Aortic Origin of the RPA: Immediate Resolution of Severe Pulmonary Artery Hypertension by Surgical Repair

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SUMMARY. Aortic origin of the right pulmonary artery (AORPA) is a rare condition requiring a high index of suspicion for diagnosis as the usual signs of structural heart disease are often absent. Ultrasound examination can strongly suggest the condition but can easily be misinterpreted. Infants should be considered "operable" even when the resistance calculations predict otherwise as there appears to be a large component of immediately reversible pulmonary artery hypertension. With technical skills learned from arterial switch operations (for transposition of the great arteries), surgical repositioning of the RPA should be possible in nearly all cases without the use of a tube graft.

KEY WORDS: Congenital heart surgery — Pulmonary vascular obstructive disease — Absent pulmonary artery

Aortic origin of the right pulmonary artery (AORPA), first described in 1868 by Fraentzel [8], has been variably called "congenital absence of the RPA" [2, 3, 5, 6, 12, 14, 18] or even hemi-truncus. It is a condition where the intracardiac anatomy is generally normal and a well-formed RPA originates from the ascending aorta [4, 6, 15, 16, 19, 22]. Though readily reparable with modern techniques [7, 10, 11], mortality has been high because it may go undiagnosed and is usually associated with severe bilateral pulmonary artery hypertension, the pathogenesis of which is unclear. Our recent experience with this condition indicates that an extreme level of pulmonary artery hypertension may be readily reversible and despite hemodynamic calculations suggesting an inoperable status, a successful outcome can be achieved with surgical repair.

Case Report

A 3-month-old black male infant presented with breathing difficulties for several weeks. He was acyanotic (room air O₂ saturation = 95%) and in moderate respiratory distress with a respiratory rate of 48–60/min. His weight (3.85 kg) was less than the 5th percentile. The lungs were clear to auscultation. Abdomen was soft with a 3-cm firm, tender liver edge palpable at the mid-clavicular line below the right costal margin. Peripheral pulses were strong and equal. The precordial maximal impulse at the 5th left intercostal space was 1 cm outside the mid-clavicular line with a strong parasternal heave. The second heart sound was single and loud. A grade ¾ nonspecific murmur was heard along the left lower sternal border. Diastole was clear and the cardiac rhythm was normal.

Electrocardiogram showed a normal sinus rhythm with right axis deviation (95°), right atrial and right ventricular hypertrophy, and normal ST and T waves. Chest x-ray showed moderate generalized cardiomegaly; pulmonary vascularity seemed mildly increased on the right and normal on the left. Two-dimensional echo Doppler study demonstrated dilated right atrium (RA) and right ventricle (RV) with tricuspid regurgitation (5 m/s) suggesting RV hypertension. The RV contractility was markedly diminished. The great artery arising from the left ventricle appeared to bifurcate (Fig. 1A) soon after its origin, giving the impression of a main pulmonary artery (MPA) and, on para-sternal short-axis view, another great artery arose from the RV apparently parallel to the posterior great artery (Fig. 1B).

Angiocardiography revealed origination of the right pulmonary artery (RPA) from the ascending aorta (Fig. 2B); the left pulmonary artery (LPA) arose normally from the main pulmonary artery. RV trabeculae were markedly hypertrophied and RV contractility was severely reduced. Systolic pressure in the RPA was equal to aortic pressure and the LPA systolic pressure was suprasystolic (see Table 1). Administration of oxygen had...
wall; the resulting aortic defect was reconstructed with a patch of gluteraldehyde-fixed autologous pericardium. The RPA was mobilized well into the hilum and passed posterior to the ascending aorta. The MPA was opened longitudinally and enlarged anteriorly to form a flap on the anterior aspect of the MPA arteriotomy. Direct end-to-end anastomosis was performed with running 6-0 PDS suture without tension. At the end of the procedure the RPA and LPA systolic pressure was 28 mmHg, while the simultaneous systemic arterial systolic pressure was 90 mmHg. RV dimensions and contractility seemed immediately improved by visual inspection, although right atrial pressure remained -4 mmHg higher than left atrial pressure for several hours after surgery.

The postoperative course was benign. Prior to hospital discharge a follow-up cardiac catheterization was done (Fig. 2 and Table 1). Pulmonary perfusion scan performed 3 months after surgery showed 35% of RV ejection going to the left lung, compared to 25% noted 1 week after operation (Table 1). The patient continues to do well 6 months after surgery.

Discussion

Our patient had AORPA; the RPA was not “absent” and there were no abnormalities of the conotruncus to support a label of “hemi-truncus.” Based on the echocardiographic picture (Fig. 1), one might be tempted to suggest “hemi-transposition,” as one pulmonary artery is connected to the left ventricle. Clearly, the best name is AORPA which is precise, accurate, and unambiguous.

Why is this condition so hard to diagnose, especially with modern echocardiographic capabilities? The answer lies in the difficulty in recognizing the presence of significant congenital heart disease. The usual hallmarks—cyanosis, murmur, or overt congestive heart failure—generally are absent. As the pulmonary vascular resistance is usually elevated, the “shunt” flow through the right lung may not be high enough to produce volume overload of the left heart. Indeed, until there is right heart failure from pressure overload, these children may do reasonably well without signs of cardiac congestion. The clinical signs usually present are tachypnea, narrowly split-second heart sound with accentuated P2 and failure to thrive.

Current embryologic thinking indicates that the proximal portions of the pulmonary arteries derive from the sixth aortic arches while the more distal (intraparenchymal) pulmonary arteries originate from the lung buds. There are two theories extant to explain AORPA: one [20] suggests malattachment of the sixth aortic arch, connecting to the aortic side rather than the pulmonary artery side of the truncus, while the other by Wagenvoort and Wagenvoort [21] proposes atresia of the right sixth arch with the RPA derived from the right dorsal aorta and the right ductus arteriosus and lung-bud derived from the distal RPA.

The mechanism of pulmonary artery hyperten-