Chapter 8
Pulmonary Atresia, Ventricular Septal Defect and Major Aorto-Pulmonary Collateral Arteries

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Abstract The term pulmonary atresia and ventricular septal defect (VSD) covers a spectrum of conditions with various degrees of severity. The terminology of pulmonary atresia, VSD and major aortopulmonary collateral arteries (MAPCAs) is usually restricted to patients with diminutive central pulmonary arteries whose pulmonary circulation is dependent from systemic-pulmonary collateral arteries. It therefore excludes patients with ductus-dependent pulmonary circulation. The natural history of the condition is severe. In the 90s, only a fifth of the patients born with pulmonary atresia, VSD and MAPCAs survived to 30 years of age.

Significant progress has been made in the management of patients with pulmonary atresia, VSD and MAPCAs. Our understanding of the variations in pulmonary vasculature has evolved, although there remains debate about the best methods of classification. There is an ongoing debate on optimal surgical strategies between surgical rehabilitation of the native hypoplastic PA that our group favors and unifocalisation of MAPCAs early in infancy. There continues to be debate on the most effective strategy to provide long-term survival in this patient group.

Keywords Pulmonary atresia • Ventricular septal defect • Major aortopulmonary collaterals • Unifocalisation • Rehabilitation of pulmonary arteries • Congenital heart defect

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Background

In the 70s, attempts had been made to shunt the small native vessels of these patients with poor outcomes [1]. Subsequently, strategies of unifocalisation were developed whereby collateral vessels were joined to either native pulmonary vessels that were previously shunted to increase their size, or to centrally reconstructed vessels. Initially, unifocalisation was performed as a staged procedure with some via sternotomy to enlarge the size of the central native pulmonary arteries, and others performed by thoracotomy to anastomose collateral arteries to centrally reconstructed pulmonary vessels [2]. Later these unifocalisation procedures were completed in one stage via a sternotomy, even at an early age [3]. Doubt has been cast on the long-term benefits of the translocation of collateral arteries by unifocalisation procedures [4–6].

Today, treatment strategies are divided between two diverging approaches: rehabilitation of the native pulmonary vessels and unifocalisation [7–10]. Some have proposed the use of both approaches following the patients' own anatomic characteristics [11].

Rehabilitation Strategy

Supporters of this approach claim that central pulmonary arteries are present in the majority of neonates [12]. Before rehabilitation of these central vessels is attempted, the exact distribution of these vessels to lung fields should be evaluated. It is believed that these central pulmonary vessels have a better chance to grow if procedures are aimed at increasing flow in early life [9, 13]. The initial procedure is performed at 2–3 weeks of life, even if the patients are well saturated. In patients showing signs of heart failure related to pulmonary over-circulation, the procedure may be delayed if symptoms can be controlled with medical therapy. The rehabilitation approach requires multiple interventions upon the central pulmonary arteries. The second procedure usually consists of the insertion of a right ventricle (RV) to pulmonary artery (PA) conduit. The conduit is gradually increased in size during repeat procedures performed during the first years of life. Almost all patients necessitate one or more procedures involving patching of the central pulmonary arteries, generally from hilum to hilum. Complete repair, including the closure of the VSD and the insertion of a valved conduit between the right ventricle and the pulmonary arteries, is only performed when the clinicians have the subjective impression that the pulmonary vasculature is developed enough and the central vessels are connected to enough of the overall lung parenchyma. This type of repair may be performed as a second procedure or be delayed until the third, fourth or even fifth procedure.

Exception to this strategy includes patients who present with near normal native pulmonary vasculature in both hila. These patients tend to have one or two MAPCAs feeding these vessels. A single stage repair procedure in early life will connect the hilar vessels to either native central vessels enlarged with patches, pericardial tubes or Gore-Tex grafts. Additional procedures may be required in cases where native lobar branches fed by MAPCAs, are not in continuity with the central vessels. These

near-normal lobar branches can then be connected to the rehabilitated central vessels either directly or via an interposition graft.

The benefit of the rehabilitation strategy is its relative safety. As long as the MAPCAs are left undisturbed, the pulmonary blood flow is maintained. We have observed in some instances the spontaneous regression of these collateral vessels while in others there is increased pulmonary blood flow resulting from the growth of the native vessels. This has necessitated surgical ligation of collateral vessels or occlusion by interventional catheterisation procedures.

Unifocalisation

In its early development, unifocalisation was performed in stages [2]. Procedures were performed with the aim to rehabilitate central pulmonary vessels via a sternotomy and to translocate collateral vessels to the central vessels via thoracotomy. This strategy has now been virtually abandoned. Today most teams would proceed with one-stage unifocalisation of all or most collateral arteries [10, 14]. The principles of this approach are seemingly clear but in most series, the unifocalisation strategy is applied with some variation between individual surgeons and patients within the one institution. Some patients have had previous procedures on the central pulmonary vessels in an attempt to develop these vessels prior to unifocalisation, while others were late referrals to a "unifocalising" team.

The collaterals are detached from their insertion, mainly the descending aorta and the subclavian vessels, and the most distal segment is reattached to the centrally rehabilitated or reconstructed vessels. Practices in unifocalisation procedures vary from the translocation of all identified collateral vessels, to the translocation of selected collaterals deemed to be the most important contributors to the pulmonary vasculature. The decision of the suitability for complete repair varies between teams. Most appear to base their decision on the subjective appreciation of the adequacy of the reconstructed pulmonary vessels and degree of peripheral arborization. It has been suggested to cannulate the reconstructed pulmonary circuit, and measure the pressure while maintaining a full cardiac output in the pulmonary arteries. This can be achieved by an independent bypass circuit and is thought to enable prediction of the pulmonary artery pressures post repair [8].

Anatomical Classification of the Pulmonary Arterial Supply

Brawn and Hanley, two pioneers of this field with the largest described experience, vary in their classification of the anatomy of the pulmonary vasculature. Brawn identifies patients as having "confluent intra-pericardial pulmonary arteries, confluent intra-pulmonary arteries and non-confluent intra-pulmonary arteries". While Hanley described four patient groups; "large caliber MAPCAs without significant segmental level stenoses, small-to-moderate caliber MAPCAs without segmental level stenoses,

centrally confluent fully arborizing hypoplastic true pulmonary arteries with dual supply MAPCAs, and MAPCAs with extensive segmental level stenoses" [8, 15]. While there is no clear consensus on a single, specific classification, there is general agreement that pulmonary atresia, VSD and MAPCAs is spread across a spectrum of severity. In its most benign form, there are central pulmonary arteries which are small and connected to the majority of the lung fields. At the other extreme, there are no visible native central pulmonary vessels or they are severely hypoplastic, with the main source of pulmonary blood flow arising from multiple small collateral vessels which have no resemblance to normal intra-pulmonary vasculature. Pulmonary blood flow distribution may not be uniform across both lungs. In the extreme, one lung vasculature may be normal and dependent upon a ductus or a hemitruncus, while the other lung depends exclusively on diminutive collateral vessels.

In the absence of an agreed classification, we encourage clinicians to identify the characteristics that will be the most important to their individual management strategy.

Central Pulmonary Arteries

Central pulmonary arteries are incorporated in both the rehabilitation and unifocalisation strategies. This may be as a unique target of revascularization, or as a central point of reconstruction. The discrepancy in the appreciation of the proportion of patients with native central pulmonary arteries may be explained by the way they are managed. A strategy of early surgery is more likely to identify the native central pulmonary arteries while unifocalisation may not allow identification of these vessels especially if they are referred late [8, 12].

Near-Normal Intra-Pulmonary Pulmonary Arteries or Pulmonary Lobar Branches

MAPCAs may be connected to intra-pulmonary pulmonary vessels that appear to have a normal hilar distribution. They can also be directly connected to individual lobar branches with a normal appearing intra-pulmonary course. The goal is to identify these vessels and connect them to the central pulmonary circulation.

Multiple, Small Diminutive MAPCAs

Some collaterals do not connect to any vessels that bears resemblance to native pulmonary vasculature. They can be identified by their abnormal course (intimate contact with the bronchial tree), tortuous or tubular appearance, and their limited distribution to small lung segments with a lack of normal pulmonary arborization.

These vessels would be targeted in aggressive unifocalisation strategies and neglected in rehabilitation strategies. There is a general agreement that these patients are at the worst end of the spectrum of the condition [5, 16] and very often, have diminutive central pulmonary arteries.

Dual Pulmonary Blood Supply

Pulmonary segments or lobes may be vascularized by both native pulmonary vessels and MAPCAs. In rehabilitation strategies, dual blood supply can be the source of competitive flow. This has been identified to be a potential source of failure during rehabilitation of lobar branches. Dual blood supply can lead to pulmonary overcirculation and heart failure, precipitating the need for ligation or catheter occlusion. It is important to appreciate the presence of dual blood supply and consider preemptive MAPCA ligation at the time of surgical intervention. Those undertaking a unifocalisation strategy may decide to translocate these collateral vessels regardless of whether or not the territories have dual supply.

Diagnosis and Imaging

At birth, the intra-cardiac diagnosis is usually made by *transthoracic echocardiography*, but more accurate definition of the pulmonary blood supply is required pre-operatively.

Historically, all patients bearing this diagnosis underwent *cardiac catheterisation and selective angiographic examination.* Selective injection of contrast into collateral vessels allowed the most accurate demonstration of the sources of pulmonary blood supply to each lung segment. Today, this investigation is no longer performed at birth because of the need for vascular access, general anaesthesia and radiation exposure. Those opting for a rehabilitation strategy will attempt an early central shunt notwithstanding the quality of distal vessels. Those attempting aggressive unifocalisation will need to identify only the proximal portion of the collateral arteries and the timing of their procedure will be based on the clinical status of the patients rather than the morphology of their pulmonary circulation. Imaging of the distal vessels may be performed at an older age, before the planned unifocalisation.

Cardiac magnetic resonance imaging (cMRI) and Computerized Tomographic Angiography (CTA) are the more common, less invasive techniques employed for imaging the pulmonary vessels. While image quality may not be as accurate as selective catheter-based angiography, the anatomical definition provided is more than adequate for surgical planning. General anaesthesia is still required for cMRI due to the amount of time required to obtain the images and CTA exposes the patient to radiation but may not necessarily need general anaesthesia. Clinicians must

weight up the risk-benefits in each individual patient and institution, but cMRI would be the preferred technique if available.

After the initial rehabilitation procedure, the progress in the growth of the central pulmonary arteries can usually be followed by echocardiography. More accurate measurements and extensive imaging of the distal vessels would be ideally be performed by cMRI or CTA, again depending on availability and institution preference.

Before repair or unifocalisation, cardiac catheterisation and extensive angiography is necessary. This allows for haemodynamic assessment of each pulmonary segment and accurate definition of the distribution of the central and collateral vessels, and their peripheral arborization. In particular, aortic root and ascending aorta angiography will demonstrate rare coronary collaterals and more proximal MAPCAs. The descending aorta should be injected with distal occlusion to allow back-filling of collaterals. Both subclavian vessels should be selectively injected because they are commonly a source of MAPCAs, especially on the right side. The superior portion of the abdominal aorta may similarly give rise to collateral vessels. Each identified MAPCA should be injected selectively.

Checklist Before Surgery

Mapping of PA and MAPCAs based on selective Angiogram and/or CTA-MRI
Nakata index or total neo-pulmonary artery index
Material to be ready:

- Gore-Tex shunts,
- Patch enlargement material: 0.4 mm Gore-Tex, autologuous pericardium (fresh and gluteraldehyde tanned)
- RV to PA valved conduit: pulmonary or aortic homografts, Bovine Jugular Vein conduit (Contegra)
- If perfusion of the main pulmonary artery is planned, the necessary circuit has to be prepared.
- Potential Intra-operative stenting

Surgical Techniques

Timing of Surgery

The timing of the surgery varies depending on the type of strategy employed. In a rehabilitation strategy, the first procedure is recommended to take place at 2–3 weeks of age [9]. Patients directed to a unifocalisation strategy are usually offered their first surgery from 3 to 9 months of age [7, 8]. As most patients with well-developed

MAPCAs are initially only moderately desaturated, there is no clinical need for the patients to be operated early. There appears to be a general consensus on clinical indications for early surgery: the rare cases of patients in heart failure despite medical treatment, and those with a ductus or a hemitruncus providing circulation to one lung.

The type of surgery depends of the anatomy of the patients. Patients with adequate intra-pulmonary branches should be offered a definitive repair. Those with a well-developed pulmonary circulation on one side and diminutive collateral arteries on the contralateral lung should undergo a connection between the right ventricle and the well-developed lung circulation and a rehabilitation procedure in the contralateral lung [17].

The decision to proceed with the definitive repair and ultimately the closure of the VSD depends of the appreciation of the development of the ultimate pulmonary circulation. Traditionally the measurements of the Nakata index of the reconstructed pulmonary circulation has been the predominant factor guiding this decision. An index superior to 150 mm/m^2 is usually considered as suitable to achieve a repair [18]. The total neopulmonary artery index, a calculation of the total surface of the pulmonary blood vessels supplying the lungs notwithstanding their origin has been quoted to equally be a good estimate of the postoperative pulmonary pressure with similar values being considerate adequate for repair [11, 19]. Others have suggested to base their decision on preoperative MRI estimates of the pulmonary blood flow calculated as the total pulmonary venous return or by subtracting superior vena cava and descending aortic flow from ascending aorta flow [19]. Apart for some rare teams, decision to proceed to repair is no longer based on the number of bronchopulmonary segments perfused by the reconstructed or rehabilitated pulmonary vasculature [20]. In borderline cases of patients undergoing unifocalisation, intraoperative flow testing of the pressure achieved in the pulmonary circulation has been maintained by some teams who favours unifocalisation procedures [21, 22].

Rehabilitation of Native Pulmonary Arteries

Central shunt following Laks technique (Fig. 8.1) The creation of a shunt between the ascending aorta and the main pulmonary artery appears to be the best way to provide increased pulmonary blood flow to the small native central pulmonary arteries without distorting the branches. With this technique, the procedure can be performed without the need for cardio-pulmonary bypass. The branch pulmonary arteries are gently snared. A single 5/0 suture is passed at the base of the main pulmonary arteries. Traction on this suture and the two snugger's provide optimal exposure of the main pulmonary artery. A longitudinal incision is made on the main pulmonary artery. This incision is central and not directed towards either of the branches. The largest possible shunt is inserted at this level, but in the majority of patients (based on age/weight at the time of procedure), this is usually limited to a

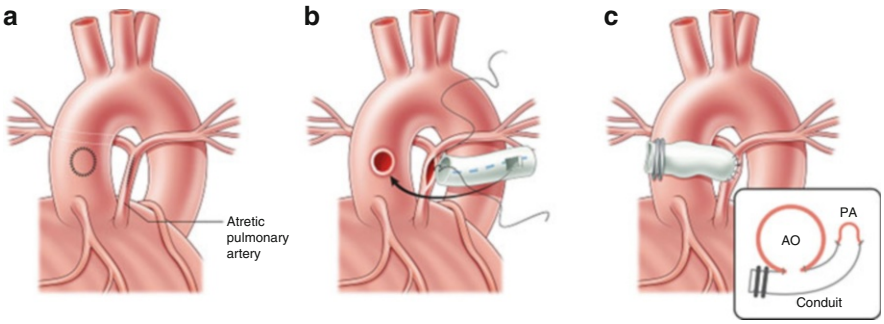

Fig. 8.1 (**a**, **b**, **c**) Central shunt following Laks technique

3.0 or 3.5 mm diameter shunt. Most of the time the extremity of the shunt does not need to be beveled as a transverse cut fits adequately to the vessel opening. After relieving traction on the suture, the length of the shunt is estimated. A slight upwards or transverse course gives the best curvature to the shunt. A rectangular slit incision is made on the shunt and a side-biting clamp is applied on the ascending aorta. The proximal orifice is best made with a punch of 2.8 or 3.5 mm diameter. After the completion of this side-to-side anastomosis, the shunt is de-aired and two clips are applied on the proximal end. Care should be taken to avoid positioning any chest drains across the path of the shunt.

Alternative techniques It has been recommended to divide the proximal end of the main pulmonary artery and re-implant the main pulmonary artery directly on the side of the aorta [16]. We have found this technique to be unreliable. In our review of this technique, close to half of the patients developed right pulmonary artery stenosis [4]. Invariably, these patients have a large aorta and we have found that after this technique, the right pulmonary artery may become adhered to the back of the aorta. Others have proposed to perform an opening and patching of the occluded right ventricular outflow tract [18]. This technique is more technically demanding and extensive patching may increase the risk of distortion of the proximal pulmonary arteries compared to performing a central shunt. Additionally, this procedure requires cardio-pulmonary bypass with cardioplegic arrest.

RV to PA conduit (Fig. 8.2) The second procedure of a rehabilitation strategy is commonly a definitive repair utilising an RV to PA conduit. This is performed as the infant outgrows the shunt and requires increased pulmonary blood flow to the central pulmonary vessels. The conduit is positioned between the right ventricle and main pulmonary artery. It provides a large amount of pulmonary blood flow in additional to giving the interventional cardiologist access to distal pulmonary arteries. At this stage, the central pulmonary vessels have increased to a size that decreases the risk of any distortion of the branch pulmonary arteries. We have found a 6 mm Gore-Tex conduit to be the ideal size around the age of 6 months. Later in life, we use the smallest size Contegra conduit of 12 mm diameter. By this stage, the central

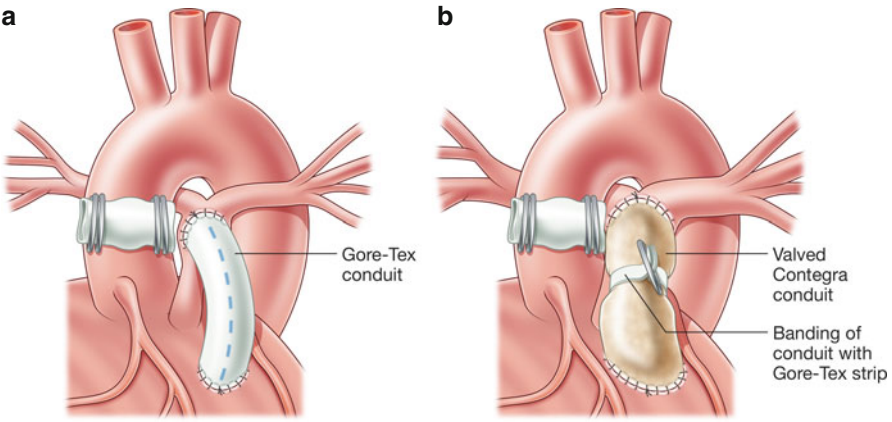

Fig. 8.2 Creation of a RV to PA conduit with (**a**) a Gore-Tex conduit and (**b**) with a Contegra conduit banded with a strip of Gore-Tex

pulmonary arteries have developed and patients are at risk of pulmonary over-circulation. Our strategy is to band all valved conduits while monitoring their pulmonary pressure. Banding of the conduit is performed with strips of Gore-Tex, secured with clips, to allow subsequent balloon dilatation of the conduit if required, which loosens the band and increases pulmonary blood flow.

Pulmonary artery patching (Fig. 8.3) During any of the procedures following the initial central shunt, it may be necessary to proceed with the patching of the central pulmonary arteries. Our preference has been to always enlarge both origins of the pulmonary arteries with an incision cephalad to the bifurcation. Patching is then extended to as far as necessary, sometimes crossing the origin of a pulmonary lobar branch. Patching of the pulmonary artery bifurcation results in the alteration of the shape of the bifurcation from a 'V' to a 'T' configuration.

Repair Repair is ultimately performed when there has been adequate growth of the pulmonary arterial branches and the central pulmonary vessels are connected to enough lung segments. In addition, lobar branches connected to MAPCAs, can be integrated into the native pulmonary circulation. It is very frequent to have to proceed with additional patching of the central pulmonary arteries concomitantly with the repair. Central pulmonary arteries which have been patched repeatedly have impaired long-term growth. In these circumstances, we favor the interposition of an adult size Gore-Tex graft conduit between both hila whenever possible (Fig. 8.4). We proceed with the repair of the central pulmonary arteries on bypass with the beating heart before proceeding to the VSD closure. The distal end of the RV to PA conduit is anastomosed to the pulmonary artery bifurcation before closing the VSD. In our experience, transection of the aorta is never necessary to access the pulmonary arteries. They can be accessed by gentle retraction of the aorta on both sides. After cardioplegia is administered the right ventriculotomy is performed. A

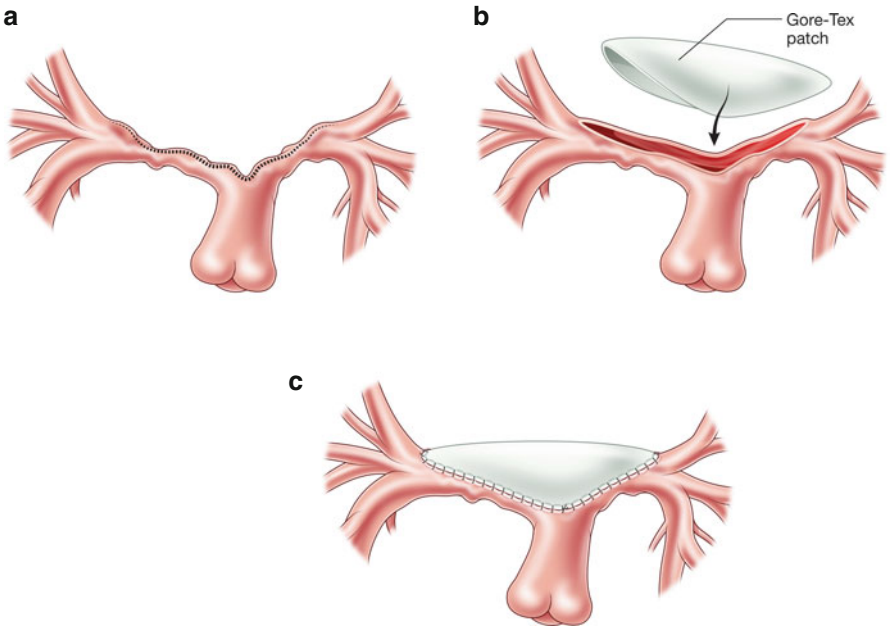

Fig. 8.3 (**a**, **b**, **c**) Patch reconstruction of the central pulmonary arteries

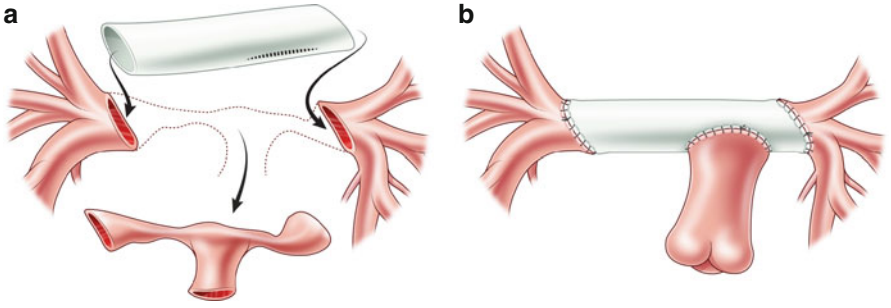

Fig. 8.4 (**a**, **b**) Replacement of the central pulmonary vessels with an interposition Gore-Tex graft

circumferential opening is made in the best identified location, usually below the level of infundibular obstruction. The area for the ventriculotomy is identified by examining the ventricular free wall from the outside and through the tricuspid valve, taking great care to stay away from the left anterior descending coronary artery and the aortic valve. After closure of the VSD, the proximal anastomosis of the conduit is completed. When there are concerns about the suitability of the pulmonary circulation, we have proceeded with closure of the VSD using a fenestrated flap patch or alternatively we have left a patent foramen ovale.

Additional procedures MAPCAs may need to be ligated if there is dual blood supply resulting in pulmonary over-circulation and heart failure, or because competitive flow results in a lack of growth of the rehabilitated pulmonary artery branches. The vascularization of the trachea and the bronchial tree is dependent upon some of these collateral vessels and tracheal necrosis has been described after the ligation [23]. We therefore avoid the systematic ligation of collaterals unless needed. Intra-pulmonary arterial branches may be reconnected to the centrally reconstructed circulation. There are two ways to access these branches. Whenever these branches are in continuity with a hypoplastic central vessel, dissection can proceed from this vessel and continue until the intra-pulmonary branch is adequately exposed. Alternatively, the dissection can proceed from the proximal segment of the MAPCA feeding the vessel, and be followed as distally as possible. These intra-pulmonary branches may then be connected to the pulmonary circulation directly, or by an interposition Gore-Tex graft.

Unifocalisation

Historically, unifocalisation procedures have been performed as staged procedures with multiple thoracotomies required at times. These techniques are now obsolete and most teams would proceed with a single-stage unifocalisation procedure. The most appropriate timing for the procedure will vary between teams and their preferred management strategy.

The procedure starts with an extensive time of dissection of the collaterals. It is possible at times to access these collaterals directly from the posterior mediastinum or directly in the hilum. At the best of times, these approaches are difficult and the identification of the vessels is not straightforward. It is therefore easier to identify the MAPCAs from their origin. Dissection proceeds from the transverse arch and is continued along the descending aorta. The dissection is made from an incision of the posterior mediastinum located on the left side of the SVC. The incision is extended caudally under the carina and may require the retraction of the roof of the left atrium. In rare circumstances, dissection requires the opening of the posterior mediastinum on the left side of the aorta, lifting up the heart to the right, while on cardio-pulmonary bypass. All attempts should be made to anastomoses these vessels as distally as possible as it is likely that the proximal segment of these collaterals will not allow adequate growth.

The team of Dr Hanley suggested proceeding with the isolated perfusion of the newly recreated pulmonary circulation with invasive pressure measurements in the central pulmonary vessels to identify whether the patients were suitable for the closure of the VSD. The cut-off of achieving a pulmonary flow of 3 l per minute per square meter and maintain a pulmonary to aortic pressure ratio of less than 0.4 has been suggested [16]. Others have proposed using the pre-operative cMRI evaluation of pulmonary blood flow [19].

Outcomes

To date, there have been no in-depth evaluations of the late functional capacity of patients born with pulmonary atresia, VSD, and MAPCAs. In reality the success of the various strategies can only be evaluated using three main outcome parameters: survival, number of patients palliated without complete repair, and right ventricular pressure after complete repair.

Survival

There is no data on the natural history of the disease. In a seminal manuscript of patients from the UK undergoing varied surgical strategies, survival to the age of 30 years was barely 20 % [24]. Following a strategy of staged procedures with unifocalisation, our team achieved a 30 year survival of 57 % [4]. More recently, groups performing unifocalisation have reported 3 year survival of 89 % and 5 year survival of 85 % [8, 20]. In a much smaller series of 25 patients using a rehabilitation strategy, we encountered only one unrelated death [12]. On the basis of these preliminary results, our institution believes higher survival may be achieved by pursuing a rehabilitation strategy. As long as the collateral arteries are not compromised and pulmonary blood flow is adequate, survival may be improved.

Palliation Without Complete Repair

Review of all strategies, demonstrates that some patients will not achieve a complete repair. In a historical series of staged palliation, 35 % of patients were palliated [4]. The team of Dr Hanley and Dr Brawn have reported this rate to be 10 and 44 % respectively [8, 20]. In our recent small series, 10 % of the patients were palliated after being deemed unsuitable for complete repair [12].

Right Ventricle Pressure After Complete Repair

This pressure is most often a reflection of the quality of the pulmonary circulation. While low physiological pressure is desirable, the minimum acceptable level of pressure for the right ventricle to provide adequate functional capacity over the course of an entire life-time is still unknown. It has been suggested that right ventricular systolic pressure should be less than 0.4 of the left ventricular systolic pressure. The Hanley team demonstrated that in their patients undergoing repeat conduit replacement, both early post-repair and late after replacement, the ratio between the

right and left ventricular systemic pressures were below 0.4 [25]. In our small series describing the rehabilitation strategy, the ratio between the right and left to ventricular pressures was 0.57, and between the pulmonary artery and the left ventricle was 0.4 [12].

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