IMAGES **in PAEDIATRIC CARDIOLOGY**

Management of Tetralogy of Fallot with Pulmonary Atresia

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Abstract

Tetralogy of Fallot with Pulmonary Atresia is an extreme form of tetralogy characterized by absence of flow from the right ventricle to the pulmonary arteries. Pulmonary blood flow is derived from a variety of sources, including native pulmonary artery branches and aorto-pulmonary collaterals with significant variability from patient to patient. Management must therefore be individualized to each patient's anatomy and physiology. Cardiac catheterization plays a crucial diagnostic and therapeutic role in this group of patients. This article is a concise review of the spectrum of anatomic variability seen in this lesion with an emphasis on diagnostic and therapeutic catheterization . It also highlights our staged surgical approach to this lesion and provides data on long-term outcome after complete intracardiac repair.

MeSH: Tetralogy of Fallot, Pulmonary Atresia

Background

Tetralogy of Fallot with Pulmonary Atresia (TOF/PA) is a complex lesion with many different anatomic variants. The intracardiac anatomy is that of a typical Tetralogy of Fallot with a large subaortic ventricular septal defect and anterior malalignment of the conal septum resulting in right ventricular outflow tract obstruction. The pulmonary valve is often absent with a muscular pouch in its place. Occasionally, it presents as a blind membrane. The primary source of variability in this lesion is in the anatomy of the pulmonary arteries, with the spectrum ranging from well-formed, confluent pulmonary artery branches (Fig. 1A,B) to completely absent native pulmonary arteries and major aorto-

pulmonary collaterals (MAPCA's) providing all the pulmonary blood flow (Fig. 2).[1]

The majority of cases fall somewhere in between these two ends of the spectrum, with the total pulmonary blood flow provided by a combination of diminutive native pulmonary arteries and multiple MAPCA's (Fig. 3A,B). Different segments of the lungs are fed by either a native pulmonary artery or a MAPCA, and sometimes by both, referred to as "dual supply".[2] Frequently these patients have underperfused lung segments with little or no flow from the native pulmonary arteries or the collateral circulation, referred to as arborization defects (Fig. 4A,B).

Figure 1 Angiogram in the left BT shunt in anterior/posterior (A) and lateral (B) projections demonstrates good size right and left pulmonary arteries. This patient had ductal dependent pulmonary circulation and underwent placement of the BT shunt shortly after birth. BT = Blalock Taussig.

Figure 2 Descending aortogram showing two large MAPCA's, one to each lung, and absent native central pulmonary arteries. The patient has a right aortic arch.

Figure 3 A. Descending aortogram in an infant showing multiple MAPCA's to both lungs. B. Selective injection into one of the left-sided MAPCA's fills the native central pulmonary arteries. Note the very stenotic intrapulmonary connection between the MAPCA and the native pulmonary arteries.
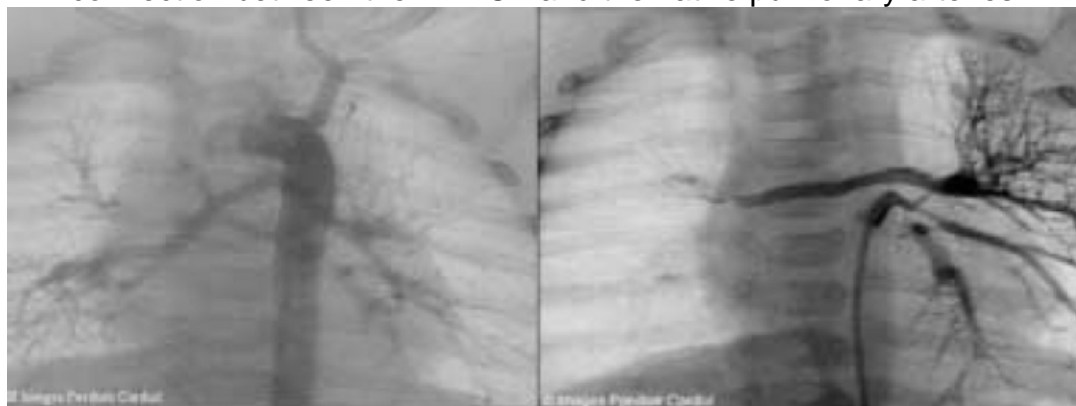

Figure 4 A. Injection into a reconstructed right pulmonary artery in a patient who has undergone a right unifocalization. The catheter is inserted via a right modified Blalock Taussig shunt. Note the lack of perfusion to segments of the right upper and right lower lobes in the anterior-posterior projection. B. In the lateral projection the reconstructed right pulmonary artery is seen to perfuse primarily right middle lobe anterior segments.

Over the last few decades, the management of TOF/PA has evolved from a conservative approach of no intervention or limited palliation when mandated by severe cyanosis or unremitting heart failure, to one of early intervention in the hope of achieving a divided circulation with acceptable right ventricular pressure in childhood. There are still multiple surgical approaches to this lesion. Long-term data are needed to determine the optimal form of management.

It should also be noted that there is a small subset of patients with true tetralogy of Fallot, i.e. with forward flow from the right ventricle to the pulmonary arteries, whose pulmonary artery anatomy resembles that of patients with TOF/PA. These patients have relatively hypoplastic central

pulmonary arteries and their pulmonary blood flow is augmented by MAPCA's. Such patients are subjected to a similar management strategy as that for patients with TOF/PA. However, if they present early on with inadequate pulmonary blood flow, the initial palliation would be a Blalock-Taussig shunt rather than a central shunt. The MAPCA's are dealt with in a similar fashion as in patients with TOF/PA.

## Management strategies – initial stage

In order to plan surgical management, the anatomy of the native pulmonary arteries and MAPCA's must be clearly defined. The first question to be answered in a newborn with TOF/PA is whether the pulmonary flow is dependent on a patent ductus. Often this question can be answered satisfactorily with echocardiography, but when in doubt, cardiac catheterization is mandatory. Patients with ductal dependent pulmonary circulation almost always have good size, confluent pulmonary arteries and no significant MAPCA's (Fig. 1).

In the majority of cases of TOF with pulmonary atresia, the native pulmonary arteries are confluent but hypoplastic, sometimes measuring no more than 1-2 mm in diameter. Flow into them is derived from MAPCA's, typically via small intrapulmonary communications (Fig. 3B). It is often possible to see these small pulmonary arteries and multiple MAPCA's arising from the descending aorta by echocardiogram. Less frequently, MAPCA's arise from the ascending aorta or head and neck vessels. MAPCA's arising from the head and neck vessels most commonly arise from the subclavian arteries and typically perfuse a single lobe (Fig. 5), however, they can arise from any vessel including the coronary arteries (Fig. 6).

Figure 5 Selective angiogram of a large MAPCA originating from the left subclavian artery and supplying the left upper lobe in a patient with a right aortic arch.

Figure 6 A. Selective injection in this patient's single coronary artery demonstrating a large MAPCA arising from the proximal left coronary artery branch and perfusing segments of the right upper and right lower lobes. B. Lateral projection of the same injection. MAPCA = major aortopulmonary collateral artery.
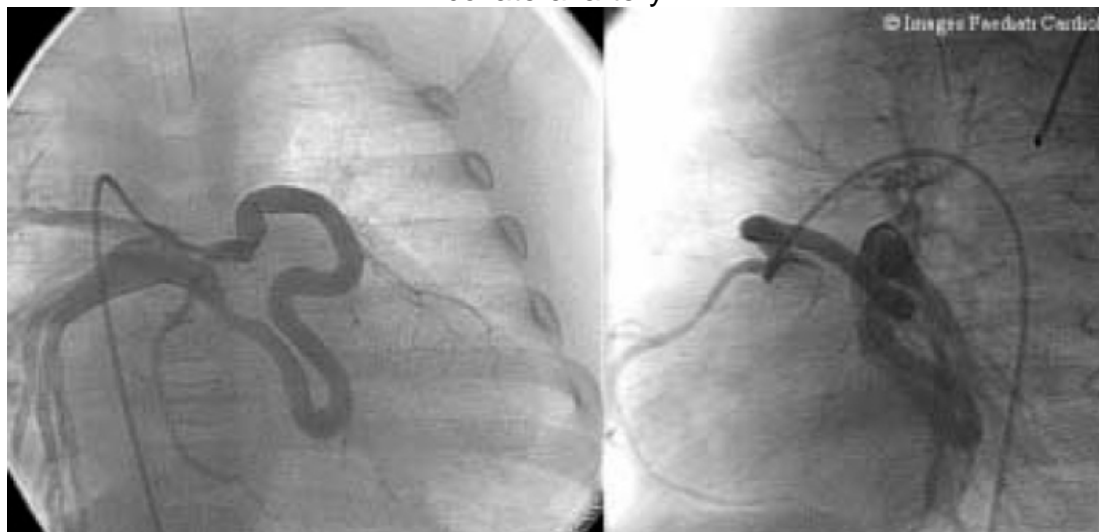
If it is clear by echocardiography that there are large collateral vessels and the pulmonary circulation is not ductal dependent, and the patient's oxygenation is adequate with saturations in the high 70's to 80's, catheterization can be deferred beyond the immediate newborn period. In the majority of these cases, the patient will have either adequate or excessive pulmonary blood flow requiring anti-congestive therapy for the first several weeks of life. If no intervention is performed, these patients can remain well oxygenated for a variable period. The natural history of this lesion, however, is that of eventual progressive cyanosis, either due to development of stenosis in the aortopulmonary collaterals (Fig. 7), or development of pulmonary vascular disease in segments where unrestricted flow from large MAPCA's has been present (Fig. 8).[3,4] In patients with diminutive native pulmonary arteries, we recommend initial intervention in the first few months of life to optimize the potential for growth of the native pulmonary arteries.

Less commonly, the native central pulmonary arteries are completely absent and all the pulmonary blood flow is derived from MAPCA's (Fig. 2). In these cases, the timing of initial intervention is less crucial, since there are no native pulmonary arteries to induce growth of. Intervention can then be based on the patient's physiologic status. However, if the patient is well balanced with saturations in the 80's and intervention in the first few months of life does not appear necessary, it is important to be vigilant if any of the MAPCA's are large without significant stenosis. Any unprotected lung segment is at risk of developing pulmonary vascular disease by as early as four to six months of age. In order to defer intervention beyond this point, cardiac catheterization must be performed to document that all lung segments are protected. If it is not clear that a MAPCA is pressure restrictive, a catheter should be inserted distally into the vessel to record pressure. Not infrequently these patients have large, hypertensive MAPCA's arising from the mid thoracic aorta and

present early in congestive heart failure necessitating surgical intervention in the first few weeks of life.

Ductal dependent circulation of one or both pulmonary artery branches

Our approach in patients with ductal dependent pulmonary circulation is to place a Blalock-Taussig (BT) shunt via a lateral thoracotomy from either the right subclavian artery in the case of a left aortic arch, or the left subclavian artery if a right aortic arch is present. In the absence of significant MAPCA's, complete intracardiac repair can then be performed at around one year of age, or sooner if the patient's oxygenation becomes inadequate with saturations below mid to high 70's. In a small percentage of cases with non-confluent native pulmonary arteries, one of the pulmonary artery branches is "ductal dependant" (Fig. 9A,B), while the other lung is supplied by MAPCA's. In these cases, although there may be enough pulmonary flow from the MAPCA's for the patient to have adequate oxygenation, it is necessary to intervene early to prevent interruption of flow to the ductal dependant branch and assure that it will continue to grow normally. The PDA is ligated and a BT shunt is inserted into that branch as the first procedure.

Figure 7 Descending aortogram in a ten-year-old patient with TOF/PA who had undergone a central shunt as an infant at an outside institution. Note the significant stenosis in the MAPCA perfusing the left lower lobe, as well as in some of the MAPCA's perfusing the right lung.

Figure 8 Selective angiogram into a large, unrestrictive MAPCA perfusing multiple segments of the left lung as well as some segments of the right lung. This patient is at risk of developing early pulmonary vascular disease if left untreated.

Figure 9 A. Selective angiogram in the patent ductus of a three week old patient who presented with severe cyanosis. The ductus arises from an anomalous left subclavian artery in this patient with a right aortic arch. It supplies the entire left pulmonary artery. At presentation at three weeks of age the ductus was nearly closed and did not respond to prostaglandin. The patient had a stenotic MAPCA to the right lung and saturations in the 40's. B. Following emergent balloon dilation of the ductus, saturations increased into the 70's. A left modified BT shunt was placed ten days later and the ductus was ligated.
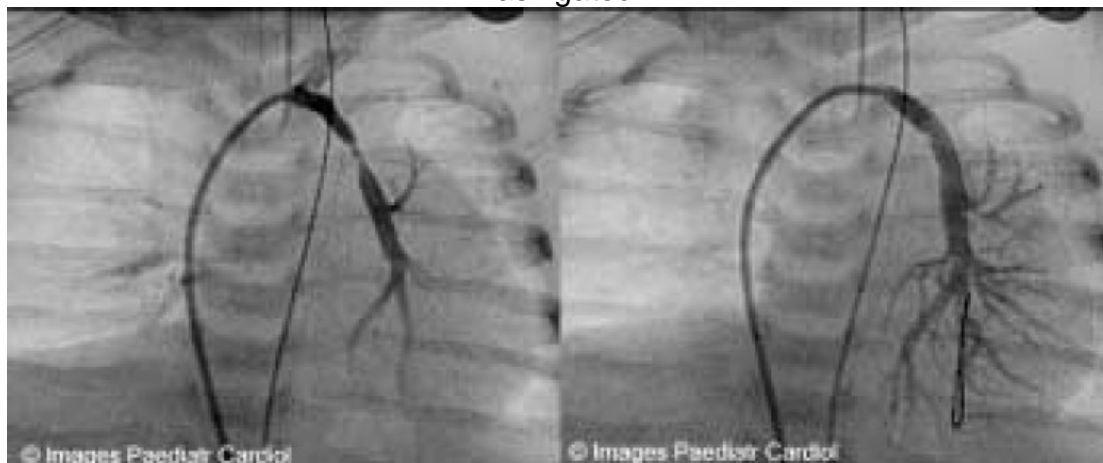
Diminutive, confluent pulmonary arteries and MAPCA's

At our institution, the majority of patients with tetralogy of Fallot and pulmonary atresia who have diminutive, confluent pulmonary arteries and MAPCA's undergo a staged reconstruction leading to complete repair by one to two years of age.[5] Timing of the initial surgical intervention is determined by the physiologic status of the patient, but typically occurs sometime between three and six months of age.[6] The initial procedure consists of a direct anastomosis between the diminutive native pulmonary arteries and ascending aorta, referred to as a Melbourne shunt. The objective is to promote growth of the hypoplastic native pulmonary arteries by providing uniform blood flow through both central pulmonary arteries at substantial enough pressure to encourage growth (Fig. 10A–F). As the native pulmonary arteries grow, pulmonary blood flow increases and patients may develop congestive heart failure. However, the orifice of the shunt typically remains small in comparison to the size of the pulmonary arteries and therefore pressure restrictive. Whether cardiac catheterization is performed prior to this initial operation depends on how well the native pulmonary arteries can be imaged by echocardiography. If there is no question that they are present and confluent, catheterization can be obviated. If there is any question about the presence or confluency of the native pulmonary arteries, cardiac catheterization must be performed before surgical intervention. Angiography in the ascending and descending aorta is performed to identify all the MAPCA's. Often from these injections, it is possible to identify the diminutive native pulmonary arteries as they are filled by collateral flow. These have the typical appearance of a "seagull" in the anterior-posterior projection and are best seen with cranial angulation (Fig. 10A). On the lateral projection, the central pulmonary confluency extends anterior to the airway with a beak-like appearance that moves in conjunction with the heart. Selective injection into each of the MAPCA's is performed to define their anatomy and determine how many of the bronchopulmonary segments are perfused. The selective injections also demonstrate intrapulmonary connections between the MAPCA's and native pulmonary arteries, and often allow optimal definition of the native pulmonary artery anatomy (Fig. 3). If the native pulmonary arteries are not opacified by any of these injections, they are most likely absent (Fig. 2). To know this with certainty, pulmonary vein wedge angiography should be performed in both right and left pulmonary veins (Fig. 11).

Although this initial catheterization is almost always diagnostic with no need for catheter intervention, it requires a significant level of expertise. It is typically performed in very small and sometimes fragile infants who may be either very cyanosed or in heart failure. It is not always easy to selectively engage each and every MAPCA, yet this is often necessary to accurately define the native pulmonary artery anatomy. In patients with absent native pulmonary arteries, a transseptal puncture may need to be performed if the foramen ovale is no longer patent in order to access the pulmonary veins for wedge angiography.

Caution should be exercised when catheterizing each MAPCA selectively. These vessels are often tortuous and may be thin walled. Care should be taken not to apply too much force when advancing the catheter around sharp bends so as to avoid vessel dissection. In a patient dependant on MAPCA's

for pulmonary blood flow injuring one of these vessels may lead to life-threatening cyanosis. Contrast injections should be performed by hand, slowly at the beginning of the injection while one appreciates the caliber of the vessel being injected as well as the position of the catheter, then increasing the rate and power as needed until the vessel is well opacified. If the tip of the catheter appears to be lodged against the vessel wall its position should be adjusted, since a forceful injection of contrast could result in dissection. When measuring pressure in a MAPCA distal to a significant stenosis or a branch point, it should be realized that the catheter itself may be large enough to cause obstruction and falsely lower the measured distal pressure.

Figure 10 A. Selective angiogram in a MAPCA in a three month old shows the severely hypoplastic native pulmonary arteries, which measure at most 1 mm in diameter. 10B: Lateral projection of the same angiogram showing the tiny central pulmonary arteries extending anteriorly. 10C: Pulmonary artery angiogram in the same patient at 18 months of age after a Melbourne shunt at three months of age and left unifocalization with placement of a 4 mm left BT shunt at 11 months of age. 10D: Lateral projection of the same angiogram performed via the left BT shunt. Note the dramatic increase in size of the central pulmonary arteries. The catheter course is from the left BT shunt into the central pulmonary arteries, and then through the Melbourne shunt into the ascending aorta. The catheter is pulled back towards the pulmonary arteries during the injection. The patient underwent complete repair at 2 ½ years of age. 10E, F. Pulmonary angiogram in the same patient at 3 ½ years of age, one year after complete repair. She underwent dilation of left middle and left lower lobe branches with RV pressure decreasing from ¾ systemic to 2/3 systemic. BT = Blalock-Taussig, MAPCA = major aortopulmonary collateral artery, RV = right ventricle.
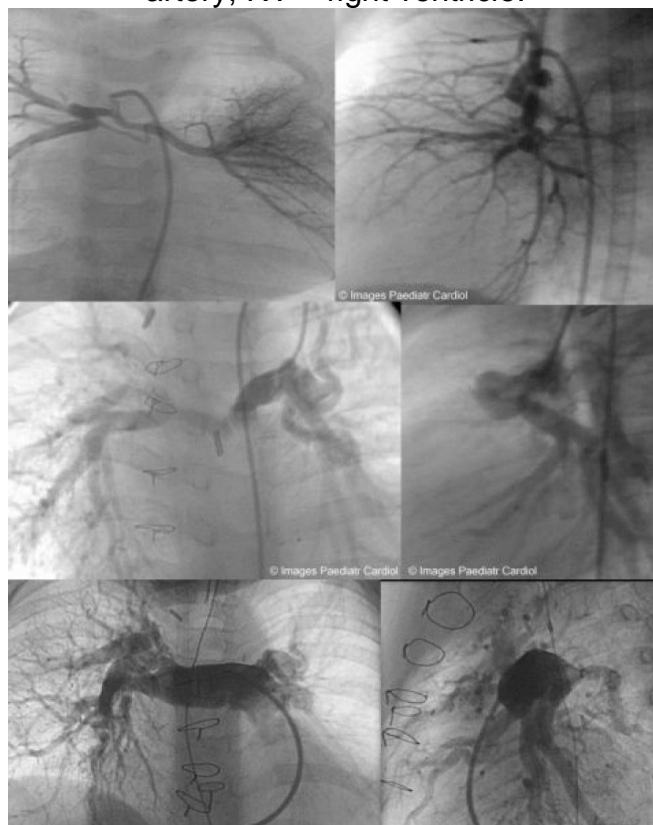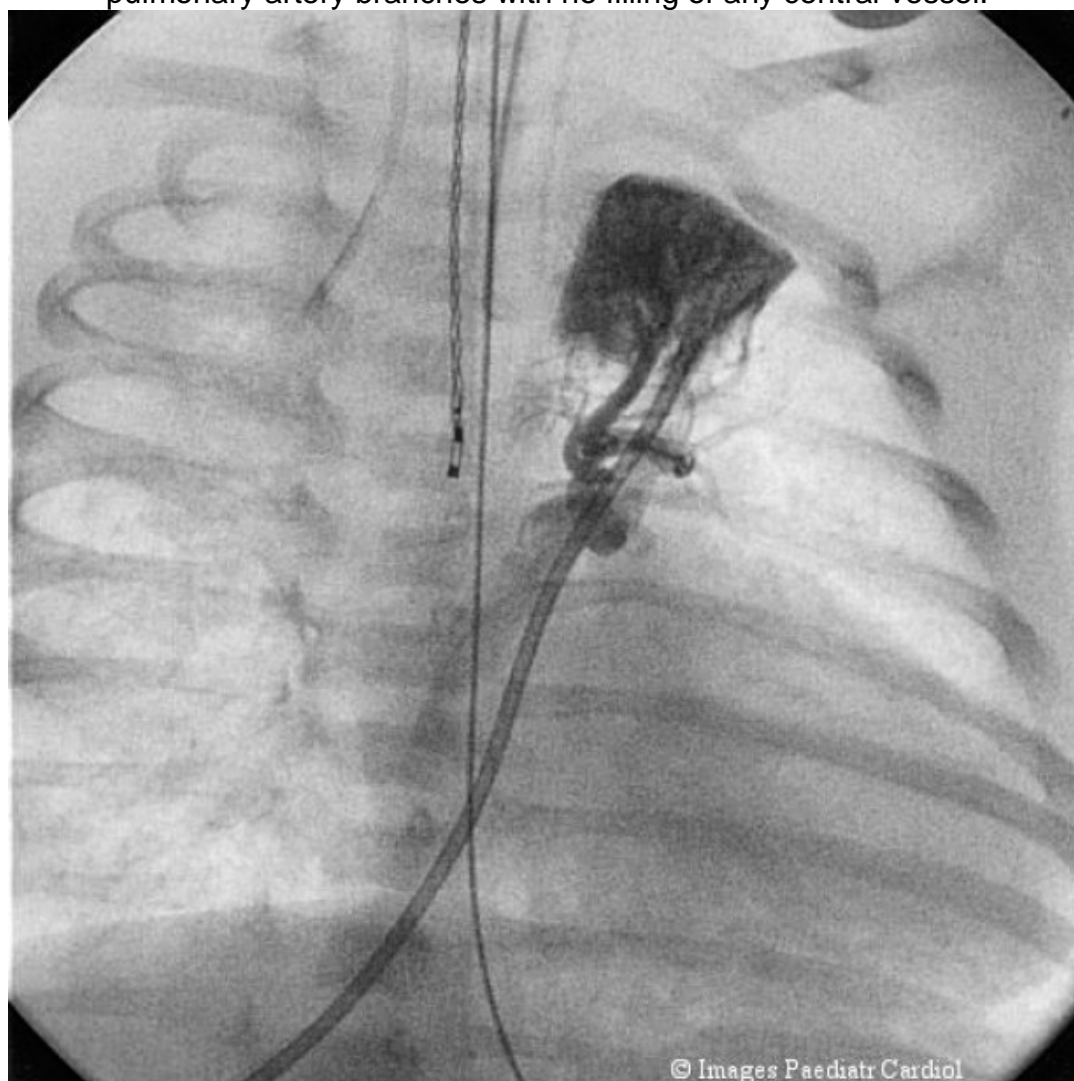
Figure 11 Wedge angiography in the left upper pulmonary vein reveals absence of a central left pulmonary artery. Note the intraparenchymal left pulmonary artery branches with no filling of any central vessel.


Unifocalization

After the initial central (Melbourne) shunt, recruitment of the MAPCA's into the pulmonary circulation is achieved via staged unifocalizations of the right and left lungs. A detailed cardiac catheterization must be performed before each of these stages to provide the surgeon with an accurate anatomic roadmap of the collaterals to be unifocalized and their relationship to the native pulmonary arteries. The Melbourne shunt must be engaged and each branch pulmonary artery selectively accessed. Biplane angiography is performed in each branch pulmonary artery (Fig. 12A,B). Selective angiography in each branch is particularly important in the lateral projection, where there is too much overlap from the left and right pulmonary artery branches to define their individual anatomy. If dual blood supply to a lung segment from the native pulmonary arteries and MAPCA('s) is documented, the MAPCA('s) can be occluded in the catheterization laboratory or ligated at the time of unifocalization. To be certain that there is adequate dual supply it may be necessary to balloon

occlude the MAPCA while performing an angiogram in the native pulmonary artery.

Figure 12 A,B. Selective angiogram in the left pulmonary artery (A) and right pulmonary artery (B) accessed via the Melbourne shunt. Note that a wire has been inserted into the distal right pulmonary artery in order to maintain catheter position in the proximal right pulmonary artery.
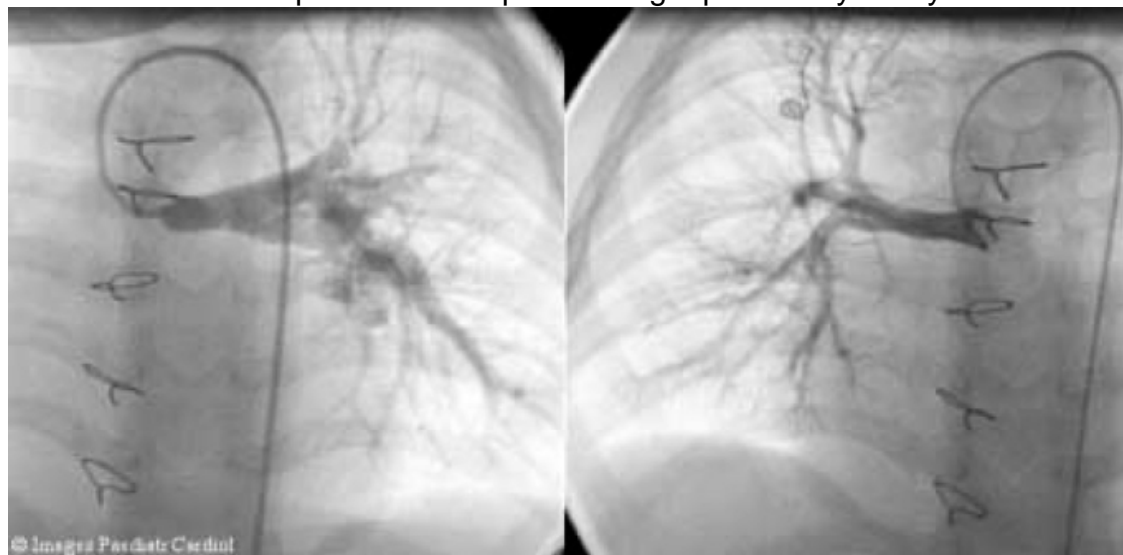

Once again, expertise in the catheterization laboratory is necessary to obtain the necessary information. The Melbourne shunt is short and narrow, and typically originates from the posterior ascending aorta. It is almost always necessary to heat shape a catheter in order to successfully engage the shunt and direct a guidewire into one of the pulmonary artery branches. Once this is accomplished, it is not always possible to advance the heat-shaped catheter, which is usually relatively stiff, through the shunt, and it must be carefully exchanged for a softer catheter. More often than not, the left pulmonary artery is more easily engaged than the right, and extensive manipulation may be necessary to make the more acute turn from the shunt into the right pulmonary artery. To avoid the need for repeat procedures, it is often preferable to have the catheterization performed in the same center performing the surgery to make sure all the required information is obtained, as well as to have the option of reviewing the angiograms with the surgeon, if there is any doubt, while in the catheterization laboratory.

Unifocalization is performed via a lateral thoracotomy. Each MAPCA is detached from the aorta and joined to the native pulmonary arteries. Although some MAPCA's have intrapulmonary connections to the native pulmonary arteries, these are often small and restrictive. The objective of the unifocalization is to recruit as many of the perfused lung segments as possible and maximize the cross-sectional area of the pulmonary vascular bed. At the same time, by unifocalizing unobstructed MAPCA's, those lung segments are protected from the development of pulmonary vascular disease. Conversely, stenoses within the MAPCA's are surgically relieved as much as possible, at the same time that these vessels are anastomosed to the native pulmonary arteries (Fig. 13A-D). The use of synthetic material is avoided, and it is almost always possible to join the MAPCA's directly to the native pulmonary arteries

or to each other. When an interposition graft is required due to excessive distance from a MAPCA, the patient's azygos vein is used to create the anastomosis.[7] The timing is again dictated by the patient's physiology, but can typically be performed approximately six months after the Melbourne shunt. If the patient is in congestive heart failure at the time of unifocalization, the side with the most unobstructed MAPCA's is attacked first. On the other hand, if the physiology is that of significant cyanosis, the side with the most stenotic collaterals is unifocalized first. At the time of unifocalization, a modified Blalock Taussig shunt may be placed if pulmonary blood flow needs to be enhanced. The timing between right and left unifocalizations is also typically around six months.

Figure 13 A-D. Right unifocalization. A. Selective injection in the right pulmonary artery via a left BT shunt demonstrates a good size branch to the right lower lobe and very small branch to the right upper lobe. B. Selective injection in a MAPCA to the right lower lobe in the same patient. Note the presence of severe mid segment stenosis. C. Additional MAPCA from the underside of the aortic arch to the right upper lobe with evidence of proximal stenosis. D. Selective injection in the right pulmonary artery following unifocalization of the two right-sided MAPCA's showing significantly improved flow to the right upper lobe, as well as increased perfusion to segments of the right lower lobe. There are no significant peripheral stenoses visible. Click to view angiogram. BT = Blalock-Taussig, MAPCA = major aortopulmonary collateral artery.
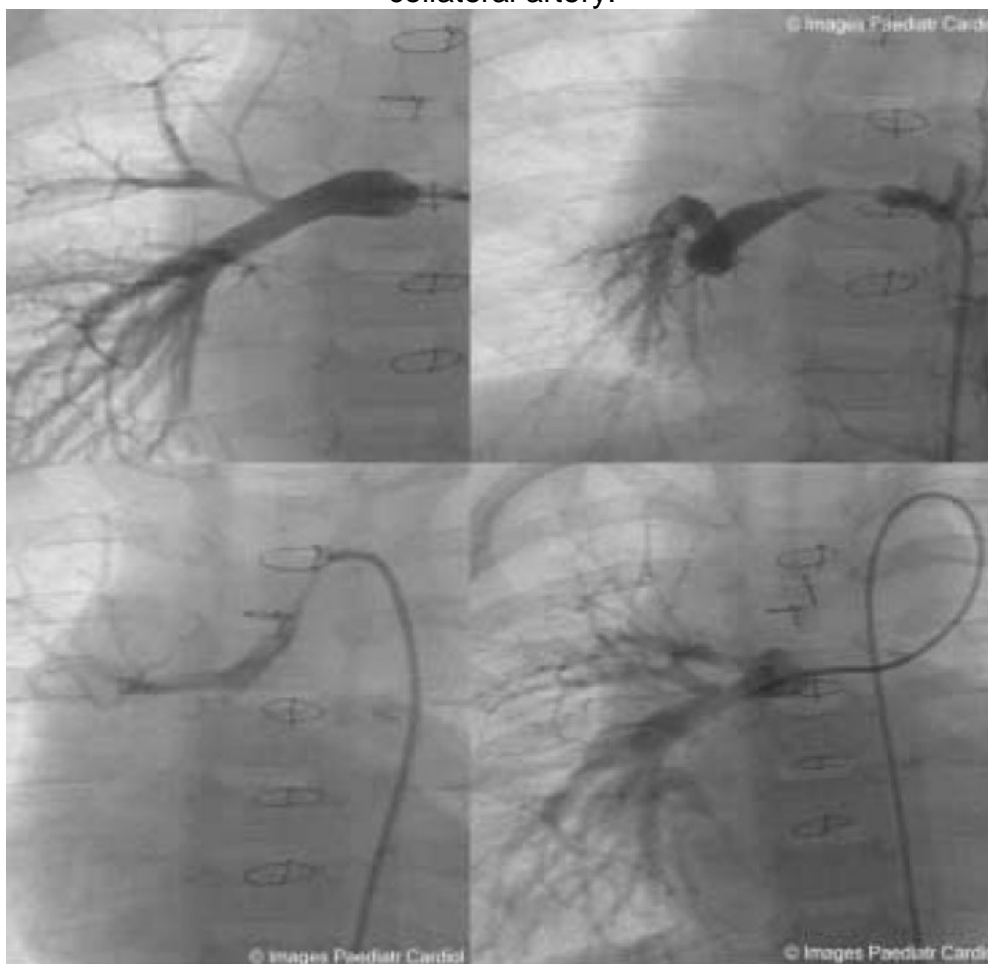
Absent native central pulmonary arteries

If the native central pulmonary arteries are absent, sequential unifocalizations are performed by detaching the MAPCA's from the aorta and surgically constructing a central pulmonary artery using a roll of autologous pericardium or pulmonary homograft to anastomose with MAPCA's. The reconstructed neo-pulmonary artery is brought into the mediastinum via a pericardial window and tacked to the ascending aorta (Fig. 14 A–D), and a Blalock Taussig shunt is inserted from the ipsilateral subclavian artery to the neo-pulmonary artery.

Figure 14 Angiography in the right pulmonary artery in anterior/posterior (A) and lateral (B) projections in a patient with absent native central pulmonary arteries following bilateral unifocalizations. A neo right pulmonary artery has been constructed into which the BT shunt is inserted. Note extension of the neo right pulmonary artery anteriorly in front of the airway in the lateral projection. The reconstructed right pulmonary artery has been brought into the mediastinum and tacked to the ascending aorta. (C), (D). Angiography in the left pulmonary artery in the same patient. A left BT shunt was inserted into the unifocalized LPA after anastomosing a large MAPCA to the LPA remnant within the lung. The patient underwent complete repair at 16 months of age using a pericardial roll to join the reconstructed right and left pulmonary arteries and a Hancock conduit was placed from the right ventricle to the pericardial roll. 14E,F. Angiogram in AP/cranial (E) and lateral projections (F) in the RV outflow tract 1 ½ years after complete repair. RV pressure was 2/3 systemic. 14G 14H 14I 14J . Peripheral stenoses in the right upper lobe branch and in the distal main RPA (G – anterior/posterior, H – lateral) were dilated with RV pressure decreasing to 55% systemic post dilation (I – anterior/posterior, J – lateral). AP = anterior/posterior, BT = Blalock-Taussig, LPA = left pulmonary artery, MAPCA = major aortopulmonary collateral artery, RPA = right pulmonary artery, RV = right ventricle. Click to view angiogram.

Final stage – complete intracardiac repair

The final stage following bilateral unifocalizations is complete intracardiac repair with closure of the ventricular septal defect and right ventricle to pulmonary artery conduit (Figs. 10,14). At the time of complete repair, pulmonary artery reconstruction is also performed as needed, relieving any stenoses that are surgically accessible. The patient's candidacy for complete repair must be carefully assessed by way of a detailed cardiac catheterization. The anatomy and physiology of the pulmonary vasculature is examined with careful measurements of pulmonary artery pressure and resistance. Not infrequently, significant distal pulmonary artery stenoses are identified in either the native pulmonary arteries and MAPCA's, or the surgically created connections between these two structures. Stenoses that appear surgically inaccessible from a median sternotomy are treated with balloon angioplasty to optimize the patient for complete repair (Fig. 15A–D). Angiography in the descending aorta should also be performed prior to complete repair to exclude the presence of any remaining collaterals. If any collaterals are found, they may need to be occluded in the catheterization laboratory.

If the calculation of pulmonary resistance suggests that the postoperative right ventricular pressure after complete repair will be prohibitively elevated, one must carefully examine the reasons why. If it is due to surgically inaccessible pulmonary artery stenoses, balloon angioplasty is performed - often more than once. Cutting balloons and high pressure balloons need to be used to achieve optimal results. After sequential ballooning over a period of 6-18 months, it is often possible to proceed to complete repair, at which time further pulmonary artery reconstruction can be performed as needed. If the reason for the elevated resistance is primarily arborization defects with large underperfused lung segments, complete repair may not be possible, or it may be necessary to fenestrate the VSD patch postoperatively to maintain adequate cardiac output.

The information obtained at catheterization prior to this final stage is crucial, as it will determine whether a complete repair can be safely performed. At times, it may be difficult for the physician doing the catheterization to know whether the surgeon can reach a specific area of peripheral stenosis or whether balloon angioplasty should be undertaken. When balloon angioplasty is necessary, access to the peripheral stenosis must be through an aorto-pulmonary shunt, with its inherent technical difficulties. It may also be challenging to decide whether a remaining MAPCA that failed to be unifocalized via a previous lateral thoracotomy should be occluded, or could be unifocalized via a median sternotomy at the time of complete repair. For these reasons, we often recommend that patients undergo catheterization at the institution where complete repair is to be performed, so that the information can be shared with the surgeon as it is obtained, and interventional decisions made jointly in the patient's best interest.

These patients are of course facing further surgical procedures typically some years after "complete repair," most commonly replacement of the right ventricle to pulmonary artery conduit because of stenosis, insufficiency or a combination of the two. It will again be important to evaluate the pulmonary artery anatomy carefully before surgical intervention, in order to alert the surgeon to any further pulmonary artery reconstruction that may be necessary at that time. These patients are typically older, and therefore can cooperate

with such non-invasive imaging modalities as magnetic resonance imaging (MRI). MRI may offer excellent anatomic detail of the pulmonary arteries at this stage, when the vessels are typically larger and therefore well within the margin of resolution of this technique. However, when physiologic data is needed, such as in patients with persistent elevation of the pulmonary artery pressure, cardiac catheterization should be performed. Catheterization is of particular importance in patients with distal pulmonary artery stenoses inaccessible to the surgeon, where balloon pulmonary angioplasty or stenting may be indicated. MRI can still be a very useful adjunct to the patient's evaluation by demonstrating the anatomy prior to catheterization. A joint review by the interventionalist and the surgeon can then lead to a plan of action best suited to the specific patient's anatomy.

The extensive anatomic variability encountered in this lesion makes each one of these patients virtually unique. It is important to approach each patient, and each stage of their reconstruction, with extreme attention to detail. The ultimate goal should be that of preserving perfusion to as many lung segments as possible and avoiding irreversible changes in the pulmonary vascular bed. The outcome of these complex patients is dependent not only on their underlying anatomic and physiologic substrate, but also largely on the surgical and interventional expertise brought to their care. It is therefore crucial that each procedure they are subjected to be performed in an experienced institution by an expert team, optimally with the interventional cardiologists and surgeons working together.

Outcome

Our institutional experience with this lesion has been recently published.[8] From a total of 46 consecutive patients, there have been no hospital deaths, and one late death due to bronchopulmonary dysplasia. Over 90% of patients in our series have either successfully undergone complete repair or are currently being staged and are considered good candidates for eventual complete repair. In the operating room, post-operative mean pulmonary artery pressure to mean systemic arterial pressure ratio had a median value of 0.36 (range 0.19-0.58). Two patients required VSD fenestration 2 and 3 months after complete repair due to a rise in right ventricular pressure without the development of new peripheral pulmonary artery stenosis. This compares favorably to other published series with an early mortality rate of 10.6% and overall mortality of 18.8%.[9] The surgical approach in this latter series was an attempt at single stage unifocalization and full intracardiac repair. We believe a staged surgical reconstruction optimizes the recruitment of collateral vessels in a fashion that is matched to the patient's physiology at each stage. The crucial decision of whether to proceed to complete intracardiac repair is made at a time when the pulmonary anatomy and physiology have been optimized and can be well assessed. The sources of pulmonary blood flow are controlled early to avoid exposure of unprotected lung segments to high pressure.

During long-term follow-up over a median of 40 months, the right ventricular to left ventricular pressure ratio has averaged 0.51 + 0.24. Over 60% of these patients have required further interventions, most commonly balloon angioplasty of distal pulmonary artery stenosis[8] (Figs. 14,16). Other series

have reported similar reintervention rates.[5,9,10] These patients require ongoing close follow-up with particular attention to non-invasive estimation of right ventricular pressure and distribution of pulmonary blood flow. Cardiac catheterization should be performed if a progressive increase in right ventricular pressure is documented or if there is marked asymmetry of flow to the right and left lungs, and pulmonary artery angioplasty and/or stenting undertaken if warranted. Longer-term follow-up should continue so that we can better assess the optimal management strategies among the various currently advocated algorithms.

Figure 15 A. Right pulmonary artery angiogram demonstrating significant long segment stenosis. The stenosis extends fairly distally into the hilum, best appreciated in the lateral projection (B). Initial angiography was performed with a catheter advanced retrograde through the central shunt. C, D. Following balloon angioplasty, there is marked improvement in the stenosis. Note the catheter is now accessing the shunt in a prograde fashion from the right ventricle to the ascending aorta and through the central shunt. By accessing the pulmonary artery from a venous approach to perform balloon angioplasty, it was possible to avoid placement of a large sheath in the femoral artery.
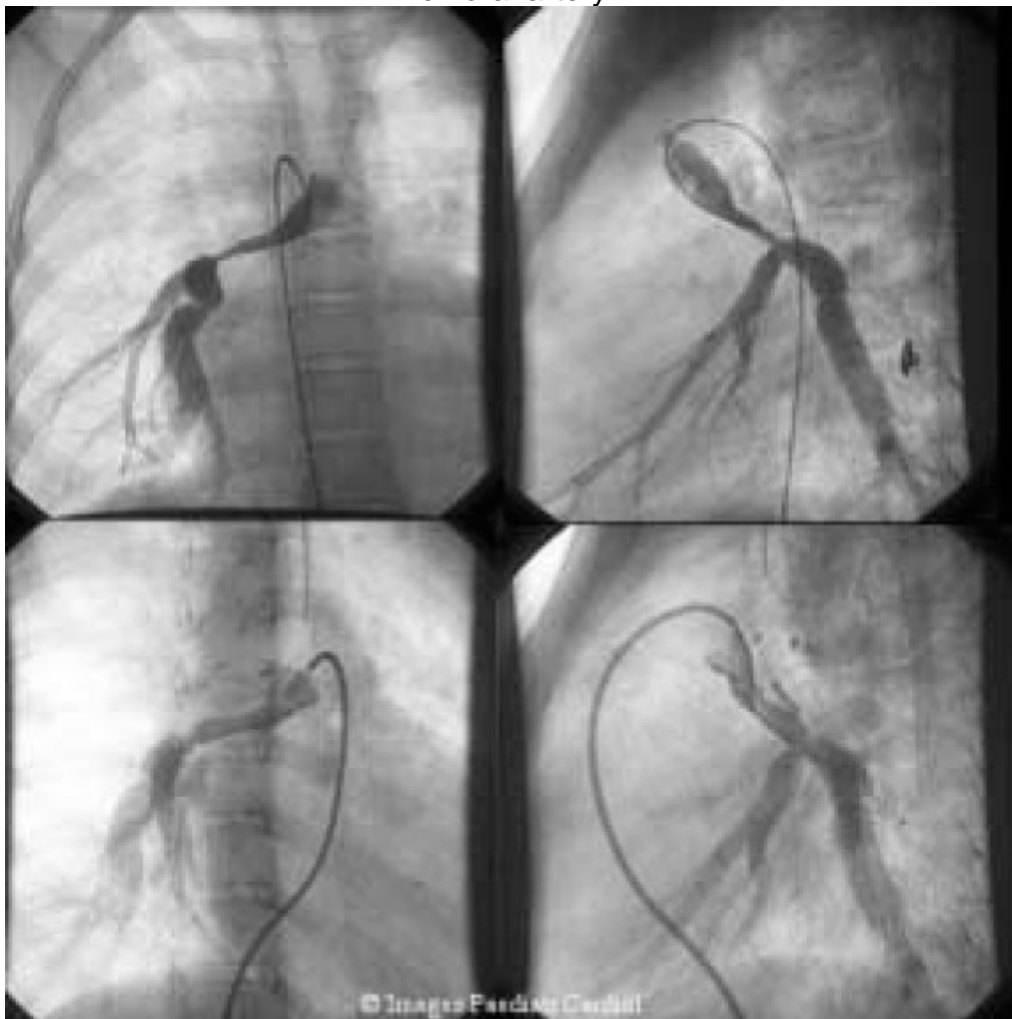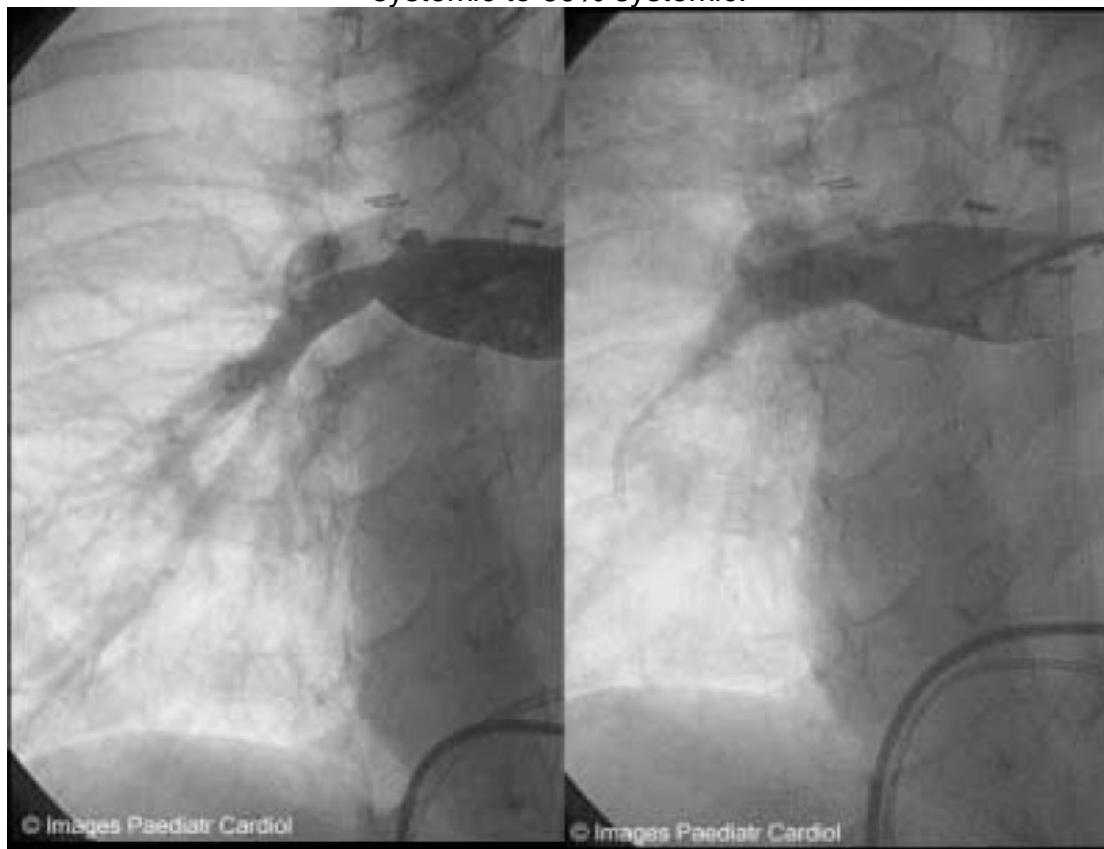
Fig. 16 Right pulmonary artery angiogram before (A) and after (B) balloon angioplasty in a patient two months after complete repair. The left pulmonary artery was also dilated. Right ventricular pressure decreased from 80% systemic to 60% systemic.

Acknowledgments

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