Reoperation in Congenital Aortic Stenosis
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ABSTRACT Over a 22-year period, 81 patients underwent initial operations for critical aortic stenosis at our institution. Their ages ranged from 3 days to 20 years (mean, 7.3 ± 5.9 years). Fourteen (17%) were infants less than 1 year old. Three children died perioperatively (3.7%). We have followed the survivors and 3 children who underwent initial operations elsewhere for a mean of 9.0 ± 6.8 years (range, 2 to 23 years). To the present, 27 patients have undergone one reoperation (24 of our initial survivors) at a mean interval of 7.3 years, with 2 perioperative deaths. Ten of these patients have required a second reoperation at a mean interval of 3.7 years, with 2 deaths perioperatively. There were 3 late cardiac deaths after the initial procedure and 1 after a third operation. Actuarial reoperation-free survival is 56.7% at 10 years. While overall survival is 88.6% at 10 years, we find a significantly poorer survival among those patients with valvular stenosis compared with those with subvalvular lesions (p = 0.03). We believe that for children with all levels of aortic stenosis, good functional results and survival can best be obtained by follow-up, recatheterization, and reoperation.

The palliative nature of operative attempts to treat congenital aortic stenosis in childhood was well documented in 1978 by Salomon and colleagues [1] at Stanford. They reported an actuarial reoperative rate of 24% 10 years following the initial procedure. While it has been estimated that all children with valvular stenosis treated by valvotomy will eventually require reoperation [2, 3], others note that discrete subvalvular stenosis should be cured by resection [4]. In view of these data, we examined our experience with all levels of aortic stenosis, with particular reference to the incidence of reoperation, the survival thereafter, and the functional results.

Material and Methods
All patients operated on for critical aortic stenosis from 1961 to 1983 were included in this retrospective analysis of our experience. No patient was excluded because of associated problems. Eighty-one patients had the initial operation at the Oklahoma Teaching Hospitals, and an additional 3 underwent the first valvotomy at other institutions. The latter 3 are included in our reoperative mortality and follow-up evaluations. The chart of each patient was examined for preoperative gradient, operative procedure, intraoperative gradient, postoperative complications, and outcome.

All operations were performed by one of four staff surgeons. Valvotomies were done by carefully opening fused commissures so as to avoid insufficiency. Mild residual stenosis was preferred over aortic resection. Valve replacement was accomplished with the valve of the surgeon's choice. Bioprosthetic valves have been avoided since 1980. Deaths were categorized as perioperative if they occurred within thirty days of the procedure. Subsequent deaths were classified as late, related or late, unrelated.

Current follow-up was obtained over a 4-month period in 1984, by telephone contact with the patient, parent, or cardiologist. New York Heart Association (NYHA) Functional Class was assessed for patients older than five years. The presence of recurrent aortic stenosis and insufficiency and the decision for recatheterization were determined by either of two of the investigators (J. D. R. or W. M. T.). Recatheterization was dictated primarily on clinical grounds, such as a change in murmur during rapid physical growth. Electrocardiographic changes indicating an increase in hypertrophy or strain and more recently, echocardiographic evidence of ventricular chamber size have also been used to indicate the need for recatheterization. Reoperation was based on valve gradients greater than 65 mm Hg or increased symptoms. All reoperations up to October, 1984, are included.

Survival was assessed using the Kaplan-Meier actuarial method. Simple survival was assessed exclusive of unrelated late deaths. Reoperation-free survival curves were similarly obtained. We made comparisons of these curves based on the level of initial obstruction as well. Means are expressed as ± 1 standard deviation.

Initial Operation
Over the 22-year period of this review, 81 patients underwent an initial operation for aortic stenosis at the Oklahoma Teaching Hospitals. They had gradients ranging from 50 to 180 mm Hg (mean, 84.7 mm Hg). They ranged in age from 3 days to 20 years (mean, 7.3 ± 5.9 years); 14 (17%) were infants (age of less than 1 year) (Fig 1). At operation the level of obstruction was valvular in 52 patients (64%), subvalvular in 21 (26%), supravalvular in 3 (4%), and combined in 5 (6%) (1, valvular plus...
Fig 1. Age distribution of patient population at initial operation.

supravalvular and 4, valvular plus subvalvular). Of the 21 pure supravalvular stenoses, 16 were described as discrete membranes and 5, as more thickened "collars" or diffuse narrowing.

Operation consisted of valvotomy in all but 3 of our patients with valvular stenosis. Prior to 1978, bioprosthetic valve replacement was accomplished in 2 patients aged 17 and 20 years. A Konno procedure [5] with a 19-mm bileaflet tilting-disc prosthesis was utilized in a 2½-year-old child with a gradient of 100 mm Hg, a small aortic root, and a unicusps valve. It was thought that the valve could not be opened without unacceptable insufficiency. All of the patients with supravalvular stenosis had resection. In 4 patients with more diffuse than discrete narrowing, a septal splitting maneuver was accomplished to dilate the outflow tract. The 3 patients with supravalvar obstruction underwent patch aortoplasty. The patient with both supravalvular and valvular stenosis was treated with valvotomy and patch aortoplasty. Three patients with subvalvar and valvular lesions had resection of the former and commissurotomy of the latter. The other such patient (18 years old) had resection and annuloplasty with prosthetic valve replacement.

Three of these patients died perioperatively; 2 were infants (4 weeks and 3 months old) and 1 was a 9-year-old child who died as a consequence of cerebral emboli. All had valvular aortic stenosis. Excluding 6 hospital survivors lost to follow-up after a mean follow-up of 16 months (2 to 24 months), 72 of our patients were available for follow-up to a possible reoperation (Fig 2). They have been followed for a mean of 9.0 ± 6.8 years.

There were 4 late deaths after initial operation, 2 in children who underwent operation as infants and all in patients who had valvular stenosis. A hydrocephalic child died 2 months postoperatively of a cerebral hemorrhage. A 2½-year-old child with multiple other anomalies died suddenly at home 2 years after operation, and a 22-year-old man who had had valve replacement 5 years earlier, complicated by an atrioventricular block and pacemaker insertion, died suddenly. The fourth late, unrelated death occurred in a 14-year-old boy who was killed in an automobile accident 10 years after operation.

Results

Second Operation

Twenty-four (33.3%) of the hospital survivors whom we have been able to follow for more than 2 years have required a second operation, as have 3 children operated on elsewhere initially. The mean interval to this reoperation was 7.3 years. Recurrent aortic stenosis was the sole indication in 21 of these 27 patients. Valve gradients ranged from 65 to 250 mm Hg (mean, 104 mm Hg). Six patients also had some degree of preoperative aortic insufficiency. Two of them had been treated medically for bacterial endocarditis since the first operation (one a valvotomy and one a bioprosthesis replacement), and they had the only clinically significant aortic regurgitation.

The second operation was a repeat of the first procedure in 5 children who had previously undergone valvotomy and in 6 who had had a subvalvular resection. Similarly, recurrent supravalvular stenosis was treated by repatching the aorta, and the child with recurrent subvalvular and valvular obstruction simply had another valvotomy with resection. Some form of valve replacement was accomplished in the other 14 patients, including 5 of the 6 children who had a degree of aortic insufficiency. The ages at valve replacement ranged from 3 to 25 years. Annuloplasty was required in a 12-year-old patient to place a 19-mm porcine valve and in an 8-year-old child to place a 19-mm tilting-disc valve. A Konno procedure was performed in a 3-year-old child to
accommodate a 19-mm bileaflet tilting-disc prosthesis.

Table 1 lists the incidence of reoperation, interval to reoperation, operation required, and reoperative mortality by initial level of obstruction. Three patients with subvalvular obstruction had no evidence of valvular abnormality at their first operation, but 7, 12, and 17 years later, they had sufficient gross valvular pathology to require valve replacement.

There were several complications following reoperation. One child had excessive mediastinal hemorrhage, which necessitated the reopening of the sternotomy. There were 2 instances of postpericardiotomy syndrome requiring salicylate therapy. Slight aortic insufficiency was noted prior to the discharge of a 6-year-old child who had undergone valvotomy. Atrioventricular block occurred only in the 1 child who had the Konno procedure, and resulted in intermittent pacemaker dependence.

Table 1. Data on First Reoperations

<table>
<thead>
<tr>
<th>Variable</th>
<th>Valvar (N = 14)</th>
<th>Subvalvular (N = 9)</th>
<th>Supravalvular (N = 1)</th>
<th>Combined (N = 3)</th>
<th>Total (N = 27)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Incidence of reoperation&lt;sup&gt;b&lt;/sup&gt;</td>
<td>11/37 (29.7)</td>
<td>9/21 (42.6)</td>
<td>1/3 (33)</td>
<td>3/5 (60)</td>
<td>24/66 (36.4)</td>
</tr>
<tr>
<td>Interval to reoperation (yr)</td>
<td>8.9</td>
<td>6.9</td>
<td>3.8</td>
<td>2.7</td>
<td>7.3</td>
</tr>
<tr>
<td>Procedure</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Repeat procedure&lt;sup&gt;c&lt;/sup&gt;</td>
<td>5</td>
<td>6</td>
<td>1</td>
<td>1</td>
<td>13</td>
</tr>
<tr>
<td>Valve replacement</td>
<td>9</td>
<td>3</td>
<td>0</td>
<td>2</td>
<td>14</td>
</tr>
<tr>
<td>Perioperative mortality</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>2 (7.4)</td>
</tr>
</tbody>
</table>

<sup>a</sup>Numbers in parentheses are percentages.
<sup>b</sup>Incidence of reoperation reflects the number of reoperations as a percentage of our hospital survivors and late deaths (excludes those lost to follow-up before 2 years).
<sup>c</sup>Repeat procedure indicates valvotomy for valvar and resection for subvalvular stenosis.

There were 2 perioperative deaths among the initial reoperations, both in children who had had valve replacement. A 14-year-old patient who had undergone valvotomy 7 years earlier had a 21-mm bioprosthetic valve placed. Cardiac arrest occurred in the operating room. Autopsy revealed a strut obstructing the left coronary ostium. A 3-year-old child had had a resection of a subvalvular membrane and valvotomy 2 years previously. At reoperation, the Konno procedure was done to insert a 19-mm tilting-disc prosthesis. Postoperatively he experienced third-degree atrioventricular block requiring temporary pacing. Pacemaker failure two days after operation was associated with cardiac arrest.

Third Operation

The 25 hospital survivors were available for follow-up. One child, who had subvalvular stenosis initially and repeat resection 2 years later, was lost to further follow-up after his first postoperative clinic visit. Fourteen have been followed for a mean of 4.7 years without requiring further operative intervention. There have been no late deaths among those who have not required reoperation. A second reoperation was required in 10 children, representing 11% of our initial group and 42% of those who...
survived a first reoperation. The interval to the second reoperation was 3.7 years.

The indications for reoperation were related closely to the previous operation. All 4 of the patients with subvalvular stenosis who had previously undergone resection had recurrent stenosis with gradients ranging from 60 to 110 mm Hg. Four patients who had had valve replacement as the second operation following an initial valvotomy required a third operation for prosthesis "relative" stenosis in 2 (6 and 8 years after valve replacement) and valve complications in 2 (perivalvular leak in 1 and porcine valve degeneration in a 15-year-old patient who had received his valve 3 years earlier). A child who had had repeat valvotomy as a second procedure at the age of 6 years required valve replacement 3 years later when his valve gradient was 100 mm Hg. Another patient had supravalvular and valvular stenosis. Bacterial endocarditis, involving a bioprosthesis placed 3 years earlier when the patient was 12 years old, profound aortic insufficiency developed and necessitated reoperation.

The operations performed included valve replacement in every instance. The two prosthetic valvular stenoses were corrected by seating larger prostheses in conjunction with root enlargement by annuloplasty (17-mm tilting-disc valve to a 23-mm valve after 8 years) and Konno annuloplasty (19-mm tilting-disc valve to a 23-mm valve after 6 years). Two of the 4 children with recurrent subvalvular stenosis had a Konno annuloplasty (a 7-year-old received a 21-mm and a 14-year-old, a 23-mm bileaflet tilting-disc prosthesis). 1 had placement of a valved conduit (14-mm porcine bioprosthesis at 5 years of age), and 1 had resection of a membrane with valve replacement for resulting insufficiency (23-mm porcine bioprosthesis at 24 years of age). Reoperation in a young man with porcine valve dysfunction required a mechanical replacement, and the perivalvular leak was corrected similarly. Table 2 displays the data related to the incidence, interval, procedures performed, and operative mortality for the second reoperations.

Complications of the third operations included postoperative cerebral hemorrhage, from which recovery was complete (1 patient), postoperative right bundle-branch block following valve replacement without annuloplasty (1), and postpericardiotomy syndrome requiring salicylates (1) and pericardiotomy (1). There were 2 perioperative deaths. One occurred in the 35-year-old man with the periprosthetic leak; he had uncontrollable postoperative ventricular fibrillation. An 18-year-old patient who had valve replacement also experienced ventricular tachyarrhythmias unresponsive to medical therapy immediately postoperatively. Autopsy revealed early infarction changes. There was 1 late death: a teenager was running wind sprints when he experienced an arrest.

Fourth Operation

A fourth operation was necessary in 1 patient, a 16-year-old boy. One year after the third procedure, a third valve replacement was performed when valvular endocarditis involving a mechanical valve developed. He has done quite well for more than 3 years.

Summary of Operative Experience

A schematic representation of our experience is provided in Figure 2. Forty-four patients have survived one operation and have not required another after a mean follow-up of 9.2 years. Twenty-eight of them (63.6%) had valvular stenosis, 12 (27.3%) subvalvular, 2 (4.5%) supravalvular, and 2 (4.5%) valvular plus subvalvular stenoses. The actuarial incidence of reoperation is depicted in Figure 3 and reflects a similar rate of reoperation for valvular and subvalvular obstruction. The incidence of reoperation for patients initially seen with more than one level of stenosis appears higher than for those with isolated levels of obstruction, but the small number of patients precludes a valid actuarial analysis. The actuarial survival data are displayed in Figure 4, as is the survival of the patients with valvular and subvalvular stenosis. The subvalvular group, although reoperated on at the same rate as the valvular group, fared significantly better (p = 0.03).

Table 2. Data on Second Reoperations

<table>
<thead>
<tr>
<th>Variable</th>
<th>Valvarular (N = 5)</th>
<th>Subvalvular (N = 4)</th>
<th>Supravalvular (N = 0)</th>
<th>Combined (N = 1)</th>
<th>Total (N = 10)</th>
</tr>
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<tr>
<td>Incidence of second operation</td>
<td>5/13 (38.5)</td>
<td>4/8 (50)</td>
<td>...</td>
<td>1/2 (50)</td>
<td>10/24 (41.7)</td>
</tr>
<tr>
<td>Interval to second op (yr)</td>
<td>4.4</td>
<td>3</td>
<td>...</td>
<td>3.4</td>
<td>3.7</td>
</tr>
<tr>
<td>Procedure</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Valve replacement</td>
<td>2</td>
<td>1</td>
<td>...</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Annuloplasty with valve</td>
<td>3</td>
<td>2</td>
<td>...</td>
<td>...</td>
<td>5</td>
</tr>
<tr>
<td>Valved conduit</td>
<td>...</td>
<td>1</td>
<td>...</td>
<td>...</td>
<td>1</td>
</tr>
<tr>
<td>Perioperative mortality</td>
<td>2</td>
<td>0</td>
<td>...</td>
<td>0</td>
<td>2 (20)</td>
</tr>
</tbody>
</table>

*Numbers in parentheses are percentages.

*Denominator reflects 1 patient lost to follow-up after a month.
Fig 3. Actuarial plot of reoperation-free survival, depicting the percentage of patients surviving without reoperation at any given mean interval from initial operation. Shown separately are curves for the total population (TOT), those with valvular stenoses (V), and those with subvalvular stenoses (SUB). (NS = not significant.)

Functional Recovery
Functionally, 50 (77%) of 65 survivors have a good result (NYHA Functional Class I if older than 5 years or are developing normally without apparent need for reoperation if less than 5 years of age), and 15 (23%) have a satisfactory result (NYHA Class II if older than 5 years, if less than 5 years old, following a regimen of digoxin, catheterization planned in the next 6 months, or limited play activity noted by parents). All 4 children who have had three operations for subvalvular stenosis are in the group with only satisfactory functional results. Overall, good functional results are present in 31 (81%) of those who survived operation for valvular lesions, 12 (60%) of those who had subvalvular lesions, and all of those who had operation for supravalvular and combined obstructions.

It is notable that 10 (35.7%) of the 28 patients with valvular lesions who have not yet required reoperation are in the third or fourth decade of life (20 to 38 years of age) with good results at a mean of 16.9 years after the initial operation (range, 13 to 20 years). Seven (58%) of 12 patients with subvalvular disease are in a similar age group (20 to 33 years old) with a mean follow-up of 12.8 years. Five of the 7 have functionally good results. Two have clinically mild to moderate aortic insufficiency, although they remain asymptomatic; 1 is fully employed, and the other is a full-time college student.

Comment
Infants who require operation for critical aortic stenosis do less well than their counterparts who can undergo operation later in life. Dobell and associates [6] from Montreal reported a perioperative mortality of 14%, with 86% of these deaths occurring in infants less than 2 months of age. In our study, 16 patients had the initial operation when they were less than 1 year old (2 at other institutions). Six (37.5%) died: 2 after the first operation (2 months and 2 years postoperatively) and 2 each after the second or third operation. Ten of these 16 children are alive with a mean follow-up of only 4.5 years, and 3 have less than good results.

Critical aortic stenosis in infancy generally represents valvular obstruction [1]. All but 1 of the infants in our series had valvular stenosis (1 in combination with subvalvular), accounting for 6 of our 11 cardiac deaths. This explains in part the poorer survival of our patients with valvular stenosis compared with subvalvular obstruction. The mortality among much of the remainder of the valvular group is related to the difficulty of valve replacement in children [7]. Twenty-two patients in our series have required valve replacement at one or more operations. Fifteen of these patients were less than 18 years old at the first implantation (range, 2.5 to 17 years). Four (27%) of these 15 have had a second (and 1 a third) replacement for a total of twenty valves (including one conduit) in children less than 18 years old. There were 14 year old patients who had 4 conduit transplants.
were 3 perioperative deaths in children aged 3, 10, and 14 years (none had had a second replacement), for an operative mortality of 15% for valve implantation in this age group. There were the 2 late, sudden deaths in children who had good functional results. Ten of the 15 patients are alive at a mean follow-up of 4.9 years (8 months to 13 years) after the initial replacement. Six are considered to have good functional results.

We note with some concern the subgroup of patients with subvalvular stenosis who require reoperation. Half of the children who required a second operation underwent a third after a relatively short interval (see Table 2). At the initial operation, 5 of 21 patients had more diffuse, thickened subvalvular obstruction. All 5 of them had to be reoperated on at a mean of 6.7 years later, as did 4 other children; each of these 4 had distinct membranous obstruction initially, although 1 had a hypoplastic annulus. Three of the 5 with nonmembranous obstruction, plus the child with a small root, required a third operation a mean of 3 years later. Placement of a valved conduit, two Konno procedures, and a valve replacement were necessary.

As a result of this experience, we now believe that although membranous subvalvular stenosis may occasionally require reoperation, one of the more extensive processes [8] is likely the problem when there is recurrent stenosis. Recognition of this might dictate earlier use of a valved aortic conduit or a Konno procedure, but if a satisfactory reduction in the gradient can be demonstrated intraoperatively, we would still be inclined to delay these procedures albeit in favor of a possible reoperation. Like others [1, 2], we noted an increase in mortality with reoperation (initial operation, 3.7%; reoperation, 7.4%; second reoperation, 20%), but it would seem that the greater mortality for reoperation is the result of the underlying pathological condition, since the survival of our patients with subvalvular stenosis is not affected by an incidence of reoperation similar to that of the valvular group (see Figs 3, 4).

If we adhere to the concept that operative intervention can positively alter the natural course of aortic stenosis in childhood [9], then the management of these patients offers us a great challenge. There is neither a single curative procedure nor a definitively curable level of obstruction. In our experience, the incidence of recurrent obstruction in children with all levels of left ventricular outflow tract obstruction mandates a vigilant follow-up with liberal catheterization when indicated by clinical or noninvasive criteria. This becomes quite apparent as these patients reach adolescence and attempt to undertake the physically vigorous life-style of their peers. Later when they reach young adulthood and leave their parents and homes, they must understand the need to continue a regular surveillance program.

Although we do not have sufficient data to be certain, we are hopeful that routine use of noninvasive techniques will assist in this effort and more objectively guide catheterization. When reoperation is indicated
by a substantial gradient, we believe that repeat valvotomy or resection or both should be accomplished in preference to valve replacement. If circumstances such as resultant insufficiency or inadequate gradient reduction dictate replacement in a growing child, we, like others [10], favor the use of the Konno annuloplasty. The newer annuloplasty techniques offer a means to avoid the problems we have had with simple valve replacement earlier in our experience. In summary, we think that with aggressive follow-up and judicious reoperation, quite acceptable results in terms of survival and function can be obtained.

References

Avoiding Disrupt
Joseph M. B. Woodfill

ABSTRACT
mortality for percutaneous balloon angioplasty (15% to 20%)—as tricular dysrhythmia in 3 or more midventricular segments in 1 or more patients. At 6 months, 7 of the 19 patients treated in the first 241 procedures have had stable hemodynamics without significant ventricular block. The procedure was well tolerated by all patients. The only significant procedural complications were catheter-related infections in 3 patients who underwent multiple procedures. The success rate was 94%, with 19 of the 20 patients remaining asymptomatic and active. The procedure was effective in eliminating symptoms in the majority of patients.

From the J"ssing Thoracic Surgery Clinic, A
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