

Pulmonary Atresia, VSD and Mapcas: Repair Without Unifocalization

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The last three decades have seen considerable development in the management of infants with pulmonary atresia, ventricular septal defect, and major aorto-pulmonary collaterals. The technical difficulty of surgical treatment lies in the fashioning of a pulmonary vascular bed capable of providing suitably low right ventricular pressure for repair of the intracardiac defect. With the current trend toward early and complete unifocalization of major aortopulmonary collateral arteries (mapcas), we describe an alternative approach - "repair without unifocalization" - and explain our reasoning for pursuing this technique.

Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 12:139-144 Crown Copyright © 2009
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The cardiac surgery unit in Melbourne has been at the forefront of the surgical treatment of pulmonary atresia, ventricular septal defect (VSD), and major aortopulmonary collateral arteries (mapcas) from the late 1970s to the mid 1990s. Iyer and Mee¹ introduced and promoted the staged recruitment of lung segments that were initially or secondarily isolated from the pulmonary arteries and solely supplied by mapcas. The preparatory procedures consisting of central shunting and unifocalization of mapcas to native pulmonary artery (PA) branches were performed by multiple sternotomies and thoracotomies. If no central PA branches were found or were not suitable to receive unifocalization, they were reconstructed with autologous pericardial rolls. Once all or most of the pulmonary segments were connected to the PA tree, patients were offered a complete repair with trans-ventricular closure of the VSD and implantation of a valved conduit between the right ventricle and the PA.

This approach has led to significant success in terms of operative mortality. However, the complexity of the approach and aspects of the pulmonary vascular bed after completion of repair led us to revisit this strategy. For a while, several different strategies were used simultaneously in the unit; including one-stage repair from the front in infancy,² as well as central shunting to promote growth of the diminutive central pulmonary arteries, and our own traditional ap-

proach described above. In 2003, we initiated a review³ of the long-term outcome for the patients who underwent the multi-stage approach with unifocalization.

We found the multi-stage approach generated a radical improvement in long-term survival compared with the natural history of the disease, with a greater than two-fold increase in the probability of survival at 30 years of age.³ However, other results of the retrospective review have been disappointing: the survival probability of the patients who had received a correction was identical to those who remained palliated, although the exercise ability was improved. Moreover, the study of serial angiographies of the unifocalized mapcas has demonstrated that of the 60 unifocalized mapcas identified on serial angiographies in 31 patients, 26 had thrombosed, 12 had stenoses greater than 50%, and 22 failed to grow ($P = .25$).³

The retrospective analysis suggested that although the increased vascular bed recruited by unifocalization was sufficient to allow a safe repair shortly after the completion of unifocalization, it subsequently failed to achieve adequate growth. Progressive right ventricular failure ensued, leading to late death or poor exercise ability. The probability of being alive and in NYHA class I 12 years after the initial procedure was $32\% \pm 10\%$. This suggested as well that the quality of the long-term repair was dependant on the size and compliance of the central pulmonary arteries, as well as the number of segments included in the repair. Review of the angiograms not only failed to demonstrate any growth of the unifocalized mapcas, but also inferred that growth of the PA branches themselves were compromised by the unifocalization process.

The following postulate summarizes the lessons learned from our retrospective review:

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- The long-term hemodynamic quality of the repair is based on the size and compliance of the native pulmonary arteries.
- The unifocalized mapcas generally do not grow and the surgical process may in some instances compromise the growth of the PA on which the mapca is implanted.
- The reconstructed pulmonary arteries are very stiff, as suggested by the poor tolerance of pulmonary regurgitation from the conduit after the repair.

Furthermore, simultaneously with Martin Norgaard, we studied 238 initial angiograms of 61 patients with pulmonary atresia and mapcas.⁴ Mapping of all the mapcas identified were compared with the mapping of the bronchial arteries according to the totality of the literature available on bronchial artery anatomy. Distribution of the different branching patterns of mapcas arising from the aorta was similar to the distribution of bronchial arteries described in previous angiographic studies ($P = .32$ and $P = .24$). Mapcas with anatomy similar to the right intercosto-bronchial artery were found in 87% of the patients. Fifty percent of the patients had mapcas originating from the subclavian artery regions. These numbers were all similar to those previously described for bronchial arteries.⁴ All mapcas had anatomy similar to bronchial arteries. Our conclusions were that mapcas are dilated bronchial arteries and that this inferred that their growth potential, if any, was very limited, and that somehow the mapca tissue itself was unstable.

Logically, the approach designed in the late 1970s had to be radically modified. The focus of the alternative strategy was aimed at growth of the native pulmonary arteries rather than recruitment of mapcas. Several dogmas, including some coming from our own unit, had to be transgressed to set this strategy:

Large mapcas Left Untouched Lead to Rapid and Irreversible Pulmonary Vascular Disease in the Lung Segments Connected to That mapca

We believe that the natural evolution of most mapcas is toward stenosis of the mapca origin, even if only mild, and in any case, progression toward pulmonary hypertensive vascular disease is slow and exceedingly rare. Echocardiographic studies, computed tomography (CT) scan, and magnetic resonance imaging (MRI) follow-up have demonstrated this evolution in our patients.

A Significant Number of Patients With PA VSD and mapcas Have no Central PA

On the contrary, we believe that the presence of a central PA is almost the rule. Only in the presence of bilateral ducts do we see no central PA. In that case, both branches have harmonious arborization.

Very Diminutive Pulmonary Arteries do not Grow

In our experience, all central pulmonary arteries grow, even in the very small pulmonary arteries (less than 2 mm), growth can be significant and sometimes leads to the development of tiny

antegrade flow, which can be dilated to generate further PA blood flow.

Most of the Very Diminutive Pulmonary Arteries have an Incomplete PA Arborization

On the contrary, we believe that most PA branches are potentially connected to all pulmonary segments.

Our strategy aims at growing the native PA branches every time they are diminutive, regardless of how small they are and regardless of the number, shape of branching pattern, distribution, and size of the collaterals. This is done with a surgically placed central shunt. This strategy is not new and has been described and used by several teams in selected cases for more than a decade^{5,6} and was initially described by Haworth et al⁷ in 1981. Most of the previous authors, however, completed the initial operation by a series of unifocalizations of the collaterals into the PA branches once they had started to develop. We do not use unifocalization in the management plan; on the contrary, we replace the central shunt after several months with another source of central blood flow to continue pursuing the goal of growing pulmonary branches. In fact, we believe that unifocalization may hinder the growth potential of the pulmonary branches. According to our current thinking, the ideal chronology of the multi-stage approach would be the following (this is a work in progress and it may evolve in the future):

Around the first week of life. Initial exploration, now mostly performed with a contrast CT scan,⁸ but previously with selective angiography (Figs. 1, 2), to demonstrate the size of the PA branches, and the number, size, and position of the mapcas. It has at times failed to reveal the presence of a central PA that we were able to find intraoperatively.

Within the first 4 to 6 weeks. A central shunt usually designed according the Hillel Laks modification,⁹ constructed without mobilization of the central PA immediately on the central PA origin where it dilates into a slight bulb (Fig. 3). This shunt is usually constructed before the end of the first month. It is done in the first few days in cases of cyanosis. The shunt is even performed when there is a high flow situation, providing that central pulmonary arteries are diminutive. Our aim is central PA growth and we believe that the shunt will not generate significant over-circulation because the small PA branches initially restrict flow (Figs. 4, 5). The alternative to the central shunt according to Laks is the left modified BT shunt from a left innominate artery when there is a right aortic arch. The shunts can be constructed with or without bypass. We do not use the aorto-pulmonary window creation advocated by Mee^{5,10} or Hanley.⁶ The retrospective review of the patients who have received this technique in Melbourne demonstrated that stenosis or thrombosis of the right PA origin was almost the rule. Interestingly, with the modified central shunt, we have seen significant growth of the origin of the main PA in several instances, to the extent that follow-up echocardiographic studies demonstrate forward flow. One of these pulmonary valves has been dilated, representing an alternative to the second operation; as well as an easier final repair.

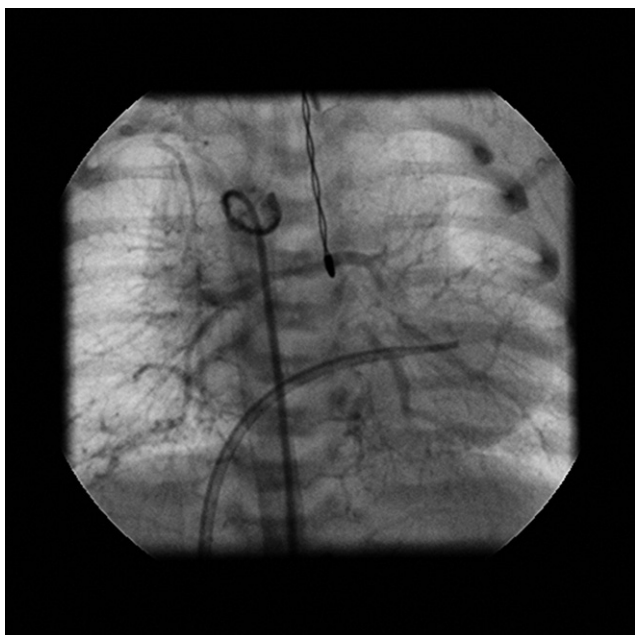


Figure 1 Neonatal imaging of pulmonary atresia VSD and mapcas. Injection in the descending aorta, late visualization of confluent very diminutive central pulmonary arteries.

A second evaluation at 3 to 4 months of age is performed either with a CT scan, an MRI study, or a catheter study. The aim is to evaluate the response of the pulmonary arteries to the increased flow and pressure centrally, and to determine whether localized stenoses or hypoplasia have to be dealt with at the time of the first reoperation (Fig. 6).

Second operation, shortly after the second evaluation, between 4 and 6 months of age. The central shunt is taken down and a



Figure 2 Neonatal imaging of pulmonary atresia VSD and mapcas. Same examination as Figure 1. Injection in the descending aorta: visualization of mapcas from the descending aorta.



Figure 3 Intraoperative photograph of a central shunt constructed according to Hillel Laks.

right ventricle (RV) to PA conduit inserted. This can be a valveless conduit of 6 to 8 mm or a banded 12 mm valved conduit (Fig. 7). The rationale is to further augment the central pulmonary pressure and flow, which continues to promote growth of the central pulmonary arteries, as the central

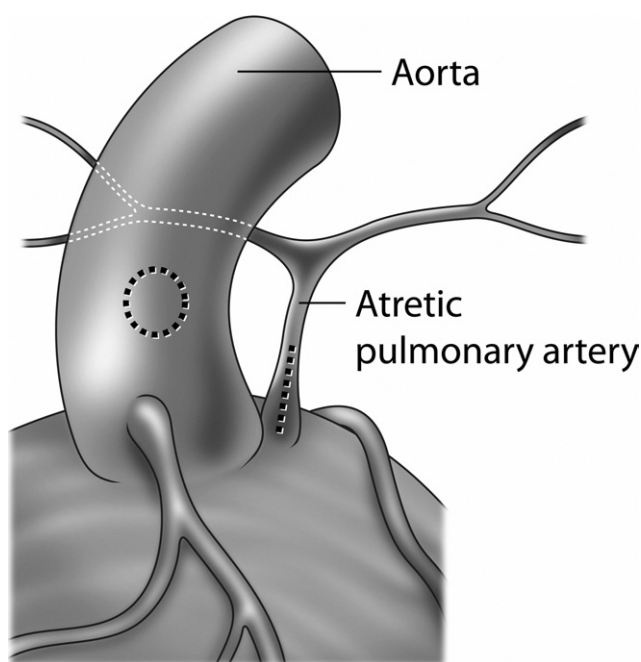


Figure 4 Schematic representation of diminutive pulmonary arteries showing the site of implantation of the central shunt.

shunt may rapidly become restrictive. Also, the RV to PA conduit provides easy access to the PA branches for angiography and balloon intervention if necessary.

The third operation is either a complete repair or a second conduit with or without PA branch surgical enlargement using autologous pericardium or ePTFE patches. Some patients at this stage have had ligation of a persisting mapca, but the rule is that most mapcas show significant involution. The repair always includes a valved conduit (Figs. 8, 9).

Technical Aspects

The central shunt is constructed along the recommendations of Hillel Laks.⁹ The PA bifurcation is carefully dissected and

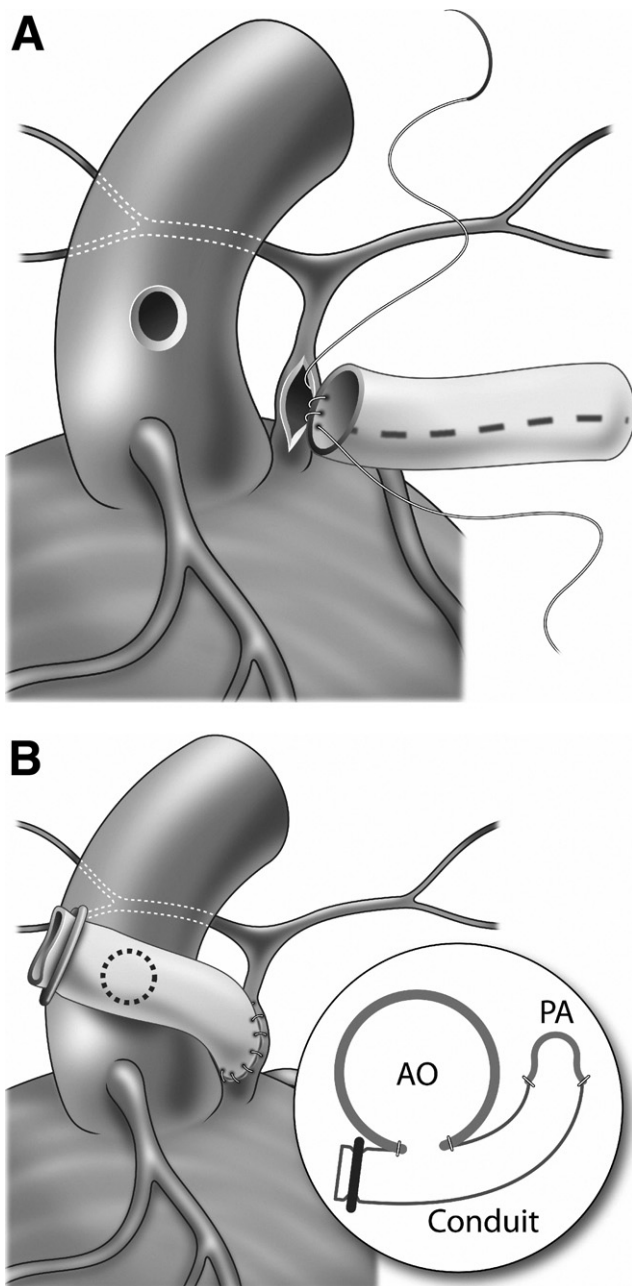


Figure 5 Construction of the central shunt.

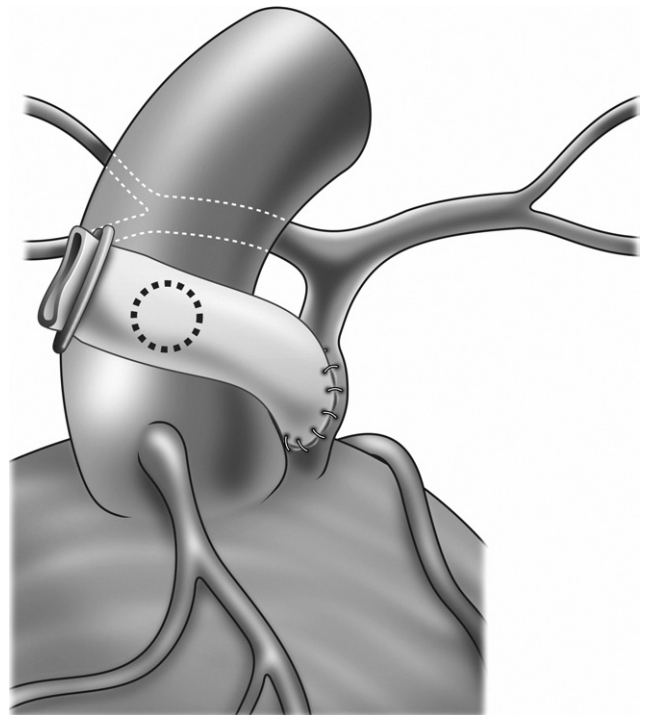


Figure 6 Within 2 to 3 months the central shunt has generated some growth of the central pulmonary arteries.

the origin of the main PA is exposed. This may require the removal of epicardial tissue adhesions and overlapping thin layers of infundibular muscle. When this is done, the bulb of the sinuses of the native main PA is exposed. It can be extremely small, but is usually significantly larger than the di-

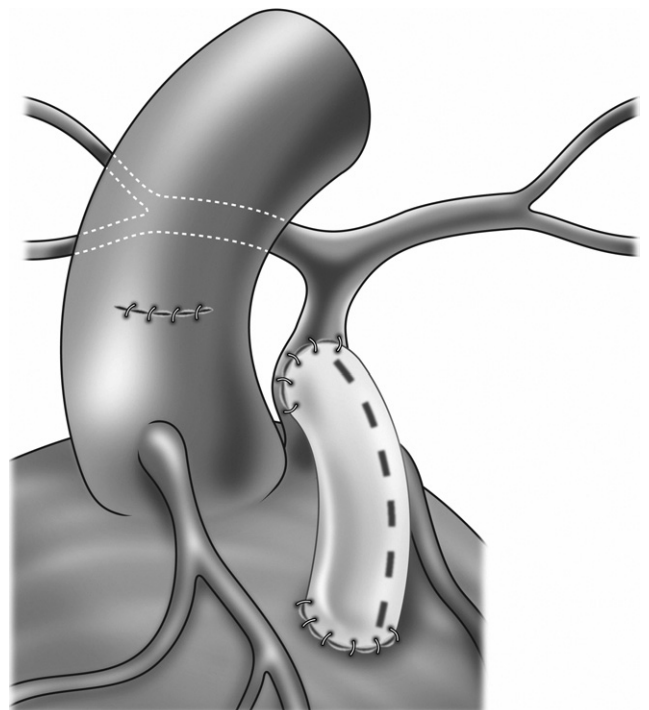


Figure 7 The central shunt is replaced by a restrictive RV to PA conduit to promote further growth.

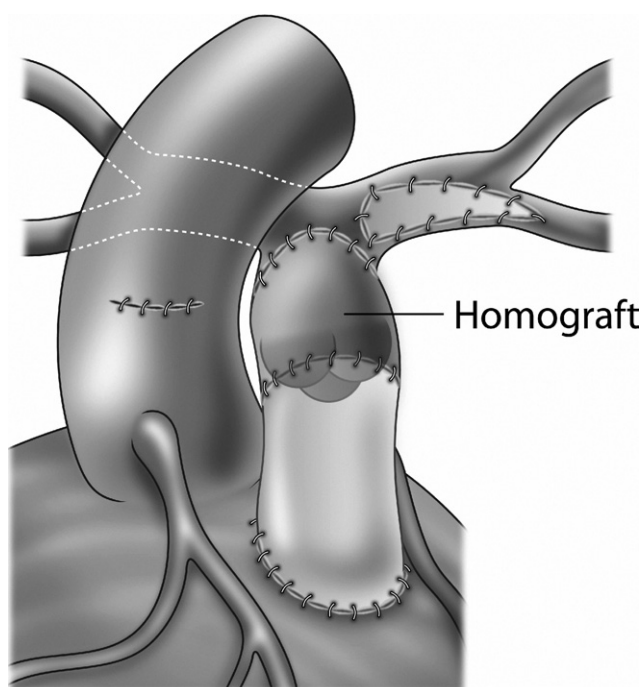


Figure 8 The repair completed.

ameter of the branches. This allows construction of the shunt without any distortion of the origin of the branches. The end-to-side anastomosis is performed at the very origin of the main PA, usually with a 3.5 or 4 mm Gore-Tex stretched conduit. This is done with full heparinization if bypass is required or 100 UI/Kg otherwise. The implantation is at 90° with the main PA in all directions. Then a side to side anastomosis is performed after a punch hole is created on the facing aspect of the ascending aorta and the lateral aspect of the conduit. The conduit has to be stretched gently to avoid any kinking. The aorta is either totally cross clamped with cardioplegia or laterally excluded. A photograph of the coronary anatomy is taken to facilitate further procedures and the chest is closed over an ePTFE pericardial substitute.

The RV to PA conduit is constructed with cardiopulmonary bypass and aortic cross clamp. The size of the conduit is 6 or 8 mm ePTFE, or a 12 mm valved conduit reduced with banding. The 8 mm conduit can also be reduced with hemoclips applied laterally. The distal anastomosis is constructed at the pulmonary site of the central shunt. The proximal anastomosis is performed as high as possible on the infundibulum and away from the LAD. At the end of the procedure, during systole, the pressure can be near systemic immediately distal to the conduit. The conduit is not reduced if the mean pressure in the hilum is low. We avoid performing branch plasty at this stage unless very significant asymmetry between branches is demonstrated. Localized narrowing of the branch pulmonary arteries can be dealt with dilation 3 to 4 weeks before the next procedure. Harmonious hypoplasia is dealt with patch augmentation at the time of the second procedure. Autologous pericardium treated with glutaraldehyde is used or 0.4 mm ePTFE.

The last stage is the complete repair and is performed when the pulmonary arteries are a suitable size at the hilum. Patch augmentation of the intra-pericardial branch pulmonary arteries may be required at this stage. Repairs have been done between 15 and 40 months of age. Valved conduits are always used.

Patients

From 1999 to 2003, 15 patients were treated with a central shunt and various combinations of the unifocalization process; including two patients in whom the unifocalization process was abandoned due to either very small size of the pulmonary arteries initially or the spectacular response to the increased pulmonary blood flow. These 15 patients are not included in this review. Since 2003, 18 patients with diminutive pulmonary arteries have been treated with the protocol described above. One other patient with discontinuous pulmonary arteries, two ducts, and one mapca, was repaired in one stage.

Median age at the time of the first procedure was 3.5 weeks (range, 0.8 to 17 weeks); median peripheral saturation was 85% (range, 69% to 95%). Fourteen patients received a central shunt via the Laks technique (3 mm, n = 2; 3.5 mm, n = 11; and 4 mm, n = 3), three patients received a modified BT shunt or a combination of central shunt and modified BT shunt for discontinuous pulmonary arteries; one patient received a restrictive 4 mm RV to PA conduit. Fourteen patients had a second procedure after a mean interval of 6.5 months (range, 3.5 to 19.7 months) from the initial procedure. With regard to age, the deviation from the protocol detailed above was due to:

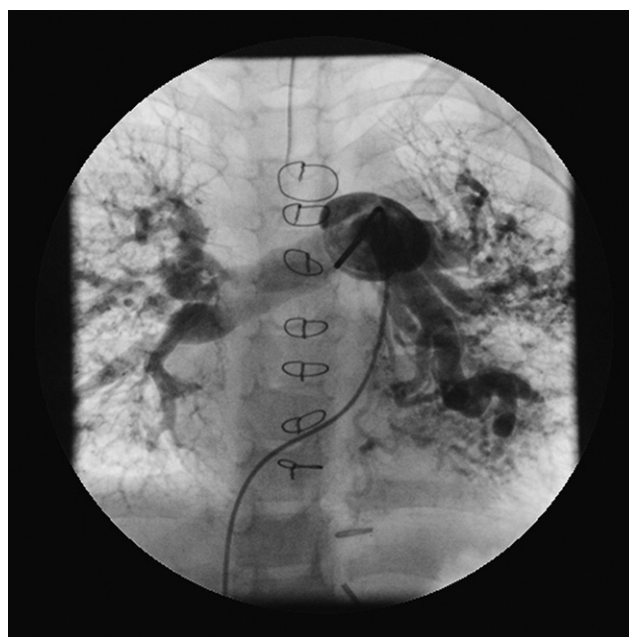


Figure 9 Same patient as shown in Figures 1 and 2. Injection in the PA bifurcation. Angiographic control 2 months after the repair.

- Adequate growth of the central pulmonary arteries, patient ready for repair
- Inadequate but harmonious growth at the time of exploration with a central shunt well patent
- Logistic delay
- Patient age outside the neonatal protocol

One patient died of sepsis 3 months after central shunt placement. The patient had DiGeorge syndrome and was 17 months old at the time of referral.

The second operations were either: complete repair with VSD closure and conduit insertion (n = 2, age 11 and 19 months; with mapca ligation, n = 2), central shunt take down and RV to PA conduit (n = 10; associated with PA surgical repair n = 6) or left BT or right BT shunt for discontinuous PAs (n = 2; age 4 and 4.5 months, with mapca ligation n = 1).

The third operation was either a complete repair (n = 3, age 40, 36, and 20 months) or a conduit upgrade n = 2 and mapca ligation n = 1.

The fourth operation was complete repair (n = 2, age 48 and 20 months).

Seven patients have had a complete repair, eight patients are awaiting repair, while one patient is unlikely to ever be suitable for repair and has the VSD left intact.

Five of the seven repaired patients have had a catheter study at a median of 13 months (range, 2 to 19 months) after the repair, with median RV pressure 59% (range, 37 to 64%) of systemic. The two other patients had an RV pressure estimate on their latest echo using the tricuspid regurgitation envelope of one half and two thirds systemic pressure.

So far, four mapcas have been ligated in 17 patients, none have been unifocalized.

Conclusions

The recent modification of our protocols for the approach of PA, VSD, and mapcas in Melbourne is still a work in progress. So far, however, we have been able to enroll all patients presenting to our unit and the aim of achieving growth of the pulmonary arteries without the use of unifocalization is being achieved. The patients who have reached repair demonstrate sub-systemic RV pressure and very few mapcas have required

ligation at the stage of repair or earlier, somehow demonstrating that the fate of most mapcas in a competitive flow environment is essentially toward reduction and possibly disappearance of any significant flow. Seven patients out of eight who have reached the end of the preparation program have had a repair within the age window that had been set. The other patients are still in the preparative stage.

Only longer follow-up and larger numbers of patients will confirm the adequacy of our approach. We shall be looking at the evolution of the RV and PA pressures, and the tolerance of pulmonary regurgitation. For patients who have not been able to be repaired, the exercise ability and oxygen saturation at rest will be the main criteria of follow-up.

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