Repair of Coarctation of the Aorta in Neonates and Infants: A Thirty-Year Experience

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Between January 1962 and December 1991, 179 children less than 1 year of age underwent repair of coarctation of the aorta. Group I (1962 to 1971) consisted of 19 patients, group II (1972 to 1981) of 57 patients, group III (1982 to 1991) of 103 patients. Neonates (<30 days old) made up 60% of group I, 57% of group II, and 70% of group III. The proportion of infants with associated complex cardiac abnormalities was 7% in group I, 23% in group II, and 39% in group III. Techniques of repair included resection with end-to-end anastomosis (n = 65), subclavian flap repair (n = 85), patch aortoplasty (n = 18), and other procedures (n = 11). The early mortality (<30 days) was lowest in group III (group I, 21%; group II, 21%; and group III, 7%; p < 0.05), but the late mortality was similar in all groups (group I, 11%; group II, 13%; and group III, 15%). The overall actuarial survival was 57.7% ± 0.15% at 27.1 years in group I, 65.7% ± 0.07% at 19.7 years in group II, and 77.5% ± 0.04% at 9.3 years in group III (p = not significant). Twenty-five restenoses requiring intervention occurred in 23 patients, for an overall restenosis rate of 16.4%. The incidence of restenosis was 23% for the patients who underwent end-to-end anastomosis, 11% for those who underwent subclavian flap repair (p < 0.01), and 27% for those who underwent patch aortoplasty (p < 0.01). Balloon angioplasty was successful in relieving II of the 12 restenoses in groups II and III. The mean interval (± the standard deviation) between repair of coarctation of the aorta and definitive intracardiac repair decreased from 61.5 ± 43.5 months in group I to 41.8 ± 45.5 months in group II and 10.3 ± 13.7 months in group III (p < 0.001). Twenty-eight variables (various patient characteristics, presenting signs and symptoms, management and operative variables, and severity of disease) were subjected to a Cox proportional hazards multivariate regression analysis to determine predictors of restenosis and mortality. Only patch aortoplasty was significantly associated with restenosis (p < 0.01). Increasing age at operation and the use of monofilament nonabsorbable suture were significantly associated with freedom from restenosis (p < 0.02). Younger age at operation, the need for concomitant pulmonary artery banding, and the existence of associated cardiac abnormalities were significantly associated with early mortality (p < 0.01). This retrospective review revealed that (1) subclavian flap aortoplasty is associated with the lowest rate of restenosis after repair of coarctation during infancy, and, conversely, patch aortoplasty is significantly associated with restenosis; (2) the restenosis rate is significantly lower in association with the use of monofilament nonabsorbable suture than with the use of other suture material; (3) the early mortality after coarctation repair has decreased significantly in the current era, despite a higher proportion of infants with complex cardiac malformations; and (4) late mortality is associated with younger age at operation and the presence of severe associated cardiac anomalies, and this has remained constant during the three decades covered by this study. (Ann Thorac Surg 1995;59:33-41)

Since 1945 when the first successful repair for coarctation of the aorta (CoA) was performed [1, 2], surgical repair has remained the preferred and primary therapy for CoA. Despite satisfactory results with end-to-end anastomosis (ETA) [3-6], a small but persistent rate of restenosis [3-5] has prompted the further development of repair techniques, such as the subclavian flap aortoplasty (SFA) [7] and patch angioplasty [8, 9], as well as the use of absorbable suture [10] and the application of balloon angioplasty for the treatment of restenosis [11]. Concurrently, there have been advances in the surgical management of the associated cardiac abnormalities, as well as improvements in the approach to perioperative management. The use of prostaglandin E1 (PGE1) to maintain ductal patency has led to an improvement in the preoperative condition in certain patients [12]. Improvements in diagnostic technology have made possible a more accurate evaluation of each patient and a better understanding of the contribution of associated cardiac anomalies to the patient’s condition. Despite these changes, significant perioperative morbidity and mortality still attend CoA repair, particularly in neonates [13-16]. This population is at higher risk for complications because of the acute nature of the presentation, the presence of heart failure, and a higher prevalence of other cardiac malformations.


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were present in 19% (34/175), the most common being by direct communication with the patient, family, or local ison, patients were divided into three groups: group I, patients; and group III, 1982 to 1991, 103 patients. The During the 30 years from January 1962 to December 1991, we reviewed three decades' worth of experience with CoA repair in infants and neonates at The Johns Hopkins Hospital to identify changes in the management of patients with CoA and the technique of repair, and to determine whether these changes have resulted in improved survival and restenosis rates.

**Material and Methods**

During the 30 years from January 1962 to December 1991, 179 patients less than 1 year of age underwent repair of CoA at The Johns Hopkins Hospital. All hospitalization charts, operative notes, and clinic visit charts were reviewed retrospectively by the same reviewer to ensure consistency in data collection. Follow-up was carried out for 89% (17/19) of the procedures, compared with 58% (105/175) for group I to 22 ± 41 days in group II and 27 ± 53 days in group III (p = NS). However, the mean age at the time of operation decreased from 58 ± 93 days in group I to 54 ± 89 days in group II and 45 ± 67 days in group III (p = NS). The mean patient weight at the time of operation was not significantly different among the three groups: group I, 4.2 ± 1.7 kg; group II, 3.8 ± 1.7 kg; and group III, 3.9 ± 1.5 kg (p = NS). The frequency of associated cardiac abnormalities varied among the groups (Table 2). Isolated CoA was present in 13% (2 patients) in group I, 19% (10 patients) in group II, and 18% (19 patients) in group III (p = NS). Patent ductus arteriosus (PDA) was more prevalent in group I (27%, 4 patients) than it was in groups II (19%, 10 patients) or III (17%, 17 patients) (p = NS). The frequency of ventricular septal defects (VSDs) with or without PDA decreased over the three decades from 53% (8/15) in group I to 38% (20/53) in group II and 26% (27/103) in group III (p < 0.05). The proportion of patients with complex congenital cardiac abnormalities increased from 7% (1/15) in group I to 17% (25/153) in group II and 39% (40/103) in group III (p < 0.05). A bicuspid aortic valve was present in 35% (60/171) of all the patients. A family history of coarctation or congenital heart disease existed in 68 patients, of whom 17 (25%) had first-degree or second-degree relatives with congenital cardiac abnormalities.

The presenting signs and symptoms for CoA were absence of femoral pulses, tachypnea, cyanosis or lower extremity mottling, diaphoresis at feeding, and upper extremity hypertension (Figs 1, 2). The mean gradient between the systolic blood pressure in the upper and lower extremities was similar among the groups; group I, 65 ± 31 mm Hg; group II, 77 ± 52 mm Hg; and group III, 55 ± 30 mm Hg (p = NS). The preoperative use of PGE1 was begun in early 1976; relatively few patients in group I (9%, 5/55) received prostaglandin treatment, compared with group III (55%, 57/103) (p < 0.01). Digoxin and diuretics were the mainstay of therapy in groups I and II.

The preferred repair technique has changed over the past three decades (Table 3). Resection of the CoA with ETE was the primary mode of repair in group I, accounting for 89% (17/19) of the procedures, compared with 58%
Fig 1. Presenting signs at time of initial diagnosis. (CHF = congestive heart failure.)

(33/57) of the repairs performed in group II and 14.5% (15/103) of those done in group III (p < 0.05). Subclavian flap aortoplasty was first performed in our institution in 1978, and constituted 23% (13/57) of the repairs done in group II and 70% (72/103) of the repairs done in group III (p < 0.05). The remaining procedures consisted of patch angioplasty (18 patients), a flap–patch combination (7 patients), or extraanatomic bypass (4 patients). Absorbable suture was introduced into use during the group III period and was used in 39% (39/100). All procedures were performed through a left thoracotomy.

Isolated CoA repair was performed in 20% (3/15) of group I, 27% (15/56) of group II, and 20% (21/103) of group III. Repair of the CoA plus PDA ligation was carried out in 60% (9/15), 46% (26/56), and 54% (56/103) of groups I, II, and III, respectively. Repair of the CoA and pulmonary artery banding (with or without PDA ligation) was performed in 20% (3/15) of group I, 27% (15/56) of group II, and 25% (26/103) of group III (all p = NS).

All surviving patients had postoperative clinic visits every month for 3 months, and then every 3 months for 1 year. Thereafter, most were seen on a yearly or twice-yearly basis. Follow-up information was obtained from clinical records, as well as from direct contact with the patient or local physician.

Twenty-eight clinical variables were subjected to an initial univariate analysis to screen for positive or negative predictors of restenosis and mortality. Variables identified by univariate testing with a p value of less than 0.15 were subjected to a Cox proportional hazards regression analysis. The following variables were examined:

<table>
<thead>
<tr>
<th>Table 3. Repair Techniques for Coarctation of Aorta</th>
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<tbody>
<tr>
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<tr>
<td>Resection/end-to-end anastomosis</td>
</tr>
<tr>
<td>Subclavian flap aortoplasty</td>
</tr>
<tr>
<td>Patch aortoplasty</td>
</tr>
<tr>
<td>Extraanatomic bypass</td>
</tr>
<tr>
<td>Subclavian flap–patch aortoplasty</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>

* p < 0.01.  b p < 0.05.
Actuarial survival was determined by the Kaplan-Meier method. Comparisons between groups were performed by an analysis of variance of repeated measures and $\chi^2$ tests. All data are represented as the mean $\pm$ the standard deviation or as percentages. Actual numbers are included in parentheses. Statistical significance was assigned to differences with a $p$ value of less than 0.05.

Results

Preoperative Data
The presenting symptoms in decreasing order of frequency were: tachypnea (49%, 71/144), cyanosis (32%, 53/165), abnormal feeding patterns (23%, 38/166), and a listless or moribund state (13%, 22/167). The most prevalent sign was absence of femoral pulses (92%, 153/166). Upper extremity hypertension (defined as $>100$ mm Hg in neonates and $>110$ mm Hg in infants) was observed in 44% (60/136). Congestive heart failure was noted on chest x-ray studies in 54% (90/168). Nineteen percent (30/162) required intubation on presentation. The prevalence of the presenting signs and symptoms was similar among the three groups (see Figs 1, 2).

In groups I and II, 100% (10/10) and 81% (44/54), respectively, were given digitalis treatment preoperatively, compared with only 15% (15/100) in group III ($p < 0.05$, group III versus groups I and II). Diuretic use was similar in the groups (group I, 20%, 2/10; group II, 40%, 17/42; and group III, 36%, 36/100) ($p = NS$). Prostaglandin $E_1$ was administered in 9% (5/57) of the group II patients and 57% (55/103) of the group III patients ($p < 0.01$).

Operative and Postoperative Data
In the total series, the intraoperative mortality rate was 3.9% (7/179) and the postoperative mortality rate was 8.9% (16/179), for a total perioperative mortality of 12.8%. The perioperative mortality was 21% (4/19) for group I, 21% (12/57) for group II, and 7% (7/103) for group III ($p < 0.05$, group III versus groups I and II). Diuretic use was similar in the groups (group I, 20%, 2/10; group II, 40%, 17/42; and group III, 36%, 36/100) ($p = NS$). Prostaglandin $E_1$ was administered in 9% (5/57) of the group II patients and 57% (55/103) of the group III patients ($p < 0.01$.

Postoperative complications. Persistent congestive heart failure after CoA repair occurred in 27% (47/174). Minor complications occurred in 21% (4/19), 26% (15/57), and 23% (24/103) of groups I, II, and III, respectively ($p = NS$). Twenty-four (14%, 24/174) patients had hypertension postoperatively. Left hemidiaphragm paralysis occurred in 6 patients; right-sided paralysis opposite the CoA was observed in 1. One patient had vocal cord paralysis. Chylotrachea developed in 4 and reoperation to manage it was required in 2. Two patients required reoperation for the control of postoperative bleeding. There were no episodes of extremity necrosis on the side of the subclavian artery ligation. There were 21 infections (9 cases of pneumonia, 6 cases of systemic sepsis, and 6 cases of wound infection). There were no cases of paraplegia in this series.

There were no intraoperative or postoperative deaths nor postoperative complications among the patients undergoing isolated CoA repair. Two patients with CoA and PDA died postoperatively. All other deaths occurred in patients with VSD or complex cardiac abnormalities, or with a combination of both.

Among the patients receiving preoperative PGE$_1$ therapy, the operative mortality was 11% (7/62), compared with 15% (23/172) among those not given PGE$_1$ therapy during the same period ($p = NS$). Of the 62 patients treated with PGE$_1$, 8% (5 patients) had an isolated CoA, 16% (10 patients) had CoA and PDA, 25% (14 patients) had VSD (with or without PDA), and 53% (33 patients) had additional complex intracardiac abnormalities. During the same period, 72 patients were not treated with PGE$_1$, preoperatively. Twenty-six percent (19 patients) had an isolated CoA, 15% (11 patients) had PDA, 42% (30 patients) had an isolated VSD, and 17% (12 patients) had additional complex cardiac abnormalities. The preoperative use of PGE$_1$ was not found to be a predictor of survival (Table 4).

ADDITIONAL SURGICAL PROCEDURES. Fifty-five patients (approximately a third of the population with CoA) required subsequent cardiac surgical procedures. Twelve patients required VSD repair and pulmonary artery debanding. Closure of a VSD with or without closure of an atrial septal defect was performed in 10 patients; VSD and mitral valve repair was performed in 1. Four patients with banded pulmonary arteries experienced spontaneous VSD closure, and later underwent band removal and reconstruction of the pulmonary artery. Eleven patients required aortic valve or left ventricular outflow tract enlargement procedures. Five patients underwent a series of operations culminating in the modified Fontan procedure. Two arterial switch procedures and one Mustard operation were performed. Three patients underwent repair of an atrioventricular canal malformation. Additional procedures included two atrial septal defect closures, one mitral valve replacement, one repair of a total anomalous pulmonary venous return, one orthotopic heart transplantation for treatment of the hypoplastic left heart syndrome, and one pacemaker insertion.

Forty percent of the group I patients (4/10) required subsequent operations, as did 32% (17/52) of those in group II and 33% (34/103) of those in group III. There were one intraoperative and four postoperative deaths associated with these procedures in group II and two intraoperative and four postoperative deaths in group III. The mean interval between the time of the initial CoA repair and the subsequent secondary procedure was 61.5 $\pm$ 43.5 months in group I, 41.8 $\pm$ 45.5 months in group II, and 10.3 $\pm$ 13.7 months in group III ($p < 0.001$).

RESTENOSIS. Twenty-five clinically significant restenoses (arm-leg gradient $>30$ mm Hg in the resting state) occurred in 23 patients, for an overall restenosis rate of 16.4% (25/140). (The threshold arm-leg gradient for re-
Table 4. Univariate and Multivariate Analysis of Factors Relating to Overall Survival

<table>
<thead>
<tr>
<th>Variable</th>
<th>Restenosis (p value)</th>
<th>Survival (p value)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at operation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sex</td>
<td>0.11</td>
<td>0.0001</td>
</tr>
<tr>
<td>Age at diagnosis</td>
<td>0.24</td>
<td>0.0023</td>
</tr>
<tr>
<td>Weight</td>
<td>0.93</td>
<td>0.0014</td>
</tr>
<tr>
<td>Lack of femoral pulses</td>
<td>0.46</td>
<td>0.95</td>
</tr>
<tr>
<td>Tachypnea</td>
<td>0.91</td>
<td>0.92</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>0.95</td>
<td>0.045</td>
</tr>
<tr>
<td>Congestive heart failure</td>
<td>0.62</td>
<td>0.37</td>
</tr>
<tr>
<td>Abnormal feeding patterns</td>
<td>0.025</td>
<td>0.45</td>
</tr>
<tr>
<td>Shock</td>
<td>0.97</td>
<td>0.002</td>
</tr>
<tr>
<td>Upper extremity pressure</td>
<td>0.70</td>
<td>0.0002</td>
</tr>
<tr>
<td>Upper-to-lower extremity gradient</td>
<td>0.83</td>
<td>0.072</td>
</tr>
<tr>
<td>Digitalization</td>
<td>0.49</td>
<td>0.23</td>
</tr>
<tr>
<td>Diuretic use</td>
<td>0.97</td>
<td>0.42</td>
</tr>
<tr>
<td>Prostaglandin use</td>
<td>0.10</td>
<td>0.25</td>
</tr>
<tr>
<td>Resection and end-to-end anastomosis</td>
<td>0.97</td>
<td>0.45</td>
</tr>
<tr>
<td>Subclavian flap aortoplasty</td>
<td>0.51</td>
<td>0.057</td>
</tr>
<tr>
<td>Patch aortoplasty</td>
<td>0.13</td>
<td>0.0019</td>
</tr>
<tr>
<td>Braided nonabsorbable suture</td>
<td>0.045</td>
<td>0.070</td>
</tr>
<tr>
<td>Monofilament nonabsorbable suture</td>
<td>0.011</td>
<td>0.50</td>
</tr>
<tr>
<td>Monofilament absorbable suture</td>
<td>0.20</td>
<td>0.0001</td>
</tr>
<tr>
<td>Pulmonary artery banding</td>
<td>0.60</td>
<td>0.0001</td>
</tr>
<tr>
<td>Patent ductus ligation</td>
<td>0.73</td>
<td>0.07</td>
</tr>
<tr>
<td>Bicuspid aortic valve</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complexity of associated malformations</td>
<td>0.41</td>
<td>0.00001</td>
</tr>
<tr>
<td>Postoperative complications</td>
<td></td>
<td>0.00001</td>
</tr>
<tr>
<td>Need for later definitive procedure</td>
<td>0.31</td>
<td>0.001</td>
</tr>
<tr>
<td>Era of operation</td>
<td>0.52</td>
<td>0.19</td>
</tr>
</tbody>
</table>

Restenosis occurred in 18% (7/39) of the group II patients, but group III had the lowest rate of 12% (11/94) (p < 0.05). The mean time to the diagnosis of restenosis was 6.0 ± 5.3 years in group I, 7.0 ± 4.3 years in group II, and 0.39 ± 0.26 years in group III (p < 0.05).

In group I, 4 patients with restenoses required operation: 2 underwent re-resection with an ETE, 1 had an extraanatomic bypass, and 1 underwent patch repair with prosthetic material. One patient refused reoperation and was lost to follow-up. The patient with restenosis repaired by the ETE technique suffered a second restenosis 10 years later at 24.3 years of age and underwent successful balloon angioplasty. Successful balloon angioplasty was defined by the following two features: no angiographically apparent stenosis, and a residual gradient between the ascending and descending aorta of less than 30 mm Hg. Among the patients in group II, 3 patients with restenoses underwent surgical repair with a patch technique and 4 underwent balloon angioplasty. Balloon angioplasty was used for dilation of eight of the 11 restenoses in group III. Seven balloon procedures were successful; the 1 patient in whom it failed required patch angioplasty for surgical repair 3 months later. Overall, the ages of the patients undergoing balloon angioplasty ranged from 58 days to 24.3 years. Three group III patients with restenosis underwent surgical repair (two patch repairs and one ETE procedure) because the length of the stenosis or aortic tortuosity precluded the performance of balloon angioplasty. No patient in the series required a third CoA repair.

There were no early deaths among those patients who underwent reoperation or balloon angioplasty for the relief of restenosis. One late death occurred during a completion Fontan procedure 5 years after the patient had undergone an operation for relief of the restenosis.

From the standpoint of the risk of restenosis in relation to the surgical repair technique, restenosis occurred a mean of 6.5 ± 5.1 years postoperatively in 23% (12/52) of the patients who underwent ETE repair; this compares with 11% (9/77; p < 0.1) of those who underwent SFA at 0.63 ± 1.0 years and 27% (31/113; p < 0.01) of those who had patch angioplasty at 2.2 ± 2.4 years. From 1962 to 1978 (before SFA was introduced into use at our institution), the restenosis rate was 30% (10/33) among long-term survivors of ETE. Although the overall restenosis rate was lower in association with SFA than with ETE (p < 0.1), neither proved to be significant predictors of freedom from restenosis based on the results of multivariate analysis. However, patch aortoplasty was significantly associated with a risk of restenosis (p < 0.01) (see Table 4). All but 1 patient with restenosis who had undergone...
SFA were originally operated on when they were less than 20 days of age, and the great majority of restenoses occurred within 1 year of operation. The only late restenosis among the patients who underwent SFA occurred at 3.4 years in a patient operated on at 18 days of age. Although the follow-up was longer in the ETE group, only three restenoses developed within 1 year of repair (69, 171, and 323 days postoperatively). Younger age at operation was shown by multivariate analysis to be significantly associated with restenosis (p < 0.02) (see Table 4). Also examined was the risk of restenosis in relation to the type of suture used.

The types of suture used for repair in group I were silk and braided polyester. The two types of suture used over the past two decades have been polydioxanone absorbable suture and polypropylene. The restenosis rate by suture type was 18% (6/33) for absorbable suture (4 cases after SFA and 2 after ETE) versus 11% (9/82) for polypropylene (5 cases after SFA, 2 after ETE, and 2 after patch angioplasty, and 42% (8/19) for braided nonabsorbable suture (7 cases after ETE and 1 after patch angioplasty). The use of monofilament nonabsorbable suture (polypropylene) was significantly associated with freedom from restenosis (p < 0.02). The era of operation did not affect the restenosis rate (see Table 4).

**Follow-up Findings**

In group I, early follow-up information was obtained in 15 patients, and late follow-up data in 10 patients, of whom 9 were long-term survivors. The mean follow-up was 13.8 ± 10.4 years. Three patients had persistent hypertension, and 2 of them required medication. One of these patients suffered leg cramping with exercise, but exhibited no upper-to-lower extremity systolic blood pressure gradient. There was one late mortality (11.3 years postoperatively) stemming from ventricular fibrillation in a patient with aortic valve disease and third-degree heart block.

In group II, the mean follow-up in 39 long-term survivors was 10.9 ± 5.7 years. Three patients had significant hypertension; 1 had undergone successful balloon angioplasty and none were on medication. One patient had persistent leg claudication 20 years after the CoA repair. One patient had moderate congestive heart failure, defined by a need for medication. One patient suffered minor limb length discrepancy after SFA. Limb length differences were deemed to exist if the physician noted a discrepancy during examination at serial follow-up visits. The smallness of the left upper extremities did not preclude near-normal use of the arm in any of the patients involved. There were five late deaths, occurring an average of 2.7 ± 2 years after initial operation, and all occurred at the time of subsequent cardiac operations. One patient could not be weaned from cardiopulmonary bypass, 1 died from a left atrial monitoring line complication, 1 had fatal postoperative bleeding, and 2 died from cardiac failure.

In group III, the mean follow-up was 3.0 ± 2.9 years for 94 long-term survivors among 99 patients. Three patients had significant hypertension; 1 of whom was on medication. Shortening of the left arm was noticeable in 4 patients (4%, 4/94) who had undergone SFA. Leg claudication remained symptomatic in 10 patients despite the absence of a pressure gradient. One patient remained in congestive heart failure. There were 14 late deaths occurring an average of 0.79 ± 0.63 years after CoA repair. Six were associated with subsequent definitive cardiac surgical procedures; all patients involved had complex cardiac abnormalities. Eight late deaths occurred in patients with complex cardiac abnormalities before a definitive surgical repair procedure could be performed; six were due to cardiac failure.

The overall late mortality was 11% (1/9) in group I, 13% (5/39) in group II, and 15% (14/94) in group III (p = NS). The early and late mortalities were significantly higher in patients with other cardiac abnormalities. In patients with isolated CoA or with CoA and PDA, there was a 1.6% (1/62) hospital mortality and no late deaths. In those with VSD, the early mortality was 21% (11/53) and the late mortality was 1.9% (1/53). In those with complex intracardiac abnormalities, the early mortality was 20% (10/51) and the late mortality was 37% (19/51) (p < 0.05 compared with the other categories). For the patients treated in the most recent decade (group III), the hospital mortality for those with CoA and VSD was 5.1% (2/39) and there have been no late deaths. In those patients with complex malformations, the hospital mortality was 19% (5/26), but the late mortality rose to 54% (14/26). Based on the results of multivariate regression analysis, those variables associated with late mortality were younger age at operation, the concurrent performance of pulmonary banding, and the presence of other complex cardiac anomalies (p < 0.01). Surprisingly, the era of operation was not significantly associated with late survival (see Table 4). The overall actuarial survival was 57.7% ± 0.15% in group I at 27.1 years, 65.7% ± 0.07% at 19.7 years in group II, and 77.5% ± 0.04% in group III at 9.3 years (p = NS). The three survival curves were parallel throughout the long-term follow-up period (Fig 3).

**Comment**

The optimal method of CoA repair for infants and neonates remains controversial [17–20]. Resection with ETE was used during the first several decades of CoA repair, but was associated with a significant rate of late restenosis [4, 5]; this was seen in our series as well. Subclavian flap aortoplasty became a popular repair technique after 1978. As also seen for many other series of infants undergoing CoA repair, the incidence of restenosis in our series has been lower in association with SFA than it has been with other techniques. Of note, however, restenosis consistently occurs earlier after SFA than it does after other types of repair. Jonas [21] and Sanchez and associates [22] implicated involution of the remaining circumferential ductal tissue as the source of this early restenosis. That early age at operation is a risk factor for restenosis favors this hypothesis; indeed, in our series, multivariate analysis showed that younger age at operation was associated with restenosis. Patients undergoing an SFA at less than 20 days of age exhibited a significantly higher rate of restenosis. Late restenosis (>1 year postoperatively) es-
sentially did not occur in those who underwent SFA. Indeed, among the patients undergoing SFA, the rate of restenosis was slightly higher in association with the use of absorbable suture than it was in association with non-absorbable suture. The occurrence of late restenosis suggests failure of anastomotic growth. In this series, late restenosis appeared to be related to the type of suture used. The highest restenosis rate was seen in patients undergoing ETE in which nonabsorbable braided suture was used. The use of braided suture was associated with a markedly higher restenosis rate, but did not prove to be a significant predictor of restenosis when subjected to multivariate analysis. Although this suture incites a strong fibrous reaction near the anastomosis compared with the effects of monofilament permanent or absorbable suture [23], its association with restenosis may have more to do with its near-exclusive use in patients undergoing resection procedures involving the use of ETE early in the series. The use of monofilament nonabsorbable suture subsequently proved to be significantly associated with a reduction in the incidence of restenosis. Monofilament polypropylene is a relatively inert substance and incites minimal adjacent inflammatory reaction. There is evidence suggesting that the use of polydioxanone suture results in even less long-term fibrosis and in improved growth of vascular anastomoses [10]. However, our study findings do not corroborate this and suggest an unexpected benefit from monofilament non-absorbable suture for CoA repair in this age group.

Our results show that SFA was associated with the lowest rate of restenosis. Almost all of the restenoses in these patients occurred within 1 year of operation, and there were virtually no late restenoses in this group. In contrast, ETE was associated with a high rate of late restenosis. The difference in the timing of restenosis is likely real and not an artifact stemming from the earlier diagnosis now possible in the current era. All patients are monitored by serial blood pressure gradient examinations. Before the advent of ultrasonic technology, when restenoses were suspected, aortography and directly measured pressure gradients were used for confirmation. In the current era, echocardiography is done if a restenosis is suspected. Multivariate analysis revealed that neither type of repair was significantly associated with restenoses, even though the incidence of restenosis tended to be lower in association with SFA. Patch aortoplasty, however, was associated with a significantly higher rate of restenoses. Thus, the findings from our study support SFA and ETE as the preferable repair procedures for infants.

Late symptoms were unrelated to the type of repair performed (except for limb length discrepancy after SFA). The frequency of limb length discrepancy was similar for both infant and neonate repairs. In previous reports, both shorter arm length and brachial plexus injuries have been described as consequences of SFA [24, 25]. The decreased arm length observed in our series was minor and did not interfere with normal use of the arm.

Balloon angioplasty is known to be an effective treatment for restenosis [26–28], but it has proved to be less effective in the treatment of primary CoA [29]. Long-term freedom from restenosis has been demonstrated in patients who undergo balloon angioplasty [11]. In our series, balloon angioplasty was associated with an early success rate of 92% (11/12), a figure comparable to that observed for redo CoA repair. The procedure was effective in achieving restenosis regardless of the type of primary surgical repair performed and over a wide age range (58 days to 24.3 years). The lack of postangioplasty mortality (early or late) and morbidity in our series supports the further use of the technique. Reoperation consisting of patch angioplasty, ETE, or an extraanatomic bypass has also been associated with a 96% success rate without mortality [30]. Similarly, Pollack and colleagues [31] observed no perioperative morbidity or mortality in their series and favored the patch technique. We obtained comparable results with reoperation, with only 1 case of late restenosis among 11 cases (9%). Operations for the relief of restenosis have still been necessary despite the availability of balloon angioplasty, because the length and tortuosity of the stenotic aortic segment can render balloon angioplasty inapplicable.
Elevated early mortality and perioperative complications rates have been associated with CoA repair in infants [13, 18, 32]: an early mortality of 20% to 43% has been reported as well as a significant frequency of postoperative complications such as hypertension, post-cardiomyotomy syndrome, pulmonary insufficiency, infection, bleeding, and neurologic injury [13]. The reasons for this higher complication rate among infants include the acute nature of the presentation in severely ill patients, poor response to attempts at medical management, and the need for emergent repair. Acidosis secondary to poor tissue perfusion distal to the CoA often contributes to causing poor cardiac function. The early mortality rates have been even higher in patients with other cardiac abnormalities (25% to 44%) [33]: in our study, virtually all early deaths occurred in patients with additional cardiac abnormalities. Younger age at operation was also a factor associated with higher mortality, because younger infants were more likely to present in severe distress.

The advent of PGE, therapy in 1976 was temporally associated with improved perioperative survival, even though the prevalence of complex intracardiac abnormalities was greater in these patients than it was in those treated in the earlier era. However, the long-term survival has remained unchanged because of the deaths from cardiac failure that occur while the patients are awaiting a definitive surgical procedure for the repair of complex cardiac malformations. Because of advances in cardiac surgical procedures for the treatment of congenital heart disease, successful repair in small infants is now possible. The interval between CoA repair and definitive repair has also decreased over the past 30 years. In our study, the early mortality was found to be significantly decreased between the first two decades and the third decade. However, the late mortality was higher in the third decade, resulting in no statistical difference in the overall survival among the patients in the three groups. Fifty percent (7/14) of the late deaths in group III occurred before anticipated definitive repair was performed. This shift points up the continued improvement in operative and perioperative management techniques in the current era.

The most significant recent improvement in late results has been observed in patients with CoA and VSD. There was a marked decrease in early mortality, unrelated to changes in the pulmonary artery banding practice. Other authors have advocated either avoiding pulmonary banding in favor of tolerating a moderate ventricular septal defect, or delaying surgery until the volume is larger. However, the primary determinant of postoperative survival in this group of patients.

The design of this study is limited by the fact that it is retrospective and spans three decades. There is also no comparable control group. These factors raise the possibility that there may have been changes in the definitions of the variables studied in the precision with which they were measured over the course of the three decades. However, the variables assessed have been largely unaffected by changes in technology. The significant associations with restenosis and mortality therefore need to be interpreted in this light. In addition to documenting the evolution in the treatment of CoA over the course of three decades, the findings in our series confirm the excellent early and late survival rates associated with repair of isolated CoA in neonates and infants; they also suggest that SFA and ETB repairs performed with polypropylene suture achieve results superior to those achieved with patch aortoplasty in terms of preventing late restenosis. In the current era, the early and late mortality in infants and neonates undergoing CoA repair depends mainly on the presence or absence of other cardiac malformations.

References


DISCUSSION

DR ROBERT M. SADE (Charleston, SC): This excellent paper reached some interesting conclusions, and I congratulate the authors for putting together a very nice series. There were several variables that you were looking at all at the same time, suture type, the decade of operation, the type of repair, as well as others, and I did not see that you had dissected out the effect of each of those independently of the others. For example, the suture type probably changed over time, being different in the first decade than it was in the first follow-up year. All of the restenoses occurred in the early modern era regardless of the type of repair.

Subclavian flap aortoplasty repair was used more frequently in recent repairs. All of the restenoses occurred in the early follow-up period. There was only one restenosis occurring after 1 year in the subclavian flap aortoplasty group. In the group having resection with end-to-end anastomosis there were only three restenoses occurring within the first follow-up year. All others occurred later. Thus era of repair and subsequent difference in follow-up period should not influence the conclusion that this repair is more favorable in infants and neonates.

Further analysis using multivariate technique will be helpful in dissecting out covariable influences.