Results of Surgical Treatment for Congenital Aortic Stenosis

By F. Henry Ellis, Jr., M.D., Patrick A. Ongley, M.B., and John W. Kirklin, M.D.

Congenital aortic stenosis is a potentially lethal malformation that is amenable to surgical treatment. Some surgeons have employed moderate hypothermia and inflow stasis, but the added time for repair provided by perfusion technics clearly recommends their use. Low hospital mortality rates generally are achieved. Data are few, however, concerning the late results, and indications for operation vary.

The experience presented herein supports the view that operation should be done before the development of severe left ventricular hypertension and its sequelae. In general, late results have been good in spite of the common occurrence of residual transvalvular systolic pressure gradients at operation after repair.

Material

Forty-seven patients with congenital aortic stenosis underwent operation at the Mayo Clinic between April 1955 and July 1, 1960 (tables 1 and 2). The three patients with supravalvular stenosis have already been reported on by McGoon and associates and will not be included in tables 3 and 4 or in the remainder of the text.

The ages of the 32 males and 12 females ranged from 10 months to 36 years. The usual symptoms exhibited by patients with aortic stenosis were noted among the 34 symptomatic patients of this group, including angina, syncope, breathlessness on exertion, and fatigue. Ten patients were without symptoms. The symptomatic patients were classified according to the New York Heart Association classification as follows: 15, class II; 18, class III; and 1, class IV.

All of the asymptomatic patients had as indications for operation one or more of the following findings: an enlarged heart, electrocardiographic evidence of advanced left ventricular hypertrophy, or in the absence of these findings, a systolic pressure gradient across the aortic valve of 50 mm. mercury or more.

All patients were operated on by an open technique with normothermic whole-body perfusion. Until late in 1958, cardiac arrest was produced by the injection of potassium citrate; later it was produced by ischemia. Direct perfusion of the coronary arteries has not been used. Patients with stenosis of the aortic valve were treated in general by incision of fused commissures or raphes. Those with a localized subaortic obstruction located just below the valve underwent transaortic excision of the lesion. Care was taken to avoid damage to the aortic leaflet of the mitral valve. Several methods were used for treatment of the diffuse muscular type of aortic obstruction, the successful one of resection through an incision in the left ventricle having been described recently.

Results

Mortality

Two of the 33 patients with valvular aortic stenosis died in the hospital (table 2). In one patient, a patent ductus arteriosus was not recognized before perfusion and the child died shortly after operation. Another patient died suddenly 12 hours postoperatively after good relief of aortic valve obstruction had been obtained. Death has not occurred among the last 29 patients operated on since August 1959.

One patient with localized subvalvular aortic stenosis died of complications due to staphylococcal infection at the site of aortotomy. Another patient with combined subvalvular aortic and pulmonary stenosis and a ventricular septal defect failed to survive. The aortic valve had been incompetent prior to operation and incompetence persisted to produce left ventricular dilatation and cardiac failure after release of the aortic clamp. Two patients with diffuse subvalvular aortic

From the Mayo Clinic and the Mayo Foundation, Rochester, Minnesota.
Before Valvotomy

Pressure measurements (11-year-old boy) obtained at operation by the withdrawal of a catheter from the aorta across the aortic valve to the left ventricle. Prior to operation the left ventricular systolic pressure exceeded 250 mm. Hg while the aortic systolic pressure was approximately 90 mm. Hg. After valvotomy, on withdrawal of the catheter in a similar manner, the systolic pressures in the ascending aorta and in the left ventricle were closely similar at 70 and 85 mm. Hg, respectively.

Figure 1

Postoperative Aortic Valve Incompetence

Only one patient with subvalvular stenosis has evidence of moderate aortic valve incompetence after operation. This was not present in the early postoperative period but had developed 1 year later. Some scarring of the aortic leaflets may have developed.

The occurrence of aortic valve incompetence after treatment for congenital valvular aortic stenosis is shown in table 3. Fifteen patients had none. Nine patients had normal pulse pressures, but audible grade-I (I-VI) or grade-II (I-VI) diastolic murmurs in the aortic area. They are considered to have mild incompetence. Five patients (18 per cent of the total group) are considered to have moderate incompetence, without cardiomegaly and with a diastolic blood pressure of about 60 mm. Hg, but with wide pulse pressure and aortic diastolic murmur. No surviving patient has severe aortic valve incompetence.

Pressures after Repair*

Pressures were measured in the left ventricle and aorta (or one of its branches) in

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*We are grateful to Dr. H. J. C. Swan for his help in analyzing the hemodynamic data.
Partial relief of valvular aortic stenosis by aortic valvotomy in 9-year-old girl. In the left panel, pressures in the left ventricle and aorta reveal a peak pressure difference of approximately 130 mm. Hg. After valvotomy a pressure gradient of approximately 55 mm. Hg persisted across the aortic valve.

Table 1

Anatomic Findings in Patients Operated on for Congenital Aortic Stenosis (April 1955 to July 1960)

<table>
<thead>
<tr>
<th>Aortic stenosis, type</th>
<th>Number of cases</th>
<th>Associated defects</th>
<th>Number of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Valvular</td>
<td>33</td>
<td>Patent ductus</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Ventricular septal defect, tiny</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Valvular pulmonary stenosis</td>
<td>1</td>
</tr>
<tr>
<td>Supravalvular</td>
<td>3</td>
<td>Infundibular pulmonary stenosis</td>
<td>1</td>
</tr>
<tr>
<td>Subvalvular</td>
<td>11</td>
<td>and ventricular septal defect</td>
<td>1</td>
</tr>
<tr>
<td>Localized</td>
<td>7</td>
<td>Infundibular pulmonary stenosis, ventricular septal defect, and severe aortic insufficiency</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Patent ductus</td>
<td>1</td>
</tr>
<tr>
<td>Diffuse</td>
<td>4</td>
<td></td>
<td>9</td>
</tr>
<tr>
<td>Total</td>
<td>47</td>
<td></td>
<td>9</td>
</tr>
</tbody>
</table>

the operating room before and after repair, and in eight cases in the late postoperative period. Since cardiac output was not measured, only limited conclusions can be drawn from these data (table 4).

The nature of the surgical procedure assured a larger valve orifice after repair in those patients with valvular stenosis. Yet in the operating room after repair, a systolic pressure gradient between left ventricle and aorta was absent or mild in only 13 of 31 patients so studied (42 per cent) (fig. 1). Eight
patients (26 per cent) had a moderate residual gradient. All of these patients, two of whom were severely handicapped prior to operation, have an excellent symptomatic result. Ten patients (32 per cent) had severe residual gradients (fig. 2) averaging 76 mm. Hg. In all but two of the 10 patients this represented a significant reduction when compared with the gradient present at operation prior to repair. The preoperative gradient averaged 116 mm. Hg in this group, and in seven of the 10 patients it exceeded 100 mm. Hg. Of the 21 patients of the other three groups, only four had gradients before operation in excess of 100 mm. Hg and the average was 81 mm. Hg. One of these had no residual gradient when studied 20 months after operation (fig. 3). Patients with severe residual gradients were usually symptomatic prior to operation, and all have been improved in this regard.

In the group with localized subaortic stenosis it is more difficult to be certain, from an anatomic standpoint, of increasing the size of the orifice subsequent to operation. One patient with a moderate residual gradient in the operating room had a severe gradient of 106

Table 2

<table>
<thead>
<tr>
<th>Aortic stenosis</th>
<th>Hospital deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type</td>
<td>Number</td>
</tr>
<tr>
<td>Valvular</td>
<td>33</td>
</tr>
<tr>
<td>Subvalvular</td>
<td>11</td>
</tr>
<tr>
<td>Localized</td>
<td>7</td>
</tr>
<tr>
<td>Diffuse</td>
<td>4</td>
</tr>
<tr>
<td>Supravalvular</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>47</td>
</tr>
</tbody>
</table>
mm. Hg when studied 4 months later and died 32 months after operation. Yet another patient with a severe residual gradient at operation had virtually none when studied 14 months later (fig. 4).

Postvalvotomy transvalvular gradients were measured in only two of the four patients with diffuse subaortic stenosis. The residual gradient was mild in one and virtually absent in the other (fig. 5). The latter patient is now asymptomatic more than a year after operation.

Eight patients have had late postoperative measurement of transaortic valve pressures. Reference to two of these patients has already been made (figs. 3 and 4). Six of the eight had valvular aortic stenosis, and late studies are of interest concerning the ultimate hemodynamic status of patients who demonstrate a residual gradient at operation after valvotomy. Five of these patients had residual gradients at operation, and the two studied less than a year later did not show further reduction in this gradient. However, the remaining three studied 20 to 33 months later all showed a significant drop in gradient as compared to their immediate postvalvotomy operative pressures.

### Table 3

**Aortic Valve incompetence Following Repair of Valvular Aortic Stenosis**

<table>
<thead>
<tr>
<th>Degree</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>15</td>
</tr>
<tr>
<td>Mild</td>
<td>9</td>
</tr>
<tr>
<td>Moderate</td>
<td>5</td>
</tr>
<tr>
<td>Severe</td>
<td>0</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>29</strong></td>
</tr>
</tbody>
</table>

*Surviving patients only.*
Before Operation

\textbf{AORTA (mm. Hg)}

\begin{tabular}{c|c|c}

\hline

\textbf{Before Operation} & 100 & 50 \\

\hline

\textbf{LEFT VENTRICLE (mm. Hg)} & 200 & 100 \\

\hline

\end{tabular}

\textbf{Base Line}

\begin{tabular}{c|c|c|c}

\textbf{After Valvotomy} & 100 & 50 & 0 \\

\hline

\textbf{1 sec.}

\end{tabular}

\textbf{Figure 5}

Arterial and left ventricular pressures at operation in a 17-year-old boy with diffuse subvalvular stenosis. In the left panel, pressures obtained before subvalvular resection indicated a pressure gradient of approximately 90 mm. Hg. After resection the pressure gradient was virtually eliminated.

\textbf{Table 4}

\textit{Residual Systolic Pressure Gradient (at Surgery) after Operation for Congenital Aortic Stenosis}

\begin{tabular}{|c|c|c|c|}

\hline

\textbf{Gradient (mm. Hg)} & \textbf{Valvular} & \textbf{Localized} & \textbf{Diffuse} \\

\hline

\textbf{Absent (< 5)} & 4 & 1 & 1 \\

\textbf{Mild (6 to 25)} & 9 & 1 & 1 \\

\textbf{Moderate (26 to 50)} & 8 & 2 & 2 \\

\textbf{Severe (> 50)} & 10 & 2 & 2 \\

\hline

\textbf{Total} & 31* & 6† & 2* \\

\hline

*No pressure measurements in two additional cases. \\
†No pressure measurements in one additional case.

\textbf{Clinical Results}

Patients were considered to have a good result when late follow-up disclosed no symptoms, absent or mild aortic valve incompetence, and regression of the electrocardiographic evidence of left ventricular overload. Patients were considered to have a fair result when these criteria were present, except that there was moderate aortic valve incompetence without progression. Unimproved patients were those with symptoms (table 5).

Two patients with valvular stenosis died after dismissal from the hospital. One patient had a Pseudomonas infection. The other had severe left ventricular systolic hypertension (300 mm. Hg) prior to repair and only a mild residual gradient at operation after repair. There was no aortic valve incompetence. He died abruptly while playing 3 months after operation, having been entirely asymptomatic up to that moment. One patient with localized subaortic stenosis died as indicated 32 months after operation.

Of the 31 patients surviving valvotomy, 90 per cent had fair or good results; 80 per cent of the five patients surviving resection of localized subaortic stenosis had fair or good results. The one patient having a successful definitive surgical procedure for diffuse subvalvular obstruction had a good result.
Roentgenograms

Cardiac enlargement was noted on the preoperative thoracic roentgenograms of 22 of the 35 surviving patients. Follow-up roentgenographic studies were available for review in 14 of these patients. In five there had been a noticeable decrease in the size of the cardiac silhouette (fig. 6), although two were classified as having only a fair result because of moderate aortic insufficiency. Both patients whose hearts seemed to have enlarged slightly had an excellent clinical result as did four of the seven patients whose hearts remained the same size. One of the remaining three patients whose roentgenograms showed no change died later, one was unimproved, and one had a fair result.

Electrocardiograms

Electrocardiograms made before operation were grossly abnormal for all patients. The findings were those of left ventricular overload with S-T depressions and T-wave inver-

Figure 6

Thoracic roentgenograms of two patients with congenital valvular aortic stenosis. a. Preoperative ray of 10-month-old boy; b. 16 months after operation; c. Preoperative ray of 2-year-old girl; d. 9½ months after operation.
sions in leads reflecting left ventricular voltage. One patient who also had pulmonary stenosis showed combined right and left ventricular overload, and a second patient showed right bundle-branch block.

Late postoperative electrocardiograms made from 6 months to 2 years after operation are available on 22 patients. Eighteen of these patients had valvular stenosis and all but two showed definite evidence of improvement of the abnormal S-T and T-wave changes (fig. 7). One of these two patients had a good clinical result and one had a fair result. No postoperative tracing is available on the unimproved patient with valvular stenosis. Improvement in the appearance of the electrocardiogram may require a considerable lapse of time as seen in figure 8.

Late follow-up electrocardiographic tracings were available in four patients with subaortic stenosis. No improvement was seen in the tracing of one patient with localized subaortic stenosis who died 14 months after operation. Definite improvement was noted in two other patients with localized subaortic stenosis, whereas the patient with diffuse subvalvular stenosis operated on through a left ventriculotomy shows left bundle-branch block a year after operation (fig. 9).

**Discussion**

With the technical problems of repair now well understood, hospital mortality rates can be expected to remain low after surgical treatment for congenital aortic stenosis of all types. In general, late results are good. A few points bear additional discussion.

A moderate or severe residual systolic pressure gradient between left ventricle and aorta existed during operation after valvotomy in 58 per cent of this group. Although in most instances this was a significant improvement over the gradient present prior to valvotomy, its presence is disturbing. Whether the valve

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**Table 5**

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Good</th>
<th>Fair</th>
<th>Unimproved</th>
<th>Death</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Valvular stenosis</td>
<td>21</td>
<td>7</td>
<td>1</td>
<td>2</td>
<td>31</td>
</tr>
<tr>
<td>Subvalvular</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Localized</td>
<td>3</td>
<td>1</td>
<td>1†</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Diffuse‡</td>
<td>1</td>
<td></td>
<td></td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>

*Follow-up period 6 to 39 months; average 18.2 months.
†Residual severe gradient.
‡One patient who had exploration only is not included.
or the subvalvular area or both are responsible is not known. Although we have made as wide a valvotomy as possible without producing incompetence, the stiffness and abnormal configuration found in most of these valves may account in part for the frequency of residual gradients. The regression of the gradient in a few patients during the months after operation suggests resolution of a secondary subvalvular obstruction. All three patients with valvular stenosis who had residual gradients at operation and who were studied more than a year later showed regression of the transvalvular gradient. Of interest is the fact that patients with unusually high pressure gradients prior to operation tended to have high residual gradients immediately after repair even though an excellent clinical result was usually achieved.

When proper care is exercised while incising rudimentary commissures and raphe, significant aortic valve incompetence can now be avoided. The development of moderate aortic valve incompetence in the first year after resection of localized subvalvular obstruction in the patient suggests scarring of the previously normal valve leaflets. This emphasizes the need for a gentle surgical technic when working on or through valve leaflets.

On the basis of these results it seems wise to advise operation when significant aortic stenosis is found to exist, without awaiting...
the development of severe left ventricular hypertension and its sequelae.

Summary
Open operation by means of extracorporeal circulation has been used on 47 patients with congenital aortic stenosis at the Mayo Clinic between April 1955 and July 1, 1960; 33 patients had valvular stenosis, 11 had subvalvular stenosis, and three had supravalvular stenosis. The over-all operative mortality rate was 13 per cent; 6 per cent of patients with valvular aortic stenosis died in the hospital.

Severe aortic insufficiency did not develop in any patient operated on for valvular aortic stenosis although a diastolic murmur was present after operation in some. Rarely, however, was the transvalvular gradient completely eliminated by operation although 90 per cent of the surviving patients were either asymptomatic or had definite clinical improvement. Late follow-up hemodynamic studies suggest that a persistent transvalvular gradient may regress in time.

Relief of obstruction caused by subvalvular variety of aortic stenosis is difficult whether stenosis is localized or diffuse. Even in this group, however, it has been possible to achieve good results with careful attention to certain technical details.

Patients with congenital aortic stenosis should be operated on before the development of the sequelae of severe left ventricular hypertension.

References

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