Aortic coarctation with hypoplastic aortic arch

Results of extended end-to-end aortic arch anastomosis

Between 1980 and 1986, 80 infants (≤3 months old) with symptomatic aortic coarctation and associated severe tubular hypoplasia of the transverse aortic arch underwent surgical treatment. Extended end-to-end aortic arch anastomosis was used in an attempt to correct both the isthmic stenosis and the hypoplasia of the transverse arch. After complete excision of the coarctation tissue, a long incision was made in the inferior aspect of the aortic arch, which was then anastomosed to the obliquely trimmed distal aorta. Pure coarctation was present in 17 patients (group I); 24 infants had an additional ventricular septal defect (group II), and 39 patients had associated complex heart disease (group III). The overall early mortality rate was 26% (confidence limits 21% to 32%) (18% in group I, 17% in group II, and 36% in group III). The early risk declined with time and was 18% (confidence limits 12% to 26%) for the last 2 years (seven deaths in 39 patients). Follow-up was 100% for a mean of 19 months. Actuarial survival rate at 3 years was 82% for group I, 78% for group II, and 32% for group III. Recurrent coarctation (gradient ≥ 20 mm Hg) occurred in six operative survivors (10%, confidence limits 6% to 16%) and necessitated reoperation in three. Freedom from recoarctation at 4 years was 88%. Because extended end-to-end aortic arch anastomosis provides adequate correction of the aortic obstruction and entails a low risk of restenosis, it is our procedure of choice in infants with coarctation and severe hypoplasia of the aortic arch.

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Many patients with symptomatic aortic coarctation in early infancy have a more diffuse narrowing caused by hypoplasia of the transverse aortic arch, in addition to the localized isthmic stenosis. There is still a considerable debate as to the optimal surgical management of this common congenital anomaly. Procedures in which the hypoplastic arch is left intact are usually advocated: resection of the isthmic stenosis with end-to-end anastomosis or subclavian flap angioplasty. However, there is some evidence to suggest that both the early mortality and the incidence of recoarctation may be increased if the hypoplastic segment is disregarded. Therefore, various procedures that provide surgical correction of the tubular hypoplasia have been described.

Since 1980 we have tried to correct hypoplasia of the transverse aorta in all infants with coarctation and tubular hypoplasia by using a modification of the end-to-end anastomosis. The early and late results of this experience are reported herein.

Methods

Patient population. Between January 1980 and December 1986, 80 infants (≤3 months old) underwent surgical repair of aortic isthmic coarctation and associated tubular hypoplasia of the aortic arch. Tubular hypoplasia was defined as a narrowing of the transverse aorta commencing at the innominate or the left common carotid artery and extending up to the entry of the ductus arteriosus, in which the diameter of the hypoplastic segment was less than 50% of that of the descending thoracic aorta.

There were 53 boys and 27 girls. The mean (± standard deviation) weight was 3.2 ± 0.6 kg. Fifty-four (67.5%) patients were less than 1 month old. Seventeen (21%) infants had pure coarctation and no major intracardiac lesion (group I). Group II included 24 (30%) patients with coarctation and an additional ventricular septal defect. Thirty-nine (49%) children had coarctation and associated complex intracardiac anomalies (group III) (Table I). A patent ductus arteriosus was present in 52 (65%) patients.
Table I. Associated cardiac anomalies in group III (46 anomalies in 39 patients)

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital mitral valve anomaly</td>
<td>12</td>
</tr>
<tr>
<td>Valvular aortic stenosis</td>
<td>9</td>
</tr>
<tr>
<td>Subvalvar aortic stenosis</td>
<td>4</td>
</tr>
<tr>
<td>Transposition plus ventricular septal defect</td>
<td>11</td>
</tr>
<tr>
<td>Transposition plus intact ventricular septum</td>
<td>1</td>
</tr>
<tr>
<td>Corrected transposition</td>
<td>2</td>
</tr>
<tr>
<td>Single ventricle</td>
<td>4</td>
</tr>
<tr>
<td>Double-outlet right ventricle</td>
<td>2</td>
</tr>
<tr>
<td>Complete atrioventricular canal</td>
<td>1</td>
</tr>
</tbody>
</table>

Operative technique. The operation is performed through a standard left posterolateral thoracotomy. The aorta is completely mobilized, including the transverse arch, the proximal portion of the arch vessels, and the upper half of the descending thoracic aorta. The intercostal arteries originating from the descending aorta are fully mobilized but usually not divided. If present, a ductus arteriosus is ligated and divided. Occasionally, the pulmonary side of a large ductus is oversewn.

The type of coarctation repair is then chosen according to the operative findings. When the hypoplastic segment is located between the origin of the left common carotid artery and the entry of the ductus (Fig. 1), vascular occlusive clamps are placed on the distal portion of the arch across the origin of the left common carotid artery and on the descending thoracic aorta distal to the coarctation. All macroscopically abnormal periductal tissue is excised. The proximal incision is made across the aorta just distal to the origin of the left subclavian artery and is extended into the inferior aspect of the distal transverse arch. The distal aortic incision is made into the wide postcoarctation segment at an appropriate angle to match the orientation of the proximal incision and offer the widest possible aortic diameter. The anastomosis is performed with a single running suture of 7-0 polypropylene or 7-0 polydioxanone.

In a less common situation, when the hypoplastic segment extends from the innominate artery up to the entry of the ductus or when the portion between the left common carotid artery and the left subclavian artery is very long (Fig. 2), the surgical technique is changed. The left subclavian artery is ligated. The proximal vascular clamp is placed proximal to the origin of the left common carotid artery, often slightly across the origin of the innominate artery. All abnormal periductal tissue is excised. The hypoplastic transverse arch is divided between the left common carotid artery and the left subclavian artery. The aortic incision is then extended into the inferior aspect of the arch as proximally as possible. The distal aorta is appropriately trimmed and anastomosed with a single running suture.

Surgical management

Initial operation. Sixty-one (76%) patients underwent a type 1 procedure (Fig. 1). Mean (± standard deviation) peak systolic pressure gradient across the repair area, assessed in 49 patients at the end of the operation, was 5.0 ± 6.8 torr. The type 2 operation (Fig. 2) was used in 19 patients. Mean (±
Table II. Causes of early deaths (21 deaths in 80 patients)

<table>
<thead>
<tr>
<th>Cause</th>
<th>Pure coarctation (n = 17)</th>
<th>Coarctation + VSD (n = 24)</th>
<th>Coarctation + complex anomaly (n = 39)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Associated cardiac anomalies</td>
<td>0</td>
<td>1</td>
<td>14</td>
</tr>
<tr>
<td>Acute declamping syndrome</td>
<td>1</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Congestive heart failure</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td><strong>p = 0.16</strong></td>
<td><strong>3 (18%)</strong></td>
<td><strong>4 (17%)</strong></td>
<td><strong>14 (36%)</strong></td>
</tr>
<tr>
<td></td>
<td><strong>(8% - 32%)</strong></td>
<td><strong>(9% - 28%)</strong></td>
<td><strong>(27% - 45%)</strong></td>
</tr>
</tbody>
</table>

Table III. Causes of late deaths (12 deaths in 59 patients)

<table>
<thead>
<tr>
<th>Cause</th>
<th>Pure coarctation (n = 14)</th>
<th>Coarctation + VSD (n = 20)</th>
<th>Coarctation + complex anomaly (n = 25)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congestive heart failure</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Associated cardiac anomaly</td>
<td>0</td>
<td>0</td>
<td>5</td>
</tr>
<tr>
<td>Reoperation</td>
<td>0</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td>Palliation</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Complete repair</td>
<td>0</td>
<td>1 (5%)</td>
<td>11 (44%)</td>
</tr>
<tr>
<td><strong>p &lt; 0.001</strong></td>
<td><strong>0 (0%)</strong></td>
<td><strong>1 (5%)</strong></td>
<td><strong>11 (44%)</strong></td>
</tr>
<tr>
<td></td>
<td><strong>(0% - 12%)</strong></td>
<td><strong>(1% - 16%)</strong></td>
<td><strong>(33% - 56%)</strong></td>
</tr>
</tbody>
</table>

standard deviation) peak gradient was evaluated in 15 patients and was 2.7 ± 4.2 torr. Polypropylene suture was used in 19 patients and polydioxanone monofilament in 61 patients.

Associated procedures were performed in 39 (49%) patients. Eleven (46%) infants of group II underwent pulmonary artery banding. In group III, 28 (78%) patients underwent a concomitant operation: pulmonary banding in 26 and closed aortic valvotomy in two.

Subsequent operations. Late cardiovascular operations were performed 3 to 29 months (mean 10 ± 7 months) after the coarctation operation in 22 patients: a palliative procedure in two, complete intracardiac repair in 17, recoarctation repair in two, and complete repair with concomitant recoarctation repair in one.

Follow-up. Follow-up data were collected during a 1-month closing interval (January 1987). All patients were traced. Children not followed up at this institution were contacted through referring physicians or parents. Mean (± standard deviation) follow-up was 19 ± 17 months (range 2 months to 5.4 years).

Continuous data were presented as mean ± one standard deviation and crude ratios with 70% confidence limits (CL). Standard actuarial techniques were used to study patient survival and freedom from recoarctation; actuarial probabilities were expressed as mean ± one standard error. Statistical comparison was performed by standard techniques.

Results

Early results (within 30 days after operation). The early mortality rate was 26% (CL 21% to 32%). The causes of death are listed in Table II. Mortality rates were not significantly different among the subgroups (group I, 18%; group II, 17%; group III, 36%; p = 0.16) and were increased, but not significantly so, by very young age (<1 month) at operation (p = 0.08).

Early mortality was increased by earlier date of operation, although the difference did not reach statistical significance (p = 0.07). During the last period (1985 to 1986), among 39 patients there were seven early deaths (18%, CL 12% to 26%): one in group I, one in group II, and five in group III (Fig. 3).

Postoperative complications occurred in 12 patients (15%). The most frequent was paradoxical hypertension (six patients) necessitating intravenous infusion of vasodilators.

Late results

Late mortality. Among the 59 operative survivors, there were 12 late deaths (late mortality rate 20%, CL 15% to 27%). These deaths occurred 2 to 14 months (mean 5 ± 3 months) postoperatively. Most of them (10/11) were observed in group III and were related to either associated cardiac anomalies or reoperation (Table III). Late mortality was not influenced by age at operation.

Survival. The overall actuarial probability of survival at 4 years was 56% ± 6%. The probability of survival was significantly lower in group III patients than in the other two groups (Fig. 4).

Recoarctation. Recurrent coarctation was defined as the presence of a resting peak systolic pressure gradient exceeding 20 mm Hg across the repair area. Recoarcta-
Fig. 3. Early mortality rates (with 70% confidence limits) according to date of operation. VSD, Ventricular septal defect; CoA, coarctation of aorta.

Fig. 4. Actuarial survival rates for patients with pure coarctation (pure CoA), coarctation with ventricular septal defect (+ VSD), and coarctation with complex cardiac anomaly (complex).

Fig. 5. Actuarial probabilities of recurrent coarctation and reoperation for recoarctation.

The overall actuarial probability of freedom from recoarctation was 88% ± 5% at 4 years. The probability of freedom from reoperation for recoarctation was 90% ± 7% (Fig. 5).

The recoarctation rate was 13% (CL 8% to 20%) after type 1 repair and 0% (CL 0% to 14%) after type 2. The use of polypropylene material for suturing was associated with a 20% (CL 7% to 41%) rate of restenosis, whereas this rate was 8% (CL 4% to 14%) when polydioxanone monofilament was used.

Evaluations of the peak systolic pressure gradient, immediately after repair and late postoperatively, were available in 50 patients. The results, according to the type of repair, are summarized in Fig. 6.

Comments

The association of tubular hypoplasia of the transverse aortic arch to isthmic coarctation is common in infants. This is particularly true in the presence of concomitant ventricular septal defect or intracardiac complex anomaly but may also occur in patients with coarctation without an associated major cardiac defect. During the period 1980 to 1986, 178 infants with aortic coarctation were operated on at our institution. Severe tubular hypoplasia was present in 65%.

When the hypoplasia is moderate, there is usually no pressure gradient across the hypoplastic area and this lesion can be left intact at the time of coarctation repair without deleterious effect. If the hypoplasia is severe, as it was in all patients in the present report, there is usually a mild pressure gradient across the hypoplastic segment. Thus the surgical correction of the isthmic coarctation alone, even if optimal, leaves a mild obstruc-
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Fig. 6. Evaluations of peak systolic gradient across the aortic repair at the end of operation (early postop) and late postoperatively. 1, After type 1 procedure. Three patients (asterisk) underwent reoperation for recoarctation. 2, After type 2 procedure.

Fig. 7. Angiographic aspect (A) before operation and (B) 30 months postoperatively.

tion of the left ventricular outflow. This may compromise a successful early outcome in many patients: (1) Complete relief of the aortic obstruction is mandatory if left ventricular dysfunction is present; (2) another concomitant left-sided obstacle (subvalvular or valvular aortic stenosis) may increase the total left ventricular strain; (3) the need for associated pulmonary artery banding would leave an obstacle on each cardiac outflow tract. In our series of 178 consecutive infants, the early mortality rate was significantly increased by severe tubular hypoplasia (without hypoplasia, 8%; with hypoplasia, 26%; \( p < 0.01 \)).\(^{17}\) Besides, tubular hypoplasia has been said to increase the prevalence of recoarctation.\(^{11}\) For all these reasons, we believe that severe hypoplasia of the transverse aortic arch should be corrected, provided the procedure can be done without increasing the operative risk or the incidence of restenosis.

The technique of aortic arch reconstruction used in the present series has been previously reported.\(^{1,16}\) The principle is to excise completely the coarctation tissue and to restore a normal-sized aortic arch by creating an extended end-to-end anastomosis to the inferior aspect of the transverse aorta. Extensive mobilization of the aortic arch and descending aorta is necessary to allow adequate resection and reanastomosis without undue tension; this usually can be done without division of intercostal vessels. In most cases the arch vessels are left intact although sacrifice of the left subclavian artery
may be necessary in selected patients. Our data (Fig. 6) and those from other reports\textsuperscript{1-3,16} show that extended end-to-end anastomosis of the aortic arch provides complete relief of the aortic obstruction in almost all cases, whatever the operative anatomic findings are (Fig. 7).

The overall early mortality rate noted in the present series is high (26%, CL 21\% to 32\%). In our complete series of infants with coarctation and hypoplastic aortic arch, surgical repair of the hypoplasia did not alter the early mortality rate (with repair, 26\%; without repair, 26\%).\textsuperscript{17} These results arouse some comments. On one hand, infants with coarctation and associated severe tubular hypoplasia constitute a high-risk subset. Many of them (49\% in our series) have associated major complex intracardiac anomalies, a well-defined risk factor of early death.\textsuperscript{1,5,7,9} In the remaining patients, particularly those without a ventricular septal defect, the left side of the heart is usually small.\textsuperscript{13} On the other hand, the operative mortality declined with time (18\%, CL 12\% to 26\% for the past 2 years). This decline may be due in part to better surgical mastery of the technique but also to improvement in the perioperative management and especially to the introduction of prostaglandin E\textsubscript{1}.\textsuperscript{3}

Because reports dealing specifically with this subgroup of patients are scarce, comparisons with other techniques are difficult to make. Our current early mortality rate is not significantly different from that rate observed in recent series involving subclavian flap angioplasty\textsuperscript{6-10} or classic end-to-end anastomosis\textsuperscript{15} in which all infants were included, irrespective of the association of tubular hypoplasia.

The incidence of recoarctation after extended end-to-end aortic arch anastomosis is low (10\%, CL 6\% to 16\%) and compares favorably with the risk of restenosis noted after classic end-to-end anastomosis\textsuperscript{1-5} or subclavian flap angioplasty.\textsuperscript{6-10} The coarctation tissue and the surrounding aortic wall consist partially or totally of ductal material.\textsuperscript{19,20} This abnormal tissue may involute or constrict and play an important role in molding this aortic area over the first 3 months of life.\textsuperscript{4,5,21} This continuing development may explain most of the recoarctations after subclavian flap angioplasty (in which the ductal material is left in place)\textsuperscript{4,5,21} or after classic end-to-end anastomosis (in which the ductal tissue is not always completely excised).\textsuperscript{3} This emphasizes the need, at least before 3 months of age, to remove totally the periductal tissue.

Complete excision of the ductal material is part of the extended end-to-end aortic arch anastomosis procedure. The surgical resection cuts back on both sides so that the anastomosis is done on the normal aortic wall. Evidence that such a circumferential aortic suture line per se has no restricted growth potential is accumulating from the neonatal arterial switch experience for transposition of the great arteries.\textsuperscript{3,22}

Provided complete excision of the ductal material is achieved, recoarctation can be related to immediate unsatisfactory surgical repair or to secondary unexplained growth failure. Incomplete relief of the aortic obstruction is unusual after extended end-to-end anastomosis; in the present series, among the six patients with recoarctation, only one had an immediate postrepair gradient equal to 20 mm Hg. The type of suture material may play an important role in the occurrence of secondary growth failure.\textsuperscript{3} Our results suggest that the risk of recoarctation may be lower when polydioxanone monofilament is used. Although further evaluation is mandatory, the introduction of this suture material may be a significant advance.\textsuperscript{23}

In conclusion, the optimal surgical management of infants with aortic coarctation and concomitant hypoplastic aortic arch remains controversial. Whether aortic arch hypoplasia should be repaired remains to be determined. Our data show that extended end-to-end anastomosis of the aortic arch provides adequate relief of the aortic obstruction and entails a low risk of recurrent coarctation but fails to reduce the high operative risk noted in this subset of patients. However, reports dealing specifically with this difficult group of infants and further experience are necessary before precise recommendations can be made.

We express our appreciation to Corinne Pasquet for her secretarial assistance.

REFERENCES