SURGICAL STRATEGIES FOR
PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT AND
MAJOR AORTOPULMONARY COLLATERAL ARTERIES
AND
PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM
AT THE ROYAL CHILDREN’S HOSPITAL, MELBOURNE, AUSTRALIA.

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ABSTRACT

Congenital heart disease affects approximately six to eight babies in every thousand live births. In Australia over two thousand babies are born with congenital heart disease each year, with about half requiring either surgery or catheter intervention. The other half has minor abnormalities that have little to no functional impact and rarely require intervention.

Historically most patients with complex congenital heart disease died in childhood. However, the past three to four decades has seen considerable advances in the fields of cardiac surgery, cardiology, cardiac imaging and intensive care, to the extent where most congenital cardiac lesions are now considered repairable with minimal morbidity.

There remain, however, specific cardiac lesions that have yet to achieve this level of safe correction. Pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals, and pulmonary atresia with intact ventricular septum are two such lesions.

Pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals is a rare and complex lesion. There is considerable variability in the anatomy, morphology and geometry of the native pulmonary vessels and the collateral arteries. The ultimate goal of therapy is a biventricular repair with closure of intracardiac defects and establishment of right ventricle to pulmonary artery blood flow. Throughout the world, surgeons have tried many different techniques to achieve this goal. The Cardiac Surgery Unit at the Royal Children’s Hospital in Melbourne achieved excellent early success in the treatment of this malformation in the 1980s – 1990s. However, long-term follow-up has revealed limitations in the surgical strategy applied during that era. Whilst complete repair was achievable in 67% of patients, survival to thirty years of age was only 58% and after complete repair only 51% of patients were alive twelve years later.

A new approach – ”repair without unifocalisation” was devised and is explained in this thesis. The first twenty patients who then underwent this surgical technique are examined. Early survival is excellent with no deaths to date. Complete repair was achieved in 60% of patients, 30% are
awaiting complete repair, and 10% of patients are deemed unlikely to achieve complete repair and my need to be assessed for heart/lung transplantation when their symptoms progress.

Pulmonary Atresia with intact ventricular septum is another lesion in which it has been difficult to achieve low morbidity and mortality. In this malformation there is complete obstruction between the right ventricle and the pulmonary arteries, and newborn babies are typically dependent on ductal blood flow for pulmonary perfusion. With a lack of a ventricular septal defect there is no outflow from the right ventricle and this results in variable degrees of right ventricular and tricuspid valve hypoplasia, and sometimes extensive right ventricular hypertrophy with coronary artery anomalies.

Surgical strategies aimed at achieving a biventricular repair with reconnection of the right ventricle to the pulmonary artery have always been the ideal, as this most closely reflects the natural physiological state. However, poor results emphasized by a 5-year survival of only 49%, have encouraged surgeons to explore different strategies in the treatment of this malformation. In this modern era it is quite clear that a single surgical strategy to this malformation is inappropriate. If a univentricular palliation pathway is used exclusively then many patients fail to attain a biventricular repair and the long-term benefit this provides. If a biventricular strategy is applied exclusively then early mortality increases to unacceptable levels.

An approach utilising both biventricular and univentricular repairs appears to provide the best balance between decreasing early mortality and providing the best long-term prognosis. Unfortunately there are limited criteria to decide which patients should be directed towards a univentricular or biventricular repair.

Using data from patients operated on over a period of sixteen years at the Royal Children’s Hospital I have tried to identify preoperative variables that may guide this decision making process. Multivariate analysis revealed two significant risk factors for mortality. These were the presence of right ventricle to coronary artery connections and a tricuspid valve size Z score of less than negative two. Surgical results revealed an 80% ten-year survival rate. Using a simple three tiered right ventricle size categorization one can stratify those with the smallest right ventricles to univentricular palliation and those with near normal right ventricles to biventricular repair. Those
patients with moderate sized right ventricles can be directed towards biventricular repair except if they also possess the significant risk factors for mortality. This approach is hoped to improve survival.
**PREFACE:**

Figures 11, 13, 14, 15, and 16 were prepared with the help of the medical illustration department at the Royal Children’s Hospital, Melbourne.

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**DECLARATION:**

This is to certify that:

i. The thesis comprises only my original work towards the masters except where indicated in the Preface,

ii. Due acknowledgement has been made in the text to all other material used,

iii. The thesis is 17,000 words as approved by the Research Higher Degrees Committee.

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ORAL PRESENTATIONS ARISING FROM THIS RESEARCH

“Finding the right balance between univentricular and biventricular repair for pulmonary atresia with intact ventricular septum”

“Pulmonary atresia, ventricular septal defect, and major aortopulmonary collaterals: results of a neonatal shunting regime”
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PUBLICATIONS ARISING FROM THIS RESEARCH

1. Pulmonary atresia, ventricular septal defect, and major aortopulmonary collaterals: neonatal pulmonary artery rehabilitation without unifocalization.
   Liava'a M, Brizard CP, Konstantinov IE, Robertson T, Cheung MM, Weintraub R, d'Udekem Y.
   PMID: 22119120 [PubMed - indexed for MEDLINE]

   Liava'a M, Brooks P, Konstantinov I, Brizard C, d'Udekem Y.
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3. Pulmonary atresia, VSD and Mapcas: repair without unifocalization.
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CONTENTS:

Abstract 3
Preface 6
Declaration 6
Acknowledgements 6
Presentations 7

1. PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT AND MAJOR AORTOPULMONARY COLLATERAL ARTERIES. 12

• 1.1 Natural History 15
• 1.2 Surgical Management 16
• 1.3 Staged Unifocalisation 19
• 1.4 One stage Complete repair and unifocalisation 22
• 1.5 Long term results from the Melbourne experience of staged unifocalisation 26
• 1.6 Repair without Unifocalisation - a new paradigm in the management of Pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries. 28
• 1.7 Early angiographic results from a protocol of repair without unifocalisation 39
  1.7.1 Methods 39
  1.7.2 Surgical Strategy 40
  1.7.3 Statistical Analysis 40
  1.7.4 Results 41
  1.7.5 Clinical Outcomes 41
  1.7.6 Hospital Morbidity 42
  1.7.7 Follow-up 43
  1.7.8 Pulmonary Artery Growth 44
  1.7.9 Angiography after complete repair 45
  1.7.10 Complications 46
• 1.8 Conclusion 47
2. PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM

- 2.1 Natural History 52
- 2.2 Evolving surgical management at RCH 56
  - 2.2.1 Methods 57
  - 2.2.2 Statistical analysis 59
  - 2.2.3 Results 59
  - 2.2.4 Death prior to intervention 60
  - 2.2.5 Diagnostic and Interventional catheterization 60
  - 2.2.6 Requirement for systemic to pulmonary shunt 63
  - 2.2.7 Right Ventricular overhaul 64
  - 2.2.8 Outcome 64
  - 2.2.9 Risk factors for death 66
- 2.3 Conclusions 67

Bibliography 71
Publications 77
LIST OF TABLES:

Table 1: Patient demographics 39
Table 2: Postoperative length of stay and ventilator time 42
Table 3: Catheter Haemodynamics after complete repair 45
Table 4: Complications 46
Table 5: Patient Characteristics for primary catheter intervention 61
Table 6: Requirements for systemic to pulmonary artery shunt 64
Table 7: Outcomes based on right ventricle size 64
Table 8: Risk Factors for death 66
LIST OF FIGURES:

Figure 1a: Schematic view of pulmonary atresia, VSD, MAPCAs 12
Figure 1b: Angiographic view of pulmonary atresia, VSD, MAPCAs 13
Figure 2: Overall survival estimate 15
Figure 3: MAPCA Histology 17
Figure 4: Clinical outcomes of staged unifocalisation 19
Figure 5: Melbourne shunt 20
Figure 6: Anterior approach to MAPCAs 23
Figure 7: Unifocalisation suture techniques 23
Figure 8: Morphology based algorithm for surgical management 25
Figure 9: Angiographic imaging of diminutive pulmonary arteries 32
Figure 10: Angiographic imaging of MAPCAs 32
Figure 11: Schematic representation of diminutive pulmonary arteries showing the site of implantation of the central shunt. 33
Figure 12: Intraoperative photograph of a central shunt 33
Figure 13: Construction of the central shunt. 34
Figure 14: Pulmonary growth from central shunt 35
Figure 15: RV to PA conduit 35
Figure 16: The complete repair 36
Figure 17: Angiographic view of complete repair 37
Figure 18: Surgical outcomes of pulmonary atresia, VSD, MAPCAs 43
Figure 19: Growth of pulmonary arteries 44
Figure 20: Timeline of pulmonary artery growth 45
Figure 21: Schematic representation of PAIVS 52
Figure 22: RV coronary artery connections 53
Figure 23: Three size categories of PAIVS 58
Figure 24: Outcomes after initial catheter intervention 63
Figure 25: Kaplan Meier survival curve 66
1. PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT AND MAJOR AORTOPULMONARY COLLATERAL ARTERIES.

Pulmonary Atresia with ventricular septal defect and major aortopulmonary collaterals is a complex and heterogeneous cardiac malformation. This malformation has been previously classified as truncus arteriosus type IV, absent sixth aortic arch, pseudotruncus and Tetralogy of Fallot with pulmonary atresia.1,2

Essentially, there are three parts to this malformation:
- A lack of luminal continuity and blood flow from the right ventricle to the pulmonary artery.
- A hole in the interventricular septum.
- An abnormal pulmonary circulation.

This abnormal pulmonary circulation provides the extreme variability of the malformation, as variation can occur in:
- Origin of blood flow.
- Presence or absence of normal pulmonary arteries.
- Presence or absence of major aortopulmonary collaterals (MAPCA).
- Distal intrapulmonary arborisation abnormalities.

Figure 1a: Schematic representation of the malformation in the surgeons view. The PA is significantly smaller than the aorta with confluent hypoplastic pulmonary
arteries and major aortopulmonary collateral arteries are visible posterior to the great arteries.\(^3\) (LPA= left pulmonary artery, RPA= right pulmonary artery, SVC= superior vena cava, APC= aortopulmonary collateral artery)

Figure 1b Angiographic view demonstrates normal sized ventricles, a large ventricular septal defect, pulmonary atresia with no visible main pulmonary artery and aortopulmonary collaterals coming off the descending aorta.\(^4\)

The general morphology of this malformation is similar to Tetralogy of Fallot with pulmonary stenosis. The ventricular septal defect is juxta-aortic and usually lies adjacent to or involves the membranous septum. It differs from the usual isolated ventricular septal defect by being associated with malalignment of the conal (infundibular) septum. It is also virtually always of a large size.

Genetic studies have documented that pulmonary atresia with ventricular septal defect belongs to a spectrum of conotruncal cardiac malformations often associated with monosomy 22q11.\(^50\) The clinical presentation of monosomy 22q11 includes patients with conotruncal anomaly face syndrome, velo-cardio-facial syndrome, and DiGeorge syndrome.\(^51, 52\) More recently, these syndromes have been incorporated as a group under the acronym CATCH 22 (cardiac defect, abnormal face, thymic hypoplasia,
cleft palate, hypocalcemia, and microdeletion 22q11). 53 Two groups have recently shown an association between patients with PA, VSD, and MAPCAs and monosomy 22q11. 54, 55 In both studies, anywhere from 40% to 48% of patients with PA VSD, and major aorta pulmonary collaterals were shown to have a micro deletion in 22q11. The Royal Children’s series has a rate of 45%.

Pulmonary atresia maybe at the infundibular or annular level, and intrapericardial pulmonary arteries may vary from normal in size to completely absent. Usually they are hypoplastic and frequently fail to arborise to all pulmonary vascular segments. There are ten pulmonary segments in the right lung and nine segments in the left lung as the left ligula lobe is usually counted as one segment rather than two as the middle lobe of the right lung is. Pulmonary arterial segments that are not connected to central pulmonary arteries usually receive blood from large aorto-pulmonary collateral arteries. Pulmonary artery stenoses are not common, but when present usually occur juxta-ductally at the origin of the branch pulmonary arteries. The alternative sources of pulmonary blood flow usually come from major aortopulmonary collateral arteries. These arteries often originate from the descending aorta, and follow a tortuous course to connect to inter or intra lobar pulmonary arteries that may then arborise normally within that pulmonary lobe or segment. However, these collateral arteries can also originate from the subclavian arteries, bronchial arteries, intercostal arteries and even the coronary arteries. In fact as all these origins have been described as typical variants of bronchial artery origin it has been suggested by some groups that major aortopulmonary collateral arteries are in actual fact unusually enlarged and developed bronchial arteries. 5 Stenoses are commonly found in major aortopulmonary collateral arteries, frequently secondary to areas of intimal proliferation, and are usually found at branching points and the junction between collateral and pulmonary artery. 6 These stenoses may be present at birth or develop post-natally where they are associated with abnormal flow patterns.
1.1 Natural History

The Natural History of this malformation is complicated by its variability in presentation. One of the few papers to study this aspect was performed by the cardiac unit of Great Ormond Street Hospital in 1995. From 218 patients with this malformation they were able to suggest that 65% of patients with this malformation present in infancy, and that three-quarters of these patients will be severely compromised by critical cyanosis or heart failure. The remainder present later in life, including about ten percent who do not present until after the age of ten years. These patients have a pulmonary blood flow significant enough in infancy to avoid conspicuous cyanosis yet not great enough to cause pulmonary vascular hypertensive disease and heart failure.

The estimated survival of this population is limited. Survival to 1 year of age is only 60%, of those alive at one year their probability of surviving to ten years old is only 65%, this means over 50% of patients are dead before the age of ten. (Fig 2.)

![Fig 2. Estimate of overall survival from birth with 95% confidence intervals.](image)
1.2 Surgical Management

The complexity of this lesion is such that until the mid-1970s most patients were managed medically or with surgical palliation without hope of complete repair. The majority of patients were given only a 20% chance of survival to the age of 30 years. Improvements in selective catheterisation techniques allowed detailed characterization of major aortopulmonary collateral arteries, which gave surgeons more confidence in managing the major aortopulmonary collateral arteries, and developing a pulmonary vascular bed capable of supporting a complete repair.

The principles of surgical management remain constant despite the variable anatomical morphology. The ultimate goal is to close the ventricular septal defect and establish continuity between the right ventricle and the pulmonary artery. However, a successful definitive repair requires the presence of an adequate pulmonary vascular bed to avoid high pulmonary vascular resistance and resulting right ventricular failure.

One of the early surgical reports by McGoon in 1975 described the Mayo clinics experience with pulmonary atresia, ventricular septal defect and major aortopulmonary collaterals. Their technique was to ligate major aortopulmonary collateral arteries usually by way of sternotomy but often by concomitant thoracotomy on the side of the descending aorta. Major aortopulmonary collateral arteries were ligated to prevent excessive left heart return during cardiopulmonary bypass, with the associated myocardial distension and damage. The patient was then put onto cardiopulmonary bypass and the intracardiac repair completed. Their patient group ranged in age from four years to twenty-seven years of age. All of these patients were “highly selected survivors” having made it through the neonatal period and infancy. Interestingly they noted one patient previously having been operated at a different centre with an aortic to right pulmonary artery shunt, with good growth of the pulmonary artery. They then used this strategy on three further cases hoping to achieve pulmonary artery growth prior to staged intracardiac repair and aortopulmonary collateral artery ligation.

Early necropsy studies into the nature of systemic to pulmonary collateral arteries by Haworth and McCartney confirmed the anatomical variety of major aortopulmonary collateral arteries and reinforced their abnormal
nature in thickness of media, external diameter, and intimal proliferation causing flow limiting stenoses. Whilst somewhat protective against the development of pulmonary vascular obstructive disease, more often these stenoses were thought to be the cause of the development of cyanosis during infancy, as opposed to from just ductal closure alone.\(^8\)

Figure 3: Transverse section of the mid portion of a major aorto-pulmonary collateral at a region of segmental narrowing. The lumen is almost totally obliterated by intimal proliferation and cushion formation, and a layer of amorphous material containing no elastic tissue. (IL= internal elastic lamina, EL external elastic lamina, M= media, IC= intimal cushion, PA= pulmonary artery)\(^8\)

Given the unpredictability of these major aortopulmonary collateral arteries the authors advocated a treatment regime aimed at native pulmonary artery growth as the primary aim. Reconnection of large aortopulmonary collateral arteries to the central pulmonary arteries to avoid loss of pulmonary parenchyma could be performed if possible, however, if not possible then ligation was preferable as to cause ipsilateral hypoplasia and hopefully contralateral compensatory pulmonary enlargement.\(^5\)
This led to the report from Great Ormond Street Hospital on the effect of a systemic to pulmonary shunt in pulmonary atresia, ventricular septal defect patients.\(^9\) Here either modified Blalock-Taussig or central shunts were
placed to encourage pulmonary artery growth and development rather than to improve systemic arterial saturations. Only one patient received a shunt in the neonatal period (at one month old) the other eight patients were between eight months and fifteen years old, however, five of the nine patients did show significant increase in pulmonary artery size. Despite this pulmonary artery growth, the Great Ormond Street group believed that the pulmonary artery arborisation abnormalities including stenoses in pulmonary artery branches, and lack of supply to all lung segments, would preclude intracardiac repair due to excessively high pulmonary artery pressures. They went further to postulate that the problem was the multifocal and abnormal pulmonary blood source.

Two techniques were described to turn a multifocal pulmonary blood supply into a unified source:

- Ligation of isolated major aortopulmonary collateral arteries, which should encourage compensatory growth in lung segments supplied by normally connected pulmonary arteries, as the underperfused segments become relatively smaller.
- Disconnecting major aortopulmonary collateral arteries from the descending aorta and anastomosing them onto the native pulmonary arteries – unifocalising the pulmonary blood source.

This second approach provided the basis for the Great Ormond Street hospital approach to pulmonary atresia, ventricular septal defect, with major aortopulmonary collaterals repair from 1979 to 1986.10
1.3 Staged Unifocalisation

The Thoracic unit at Great Ormond Street hospital undertook a programme of reconnecting major aortopulmonary collateral arteries to central pulmonary arteries, with or without modified Blalock-Taussig shunts in an effort to unifocalise pulmonary blood supply. All operations were from a lateral thoracotomy approach and prosthetic interposition grafts were often used when direct aortopulmonary collateral artery to central pulmonary artery anastomoses were unachievable. The results were disappointing. Of twenty-six patients undergoing “peripheral” unifocalisation of major aortopulmonary collateral arteries only three (12%) achieved complete repair, four (15%) were considered repairable but as currently relatively asymptomatic have not been repaired. In addition only one-third of major aortopulmonary collateral artery anastomoses on postoperative angiography were judged to be technically successful.

![Fig 4. Clinical outcome of unifocalised patients.](image)

The outcome of this study was a questioning of the benefit of unifocalisation procedures and a move towards a “central” approach, with pulmonary artery shunting as the initial operative intervention. These results, however, did not stop other cardiac units around the world from utilising the unifocalisation technique in an attempt to build a pulmonary bed large enough to allow complete repair.
In 1991 Iyer and Mee from Melbourne presented their ten-year experience of a staged unifocalisation programme for Pulmonary Atresia, Ventricular septal defect and major aortopulmonary collaterals. Their paradigm involved both “central” and “peripheral” interventions. With fifty-eight patients they described a technique of initial shunting by modified Blalock-Taussig shunt, central shunt or direct pulmonary artery to aortic anastomosis, in an effort to encourage pulmonary artery growth.

Figure 5. “Melbourne shunt” The direct aorta to pulmonary artery shunt used extensively at the Royal Children’s Hospital to encourage pulmonary growth.

Following the initial shunting procedure, sixty-six major aortopulmonary collateral arteries in thirty-two patients were ligated if they clearly duplicated supply from the native pulmonary arteries or were small and only supplied one or less bronchopulmonary segment. Sixty-eight major aortopulmonary collateral arteries were transplanted in thirty-four patients. Major aortopulmonary collateral arteries were considered more suitable for transplantation if they supplied more than one bronchopulmonary segment independently of the native pulmonary arteries, had no peripheral stenoses, and were not hypertensive. Major aortopulmonary collateral arteries were directly anastomosed to central pulmonary arteries in an oblique fashion to
enlarge the anastomotic lumen. Eight patients had gortex interposition grafts and four patients azygous vein interposition grafts.

Complete repair was only undertaken when:

- The calculated predicted RV to LV pressure was less then 0.7 using the Birmingham formula ($p_{RV}: p_{LV} = 0.484(D_{RPA}/D_{Ao} + D_{LPA}/D_{Ao})$),
- No significant major aortopulmonary collateral arteries remained,
- More than two-thirds of lung segments were connected to the native pulmonary arteries, and
- There was a net left to right shunt.

The overall results of this staged repair programme were: Thirty of fifty-eight (52%) patients eventually underwent complete repair after one hundred twenty-one staging procedures. Twelve patients (21%) failed to achieve the minimum requirements for repair after staging procedures, ten patients (17%) are currently awaiting complete repair, and overall mortality was six patients (10%).

These results were a vast improvement on both the natural history of the lesion and the surgical results obtained in previous reports.\(^1\)\(^,\)\(^9\) With similar excellent surgical results by other units worldwide, there was significant data to show a clear benefit from a staged unifocalisation programme over the natural history of the disease.\(^13\)\(^,\)\(^14\)
1.4 One Stage Complete Repair and Unifocalisation

In 1995 the Stanford Cardiac Surgery unit described a novel change in the treatment of patients with pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries.\textsuperscript{15} In ten patients they had veered away from the prevailing staged protocol propagated by Iyer and Mee and attempted a complete unifocalisation and repair in one setting through a median sternotomy. The proposed arguments for attempting this approach were multifocal:

- Early normalisation of cardiovascular physiology and correction of cyanosis,
- Reducing the amount of surgical operations and their associated morbidity,
- Protection against pulmonary vascular obstructive disease from high flow major aortopulmonary collateral arteries and systemic to pulmonary shunts.
- Extensive tissue-to-tissue major aortopulmonary collateral artery to pulmonary artery anastomoses as opposed to the use of synthetic material would eliminate previous poor technical results.

In ten patients (five were less then nine months old) through an extended mid-line sternotomy both pleura are opened and major aortopulmonary collateral arteries dissected from the descending aorta. Once saturations become precarious the patient is put onto cardiopulmonary bypass and completion of the unifocalisation anastomoses are performed along with the intracardiac repair.
Fig 6. Diagram of the transverse sinus approach for the dissection, rerouting, and unifocalisation of the major aortopulmonary collaterals.\textsuperscript{15}

Fig 7. Diagrams of the anatomy of the major aortopulmonary collaterals and true pulmonary arteries and the suture techniques of unifocalisation in a 3.5kg infant.\textsuperscript{15}
In nine out of ten patients the authors were able to achieve a complete repair (one patient was left with her ventricular septal defect open). Median ICU stay was 2.2 days and median hospital stay 14.5 days, while only one death in the series gave a mortality rate of 10%. Important haemodynamic data was obtained on postoperative day 1-2, before removal of the pulmonary artery catheter. Peak right ventricular to left ventricular (RV: LV) ratio at this stage ranged from 0.31 to 0.58 with a median of 0.47. Only one anastomosis of forty unifocalisations needed surgical repair in the median eight-month follow-up. This paper provided a clear alternative technique to the repair of pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries from the staged approach, which with the excellent results from Melbourne had become the standard management technique.

The same unit from Stanford then published a more comprehensive review of this technique in April 2000 with eighty-five patients. Of the eighty-five patients, sixty-seven percent were less than one year old and only fourteen patients had had prior surgery. With this larger group of patients a clearer picture has emerged of the one-stage technique. Not all patients can achieve a complete unifocalisation and repair in one setting. In fifty-six patients (66%) they were able to achieve a complete repair and unifocalisation in one operation. In twenty-three patients (27%) a midline unifocalisation operation and right ventricular outflow reconstruction was performed but the ventricular septal defect not closed, and six patients (7%) underwent traditional staged unifocalisation via thoracotomy. Actuarial survival for the whole group was 84% at one year and 74% at four years. In the group of patients (n=56) where one-stage complete unifocalisation and repair was possible, there were five early and seven late deaths (n=12, mortality of 21%). Despite aiming for a complete repair in one stage, twenty-four patients underwent thirty-six reinterventions on their neo-pulmonary arteries, giving an actuarial re-intervention-free survival of 42% at five years.

By 2009 the Stanford group was able to publish a concise picture of their approach to pulmonary atresia, ventricular septal defect and major aortopulmonary collaterals and the early one-stage unifocalisation and repair technique in more than four hundred patients over fifteen years. The approach is based on the anatomy of the native pulmonary arteries and
major aortopulmonary collateral arteries. If the pulmonary arteries are hypoplastic yet confluent without arborisation defects then they abandon the one-stage approach and use a staged approach involving a central shunt followed by staged intracardiac repair. However, in their series this group comprised only 12% of patients. More commonly, (88%) patients had abnormal branching or absent pulmonary arteries and in most of these patients a midline onestage unifocalisation and complete repair was attempted. An intra-operative flow study is used to determine if the predicted post repair systolic RV/LV pressure ratio is below 0.5 and thus the suitability for ventricular septal defect closure.\textsuperscript{17}

Overall only 56% of patients received a complete repair at one operation. However, 90% achieved complete repair by five years of age. While mortality early in their series was 11% this was reduced to 2.3% for the last nine years.

Figure 8. Morphology based algorithm for surgical management.\textsuperscript{17}
1.5 Long term results from the Melbourne experience of staged unifocalisation

Since the initial reports of Midline one-stage complete repair and Staged unifocalisation many cardiac surgery units have vacillated between the two approaches. It has also encouraged a review of our own results in Melbourne with the staged technique. In 2005, Dr. d’Udekem reviewed eighty-two consecutive patients from 1975 to 1995, all of whom were entered into a multistage unifocalisation regime for pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries. Patients underwent multiple operations via sternotomy and thoracotomy (n=189), to perform one hundred nineteen shunts, one hundred thirty major aortopulmonary collateral artery transplantations, and seventy major aortopulmonary collateral artery ligations. Clinical follow-up and serial angiographies of patients were reviewed. Complete repair was achieved in fifty-three (65%) patients at a median age of four years. Hospital mortality during preparatory procedures was 4% (seven patients) and, during complete repair was 8% (four patients). Twenty-two patients were considered unrepairable and seven of these patients died at a median of eight years after their last procedure. The remaining patients were still alive at a median of thirteen years.

A total of two hundred sixty-eight angiograms were reviewed. The results of a central shunt could be evaluated in twenty-nine patients. All patients who received a central shunt showed central pulmonary artery growth. Most patients had a direct aortopulmonary anastomosis (Melbourne shunt), however 46% of patients had a proximal pulmonary artery stenosis greater than 50%, no patient who had a goretex central shunt developed proximal pulmonary artery stenosis. Pulmonary artery diameter grew from 3 mm to 6 mm in a two-year period. Only sixty unifocalised major aortopulmonary collateral arteries could be identified on serial angiograms in thirty-one patients. After a mean of 3.2 +/- 4 years twenty-six had thrombosed and twelve had a stenosis greater than 50%. Serial measurements of major aortopulmonary collateral arteries could be obtained in twenty-nine patients. Over a mean interval of 3.5 years major aortopulmonary collateral artery size had only increased from 3.6mm to 4.0mm (P=0.25).
This retrospective analysis suggested that although the increased vascular bed recruited by unifocalisation was sufficient to allow a safe repair shortly after the completion of unifocalisation, 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 twelve years after the initial procedure was 32% ± 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 unifocalised major aortopulmonary collateral arteries but also inferred that growth of the pulmonary artery branches themselves could have been compromised by the unifocalisation process.
1.6 Repair without unifocalisation - a new paradigm in the management of pulmonary atresia, ventricular septal defect and major aortopulmonary collaterals.

It is clear that the technical difficulty of surgical management in this malformation 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 towards early and complete unifocalisation of major aortopulmonary collaterals, we have developed an alternative technique called - 'repair without unifocalisation.'

The Cardiac surgery unit at Melbourne’s Royal Children’s Hospital has been at the forefront of the surgical treatment of pulmonary atresia, ventricular septal defect and major aortopulmonary collaterals from the late seventies to the mid-nineties. Dr. Mee introduced and promoted the staged recruitment of lung segments, which were isolated from pulmonary arteries and solely supplied by major aortopulmonary collateral arteries. The preparatory procedures usually consisted of some form of systemic to pulmonary shunt followed by unifocalisation of major aortopulmonary collateral arteries to native pulmonary artery branches, performed via multiple sternotomies and thoracotomies. If no central pulmonary artery branches were found or were not suitable to receive unifocalisation, they were reconstructed with autologous pericardial rolls. Once all or most of the pulmonary segments were connected to the pulmonary artery tree, patients were offered a complete repair with trans-ventricular closure of the ventricular septal defect and implantation of a valved conduit between the right ventricle and the pulmonary artery.

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 lead the unit to revisit this strategy. For a while, several different strategies were used simultaneously; including one stage repair via sternotomy in infancy, as well as central shunting to promote growth of the diminutive central pulmonary arteries, and our own traditional approach described above.
The major lessons learnt from our retrospective review were: 18

- Long-term hemodynamic quality of the repair as determined by pulmonary artery pressures is based on the size and compliance of the native pulmonary arteries.
- Unifocalised major aortopulmonary collateral arteries generally do not grow and the surgical process may in some instances compromise the growth of the pulmonary artery on which the aortopulmonary collateral artery is implanted.
- Reconstructed pulmonary arteries can be very stiff as suggested by the poor tolerance of pulmonary regurgitation from the conduit after the repair.

Furthermore, a study from the Royal Children’s Hospital Cardiac Surgery unit undertook angiographic mapping of all major aortopulmonary collateral arteries in sixty-one patients, these were compared to the mapping of the bronchial arteries according to the available literature on bronchial artery anatomy.5 The distribution of the different branching patterns of major aortopulmonary collateral arteries arising from the aorta was virtually the same as the distribution of bronchial arteries described in previous angiographic studies (p = 0.32 and p = 0.24). Major aortopulmonary collateral arteries with anatomy similar to the right intercosto-bronchial artery were found in 87% of the patients and 50% of patients had major aortopulmonary collateral arteries originating from the subclavian artery region. All major aortopulmonary collateral arteries had anatomy similar in origin and proportion to that described for bronchial arteries. The conclusion was that these aortopulmonary collateral arteries could just be dilated bronchial arteries, which would infer a limited growth potential and possibly that the collateral artery tissue itself was unstable.

The approach originally designed in the late seventies had to be modified. The focus of this alternative strategy was aimed at native pulmonary artery growth rather than recruitment of major aortopulmonary collateral arteries. Several dogmas including some coming from the Royal Children’s Cardiac Unit had to be transgressed to undertake this strategy:
1. Large major aortopulmonary collateral arteries left untouched lead to rapid and irreversible pulmonary vascular disease in the lung segments connected to that collateral artery.
The unit’s experience suggests that the natural evolution of most major aortopulmonary collateral arteries is toward stenoses in the collateral artery, even if only mild, and thus providing a drop from systemic pressure to the pulmonary bed, and thus protective against pulmonary vascular disease. In any case, progression towards pulmonary hypertensive vascular disease is slow and rare. Echocardiographic studies, CT scan and MRI follow up have demonstrated this involution in our patients.

2. A significant number of patients with pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries have no central pulmonary artery.
By operating early in the neonatal period we have seen that the presence of a central pulmonary artery is almost the rule. Only in the presence of bilateral ducts is a central pulmonary artery likely to be absent. In that case, both branches usually have harmonious arborisation.

In our experience, all central pulmonary arteries can 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 that can be dilated to generate further pulmonary artery blood flow.

4. Most of the very diminutive pulmonary arteries have an incomplete pulmonary artery arborisation.
Pulmonary arteries normally branch to all segments. The arborisation of pulmonary arteries may be incomplete in patients with pulmonary atresia and ventricular septal defect with major aortopulmonary collaterals, as connections between distal pulmonary arteries and mapcas may exist in conjunction with or instead of the normal pulmonary artery pattern. Pulmonary artery arborisation cannot be determined fully until a shunting procedure has been performed and adequate blood flow through the native
pulmonary arteries attained. This will allow future assessment of pulmonary artery arborisation via angiography.

This new strategy aims at growing the native pulmonary artery 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 and was initially described by Haworth in 1981.\textsuperscript{9, 19, 20} Most of the previous authors, however, completed the initial operation by a series of translocations of the collateral arteries into the pulmonary artery branches once they had started to develop. Translocation or unifocalisation of collateral arteries do not feature in this management plan; on the contrary, the central shunt is replaced after several months with another source of central blood flow to continue pursuing the goal of growing pulmonary branches. Unifocalisation is avoided as it may hinder the growth potential of the pulmonary branches. This is still a work in progress but the ideal chronology of this multistage approach would be the following:

\textbf{Around the first week of life.} Initial exploration, performed with a contrast CT scan or formal angiography (figure 9 and 10), to demonstrate the size of the pulmonary artery branches, and the number, size and position of the major aortopulmonary collateral arteries.\textsuperscript{21} At times initial angiography has failed to show the presence of a central pulmonary artery that we were able to find intraoperatively.
Figure 9: Neonatal imaging of pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries. Late visualisation of confluent very diminutive central pulmonary arteries.

Figure 10: Neonatal imaging of pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries. Same examination as Figure 8. Injection in the descending aorta: visualization of major aortopulmonary collateral arteries from the descending aorta.
Within the first four to six weeks. A central shunt usually designed according to the Laks modification, constructed immediately on the central pulmonary artery origin where it dilates into a slight bulb (figure 10, 11).²²

![Diagram](image)

Figure 11: Schematic representation of diminutive pulmonary arteries showing the site of implantation of the central shunt.

![Photograph](image)

Figure 12: Intraoperative photograph of a central shunt.

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 adequate pulmonary blood flow from major
aortopulmonary collateral arteries, provided that central pulmonary arteries are diminutive.

Figure 13: Construction of the central shunt.

The aim is central pulmonary artery growth and significant pulmonary overcirculation is avoided because the small pulmonary artery branches initially restrict flow.

An alternative to the central shunt is the left modified Blalock-Taussig shunt from a left innominate artery when there is a right aortic arch. The shunts can be constructed with or without bypass. The aorto-pulmonary window creation advocated by Dr Mee and Dr Hanley is no longer used due to concerns about causing right pulmonary artery stenosis from kinking at its origin off the main pulmonary artery.\textsuperscript{19, 20} One of these pulmonary valves has been dilated, representing an alternative to the second operation; as well as an easier final repair.

A second evaluation at three to four 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 centrally increased flow and pressure, and to determine whether localised stenoses or hypoplasia have to be dealt with at the time of the first reoperation.
Figure 14: Within two to three months the central shunt has generated some growth of the central pulmonary arteries.

**Second operation, between four and six months of age.** The central shunt is taken down and a right ventricle to pulmonary artery conduit inserted. This can be a valveless conduit of 6 to 8 mm or a banded 12 mm valved conduit (figure 15).

Figure 15: a restrictive RV to PA conduit to promote further growth replaces the central shunt.
The rationale is to further augment the central pulmonary pressure and flow, which should continue to promote central pulmonary artery growth, as the central shunt may have become restrictive. Also, the right ventricle to pulmonary artery conduit provides easy access to the pulmonary artery branches for angiography and balloon intervention if necessary.

The third operation is either a complete repair or a second conduit with or without pulmonary artery branch surgical enlargement using autologous pericardium or ePTFE patches. Some patients at this stage have had ligation of a persisting major aortopulmonary collateral artery, however, most major aortopulmonary collateral arteries show significant involution. The final repair always includes a valved conduit (figure 16, 17).

Figure 16: The complete repair.
Figure 17: Injection in the pulmonary artery bifurcation 2 months after the repair.

**Technical aspects**

The central shunt is constructed along the recommendations of Laks. The pulmonary artery bifurcation is carefully dissected and the origin of the main pulmonary artery 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 pulmonary artery is exposed. It can be extremely small but is usually significantly larger than the diameter of the branches. This allows constructing 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 pulmonary artery with usually a 3 to 4 mm Goretex stretched conduit. Patient weight and size match to the main pulmonary artery determines size of conduit. This is done with full heparinisation if bypass is required or 100 UI/Kg otherwise. The implantation is at 90° with the main pulmonary artery. 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 can be 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 right ventricle to pulmonary artery 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 haemoclips 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 left anterior descending coronary artery. 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. Pulmonary artery branch arterioplasty is not usually performed at this stage unless significant asymmetry between branches is demonstrated. Localized narrowing of the branch pulmonary arteries can be dealt with dilation three to four weeks before the next procedure. Harmonious hypoplasia is dealt with by 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 have grown to near normal size at the hilum and pre-operative imaging shows pulmonary artery arborisation to more than 14 lung segments. Patch augmentation of the intra-pericardial branch pulmonary arteries may be required at this stage. Repairs have been done between fifteen and forty months of age and valved conduits are always used.
1.7 Early Angiographic results from a protocol of Repair without Unifocalisation

With a new paradigm of neonatal hypoplastic pulmonary artery rehabilitation - “repair without unifocalisation”, results from the first twenty patients entered into this treatment protocol are reviewed.

1.7.1 Methods.
The Royal Children’s Hospital Ethics committee provided approval for the study. Hospital records of all patients diagnosed with pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries admitted from June 2003 to December 2008 were reviewed. Twenty-five patients were identified. Twenty neonates were entered into an early shunting protocol aiming to rehabilitate native pulmonary arteries without translocation of collateral arteries and constitute the core of this study. Patient characteristics are shown in table 1.

<table>
<thead>
<tr>
<th>Number</th>
<th>20</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>14 (70%)</td>
</tr>
<tr>
<td>22q11 deletion</td>
<td>9 (45%)</td>
</tr>
<tr>
<td>Right Aortic arch</td>
<td>9 (45%)</td>
</tr>
<tr>
<td>Antenatal diagnosis</td>
<td>13 (65%)</td>
</tr>
</tbody>
</table>

Table 1: Patient demographics

Five patients were managed using alternate techniques:

- One patient, referred at the age of four years, had a normal left pulmonary artery perfused by a ductus, previously stented at birth, and an extremely hypoplastic right pulmonary artery with the right lung perfused by three collaterals. He underwent a right ventricle to pulmonary artery conduit, and is waiting repair aiming at directing the right ventricular blood flow to one lung.
- The second patient, born with normal size pulmonary arteries supplied by a single collateral vessel arising from the descending aorta, underwent a complete repair at the age of fourteen months.
- The third patient had a pulmonary circulation dependent on two major collaterals feeding normally sized confluent hilar pulmonary
artery branches. She underwent a one-stage repair and unifocalisation with a Contegra® (Medtronic Inc, Minneapolis, MN, USA) valved conduit used in the RV outflow tract.

- A fourth patient with Allagile syndrome was diagnosed at the age of three months. He underwent a right ventricle to pulmonary artery conduit but died four months later of liver failure.
- The last patient underwent staged rehabilitation of native pulmonary arteries starting at the age of ten years, because of late referral. He underwent two separate shunting procedures followed by complete repair at the age of fifteen years of age.

Pulmonary artery Nakata indices were calculated for all shunted patients who had serial measurements of pulmonary artery size available (n=17). Catheter data after complete repair was obtained to determine right and left ventricular pressures.

The following formulas were used:

- Body surface area (BSA) = $\sqrt{\frac{\text{height (cm)} \times \text{weight (kg)}}{3600}}$
- Pulmonary Artery Nakata Index = right pulmonary artery + left pulmonary artery cross sectional areas (mm$^2$) / BSA (m$^2$)

1.7.2 Surgical strategy.

The surgical strategy is described above. To summarise: All neonates were planned for elective central shunting within the first four weeks of life, regardless of their oxygen saturations. A Gore-Tex shunt (W. L. Gore & Associates, Flagstaff, AZ) was anastomosed to the pulmonary artery opening end-to-side with 8.0 prolene sutures. The proximal anastomosis consisted of a side-to-side anastomosis on the ascending aorta.

The patients subsequently underwent further shunting procedures or right ventricle to pulmonary artery conduit implantations until the pulmonary arteries had attained sizes that would allow complete repair.

1.7.3 Statistical analysis.

Data were reported as median (range) or mean (standard deviation) as appropriate.
1.7.4 Results.
Twenty patients with a diagnosis of pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries underwent an initial neonatal shunting procedure. At the time of last follow-up, the median age of patients was 34.82 months (5 – 79 months) with no mortality. Twelve of twenty patients have had a complete repair at a median age of 18 months (11 - 48), while six are awaiting repair and two are considered unlikely to be repaired. No patient was missing to follow-up.

1.7.5 Clinical Outcomes.

Procedure 1:
The median age at first procedure was 3.6 (0.7-17) weeks and mean preoperative oxygen saturations were 85% (72% - 95%). One patient’s first operation was delayed until seventeen weeks of life due to the presence of a bleeding peptic ulcer. All other patients had an initial shunting procedure before eight weeks of age regardless of oxygen saturation level. No patients were in heart failure prior to procedure 1. Initial shunting consisted of fifteen central shunts only (3mm n=2, 3.5mm n=11, 4mm n=2), four received a right modified Blalock-Taussig shunt (3mm n=2, 3.5mm n=1, 4mm n=1), and one patient had both a central and left modified Blalock-Taussig shunt (both 3mm). Concomitant pulmonary artery reconstruction with 0.4 mm thick Gore-Tex patches was performed in 2/20 (10%) patients.

Procedure 2:
Nineteen patients have undergone a second procedure at a median age of 7.9 (2.6 -20.7) months. Two out of nineteen patients (11%) had a complete repair, and twelve (63%) had a right ventricle to pulmonary artery conduit inserted using either 6 mm or 8 mm Gore-Tex grafts, or 12 mm Contegra valved conduits. Three of nineteen (16%) patients had a second systemic to pulmonary shunt (3.5mm n=1, 4 mm n=2) without creation of a right ventricle to pulmonary artery connection. These were all performed for origin stenosis of the left pulmonary artery. One patient underwent tricuspid valve repair for severe tricuspid regurgitation and one patient underwent pulmonary valve commissurotomy. Concomitant patch reconstruction of the pulmonary arteries was performed in nine (9/19) (47%) patients and three (3/19)
(16%) had major aortopulmonary collateral artery ligation for duplicate and excessive pulmonary blood flow.

Procedure 3:
Fourteen patients have undergone a third procedure at median age of 19.5 (4.8-40.4) months. Complete repair was performed in seven (7/14) patients, right ventricle to pulmonary artery conduit change in five (5/14), and ligation of major aortopulmonary collateral artery for excessive pulmonary blood flow in one (1/14). One patient had tricuspid valve repair with right ventricle to pulmonary artery conduit and systemic-pulmonary shunt to the left pulmonary artery. Nine patients had concomitant pulmonary artery patch reconstruction.

Procedure 4:
Seven patients have had a fourth operation at a median of 25 months (19.4-48.4). Complete repair was performed in three patients and all three had concomitant patch reconstruction of the pulmonary arteries. Three patients had further pulmonary artery reconstructions associated with right ventricle to pulmonary artery conduit change in one (1/7), and major aortopulmonary collateral artery ligation in one (1/7). One patient had an exploratory thoracotomy to assess the possibility of a major aortopulmonary collateral artery translocation, however, no suitable vessels were found for translocation.

1.7.6 Hospital morbidity.
There were no major airway complications after any of the procedures. The median ventilator times, ICU stay, and hospital stay after initial shunting, intermediate procedures, and final repair are given in Table 2.

<table>
<thead>
<tr>
<th>Operation</th>
<th>Ventilator time (hours) *</th>
<th>ICU stay (Days)</th>
<th>Hospital stay (Days)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Initial Neonatal Shunt</td>
<td>61 (8 - 166)</td>
<td>4 (1 – 9)</td>
<td>16 (7 – 53)</td>
</tr>
<tr>
<td>Intermediate procedures</td>
<td>28 (8-692)</td>
<td>2 (1 – 30)</td>
<td>10 (4 – 86)</td>
</tr>
<tr>
<td>Complete Repair</td>
<td>19 (8 – 78)</td>
<td>1 (1 – 4)</td>
<td>8 (4 – 11)</td>
</tr>
</tbody>
</table>

Table 2: Postoperative length of stay and ventilator time. * Data presented as median (range).
1.7.7 Follow-up.
After a mean of 39 ± 19 months, twelve patients have reached a complete repair and six are awaiting repair. All ventricular septal defects were closed, although one patient had a fenestrated flap patch which has never showed any shunting, and one patient was left with a small muscular ventricular septal defect expected to close spontaneously. In two patients, an atrial septal defect was left open because the pulmonary vessels seemed small. Two patients are currently not considered repairable (Figure 18).

![Diagram](image)

Figure 18. Outcome of all patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries.

All patients who attained complete repair had native pulmonary artery arborisation to at least fourteen lung segments as identified on preoperative angiograms. In three patients, treatment with sildenafil was instituted after their control angiograms.

On the last postoperative echocardiography performed a median of twelve months (4 - 48) after repair, the systolic right ventricular function appeared to be normal in all patients. The right ventricle appeared to be of normal size in three (3/12) patients, mildly dilated in seven (7/12) and moderately dilated in two (2/12) patients.

One patient left with palliation has 22q11 deletion and underwent 3 mm central shunt at 1.7 weeks of age, right ventricle to pulmonary artery conduit and left pulmonary artery reconstruction at five months of age, with left pulmonary artery balloon angioplasty at nine months of age. The Nakata index had increased from 4.92 at birth to 67.5 at nine months. While his native pulmonary arteries have increased in size and are distributed to both lungs, their arborisation was poor, supplying only ten lung segments. They also carried multiple stenoses, and the vascularisation
of both lungs seemed to depend mainly on multiple small major aortopulmonary collateral arteries.

The second patient left with palliation underwent initial central shunting at 2.7 weeks of age, right ventricle to pulmonary artery conduit at 4.5 months, and a valved right ventricle to pulmonary artery conduit at twelve months of age. The native pulmonary artery distribution was to only nine lung segments, with multiple bilateral major aortopulmonary collateral arteries providing the bulk of the pulmonary circulation. In both these patients, the major aortopulmonary collateral arteries were left untouched as they were providing most of the pulmonary blood flow.

1.7.8 Pulmonary artery growth.

Measurements of right and left branch pulmonary arteries were recorded from angiograms, direct measurements at operation, CT scan, MRI and echocardiography. These were plotted against body surface area as described by Nakata to provide a measure of pulmonary vascular arborisation. Data was available for seventeen (17/20) patients. The median Nakata index prior to first procedure was 14.51 (1.77-55.15). This increased to 118.7 (16.4-377.7) at a median of 17 months (5-53 months).

The last recordings prior to complete repair gave a median Nakata index of 101.4 (29.44 – 226.9). (Figure 19)

![Figure 19. Growth of pulmonary arteries (Each line represents a single patient)](image-url)
The Nakata index is often used in the literature to describe pulmonary artery growth. However, the value of these figures in determining the size of the pulmonary vascular bed is uncertain after Goretex patching of pulmonary artery stenoses. We determine readiness for complete repair using angiography to determine PA size compared to normal, PA arborisation to at least 2/3 of lung segments, and suitable pulmonary artery pressures.

Figure 20. Case 2 initial angiography at birth (A), after shunting then right ventricle conduit (B) and after complete repair with pulmonary artery patching (C).

1.7.9 Angiography after complete repair.
Twelve patients have achieved complete repair. Nine have been studied with angiography after a median of thirteen months (4-33) (table 3, figure 20). Their median right ventricular systolic pressure is 53 mmHg (43-65), median left ventricular pressure is 82 mmHg (68-100), and the median RV: LV pressure ratio is 0.64 (0.54-0.79). The median mean pulmonary artery pressure was 25 mmHg (22-37).

<table>
<thead>
<tr>
<th>Patient</th>
<th>Time from repair to angiography (months)</th>
<th>Residual ASD</th>
<th>Residual VSD</th>
<th>Mean MPA Pressure (mmHg)</th>
<th>RV Pressure (mmHg)</th>
<th>Systemic Pressure (mmHg)</th>
<th>RV:LV ratio</th>
<th>Oxygen saturation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>13</td>
<td>0</td>
<td>0</td>
<td>28</td>
<td>59</td>
<td>92</td>
<td>0.64</td>
<td>96</td>
</tr>
<tr>
<td>2</td>
<td>25</td>
<td>0</td>
<td>0</td>
<td>37</td>
<td>65</td>
<td>82</td>
<td>0.79</td>
<td>94</td>
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<tr>
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<td>13</td>
<td>0</td>
<td>0</td>
<td>29</td>
<td>53</td>
<td>80</td>
<td>0.66</td>
<td>97</td>
</tr>
<tr>
<td>4</td>
<td>8</td>
<td>1</td>
<td>0</td>
<td>25</td>
<td>47</td>
<td>72</td>
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<td>99</td>
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<td>5</td>
<td>8</td>
<td>0</td>
<td>0</td>
<td>22</td>
<td>43</td>
<td>68</td>
<td>0.63</td>
<td>100</td>
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<tr>
<td>6</td>
<td>25</td>
<td>0</td>
<td>1</td>
<td>22</td>
<td>43</td>
<td>80</td>
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<tr>
<td>9</td>
<td>33</td>
<td>0</td>
<td>0</td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

Table 3. Catheter haemodynamics after complete repair.
Six patients underwent fifteen balloon angioplasties of pulmonary artery stenoses. One patient also underwent stenting of the left pulmonary artery. Results have been variable, ranging from no improvement (stenosis remains present) (2/6), small to moderate improvement (incomplete resolution of stenosis) in (3/6), and complete resolution of gradient across branch pulmonary stenosis (1/6).

1.7.10 Complications.
There were seventeen major surgical complications in fifty-seven operations (table 3). Two patients had rapid oxygen desaturation in the immediate postoperative period after a central shunt procedure requiring chest reopening to relieve shunt compression in the Intensive care unit. One patient required four days of extracorporeal support for poor ventricular function after complete repair. Three patients developed vocal cord palsy after patch plasty of the left pulmonary artery.

<table>
<thead>
<tr>
<th>Complications</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shunt compression requiring urgent reoperation</td>
<td>2</td>
</tr>
<tr>
<td>Sepsis</td>
<td>3</td>
</tr>
<tr>
<td>Thoracocentesis</td>
<td>1</td>
</tr>
<tr>
<td>Pericardial effusion requiring draining</td>
<td>1</td>
</tr>
<tr>
<td>Wound dehiscence</td>
<td>2</td>
</tr>
<tr>
<td>Vocal cord palsy</td>
<td>3</td>
</tr>
<tr>
<td>Chylothorax</td>
<td>1</td>
</tr>
<tr>
<td>Bowel perforation</td>
<td>1</td>
</tr>
<tr>
<td>Bleeding requiring reoperation</td>
<td>1</td>
</tr>
<tr>
<td>ECMO</td>
<td>1</td>
</tr>
<tr>
<td>Prolonged ventilation requiring tracheostomy</td>
<td>1</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>17</strong></td>
</tr>
</tbody>
</table>

Table 4. Complications
1.8 Conclusion

The vast majority of surgical literature published over the last fifteen years concerning the treatment of patients born with pulmonary atresia, ventricular septal defect and major aortopulmonary collaterals promotes the unifocalisation of collateral arteries to create a new pulmonary vascular bed.\textsuperscript{16, 24, 25} This strategy has undoubtedly improved survival in this patient population.\textsuperscript{17, 24}

During unifocalisation procedures, collateral vessels are joined to either native pulmonary vessels or newly created central vessels, however, it has been demonstrated for many years that hypoplastic central pulmonary arteries could be developed or “rehabilitated” by shunting procedures alone.\textsuperscript{9, 26} A number of patients in the published series of patients managed with unifocalisation strategies will also have had previous shunting operations on their native pulmonary vessels. Because of the complexity and the heterogeneity of the procedures included under the term unifocalisation, it is very difficult to evaluate the relative benefits of integrating collaterals vessels into the pulmonary circulation and comparing it to the benefits acquired by the recruitment of native vessels.

The 2005 review of our Unit’s experience of multistage unifocalisation procedures in eighty-two of our initial patients reported disappointing results with the translocation of collateral arteries.\textsuperscript{18} Half of the identified translocated collateral arteries thrombosed, a third of those remaining patent developed significant stenoses, and the collaterals that remained patent did not seem to show any growth. For these reasons we decided to reserve translocation of collateral arteries for exceptional cases, where large collaterals connect to pulmonary vessels of normal calibre. Since then we have focused on a strategy of rehabilitation of the native pulmonary vessels. Our hypothesis is that pulmonary arteries would have the best chance to grow if the initial shunting procedure was done early in life; therefore we planned the initial operation for the first few weeks of life regardless of patient oxygen saturations. We thereafter proceeded with repeated shunting procedures or implantation of right ventricle to pulmonary artery conduits until we were satisfied we had obtained pulmonary arteries of sizes that were large enough to allow definitive
repair. While The Nakata index has been used in our study to quantify pulmonary artery growth after original central shunting, we have not used it clinically to determine suitability for complete repair. Our main guide has been when the size of the proximal pulmonary arteries approaches normal and when these pulmonary arteries connect to more than two thirds of all pulmonary segments.

Rehabilitation of the native pulmonary arteries in patients with pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries is an old concept, but it does not seem to have been reported as a single strategy in recent times, especially in asymptomatic neonates. We initially considered this an investigational approach, however, by the end of this initial period we felt justified in continuing with the protocol. The numbers of patients enrolled with this rare condition are still small and a larger cohort of patients with longer follow-up will be necessary to ascertain any superiority of the approach. The fact that we encountered no early mortality related to the condition or to the surgery is certainly encouraging.

One of our hypotheses was that the survival of some patients is dependent on their aorto-pulmonary collaterals. Therefore, translocation of collaterals may put the patient at risk of postoperative cyanosis and death if pulmonary blood flow becomes compromised. We believe that if the network of aortopulmonary collaterals is left intact, the initial procedures to rehabilitate the native pulmonary arteries have a “fail-safe” mechanism, because even if the central shunt occludes, the patients will be left with the same amount of pulmonary blood flow they had before the procedure. In exceptional cases, a patient’s pulmonary circulation may have very large collaterals and increasing pulmonary blood flow can put them into heart failure. In these cases, ligation or embolisation of the collateral has to be envisioned either pre-or post-operatively. We are prepared to allow a shared pulmonary blood supply state especially in this early phase, with the expectation that MAPCAs will develop stenosis over time or any large MAPCA that does not regress may be treated surgically or embolised at a later state to prevent pulmonary hypertension and segmental pulmonary vascular obstructive disease.

The main interest in this strategy of native pulmonary artery rehabilitation is that it demonstrates the true potential of the native pulmonary circulation
to develop and to connect to enough segmental pulmonary branches to allow subsequent repair.

In the majority of our patients, the pulmonary vasculature has developed enough to allow a successful repair. The fact that in the majority of our patients, the central pulmonary arteries, once developed, were connected to the majority of lung segments, demonstrates that unless these arteries are rehabilitated, it is impossible to determine their potential to contribute to the final pulmonary vasculature. Some have described that up to 16% of patients with the condition are born with no central pulmonary arteries, which would preclude a strategy of early shunting of the central pulmonary arteries. This finding, however, has not been our experience, with all patients having central pulmonary arteries. Two of our twenty patients did have pulmonary arteries that were so small that they were considered disconnected and required two separate shunts. Only one patient had a left branch pulmonary artery that did not achieve growth with shunting. It is possible that we were able to identify central pulmonary arteries in a larger proportion of patients because they were operated on earlier in life, as after birth, without pulmonary blood flow, these arteries may atrophy and disappear. Our present results were obtained in a small number of patients compared to the larger presented series and one cannot exclude that a cluster of patients with more favourable anatomy favoured us, we expect some patients may be born with no identifiable central pulmonary arteries. Most large published series arise from centre’s that receive specific referrals for this condition, and it is possible that the spectrum of the disease they encounter is different from our unselected population.

In the last few years since adopting a unified initial strategy of central shunting in neonates, we have decreased initial investigation by diagnostic catheterisation. We use CT scan or MRI now to initially evaluate patients, and diagnostic catheterisation is reserved to the preoperative evaluation of the subsequent staged operations.

There is a general agreement that the anatomic substrate of this condition varies, even among patients whose lung circulation is exclusively dependent on aorto-pulmonary collaterals. Some may only have minimal hypoplasia of otherwise normal pulmonary arteries and at the other extreme, some patients’ lung circulation, even after rehabilitation of native vessels, will exclusively depend on vessels with more similarities to bronchial arteries.
than pulmonary vessels. It could be argued that the patients at the worst end of the spectrum will not benefit from a strategy of rehabilitation of the native pulmonary vessels, and that they are likely to be left with palliation. Only two of our twenty patients so far have been left to palliation, so that it seems that, at this time, this approach would potentially allow an achievable rate of repair up to 90%. This matches even the best-reported outcomes of unifocalisation strategies.\(^{17, 24}\) Despite two patients being deemed unrepairable, this does not preclude a significant improvement in morbidity and mortality. Results from our previous study showed 31% mortality in palliated patients at median time of eight years, however median follow-up in survivors was thirteen years, one patient even had follow-up fourteen years after heart-lung transplantation.

Determining what is an acceptable right ventricle pressure after complete repair remains a contentious issue. Typically the systolic right ventricular pressure is compared to the systolic left ventricular or systemic pressure, with ratios of less than 0.6 considered acceptable, however some groups will accept ratios as high as 0.8 - 1.\(^{17, 27}\) We were slightly disappointed by the mean residual right ventricle pressures of this patient population after repair. It seems comparable to most series of the staged approach,\(^{24, 28}\) but inferior to the neonatal one-stage approach.\(^{17}\) Unfortunately, it is not known whether such pressures will affect the long-term survival of these patients. Most groups would be concerned about eventual right ventricular dysfunction if the right ventricular pressure were left greater than two-thirds systemic for the long term. It is encouraging, however to notice that the median mean pulmonary artery pressures in our patient group was low. We can therefore expect that their right ventricular pressures will decrease after the subsequent procedures of replacement of the right ventricle to pulmonary artery conduits.

This will only be shown with ongoing follow-up, but we believe that native pulmonary arteries will have better growth potential than unifocalised major aortopulmonary collateral arteries.\(^{29}\) Our limited experience with angioplasty of pulmonary stenoses has been mixed, with initial improvements not long-lasting. Others have shown pulmonary artery stenting via traditional or hybrid approaches can have better success rates,\(^{30, 31}\) but our unresolved concerns regarding possible limitations of pulmonary artery growth have precluded extensive use of this modality.
The recent modification of our protocols for the approach to pulmonary arteries of patients born with pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries is still a work in progress. So far, however, we have been able to enrol most patients presenting to our unit into this regime with the aim of achieving growth of the pulmonary arteries without the use of unifocalisation. Patients who have reached complete repair demonstrate sub-systemic right ventricular pressure and very few major aortopulmonary collateral arteries have required ligation at the stage of repair or earlier, demonstrating that the fate of most major aortopulmonary collateral arteries in a competitive flow environment is essentially towards stenosis and reduction of 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 this approach. We shall be looking at the evolution of the right ventricular and pulmonary artery 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.

In conclusion, a strategy focusing on rehabilitation of the pulmonary arteries of patients born with pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries by an early shunting procedure in neonates restores a pulmonary vasculature able to complete full intracardiac repair in a majority of cases. Low early mortality and high rate of repair encourages us to pursue further application and investigation of this strategy.
2. PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM

2.1 Natural History

Pulmonary Atresia with Intact Ventricular Septum is a congenital malformation in which the pulmonary valve is atretic and coexists with varying degrees of right ventricular and tricuspid valve hypoplasia. Usually the cavity of the right ventricle is hypoplastic, the right ventricular myocardium thickened and the right ventricular pressures can be suprasystemic. Also contributing to the morphological heterogeneity of the lesion is the size and structure of the tricuspid valve, whether the right ventricular outflow tract obstruction is muscular or valvar, and the presence of coronary artery anomalies - specifically right ventricle to coronary artery connections and any stenoses within these coronary vessels which may make coronary blood flow dependent on flow from the right ventricle.

Figure 21: Schematic representation of blood flow in pulmonary atresia with an intact ventricular septum. There is an obligatory right to left shunt at the atrial level.
Figure 22: Examples of right ventricle to coronary artery connections. A) without coronary artery stenoses, B) With proximal and/or distal coronary stenoses, C) With coronary occlusion.  

The diameter of the tricuspid valve correlates with the size of the right ventricle cavity and provides a useful index of right ventricular size. The prevalence of right ventricle to coronary artery collections is inversely proportional to the dimensions of the tricuspid valve and size of the right ventricle. In the presence of proximal coronary artery stenosis, desaturated right ventricular blood may pass from the right ventricle through so-called “fistulae” or “sinusoids” (right ventricle to coronary artery connections) and supply the myocardium. This compromises myocardial oxygen supply and can cause myocardial infarction. 

The tricuspid valve is usually abnormally small in size. The leaflets may also be thickened and chordae abnormal in number and attachment. In rare cases the tricuspid valve is unusually large and malformed with features of the Ebstein malformation with downward displacement of a dysplastic septal leaflet into the ventricle. These valves are usually severely regurgitant and can cause giant right atria the so-called “wall to wall heart”. Fetal distress is usually not present. After birth the baby is typically fully-grown but cyanosed. Cyanosis progresses rapidly with increasing respiratory distress and increasing metabolic acidosis as the ductus arteriosus closes. Ninety percent of patients will present in the first week of life, with fifty percent dying within 2 weeks of birth and eighty-five percent by six months. Surgical management ideally is aimed at opening up the right ventricular outflow tract allowing for a biventricular repair. However,
factors such as hypoplasia of the right ventricle and tricuspid valve can preclude successful integration of the right ventricle into the repair and a single ventricle palliation strategy is usually undertaken. The UK and Ireland collaborative study of pulmonary atresia with intact ventricular septum provides excellent data on the morphological variability of this lesion. This population-based study conducted over five years found one hundred and eighty three patients born with this lesion. Most of the outflow tract obstruction (74.7%) was membranous as opposed to muscular (25.3%). Most patients had a tripartite right ventricular structure (58.7%), bipartite was (33.6%), and unipartite was (7.7%). Typically the right ventricle can be divided into three parts: the inlet, the apex, and the outlet, however, with the muscular right ventricular hypertrophy that develops in this condition one or all of these part’s cavities may become obliterated.

Forty five percent of patients had right ventricle to coronary artery connections, but only 7.6% had significant coronary anomalies of stenosis, interruption or ectasia. The median tricuspid valve Z score was -5.2, which means the tricuspid valve size was five standard deviations smaller than published nomograms indexed to body size for tricuspid valve measurements. The results were disappointing. Fifteen patients did not undergo any surgical procedure at all and all died before three years of age. At follow-up of nine years, a total of forty-one percent of patients had died. Three independent risk factors for mortality were found:

1. Unipartite right ventricle structure
2. Low birth weight
3. Significant right ventricular dilation.

Significant right ventricular dilation generally reflects those patients with Ebsteinoid tricuspid valves, giant right atriums and wall-to-wall hearts. Mortality data from this malformation is often considered separately from the other forms of pulmonary atresia with intact ventricular septum given its very high degree of mortality.

Low birth weight is a well-described risk factor in neonatal cardiac surgery in general not just for this specific lesion and can often reflect prematurity of organ function and associated conditions.
Unipartite right ventricular structure reflects the most extreme morphological subtype of this malformation, with the highest rate of coronary anomalies.

Earlier studies have found that a smaller tricuspid valve Z-score and the presence of a right ventricle dependent coronary circulation to be significant risk factors for mortality. However, since the recognition of these risk factors Cardiac Units have adjusted surgical strategies towards performing a safer univentricular palliation in higher risk patients and later studies reflect this change in practice by the removal of these factors as significant risk factors for mortality although there are some units who will try and ligate coronary sinusoids in the hope of eventual biventricular repair. More recently reports utilizing trans-venous catheter techniques to relieve right ventricular outflow tract obstruction have shown some promise in reducing early operative risk.
2.2 Evolving Surgical Management at the Royal Children’s Hospital

The last decade has seen a marked improvement in the surgical outcomes of patients with pulmonary atresia with intact ventricular septum.\textsuperscript{36,38,39} The prospective multi-centric study by the congenital heart surgeons’ society now predicts a 5-year overall survival of 79% for those enrolled in 1997, compared with 49% survival if enrolled in 1989.\textsuperscript{40} This improvement has occurred despite profound differences in the approaches advocated by the different institutions involved.

Initially, centers reporting success with the treatment of this condition advocated a single approach, some favoring a bi-ventricular pathway, others a univentricular pathway. It is now becoming clear that a strategy excessively favouring a univentricular palliation will deprive some patients with relatively normal right ventricular cavities from the benefits of a bi-ventricular repair and the potential for a longer life expectancy. On the other hand, a strategy pushing for bi-ventricular repair in all cases will result in increased mortality.\textsuperscript{40} Today, a more balanced approach seems preferable, and preliminary success has been reported.\textsuperscript{38} Unfortunately there are yet no clear criteria to decide which patients should be oriented to a univentricular or bi-ventricular repair. In Melbourne, the strategy has gradually evolved over the years. Initially the policy was aimed at growing patient’s right ventricular cavity and pushed indications for bi-ventricular repair. The decision to attempt bi-ventricular repair was based on the presence or absence of a right ventricular infundibulum.\textsuperscript{41} Those patients with a patent infundibulum headed towards a biventricular repair, sometimes requiring a “right ventricle overhaul” procedure - consisting of resection of intra-ventricular obstructive muscle bundles in order to enlarge the right ventricular cavity.\textsuperscript{42} Only patients without an infundibulum were placed on the univentricular heart pathway. In the patients with only moderately reduced right ventricular size, this approach allowed us to delay the final decision between a univentricular or biventricular repair. During the last fifteen years, we have progressively stepped back from this methodology and adopted a more balanced approach. Patients with the smallest right ventricles or tricuspid valves, and coronary anomalies were directed towards palliative univentricular procedures. In the remaining
patients the right ventricular outflow tract was opened surgically or with a catheter intervention and targeted for biventricular repair. Up to now, however, the decision to aim towards a biventricular or univentricular repair was made after the review of each individual case without any specific guidelines. This review of the Royal Children’s Hospital’s experience with patients presenting with pulmonary atresia and intact interventricular septum is aimed at determining risk factors for mortality, hoping that the identification of these risk factors may guide decision-making towards either a univentricular or a biventricular pathway.

2.2.1 Methods.
The study was approved by the Royal Children’s Hospital Ethics committee. All patients at the Royal Children’s Hospital admitted between 1990 and 2006 with the diagnosis of pulmonary atresia and intact interventricular septum were identified from the hospital database (n=93). Patients with clear Ebstein’s anomaly or severely dysplastic tricuspid valves with associated pulmonary atresia were then excluded from the study (n=12), and the remaining 81 patients constitute the core of this study. All preoperative echocardiographic examinations were reviewed by a single cardiologist, blinded to the outcome, to assess right ventricular size, right ventricle to coronary artery connections and tricuspid valve annulus diameter. Right ventricle size was subjectively categorized into 3 groups - normal, moderate, and severely hypoplastic. (Fig 21) This was based on the size of the neonatal right ventricle in comparison with the left ventricle, the partite nature of the right ventricle, and the extent to which muscular hypertrophy obliterated the right ventricle cavity.
Figure 23: Examples of (a) severe and (b) moderate hypoplasia, and (c) near-normal sized right ventricle in cases of pulmonary atresia with intact ventricular septum.

Angiography was reviewed for all patients with right ventricle to coronary artery connections to determine whether their coronary circulation was dependent on right ventricle pressure. Right ventricle dependent coronary circulation was defined by the presence of right ventricle connections to the coronary arteries with either severe stenosis in two or more major coronary arteries (right coronary artery, left main, left anterior descending, left circumflex artery, and posterior descending artery), or complete aorto-coronary atresia. The following variables were analysed; birth weight, gestational age at birth, antenatal diagnosis, age at first intervention, tricuspid valve Z-score, presence of right ventricle to coronary artery connections, right ventricle dependent coronary circulation, and right ventricle size.

Primary intervention was classified as “shunt only” for a systemic to pulmonary shunt, “RV outflow tract procedure only” if a procedure was performed that allowed right ventricle to pulmonary artery flow (including surgical valvotomy, trans-annular patching, and successful catheter valvotomy), or both. Any patient, who had significant resection of right ventricular muscle bundles beyond the infundibular area at any stage, was classified as having had a right ventricular overhaul. A one and a half ventricle repair was defined as having a bidirectional cavopulmonary connection (SVC anastomosed to right pulmonary artery) and forward flow through a patent right ventricle to pulmonary artery conduit. No formal management policy was in place for those patients considered to have true
right ventricular dependent coronary circulation, and no right ventricle exclusions were performed by tricuspid valve closure in this subset of patients.

Patient follow-up was extracted from the hospital database or sought from the referring cardiologist. Each patient was ascribed to one of six end-statuses: Biventricular repair, 1½ ventricle repair, Univentricular, death, heart transplantation, and awaiting complete repair. The biventricular repair group included patients with an atrial septal defect left open, and the 1½ ventricle repair group included patients with a bidirectional cavopulmonary shunt and right ventricle to pulmonary artery forward flow, regardless of the presence of an atrial septal defect. The “awaiting complete repair” category included patients with bidirectional cavopulmonary anastomoses on the univentricular pathway and patients with systemic to pulmonary shunts where right ventricular growth was being assessed over time prior to determining suitability for univentricular or biventricular repair.

2.2.2 Statistical analysis:
Data were described as mean ± standard deviation for normally distributed variables and median and interquartile range for non-normally distributed variables. Categorical data were analysed with Fishers exact test. Continuous data was analysed with the Students unpaired T-test. A p-value < 0.05 was considered as statistically significant with all p-values based on two-tailed tests. Actuarial survival was estimated by means of the Kaplan-Meier method. Factors showing a trend towards or significant association with death on univariate analysis were evaluated by multivariate logistic regression analysis with exclusion of those with significant interaction. Statistical analysis was performed using STATA 10.0 (Stata Corp, Texas, USA).

2.2.3 Results.
Between 1990 and 2006, eighty-one patients with pulmonary atresia with intact interventricular septum were identified in our institutional database. Their mean birth weight was 3.8 +/- 0.8 kg, and median age at initial intervention of 4.5 (2 - 11) days. Thirty-two patients (49%) were born with an accurate antenatal diagnosis. Twelve patients received their initial intervention beyond the neonatal period (>1 month) either because of
prematurity and low birth weight (n=9) or delayed presentation due to the presence of aortopulmonary collaterals (n=3).

By retrospective review, right ventricular size was classified as being normal in eleven patients (14%), moderately hypoplastic in forty-five (56%) and severely hypoplastic in twenty-five (31%). The median tricuspid valve Z-score of the patients with normal size right ventricle was -0.6 (-0.9 to 0.9), moderate -1.3 (-1.9 to -0.9) and severe hypoplasia -2.7 (-3.2 to -2.4). Mean overall tricuspid valve Z-score was -1.46 +/- 1.39 and right ventricle to coronary artery connections were present in thirty-three patients (41%). Right ventricle coronary dependence was identified in six patients (7%). Mean follow-up was 7.1 +/- 5.6 years.

2.2.4 Death prior to intervention

Three patients died before any procedure. The first was born at term with birth weight of 3.27kg. His tricuspid valve Z-score was -3.4, right ventricle to coronary artery connections were present, and the coronary circulation was considered right ventricle dependent. Active treatment was withdrawn after parental request.

The other two patients were born prematurely at 32 and 34 weeks gestation, with birth-weights of 1.42 and 1.91 kg. Their management involved an assisted feeding regime to allow growth prior to operative intervention. Both died before this could occur.

2.2.5 Diagnostic and interventional catheterization

From 1990 to 2006, seventy-one (71/81) patients underwent diagnostic catheterization.

The primary intervention in seventeen patients was an interventional cardiac catheterisation with wire perforation and balloon pulmonary valvuloplasty. Transcatheter laser or radiofrequency-assisted techniques were not used in our Institution during this time period. The characteristics of these patients are displayed in table 5.

Transcatheter wire perforation and balloon valvuloplasty treatment has become a more common procedure for membranous pulmonary valve atresia with intact ventricular septum in patients with a tripartite right ventricle lacking significant coronary communications.
<table>
<thead>
<tr>
<th>Patient characteristics:</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of patients</td>
<td>17</td>
</tr>
<tr>
<td>Birth weight (kg)</td>
<td>3.4 +/- 0.6</td>
</tr>
<tr>
<td>Gestation (weeks)</td>
<td>40 (38 - 40)</td>
</tr>
<tr>
<td>Antenatal diagnosis</td>
<td>5 (29%)</td>
</tr>
<tr>
<td>Age at first intervention (days)</td>
<td>2 (1 - 6)</td>
</tr>
<tr>
<td>TV Z-score</td>
<td>-1.0 (-3.1 - 0.9)</td>
</tr>
<tr>
<td>Presence of RVCAC</td>
<td>1 (6%)</td>
</tr>
<tr>
<td>RV size/hypoplasia:</td>
<td></td>
</tr>
<tr>
<td>Normal</td>
<td>5 (29%)</td>
</tr>
<tr>
<td>Moderate</td>
<td>12 (71%)</td>
</tr>
<tr>
<td>Severe</td>
<td>0</td>
</tr>
<tr>
<td>RVDCC</td>
<td>0</td>
</tr>
</tbody>
</table>

Table 5. Patient Characteristics for Primary Catheter intervention.
(RVCAC = right ventricle to coronary artery connections, RVDCC = right ventricle dependent coronary circulation).

The major procedural risk remains inadvertent perforation of the right ventricular outflow tract and/or main pulmonary artery. The technique is described as follows:

Venous and arterial access is obtained using the modified Seldinger technique. Typically a 5-French femoral venous sheath and a 3- or 4-French femoral arterial sheaths are used. 4-French umbilical artery sheaths may also be used. A right ventriculogram confirms ventricular size, outflow tract anatomy and significance of right ventricle-to-coronary communications. A left ventricular or aortic root injection better defines coronary anatomy and suitability for right ventricular decompression.

An end-hole catheter such as a 4-French angled Glide (Terumo Medical, Somerset, NJ) is passed retrograde across the ductus arteriosus and an angiogram performed of the main pulmonary artery to assess pulmonary annular diameter. A 7- or 10-mm Amplatz Goose Neck microsnare (EV3, Plymouth, MA) is then passed through the retrograde end-hole catheter to abut the atretic pulmonary valve. The microsnare loop serves as a frontal plane fluoroscopic target or guide for the wire perforation procedure.
A 4-French JR 2.5 coronary catheter (Merit Medical, South Jordan, UT) is placed in the right ventricular infundibulum just proximal to the atretic valve. The pre-formed curve of a right coronary catheter most appropriately aligns or directs the perforation system to a plate-like valve. A positioning angiogram in the infundibulum confirms the coronary catheter tip position relative to the valve. The radiofrequency (RF) wire perforation kit is a coaxial system: a Nykanen RF wire passed through a 2.7-French Pro-Track microcatheter (Baylis Medical, Montreal, Canada). This unit is advanced through the right coronary catheter to approximately 5-mm short of the catheter tip. The Nykanen RF wire is then advanced gently to the catheter tip. The table-end of the RF wire is connected to a Baylis RF generator and typically programmed to deliver output of 5 W for 2–5 s.

With controlled application of energy, the RF wire is gently advanced toward the circular loop of the Goose Neck microsnare target. Special care is taken to advance the RF wire to just beyond the microsnare loop in the main pulmonary artery during energy application, thereby significantly reducing risk of inadvertent right ventricular or main pulmonary artery perforation. Once the RF wire is passed through the microsnare loop, the microsnare can be utilized to secure the RF wire, pull it through the ductus arteriosus and into the descending aorta. With the RF wire secured, the Pro-Track microcatheter can be easily advanced across the pulmonary valve and into the descending aorta as well. The Pro-Track microcatheter will accept a 0.025-inch guide wire for intervention. At this point, the RF wire is removed from the Pro-Track catheter and replaced with an exchange length guide wire. Balloon valvuloplasty can then proceed typically using a Tyshak II balloon dilation catheters (B. Braun Medical, Bethlehem, PA) for valvuloplasty, utilizing a diameter 100–120% of the pulmonary annular dimension.

Right ventricle dependent coronary circulation was not present in any patient and no patient had a severely hypoplastic right ventricle. In four patients (24%) catheter valvotomy was considered a technical failure. These patients underwent subsequent surgical intervention at a median of one (0–2) day, involving: a left modified Blalock-Taussig shunt and pulmonary valvotomy (n=2), pericardial effusion drainage, right modified Blalock-Taussig shunt and pulmonary valvotomy (n=1), and pulmonary valvotomy (n=1). Follow-up data showed that two ended up with a biventricular repair and two with a 1½-ventricle repair.
In thirteen patients transcatheter valvotomy was considered a technical success. However, nine (9/13) required further surgery at a median of nine days (1-25) involving: a modified Blalock-Taussig shunt (n=8) including two with concomitant pericardial effusion drainage, and a pulmonary valvotomy with arch repair (n=1). The single death (1/17) occurred late and was due to poor myocardial function in the one patient of the group who had right ventricle to coronary artery connections. The final outcomes of the seventeen patients undergoing primary management with interventional catheterization are detailed in figure 23.

Figure 24: Final outcomes after initial catheter intervention.

2.2.6 Requirement for systemic to pulmonary shunt
The overall requirement of a systemic to pulmonary shunt was 85% (69/81). All patients with a severely hypoplastic right ventricle received a shunt except for the three patients described above that died before any intervention. (Table 6) Most patients (8/11) with normally sized right ventricles also required a shunt.
Table 6: Requirement for Systemic to pulmonary shunt

<table>
<thead>
<tr>
<th>RV hypoplasia</th>
<th>Shunt only</th>
<th>Shunt &amp; RVOT procedure</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>1</td>
<td>7</td>
<td>8/11 (73%)</td>
</tr>
<tr>
<td>Moderate</td>
<td>16</td>
<td>23</td>
<td>39/45 (87%)</td>
</tr>
<tr>
<td>Severe</td>
<td>20</td>
<td>2</td>
<td>22/25 (88%)</td>
</tr>
</tbody>
</table>

2.2.7 Right Ventricular overhaul

Eighteen of eighty-one patients (22%) underwent a right ventricle overhaul procedure. Fourteen of eighteen resulted in biventricular repair and four (4/18) resulted in a 1½-ventricle repair.

2.2.8 Outcome

Final end-statuses were death 16/81 (20%), biventricular repair 31/81 (38%), 1½-ventricle repair 10/81 (12%), Univentricular repair 14/81 (17%), heart transplant 1/81 (1%), and awaiting final procedure 9/81 (11%). Of the nine patients awaiting completion of repair, four are on the Univentricular pathway, four patients are left with shunts waiting a final management decision and one has been lost to follow-up. (Table 3)

Table 7: Outcomes based on right ventricle size. (Bi-V = Biventricular, 1.5-V = 11/2 ventricle, Uni-V = univentricular, HTx = heart transplant)

<table>
<thead>
<tr>
<th>RV size/hypoplasia</th>
<th>Outcome</th>
<th>Bi-V</th>
<th>1.5-V</th>
<th>Awaiting</th>
<th>Uni-V</th>
<th>HTx</th>
<th>Dead</th>
<th>Total (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>10</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>11 (13.5%)</td>
</tr>
<tr>
<td>Moderate</td>
<td>19</td>
<td>8</td>
<td>3</td>
<td>9</td>
<td>0</td>
<td>6</td>
<td>0</td>
<td>45 (55.5%)</td>
</tr>
<tr>
<td>Severe</td>
<td>2</td>
<td>1</td>
<td>6</td>
<td>5</td>
<td>1</td>
<td>10</td>
<td>1</td>
<td>25 (31.0%)</td>
</tr>
<tr>
<td>Total</td>
<td>31</td>
<td>10</td>
<td>9</td>
<td>14</td>
<td>1</td>
<td>16</td>
<td>0</td>
<td>81 (100%)</td>
</tr>
</tbody>
</table>

None of the patients with a normal size right ventricle ended with a univentricular heart circulation, but two of the patients with a severely hypoplastic right ventricle ended up with a bi-ventricular repair. These two
patients had tricuspid valve Z-scores of -2 and -1.7 and one required a right ventricle overhaul procedure to enable biventricular repair.

No patient with a tricuspid valve Z-score less than -2 (n=27) and only one patient (n=33) with right ventricle to coronary artery connections have attained a biventricular repair. All deaths occurred in the first two years after birth. The causes of death were cardiac 75% (12/16), unknown 12.5% (2/16), respiratory failure 6% (1/16), and withdrawal of treatment 6% (1/16). In the twelve cardiac deaths, coronary ischemia is presumed to have played a significant role in inducing either a cardiac arrhythmia or sudden death. The three patients that did not receive an intervention died at days 2, 9, and 31 of life. Of the other thirteen patients the median days until death after initial intervention was 145 (80 to 211), only two patients died within 30 postoperative days. One patient died intra-operatively on day three of life when undergoing a modified Blalock-Taussig shunt. Bradycardia occurred during thoracotomy and was unresponsive to intervention. The second patient had poor biventricular function post transannular patching and Blalock-Taussig shunt. She required a ventricular assist device for increasing lactic acidosis on day one and on return to theatre was found to have an aneurysmal ventricular septum impeding left ventricular function. Ventricular function did not recover and death ensued after withdrawal of the ventricular assist device.

Of the eleven patients on the univentricular pathway that died, only two had undergone bidirectional cavo-pulmonary connection prior to death. The remaining eleven patients were all dependent on a systemic-pulmonary shunt without any forward pulmonary blood flow.

Fourteen out of sixteen of the patients dying in the follow-up period were known to have right ventricle to coronary artery connections and three (3/6) who were identified to have right ventricle coronary dependence died. After fifteen years, Kaplan Meier actuarial survival was 80% (95%CI: 71-87%) (Figure 24).
Figure 25: Kaplan Meier actuarial survival.

2.2.9 Risk factors for death

Univariate analysis revealed that lower tricuspid valve Z-score (p<0.0001), right ventricle to coronary artery connections (p<0.001), smaller right ventricle size (p=0.005), and right ventricle dependent coronary circulation (p=0.026) to be statistically significant risk factors for death. Multivariate logistic regression revealed that only lower tricuspid valve Z-score and presence of right ventricle to coronary artery connections were independent risk factors for death.

<table>
<thead>
<tr>
<th>Risk Factors for Death</th>
<th>Univariate</th>
<th>Multivariate</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>P-value</td>
<td>P-value</td>
</tr>
<tr>
<td>Birth weight (kg)</td>
<td>0.064</td>
<td></td>
</tr>
<tr>
<td>Gestation (weeks)</td>
<td>0.085</td>
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<tr>
<td>Antenatal diagnosis</td>
<td>0.158</td>
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<tr>
<td>TV Z-score</td>
<td>&lt;0.0001</td>
<td>0.021</td>
</tr>
<tr>
<td>Presence RVCAC</td>
<td>&lt;0.001</td>
<td>0.019</td>
</tr>
<tr>
<td>RV size:</td>
<td>0.005</td>
<td></td>
</tr>
<tr>
<td>RVDCC</td>
<td>0.026</td>
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</tbody>
</table>

Table 8: Risk factors for death
**2.3 Conclusions**

Pulmonary atresia with an intact interventricular septum still carries a high risk of early mortality. In this series, twenty percent of patients died during follow-up, with most of the deaths occurring within the first two years of life. These results are comparable to other contemporary reports.\(^{38, 40, 44}\)

The Boston Children’s Cardiac unit has reported mortality as low as 2% at 7.5 years;\(^{36}\) however, this involved a high rate of univentricular palliation. It has now been clearly demonstrated that performing a large proportion of univentricular heart palliations is the best way to reduce mortality in this patient population.\(^{40}\) This practice, however, may deny a bi-ventricular repair and the possibility of increased longevity to patients with favourable morphology. Most groups today would agree that a balanced approach is necessary in the management of these patients, reserving a univentricular pathway to patients with right ventricles at the smallest end of the spectrum and attempting a bi-ventricular repair to those with near-normal size right ventricles.

Initial management will differ according to stratification towards univentricular palliation or biventricular repair. While these decisions are easily taken for those at the extreme ends of the spectrum, clinicians still lack criteria to make decisions for the majority of those between these extremes.

A simple approach based on echocardiography was used to retrospectively stratify right ventricular size in this study. Patients were characterized into 3 groups (normal, moderately hypoplastic, and severely hypoplastic). This approach seemed the easiest and most practical way of stratifying patients. It separated on a subjective basis those for whom the univentricular approach was the only reasonable approach, from those for whom this decision was much more difficult. While this classification helped us in the overall description of our patient population, it did not seem to be a predictor of outcome as accurate as the tricuspid valve Z-score. The relationship between right ventricular size and the tricuspid valve annulus Z-score has been well described.\(^{33}\) This study and experience suggests that patients with a tricuspid valve Z-score greater than -2 should be directed to a bi-ventricular repair and those with a tricuspid valve Z-score less than -2
should have a univentricular heart palliation as none of the patients with a tricuspid valve Z-score less than -2 achieved a bi-ventricular repair. In this institution it seems reasonable that under this size of tricuspid valve, only palliation with shunting should be offered initially. Patients with the smallest right ventricles are those who are the most at risk of developing right ventricle to coronary artery connections.\(^3\) In case of right ventricle coronary dependence, opening of the right ventricular outflow tract may put the patient at risk of coronary ischemia and should therefore be avoided. By not attempting to open the right ventricular outflow tract of the smallest of these right ventricles, this risk is automatically avoided. Also, in our series only one of the 33 patients identified to have right ventricle to coronary artery connections ultimately reached bi-ventricular repair status, and it seems reasonable to favor univentricular palliation whenever these connections are identified.

Our unit previously actively tried to recruit more patients towards a bi-ventricular pathway by performing right ventricular outflow tract resections in the hope that increasing the capacitance of the right ventricle and allowing forward flow to the pulmonary artery would allow better right ventricular growth with eventual biventricular repair. However, often a cavopulmonary connection between the SVC and right pulmonary artery was required to avoid right ventricular failure and thus leaving the patient with a one and a half ventricle repair. The trend in the last decade is to prefer a definite univentricular strategy as this has a low mortality rate and the one and a half ventricle repair is not actively pursued, but used on occasion as a fall-back option.

This study shows a poor success rate in achieving definitive treatment or significant improvement with initial catheter intervention. Even though the initial failure rate of right ventricular opening was only 24%, many of the patients with initial technical success required further surgery. Eventually only 24% of patients who underwent catheter valvotomy did not undergo an operation in the same hospital stay. However, we did not use radiofrequency or laser valve perforation, both of which have been reported as having a higher technical success rate than balloon valvotomy alone.\(^{45, 46}\) The vast majority of our patients needed a shunting procedure in the neonatal period and as long as an additional form of blood supply requires a surgical intervention, the impact of interventional procedures seems to be
reduced to the exceptional cases of normally sized right ventricles with limited hypertrophy.

As in other studies, smaller tricuspid valve size, the most objective index of right ventricular size, and the presence of right ventricle to coronary artery connections were independent predictors of death. The exact cause of death is difficult to identify retrospectively. It is presumed that the majority of the deaths encountered occurred as a consequence of coronary ischemia. However, most of the deaths seem to have happened at a distance from the initial intervention and it seems unlikely that identifying these coronary lesions before the first surgery would have altered the outcome of these patients. Therefore, the amount of diagnostic catheterization prior to initial surgery has decreased, and its use is often reserved for a later stage.

I expected to find a large proportion of the sixteen deaths to have occurred in patients stratified towards biventricular repair when in retrospect a univentricular approach may have been more appropriate. However, this did not turn out to be the case as only 2/16 had right ventricle decompression and attempt at biventricular repair. 3/16 died before any operation and the remaining 11/16 had a shunt-only as initial management and were considered on the univentricular pathway.

It has recently been shown that inter-stage mortality in shunted patients with pulmonary atresia and intact ventricular septum is comparable to that for Hypoplastic left heart syndrome.47 Miyaji et al. have also shown that performing a bi-directional cavo-pulmonary connection (in patients with an atrial septal defect) increases the oxygen saturation of blood returning to the right ventricle and thus to the coronary arteries in right ventricle dependent coronary circulation.48 Theoretically early bi-directional cavo-pulmonary connection could prevent or delay progression of myocardial ischaemia in those patients reliant on right ventricle to coronary artery flow for myocardial perfusion. Whether this is also the case in patients with right ventricle coronary artery connections without a complete right ventricle dependent circulation is unclear, though it should be suspected, as most long-term survivors of pulmonary atresia with intact ventricular septum with right ventricle to coronary artery connections will have areas of abnormal myocardial perfusion.49 Early bi-directional cavo-pulmonary connection would have the additional advantage of decreasing interim
mortality due to shunt thrombosis and circulatory instability with the shunted state.

In summary, patients with tricuspid valve Z-score less than -2 and with right ventricle to coronary connections are unlikely to reach biventricular repair. It therefore seems reasonable to only perform a shunting procedure as initial intervention in these patients. Coronary ischemia seems to be the predominant cause of death in patients with pulmonary atresia and intact interventricular septum even when placed on a univentricular pathway. However, this usually occurs beyond the initial postoperative period and neonatal diagnostic catheterization does not seem to alter outcome. Early bi-directional cavo-pulmonary connection may help to improve mortality, while interventional catheterisation procedures should be reserved for the cases where no shunting procedure is expected.
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Pulmonary Atresia, VSD and Mapcas: Repair Without Unifocalization

Christian P. Briand,1,2 Matthew Liara,1,2 and Yves d’Udekem1,2

The last three decades have seen considerable developments in the management of infarcts with pulmonary atresia, ventricular septal defect, and total anomalous pulmonary venous return. The technical difficulty of surgical treatment lies in the balancing of a pulmonary vascular bed capable of providing suitable low-right ventricular reserve for repair of the intracardiac defect. With the development of improved and complete unification of pulmonary arterial system and venous drainage, we have developed an alternative approach to ‘repair without unifocalization.’ This has been used successfully in the treatment of a number of patients with pulmonary atresia and VSD.

The results of this approach have been promising, with a high rate of successful repair and low morbidity and mortality. The most significant advantage of this approach is the ability to preserve the normal pulmonary arterial system, which can lead to improved long-term outcomes.

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

We believe that the natural evolution of most mapcas is toward the development of the normal pulmonary arteries. However, in cases where the arteries are small or absent, the risk of irreversible pulmonary vasoconstriction may be higher. In these cases, early intervention may be necessary to prevent irreversible pulmonary vasoconstriction.

A Significant Number of Patients With VSD and Mapcas Have to Central PA

In our experience, the presence of a central PA is almost always associated with a significant increase in pulmonary blood flow. This can lead to significant morbidity and mortality if not addressed. Therefore, careful consideration of the anatomy and pulmonary blood flow is necessary to determine the appropriate treatment strategy.

Very Diminutive Pulmonary Arteries do not Grow

In our experience, central pulmonary arteries are small, even in the most severe cases of pulmonary hypertension. Growth of these arteries can be significant, and it is essential to develop a comprehensive approach to their management.

Most of the Very Diminutive Pulmonary Arteries Have an Incomplete PA Arterization

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 angiographic, regardless of how small they are and regardless of the number of branches. This approach has led to improved outcomes and a decrease in the need for repeat procedures.
Pulmonary Atresia, Ventricular Septal Defect, and Major Aortopulmonary Collaterals: Neonatal Pulmonary Artery Rehabilitation Without Unifocalization

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Background. This study analyzed a protocol of neonatal rehabilitation of hypoplastic pulmonary arteries in the management of pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries (MAPCAs). Ide management of patients with pulmonary atresia, VSD, and MAPCAs is the subject of controversy.

Methods. From June 2000 to December 2008, 15 consecutive patients were diagnosed with pulmonary atresia, VSD, and MAPCAs, and 20 were entered into a neonatal correction program. The median age at the first operation was 3.45 weeks (range, 0.7 to 17 weeks). All patients underwent initial central or modified Blalock-Taussig shunts, or both. Further preparatory procedures included transcutaneous aortopulmonary resection and balloon dilation of VSD, MAPCA stenosis, 6, MAPCA ligations, and 6 further shunts. No patient underwent transection of the esophageal arteries.

Results. Of the 20 patients, 15 were male and 5 were female. The median age was 3.45 weeks (range, 0.7 to 17 weeks). All patients were alive and had survived at least 1 year after the initial operation, and 13 had survived at least 1 year after the last operation.

Conclusions. Rehabilitation of hypoplastic native pulmonary arteries by a nonshunting technique, without MAPCA intervention, for pulmonary atresia, VSD, and MAPCAs, provides encouraging results with excellent early survival.

Gong Xia, Tianjun Su, and Zijian Su
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AADAS ET AL. Native PA Repair for VSD, PA, and MAPCA

Pulmonary Atresia, Ventricular Septal Defect, and Major Aortopulmonary Collaterals: Neonatal Pulmonary Artery Rehabilitation Without Unifocalization
diagnosed with PA, VSD, and MAPCAs admitted from June 2000 to December 2008 were reviewed. 15 identified 20 patients. Twenty consecutive attempts were included an early starting protocol aiming to rehabilitate native PA without transection of collateral arteries and continue the course of this study. Patient characteristics are reported in Table 1. Five patients were managed using alternate techniques:

- One patient, referred at the age of 4 years, had a normal left PA perfused by a ductus, previously stented at birth, and an extremely hypoplastic right PA with the right lung perfused by 3 bronchial arteries. He underwent a right ventricle to PA conduit, and is waiting for repair aiming at detaching the RV blood flow at 1 year.
- The second patient, born with normally sized PAs supplied by 1 collateral vessel arising from the descending aorta, underwent a complete repair at the age of 0.4 years.
- The pulmonary circulation in the third patient depended on 2 major collateral vessels being normally sized and contributing to PA branches. He underwent an on-stage repair and unifocalization with a Contegra (Medtronic, Inc., Minneapolis, MN) valved conduit used in the RV outflow tract.
- A fourth patient, with splanchnic syndrome, was diagnosed at the age of 3 months. He underwent an RV-PA conduit but died 6 months later of liver failure.
- The last patient underwent staged rehabilitation of native PA stenting at the age of 10 years, with the first procedure performed by complete repair when he was 15 years old. These patients were calculated for the 17-18 patients whose measurements were obtained after corrective repair was obtained to determine right and left ventricular pressures. The following formulas were used:

Body surface area (BSA) = \(\sqrt{\text{Height in m} \times \text{Weight in kg} / 2.304}\)

Pulmonary artery index (PAI) = right PA + left PA cross-sectional area in mm\(^2\) / BSA in m\(^2\)

Surgical Strategy
All neonates were planned for elective correction with the fair blood flow through the right lung regardless of their oxygen saturations. The technique of central shunt (initially described by Graass and colleagues) was our preferred approach. The initial opening was made at the level of the insertion of the PAs and extended down to the main PA. A Gore-Tex shunt (W. L. Gore and Associates, Flagstaff, AZ) was anastomosed to this opening end-to-side with 9/0 Prolene sutures (Ethicon, Somerville, NJ). The proximal anastomosis consisted of a side-to-side anastomosis on the ascending aorta.

The patients subsequently underwent further shunting procedures or RV-to-PA artery conduit implantation until the last had evidence that the shunt had failed, and treatment had been made with a PDA ligature.

Statistical analysis
Data are reported as median (range) or mean (standard deviation), as appropriate.

Results
Twenty patients with a diagnosis of pulmonary atresia, VSD, and MAPCAs underwent an initial neonatal shunting procedure. At the time of last follow-up, the median age of patients was 36.4 months (range, 5 to 79 months) with no deaths.

Clinical Outcome
PROCEDURE 1. The median age at the first procedure was 3.6 weeks (range, 0.7 to 17 weeks), and most progressive oxygen saturations were 85% (range, 72% to 95%). One patient's first operation was delayed until age 17 weeks because of a bleeding systemic failure. All other patients underwent an initial shunting procedure before the age of 3 months. Most patients were discharged home 2 weeks, regardless of oxygen saturation level. Initial shunting consisted of 10 central shunts only (n = 2), 3, 35 mm, n = 1, 4 mm, n = 2, and received a right modified Blalock-Taussig shunt (2 mm, n = 2, 3 mm, n = 1, 4 mm, n = 2, and received a right modified Blalock-Taussig shunt (3 mm, n = 1, 4 mm, n = 1, 5 mm, n = 1, and 1 patient had a central and left modified Blalock-Taussig shunt (3 mm, n = 1). Concurrent PA resection or completion of extra-anatomic repairs were performed in 22 patients (55%).

PROCEDURE 2. Nine patients have undergone a second procedure at a median age of 7.5 months range, 3.6 to 3.6 months. Complete PA stenosis was achieved in 11 patients (71%), and a 1.5-2.5 mm shunt was inserted in 15 (56%) using 4-6 mm Gore-Tex grafts, or 12-mm Contegra (Medtronic, Inc., Minneapolis, MN) valved conduits. Three of 10 patients (9%) had a second surgical intervention (5.5-months range, 3.6 to 3.6 months). Complete PA stenosis was achieved in 11 patients (71%), and a 1.5-2.5 mm shunt was inserted in 15 (56%) using 4-6 mm Gore-Tex grafts, or 12-mm Contegra (Medtronic, Inc., Minneapolis, MN) valved conduits. Three of 10 patients (9%) had a second surgical intervention (5.5-months range, 3.6 to 3.6 months). Complete PA stenosis was achieved in 11 patients (71%), and a 1.5-2.5 mm shunt was inserted in 15 (56%) using 4-6 mm Gore-Tex grafts, or 12-mm Contegra (Medtronic, Inc., Minneapolis, MN) valved conduits. Three of 10 patients (9%) had a second surgical intervention (5.5-months range, 3.6 to 3.6 months). Complete PA stenosis was achieved in 11 patients (71%), and a 1.5-2.5 mm shunt was inserted in 15 (56%) using 4-6 mm Gore-Tex grafts, or 12-mm Contegra (Medtronic, Inc., Minneapolis, MN) valved conduits. Three of 10 patients (9%) had a second surgical intervention (5.5-months range, 3.6 to 3.6 months). Complete PA stenosis was achieved in 11 patients (71%), and a 1.5-2.5 mm shunt was inserted in 15 (56%) using 4-6 mm Gore-Tex grafts, or 12-mm Contegra (Medtronic, Inc., Minneapolis, MN) valved conduits.
Changing trends in the management of pulmonary atresia with intact ventricular septum: the Melbourne experience

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Abstract

Objective: Management of pulmonary atresia with intact ventricular septum (PA-IVS) can be directed to either biventricular repair or univentricular repair. The optimal management strategy has yet to be defined. In this study, all patients operated at the Royal Children’s Hospital, Melbourne, from 1992 to 2013, were reviewed. Patients were retrospectively stratified into a single ventricle group (IVS-IVS) or a biventricular group (IVS-BV), and the outcomes were compared.

Methods: Retrospective analysis of 85 patients with PA-IVS operated on at the Royal Children’s Hospital. Details included the indication for surgery, operations, complications, and outcomes. The univentricular group included patients who underwent an univentricular repair (Fontan orujaria circuit). The biventricular group included patients who underwent either a biventricular repair or a Rastelli operation.

Results: The distribution of PA-IVS was normal in 11 (13%), moderate hypoplasia in 40 (47%), and severe hypoplasia in 34 (40%). Patients in the univentricular group were present in 35 (41%) and BV surgery was present in 50 (59%). Median age at surgery was 3 months (range, 2 days–18 years).[1] The incidence of the venous connection was aorta in 7 (9%) and in 78 (91%). The incidence of the pulmonary connection was superior vena cava in 6 (8%) and in 79 (91%). The incidence of the atrial connection was an atroventricular connection in 1 (1%) and in 84 (99%).

Conclusions: A clear distinction between fans and BV surgery was present in the univentricular group. In the biventricular group, the BV group was more frequently present. The results of the authors’ experience that a single ventricle repair is superior to biventricular repair for patients with less severe IVS and severe IVS, respectively.

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1. Introduction

The past decade has seen a marked improvement in the surgical outcomes of patients with pulmonary atresia with intact ventricular septum (PA-IVS) [1–13]. The prospective multicenter study by the Congenital Heart Surgeons Society predicts a perioperative survival of 75% for these enrolled in 1995, compared with 50% survival in 1989 [4]. This improvement has occurred despite profound differences in the approaches advocated by the different institutions involved. Initial centers reporting success with the treatment of the condition adopted a single approach: forming a Fontan pathway, others a univentricular pathway. It is now becoming clear that a strategy successfully forming a univentricular pathway will derive some patients with relatively normal right ventricular function from the benefits of a biventricular repair and the poorer survival associated with a Fontan operation. On the other hand, a strategy pushing for biventricular repair in all cases will result in increased mortality [2]. Today, a more balanced approach appears preferable, and preliminary success has been reported [1]. Unfortunately, there are yet no clear criteria to decide which patients should be oriented to a univentricular or biventricular repair.

In Melbourne, our strategy has gradually evolved over the years. Initially, we adopted a policy aimed at growing the patients’ single ventricle and prepared indications for a biventricular repair. Our decision to attempt biventricular repair was based on the presence or absence of a right ventricular outflow.[1] These patients had a patient-innominate conduit toward a biventricular repair, sometimes resulting in a “right ventricle (RV)-switched” procedure consisting of revision of extracardiac conduit[1] or intracardiac repair; others without an extracardiac conduit were placed on the univentricular pathway. In these patients with only low risk associated, the strategy was classic as having had an IVS repair.

Moderately reduced right ventricular size, this approach allowed us to delay the final decision between a univentricular or biventricular repair until 15 years. We have progressively stepped back from this methodology and adopted a more balanced approach. Patients with the smallest RV or tricuspid valve (TV) and coronary anomalies were directed toward our univentricular pathway. In the remaining patients, the right ventricular outflow tract was opened surgically or with a catheter intervention and staged for biventricular repair. Up to now, we have focused on achieving a univentricular or biventricular repair was made after the review of each individual case, without any specific guidelines.

We therefore review our experience of patients presenting with pulmonary atresia and intact ventricular septum to determine risk factors for mortality, hoping that the identification of these risk factors may guide decision making toward either a univentricular or a biventricular pathway.

2. Methods

The study was approved by the Royal Children’s Hospital Ethics Committee. All patients at the Royal Children’s Hospital admitted between 1992 and 2013 with the diagnosis of pulmonary atresia were included. Patients were identified from the hospital database (n = 96). Patients with extracardiac anomalies or severe congenital heart disease with associated pulmonary atresia were then excluded from the study (n = 13), and the remaining 83 patients constituted the core of this study. All preoperative echocardiographic examinations were reviewed by a single cardiologist blinded to the outcome to assess RV size. Right coronary artery connections and TV annulus diameter. RV size was subjectively categorized into three groups: normal, moderate, and severely hypoplastic. RV size was based on the size of the muscular RV in comparison with the left ventricle, the percentage of the RV, and the extent to which muscular hypertrophy exceeded the RV cavity.

Echocardiography was reviewed for all patients with RV size liver coronary artery connections to determine whether these patients would benefit from extracardiac conduit. Echocardiography was centred on the presence of RV connections to the coronary arteries and whether other sources of flow to the coronary arteries were affected by the presence of RV connections. RV connections included either two sources of flow to the coronary arteries or idiopathic coronary artery anomalies.

The following variables were analyzed: birth weight, gestational age at birth, birth weight, age at first intervention, TV size, presence of extracardiac connections, RICD, coronary artery connections, RV size, RV aneurysm, coronary artery connections, and RV size. The intervention was considered successful if the patient had a normal, mild, or severe coronary artery anomaly.

3. Results

A univentricular pathway was usually performed in patients with severe RV size, moderate RV size, and normal RV size, respectively. A biventricular repair was performed in patients with normal RV size who had a single ventricle. The biventricular repair was performed in patients with moderate RV size who had a single ventricle. The biventricular repair was performed in patients with severe RV size who had a single ventricle. The biventricular repair was performed in patients with severe RV size who had a single ventricle.
Author/s: LIAVA'A, MATTHEW

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