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 fits 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 concentra-
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 phenobarbitone 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-
reviewed by Bharati and associates,4 only one patient had anatomy similar to our patient.

Bilateral origin of the pulmonary arteries from the homolateral ductus may arise because of either disappearance or failure of the ventral bud of the sixth aortic arch to form on each side simultaneously, with persistence of the dorsal bud of the sixth arch on each side, and continued continuity with the postbrachial pulmonary plexus. Therefore, the source of arterial supply to each lung was that part of the sixth arch designated as the distal end of the ductus arteriosus.1

Cyanosis was a prominent clinical feature in the two previous cases2 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.5 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.

References

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