Coarctation of the Aorta Associated with Patent Ductus Arteriosus

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The combination of patent ductus arteriosus and coarctation of the aorta poses special anatomic, physiologic and surgical problems. An experience with 14 such cases is presented, which suggests that pulmonary hypertension occurs frequently in these cases. Physiologic data in cases having coarctation of the aorta, patent ductus arteriosus and pulmonary hypertension that underwent cardiac catheterization are presented. Clinical, anatomic, physiologic and surgical implications of this combination are discussed.

The combination of coarctation of the aorta and patent ductus arteriosus may present certain special problems in surgical management. In order to survey critically the challenge posed by these relatively uncommon cases, the surgical experience with them at the Mayo Clinic has been reviewed.

Historically, the combination of coarctation of the aorta and patent ductus arteriosus has been recognized for more than a century. Craigie, in 1841, was one of the first to describe a case having these 2 anomalies. In 1862 de Almagro described a similar patient who died at the age of 19 years of a cerebrovascular accident. Other patients were recorded by Legg in 1878, Horder in 1908 and Staunig in 1913. These latter 3 patients all died of rupture of the aorta. Maude Abbott, in an extensive review of the literature on coarctation of the aorta in 1928, found that 10 per cent of 200 patients having this anomaly had an associated patency of the ductus arteriosus. Of these 200, only 21 were diagnosed ante mortem as having coarctation, while seven others were considered to have "obstruction of the aorta." In no instance was the association of a patent ductus arteriosus with the coarctation diagnosed ante mortem. In the last decade, with the advent of increased clinical acumen, reconstructive curative surgery, and development of new diagnostic technics, the two associated conditions have been diagnosed preoperatively and effectively treated surgically.

The records of 193 cases in which coarctation of the aorta was treated surgically at the Mayo Clinic were reviewed. In 14 instances (7.3 per cent of cases), the ductus arteriosus was grossly patent. A similar figure was given by Gross in a review of 100 cases of coarctation of the aorta in 1950. He noted that the ductus arteriosus was patent in seven instances. Other reported instances of this combination include case reports by Swan and associates in 1949, Taylor and associates in 1950 and Leeds and associates in 1953. Edwards and associates, in several earlier communications from the Mayo Clinic, emphasized that increased pulmonary vascular resistance could occur with the combination of coarctation of the aorta and patent ductus arteriosus. They were able to correlate this occurrence of increased pulmonary vascular resistance with anatomic changes in the pulmonary vessels, demonstrated at necropsy. One of our cases was previously reviewed in detail by them.

Classification of Cases

For many years, classification of coarctation of the aorta has been based primarily on the site at which the ductus arteriosus, whether patent or ligamentous, is attached to the aorta. This idea was first suggested by Bonnet in 1903. He felt that in those cases in which the ligamentum arteriosum inserted proximal
to the coarctation, the lesion should be called "adult," while in those cases with a hypoplastic aortic arch and a ligamentum arteriosum inserting distal to the coarctated segment, the lesion should be called "infantile." Johnson and associates, in an excellent paper on this subject, enlarged this classification to include not only the location of the ductus with respect to the coarctation, but also whether or not collateral circulation was present. They felt that if the ductus was located proximal to the coarctation, collateral circulation was necessary to maintain life. If the ductus was distal to the coarctation, however, the amount of blood flowing from the pulmonary artery into the distal portion of the aorta would be determined by the comparative pressure of the collateral circulation and that of the pulmonary artery.

The experience at the Mayo Clinic with 14 cases of coarctation of the aorta associated with patent ductus arteriosus suggests that an important feature of this combination of defects is the presence or absence of pulmonary hypertension. The association of pulmonary hypertension with patent ductus arteriosus and coarctation of the aorta has been discussed previously from this institution. The 14 cases of associated coarctation of the aorta and patent ductus arteriosus in this study have been divided into two groups: coarctation associated with patent ductus arteriosus, pulmonary hypertension and increased pulmonary resistance; and coarctation associated with patent ductus arteriosus but with normal pulmonary resistance. For these purposes, pulmonary hypertension is defined as an average pressure in the pulmonary artery in excess of 40 mm. Hg systolic, and increased pulmonary resistance represents a resistance in excess of 200 dynes sec. cm.~5.

In this series, the occurrence of patency of the ductus arteriosus without pulmonary hypertension posed only slight additional problems in the treatment of coarctation of the aorta. A patent ductus arteriosus associated with pulmonary hypertension, however, added considerably to the technical problems of operation in coarctation. It seemed of secondary importance, surgically, whether the patent ductus attached to the aorta in the coarctated area, above it or below it.

**Clinical Data**

In eight patients patent ductus arteriosus was found at operation to be associated with coarctation of the aorta in the absence of pulmonary hypertension. In six of these patients the ductus entered the aorta proximal to the coarctation while in one patient the ductus entered opposite and in another distal to it. Seven of these eight patients had well-developed collateral circulation. There was one postoperative death in this group.

Six patients had patent ductus arteriosus associated with aortic coarctation and acceptable evidence of pulmonary hypertension. The ages of the patients in this group varied between 20 months and 27 years. In 1 of the 6 patients with coarctation, patent ductus arteriosus and pulmonary hypertension, the ductus was divided but the coarctation was not resected. In the five cases in which a complete repair was done, there were two postoperative deaths.

The location of the ductus with respect to the coarctation was different in the cases in which pulmonary hypertension was present from those in which the pulmonary pressure was normal. While 6 of the 8 patients in the group without pulmonary hypertension had a ductus joining the aorta proximal to the coarctation, 1 of the 6 in the pulmonary hypertension group had a ductus joining the aorta proximal to the coarctation, while in three the ductus arteriosus entered exactly at the coarctation and in two it entered the aorta distal to the coarctation. Table 1 includes a summary of the clinical data in these six cases in which pulmonary hypertension was present.

**Physiologic Data**

In only 4 of the 6 patients with pulmonary hypertension were preoperative catheterization data available (table 2). In the other 2 cases, acceptable evidence of severe pulmonary hypertension was obtained at opera-
TABLE 1.—Clinical Data on 6 Patients with Coarctation of the Aorta, Patent Ductus Arteriosus, and Pulmonary Hypertension

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Anatomy</th>
<th>Shunts (by catheterization)</th>
<th>Operative procedure</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>4 yr.</td>
<td>M</td>
<td>Ductus at coarctation, poor collateral circulation</td>
<td>On air: R → L and L → R</td>
<td>Ductus divided, coarctation resected</td>
<td>Died several hours postoperatively</td>
</tr>
<tr>
<td>2</td>
<td>27 yr.</td>
<td>M</td>
<td>Ductus proximal to coarctation, good collateral circulation</td>
<td>On O₂: L → R only</td>
<td>Ductus divided, coarctation resected</td>
<td>Good</td>
</tr>
<tr>
<td>3</td>
<td>12 yr.</td>
<td>M</td>
<td>Ductus at coarctation, poor collateral circulation</td>
<td>On air: L → R only</td>
<td>Ductus divided, coarctation resected</td>
<td>Good, regression, pulmonary hypertension</td>
</tr>
<tr>
<td>4</td>
<td>20 mo.</td>
<td>F</td>
<td>Ductus at coarctation, poor collateral circulation</td>
<td>No oxygen studies</td>
<td>Ductus divided, coarctation resected</td>
<td>Good</td>
</tr>
<tr>
<td>5</td>
<td>16 yr.</td>
<td>M</td>
<td>Ductus distal to coarctation</td>
<td>No catheterization data</td>
<td>Ductus divided, coarctation resected</td>
<td>Fair</td>
</tr>
<tr>
<td>6</td>
<td>7 yr.</td>
<td>F</td>
<td>Ductus distal to coarctation, poor collateral circulation</td>
<td>No catheterization data</td>
<td>Ductus divided, coarctation resected</td>
<td>Died suddenly on thirteenth postoperative day</td>
</tr>
</tbody>
</table>

TABLE 2.—Physiologic Data from Cardiac Catheterization on 4 Patients with Coarctation of the Aorta, Patent Ductus Arteriosus, and Pulmonary Hypertension

<table>
<thead>
<tr>
<th>Case</th>
<th>Pressure, mm. Hg</th>
<th>Flow, L/min.</th>
<th>Shunts, %</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Femoral artery</td>
<td>Radial artery</td>
<td>Right ventricle</td>
</tr>
<tr>
<td>1*</td>
<td>67/53</td>
<td>L.: 65/53</td>
<td>98/8</td>
</tr>
<tr>
<td>2</td>
<td>109/77</td>
<td>182/70</td>
<td>61/13</td>
</tr>
<tr>
<td>3*</td>
<td>115/63</td>
<td>140/50</td>
<td>109/7</td>
</tr>
<tr>
<td>4*</td>
<td>59/46</td>
<td>110/68</td>
<td>124/8</td>
</tr>
</tbody>
</table>

* Catheterized under general anesthesia.
† Descending aorta.
‡ These values for pulmonary blood flow and left → right shunt are subject to large error, since they are based on an arteriovenous difference of only 0.5 volume per cent.

Amputation by the surgeon’s noting a very tense pulmonary artery in which the pressure seemed equal to that in the aorta. In 2 of the 4 patients who underwent cardiac catheterization, the pulmonary artery pressure was as high as or higher than the aortic pressure above the coarctation. The ages of these patients were 4 years and 20 months, respectively. In both cases, there was right-to-left shunting of blood across the ductus arteriosus when the patients were breathing room air, and associated desaturation of blood in the descending aorta. No such right-to-left shunting was present when the patients were breathing 100 per cent oxygen, however. Left-to-right shunting of blood was present in only one of these cases on room air and in both cases on oxygen.

Two of the four patients, ages 12 and 27 years, respectively, had pulmonary artery pressures lower than those of the aorta above the coarctation, one being 61/41 mm. Hg and the other 106/68 mm. Hg. This latter patient had right-to-left shunting of blood across the patent ductus when breathing air but no right-to-left shunt when breathing oxygen, while the former patient had no right-
to-left shunt or aortic desaturation. Both patients had a left-to-right shunt, the latter one having a 92 per cent* shunt in this direction when breathing oxygen.

Direct femoral artery pressures were measured preoperatively in all four patients who were catheterized. In each instance these pressures were considerably lower than simultaneous radial pressures (table 2).

Pulmonary and systemic flow calculations employing the Fick principle were carried out with the patient breathing in sequence air and oxygen in 2 of these 4 catheterized patients, and in a third patient with the patient breathing air only. Table 2 includes the amount of flow calculated. These flows reflect the decrease in pulmonary resistance and increase in left-to-right shunting of blood across the ductus and into the pulmonary circulation that occurs when the patient breathes oxygen. The calculation of the total pulmonary resistance and total systemic resistance was carried out on the four patients who were catheterized preoperatively. These data are included in table 3.

One of the four cases has been reviewed in detail recently by Shepherd and associates. They found the total pulmonary resistance to be 860 dynes sec. cm.−5 preoperatively. It could be reduced to 150 dynes sec. cm.−5 with the patient breathing 99.6 per cent oxygen. The pulmonary arteriolar resistance on air was also markedly elevated, being 620 dynes sec. cm.−5 Four months postoperatively, this patient was again catheterized. His excellent clinical result was reflected in the postoperative catheterization findings. Table 4 gives a brief résumé of these findings. It is to be noted that right ventricular pressure fell from a preoperative level of 109/7 to 59/3 mm. Hg postoperatively, while the pulmonary arterial "wedge" pressure fell from 30/20 to 17/11 mm. Hg. Similarly, the total pulmonary resistance fell in 4 months after operation to 596 dynes sec. cm.−5 Pulmonary arteriolar resistance fell postoperatively, being 372 dynes sec. cm.−6 This patient was clinically well and had completely recovered from the effects of operation.

**Results of Operation**

The results of operation in the group of eight patients having a patent ductus arteriosus associated with coarctation of the aorta and no pulmonary hypertension were good in every case except for the one postoperative death. Of the six patients having pulmonary hypertension, patent ductus and coarctation of the aorta, two died in the postoperative period. Of the four patients surviving operation, three had good results with adequate lowering of the systemic blood pressure. One of these underwent cardiac catheterization preoperatively and postoperatively, as previously indicated. One patient, however, had only a fair result—the one in whom the ductus was divided without repair of the coarctation.

**Anatomic and Physiologic Features**

In this group of 14 cases of combined defects the physiologic and the anatomic patterns seemed related. An endeavor was therefore made to classify each case according to the point of attachment of the patent ductus arteriosus and the aorta with reference to the coarctation. In several cases this was difficult to determine and required microscopic study for accurate classification.* Three different insertions of the patent ductus were recognized: proximal to the coarctation, at or

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* This value is subject to large error, since it is based on an arteriovenous difference of only 0.5 volume per cent.

*We are indebted to Dr. J. E. Edwards for assistance in this regard.
opposite the coarctation, and distal to the coarctation.

When the patent ductus inserted proximal to the point of coarctation, a left-to-right shunt from the aorta to the pulmonary artery was usual. This shunt was suspected in all seven of the patients with this anatomic arrangement, although it was confirmed only in the one patient who was catheterized preoperatively. Pulmonary hypertension and increased pulmonary vascular resistance were rarely found when the ductus inserted proximal to the coarctation. Only 1 of 7 patients in this category had pulmonary hypertension and this patient had no right-to-left shunt.

When the patent ductus inserted at or opposite the coarctation, a different physiologic pattern occurred. Four of the patients had this anatomic arrangement. In three of these (cases 1, 3 and 4) pulmonary hypertension and increased pulmonary resistance were demonstrated at cardiac catheterization. The other patient was not catheterized, but the surgeon noted evidence of pulmonary hypertension. All of the patients in this group studied by cardiac catheterization had bidirectional shunts. In those cases in which the ductus inserted opposite the coarctation, the aortic obstruction seemed less marked than in the usual case of coarctation (table 2). In this connection all four patients having this anatomic arrangement had little or no collateral circulation. It is probable that the ductus acts as a detour, blood flowing from the proximal portion of the aorta into the aortic end of the ductus, around the coarctation, and thence from the ductus into the distal portion of the aorta.

When the patent ductus inserted distal to the coarctation, the physiologic situation was somewhat different. In one of the patients, the coarctation was not severe and there was considerable flow directly down the aorta with little collateral circulation. This patient also had pulmonary hypertension as judged by the surgeon and was the only patient in whom resection of the coarctation was not carried out, although the ductus was divided (case 5). In another case in which the ductus inserted below the coarctation, there were severe coarctation, minimal collateral circulation and pulmonary hypertension. Although cardiac catheterization had not been done to settle the point, it is probable that the ductus was supplying a portion of the descending aortic blood. In a third case with the ductus inserting below the coarctation, there was good collateral circulation and no pulmonary hypertension. Although cardiac catheterization was not carried out, presumably this patient had a left-to-right shunt supplied by the collateral circulation to the pulmonary artery. From these examples it is apparent that the flow of blood down the descending aorta in patients in whom the ductus inserts below the coarctation is governed by several factors: the caliber of the aortic lumen at the coarctation, the status of pulmonary resistance, and the development of the collateral circulation.

### Clinical Considerations

Significant in a discussion of the surgical aspects of patients with patent ductus arteriosus and coarctation of aorta is the fact that 6 of 14 such patients had pulmonary hypertension and that of the 5 patients with pulmonary hypertension in whom the combined defect was completely repaired two died in the postoperative period. One death occurred within a few hours of operation. In this 4 year old child the wedge pressure had been elevated at the time of cardiac catheterization. At necropsy, mitral insufficiency and endocardial sclerosis of the left ventricle were
found. All suture lines were intact. The other hospital death occurred suddenly, 13 days after operation. Necropsy did not reveal the cause of death in this case.

In instances of severe pulmonary hypertension and attachment of the ductus in the region of the coarctation or distal to it, considerable amounts of the blood going into the descending aorta may be coming from the pulmonary artery, as already mentioned. The usually rich collateral circulation is sometimes poorly developed under these circumstances. Then cross-clamping of the aorta for a period long enough to allow an aortic resection and anastomosis might theoretically cause paralysis of the lower limbs. This catastrophe has not occurred in any of the cases in this series. Because of this theoretic possibility, however, the anastomosis is made with as much dispatch as possible. In the last case in which operation was performed, moderate hypothermia was utilized for protection of the spinal cord during the operation as suggested by De Bakey and Cooley.¹⁵

Another fact concerning this group of patients may be of importance in the postoperative management, namely, that high oxygen concentrations lower the pulmonary artery pressure in most people with pulmonary hypertension associated with congenital heart disease. For this reason, the patient is kept under high oxygen concentrations for a prolonged period after operation, in some instances 7 to 10 days. Though this maneuver is on an empiric basis, it may lower the pulmonary artery pressure enough to take some of the strain off the right ventricle during this critical period.

Obviously, since the technical problems of dividing the ductus in a patient with a tense, often thin pulmonary artery are great, care must be taken during the procedure itself. It is of value, after the dissection has been entirely completed, to plan the rest of the operation carefully. It is usually best to place the occluding clamp on the ductus in such a position that there is a small but definite space between it and the pulmonary artery. The aorta can be cross-clamped in the usual fashion for resection of the coarctation and divided above and below the coarctated area. This procedure leaves the coarctated portion of the aorta attached to the patent ductus arteriosus. After completing the end-to-end aortic anastomosis in the usual fashion and opening the anastomosis, one can trim away enough of the coarctated portion of the aorta so that the patent ductus is left in a condition suitable for closure. In this way, an adequate cuff for one row of fine interrupted silk sutures is left and a satisfactory closure can be obtained.

**Summary**

The association of a patent ductus arteriosus with coarctation of the aorta has occurred in about seven per cent of cases of coarctation of the aorta in which surgical treatment was used. In many cases the patency of the ductus arteriosus is small and unimportant physiologically and is discovered at the operating table. This group presents little trouble, either technically or prognostically to the surgeon. The group in which a patent ductus arteriosus is associated with pulmonary hypertension, however, is important to recognize preoperatively, both because of the greater risk of operation and because of the guarded early and late prognosis.

The pathologic physiology, surgical technic, and postoperative care of the patient with coarctation, patent ductus, and pulmonary hypertension are discussed.

**Summario in Interlingua**

Le association de patente ducto arteriose con coarctation del aorta ocurredva in circa 7 pro cento del casos de coarctation del aorta in que un operation chirurgic eseva effectuate. In multe casos le patentia del ducto arteriose es pauso extense e sin importantia physiologic de manera que illo es discoperite solmente durante le operation. Iste grupo de casos presenta al chirurgo nulle extraordinari problemas technic o prognostic. Tamen, le grupo de casos in que un patente ducto arteriose es associate con hypertension pulmonar implica plus grande riscos operatori e require un plus alte grado de circumspection in le prognose tanto immediate
como etiam a longe durantia de manera que lor recognition ante le operation deveni un desiderato molto importante.

Es discutite le physiologia pathologic, le technica chirurgie, e le maneamento post-operatori de patientes con coarctation, patente ducto arteriose, e hypertension pulmonar.

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