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 surgeons1,2 have employed moderate hypothermia and inflow stasis, but the added time for repair provided by perfusion technics clearly recommends their use.2-4 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 associates5 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 technic with normothermic whole-body perfusion.6 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.7 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.8

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...
Before Valvotomy

AORTA (mm. Hg)

At Valve

Just Below Valve

Low L.V.

AORTA (mm. Hg)

Ascending Aorta

Infundibulum

Low L.V.

After Valvotomy

Figure 1

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.

Stenosis died after operation. In one, the obstruction was not recognized at operation and the patient died. An attempt to relieve the obstruction in another case by operating across the ventricular septum was a failure because of the development of heart block.

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

*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></td>
<td></td>
</tr>
<tr>
<td>Subvalvular</td>
<td>11</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Localized</td>
<td>7</td>
<td>Infundibular pulmonary stenosis and ventricular septal defect</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td></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></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
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AORTIC COMMISSUROTOMY

Before

After

20 Months Later

AORTA (mm. Hg)

LEFT VENTRICLE (mm. Hg)

Figure 3

Arterial and left ventricular pressures during and after operation for valvular aortic stenosis (1½-year-old boy). The pre-valvotomy transvalvular gradient of 129 mm. Hg was reduced to 25 mm. at the operation (middle and left panels). Twenty months later there was virtually no transvalvular gradient (right panel).

Table 2

Hospital Mortality Rate in Patients Operated on for Congenital Aortic Stenosis (April 1955 to July 1960)

<table>
<thead>
<tr>
<th>Type</th>
<th>Number</th>
<th>Number</th>
<th>Per cent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Valvular</td>
<td>33</td>
<td>2</td>
<td>6.1</td>
</tr>
<tr>
<td>Subvalvular</td>
<td>11</td>
<td>4</td>
<td>36.4</td>
</tr>
<tr>
<td>Localized</td>
<td>7</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Diffuse</td>
<td>4</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Supravalvular</td>
<td>3</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>47</td>
<td>6</td>
<td>12.8</td>
</tr>
</tbody>
</table>

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
Arterial and left ventricular pressures before, during and after operation for localized subvalvular aortic stenosis (21-year-old man). The pressure gradient before valvotomy is of the order of 160 mm. Hg. At the time of operation, partial relief was obtained but a residual systolic gradient of 80 mm. Hg persisted. Fourteen months after operation (right panel) the radial arterial systolic pressure is within 20 mm. of mercury of that in the left ventricle.

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

<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

\[ \begin{align*}
\text{AORTA (mm. Hg)} & \quad 200 \quad - \\
\text{LEFT VENTRICLE (mm. Hg)} & \quad 0 \\
\end{align*} \]

After Valvotomy

\[ \begin{align*}
\text{AORTA (mm. Hg)} & \quad 200 \quad - \\
\text{LEFT VENTRICLE (mm. Hg)} & \quad 0 \\
\end{align*} \]

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.

Table 4

*Residual Systolic Pressure Gradient (at Surgery) after Operation for Congenital Aortic Stenosis*

<table>
<thead>
<tr>
<th>Gradient (mm. Hg)</th>
<th>Valvular</th>
<th>Localized</th>
<th>Diffuse</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absent (&lt; 5)</td>
<td>4</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Mild (6 to 25)</td>
<td>9</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Moderate (26 to 50)</td>
<td>8</td>
<td>2</td>
<td>-</td>
</tr>
<tr>
<td>Severe (&gt; 50)</td>
<td>10</td>
<td>2</td>
<td>-</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>31</strong></td>
<td><strong>6</strong></td>
<td><strong>2</strong></td>
</tr>
</tbody>
</table>

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

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.
CONGENITAL AORTIC STENOSIS

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 7

Electrocardiograms of a 5-year-old boy with valvular aortic stenosis. The transvalvular gradient was eliminated at operation and an excellent clinical result ensued. a, Preoperative tracing. b, Tracing made 1 year after operation. Note improvement in abnormal ST and T-wave changes.

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

| Table 5 |
| Late Results in Patients Treated Surgically for Congenital Aortic Stenosis* |
| --- | --- | --- | --- | --- |
| Lesion | Good | Fair | Unimproved | Death | Total |
| Valvular stenosis | 21 | 7 | 1 | 2 | 31 |
| Subvalvular Localized | 3 | 1 | 1 | 5 | |
| Diffuse† | 1 | | | 1 | |
| *Follow-up period 6 to 39 months; average 18.2 months. |
| †Residual severe gradient. |
| †One patient who had exploration only is not included. |
Figure 8
Electrocardiograms of 14-year-old girl operated on for valvular aortic stenosis on November 13, 1957. A residual gradient was present after operation. There was no improvement in the tracing taken 1 year after operation whereas the tracing made 33 months after operation showed considerable improvement in the T waves. Clinically the patient is classified as having a fair result.

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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