Aortic Stenosis Surgery in Infancy

By John F. Keane, M.D., William F. Bernhard, M.D.,
and Alexander S. Nadas, M.D.

SUMMARY

During the past 14 years, 28 infants (23 males) underwent valvotomy for severe aortic stenosis at the Children's Hospital Medical Center in Boston. The median age was two months and the oldest patient was six months old. Congestive heart failure was present in all but two babies. Preoperatively, the electrocardiogram was abnormal in all, with left ventricular hypertrophy and a strain pattern being present in 19. At cardiac catheterization, the peak systolic ejection gradient (PSEG) ranged from 35-130 mm Hg (median 90 mm Hg). Associated cardiac lesions were present in 39%.

Twenty-four infants underwent valvotomy with inflow occlusion. Four patients were operated upon using cardiopulmonary bypass. There were eight early and two late deaths.

The 18 survivors have been followed from six months to 11 years (median five years). Only four are symptomatic. Mild aortic regurgitation developed postoperatively in six patients, moderate in one and severe enough to require valve replacement in another one. The electrocardiogram improved postoperatively in 15 patients, but became entirely normal only in one. Five children required a repeat valvotomy for residual stenosis 1-10 years after the original procedure (median four years). At this second operation, the majority of the valves were flexible and noncalcified, and valvotomy was accomplished without difficulty. One child who underwent valve replacement for aortic regurgitation at age two years is well seven years later.

The incidence of Aortic Stenosis among children with congenital heart disease ranges from 3 to 5.5%. From those with valvar stenosis, the commonest variety, there occurs a group with critical obstruction who usually present in early infancy with congestive heart failure. In these babies, prolonged medical treatment is almost invariably ineffective; thus, surgical intervention is mandatory for survival.

It is the purpose of this report to present the surgical results and subsequent clinical course of 28 infants with critical aortic stenosis who underwent valvotomy at the Children's Hospital Medical Center between 1959 and 1974. Critical obstruction in these babies is defined as valvar aortic stenosis associated with marked cardiomegaly with either congestive heart failure or left ventricular hypertrophy (LVH) with a strain pattern or both. Seventeen of these patients were reviewed in 1967, including our initial attempts to provide surgical relief.

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Preoperative Evaluation

The 28 infants (23 males) ranged in age from four days to six months (median two months) at operation. Congestive heart failure was present in all but two, both of whom had a large heart on X-ray and LVH with strain in the electrocardiogram (ECG). Systolic ejection murmurs, grade II-IV/V1 in intensity, were present in 26 babies. No murmur was initially detected in the other two infants but appeared following initiation of anticongestive measures. Systolic thrills were described in only six and ejection clicks in 14 patients, these being considerably less frequent than in older children with significant valvar stenosis. Left ventricular hypertrophy was diagnosed 1) electrocardiographically using the ninety-fifth percentile values of Cassels and Ziegler and 2) vectorcardiographically when the left maximum spatial voltage was increased. The ECG was abnormal in all and details are outlined in table 1. LVH with a strain pattern was present in 19 infants and right ventricular hypertrophy alone was noted in three infants, two of whom had hypoplastic left ventricles (fig. 1).

Cardiac catheterization was performed in all but two infants who were considered too critically ill to undergo this procedure. The left ventricle was entered in 17 of the 26 patients, via the mitral valve in 13, by the retrograde approach in three and by transthoracic puncture in the remaining patient. The peak systolic ejection gradient (PSEG) across the aortic valve ranged from 35-130 mm Hg (median 90 mm Hg); the only gradients less than 70 mm Hg were 53, 40 and 35 mm Hg and were associated with left ventricular end-diastolic pressures of 20, 24 and 22 mm Hg, respectively. The systemic cardiac index, calculated in 21 babies (using an assumed oxygen consumption in 18 of these) ranged from 2.1-5.5 L/min/m² (median 3.8). The aortic valve index measured in nine infants ranged from 0.2-0.6 cm²/m² (median 0.3). Left-to-right shunting at the arial level was present in seven infants (median age 17 days) whose left ventricular end-diastolic pressures ranged from 12 to 28 mm Hg (median 20). The pulmonary-systemic
AORTIC STENOSIS SURGERY IN INFANCY

Table 1

<table>
<thead>
<tr>
<th>Electrocardiographic Findings</th>
<th>Preoperative</th>
<th>Postoperative</th>
</tr>
</thead>
<tbody>
<tr>
<td>LVH With strain pattern</td>
<td>19</td>
<td>13</td>
</tr>
<tr>
<td>LVH Without strain pattern</td>
<td>0</td>
<td>11</td>
</tr>
<tr>
<td>Strain pattern alone</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>RVH alone</td>
<td>3 (2 with hypoplastic LV)</td>
<td>0</td>
</tr>
<tr>
<td>Decreased anterior forces</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Myocardial infarction</td>
<td>1 (inferior)</td>
<td>0</td>
</tr>
<tr>
<td>RBBB</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Normal</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>28</td>
<td>18</td>
</tr>
</tbody>
</table>

Abbreviations: LVH = left ventricular hypertrophy; RVH = right ventricular hypertrophy; RBBB = right bundle branch block.

Surgical Technique

Twenty-three infants underwent aortic valvotomy with 2-3 minutes of total inflow occlusion at temperatures of 34-37°C. One patient had a valvotomy during inflow occlusion employing moderate hypothermia (30°C). Four patients were operated upon using cardiopulmonary bypass. Morphologically, the valves were rather primitive in nature with thickened and verrucous cusps and eccentric orifices. The valves were bicuspid in 11 patients and unicuspid in four, while the number of cusps was uncertain in 13. In the majority, two short incisions (anteriorly and posteriorly) were used to open the stenotic valve. In those with unicuspid valves, only one incision was made in the anterior commissure. In one of the two infants with hypoplastic left ventricles, a valvotomy via the left ventricular apex was attempted, without success, because of uncontrollable ventricular fibrillation.

Surgical Results

There were eight operative fatalities including the first four patients in the series. The two babies with hypoplastic left ventricles (table 4) also died and represent the only two deaths among the last 18 patients. Two of the 20 survivors of surgery died two and six months postoperatively, both with endocardial fibroelastosis.

Postoperative Status

The 18 survivors have been followed from 6 months

flow ratio in these babies ranged from 1.2 to greater than 4.0:1 (median 1.9:1). Pulmonary artery hypertension of varying degrees was noted in 16 infants. Left ventricular volume data were determined in seven infants (table 2). The two infants with very small end-diastolic volumes (EDV) had hypoplastic left hearts and did not survive (fig. 1). The older babies had a markedly increased left ventricular mass (LVM). Associated cardiac malformations were identified in 11 patients (39%, table 3).

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**End-diastolic lateral frames from cineangiograms of two patients with hypoplastic left ventricles and aortic valve annuli.**

EDV = 9 and 7 cm³/m² respectively (normal value 42 ± 10). Severe mitral stenosis also present in patient on right.

Circulation, Volume 52, December 1975
Ventricular septal defect
Mitral regurgitation
Patent ductus arteriosus
Mitral stenosis
2 days* 7 0.50 48
4 days 33 0.72 53
1 month 38 0.51 52
5 weeks* 9 0.62 27
6 weeks 40 0.37 131
3 months 70 0.64 213
6 months 65 0.51 186

* Died.

Abbreviations: NL = normal values; EDV = end-diastolic volume; EF = ejection fraction; LVM = left ventricular mass.

to 11 years (median 5 years). All but four children are asymptomatic (two with fatigue, one with exertional dyspnea, one with angina). Aortic regurgitation developed postoperatively in eight patients, all of whom are presently well. Six of these patients are considered to have mild regurgitation, based on the absence of symptoms, a grade II/VI diastolic murmur, pulse pressure 50 mm Hg or less (median 34) and mild cardiomegaly at most. Another asymptomatic patient is deemed to have moderate regurgitation based on a grade III/VI diastolic murmur, pulse pressure 55 mm Hg, moderate cardiac enlargement, and angiographic delineation. The remaining patient had severe regurgitation demonstrated angiographically. In addition this patient had congestive heart failure, a grade III/VI diastolic murmur, pulse pressure 62 mm Hg, marked cardiomegaly, pulmonary artery hypertension and an elevated left ventricular end-diastolic pressure. The ECG remained abnormal in all but one patient (table 1); improvement occurred in 15, the tracing remained unchanged in two and deteriorated in the remaining patient.

Of the 18 survivors, 12 who have not undergone a second operation have been followed for 6 months to

Table 2

Preoperative Left Ventricular Angiographic Data

<table>
<thead>
<tr>
<th>Age</th>
<th>EDV (cm³/m²) (NL 42 ± 10)</th>
<th>EF (NL 0.68 ± 0.05)</th>
<th>LVM (gm/m²) (NL 88 ± 12)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2 days*</td>
<td>7</td>
<td>0.50</td>
<td>48</td>
</tr>
<tr>
<td>4 days</td>
<td>33</td>
<td>0.72</td>
<td>53</td>
</tr>
<tr>
<td>1 month</td>
<td>38</td>
<td>0.51</td>
<td>52</td>
</tr>
<tr>
<td>5 weeks*</td>
<td>9</td>
<td>0.62</td>
<td>27</td>
</tr>
<tr>
<td>6 weeks</td>
<td>40</td>
<td>0.37</td>
<td>131</td>
</tr>
<tr>
<td>3 months</td>
<td>70</td>
<td>0.64</td>
<td>213</td>
</tr>
<tr>
<td>6 months</td>
<td>65</td>
<td>0.51</td>
<td>186</td>
</tr>
</tbody>
</table>

Table 3

Associated Cardiac Malformations

<table>
<thead>
<tr>
<th>Malformation</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Endocardial fibroelastosis</td>
<td>5</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>4</td>
</tr>
<tr>
<td>Hypoplastic left ventricle</td>
<td>2</td>
</tr>
<tr>
<td>Mitral stenosis</td>
<td>2</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>2</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>2</td>
</tr>
<tr>
<td>Discrete aortic coarctation</td>
<td>2</td>
</tr>
<tr>
<td>Mitral regurgitation</td>
<td>1</td>
</tr>
<tr>
<td>Endocardial and subendocardial fibris and calcification</td>
<td>1</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>1</td>
</tr>
<tr>
<td>Totals</td>
<td>22</td>
</tr>
<tr>
<td>No of anomalies</td>
<td>11 (39%)</td>
</tr>
</tbody>
</table>

Figure 2

Peak systolic ejection gradients (PSEG) in six patients before and after initial aortic valvotomy, the intervals between cardiac catheterizations ranging from 2 to 4 years (median 4 yr).

10 years (median 4 years). Cardiac catheterization has been performed in five of these, and the residual PSEG ranged from 17–60 mm Hg (median 35 mm Hg). Subvalvar stenosis was present in only one patient (PSEG = 20 mm Hg). A ventricular septal defect had closed spontaneously in another while a left-to-right shunt at the atrial level had disappeared in a further patient.

Six patients had a second operation 1–10 years (median 4 years) following the initial surgical procedure. At cardiac catheterization, prior to the second procedure, the PSEG ranged from 60–140 mm Hg (median 85 mm Hg). A repeat aortic valvotomy was performed without difficulty in five patients, and the valve was noted to be flexible and noncalcified in the majority.* Severe infundibular pulmonary stenosis was also relieved by resection in one of these children. Replacement of the aortic valve was necessary, because of severe regurgitation, in the remaining patient at age two years, and seven years later he is alive and doing well. A four-month-old brother of this patient died with aortic stenosis.

Among the 18 survivors, comparison of the pre- and postoperative PSEG was possible in six patients and details are outlined in figure 2. Left ventricular angiographic analysis was possible in six patients at cardiac catheterization 1–7 years following the original operation (table 5). The left ventricular EDV was normal in three and decreased in three children. The ejection fraction was increased in all but one, and the LVM was elevated in four patients. While endocardial fibroelastosis was diagnosed only at autopsy or surgery in our patients, the left ventricle at angiography in

*One of the five repeat aortic valvotomies was performed by Dr. F. Spencer at the University Hospital, New York City and another by Dr. H. Low at the Hartford Hospital, Hartford, Connecticut.
one of the survivors suggested this abnormality (fig. 3). This patient has a brother, now aged one year, who has a catheterization diagnosis of endocardial fibroelastosis.

Discussion

Severe aortic stenosis in infancy is an unusual form of congenital left ventricular outflow tract obstruction. Among patients who have undergone surgery for congenital aortic stenosis, the incidence of infants ranged from 4 to 13%. In contrast to older children with severe stenosis, the site of obstruction in this age group is valvar almost without exception. Congestive heart failure is present in the majority, particularly during the early months of life. Associated cardiac abnormalities (table 3) are more common in these babies than in older patients. In Hasted’s patients, some of whom we would have classified as having the hypoplastic left heart syndrome, the incidence was at least 60% among catheterized infants and 100% in those who died. The incidence of additional cardiac malformations in our series was 39%.

The treatment of severe aortic stenosis in infancy is surgical valvotomy, particularly when the lesion is accompanied by congestive heart failure. Prolonged medical management is unsuccessful. Valvotomy employing standard cardiopulmonary bypass alone has been utilized in some centers while a combination of short inflow occlusion with partial bypass has been employed by others. At the Children's Hospital Medical Center, the majority of babies have been operated upon using a simple inflow occlusion technique.

Surgical mortality in this group of infants is generally high. In recent years, however, survival rates have varied. In one series of seven babies operated upon under one month of age, there were five early and one late (six months postoperative) deaths, and it was concluded that dramatic improvement following surgery is uncommon and that long term results are poor. In contrast, of our seven patients less than one month of age at operation, there were five survivors. The oldest one of these is now 11 years old. In another recent series of seven infants,
only three of whom were less than four months of age, there were no deaths.\textsuperscript{18} The median age of our 28 patients at operation was two months, and our youngest survivor, now thriving at nine months, was four days old at the time of operation.

Our experience with hypoplastic left ventricles is similar to that of Lakier\textsuperscript{18} in that both of our infants with EDVs of 7 and 9 cm\(^3\)/m\(^2\) died following surgery, although additional cardiac lesions were also present in our patients (table 4). It would thus appear that the presence of a small left ventricle is of ominous significance. Valvotomy in these patients may not be beneficial but should probably be attempted anyway. Some of our patients with an adequate sized ventricle had a decreased ejection fraction, indicating left ventricular failure. Regardless of the presence or absence of endocardial fibroelastosis, relief of the obstruction will decrease the afterload and valvotomy should be performed in such patients irrespective of the ejection fractions. Among our six patients with postoperative angiographic data, the ejection fraction was increased in all but one, the EDV was decreased in three and the LVM was increased in four of the patients, these observations being similar to those of Graham.\textsuperscript{17}

It is important to note that residual significant aortic regurgitation is unusual, occurring in only two of our 18 survivors, one of whom required valve replacement. Our patients included three babies with unicommissural valves who have competent valves following operation, a finding not anticipated by some.\textsuperscript{18}

Although aortic valvotomy is a palliative operation, dramatic improvement occurred postoperatively initially in all of our survivors. Twelve patients followed for six months to ten years (median four years) have not yet required a further operative procedure. The median PSEG in the five patients recatheterized in this group was 35 mm Hg. In those five children who required a second valvotomy for residual stenosis 1–10 years postoperatively (median four years), valve cusps were described as flexible and noncalcified in the majority. Repeat valvotomy was accomplished without mortality, and only one patient has aortic regurgitation and this is mild in degree. This suggests a maturing of the original embryonal structure, which perhaps contributed to repeat successful valvotomy.

Our current management of the infant with aortic stenosis is as follows. The asymptomatic baby without congestive heart failure, cardiomegaly or a left ventricular strain pattern is followed medically and undergoes catheterization only when one of these complications appears. Marked cardiomegaly associated with either congestive heart failure or LVH with strain pattern or both, signify critical obstruction and are the indicators for valvotomy, as prolonged medical therapy in these babies is invariably ineffective.\textsuperscript{4, 5} Digitalis and diuretics are commenced and cardiac catheterization is performed invariably within days and occasionally within hours, to confirm the presence of valvar aortic stenosis and to identify any other associated anomalies. Measurement of the PSEG is attempted in all. While a large gradient supports the
diagnosis of critical obstruction, the finding of a small gradient in our patients, considered to be critical on clinical grounds, was associated with an elevated left ventricular end-diastolic pressure, indicating myocardial failure. Angiography in the aortic supravalvar position may be used to demonstrate the valvar stenosis in those in whom the left ventricle is not entered. An associated lesion of major significance is rare in these infants except in those with hypoplastic left ventricles. Valvotomy is performed in all these babies, currently using either the inflow occlusion technique or cardiopulmonary bypass.

References

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