OCCULT PRE-DUCTAL COARCTATION ASSOCIATED WITH A PERSISTENT DUCTUS

BY

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The simultaneous occurrence of clinical coarctation of the aorta and patent ductus arteriosus (PDA) has been reported frequently (Johnson et al., 1951; Cooley et al., 1956; Goldring et al., 1957; Engberg et al., 1959). It has been estimated that 6 per cent of clinically diagnosed patent ductus will have associated aortic coarctation. In most of the cases that have been described the presence of the coarctation was evident from simple physical examination. The remainder were diagnosed by means of cardiac catheterization and dye contrast studies. The pre-operative demonstration and localization of an aortic coarctation associated with PDA is necessary for the planning of an intelligent surgical approach.

We have recently studied two cases of PDA in which an associated coarctation could not be diagnosed either clinically, by cardiac catheterization, selective cineangiography, or indeed even by direct examination of the aorta at the time of ductal division. Since the hemodynamic relationships may be adversely altered by obliteration of the ductus, the lessons learned from these cases should be generally appreciated.

CASE REPORTS

Case 1. A 24-year-old girl was admitted to hospital for cardiac catheterization. A harsh, systolic murmur, initially thought to be due to a ventricular septal defect, had been first heard at four months of age. The child was undersized and was below the third percentile for weight and height in her age group. She suffered from frequent upper respiratory infections, bouts of pneumonia, and otitis. In addition, several episodes of cyanosis had been noted by the mother. She had been treated with digitalis from the age of nine months.

Physical examination showed a small, questionably cyanotic child, with a pulse rate of 140 a minute: the blood pressure in the left arm was 170/110, right arm 175/110, and the legs 185/130. A marked left praecordial heave was present. The point of maximal cardiac impulse was palpated in the fifth intercostal space at the left anterior axillary line. A systolic thrill was easily felt along the left  sternal border. Over the entire praecordium, as well as the left back and axilla, a harsh pansystolic murmur was heard. No diastolic murmur could be detected consistently. Femoral pulses were palpable bilaterally.

Routine X-rays demonstrated much cardiac enlargement, predominantly of the left ventricle, with bulging of the main pulmonary artery segment. Pulmonary vascular markings were prominent throughout both lung fields. The electrocardiogram (Fig. 1) suggested right ventricular hypertrophy.

Right heart catheterization was carried out via the right saphenous vein under fluothane—100 per cent O2 mixture anesthesia. The right atrium, right ventricle, main and right pulmonary arteries were entered without difficulty. In addition, it was possible on several occasions to pass the catheter through a patent ductus arteriosus (Fig. 2). Blood with a high oxygen saturation was encountered in the right atrium and

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Fig. 1.—Electrocardiogram of Case 1.

Fig. 2.—Radiograph taken during cardiac catheterization in Case 1 showing catheter passed through patent ductus arteriosus.

Fig. 3.—The coarctation at aortotomy in Case 1. The arrow points to the internal, web-like coarctation.
right ventricle, and there was a further rise in saturation as the catheter entered the main pulmonary artery (Table I). Blood from the descending thoracic aorta was not fully saturated despite the fact that the patient was inhaling 100 per cent oxygen, suggesting a right-to-left shunt. The pressure in the right atrium was normal, but that in the right ventricle and pulmonary artery was raised to systemic level.

### Table I

**Catheterization Data in Two Cases of Pre-ductal Coarctation with Patent Ductus Arteriosus**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Catheter position</th>
<th>O₂ percentage saturation</th>
<th>Pressure mm. Hg</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>IVC</td>
<td>68.8</td>
<td>(0)</td>
</tr>
<tr>
<td></td>
<td>SVC</td>
<td>88.5</td>
<td>72/4 (24)</td>
</tr>
<tr>
<td></td>
<td>RA</td>
<td>91.5</td>
<td></td>
</tr>
<tr>
<td></td>
<td>RV</td>
<td>90.0</td>
<td>64/28 (41)</td>
</tr>
<tr>
<td></td>
<td>MPA</td>
<td>95.0</td>
<td>61/30 (45)</td>
</tr>
<tr>
<td></td>
<td>RPA</td>
<td>92.0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Descend. aorta</td>
<td>95.0</td>
<td></td>
</tr>
<tr>
<td>2.</td>
<td>IVC</td>
<td>57.5</td>
<td>(5)</td>
</tr>
<tr>
<td></td>
<td>SVC</td>
<td>64.5</td>
<td>75/5</td>
</tr>
<tr>
<td></td>
<td>RA</td>
<td>60.0</td>
<td>95/19 (40)</td>
</tr>
<tr>
<td></td>
<td>RV</td>
<td>61.5</td>
<td>75/48</td>
</tr>
<tr>
<td></td>
<td>MPA</td>
<td>71.0</td>
<td>95/71</td>
</tr>
<tr>
<td></td>
<td>Descend. aorta</td>
<td>93.0</td>
<td>75/48</td>
</tr>
</tbody>
</table>

Figures in parentheses denote mean pressures.

Cine-angiocardiography was carried out using iohipaque, 90 per cent contrast medium. With the catheter tip in the aorta, filling of the descending aorta, patent ductus arteriosus, and pulmonary arteries was seen. A second injection into the right ventricle disclosed an intact ventricular septum, but on re-circulation of dye to the left heart, there was almost simultaneous filling of both the right and left atria. The catheterization diagnosis was patent ductus arteriosus and atrial septal defect with pulmonary hypertension and intermittent right-to-left shunt.

After surgical consultation it was decided that an attempt should be made to divide the ductus. At operation the ductus was seen to be 1 cm in length and larger in diameter than the aorta between it and the left subclavian artery. This portion of the aorta was of uniform diameter on the outside. The ductus was clamped and divided in the usual manner.

After returning the patient to her room, it was soon found that the blood pressure in the upper extremities was 220/110, but that no pressure was obtainable in either leg. A loud, harsh systolic murmur was now heard over the lower sternum. It was felt that some factor in the operative procedure had resulted in the altered systemic pressures, and a retrograde aortogram was carried out to see where the obstruction lay. This revealed an aortic coarctation at the approximate site of the aortic ductal stump.

Re-operation was performed the same day. The ductal stump repair was taken down and re-sutured longitudinally in an attempt to restore normal flow. Since this was unsuccessful, the aorta was cross-clamped and the aorta proximal to the ductus opened. A thin, pliable "web" attached to an obvious "medial coarctation" was seen just above the ductus (Fig. 3). The aortic lumen at this point was estimated to be 0.2 cm. As much as possible of this internal coarctation was excised and a Teflon patch inserted in the aortotomy to enlarge the lumen. Following this a strong, visible pulse was noted in the descending aorta.

After the second operation good femoral pulses were palpable bilaterally and the blood pressure in the legs was between 90–120 mm. Hg systolic. Two days post-operatively, after an unremarkable course, the baby vomited and aspirated gastric contents. Despite insertion of an endotracheal tube and cardiac massage she could not be revived.
Autopsy Findings. The heart was enlarged, mainly due to myocardial hypertrophy which doubled the normal thickness of both ventricles. Both the left atrium and left ventricle were dilated. A small defect was found in the medial cusp of the tricuspid valve which communicated with the left ventricle through a tract that passed through the membranous portion of the interventricular septum (Fig. 4): at its narrowest point, the tract measured about 4 mm. in diameter. This was considered to be a partial form of persistent common atrioventricular canal. It was no doubt responsible for a small left-to-right shunt detected at the atrial level during life.

The left atrial wall was thickened by much endocardial fibro-elastosis. There were no other cardiac abnormalities. The pulmonary artery was dilated and its wall thickened. The ligated pulmonary end of the ductus arteriosus was tightly sutured. A small, circular Teflon graft was found tightly sutured into the aorta at the site where the ductus had been ligated. It widened the area where the coarctation had been excised so that the aorta everywhere had a normal calibre. No aberrant vessels were noted.

Both lungs showed large areas of atelectasis. The tracheo-bronchial tree contained a large amount of aspirated material that resembled the gastric contents. The alveoli were filled with erythrocytes and epithelial debris. The small pulmonary arteries showed medial, muscular hypertrophy. When the pulmonary arterial muscle mass was measured by the method described by Naeye (1961a) it was found to be 2.3 times
greater than that of normal pulmonary arteries, and 15 per cent greater than the muscle mass of small systemic arteries. The remainder of the post-mortem findings were unremarkable except for vascular congestion of the liver and spleen.

Case 2. A 10-month-old girl was admitted to hospital for investigation of a suspected congenital cardiac anomaly. She had been in hospital three times previously because of intermittent cyanosis, congestive cardiac failure, recurrent respiratory infections, and pneumonia.

Physical examination showed a poorly-developed infant without evidence of cyanosis. The blood pressure in the arms was 80–90/60–70, and in the legs 90/70 mm. Hg. A Grade I systolic murmur was heard in the 3rd and 4th left intercostal spaces. No thrill was palpable. The liver edge was palpable 3 cm. below the right costal margin. The femoral pulsations were felt by many examiners.

The chest X-ray (Fig. 5) showed cardiac enlargement and an increase in the pulmonary vascular pattern. The cardiogram was interpreted as showing right ventricular hypertrophy (Fig. 6).

Digitalization was carried out and cardiac catheterization was undertaken. The catheter was advanced via the right saphenous vein into the right heart chambers and pulmonary arteries. On numerous occasions it entered the descending thoracic aorta via a patent ductus arteriosus. Intracardiac pressures and oxyhaemoglobin saturations are summarized in Table I. Pulmonary hypertension of systemic level was encountered, and a clear-cut left-to-right shunt at the level of the ductus was demonstrated.

Fig. 5.—PA chest radiograph of Case 2, showing cardiac enlargement and increased pulmonary vascularity.

Fig. 6.—Electrocardiogram of Case 2.
Cine-angiography was carried out with three injections of hypaque, 90 per cent, contrast material. The first films were made with the catheter tip in the aorta and showed filling of the ductus and pulmonary arteries. A second injection into the pulmonary artery allowed good visualization of re-circulation, during which no evidence of a shunt other than that through the ductus was detected. A third injection into the right ventricle failed to demonstrate any defect in the interventricular septum. The catheterization diagnosis was patent ductus arteriosus with associated pulmonary hypertension.

Since the child had progressed poorly and was being maintained only with difficulty, it was decided that closure of the ductus should be undertaken. At operation the surgeon noted that the portion of the aorta between the left subclavian artery and the ductus was smaller than normal, but there was no deformity to suggest aortic coarctation.

Post-operatively the patient did very well until the fifth day when signs of increasing cardiac failure became obvious. Although she had previously exhibited varying degrees of atrio-ventricular block with digitalis administration, it was decided that digitalis should be given in as large doses as possible short of inducing arrhythmia. Forty minutes after the administration of a very small dose of digitalis, the child was found dead in bed.

**Autopsy findings.** The infant was poorly nourished and poorly developed, appearing younger than her stated age. The heart was enlarged due to dilatation of all chambers and much hypertrophy of the walls of both ventricles. A tiny defect in the membranous portion of the interventricular septum was found. A cystic polyp, 0.8 cm. long, extended from its margin into the right ventricular cavity. Microscopically, this was composed of fibro-elastic tissue. The pulmonary artery was dilated and its wall hypertrophied. Both ligated ends of the ductus arteriosus were tightly sutured. A segment of the aorta between the left subclavian artery and the ductus was somewhat reduced in its external calibre. When the aorta was opened, a diaphragmatic structure completely occluded the lumen just proximal to the insertion of the ductus arteriosus (Fig. 7). This diaphragmatic structure consisted of smooth muscle and elastic tissue, which was continuous with the media of the adjacent aorta. A tiny branch of the aorta just proximal to the coarctation bypassed the constriction by connecting with the ductus arteriosus near its orifice (Fig. 7). This vessel was occluded at the ductal end by the sutures that closed the ductus. Embryologically, it may have been part of the ductus. There were no other abnormalities of the heart or great vessels except for a bicuspid aortic valve.

The lungs were congested. Their most striking abnormality was medial hypertrophy of smooth muscle in the muscular arteries, which measured approximately three times the normal pulmonary arterial muscle mass and 20 per cent more than the muscle mass of the small systemic arteries. The remainder of the post-mortem findings were not remarkable apart from passive congestion of the liver and spleen.

**Discussion**

These cases are remarkable by virtue of the fact that the hemodynamic relationships prior to division of the ductus arteriosus masked the presence of the associated aortic coarctation. It is to be noted that in both cases repeated blood pressure determinations in the arms and legs failed to reveal a pressure gradient. Likewise, there was no suggestion of differential cyanosis to alert examiners to the true diagnosis. Although both cases presented evidence at cardiac catheterization of equalized pressures in the pulmonary and systemic circulatory systems, the right-to-left shunt in Case 1 was quite small, and in Case 2 no shunt could be detected. In both cases selective angiocardiography with injection into the descending aorta, the ductus itself, and into the pulmonary artery failed to show the coarctation despite good dye concentration. This is not surprising however, since one would not expect reflux of contrast material into the aortic arch after it had entered the aortic
PRE-DUCTAL COARCTATION OF THE AORTA

stream from the ductus, but delineation of the coarctation also failed after recirculation of the dye into the proximal aorta. Finally, direct inspection of the aorta at the time of operation gave no indication of the coarctation because of its internal, “web-like” nature.

In Case 1 the coarctation came to light as a direct result of surgical closure of the PDA, for the pressure in the descending aorta was then no longer maintained by the hypertensive pulmonary circulation, and suture of the aortic stump of the ductus had increased the narrowing at the site of the coarctation. Careful re-suturing of the ductal stump however failed to re-establish pre-operative pressure levels.

Case 2 was unique in that the circulation round the coarctation was maintained by an anomalous channel intimately associated with the ductus which operation abolished. As in the previous instance, it also diverted the pulmonary pressure away from the lower aorta. Thus, in both cases failure to appreciate the anatomical and haemodynamic relationships led to grave consequences.

Approximately 100 case reports of coarctation of the aorta associated with PDA have appeared since Craigie’s original communication in 1841. In about half the cases the site of the aortic constriction was proximal to the ductus, and in the other half distal to it. We are however aware of no instance in which the malformation was entirely inside the aorta and completely invisible from the outside even at operation. On the other hand the findings of Claggett et al. (1954) suggest that a coarctation may not be visible on simple inspection of the aorta, for in their experience of 124 cases the aortic narrowing is due basically to infolding of the media so as to produce a diaphragm-like membrane across most of the aortic lumen. They consider the usual diagrammatic representation of the lesion as an hourglass deformity with a central perforation to be inaccurate. If this be true, it is not unreasonable to predict that the presence of an occult coarctation in association with PDA will not prove to be very rare.

From the haemodynamic standpoint the absence of hypotension in the lower extremities does not rule out the existence of a pre-ductal coarctation with PDA, for pulmonary hypertension, which is always present in these cases, can maintain the aortic pressure distal to the coarctation at normal or near-normal levels. The reason for the pulmonary hypertension is not completely understood, and it has been postulated that its origin is to be found before birth (Naeye, 1961b). It has been suggested that an increased left ventricular load during fetal life raises the end diastolic pressure in that chamber. Presumably this reduces the flow through the foramen ovale and so increases right ventricular output and pressure. Such infants have right ventricular hypertrophy at birth. This right ventricular hypertension is also presumably responsible for a reported increase in the pulmonary arterial muscle mass and a high pulmonary vascular resistance. After birth this pulmonary arterial muscle does not regress as in normal infants, probably because the pulmonary and systemic circulations remain in direct communication through the ductus arteriosus. In both of the current cases, the pulmonary arterial muscle mass was greater than that in the systemic bed, a gross reversal of the normal pattern.

From the consideration it appears that the finding of pulmonary hypertension with PDA must alert one to the possibility of a coexistent pre-ductal coarctation. Absence of differential cyanosis is an unreliable sign in this situation. Its lack has been reported previously (Johnson et al., 1951; Sullivan and Mangiardi, 1959) and has even been noted in complete absence of the aortic arch (Tabakin and Hanson, 1960).

**SUMMARY**

The clinical and haemodynamic features of two cases demonstrating patent ductus arteriosus with occult pre-ductal coarctation of the aorta are reviewed. Because of its endo-aortic nature the unsuspected coarctation may remain undetected despite direct aortic examination at the time of operation. In this situation division of the patent ductus may prove fatal. It is suggested that this combination of lesions may not be very rare and that the finding of pulmonary hypertension with PDA should initiate further study to rule out an associated coarctation.

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