CLINICOPATHOLOGIC CORRELATIONS

Coarctation of the Aorta Associated with Aortic Valvular Atresia

By Thomas J. Von Rueden, M.D., Laura Knight, M.D., James H. Moller, M.D., and Jesse E. Edwards, M.D.

SUMMARY
The aortas in 41 specimens of aortic atresia (ten with associated mitral atresia) were studied for the presence of coarctation of the aorta. The basic lesion, causing varying degrees of obstruction, was observed in 31 instances (75%), 24 minor, seven with major degrees of obstruction. In half, the aortic lesion lay proximal to the ductus, and in the majority of the remainder, opposite the ductus arteriosus. The location of lesion and degree of obstruction was essentially the same whether aortic atresia occurred alone or in association with mitral atresia. Angiography is a fairly reliable method of identifying coarctation associated with aortic atresia. An additional point of interest was that in aortic atresia alone there was strong dominance of the male sex, while this tendency was not observed when mitral atresia was associated with aortic atresia.

THE CLINICAL AND HEMODYNAMIC ASPECTS of aortic atresia have been extensively studied and reported.1-5 Although the pathologic details of the fundamental nature of this condition are known, the occurrence of coarctation of the aorta in association with aortic valvular atresia has been rarely described.1,3 In contrast to this experience, we found an extremely high incidence of coarctation of the aorta among 41 specimens with aortic valvular atresia, of which ten were associated with mitral atresia.

From the Cardiovascular Registry of United Hospitals-Miller Division, we reviewed the specimens of 41 instances of aortic valvular atresia. Particular attention was directed to the aortic arch and descending aorta for the presence or absence of coarctation of the aorta.

As used in this study, the term coarctation of the aorta refers to a localized medial deformity, previously described (fig. 1).6 Grossly, the medial deformity forms a ridge or curtain which protrudes toward the lumen of the aorta. As used, the term coarctation identifies the medial deformity which varies from case to case in the degree of obstruction it causes (fig. 2). In cases with significant obstruction, the superior aspect of the aorta corresponding to the medial abnormality shows a localized concavity (fig. 3). When the degree of obstruction is of minor degree, only the ridge is noted internally, while there is no external deformity of the aorta.

Observations
We divided the patients with coarctation into two groups, one showing a major degree of obstruction to the aorta and the other showing the lesion but with little or no significant obstruction, the coarctation being classified as minor.

Among the 41 specimens there were 31 in which aortic valvular atresia was associated with a mitral valve intact and ten others in which mitral valvular atresia was also present. There was a difference in sex distribution between these two categories (table 1). Among the 31 cases with aortic atresia alone, there were 27 male and three female patients, and in one the sex was unknown to us, while among the ten with coexistent mitral and aortic atresia there were five male and five female patients. Thirty of the patients had died by 13 days of age and the other two at the ages of one and two months, respectively.

Coarctation of the aorta, as defined, was found in 31 of the 41 cases. In 24 the coarctation was of minor degree, while in the other seven it was of major degree
causing significant aortic obstruction (table 1). There was no difference in the distribution of coarctation among those with only aortic atresia (74%) and those with coexistent mitral and aortic atresia (80%).

As to the location of the coarctation with respect to the aortic ostium of the ductus, there was no significant difference between aortic atresia alone as compared with coexistent aortic and mitral atresia. Among the 31 specimens with some degree of coarctation, 30 were suitable for study regarding this point. In 15 of the 30 cases the coarctation was located proximal to the ductus, in 12 opposite the ductus, and in three distal to the ductus. There was no significant difference between major and minor degrees of coarctation relative to its ductal position.

Of the 16 cases with angiographic studies (fig. 4), there were three in which coarctation was only identified later and not on the basis of the angiogram (false negatives). In two of these the coarctation was minor and, in one, major.

Among the 13 cases in which angiographic findings corresponded with later pathologic findings, there were four cases without coarctation and nine with coarctation. Of the latter, the coarctation was minor in six and major in three, as judged pathologically.

Comment

We found a very high incidence (75%) of the deformity of coarctation of the aorta in cases of aortic valvular atresia. Although an association between these two conditions has been mentioned in two earlier reports,¹ ³ neither gives the incidence of this association.

Of 77 subjects under six months of age at the time of death with coarctation of the aorta, Becker and associates⁷ found nine cases of aortic atresia.

An association between congenital bicuspid aortic valve and coarctation of the aorta is well known, the valvular deformity occurring in about 50% of cases with coarctation. A relationship between abnormalities of the aortic valve and coarctation of the aorta appears to apply for aortic atresia also. The association between malformation of the aortic valve and coarctation of the aorta suggests a developmental relation-

![Photomicrographs in two cases with coarctation associated with aortic atresia. In each, the superior wall of the aorta is sectioned longitudinally and shows a medial curtain protruding toward the lumen (below). a) Grossly, this lesion was considered to have caused minor degrees of aortic obstruction. Elastic tissue stain; × 16. b) The coarctation was major. Aortogram from this case is shown in figure 4c. Elastic tissue stain; × 23.]
COARCTATION WITH AORTIC ATRESIA

Figure 2
Gross specimens of interior of aorta in two examples of coarctation of the aorta associated with aortic atresia. a) Opposite the ductus (D.) is a ridge within the aorta (arrow) projecting into the lumen and causing a minor degree of aortic obstruction. The fold proximal to the identified ridge is an artifact. A.A. = ascending aorta; P.T. = pulmonary trunk. b) Proximal to the entrance of the ductus (D.), the aorta shows a curtain-like projection (arrow) into the lumen causing a major degree of aortic obstruction. L.S. = left subclavian artery.

Figure 3
External view of the great vessels in an example of major degree of coarctation associated with aortic atresia. Opposite the aortic entrance of the ductus arteriosus (D.) is a depression (arrow) in the superior aspect of the aorta. P.T. = pulmonary trunk; A = hypoplastic ascending aorta.

The ramification of the high incidence of coarctation of the aorta with aortic valvular atresia may be important if eventually a corrective or palliative operative procedure is devised for the treatment of aortic valvular atresia.

An additional point of interest is our confirmation of the different sex distribution in aortic atresia as compared with coexistent aortic and mitral atresia. Isolated aortic atresia occurred primarily in male patients, while combined aortic and mitral valvular atresia occurred equally in male and female patients.

Table 1
Association of Minor and Major Degrees of Coarctation with Aortic Atresia

<table>
<thead>
<tr>
<th>Coarctation</th>
<th>Valvular atresia</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Aortic only</td>
</tr>
<tr>
<td>Present</td>
<td>23</td>
</tr>
<tr>
<td>Minor</td>
<td>18</td>
</tr>
<tr>
<td>Major</td>
<td>5</td>
</tr>
<tr>
<td>Absent</td>
<td>8</td>
</tr>
<tr>
<td>Total cases</td>
<td>31</td>
</tr>
</tbody>
</table>

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Figure 4
Aortograms in three cases of aortic atresia with coarctation. a) Original reading of this aortogram had not identified coarctation, while the pathologic specimen showed a major degree of obstruction as shown in figure 3. In retrospect, absence of opacification of the descending aorta was probably caused by the coarctation and led to the false diagnosis of no coarctation. b) Coarctation identified. The pathologic specimen showed a minor degree of obstruction of the aorta. c) The coarctation is clearly identified, and the specimen confirmed the presence of major coarctation. Photomicrograph from this case shown in Figure 1b.

References
Coarctation of the aorta associated with aortic valvular atresia.
T J Von Rueden, L Knight, J H Moller and J E Ewards

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