Approaches to Pulmonary Atresia With Major Aortopulmonary Collateral Arteries

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Pulmonary atresia with major aortopulmonary collateral arteries (MAPCAs) is one of the most challenging surgical conditions to manage—not only because of the technical complexity of the surgery but also in terms of defining the anatomy of the pulmonary vasculature, the timing of surgery, and decision making on staged vs complete repair. The importance of early definition of pulmonary blood supply is paramount, establishing which areas of the lung are supplied by MAPCAs alone and which have dual supply with the native system (noting that 20% of patients have absent intrapericardial native vessels). Early unifocalization (3-6 months) is ideal, with closure of the ventricular septal defect (VSD) performed if 15 or more out of 20 lung segments can be recruited. Leaving the ventricular septal defect open with a limiting right ventricle-pulmonary artery conduit can be a useful interim or even definitive circulation in patients with borderline vasculature. Rehabilitation of small native vessels with central shunts can be very effective, but best outcomes are achieved by a combination of unifocalization of MAPCAs together with the native vessels (if present). A variety of reconstructive techniques are necessary to be able to effect these complex repairs with careful choice of materials. Ideally, surgery can be completed through sternotomy alone, but separate thoracotomies may be necessary to control and access some MAPCAs.

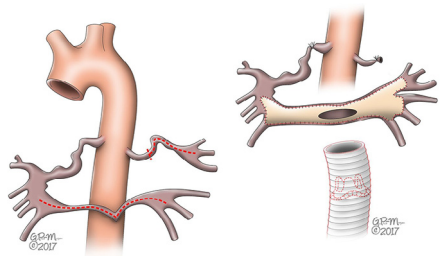
Keywords: pulmonary atresia, aortopulmonary collateral arteries, MAPCAs, unifocalization

Introduction

Within the spectrum of tetralogy of Fallot with pulmonary atresia, 20%-40% of pulmonary atresia cases will have multiple sources of pulmonary blood flow from the systemic circulation (major aortopulmonary collateral arteries [MAPCAs]). These are among the most challenging of all congenital heart lesions to manage[1] because they require detailed and exhaustive definition of the anatomy and a recognition of the great heterogeneity of MAPCAs from 1 patient to the next. Consequently, surgery needs careful preoperative planning and involves the need to operate in the posterior mediastinum among structures that are unfamiliar to most surgery in congenital heart disease. A staged approach may be necessary in more complex anatomies, and the timing and strategy for surgery is key to delivering the best outcomes for this complex and varied set of patients, with a combined approach from surgeons and interventional cardiologists.

The essential principles of management are as follows:

a. Early and thorough assessment of the anatomy of the pulmonary blood supply.
b. Focus on achieving early unifocalization (within the first 6 months of life).



Unifocalization of MAPCAs incorporating areas of dual supply and of sole MAPCA supply.

Central Message

Pulmonary atresia with MAPCA management requires early and detailed definition of pulmonary blood supply, with clear understanding of which areas are supplied by MAPCA alone and which have dual supply with the native system. Early unifocalization (3-6 months) is ideal, with closure of the VSD performed if 15 or more out of 20 lung segments can be recruited; leaving the VSD open with a limiting right ventricle-pulmonary artery conduit can be a useful interim or even definitive circulation in patients with borderline vasculature. Rehabilitation of small native vessels with central shunts can be very effective, but best outcomes are achieved by a combination of unifocalization of MAPCAs together with the native vessels (if present).

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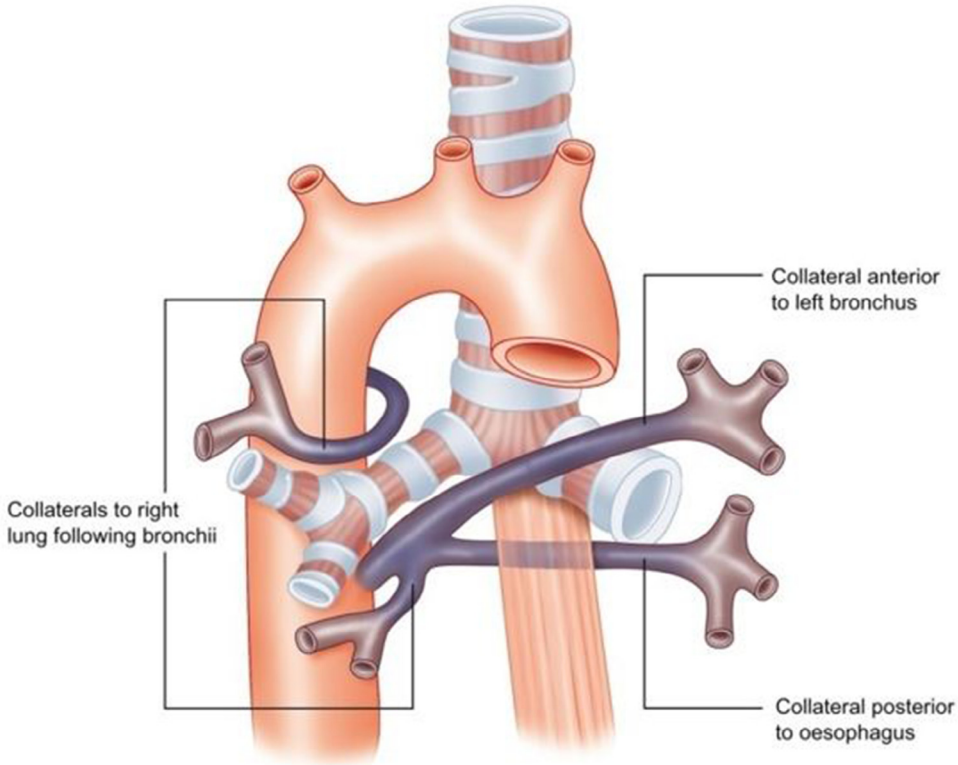

Figure 1 An example showing common relationships of MAPCAs to the trachea, main bronchi, and esophagus. (Color version of figure is available online.)

c. Understanding the relationship between native pulmonary arteries and MAPCAs to identify areas of *dual supply* and *sole supply*.
d. Unifocalization is more important than complete repair, and it can be valuable to leave the ventricular septal defect (VSD) open initially in more difficult anatomy.

One of the characteristics of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries (PA/VSD/MAPCAs) is the great heterogeneity of pulmonary blood supply, which will dictate not only the clinical condition of the patient but also the options for surgical management.[2] The MAPCAs themselves are variable in both size and number and, furthermore, are variable in their course from site of origin to their point of entry into the lung: MAPCAs most typically arise from the descending thoracic aorta at about T4-T6 level, but the next most common origin is from the underside of the aortic arch. Other common sites of origin are from the subclavian or brachiocephalic arteries and they can also arise from the distal descending aorta and even from the abdominal aorta, rising up behind the diaphragm. The extrapulmonary course of MAPCAs can be direct entry into the lung, or may be a convoluted and tortuous course, frequently with origin stenosis that can be present from birth or may develop and progress with time (reflecting the abnormal structure of the media of these vessels, which can be thickened and muscular). Because MAPCAs usually arise very posteriorly in the mediastinum, their relationship to the airways and the esophagus is also highly variable. They can pass behind, in front of, or even through the muscular wall of the esophagus and tend to run along the main bronchi—but can be above, below, anterior, or posterior to the bronchi before entering the lung (Fig. 1). A common pattern for MAPCAs arising from the mid-thoracic aorta is for them to run under the carina of the trachea and follow the course under the main bronchi to enter the lung.

The intrapulmonary course of the MAPCAs is equally variable but we have noted that MAPCAs that come to enter the lung at the hilum frequently adopt a branching pattern and appearance much like a native pulmonary artery.[3] Conversely, MAPCAs running directly unto the lung, sometimes posterior to the bronchus, can also have abnormal intrapulmonary branching patterns with stenotic areas within the lung that are not accessible to surgery.

The final and most important variation in blood supply is the relationship between native pulmonary arteries and MAPCAs. Around 80%-85% of all the patients will have confluent intrapericardial native pulmonary arteries, which are usually small and underdeveloped to a variable extent. There is normally no forward flow into these vessels (occasionally there can be a very small communication providing a wisp of forward flow), but they are filled by retrograde flow from one or more of the MAPCAs, appearing on angiography to have the appearance of a seagull in flight due to the midpoint of the vessels being connected to the heart and so moving up and down with ventricular contraction (Fig. 2). Although the central (intrapericardial) component of these native vessels is usually small, and even diminutive, the branching pattern within the lungs can be much better but is, again, variable. Regardless of the absolute size of these branches,