Hemodynamics of Aortic Valve Atresia

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SUMMARY
Hemodynamic data on 12 infants with aortic valve atresia, ranging from 1 to 3 mo of age, are presented. Peripheral arterial oxygen saturations ranged from 19 to 93%. The lowest saturations were found in patients with closed foramen ovales. Increasing age, increasing pulmonary vascular resistance, and a decrease in the pulmonary-to-systemic blood flow ratio all correlated with decreasing systemic arterial oxygen saturation. In seven of the nine infants under 1 mo of age, systemic arterial saturations ranged from 81 to 93%. Thus, we feel that the classification of this lesion as a form of cyanotic congenital heart disease is misleading and prefer to classify aortic valve atresia as an obligatory admixture lesion. Criteria for selection of patients for surgery are discussed and the minimum amount of hemodynamic data needed for such selection is outlined.

Additional Indexing Words:
Catheterization   Congenital heart disease   Ductus arteriosus   Foramen ovale
Obligatory admixture lesion   Pallor   Peripheral pulse
Surgical anastomosis of aorta to pulmonary artery

Although congenital aortic valve atresia is not a rare anomaly and accounts for slightly more than 1% of cases of congenital heart disease,1–4 cardiac catheterization data in this entity has rarely been reported. Indeed, reference to only four catheterized cases is found in the literature.5–8 This paucity of catheterization data is readily understandable since the average age of death is 4 to 5 days.5 Furthermore, the majority of these infants die within 48 hours of recognition.5 Our own experience has been similar. Of 17 infants with aortic valve atresia, five died prior to cardiac catheterization. In addition, six died within the immediate 24 hours while only six survived more than 24 hours following catheterization.

Thus, these infants are extremely fragile and at times are moribund at the time of admission. Catheterizations or angiography, or both are often kept to a minimum compatible with reaching a diagnosis and are not usually hemodynamically oriented. Because a surgical procedure recently has been proposed for these children,9 we have analyzed our catheterization data in the hopes of learning more about the selection of patients for such surgery.

Methods
Seventeen infants with aortic valve atresia were seen at our two institutions. This survey covered the years 1960 to 1969 at the University of Florida and 1964 to 1969 at the Johns Hopkins Hospital. Table 1 shows the distribution of patients according to institution and catheterization status. The 12 infants who underwent
cardiac catheterization form the basis of this
report. In 10 infants the diagnosis was confirmed
by postmortem examination (table 2).

Since injections of radiopaque contrast material
are known to cause significant hemodynamic
changes,10 data obtained following angiocardio-
draphy were used in only one instance (case 2). In
this infant, the measurements used were recorded
28 min after the angiogram.

As can be seen from table 3, varying amounts
of data were available for these patients. Oxygen
consumption was estimated by using a previously
reported regression equation.11 In one instance,
case 8b, oxygen consumption was measured by a
flow-through technic.* Wessel and associates12
used a similar method which they described in
detail. Oxygen saturations were determined by
using a Waters Xc50 cuvette and its associated
amplifier† (nine cases), an American Optical
oximeter‡ (two cases), or by the Van Slyke-Neill
method (one case). Saturations were measured
while breathing room air in all except case 7 in

whom 100% oxygen was being given by mask. In
the five instances in which pulmonary venous
oxygen saturation was not determined, it was
assumed to be 97%.

Results

As shown in table 3, right heart pressures
were markedly elevated. Pulmonary artery
mean pressure was equal to or exceeded
systemic arterial mean pressure in all seven
infants in whom both were measured. The
maximum difference occurred in one of the
youngest patients studied who had a mean
pulmonary artery pressure 14 mm Hg higher
than the systemic artery mean. This suggests
that the duc tus arteriosus was slightly
constricted in these patients. In the necropsied
patients, the duc tus arteriosus did not show
any evidence of closing.

Figure 1 illustrates the systemic arterial
oxygen saturations at various ages. With the
exception of the two patients with a closed
foramen ovale, systemic arterial saturations
tended to decrease with age (r = -0.70). A

Table 1

Distribution and Fate of Infants with Aortic Valve Atresia

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Died before catheterization</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Catheterized and died within 24 hr</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>Catheterized and survived more than 24 hr</td>
<td>4</td>
<td>2</td>
</tr>
</tbody>
</table>

Table 2

Postmortem Findings in Infants with Aortic Valve Atresia and Previous Cardiac Catheterization

<table>
<thead>
<tr>
<th>Case</th>
<th>Mitral valve</th>
<th>Foramen ovale</th>
<th>Ductus arteriosus</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>Hypoplastic</td>
<td>Closed</td>
<td>*</td>
</tr>
<tr>
<td>2</td>
<td>Hypoplastic</td>
<td>Widely patent</td>
<td>Massively dilated</td>
</tr>
<tr>
<td>3</td>
<td>Hypoplastic</td>
<td>Probe patent</td>
<td>Large</td>
</tr>
<tr>
<td>4</td>
<td>Hypoplastic</td>
<td>Large opening</td>
<td>Large</td>
</tr>
<tr>
<td>5</td>
<td>Hypoplastic</td>
<td>Patent</td>
<td>*</td>
</tr>
<tr>
<td>7</td>
<td>Hypoplastic</td>
<td>Patent</td>
<td>Widely patent</td>
</tr>
<tr>
<td>9</td>
<td>Hypoplastic</td>
<td>3-mm opening &amp;</td>
<td>Large</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1-cm primum ASD</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>Hypoplastic</td>
<td>3-mm opening</td>
<td>Large</td>
</tr>
<tr>
<td>11</td>
<td>Hypoplastic</td>
<td>Probe patent</td>
<td>*</td>
</tr>
<tr>
<td>12</td>
<td>Hypoplastic</td>
<td>2-mm opening</td>
<td>Large</td>
</tr>
</tbody>
</table>

*Not mentioned in protocol.
 Abbreviation: ASD = atrial septal defect.

†Waters Corporation, Rochester, Minnesota.
‡American Optical Co., Bedford, Massachusetts.

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Table 3

Summary of Catheterization Data

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (days)</th>
<th>Sex</th>
<th>Ht (cm)</th>
<th>Wt (kg)</th>
<th>Heart rate</th>
<th>Flow (L/min)</th>
<th>Pressures (mm Hg)</th>
<th>Art. sat. (%)</th>
<th>Peripheral pulses</th>
<th>Autopsy</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>RA mean</td>
<td>RV mean</td>
<td>PA mean</td>
<td>LA mean</td>
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<tr>
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<td>51</td>
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<td>110</td>
<td>2.02</td>
<td>0.35</td>
<td>61</td>
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<td>1</td>
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<td>48</td>
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<td>93</td>
<td>70</td>
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<td>6</td>
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<tr>
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<td>2.78</td>
<td>0.80</td>
<td>0</td>
<td>72</td>
<td>6</td>
</tr>
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</table>

*Pressures measured during withdrawal from aorta through patent ductus arteriosus to pulmonary artery or vice versa.
†Breathing 100% oxygen by mask.
‡Patient 8a was recatheterized and is also listed as patient 8b.
§Systemic and pulmonary blood flows were calculated from measured oxygen consumption; in all other instances oxygen consumption was estimated from a linear regression equation.11
**Simultaneous measurements.11

Abbreviations: S = systolic; D = diastolic; E = end-diastolic; N = normal; Decr. = decreased; Inc. = increased.
slightly better correlation was noted in the eight patients for whom it was possible to plot arterial oxygen saturation against the ratio of pulmonary blood flow to systemic blood flow ($r = 0.77$; fig. 2). Infants with the highest pulmonary-systemic blood flow ratios had the highest arterial oxygen saturations. Arterial oxygen saturation was inversely related to pulmonary vascular resistance ($r = -0.76$; fig. 3); that is, the higher the pulmonary resistance, the lower the arterial saturation. This is probably the common denominator for the other two relationships noted above, with increasing age being associated with increasing pulmonary vascular bed damage and increasing vascular resistance in turn decreasing the pulmonary blood flow.

One of the striking physical findings in aortic valve atresia is the poor quality of the peripheral pulses, especially in the presence of severe congestive heart failure. This is presumably due to a combination of lower than normal cardiac output with a resulting

**Figure 1**

Systemic arterial oxygen saturations plotted against age in our 12 catheterized infants and eight reported in the literature; one of the eight was a catheterized infant whose case was reported by Sánchez-Gonzalez and Espino-Vela, and the other seven had oxygen saturation measured by earpiece oximetry. With the exception of two patients with anatomically proved closed foramen ovale (case 1 in our group and the Sánchez-Gonzalez case7), there is an inverse correlation with age ($r = -0.70$).

**Figure 2**

When systematic arterial oxygen saturations are plotted against the ratio of the pulmonary blood flow-systemic blood flow, there is an exponential increase with increasing pulmonary flow.

**Figure 3**

Arterial oxygen saturation is plotted against pulmonary vascular resistance. Although only six patients were available for these calculations, reasonable correlation is evident between increasing pulmonary resistance and decreasing arterial saturation ($r = -0.76$).

**Figure 4**

Mean blood pressure in patients with aortic atresia plotted against age. The dashed lines represent the average and plus and minus one standard deviation of normal children. Only two patients had blood pressures which were more than one standard deviation below the normal, and only the oldest patient clearly had systemic hypotension.

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low blood pressure. As shown in table 3 and figure 4, the systemic arterial mean pressure was clearly below normal in only one infant. Systemic blood flow was within normal limits in all eight infants in whom it could be estimated, but the large errors involved in using an estimate of oxygen consumption probably negates the value of this observation. In the one infant whose oxygen consumption was measured (case 8b), systemic blood flow was 91% of the predicted value.11

Figure 5 shows that the clinical estimation of the status of the peripheral pulses correlates best with the intra-arterial pulse pressure measurements. In the five infants with weak or decreased pulses, intra-arterial pulse pressures averaged 19 mm Hg, compared to an average of 32 mm Hg in five infants with normal pulses and 50 mm Hg in one patient with "bounding" pulses.

Discussion

Although this entity has been classified among the cyanotic forms of congenital heart disease, it is more properly classified among the obligatory admixture lesions.15 In this group of cardiac anomalies, both systemic venous blood and pulmonary venous blood must pass through a common mixing chamber before being distributed to the pulmonary vascular and systemic vascular beds.

More specifically, aortic atresia should fall into that group of admixture lesions in which there is a left-to-right atrial level shunt in the presence of peripheral arterial desaturation. This forms a very special group—the other common entities being total anomalous pulmonary venous connection, mitral (or left A-V valve) atresia, and atrial septal defects with single ventricle.

The amount of blood flowing through each vascular bed is determined primarily by the resistance of that bed. The other important factor would be size of the communication between the right and left atria. If the interatrial communication is inadequate in size, or absent, this may cause severe pulmonary venous obstruction and effectively decrease pulmonary blood flow. Other causes of pulmonary venous obstruction, for example, pulmonary vein stenosis or cor triatriatum, would also reduce pulmonary blood flow. No examples of these latter conditions were found in our series of patients. Thus, as long as there is an adequate-sized atrial level communication and pulmonary resistance is significantly lower than systemic resistance, pulmonary blood flow will be proportionally greater than systemic blood flow.

The pathophysiology of aortic valve atresia has not been fully documented by catheterization data. We recognize the vagaries involved in catheterizing seriously ill infants, particularly in the absence of a truly steady state. Fortunately, the technics of gathering these data during the past decade have been very similar in both institutions.11 Collection of the above data represents a step toward establishing the principles upon which management and surgery should be based.

Although pulmonary congestion is commonly seen, there is usually no difficulty in oxygenation of the blood in these infants and fully saturated blood returns from the pulmonary veins. In the five cases in which pulmonary venous oxygen saturations were
measured in this series, they ranged from 96 to 100%. There was an excellent correlation of systemic arterial saturation with the ratio of pulmonary-to-systemic blood flow. That is, as in other admixture lesions, the lower the pulmonary vascular resistance the greater the pulmonary blood flow and the higher the arterial oxygen saturation. Similarly, arterial oxygen saturation decreased with increasing age, probably related to increasing pulmonary vascular bed damage.

Since five of our patients, as well as three of the seven earpiece oximeter readings reported in the literature, showed arterial saturations ranging from 81 to 93%, overt cyanosis may not be present. Indeed, it has been our impression these infants more often exhibit the pallor associated with poor peripheral blood flow rather than an obviously cyanotic appearance.

The two infants with the lowest arterial oxygen saturations were shown at autopsy to have a closed foramen ovale, thus allowing mixing of blood only through myocardial sinusoids, coronary arteriovenous fistulae, or a levo-atriocardinal vein. In the oldest patient of this series we were able to introduce a catheter into the aortic root while a second catheter was in the right pulmonary artery. The right pulmonary artery saturation was 41% while the simultaneously obtained root of aorta saturation was 69%. Since there was no ventricular septal defect in this patient, the most likely explanation is that there were connections between the left ventricle and aortic root via myocardial sinusoids or a minute opening in the aortic valve which did not show on the cineangiogram. The patient died approximately 2 weeks after catheterization. Autopsy permission was not granted, and thus the exact mechanism remains speculative.

**Conclusions**

In one of our patients (case 1) and in a similar reported case the foramen ovale was shown by subsequent autopsy examination to be completely closed. In patient 3 the patent foramen ovale was found to be extremely tiny at autopsy. Since all three infants had rather low arterial oxygen saturations, and in fact among the lowest saturations of this study, it seemed logical that creating or enlarging the communication between the two atria might be beneficial. Accordingly, a Blalock-Hanlon procedure was carried out in two of these infants but both died within 24 hours of operation.

Recently, a new palliative procedure was proposed for infants with hypoplastic left heart syndrome. An anastomosis from the ascending aorta to the right pulmonary artery is constructed and both pulmonary arteries are banded. The ductus arteriosus is not disturbed. In the 11-day-old infant in whom this procedure was successfully completed, there appeared to be a large intra-atrial communication. Patients to be selected for such surgery should first be evaluated in order to be certain that (1) the ascending aorta is of adequate size for the anastomosis, and (2) that a large intra-atrial communication exists. One criterion proposed by Cayler and associates is that high pulmonary precapillary vascular resistance be present, does not appear to be a limiting factor in the operation since both pulmonary arteries are to be banded. Indeed, a more desirable state of the pulmonary vascular bed would be that it be capable of altering its resistance after the bands are in place. Cayler and associates also are concerned about the adequacy of coronary perfusion and the absence of a coarctation of the aorta at the junction of the hypoplastic transverse segment with the ductus arteriosus. In our small series we have not seen any examples of coronary artery anomalies or coarctation of the aorta. Since the anastomosis is created near the base of the ascending aorta and the ductus arteriosus is not ligated, coarctation of the aorta should not be a serious hemodynamic impediment.

Since these patients are usually in poor condition by the time cardiac investigation is done, the procedure is often limited to angiography. We believe that this does not furnish adequate information for rational selection of infants for surgery. Whenever the information to be gathered must be limited to
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a minimum, we suggest (1) determination of systemic arterial oxygen saturation while the infant is breathing room air, (2) a pressure pullback from left atrium to right atrium (our experience suggests that failure to enter the left atrium from an inferior approach strongly suggests that the foramen ovale is not patent), (3) securing all measurements prior to any injections of contrast material, and (4) keeping angiography to a minimum necessary to establish the diagnosis, perhaps limited to one aortogram.

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