CLINICOPATHOLOGIC CORRELATIONS

Bilateral Ductal Origin of the Pulmonary Arteries

Systemic-Pulmonary Arterial Anastomosis as First Stage in Planned Total Correction

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SUMMARY Absence of continuity between the heart and the pulmonary arteries has the potential for total correction. When the pulmonary arteries are narrow, a palliative first stage systemic-pulmonary arterial anastomosis serves to enlarge the pulmonary arteries so that, with time, they become of adequate caliber for total correction.

In instances of absence of continuity between the heart and the pulmonary arterial system, the possibility of ultimate complete repair is present. However, the natural state of some cases is such that the caliber of the pulmonary arteries is inadequate to carry a normal volume of blood. Creation of a palliative systemic-pulmonary arterial shunt results in enlargement of the treated pulmonary artery or arteries so that at a later time total correction may be attempted. A patient we observed in whom each pulmonary artery exhibited a ductal origin fit this approach. Palliative anastomosis was done between the aorta and right pulmonary artery.

The patient was a two-week-old, American Indian boy, the product of a normal pregnancy and full term delivery. At birth the Apgar scores had been 8 and 8, at 1 and 5 min, respectively. Five days after birth the patient was discharged to the care of his mother, apparently in good health. Two weeks after birth the patient became cyanotic, hypoxic, and acidotic. He was treated with 28 mEq of sodium bicarbonate at his local hospital and then transferred to the University of Minnesota Hospitals. When he arrived, the patient was intensely centrally cyanosed and in a state of moderate respiratory distress.

Initial examination revealed a blood pressure of 44 mm Hg by the flush method, a temperature of 99°F, respiratory rate of 58, and pulse rate of 156. The pulses of the extremities were all palpable and equal and the thorax was clear to auscultation. A normal first cardiac sound and a single second sound were present. No murmurs were audible. The liver was palpable 1–2 cm below the right costal margin in the midclavicular line.

Laboratory studies revealed the hemoglobin concentration of the blood to be 15.8 g/100 ml and a hematocrit of 45.5%. Arterial blood gases while the patient was breathing 100% oxygen were pH, 7.41; pCO₂, 45; and pO₂, 19. Total leukocyte count, urinalysis, and concentrations of serum electrolytes were within normal limits.

Roentgenograms of the thorax revealed a minimally enlarged heart with an elevated apex and underdeveloped pulmonary arterial segment. The pulmonary vascularity was decreased. The electrocardiogram revealed an axis of +150 and an RS finding consistent with right ventricular hypertrophy, i.e., low RS ratio in lead V₆, flat T waves in lead I, and upright T waves in lead V₅ and right axis deviation. The RS pattern from leads V₁-V₆ suggested a single ventricle.

During cardiac catheterization, the catheter took an abnormal course into a persistent left superior vena cava. A right ventriculogram revealed a large, dilated, trabeculated ventricle and no definite left ventricle could be identified. A pulmonary arterial segment was not identified and the aorta, which was far anterior, filled directly from the identified ventricle. The aortic arch was left-sided. Small pulmonary arteries filled from the aorta in the following manner. The right pulmonary artery originated from an arterial structure coming off the innominate artery (fig. 1), probably a right ductus, and the left pulmonary artery came from the left ductus. There was no evidence of communication between the right and left pulmonary arteries. After passing over the left bronchus, the aorta crossed over the spine to the right to leave the thorax, probably through the same diaphragmatic opening as the inferior vena cava, strongly suggesting the asplenic syndrome.

The patient was explored through a right anterior-lateral thoracotomy. The pericardium was opened anteriorly to the superior vena cava. The aorta lay far anteriorly. The right pulmonary artery was located medial and posterior to the superior vena cava (fig. 2). This vessel originated from an arterial structure which could be traced superiorly to the base of the innominate artery. There was also continuation of the right pulmonary artery beneath the aorta as an atritic strand (1 mm diameter). This strand was traced medially beneath the aorta but its connection with other vascular structures could not be identified. A main pulmonary trunk...
The echocardiogram was interpreted as showing a mass, arising from the interventricular septum and extending into the left ventricular outflow tract, which obstructed the aortic valve during systole.

**Cardiac Catheterization and Angiography**

Cardiac catheterization was performed under sedation with phenobarbital and morphine. The pressure data are summarized in Table 1.

An 80 mm Hg systolic gradient was present between the left ventricle and aorta with elevated left ventricular end-diastolic and pulmonary wedge pressures. There was no evidence of a left-to-right shunt by oximetry and the cardiac output was within normal limits.

Aortography showed a large nonopacified rounded shadow filling the aortic valve in systole but disappearing from the aorta in diastole (fig. 5 A and B). The left ventriculogram revealed a large multilobulated filling defect extending up into the aortic valve in systole (fig. 6 A and B).

**Surgical and Postoperative Therapy**

Surgery was performed using cardiopulmonary bypass. A large mass was felt by palpation through the left atrium and aorta. This mass originated from the interventricular septum and protruded into the left ventricular cavity obstruct-
Figure 3. Diagrammatic representation of the operative steps. a) The right ductus arteriosus has been divided and the ascending aorta is being anastomosed to the right pulmonary artery. b) The completed anastomosis between the aorta and the right pulmonary artery. A = aorta; SVC = superior vena cava.

Cyanosis was a prominent clinical feature in the two previous cases and in this case. This suggests that, in each, ductus-dependent pulmonary flow was inadequate.

When there is absence of continuity between the heart and the pulmonary arteries, an important anatomico-surgical point is whether the two pulmonary arteries have or do not have confluent origin. In our case the atretic strand that was observed at operation to extend from the right pulmonary artery medially behind the aorta may be an atretic segment of that artery that joins the left pulmonary artery, as shown in figure 2, and this strand may also connect with an unidentified atretic strand-like pulmonary trunk. The most important finding from the practical standpoint of developing potential channels for blood flow is that the pulmonary arterial anomalies demonstrated here fall into the non-confluent category.

The next step in our case is to determine the state of the ventricular portion of the heart. If two ventricles are present, the patient would be a candidate for total correction of the conditions present. Further study is planned.

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