Coarctation of the Aorta in Infancy


SUMMARY
Material from 78 infants with coarctation and congestive heart failure during the first 6 months of life was reviewed. Three fourths of these infants had combined aortic lesions, that is, coarctation, a narrow constrictive zone of stenosis, and tubular hypoplasia of the transverse arch, a proximal, long, uniformly narrow segment.

It was the purpose of this study (1) to distinguish coarctation of the aorta from tubular hypoplasia by using pathologic and angiographic measurements, (2) to assess the relationship of these two entities and additional shunting cardiac defects with the occurrence of congestive heart failure in infancy, and (3) to evaluate the optimal therapeutic approach for symptomatic infants with coarctation.

It was found that (1) coarctation of the aorta as an isolated anomaly is a relatively uncommon cause of congestive heart failure in infancy. In contrast, coarctation associated with tubular hypoplasia of the transverse arch or with additional cardiac defects is frequently associated with severe congestive heart failure. It was found also (2) that surgical management is far more successful than medical management alone in symptomatic infants with coarctation of the aorta and associated significant cardiac defects.

Additional Indexing Words:
Congenital heart disease Tubular hypoplasia Congestive heart failure
Cardiac surgery

Coarctation of the aorta with congestive heart failure in early infancy represents a complex clinical, pathologic, physiologic, and therapeutic spectrum. This report is concerned with the interplay of the anatomy of the coarctation, the presence of associated shunting lesions, and the response of the pulmonary vascular bed in a group of 78 infants with cardiorespiratory symptoms requiring hospitalization before 6 months of age.

Coarctation of the aorta was first described by Morgagni\(^1\) in 1760 as a zone of constriction in the aorta. In 1903, Bonnet\(^2\) classified coarctation of the aorta into two types: "adult" coarctation, a sharply localized zone of constriction, and "infantile" coarctation, a long uniformly narrow segment. Subsequent authors have attempted to reclassify this malformation on the basis of combined anatomic and physiologic factors. These classifications, however, encompass both localized constriction and diffuse hypoplasia under the definition of coarctation of the aorta and lack of precise morphologic description.

Edwards and associates\(^3\) advanced a more exact anatomic description of coarctation of the aorta, defining it as a sharply localized zone of stenosis characteristically found at the junction of the transverse aortic arch and the descending aorta. This localized stenosis is produced by a curtain-like infolding of the aortic media into the lumen of the aorta. A long uniformly narrow segment, the infantile coarctation of Bonnet, was described by

\^1\) From The Children's Memorial Hospital (Willis J. Potts Children's Heart Center) and The Department of Pediatrics, Northwestern University Medical School, Chicago, Illinois.

\^2\) Work was supported in part by The Helen Fay Hunter Pediatric Cardiology Fund and Grant T12-HE-03770 from the National Institutes of Health.

\^3\) Address for reprints: Milton H. Paul, M.D., The Children's Memorial Hospital, Chicago, Illinois 60614.
Edwards and associates as tubular hyoplasia. It is characterized by a normal aortic media and is distinguished from coarctation as a separate pathologic entity. This pathologic distinction allows for a more precise evaluation of the relative roles of coarctation of the aorta and tubular hypoplasia as causes of congestive heart failure in early infancy.

We have reviewed material from 78 infants with coarctation of the aorta with particular reference to the aortic arch morphology as described by Edwards and co-workers. In all infants congestive heart failure developed before 6 months of age. It was the purpose of this study: (1) to delineate coarctation of the aorta from tubular hypoplasia by using pathologic and angiocardiographic measurements; (2) to assess the relationship of these two entities and additional shunting cardiac defects to the occurrence of congestive heart failure in infancy; and (3) to evaluate the optimal therapeutic approach for symptomatic infants with coarctation.

**Quantitative Classification of Aortic Arch Pathology (Autopsy and Angiographic Criteria)**

Criteria for tubular hypoplasia of the transverse aortic arch were developed from autopsy measurements of the frontal diameter of the transverse arch (TA), the ascending aorta (AA), and the descending aorta (DA) in 26 normal infants and 37 infants with coarctation. Measurements were made in the middle of the ascending aorta, the middle of the transverse aortic arch, and in the descending aorta 5 cm distal to the isthmic region in order to avoid the region of post-stenotic dilatation in coarctation (figs. 1 and 2). It was possible to compensate for age and other developmental variables by using the ratio of

---

**Figure 1**

*Autopsy specimen from an infant with coarctation of the aorta. Note the combination of aortic pathology; a diffusely hypoplastic segment involving the entire length of the transverse aortic arch plus a narrow constrictive diaphragm at the entry of the closing ductus arteriosus. AA = ascending aorta; TA = transverse aortic arch; PDA = patent ductus arteriosus; DA = descending aorta; PA = main pulmonary artery; CO = coarctation of the aorta.*

**Figure 2**

*Quantitative classification of aortic arch pathology. Angiocardiogram in left anterior oblique view. AA = ascending aorta; TA = transverse arch; DA = descending aorta.*

*Circulation, Volume XL, September 1969*
the transverse arch diameter to that of the ascending aorta and the ratio of the transverse aortic arch diameter to that of the descending aorta. Use of the diameter ratios has the further advantage of permitting recovery of directly comparable ratio measurements from angiocardiograms. Such in vivo measurements were obtained on angiocardiograms from 31 infants with coarctation of the aorta.

Reliable separation of a normal from an abnormal aortic arch required the use of these multiple ratio measurements. Linear discriminant analysis provided the following criteria for a normal population (fig. 3): TA/AA + 0.37 TA/DA ≥ 0.82.

By use of this discriminant function, three types of transverse aortic arches were defined in our material: type I, normal transverse aortic arch; type II, moderate tubular hypoplasia of the transverse aortic arch; and type III, severe tubular hypoplasia of the arch (fig. 4).

The anatomic malformations in our material on 78 infants with coarctation of the aorta and congestive heart failure in the first 6 months of life were reviewed. In 26 infants who are alive, the diagnosis was made by cardiac catheterization and angiography. In the remaining 52 patients, the diagnosis was based on postmortem examination. Of 78 infants whose condition was originally diagnosed as coarctation of the aorta, 71 had a segmental constrictive zone of stenosis, that is, coarctation as defined by Edwards and co-workers.3

Figure 3

Quantitative classification of aortic arch pathology: The ratio of the diameter of the transverse aortic arch (TA) to that of the descending aorta (DA) on the abscissa is plotted against the ratio of the diameter of the transverse aortic arch to that of the ascending aorta (AA) on the ordinate. The oblique line separates the normal from hypoplastic transverse aortic arch. This separation was derived by discriminant analysis of these ratio measurements. The criteria for normal transverse aortic arch are given on the illustration.
The other seven infants had only tubular hypoplasia of the transverse aortic arch and no coarctation of the aorta.

In the group of 71 infants with coarctation of the aorta, 16 had a normal (type I) transverse aortic arch, 44 had moderate tubular hypoplasia of the arch (type II), and 11 had severe hypoplasia of the transverse aortic arch (type III). Thus, in our coarctation population, 55 of 71 infants with coarctation of the aorta had combined aortic lesions, that is, a narrow constrictive zone of stenosis, coarctation, and a proximal, long, uniformly narrow segment, tubular hypoplasia of the transverse aortic arch (fig. 1).

Of the seven infants who had only tubular hypoplasia of the transverse aortic arch, the transverse aortic arch was moderately hypoplastic (type II) in three and severely hypoplastic (type III) in four.

**Associated Cardiac Defects**

The cardiac malformations associated with coarctation of the aorta and their relationship to the anatomic types of transverse aortic arch in 71 infants are listed in table I. The coarctation in all of these infants was located either at or just proximal to the entry of the ductus arteriosus.

*Figure 4*

Quantitative classification of aortic arch pathology: Histograms showing frequency distribution based on discriminant analysis of the ratio data from measurements of the transverse aortic arch, ascending aorta, and descending aorta. Data from autopsies of infants with no aortic pathologic changes are plotted on the right (normal-autopsy); data from autopsied coarctation material, on the upper left (coarctation-autopsy); and data from angiographic coarctation material, on the lower left (coarctation-angi). Three types of transverse aortic arch pathology are defined using these measurements: type I, normal transverse aortic arch; type II, moderately hypoplastic transverse aortic arch; type III, severely hypoplastic transverse aortic arch.
Table 1
Associated Cardiac Lesions in 71 Infants with Coarctation of the Aorta and with Congestive Heart Failure in the First Six Months of Life

<table>
<thead>
<tr>
<th>Aortic arch</th>
<th>Infants (total)</th>
<th>Coarctation only</th>
<th>Coarctation plus:</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Patent ductus arteriosus</td>
</tr>
<tr>
<td>Type I, normal</td>
<td>16</td>
<td>5</td>
<td>7</td>
</tr>
<tr>
<td>Type II, moderately hypoplastic</td>
<td>44</td>
<td>11</td>
<td>29</td>
</tr>
<tr>
<td>Type III, severely hypoplastic</td>
<td>11</td>
<td>0</td>
<td>10</td>
</tr>
</tbody>
</table>

In the type I transverse aortic arch group, five infants had coarctation of the aorta without other cardiac lesions. Seven infants had a patent ductus arteriosus (PDA); however, in only one patient was the ductus widely patent and hemodynamically significant. Ventricular septal defect (VSD) was found in nine infants. Atrial septal defect (ASD) was found in two, aortic stenosis in two, and mitral stenosis in one.

In the type II transverse aortic arch group, 11 had coarctation of the aorta only. The ductus arteriosus was patent in 29; in 21 it was probe patent, and in eight it was widely patent. A VSD was found in 20 patients. Four infants had transposition of the great arteries and one had total anomalous pulmonary venous return. ASD was noted in two, aortic stenosis in one, and mitral stenosis in one.

In the infants with type III transverse aortic arch, coarctation of the aorta was always associated with additional single or multiple cardiac malformations. The ductus arteriosus was patent in 10, although it was widely patent in only two. VSD was found in nine patients, transposition of the great arteries in three, ASD in two, and aortic stenosis in two.

The different cardiac malformations present in the seven infants who had tubular hypoplasia of the transverse aortic arch without coarctation of the aorta are listed in table 2. In all these infants the diagnosis was verified at postmortem examination. The ductus arteriosus was patent in all, but widely patent in only one. VSD was found in two infants and transposition of the great arteries in three, with a single ventricle in one.

Infants with particularly complex cardiac malformations were excluded from further clinical analysis because of the serious independent prognostic significance of these malformations. Of the 11 infants with complex malformations, 7 had associated Cardiac Lesions in 71 Infants with Coarctation of the Aorta and with Congestive Heart Failure in the First Six Months of Life.

Table 2
Associated Cardiac Lesions in Seven Infants with Isolated Tubular Hypoplasia

<table>
<thead>
<tr>
<th>Aortic arch</th>
<th>Infants (total)</th>
<th>Patent ductus arteriosus</th>
<th>Probe patent</th>
<th>Widely patent</th>
<th>Ventricular septal defect</th>
<th>Atrial septal defect</th>
<th>Aortic stenosis</th>
<th>Mitral stenosis</th>
<th>Transposition of the great arteries</th>
<th>Single ventricle</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type II, moderately hypoplastic</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Type III, severely hypoplastic</td>
<td>4</td>
<td>4</td>
<td>3</td>
<td>1</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>1</td>
</tr>
</tbody>
</table>

Circulation, Volume XL, September 1969
cardiac malformations, 10 had transposition of the great arteries, and one had total anomalous pulmonary venous return. Three additional infants were excluded because of incomplete clinical data.

Further analysis of the electrocardiograms, physiologic data, and the medical and surgical clinical course was directed to the remaining 64 infants: 60 with coarctation of the aorta and four with isolated tubular hypoplasia. The infants with coarctation of the aorta are grouped in table 3 according to the transverse arch anatomy (type I, normal; type II, moderate hypoplasia; type III, severe hypoplasia). Each transverse arch group is further subdivided as having isolated coarctation, coarctation with an additional extra or intracardiac lesion, or coarctation with multiple other cardiac lesions.

Of the four infants with isolated tubular hypoplasia, two had transverse aortic arches of type II and two had type III arches. In two of these a PDA was the only associated cardiac lesion. A third infant had PDA, ASD, and aortic stenosis, and the fourth infant had VSD and PDA.

**Electrocardiograms**

Electrocardiograms of 60 infants were available for review. The most frequent findings were right axis deviation (RAD) of the frontal QRS vector in 44 infants (73%) and right ventricular hypertrophy (RVH) in 38 (63%). RAD and RVH were present in all 11 infants seen before 3 months of age with isolated coarctation and a normal or moderately hypoplastic transverse arch. No consistent electrocardiographic pattern was noted in

---

**Table 3**

*Diagnosis and Aortic Arch Anatomy of 60 Infants with Coarctation of the Aorta*

<table>
<thead>
<tr>
<th>Associated cardiac lesion</th>
<th>Type I</th>
<th>Type II</th>
<th>Type III</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coarctation of the aorta only</td>
<td>5</td>
<td>10</td>
<td>0</td>
</tr>
<tr>
<td>Coarctation of the aorta with single cardiac lesion</td>
<td>5</td>
<td>12</td>
<td>1</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>1</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>3</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Other intracardiac lesion</td>
<td>1</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Coarctation of the aorta, patent ductus arteriosus, and associated intracardiac lesions</td>
<td>6</td>
<td>15</td>
<td>6</td>
</tr>
</tbody>
</table>

---

**Table 4**

*Electrocardiographic Data on 60 Infants with Coarctation of the Aorta*

<table>
<thead>
<tr>
<th>Aortic arch anatomy and type of lesions</th>
<th>Total number</th>
<th>RAD</th>
<th>NAD</th>
<th>LAD</th>
<th>XRAD</th>
<th>RVH</th>
<th>CVH</th>
<th>LVH</th>
<th>WNL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anatomy</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Type I</td>
<td>15</td>
<td>10</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>10</td>
<td>1</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Type II</td>
<td>37</td>
<td>29</td>
<td>5</td>
<td>0</td>
<td>3</td>
<td>23</td>
<td>10</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Type III</td>
<td>8</td>
<td>5</td>
<td>0</td>
<td>0</td>
<td>3</td>
<td>2</td>
<td>2</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Coarctation only</td>
<td>15</td>
<td>13</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>12</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Associated cardiac lesions</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Single</td>
<td>20</td>
<td>14</td>
<td>2</td>
<td>1</td>
<td>3</td>
<td>12</td>
<td>5</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Multiple</td>
<td>25</td>
<td>17</td>
<td>3</td>
<td>1</td>
<td>4</td>
<td>14</td>
<td>7</td>
<td>2</td>
<td>2</td>
</tr>
</tbody>
</table>

Abbreviations: RAD = right axis deviation (QRS frontal axis between +90° and +180°); NAD = normal axis deviation (0° to +90°); LAD = left axis deviation (0° to −90°); XRAD = extreme right axis deviation (+180° to −90°); RVH = right ventricular hypertrophy; CVH = combined ventricular hypertrophy; LVH = left ventricular hypertrophy; WNL = within normal limits.
Table 5

Electrocardiographic Data on 26 Surviving Infants with Coarctation of the Aorta

<table>
<thead>
<tr>
<th>Axis</th>
<th>RAD</th>
<th>NAD</th>
<th>XRAD</th>
<th>IND</th>
<th>Ventricular pattern</th>
</tr>
</thead>
<tbody>
<tr>
<td>Initial</td>
<td>21</td>
<td>4</td>
<td>1</td>
<td>0</td>
<td>RVH 15, CVH 7, LVH 4, WNL 0</td>
</tr>
<tr>
<td>Follow-up*</td>
<td>9</td>
<td>15</td>
<td>1</td>
<td>1</td>
<td>RVH 8, CVH 9, LVH 6, WNL 3</td>
</tr>
</tbody>
</table>

For abbreviations see table 4.
* Follow-up period: mean, 35 mo; range 2½ to 76 mo.

relation to the transverse arch anatomy or the various associated cardiac malformations (table 4).

Only two infants showed left ventricular hypertrophy (LVH) patterns before 3 months of age. One of these (L.P.) had VSD, PDA, and, surprisingly, marked pulmonary hypertension. The other (R.R.) had mitral insufficiency in addition to coarctation of the aorta.

Four infants more than 3 months of age showed an LVH pattern at the initial examination. One of these had isolated coarctation. One patient had an additional aortic stenosis, the third patient had mitral insufficiency, and the fourth infant had VSD and PDA.

Electrocardiograms were available in three infants with isolated tubular hypoplasia. Right

Figure 5

Electrocardiograms of three infants with isolated tubular hypoplasia and no coarctation of the aorta.

_Circulation, Volume XL, September 1969_
axis deviation was noted in all: two had combined ventricular hypertrophy, and one had RVH (fig. 5).

Analysis of serial electrocardiograms in 26 surviving infants (table 5) showed significant changes with increasing age. In infants with isolated coarctation the frontal QRS axis tended to return toward normal from right axis deviation. The RVH pattern in the precordial leads was slowly replaced by incomplete right bundle-branch block and the left ventricular potentials became more prominent in eight of 11 infants. However, in three infants with isolated coarctation, right ventricular dominance has persisted even after 3 years of age. Infants with associated cardiac lesions did not show any characteristic evolving diagnostic pattern in the electrocardiogram with progression in age.

Cardiac Catheterization

Right and left heart catheterization and cineangiocardiograms were performed on 44 infants with physiologic data from 31 infants being considered satisfactory for analysis. Pulmonary hypertension (pulmonary artery peak systolic pressure greater than 50 mm Hg) was present in all seven infants with severely hypoplastic transverse arch, in 20 of 27 (74%) with moderately hypoplastic arch, and in five of the 10 infants (50%) whose transverse aortic arch was normal. All infants with a shunting lesion had pulmonary hypertension, but only one third of those (four of 12) with isolated coarctation of the aorta had abnormally elevated right heart pressure. Three of these four infants with isolated coarctation and pulmonary hypertension had a moderately hypoplastic transverse arch (type II) and one had a transverse arch of normal caliber (type I). The latter infant (B.C.) had a pulmonary artery pressure of 67/35 mm Hg and a very high (estimated) pulmonary vascular resistance of 17 units at 4 weeks of age. Catheterization studies in this infant 1 year later revealed normal pulmonary artery pressure and pulmonary vascular resistance.

Six infants with coarctation of the aorta and PDA were catheterized. Only left-to-right shunting through the ductus arteriosus was noted by oxygen saturation data or cineangiocardiogram. Fifteen infants with PDA and VSD were catheterized. Definite evidence of a bidirectional shunt across the ductus arteriosus either by cineangiography or oxygen saturation data was noted in one case with type I transverse aortic arch, in one with type II, and in two with type III aortic arch. A small bidirectional shunt across the ductus arteriosus may have been present in three others with coarctation of the aorta, PDA, VSD, and type III transverse aortic arch, but the physiologic studies were considered inadequate for proof.

Two infants with isolated tubular hypoplasia and no segmental coarctation were catheterized. One had moderate hypoplasia and the other, severe hypoplasia of the transverse aortic arch. In both cases the ductus arteriosus could not be visualized or traversed at the time of catheterization, but the ductus was narrowly patent at autopsy. One of these infants had a VSD, and the other, aortic stenosis and a small ASD. Pulmonary hypertension and high estimated pulmonary vascular resistance were noted in both.

The diagnosis and survival of the 44 infants with physiologic studies are related to the estimated pulmonary vascular resistance in figure 6. Higher pulmonary vascular resistance was noted in infants with coarctation of the

| AORTIC ARCH | CO AORTA (1) | CO AORTA WITH SINGLE LESION (5) | CO AORTA WITH MULTIPLE LESIONS (12) | PERCENT PVR>5
|-------------|-------------|-------------------------------|----------------------------------|----------------
| NORMAL      |             |                               |                                  | 40%
| MODERATELY HYPOPLASTIC |             |                               |                                  | 76%
| SEVERELY HYPOPLASTIC |             |                               |                                  | 55%
| TOTAL PERCENT>5 |             |                               |                                  | 62%

Figure 6

Estimated pulmonary vascular resistance in units related to (1) type of coarctation (isolated or associated lesions), (2) type of aortic arch pathology (normal or hypoplastic), and (3) survival.

Circulation, Volume XL, September 1969
aorta and additional cardiac lesions, single or multiple. Overall survival was significantly lower in the group with elevated pulmonary vascular resistance.

Clinical Course and Management

Clinical data were available in 64 patients, 60 with coarctation of the aorta and four with isolated tubular hypoplasia. All but three of these infants were seen in congestive heart failure between the age of 1 day and 180 days of life (fig. 7). The three infants, first seen after 180 days of life, were included because of a definite history of onset of symptoms during the first 6 months of life. Of the 64 patients, 16 were hospitalized by the seventh day of life, and 32 by the twentieth day of life. Mortality was high among symptomatic infants needing early hospitalization, and there was no survival of infants admitted before they were 5 days old. The overall mortality (surgical and medical) in the entire group was 60% (38 of the 64).

Infants with coarctation of the aorta and an additional single cardiac defect had a mortality of 60% (11 of 18 patients). The mortality was 76% (21 of 27) in those with associated multiple cardiac lesions. Infants with isolated coarctation had a mortality of 27% (four of 15). Infants with coarctation and additional cardiac lesions were hospitalized relatively earlier in life, and no survival was noted among those admitted before 5 days of age (fig. 8).

Nonoperative management was employed with vigorous anticongestive measures in 26 of the 64 infants whose clinical data were analyzed. Survival was 31% (eight of 26) (fig. 9). Ten infants, including one with isolated tubular hypoplasia, died following early surgical intervention for noncardiac abnormalities (surgery type H, fig. 9).

Operative management of the cardiac problem was elected in the remaining 28 infants. The survival in this group was 64% (18 of 28). Eight infants with isolated coarctation underwent resection of the coarctation with six surviving. Nine infants had resection of the coarctation with division and closure of the PDA. Eight of these survived. Four of six infants with coarctation, VSD, and PDA survived surgical resection of the coarctation, ductus division, and pulmonary artery banding.

Circulation, Volume XL, September 1969
The outcome of surgical and nonsurgical management was further analyzed (fig. 10) in relation to the type of transverse arch pathology and other additional cardiac defects. The infants are grouped according to the transverse arch pathology. Each group is further subdivided as having isolated coarctation (O), coarctation with a single cardiac lesion (S) and coarctation with multiple cardiac lesions (M).

The five infants with isolated coarctation and a normal transverse aortic arch all survived with either surgical or medical management. Among the 10 patients with isolated coarctation and hypoplastic aortic arch the overall survival was less (60%). No infant with coarctation and associated single or multiple cardiac malformations survived nonsurgical management regardless of the transverse arch pathology. With surgical treatment, however, the survival in this group was encouraging since all infants with normal transverse aortic arches survived and 11 of the 14 infants (79%) with moderately hypoplastic arch survived.

Discussion

Our clinical material indicates that coarctation of the aorta presenting with congestive heart failure in early infancy is characterized by a wide spectrum of anatomic and physiologic features. Anatomically, coarctation as defined by Edwards and associates is a localized, narrow, constrictive lesion which presents histologically as an infolding of the aortic media into the lumen. In our series, 71 of 78 infants with an initial clinical diagnosis of coarctation of the aorta had this localized constriction. By contrast, tubular hypoplasia is

Figure 9

Results of medical and surgical management of 64 infants with coarctation of the aorta. Percentages of survival are given according to mode of management. Various types of surgical approach indicated on the abscissa (A to H). Non-S = medical management; Co. aorta = coarctation of the aorta; PDA = division of patent ductus arteriosus; PAB = pulmonary artery banding; Others = surgery for non-cardiac abnormalities.

Figure 10

Survival following medical management or surgical management as related to the diagnostic subgroups and the transverse aortic arch pathology in infants with coarctation of the aorta. Surgery: A = resection of coarctation; B = resection of coarctation and division and suture of patent ductus arteriosus; C = resection of coarctation, ductus arteriosus division and pulmonary artery banding. Diagnosis: O = isolated coarctation of the aorta; S = coarctation with single cardiac lesions; M = coarctation of the aorta with multiple cardiac lesions. (See text for comments and types of aortic arch pathology.)

Circulation, Volume XL, September 1969
a diffuse narrowing involving a long segment of the transverse aortic arch which does not involve the aortic media. Tubular hypoplasia as an isolated lesion was found only in seven infants. It is particularly significant to note, however, that 55 of the 78 infants had a combination of both of these elements of aortic pathology, that is, proximal tubular hypoplasia of the transverse arch ending with a localized constrictive coarctation.

In an extensive review of pathologic and surgical material, Lev has used the term "converted adult coarctation" to describe this combination of lesions. The concept of conversion implies that tubular hypoplasia, with changing physiologic factors in some infants, may convert through a transitional stage into a localized narrow zone of stenosis (coarctation). Adequate clinical evidence is not available to document this longitudinal evolution. Accordingly, we prefer to describe these two types of aortic pathology as separate entities, occurring either alone or together as noted in our pathologic and angiocardiographic material.

Infants with coarctation of the aorta and congestive heart failure in early infancy present a difficult clinical problem and have a high mortality as noted by several authors (table 6). Our clinical material consisted of infants with coarctation of the aorta, confirmed by autopsy or angiography, who were hospitalized because of congestive heart failure during the first 6 months of life. These infants form a heterogeneous group managed differently and by different surgical and medical personnel over a considerable period. Analysis of the clinical course indicated that infants with coarctation of the aorta and additional cardiac lesions became symptomatic earlier and had a considerably higher mortality than infants with isolated coarctation.

It is pertinent that 55 of the 71 infants (78%) with coarctation and early symptoms and signs of congestive heart failure showed moderate to severe tubular hypoplasia in addition to segmental coarctation. By contrast, only five infants of the entire group had isolated coarctation of the aorta, a normal transverse aortic arch, and no other additional lesion (table 1). Our material clearly demonstrates that the vast majority of infants with coarctation of the aorta who develop congestive heart failure have an additional cardiac defect, usually a significant shunting lesion, and associated tubular hypoplasia of the transverse aortic arch.

Coarctation of the aorta in early infancy can be associated with varied physiologic responses. Pulmonary hypertension and increased pulmonary vascular resistance were noted invariably in those infants who had coarctation of the aorta associated with significant shunting lesions. However, four infants with isolated coarctation of the aorta also had elevated pulmonary artery pressures, and in three of these the estimated pulmonary vascular resistance was high. Such pulmonary hypertensive responses in infants with isolated coarctation have also been documented by others.

Table 6

Mortality Among Patients with Coarctation of the Aorta in the First Year of Life as Reported in Literature

<table>
<thead>
<tr>
<th>Surgical mortality</th>
<th>Nonsurgical mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.</td>
</tr>
<tr>
<td>Glass et al. 7</td>
<td>1960</td>
</tr>
<tr>
<td>Mortensen et al. 8</td>
<td>1959</td>
</tr>
<tr>
<td>Freundlich et al. 9</td>
<td>1961</td>
</tr>
<tr>
<td>Malm et al. 10</td>
<td>1963</td>
</tr>
<tr>
<td>Adam et al. 11</td>
<td>1965</td>
</tr>
<tr>
<td>Lindesmith et al. 12</td>
<td>1966</td>
</tr>
<tr>
<td>Hartman et al. 13</td>
<td>1967</td>
</tr>
</tbody>
</table>
The electrocardiographic findings in infants less than 3 months of age did not correlate well with the different anatomic and physiologic features of coarctation or the associated cardiac lesions. Most infants less than 3 months of age showed right ventricular preponderance irrespective of the associated intracardiac lesions and the transverse arch anatomy. At times the electrocardiogram presented a misleading reflection of the physiologic status. Thus, the left ventricular preponderance in an infant with coarctation, VSD, and PDA did not reflect the systemic pressure in the pulmonary artery and the high estimated pulmonary vascular resistance (fig. 11). The right ventricular hypertrophy evident in the electrocardiogram of an infant with isolated coarctation of the aorta was difficult to reconcile with the relatively low pulmonary artery pressure and the severe systemic hypertension (fig. 12).

Management of the infant with coarctation of the aorta and congestive heart failure has been a matter of controversy. In our series, survival with anticongestive measures alone was 31% (8 of 26 patients). In contrast, our operative management of the cardiac problem resulted in 64% (18 of 28) survival.

There was no survival with medical management alone in infants with coarctation and additional cardiac defects, irrespective of the transverse arch anatomy. In contrast, with surgical intervention, survival was 78% (18 of 23) in this infant group with additional cardiac defects. The only infant with a severely hypoplastic arch who survived was surgically treated. Although this study has been retrospective in nature, the striking difference between the results of medical and surgical management appears conclusive. In agreement with others, we would recommend that early surgical intervention is indicated in the presence of significant associated shunting cardiac lesions.

There is a further controversy concerning the mode of management of infants with isolated coarctation of the aorta. In this group initial medical management alone is frequently successful. If early surgery is elected, the possibility of residual or recurrent segmental obstruction requiring subsequent additional surgery must be considered. Additional long-term follow-up studies are needed to indicate the cause and frequency of this problem. In our material, seven of the 18 surviving infants treated surgically have evidence of recurrent aortic obstruction. If infants with isolated coarctation can be managed successfully by anticongestive measures, it would seem advisable to postpone surgery until a more...
satisfactory age is reached, 3 to 8 years, when the reconstructed lumen is more likely to prove adequate for adult life.22

Acknowledgment

We gratefully acknowledge the surgical contributions to the care of these infants by Drs. Willis J. Potts, William Riker, Arthur DeBoer, Thomas G. Baffes and Farouk Idriss.

References

1. Morgagni, J. B.: De Sedibus et Causis Morborum. Epist. 18, Article 6, 1760.

Electrocardiogram and pertinent physiologic data of an infant (W.W., 16 days old) with coarctation of the aorta. AO = ascending aorta; MPA = main pulmonary artery; PVR = estimated pulmonary vascular resistance. The electrocardiogram does not reflect the severe systemic hypertension.

<table>
<thead>
<tr>
<th>Indicator</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>AO mm Hg</td>
<td>195/80</td>
</tr>
<tr>
<td>MPA</td>
<td>85/40</td>
</tr>
<tr>
<td>PVR units</td>
<td>7</td>
</tr>
</tbody>
</table>

Figure 12


