Valve replacement for rheumatic aortic incompetence in adolescents

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Weir, E K, Matisonn, R E, Mitha, A S, le Roux, B T, Rogers, N M, and Chesler, E (1978). Thorax, 33, 608–611. Valve replacement for rheumatic aortic incompetence in adolescents. The timing of valve replacement in patients with rheumatic aortic regurgitation is assessed by balancing the mortality and complications associated with the operation and the prosthetic valves against the natural history of the lesion. The time course without surgery is determined by the severity of the volume overload and the gradual deterioration of myocardial function. We wished to obtain information both on the haemodynamic recovery achieved after aortic valve replacement in young patients and also on the risks of operation in this group. Twenty patients, in whom the aortic valve was replaced at a mean age of 15 years, were reviewed. An improvement in symptoms and in the cardiothoracic ratio on the chest radiograph occurred in every case, and the voltage measurements suggestive of left ventricular hypertrophy on electrocardiogram diminished in all but two. The left ventricular end-diastolic pressure decreased in the 11 patients who were catheterised after operation. The ejection fraction improved in three patients and stayed the same in three others. While there were no operative deaths in our series the incidence of serious morbidity, in terms of myocardial damage at or after operation, was disappointingly high. Early valve replacement to preserve myocardial function is especially attractive in young patients but cannot be advised if the insertion of the prosthetic valve is associated with appreciable myocardial damage.

It is often difficult to decide when aortic valve replacement should be advised in the individual patient who has rheumatic aortic regurgitation without severe symptoms. The decision regarding operation depends on the local experience in terms of the mortality and complications from operation and also on the geographic variations in the natural history of the disease without surgery. Haemodynamic deterioration is relatively slow in the developed countries (Segal et al., 1956; Spagnuolo et al., 1971; Goldschlager et al., 1973). In less developed areas of the world there appears to be a more virulent form of rheumatic fever and a more rapid progression of rheumatic heart disease, especially in the young (Schrire, 1958; Cherian et al., 1964; Chesler et al., 1966; Selzer and Cohn, 1972). We analyse our experience of valve replacement in 20 adolescents who had long-standing rheumatic aortic regurgitation. In Europe and the USA patients under the age of 20 years rarely undergo aortic valve replacement to correct rheumatic aortic regurgitation. Unfortunately, advanced rheumatic heart disease, including aortic regurgitation, is often encountered in children in southern Africa.

Patients

Aortic valve replacement has been performed in 20 adolescents (17 male and three female) because of isolated rheumatic aortic regurgitation. The mean age at the time of operation was 15±1 years (standard error of the mean), and the range was 11 to 20 years. Twelve patients were black, seven were Indian, and one was white. The operations were performed during eight years (1968–75), and all

1Supported by a grant from the Medical Research Council of South Africa
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patients under the age of 20 who underwent aortic valve replacement to correct aortic regurgitation during this time have been included. None of the patients had appreciable mitral valve disease or a gradient across the aortic valve greater than 20 mmHg. All of them gave a history of previous rheumatic fever, but none of them had clinical evidence of continuing rheumatic activity at the time of operation. Atrial biopsies were not routinely examined for Aschoff bodies. Five patients whose effort tolerance was only moderately reduced came to cardiac catheterisation because of increasing heart size on chest radiography, or evidence of myocardial damage on the electrocardiogram. Aortic valve replacement was advised if the left ventricular end-diastolic pressure was 15 mmHg (2.0 kPa) or greater at rest and severe aortic incompetence was confirmed angiographically. Sixteen patients underwent cardiac catheterisation before operation, and eleven had a second catheterisation 25±6 months after operation. The valves used were: Bjork-Shiley, 8; Homograft, 5; Starr-Edwards, 4; Lillehei-Kaster, 2; and University of Cape Town prosthesis, 1.

Clinical data

The New York Heart Association functional classification of the patients before and after operation is given in fig 1. All but one of the patients returned to work or school after operation. Posteroanterior chest radiographs and electrocardiograms taken before and one year later were available in 19 patients. In every case there was a reduction in cardiothoracic ratio after operation. The mean cardiothoracic ratio fell from 0.61±0.01 to 0.53±0.01% (p<0.01). In all but two cases the combined voltage of the R wave in lead V₁ and the S wave in lead V₆ was also reduced one year after operation. The mean voltage fell from 76±4 to 55±4 mm (p<0.01). The effect of aortic valve replacement on these variables is shown in fig 2.

The 16 patients who underwent cardiac catheterisation before operation had a mean pulse pressure of 83±5 mmHg (11±0.7 kPa). The pulse pressure expressed as a percentage of systolic pressure was 62±2%. The mean left ventricular end-diastolic pressure was 16±1 mmHg (2.1±0.1 kPa), the cardiac index 4.7±0.2 l/min/m², and the ejection fraction measured from the left ventricular cineangiogram was 54±4%. The haemodynamic data obtained before and after operation in eleven patients are shown in the table.

There were no perioperative deaths in the 20 patients. The signs of aortic regurgitation recurred suddenly in one subject a year after operation, and he died in pulmonary oedema at another hospital. Thirteen patients are still under observation 46±7 months after operation (range 22–97 months). Two of those with Starr-Edwards valves have mild haemolytic anaemia and one of these patients has had a minor cerebral embolus. Mild
Table  Cardiac catheterisation data (11 patients)

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<thead>
<tr>
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<th>Before operation</th>
<th>After operation</th>
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<tbody>
<tr>
<td>Right atrial pressure (mmHg)</td>
<td>5 ± 1</td>
<td>5 ± 1</td>
</tr>
<tr>
<td>Pulmonary artery pressure (mmHg)</td>
<td>19 ± 2</td>
<td>19 ± 1</td>
</tr>
<tr>
<td>Left ventricular end-diastolic pressure (mmHg)</td>
<td>15 ± 1</td>
<td>11 ± 1*</td>
</tr>
<tr>
<td>Aortic systolic pressure (mmHg)</td>
<td>136 ± 6</td>
<td>119 ± 5</td>
</tr>
<tr>
<td>Pulse pressure in aorta (mmHg)</td>
<td>88 ± 7</td>
<td>38 ± 3*</td>
</tr>
<tr>
<td>Cardiac index (l/min/m²)</td>
<td>4.6 ± 0.2</td>
<td>4.8 ± 0.3</td>
</tr>
<tr>
<td>Ejection fraction % (6 patients)</td>
<td>54 ± 5</td>
<td>62 ± 4</td>
</tr>
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</table>

The data are given as the mean and standard error of the mean. * p < 0.01 (paired t test).

kPa = 1/3.5 mmHg

aortic incompetence has developed in two patients who had homografts inserted three and four years ago. Six patients have been lost to follow up, a mean period of 18±9 months after operation. Three of these six patients had electrocardiograms suggestive of subendocardial damage sustained at the time of operation, as judged by the appearance and persistence of deep symmetrical T-wave inversion, in the absence of Q waves. This occurred despite coronary perfusion in each case. None of the remaining subjects showed similar changes. A fourth member of those lost to follow-up, who had a Björk-Shiley prosthesis and who was not anticoagulated because he lived too far from a clinic to permit control, sustained a transmural anterior myocardial infarction about two months after operation, presumably from a coronary embolus. A fifth patient had left ventricular cardiomyopathy with an ejection fraction of 23% before operation. The final member of those lost to follow-up, whose aortic valve was replaced by a homograft, had normal haemodynamics on repeat catheterisation and no electrocardiographic changes. These data suggest that several of the patients lost to follow-up have died. It is difficult to obtain adequate data on patients in the rural areas who do not return to outpatient clinics.

Discussion

Left ventricular function gradually deteriorates with time in the presence of severe aortic regurgitation. The deterioration is secondary to the chronic haemodynamic load, to inadequate myocardial perfusion, and possibly to the rheumatic process in the myocardium. Initially the ventricle is enlarged because of the diastolic volume load, but the patient is asymptomatic and indices of myocardial function are normal. As the patient gets older the cardiac index is usually maintained, but the left ventricular end-diastolic pressure rises, initially on exercise and later at rest. Raised end-diastolic pressure at rest has been noted to be associated with a decrease in long-term survival after aortic valve replacement (Hirshfeld et al, 1974; Pine et al, 1976), though this is not universally accepted (Roberts et al, 1976). The incidence of early and late death after operation has also increased when the ejection fraction falls below 40% (Bolooki and Kaiser, 1976) or 50% (Fischl et al, 1977).

The progression of rheumatic heart disease, from the first bout of rheumatic fever to the onset of cardiac failure, seems to be more rapid in the less well-developed areas of the world (Cherian et al, 1964; Chesler et al, 1966). Consequently, adolescent patients with aortic regurgitation cannot look forward to 20 or 30 years of symptom-free life, which is the prospect in more developed countries (Goldschlager et al, 1973). Because of the prospect of rapid deterioration of myocardial function it is tempting to recommend relatively early aortic valve replacement in these young patients. All our patients had a normal cardiac index, and the mean was relatively high, 4.7 l/min/m², though similar to that observed by Goldschlager et al (1973) in patients under the age of 20—4.3 l/min/m². Some deterioration in myocardial function was suggested by the fact that the mean left ventricular end-diastolic pressure at rest was slightly raised at 16±1 mmHg (2.1±0.1 kPa), and five patients had an ejection fraction of less than 50%. In terms of the ratio of pulse pressure to systolic pressure, 12 of those who underwent cardiac catheterisation had moderately severe aortic regurgitation and four had severe aortic regurgitation (Goldschlager et al, 1973). The validity of this method of assessing the severity of regurgitation, however, has recently been disputed (Smith et al, 1977).

There were no operative deaths in our series, but one patient died a year after operation. In addition, the occurrence of intraoperative myocardial damage in three patients and coronary embolus in a fourth gave a disappointingly high incidence of serious morbidity. In older patients undergoing aortic valve replacement perioperative myocardial infarction, diagnosed from Q wave criteria, occurs in 10 to 12% (Rahimtoola, 1977). Even in patients without associated coronary artery disease infarcts have been recorded in 5%. Consequently the risks of operation and the complications of the prosthetic valve currently overshadow the endeavour to preserve myocardial function. Our experience supports the statement that “early valve replacement for preservation of ventricular function is not clinically indicated at present” (Rahimtoola, 1977).
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It is not surprising to find that, apart from the complications already mentioned, aortic valve replacement is associated with improved cardiac function. Figures 1 and 2 show that improvement in symptoms, reduction in the cardiothoracic ratio, and a decrease in voltage on the electrocardiogram are almost invariable, but these changes probably reflect the alleviation of a haemodynamic load rather than a recovery of myocardial function. Youth may have some advantage in that the mean reduction in cardiothoracic ratio in our series of adolescents (0·08) was double the reduction in 223 older patients after aortic valve replacement (Braun et al, 1973). A mean decrease of 0·14 has been reported in three children with an average age of 15 years (Mathews et al, 1977). It can be seen in fig 2 that all six patients whose cardiothoracic ratio was below 0·60 before operation had ratios of 0·50 or less after operation, but it does not follow that these patients will necessarily have normal left ventricular function. The left ventricular end-diastolic pressure in these six patients and the other five who were catheterised after operation fell to 15 mmHg or less, while the cardiac index remained within the normal range. Thus cardiac function was improved, though even the return of the end-diastolic pressure to a normal value does not guarantee a return to normal myocardial contractility, as measured by the velocity of circumferential fibre shortening (Gault et al, 1970). It may be reasonable to operate early in an attempt to prevent myocardial dysfunction if the operative mortality remains low and if the protection of the myocardium during operation is improved. At present, however, the myocardial damage sustained as a result of valve replacement outweighs the potential benefits of reducing gradual myocardial deterioration.

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References


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