patient with severe pulmonary stenosis and large ventricular septal defect. The only radiographic feature to suggest that this patient was not a straightforward example of Fallot's tetralogy was cardiomegaly. Prominent upper lobe pulmonary veins were evident in five patients; four of these had a history of left ventricular failure. The lung fields in case 17 with gross aortic regurgitation and pericardial effusion showed large vessels suggestive of a left-to-right shunt and also prominent upper lobe veins (fig. 4a).

Cardiac Catheterization

The hemodynamic data have been summarized in table 2. Sixteen patients had right heart catheterization, but relevant information was only available in 15. Eleven had a left-to-right shunt at ventricular level which was not found at subsequent catheterization in two (cases 6 and 15). The increase in oxygen saturation usually appeared in the outflow tract of the right ventricle and the demonstrable left-to-right shunt was usually small, but unrelated to the size of the ventricular septal defect. A right-to-left shunt was demonstrated in case 5 and no shunt was found in the rest.

A systolic gradient across the infundibular region was demonstrated in 12 of the 15

Table 2

Preoperative Hemodynamic Data from 20 Patients with Ventricular Septal Defect and Aortic Regurgitation

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Abbreviations: A = aorta; Inj = injection; LV = left ventricle; PA = pulmonary artery; PCV (m) = pulmonary capillary wedge (mean); P/s = flow ratio; RA = right atrium; RV = right ventricle; SVC = superior vena cava; Syst A = systemic artery.

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aortic regurgitation which was moderate or severe in all cases. The site of the regurgitation was difficult to localize from the angiogram, but mostly commonly appeared to occur through the anterior part of the aortic valve (fig. 5). In five patients aortic regurgitation appeared into the right ventricle as well as into the left ventricle.

Left ventricular angiocardiography was performed in five patients. Early opacification of the right ventricular outflow tract without filling of the body of the right ventricle was noted in patients with a supracristal defect (fig. 6). In spite of left ventricular cineangiography, subaortic fibromuscular stenosis was not identified before operation (case 18). Right ventricular angiocardiography in cases 2 and 20 demonstrated infundibular stenosis.

Previous Surgery

Three patients had had operations before radical correction. One patient (case 5) had a transventricular pulmonary valvotomy without benefit 7 years before total correction. In patients. In four the gradient was 50 mm Hg or more and three had obvious organic subpulmonary stenosis. Complete prolapse of the right coronary cusp through the defect contributed to the obstruction in case 5. In the rest of the patients with a gradient, the parietal band of the crista supraventricularis was displaced and hypertrophied.

Elevated pulmonary artery pressure above 60 mm Hg was found in two patients who had been in left ventricular failure. Patient 17 with elevated pulmonary artery pressure had a probable myocarditis and chronic pericarditis with effusion with subsequent pericardial constriction following pericardial biopsy. Pressure measurements taken during a radiologic investigation in case 18 failed to demonstrate the subaortic stenosis which required resection.

Angiocardiography

Aortography confirmed the presence of

**Figure 5**

Aortogram from case 16 showing aortic regurgitation without clear localization of the anatomic lesion in the aortic valve. Prominence of the right coronary sinus of Valsalva is present which was a frequent finding.

**Figure 6**

Left ventricular angiocardiogram from case 16 showing opacification of right ventricular outflow tract through a supracristal defect.
AORTIC REGURGITATION

case 20 thoracotomy was performed in 1948 to ligate a duct, and the left pulmonary artery was ligated accidentally; there was no duct. In case 17 exploratory thoracotomy had been performed to biopsy the pericardium and possibly the mediastinal glands and to ligate a duct if found.

Anatomy of the Ventricular Septal Defect

In 16 patients the ventricular septal defect was only visualized from the left side through the aortic valve; thus its exact relationship to the crista supraventricularis is uncertain. The defect was directly beneath the right coronary cusp in 10 patients, beneath the right and noncoronary cusps in eight, the noncoronary cusp in one, and the right and left coronary cusps in one.

Inspection of the right ventricular side of the defect in four patients showed that the defect was infracristal in two and supracristal in two. In 12 of the 16 defects viewed only from the left side, a probe passed through the defect led into the right ventricular outflow tract under the pulmonary valve and therefore, it was assumed that these defects were supracristal. In four the site of the defect in relation to the crista was uncertain, but the surgeon commented that it was probably infracristal in two. Thus, at least four defects were probably infracristal.

The defects were less than 2 cm in diameter in 12 patients, 2 cm in four, and 2.5 cm or more in four including two with known infracristal defects. The largest defect, 3 cm (case 5), was associated with a total prolapse of the right coronary cusp. All adults had small defects.

Aortic Valve

The aortic valve was bicuspid in three and tricuspid in 17 cases. The cusp or cusps immediately above the ventricular septal defect were the ones pathologically involved in eight cases (cases 2, 5, 8, 10, 11, 13, 19, and 20). These cusps appeared to be basically abnormal with rolled edges and thickening in the center of the cusp which was sometimes abnormally long. Prolapse of the base of the cusp appeared to be the primary cause of aortic regurgitation in only four cases. In nine, the abnormality in the aortic valve affected cusps other than those directly above the defect in the same way as when the cusp was above the defect but prolapse was not a feature. Perforation of right and noncoronary cusps occurred in case 1 whose left coronary cusp was also thickened and rolled and in case 6 with bicuspid valve. Six patients had abnormality in all aortic valve cusps (fig. 7).

Characteristically in the patients with a supracristal defect, the aortic valve was

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Figure 7
Three abnormally thickened aortic cusps removed in case 16 before homograft aortic valve replacement.

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Figure 8
Diagram to show the abnormal rotation of the aortic valve in aortic regurgitation associated with supracristal ventricular septal defect.
rotated about 20° in a clockwise direction so that the anterior commissure pointed more to the left (fig. 8).

Additional Lesions

Small ostium secundum defects in the fossa ovalis were found in two patients. In two patients there was considerable hypertrophy of the moderator band, and mobilization sometimes with resection of infundibular muscle was performed. Two other patients had a typical circular fibromuscular right ventricular infundibular stenosis below the defect. One patient had a fixed subaortic fibromuscular stenosis 2 cm below the aortic valve. Case 17 with a pericardial effusion and thickened pericardium with nonspecific inflammatory changes had severe postoperative left ventricular failure which was presumed to be myocarditis as it responded immediately to steroids. Unfortunately, no myocardial biopsy was taken in this case.

The dilated thin-walled appearance of the ascending aorta in case 20 suggested cystic medial necrosis. Dissection occurred after removal of the aortic clamp and the aorta had to be partially replaced and reinforced with Dacron.

Surgical Technic

All patients were operated on at normal temperature with use of a bubble type oxygenator and a dilute blood prime.

In every case the aorta was opened first and after confirmation of the diagnosis, bilateral coronary perfusion was set up. Coronary perfusion was not used in the early cases in the series, but it was subsequently considered necessary in patients with such diseased left ventricles, as the time on bypass varied from 50 to 130 minutes. Access to the ventricular septal defect was through the aorta after retraction of the right coronary and noncoronary cusps. The defect was closed initially with multiple interrupted sutures. When it was decided to excise the aortic valve, the adjacent right and noncoronary cusps were partially detached, folded down like a double-breasted waistcoat, and incorporated in the ventricular septal defect closure to provide additional security.

Most of the ventricular septal defects were closed transversely, but in one patient (case 9) an elongated defect ran into the commisure between the right and the noncoronary cusp. This was closed vertically by bringing the adjacent cusps together at the point of commissural attachment and over-sewing the defect. In four patients the right ventricle was opened to confirm the closure of the ventricular septal defect, and additional sutures were inserted from this aspect. Simple suturing was used in 18 patients, and in two a patch of Teflon or pericardium was used.

Efforts to conserve the aortic valve were made in patients under 20 years. When repair did not restore reasonable competence, the valve was replaced. Six patients, aged 9 to 19 years, had the aortic valve repaired. One patient had a pericardial extension of the cusps, but required reoperation later and now has a good functioning homograft. Three patients were managed by shortening and plication of the prolapsing right coronary cusp and two other patients had fascia lata extension of the cusp edges. In the remaining 14 patients the aortic valve was excised; in 13 cases it was replaced by a homograft and in one by a Starr valve. The seating of the homograft valve was often complicated by the anatomy of the base of the right coronary cusp which tended to be convex instead of concave.

Associated abnormalities like subaortic stenosis were dealt with through the aortotomy, but right ventricular infundibular narrowing and small atrial defects had to be dealt with through the appropriate ventricular or atrial chamber.

Results of Operation

One hospital death and one late death occurred. The hospital death occurred in the patient (case 5) with Fallot physiology who died the day after operation, having remained in a low output state following complete correction and insertion of a homograft; coronary perfusion was not used in this case. The other patient (case 12) died abroad 1

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year after surgery apparently in heart failure; he had persistent aortic regurgitation after repair of the aortic valve. Three patients had reoperation, one for patching a ventricular septal defect which opened after direct suture (case 7), one for resuturing a Starr valve (case 3) and one for replacement of a leaking homograft by a Starr valve (case 6). The repaired aortic valve in case 7 was regurgitant at the time of reoperation and was replaced by a homograft.

Left ventricular failure occurred in the first postoperative week in four patients and transient atrial fibrillation appeared in one. All had serious left ventricular dilatation before operation. Digoxin was given routinely after operation to all the patients who had large left ventricles, and diuretics were frequently used in the first postoperative weeks.

The follow-up of the survivors has extended for 6 months to 6 years. Seven patients have been followed less than 2 years; the rest, for longer periods. All were asymptomatic. No patient has developed a pansystolic murmur in the postoperative period; it has been assumed, therefore, that the ventricular septal defect has remained closed in all the survivors.

The fate of patients in relation to the type of operation on the aortic valve has been summarized in table 3. Of 11 survivors with aortic homografts three have aortic regurgitation. In two it is trivial and has been present since operation; in the third (case 13), it appeared dramatically with dyspnea 16 months after operation when a diastolic thrill was noted. Prior to this, no murmurs were heard or documented on the phonocardiogram. Reinvestigation confirmed important aortic regurgitation into a curious pouch-like sac (fig. 9). Presumably, this was due to tearing of the homograft away from the aortic valve. No evidence of infection was found. Six patients with repaired aortic valves were followed for 1 to 5 years. Only one had a competent aortic valve and the rest had some regurgitation with moderate widening of the pulse pressure. Moderate aortic regurgitation was present in patient 3 who had an initial Starr valve replacement and in patient 6 in whom a Starr valve was used to replace a leaking homograft.

Chest radiology showed diminution of pulmonary plethora and return to normal lung vascular markings 6 to 12 months after operation (figs. 3 and 4). The cardiothoracic ratio diminished progressively with increasing periods of follow-up. There was no evidence of calcification of the graft in any patient. Eighteen survivors are back at work or school and are symptom free. No postoperative studies have been done except in case 13 in which sudden deterioration occurred.

Left ventricular hypertrophy regressed in every patient and in five patients the electrocardiogram was normal after 2 years (fig. 2). Depressed S-T segments and T inversion disappeared from the ECGs of five patients after 1 year but remained in the ECGs of seven others. During the first 6 months after operation the T-wave changes worsened in seven patients, presumably due to myopericarditis which so frequently follows cardiac

Table 3

<table>
<thead>
<tr>
<th>Operation on aortic valve</th>
<th>No.</th>
<th>Death</th>
<th>Re-op*</th>
<th>Aortic regurgitation</th>
<th>Follow-up period (yr)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Homograft</td>
<td>13</td>
<td>2</td>
<td>1</td>
<td>7</td>
<td>No</td>
</tr>
<tr>
<td>Repair</td>
<td>6</td>
<td></td>
<td>1</td>
<td>1</td>
<td>4+</td>
</tr>
<tr>
<td>Starr</td>
<td>1</td>
<td></td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

* Reoperation on aortic valve.
† Aortic regurgitation — constant immediate diastolic murmur = widening of the pulse pressure.
‡ One patient followed for 6 years.
surgery. The heart size diminished in all. No patient has had bacterial endocarditis following operation, but strict prophylactic measures have been advised for skin sepsis and any dental manipulations because bacteria may infect the homograft or the repaired aortic valve.

**Discussion**

The combination of these lesions has been described as a ventricular septal defect and prolapsed aortic cusp. Indeed, Blumenthal and associates\(^2\) have suggested that the high ventricular septal defect is the primary lesion allowing prolapse of the adjacent superimposed right coronary cusp with subsequent development of aortic regurgitation. While this was a feature in a few cases, surgical experience in this series has shown that this view cannot satisfy all cases, for the aortic valve is often abnormal in several respects and more than one cusp is usually involved. In six of the 20 cases (30%), all aortic cusps were abnormal. This suggests that prolapse of one cusp is not necessarily the prime cause of aortic regurgitation although it is possible that once a leak has occurred because of malposition of one cusp, the others may become rolled and thickened from the trauma of the regurgitant jet.

Blumenthal's view is probably correct in some patients with aortic regurgitation and ventricular septal defect, namely in the small group in which one aortic cusp is prolapsing into the defect. Aortic regurgitation appears to increase in severity with age, and its appearance after infancy in the majority might also lend support to the "prolapse" concept. A more likely concept is that in this type of bulbar ventricular septal defect the congenital abnormality of the aortic valve is liable to bacterial infection like any other left-sided valve abnormality whose function inevitably deteriorates with age.

Closure of the ventricular septal defect alone has not been successful in curing the aortic regurgitation in this series. It could be argued that, in this relatively old group of

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**Figure 9**

Aortogram 16 months after closure of the ventricular septal defect and correction of aortic regurgitation by homograft replacement from case 13. (A) Anteroposterior view. (B) Lateral view showing regurgitation into a strange pouch in the region of the right coronary sinus. Aortic regurgitation associated with a diastolic thrill appeared suddenly after 16 months without murmurs.
patients with established and often long-standing regurgitation, it is too late to use defect closure as a means of cure or prevention and that had this been done earlier it might have had some success. This might have been possible in the four patients in whom the cusp had prolapsed into the deficient septum immediately below, but in the majority of patients the ventricular septal defect was too small to have affected the function and position of the aortic valve.

The ventricular septal defect was usually supracristal in position, but occasionally was in the membranous septum and infracristal. These patients with equally severe aortic regurgitation not due to prolapse were clinically indistinguishable from the others. Unfortunately the data are incomplete on the anatomy of the ventricular septum as the defect was often only viewed from the left side.

Because of the association of high ventricular septal defect and aortic valve abnormalities frequently involving all three cusps or complicated by a bicuspid valve, an embryologic basis for the abnormality is suggested. It could result from incomplete or arrested development of the distal bulbar cushions since these not only complete the distal bulbar septum but also give rise to the aortic and pulmonary valve cusps. An abnormality of this part of the bulbus cordis thus explain the high ventricular septal defect, valve abnormalities, and the association of infundibular and subaortic lesions as well. The typical rotation of the aortic valve also supports the view that this combination of lesions develops from a single bulbar problem.

Direct surgery on the aortic valve was always necessary for a clinically successful result and to keep down the surgical mortality. As others have found, conservative surgery with cusp plication, using methods similar to those of Frater or elongation with pericardium or fascia lata has been often not a success. In this series it was attempted in the majority of patients before valve excision and replacement. The incidence of postoperative aortic regurgitation in this series was higher after cusp repair than after insertion of the cadaver aortic homograft. There was suggestive evidence from serial examination that after repair aortic regurgitation tended to increase whereas it was usually static, if present, after the use of the homograft with the exception of the unusual behavior in case 13.

Homograft replacement has been considered to be more acceptable than valve repair and five of the homografts appear to be functioning well after 3 to 6 years and have shown no signs of deterioration. Assessment of valve calcification may be inaccurate as appropriate chest radiographs were not always taken. We believe that all patients with graft replacement of the valve should have careful radiologic assessment for calcification at 6-month intervals. It is hoped that in the future a living pulmonary autograft replacement of the abnormal aortic valve will provide a permanent living replacement which may have the potential to grow with the child, although sometimes minor abnormalities of the pulmonary valve are to be expected with this bulbar disturbance and this may preclude the use of the patient's own pulmonary valve. Valves made from the patient's own fascia lata have been used to replace the damaged aortic valve and, as living tissue, may have a useful place and be likely to function better than the fascial extensions used in some of the patients in this series. Fortunately, most of these patients can await surgery until the second decade when growth is nearly or totally completed.

Clinically and surgically the ventricular septal defect is of less importance than the aortic valve lesion. Not infrequently in these cases the ventricular septal defect is unsuspected and right heart catheterization may fail to demonstrate a shunt. Clues to the presence of a ventricular septal defect with the aortic regurgitation are early recognition of a heart murmur in infancy, a long, roaring systolic murmur at the left sternal edge in the absence of other features of aortic stenosis, evidence of right axis deviation and right ventricular hypertrophy in the electrocardiogram, and of pulmonary plethora in the chest radiograph.
Pulmonary hypertension was uncommon in our group as in other series.\(^9\), \(^10\) When present in two patients, it was related to left heart failure and not to the size of the ventricular septal defect.

Aortography, using cineangiocardiography or still films, confirmed the clinical diagnosis of aortic regurgitation but rarely gave an accurate idea of the anatomic abnormality in the valve. Regurgitation into the right ventricle from the aorta may account for some of the right ventricular dilatation and hypertrophy. Left ventricular angiography provided useful information about the anatomy of the defect, and it is recommended that this investigation be done in all suspected cases.

In view of the frequent need to replace the aortic valve to restore competence or good function, surgery should be deferred when possible until at or after the time of skeletal maturity. However, since aortic regurgitation is progressive in these patients and deterioration may be rapid,\(^2\), \(^10\), \(^11\) careful and regular supervision is required. Surgery should be undertaken quickly in the patient with gross cardiac enlargement or severe left ventricular hypertrophy irrespective of age even in the absence of symptoms.

Ventricular septal defect with aortic regurgitation is relatively uncommon but, when associated with gross cardiomegaly or symptoms, or both, demands surgical attention. Homograft or autograft replacement of the aortic valve when repair of the valve cannot produce competency is the treatment of choice. The place of fascia lata valves has yet to be explored.

Acknowledgment

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References
