Coarctation of the Aorta Complicated by Patency of the Ductus Arteriosus

Physiologic Considerations in the Classification of Coarctation of the Aorta

By Arnold L. Johnson, M.D., Charlotte Ferencz, M.D., F. W. Wiglesworth, M.D., and Donald L. McRae, M.D.

The study of the anatomic and hemodynamic relationships in a case of coarctation of the aorta complicated by patency of the ductus arteriosus and pulmonary hypertension indicated the necessity for a physiologic approach to the classification of coarctation of the aorta. The origin of existing classifications has been briefly traced and basic anatomic and physiologic considerations in this malformation have been reviewed. A classification is presented which takes into account the state of the ductus arteriosus, its relationship to the coarcted segment and the influence of the collateral circulation. The microscopic appearance of the pulmonary vessels is included.

The exact diagnosis of coarctation of the aorta is now a matter of great practical importance. No particular diagnostic problem is involved in the usual “adult” type of coarctation. There are, however, cases which are complicated by the presence of a patent ductus arteriosus. Such a complication may present diagnostic difficulties of which the case presented below is illustrative. The purpose of this paper is to discuss certain hemodynamic relationships in these cases, the means involved in diagnosis and to suggest a scheme of classification arising out of these considerations.

The classification of coarctation of the aorta has always presented considerable difficulty. The emphasis has been placed upon anatomic and embryologic considerations. It is apparent, however, that physiologic adjustments, dependent upon various arrangements of the ductus arteriosus and collateral circulation, modify the effect of the area of stenosis. Thus a classification based not only upon anatomic, but also upon hemodynamic considerations may be useful in the study of this group of malformations.

Coarctation of the aorta occurs in the region of the junction of the fourth and sixth embryonic arches, represented by the arch of the aorta and by the ductus arteriosus, respectively. Above the ductus arteriosus, lying between it and the left subclavian artery, is an important segment of the aorta through which, in fetal life, very little blood flows. Left ventricular blood is directed to the head and upper extremities above this segment, and right ventricular blood flows to the trunk and lower extremities through the ductus below it. This portion of the aorta, known as the isthmus, becomes functional when the ductus closes and it then serves to connect the cephalic and caudal arterial circulations. During fetal life the isthmus is narrow and dilates gradually after the blood flow through it is established.

These basic facts were considered by early writers attempting to clarify the pathogenesis of coarctation of the aorta. Bonnet, Hamilton and Abbott and Blackford have reviewed the various theories and it appears that these followed along three main lines of thought. Many workers favored the view that the condition was due to persistence of excessive narrowing of the isthmus and some thought that this process might be influenced by traction of the obliterating ductus. Others believed that the abnor-
mality occurred during the process of joining of the embryonic arches. The third group suggested that closure of the ductus arteriosus was responsible for the development of the condition. It was believed that a thrombus might extend from the aortic orifice of the ductus into the aorta, or that the “peculiar” tissue might either be present in the aortic wall or extend to it from the ductus and by its contraction promote an area of stenosis in the aorta.

Considering these various theories, Bonnet concluded that the existent controversy resulted from the failure to recognize two main classes of coarctation: (1) the “infantile” type, representing a persistence of the fetal condition of the isthmus, and (2) the “adult” type, being related to closure of the ductus arteriosus.

This classification clarified the study of coarctation considerably and was widely adopted. It became apparent, however, that many cases could not be allocated satisfactorily to the categories provided, and further classifications based on anatomic considerations, have been presented. Little emphasis has been placed upon important adaptive changes which significantly alter the hemodynamic relationships. Taussig pointed out the importance of the ductus arteriosus in affecting the nature of the anomaly. If, in addition, the collateral circulation is considered, it appears that a useful differentiation of types would depend upon the state of the ductus, its entry into the aorta relative to the site of coarctation, and upon the state of the collateral circulation.

This view was suggested by the correlation of hemodynamic and necropsy findings in a case of coarctation of the aorta associated with a patent ductus arteriosus. This case possessed the characteristics of both Bonnet’s “adult” and “infantile” types anatomically, but physiologically was almost entirely of the “adult” variety. The findings which initiated our interest in this problem are presented below.

**Case Report**

The presenting complaint of this 91 year old white girl was a considerable limitation of activity due to dyspnea ever since she learned to walk. There was no cyanosis or clubbing. There was no difference between the color of the fingernail and toenail beds. There was a harsh systolic murmur of moderate (grade III) intensity maximal in the fourth left intercostal space, transmitted particularly well to the left axilla and back. The murmur was not prominent over the spine. In addition there was a continuous murmur, of slight intensity, heard over a fairly localized area in the second intercostal space close to the left sternal border. The blood pressure in the arms was 108/84. The femoral artery pulsations could just be felt and the blood pressure in the legs was 96/78. Pulsations of the intercostal vessels could be felt easily in the seventh to the last intercostal space bilaterally, but no pulsations were felt beneath the medial border of either scapula. The clinical diagnosis was coarctation of the aorta associated with a patent ductus arteriosus.

On fluoroscopic examination the heart appeared to be considerably enlarged in both the posteroanterior and oblique projections. The pulmonary artery and its branches were thought to be normal in size or slightly enlarged. The left ventricle was definitely enlarged. There was questionable evidence of notching of the fourth and fifth ribs bilaterally.

The electrocardiogram showed a right axis deviation in the standard leads. The unipolar precordial leads showed a pattern which was thought to be compatible with right ventricular hypertrophy.

A finding of considerable interest was the difference noted between the percentage oxygen saturation of the femoral artery blood and that in the upper part of the body. On two occasions the saturation of blood withdrawn from the femoral artery was found to be 91.6 per cent and 92.7 per cent. Employing the oximeter on the ear, three readings were each 98 per cent. It was felt that this represented a significant difference between the oxygen saturation of blood flowing to the upper extremities and that flowing to the lower extremities.

Heart catheterization studies revealed that the pressures in the right ventricle and in the pulmonary artery were greatly increased: right ventricle 90/16 and pulmonary artery 92/72. The oxygen content of blood samples in the right auricle and right ventricle was similar and thus there was no evidence of either an auricular or a ventricular septal defect (table 1). The oxygen content of blood from the main pulmonary artery was 0.7 volumes per cent greater than any sample from the right ventricle.

At one site, distal to the main pulmonary artery, the oxygen content was considerably higher and showed a value of 18.3 volumes per cent, similar to that obtained in the femoral artery. The position of the catheter tip could not be ascertained definitely. It was felt that it could be in a branch of the pulmonary artery or possibly have passed through a ductus into the descending aorta. The latter anatomic arrangement had been suggested by the finding of slight oxygen unsaturation of the femoral
artery blood. On the other hand, it was argued that if the ductus communicated with the descending aorta, then the unsaturation would have been much greater and there should have been cyanosis of the toenail beds.

It will be apparent shortly that insufficient attention was paid to the similarity of the pressure readings in the pulmonary artery and the femoral artery. The intrafemoral arterial pressure was 102/80, and the pressure in the pulmonary artery was 92/70. Unfortunately these were not simultaneous recordings.

The angiocardiograms showed the dye filling the right auricle and ventricle and passing into the lungs through a considerably enlarged pulmonary artery. There was no evidence of over-riding of the aorta. The cavity of the left ventricle appeared to be enlarged and its wall thickened. The curvature of the ventricular septum appeared to be convex toward the right ventricle. Dye was seen in the axillary arteries but none in the descending aorta or in the left gastric or renal arteries.

It had been quite evident on clinical grounds that this patient had a coarctation of the aorta with a well developed collateral circulation. It was also thought that a patent ductus arteriosus was present. The subsequent investigation then revealed findings which were not readily explained. The electrocardiogram was not in keeping with the left ventricular enlargement expected in coarctation and demonstrated in the angiocardiogram. It was, however, compatible with the right ventricular hypertrophy considered to be present after the high right ventricular and pulmonary artery pressures had been recorded during heart catheterization. This increased pressure was a surprising finding. Was it due to a primary lung condition or secondary in some way to the presence of the ductus?

At operation the site of the coarctation appeared to be quite localized and distal to the aortic end of the ductus. The left pulmonary artery and the very short ductus were obviously dilated. During the dissection, which was a matter of considerable difficulty, uncontrollable hemorrhage occurred from a tear in the left pulmonary artery.

**Autopsy Findings**

At postmortem examination the abnormal findings were confined to the cardiovascular system. The heart was moderately to markedly enlarged and weighed 300 Gm. as compared with the normal average weight of 155 Gm. The cardiac enlargement was due almost entirely to increase in the ventricular mass. The chamber of the right ventricle was not dilated, but that of the left ventricle showed moderate dilatation. The thickness of the right ventricular wall varied from 0.4 cm. to 1.0 cm. (normal 0.15 to 0.3 cm.). The thickness of the left ventricular myocardium varied between 0.4 cm. and 1.4 cm. (normal 0.3 to 1.1 cm.). It would thus appear that the right ventricle showed a relatively greater myocardial hypertrophy than the left.

The pulmonary and tricuspid valves were normal, the aortic valve was bicuspid, but otherwise was not remarkable, and the mitral valve showed a rare anomaly, a double orifice.

On external examination of the great vessels (fig. 1) the main pulmonary artery was much larger than the aorta, being of aneurysmal proportions.

---

**Table 1.—Data from Heart Catheterization**

<table>
<thead>
<tr>
<th>Artery</th>
<th>Oxygen (vols. %)</th>
<th>Pressure (mm.Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Femoral artery</td>
<td>18.3</td>
<td>102/80*</td>
</tr>
<tr>
<td>Capacity</td>
<td>19.9</td>
<td></td>
</tr>
<tr>
<td>% Saturation</td>
<td>92.7</td>
<td></td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>14.2</td>
<td></td>
</tr>
<tr>
<td>Upper right auricle</td>
<td>14.1</td>
<td></td>
</tr>
<tr>
<td>Mid right auricle at tricuspid</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Valve</td>
<td>14.0</td>
<td></td>
</tr>
<tr>
<td>Lower right ventricle</td>
<td>14.5</td>
<td></td>
</tr>
<tr>
<td>Mid right ventricle</td>
<td>14.7</td>
<td></td>
</tr>
<tr>
<td>Upper right ventricle</td>
<td>13.9</td>
<td>90/16</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>15.4</td>
<td>92/74</td>
</tr>
<tr>
<td>Query position (see text)</td>
<td>18.3</td>
<td>84/64</td>
</tr>
</tbody>
</table>

* The pressure was recorded the day following the cardiac catheterization.
The right and left pulmonary arteries were also enlarged. The circumference of the pulmonary artery was 9.5 cm. (normal 4.0 cm.) while the circumference of the ascending aorta was 4.2 cm. The proximal aorta was of normal circumference almost to the point of coarctation. It is interesting to note, however, that the innominate artery, the left common carotid, and the left subclavian arteries showed an increased circumference, averaging at least 0.5 cm. greater than controls. The descending aorta measured 2.5 cm. in circumference (control 3.0 cm.). The mouths of the intercostal arteries did not appear to be dilated. The circumference of the internal mammary arteries (0.9 cm.) equalled that of the common iliac arteries. There was thus evidence that the mammary arteries were dilated.

A short, broad patent ductus arteriosus arose from the beginning of the left pulmonary artery and was continuous with the descending aorta (fig. 1). In the fixed state it measured 0.4 cm. in length, and 2.0 cm. in circumference. The orifice of the coarcted segment of the aorta entered the ductus at its distal margin as it became continuous with the descending aorta. The stenosed orifice measured 0.1 cm. by 0.15 cm. in diameter. On the opposite wall of the ductus arteriosus was a slightly raised, yellowish, firm, rough lesion measuring 0.4 cm. in length by 0.5 cm. in width. Grossly and microscopically this was a "jet" lesion, apparently the result of the blood flow coming from the coarcted aorta.

A second jet lesion was present on the left side of the ridge separating the right and left pulmonary arteries. This measured 0.5 by 0.2 cm. As this lesion lay only on the left side, it was presumably created by a flow of blood from the ductus into the left pulmonary artery.

The arch of the aorta between the left common carotid and the left subclavian arteries was normal in circumference, but narrowed rapidly immediately distal to the origin of the left subclavian artery. The isthmus of the aorta measured approximately 0.4 cm. in length, and funnelled to the point of actual coarctation. The isthmus terminated by turning sharply inferiorly and fusing with the superior surface of the ductus. At the actual site of coarctation no diaphragm could be seen such as described by Edwards and co-workers. The opening was a simple stenosis caused by a uniform narrowing of the wall of the aorta (so-called infantile coarctation).

Microscopic examination of the lung tissue showed vascular lesions of moderate extent. Perhaps about one-third of the pulmonary vessels showed changes and they were more numerous centrally than peripherally. The large arteries showed no atherosclerosis and only occasional plaques of intimal fibrosis similar to that to be described.

The principal arteries involved were the medium and small type, varying from about 200 to 600 microns in diameter. The lesions were essentially intimal in situation and consisted of a concentric (occasionally eccentric) fibroplastic proliferation of the intima (fig. 2). The collagenous tissue either contained numerous nuclei or was relatively acellular. The lumens showed varying reduction in size up to complete obliteration. Reduplication of the internal elastic lamina was not a prominent feature and was mostly focal. The adventitia was widened by dense connective tissue.

*Fig. 2. Small pulmonary artery (about 200 μ) showing marked intimal fibrosis (inner four-fifths of wall thickness) and moderate atrophy of the media. The internal elastic lamina can be seen as a faint wavy line at top and right side. This vessel arose from a large artery and at its origin, 100 μ proximally, was histologically normal. H. & E. × 160.*

A striking feature was the definite elastosis of both large and small arteries. The elastic fibers were thick and in stained sections under low power magnification these vessel walls appeared almost solidly black. As a result, even in those arteries showing no intimal thickening, the whole wall was thicker and heavier than normal (figs. 3A, B). It is difficult to state whether there was actual increase in the number of elastic fibers but undoubtedly they were hypertrophied. They showed slight fraying and beading. Collagen stains revealed no increase in fibrous tissue in the walls of these elastic vessels.
FIG. 3A. Pulmonary artery (about 900 μ) from a normal 11 year old child. For comparison with 3B. Weigert’s elastic tissue stain. × 380.

Fig. 3B. Pulmonary artery (about 900 μ) showing wide and heavily elastified wall. There is no fibrosis. Weigert’s elastic tissue stain. × 380.

There was no definite muscular hypertrophy in the media. A few vessels showed focal swelling and disappearance of the internal elastic lamina and occasionally there was focal fibrosis of the media with complete disappearance of the muscle. The arterioles, except for an occasional one, showed no lesion. The intimal changes of the small and medium sized arteries and the definite elastosis of all arteries are changes associated with stress, presumably hypertension. The absence of arteriolar lesions, and the fact that the intimal hyperplasia was not generalized, in the presence of generalized vascular elastosis, suggests that these lesions were the result, not the cause of the pulmonary hypertension.

Discussion

The interest of this case centers about the anatomic and hemodynamic relations between the patent ductus and the coarctation of the aorta. Here we have virtual separation of the upper and lower aortic segments at the isthmus with marked development of a collateral circulation, features of the “adult” type; and a patent ductus which is continuous with the descending aorta, a characteristic of the “infantile” type.

Taussig has made the important observation that where the descending aorta is fed mainly from the pulmonary artery, there may be cyanosis of the lower part of the body. From the evidence of our case it is apparent that it is also possible to have this free anatomic communication without cyanosis. The explanation for this lies in the pressure relationship which exists between the pulmonary artery and the descending aorta. In our case the pressure in the pulmonary artery was 92/74, and the intra-arterial pressure in the femoral artery (on the following day) was 102/80. It is evident that an efficient collateral circulation can maintain a considerable pressure in the aorta distal to the coarctation, and this pressure may be equal to or greater than that in the pulmonary artery.

The presence of a jet lesion in the ductus opposite the stenotic opening suggests that blood entered the ductus from the upper aortic segment. The finding of another jet lesion on the left side of the bifurcation of the main pulmonary artery may indicate that there was also a flow of blood from the descending aorta.
into the pulmonary artery. This explanation of the manner in which the blood flows through the ductus gains support by the demonstration of blood in the pulmonary artery with an oxygen content higher than that noted in the blood from the right ventricle. Furthermore, the slight but definite unsaturation of the femoral artery blood indicates the entry of venous blood into the descending aorta. This may occur due to the formation of eddy currents or by a change in pressure relationships. It seems probable that the main flow through the ductus was from aorta to pulmonary artery.

The high pulmonary artery pressure, the dilatation of this vessel and its main branches, and the marked right ventricular hypertrophy, have added to the difficulty of explaining the hemodynamic relationships in this case. Edwards and co-workers, in discussing the question of a systemic right ventricle supplying both the lungs and descending aorta, raise the point that one would expect the pulmonary vessels to exert a resistance similar to or greater than that in the systemic vessels. In this connection they have described changes in the small pulmonary arteries similar, in part, to those noted in our case. In addition, however, they observed arteriolar changes, which are not present in the case described. An adequate explanation for the pulmonary hypertension is not apparent.

The complicated features in the case presented led to a search for cases of coarctation in which the ductus was patent. In table 2 are listed 28 autopsied cases, over 1 year of age, which are arranged with regard to the site of the ductus and the state of the collateral circulation. The original references were not readily available for some of the earlier cases, and the data were obtained from manuscript notes in the Maude Abbott collection and from Barie's article.

The ductus was distal to the site of coarctation in 16 instances, and proximal in 5. In 4 cases the coarctation was described as occurring "at" the level of the patent ductus and in one "to the right" of it. In 2 cases the relationship was not described or was not apparent from the record. Among the 16 cases in which the ductus entered the aorta distal to the site of stenosis and communicated with the descending aorta, as in our case, there were 5 instances (table 2, cases 4, 5, 8, 16, 23), in which a collateral circulation was described. In none of these was there any mention of cyanosis in the lower extremities, and this suggests that the circulatory arrangement in these cases may have been similar to that in our case. The collateral circulation was not developed in the 2 cases (cases 27, 28, table 2) studied by Edwards. Bramwell mentions the absence of clinical evidence of a collateral circulation in his patient, but does not record the autopsy findings regarding this point. In 7 instances the collateral circulation is not mentioned and in one the ductus arteriosus was small, being almost obliterated by vegetations, and considered to be nonfunctional. The collateral circulation was well developed in all 5 cases in which the patent ductus lay proximal to the site of coarctation. Edwards points out that, if the stenosis is marked, this arrangement would be incompatible with fetal life unless collateral pathways developed early to ensure the return flow of blood to the placenta. However, this is not the sole mechanism by which collateral circulation develops because collateral circulation also occurs in cases where the ductus lies distal to the site of stenosis. In this latter group the fetal circulation is essentially undisturbed.

In the 16 cases (table 2) in which the pulmonary artery is continuous with the descending aorta through a ductus distal to the coarcted segment, there is no mention of cyanosis of the feet, and in 11 of these there is no mention of collateral circulation. If there is inadequate collateral circulation, the supply to the lower extremities would be mainly from the right ventricle and one would expect cyanosis in them. If collateral circulation is present, it does not rule out the possibility of cyanotic lower extremities, but renders it less likely. It appears quite possible that such a circulation may have been present in the above cases and yet not recognizable on physical examination or postmortem study.

Considerations arising out of the study of our present case and those cases noted above, em-
COARCTATION OF THE AORTA

Table 2.—Cases of Coarctation of the Aorta Associated with Patent Ductus Arteriosus

<table>
<thead>
<tr>
<th>No.</th>
<th>Reference</th>
<th>Date</th>
<th>Sex</th>
<th>Age</th>
<th>Rel. of C. to D.A.</th>
<th>Collaterals</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Nixon11, 12</td>
<td>1834</td>
<td>M</td>
<td>27</td>
<td>“at”</td>
<td>Not mentioned</td>
<td></td>
</tr>
<tr>
<td>2.</td>
<td>Chevers11</td>
<td>1845</td>
<td>M</td>
<td>“young”</td>
<td>above</td>
<td>Not mentioned</td>
<td></td>
</tr>
<tr>
<td>3.</td>
<td>Babington14</td>
<td>1847</td>
<td>M</td>
<td>24</td>
<td>above</td>
<td>Not mentioned</td>
<td></td>
</tr>
<tr>
<td>4.</td>
<td>Hamernik11, 12</td>
<td>1848</td>
<td>F</td>
<td>17</td>
<td>above</td>
<td>Diagnosed clinically</td>
<td></td>
</tr>
<tr>
<td>5.</td>
<td>Viaud-Grand-marais11, 12</td>
<td>1857</td>
<td>M</td>
<td>62</td>
<td>above</td>
<td>Dilated (int. mammary)</td>
<td>Two areas of stenosis</td>
</tr>
<tr>
<td>6.</td>
<td>Almagro11</td>
<td>1862</td>
<td>F</td>
<td>19</td>
<td>“at”</td>
<td>(prob. above)</td>
<td>Aorta dilated below ductus</td>
</tr>
<tr>
<td>7.</td>
<td>Peacock14</td>
<td>1862</td>
<td>M</td>
<td>30</td>
<td>above</td>
<td>Not mentioned</td>
<td>Isthmus stenosis</td>
</tr>
<tr>
<td>8.</td>
<td>Redenbacher11, 12</td>
<td>1873</td>
<td>M</td>
<td>7</td>
<td>above</td>
<td>Extensive</td>
<td>Isthmus stenosis</td>
</tr>
<tr>
<td>11.</td>
<td>Case 15</td>
<td>1905</td>
<td>M</td>
<td>21</td>
<td>“to right”</td>
<td>Not mentioned</td>
<td>D.A. wider than a goose quill</td>
</tr>
<tr>
<td>12.</td>
<td>Horder11</td>
<td>1907</td>
<td>M</td>
<td>12</td>
<td>“at”</td>
<td>Not mentioned</td>
<td></td>
</tr>
<tr>
<td>13.</td>
<td>Stauung11</td>
<td>1913</td>
<td>F</td>
<td>28</td>
<td>“at”</td>
<td>Not mentioned</td>
<td></td>
</tr>
<tr>
<td>14.</td>
<td>Meixner11</td>
<td>1922</td>
<td>M</td>
<td>20</td>
<td>below</td>
<td>Dilated</td>
<td>D.A. finely patent</td>
</tr>
</tbody>
</table>

Reifenstein, Levine and Gross' series18

<table>
<thead>
<tr>
<th>No.</th>
<th>Reference</th>
<th>Date</th>
<th>Sex</th>
<th>Age</th>
<th>Rel. of C. to D.A.</th>
<th>Collaterals</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>15.</td>
<td>devVries19</td>
<td>1918</td>
<td>M</td>
<td>3</td>
<td>above</td>
<td>Not mentioned</td>
<td>D.A. admits fine sound</td>
</tr>
<tr>
<td>16.</td>
<td>Ulrich20</td>
<td>1931</td>
<td>M</td>
<td>23</td>
<td>above</td>
<td>Dilated</td>
<td>D.A. tiny cap. lumen only</td>
</tr>
<tr>
<td>17.</td>
<td>Brochthern21</td>
<td>1939</td>
<td>M</td>
<td>33</td>
<td>below</td>
<td>Dilated</td>
<td></td>
</tr>
<tr>
<td>18.</td>
<td>Blackford1</td>
<td>Case 10</td>
<td>1928</td>
<td>F</td>
<td>23</td>
<td>above</td>
<td>Not mentioned</td>
</tr>
<tr>
<td>19.</td>
<td>Evans1</td>
<td>Case 2</td>
<td>1939</td>
<td>M</td>
<td>30</td>
<td>above</td>
<td>Not described</td>
</tr>
<tr>
<td>20.</td>
<td>Case 5</td>
<td>1939</td>
<td>M</td>
<td>8</td>
<td>above</td>
<td>Not described</td>
<td></td>
</tr>
<tr>
<td>22.</td>
<td>Tillich22</td>
<td>1941</td>
<td>M</td>
<td>17</td>
<td>?</td>
<td>Not mentioned</td>
<td></td>
</tr>
<tr>
<td>23.</td>
<td>Fagin, Shepeard &amp; Morrison19</td>
<td>1946</td>
<td>M</td>
<td>18</td>
<td>above</td>
<td>Dilated</td>
<td>I.V.S.D.</td>
</tr>
<tr>
<td>24.</td>
<td>Bramwell15</td>
<td>1947</td>
<td>F</td>
<td>23</td>
<td>above</td>
<td>No rib notching. P.M. findings not mentioned</td>
<td></td>
</tr>
<tr>
<td>25.</td>
<td>Edwards19</td>
<td>Case 1</td>
<td>1949</td>
<td>F</td>
<td>15</td>
<td>below</td>
<td>Dilated</td>
</tr>
<tr>
<td>26.</td>
<td>Case 2</td>
<td>1949</td>
<td>M</td>
<td>22</td>
<td>below</td>
<td>Dilated</td>
<td>Pulm. vascular lesions</td>
</tr>
<tr>
<td>27.</td>
<td>Case 3</td>
<td>1949</td>
<td>F</td>
<td>23 mos.</td>
<td>above</td>
<td>None</td>
<td>Pulm. vascular lesions</td>
</tr>
<tr>
<td>28.</td>
<td>Case 4</td>
<td>1949</td>
<td>F</td>
<td>7</td>
<td>above</td>
<td>None</td>
<td>Pulm. vascular lesions</td>
</tr>
</tbody>
</table>

Emphasize the desirability of a more physiologic approach to the study and classification of coarctation of the aorta. If the ductus is proximal, Edwards, as noted above, observes that collateral circulation must develop in the embryo for life to be maintained. Thus, it is not pos-
sible to have a patent ductus proximal to the stenosis in the absence of a collateral circulation. If the opening of the patent ductus lies distal to the coarcted segment two circulatory arrangements are possible. The pressure maintained in the aorta distal to the coarctation by the collateral circulation may be low and blood from the right ventricle may enter the descending aorta by way of the patent ductus. On the other hand, the pressure maintained in the descending aorta may be adequate to prevent blood entering from the ductus and may cause a flow of blood from aorta to pulmonary artery. Where the pressures in pulmonary artery and descending aorta are similar it is quite possible to visualize eddy currents which might permit a mixing of blood in the two vessels. It is evident, therefore, that when there is a patent ductus distal to the coarcted segment, the degree of saturation in the femoral arterial blood will be dependent upon these pressure relationships.

This discussion may be summarized by presenting these variations in the form of a classification of coarctation of the aorta. It is assumed that the degree of stenosis is such that there is marked interference in blood flow into the descending aorta distal to the coarcted segment. The arrangement noted below is based upon anatomic and hemodynamic considerations arising out of the state of the ductus, the relationship of the ductus to the site of coarctation, and the adequacy of the collateral circulation. It is observed that while it is possible, on clinical or pathologic evidence, to state that collateral circulation is present, the absence of a collateral circulation may not be demonstrable by clinical or pathologic means.

COARCTATION OF AORTA

I. Ductus Arteriosus Closed
   (1) With collateral circulation
   (2) Without (or with inadequate) collateral circulation* (incompatible with life)

II. Ductus Arteriosus Open
   A. Proximal to coarctation
      (1) With collateral circulation
      (2) Without (or with inadequate) collateral circulation* (incompatible with life)

B. Distal to coarctation
   (1) With collateral circulation
      (a) Pressure maintained by collateral circulation adequate to prevent flow from pulmonary artery to aorta.
      (b) Pressure maintained by collateral circulation inadequate to prevent flow from pulmonary artery to aorta.
   (2) Without collateral circulation*

   Such a scheme may be of help in assigning a case of coarctation to its proper category. If there is no suspicion of a patent ductus arteriosus, then the case falls into group I(1), which corresponds to the "adult" type. A complicated case may manifest itself by the associated signs of a ductus or by the demonstration of right ventricular hypertrophy or by cyanosis of the lower extremities. The decision as to whether the ductus lies proximal or distal to the site of stenosis may be clarified by determining the oxygen saturation of the femoral artery blood. If the ductus is proximal to the coarctation blood from the femoral artery is fully saturated. If cyanosis of the lower extremities is present the ductus is distal to the site of coarctation, and the collateral circulation is inadequate to prevent a venous-arterial shunt. If, in the femoral arterial blood, there is some unsaturation but insufficient to cause cyanosis, the ductus is also distal to the coarcted segment but the pressure, maintained by the collateral circulation in the descending aorta, approaches or may be higher than that in the pulmonary artery. It appears reasonable to suppose that when the pressure in the descending aorta is higher, femoral artery blood might be fully saturated or there might be some unsaturation due to eddy currents permitting blood from the pulmonary artery to enter the aorta.

SUMMARY

A hemodynamic and pathologic study is presented of a case of coarctation of the aorta, complicated by a patent ductus entering the aorta distal to the coarcted segment. The interest of the case centers about the pressure relations of the pulmonary artery and the aorta. Pulmonary hypertension was present and there were microscopic pulmonary vascular lesions.
which are described. The correlation of findings illustrates the importance of a more physiologic approach to the classification of coarctation of the aorta. Such a scheme is suggested, based upon the state of the ductus arteriosus, its relationship to the area of stenosis and the influence of the collateral circulation.

REFERENCES


11 Maude Abbott Memorial Collection of Congenital Cardiac Anomalies, Dept. of Pathology, McGill University, Montreal.


13 Abbott, M. E.: A statistical study and historical retrospect of 200 recorded cases, with autopsy, of stenosis, or obliteration, of the descending arch in subjects above the age of two years. Am. Heart J. 3: 574, 1928.


Coarctation of the Aorta Complicated by Patency of the Ductus Arteriosus: Physiologic Considerations in the Classification of Coarctation of the Aorta

ARNOLD L. JOHNSON, CHARLOTTE FERENCZ, F. W. WIGLESWORTH and DONALD L. MCRAE

Circulation. 1951;4:242-250
doi: 10.1161/01.CIR.4.2.242

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1951 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/4/2/242

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/