Pediatric Cardiac Surgery Annual

How We Manage Patients With Major Aorta Pulmonary Collaterals

William J. Brawn, Timothy Jones, Ben Davies, and David Barron

Patients with major aortopulmonary collateral arteries usually present in one of three ways: either with marked heart failure because of lung overflow, cyanotic because of reduced lung flow, or fairly well balanced with systemic oxygen saturations in the high 70s to low 80s. All patients require a planned cardiologic surgical approach, with careful investigation to delineate the collateral morphology. A carefully coordinated, combined approach between surgery and cardiology intervention is required throughout the treatment of these patients. The majority of these patients now enter a program of reconstruction of the collaterals to a valved right ventricular pulmonary artery conduit with or without ventricular septal defect closure. Further catheter intervention to stretch and enlarge the pulmonary arteries may be necessary, followed by staged ventricular septal defect closure. Other techniques to enlarge central pulmonary arteries or to recruit collaterals can be used. Outcomes over the last 20 years have been satisfactory, with survival of 80% over 10 years, which is a marked improvement on the natural survival in this group of patients.
Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 12:152-157 © 2009 Published by Elsevier Inc.

KEYWORDS Pulmonary atresia, Pulmonary collaterals, Major aorta pulmonary collaterals and MAPCAs

Patients with multifocal blood supply to the lungs, so called collateral vessels or major aorta pulmonary collaterals (MAPCAs), present a complex surgical problem in reconstruction and reorganization of the pulmonary artery supply to the lungs so that they can be reconnected in the normal fashion to the right ventricle (RV). In over 90% of cases this abnormal pulmonary blood supply occurs in patients with tetralogy with pulmonary atresia or extreme Fallots tetralogy, and rarely in other conditions such as pulmonary atresia with intact ventricular septum, congenitally corrected transposition, truncus arteriosus, and univentricular heart. There is great variability in the number and origin of the MAPCAs, but the majority arise from the mid descending thoracic aorta. There is marked hypoplasia of the central intrapericardial pulmonary arteries in the majority, and in a small percentage of cases there may be complete absence of intrapericardial pulmonary arteries. The distribution of intrapulmonary vessels is variable, there may be confluence between the upper and lower lobes or separate lobes and lung segments may be supplied by the collaterals without any interconnections.[1] The variability and complexity of these collaterals has made the surgical reconstruction of these vessels very difficult; it is really only in the last 20 years or so that a determined attempt has been made to repair these vessels to the RV so that the heart can be septated.[2-6]

In Birmingham, MAPCA surgery has evolved over the last 20 years from ligation and shunting of collaterals to staged hilar unifocalization and then central focalization to reconstructed intrapericardial pulmonary arteries connected to the RV with a valved conduit with or without ventricular septal defect (VSD) closure. For the patients, this has often meant multiple surgical and catheter interventional procedures; however, the outcome in the long run has been very satisfactory.

Clinical Presentation

Patients usually present with a variable degree of cyanosis that may be so mild as not to be clinically apparent and, thus, patients can present quite late because the cyanosis has not been detected. However, they can also present in severe heart failure because of lung overflow or severe dangerous cyanosis

Department of Paediatric Cardiac Surgery, Birmingham Children's Hospital, Birmingham, United Kingdom.
Address correspondence to William J. Brawn, MD, Diana, Princess of Wales Children's Hospital, Steelhouse Lane, Birmingham, B4 6NH, United Kingdom; E-mail: William.brawn@bch.nhs.uk

 1092-9126/09/$-see front matter © 2009 Published by Elsevier Inc.
doi:10.1053/j.pcsu.2009.01.018

because of inadequate blood flow to the lungs. When the pulmonary to systemic circulations are well balanced, usually with oxygen saturations 75% to 85%, no medical therapy may be needed. In the presence of congestive cardiac failure, diuretics and ACE inhibitors are necessary, and even ventilatory support with inotropes in severe cases. Similarly marked cyanosis may require supplemented oxygen. Elucidation of the intracardiac morphology is important but does not really determine the approach to the reconstruction of the pulmonary arteries and collateral vessels.

Investigation

In our center, all patients undergo cardiac catheterization and angiography at presentation. This may be repeated if the initial study does not clearly define the morphology of the MAPCAs. It is necessary to know the number, position, and origin of the MAPCAs, and their size and distribution within the lungs. The catheter will usually define any intrapericardial pulmonary arteries and potential connection to the heart. In situations were the collaterals are small and it is not possible to show any native vessels within the lung, a pulmonary venous wedge injection can be helpful to backfill the pulmonary arteries, even to within the pericardium.

The timing of the cardiac catheterization is usually within a few days of the initial diagnosis. The majority of patients now present within a few weeks of birth and have their initial investigation as a neonate or in early infancy. Patients that have severe cyanosis or marked cardiac failure may need immediate surgery following investigation.

Magnetic Resonance Imaging and Computerized Axial Tomography

Magnetic resonance imaging (MRI) and computerized axial tomography (CAT) scans are occasionally performed in our unit, but are not routine. Additional information from a CAT or MRI can help in delineating the position of MAPCAs within the mediastinum and their relationship to the trachea, main airways, and oesophagus.[7] It is helpful to know whether major collaterals pass in front of or behind the airways to determine how best to perform the reconstruction.

General Considerations

The aim of surgery in this group of patients is to improve the natural history described by Bull et al,[8] which illustrated the outcome in 218 patients, showing attrition early in life and infancy. The problem has always been not to harm patients that might survive for many years in a relatively stable condition with adequate collaterals. However, the disadvantage is that they may well have a restricted lifestyle with exercise intolerance, develop polycythemia, and be prone to infection and secondary hemorrhage. The question is whether a confluent low pressure pulmonary artery system can be reconstructed from the MAPCA morphology and native pulmonary arteries with a normal or near normal RV/LV pressure ratio. In addition, can the need for re-operation and re-interventions at cardiac catheterization be kept as low as possible?

Surgical Approach

Currently, we aim for a primary reconstruction of the pulmonary arteries to a valved conduit connected to the RV with VSD closure. The VSD may not be closed primarily because of inadequate run off into the pulmonary arteries. A staged approach may be necessary with reconstruction of the pulmonary arteries to a RV conduit with catheter interventions to dilate up distal pulmonary arteries or surgery to enlarge the pulmonary arteries, with VSD closure at a later time. If it is not possible to reconstruct the pulmonary arteries with adequate run off to the lungs, then a valved conduit reconstruction to the pulmonary arteries without VSD closure provides good long-term outcome without the risk of RV failure because of RV hypertension.

When the central pulmonary arteries are small with poor collateral development, such that reconstruction would be very difficult, enlargement of the central pulmonary arteries may be possible to facilitate later surgery. This can be achieved by direct aortic to main pulmonary artery anastomosis (Fig. 1),[9,10] or a RV outflow tract to atretic pulmonary artery connection with a patch reconstruction or interposition prosthetic tube. Growth or dilatation of the central pulmonary arteries can be achieved, but usually highlights stenoses at the hila regions or within the pulmonary parenchyma. Further surgical and catheter interventions to overcome the stenoses are needed.

Hila reconstruction of the collaterals with a systemic shunt placed from the aorta to the reconstructed hila region (Fig. 2) can be performed to stimulate growth and dilatation of the pulmonary arteries, then (depending on the development of these vessels) it may be possible to connect them across the midline, sometimes with prosthetic tubes, usually Gore-Tex.

Thus, one must be fairly inventive in the ways of connecting the repair or reconstructing the pulmonary artery tree to

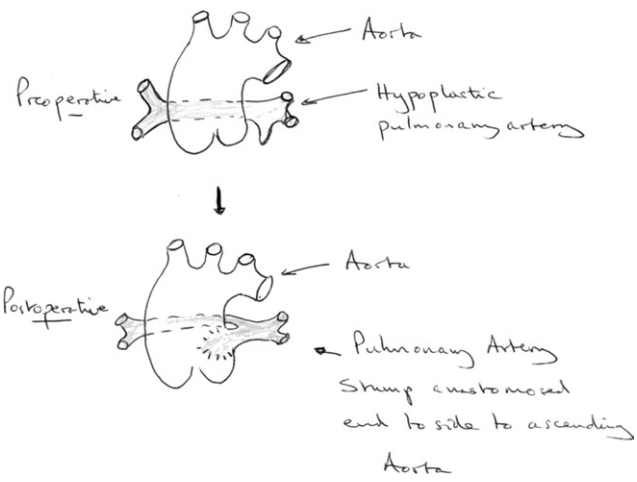

Figure 1 Main pulmonary artery stump. Aortic anastomosis to stimulate pulmonary artery growth and improve cyanosis.