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Outcomes of patients with pulmonary atresia and major aortopulmonary collaterals without intervention in infancy

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Abstract

Introduction—Treatment of pulmonary atresia with major aortopulmonary collaterals (PA MAPCA's) remains a challenge. Despite variations in surgical technique, contemporary strategies all include initial intervention in the first year of life. However, a subset of patients presents later in life, and contemporary outcomes of this group have not been reported previously.

Methods—We performed a retrospective case series of consecutive cases of PA MAPCA's who did not undergo surgery before the age of 1 year from 1/2001 and 2/2016. We describe their presenting characteristics, operative and transcatheter interventions, and outcomes.

Results—A total of 8 cases were identified from 76 children with PA MAPCA's treated over the study period. Median age at presentation was 5.9 years. 75% had confluent pulmonary arteries with a median Nakata index of 113 mm²/m². Operative intervention was performed in 5/6 cases. Two are awaiting intervention. The combination of operative and transcatheter interventions allowed for ventricular septal defect closure in 60% of cases, all of whom had sub-systemic right ventricular pressures.

Conclusion—Operative intervention is possible in some older cases with PA and MAPCA's. Though multiple operations and transcatheter therapies are necessary some can achieve operative correction of serial circulation with tolerable physiology. Subjects with ventricular hypoplasia and those without confluent pulmonary arteries are more challenging.

Keywords

pediatric cardiology; congenital heart surgery; heart catheterization; outcomes research

Introduction

The treatment of pulmonary atresia and major aortopulmonary collaterals (PA MAPCA's) is complicated by the heterogeneity of the presenting anatomy and physiology. Historically, the

prognoses was dismal, with early series estimating 20% survival at 30 years[1]. Dramatic improvement has been demonstrated in survival despite controversy regarding the optimal operative strategy[2–17].

The goal of operative correction is to recapitulate normal serial circulation, namely by 1) reconstituting the pulmonary arterial tree, 2) reconnecting the lungs to the right ventricle (RV), and 3) closure of the ventricular septal defect (VSD). An important goal during this process is maximizing the capacity of the pulmonary vascular bed and minimizing RV afterload by 1) maximizing the number of lung segments perfused, 2) minimizing obstruction in the pulmonary tree, and 3) avoiding development of pulmonary vascular disease (globally or in individual lung segments). Debate about the relative merits of different strategies for operative correction, namely single-stage versus multi-stage procedures and unifocalization of collaterals persists[2–5, 7, 8, 16, 18–20]. One aspect of commonality between current strategies is intervention in infancy. Early series from the 1970's to early 1990's reported the outcomes of intervention on older patients[21–24]. Contemporary series universally advocate an initial operative intervention in infancy, with the goals of achieving a more stable source of pulmonary blood flow (Q_p), avoiding development of heart failure, pulmonary vascular disease, and/or loss/occlusion of MAPCA segments.

However, the anatomy in some children with PA MAPCA's allows them to grow for longer periods of time without development of heart failure, pulmonary hypertension, or cyanosis. If access to cardiologists and surgeons is limited, some of these children are not intervened upon and survive to present later in life. A complex question is whether intervention at this stage will increase length and quality of life in these patients.

We sought to describe the experience at our center of subjects with PA MAPCA who did not undergo intervention until after 1 year of age, describing their presenting characteristics, interventions, and outcomes and, in an exploratory analysis, assessing potential risk factors for adverse outcome in this population.

Methods

Study population

A retrospective case series of consecutive children and adults with pulmonary atresia and major aortopulmonary collaterals between 1/1/2001 and 1/1/2016 treated at Children's National Medical Center (Washington, D.C.) was performed. The study was approved by our hospital's institutional review board. Subjects of either sex were included if they had a diagnosis of PA MAPCA's (including both tetralogy of Fallot with pulmonary atresia and other anatomic variants including those with single ventricles), and did not undergo operative intervention until after 1 year of age. After the fact, it was recognized that subjects who underwent intervention prior to 2001 represented a different era of treatment, and these were excluded from analysis. Cases were identified by direct query of center cardiac surgical, echocardiography, and catheterization databases. Eligibility was confirmed through review of electronic medical record.

Study measures

Data collected included demographics, medical history, operative and transcatheter procedures, and data from echocardiograms and catheterization procedures. Echocardiograms reviewed included the first study available and most recent study, and were used to confirm cardiac anatomy and estimation of ventricular function. Angiograms from all catheterizations were reviewed by one member of the study team (MLO). Data collected included the presence of confluent pulmonary arteries and measure pulmonary artery diameters, Nakata index (mm^2/m^2), number of segments perfused by collaterals, presence and severity of pulmonary vascular disease, and number of pulmonary segments with pulmonary vascular disease. Subjects were grouped into categories by their Nakata index (no confluent PA's, diminutive PA's (Nakata index $<90\text{mm}^2/\text{m}^2$), small PA's (Nakata index: $90\text{--}200\text{mm}^2/\text{m}^2$), moderate PA's (Nakata index: $200\text{--}300\text{mm}^2/\text{m}^2$), and normal diameter PA's (Nakata index $>300\text{mm}^2/\text{m}^2$) as described previously [2, 8, 25]. Oximetry and pressure data from all catheterizations was reviewed; the primary outcome of interest was ratio of RV to systemic arterial pressure in subjects after VSD closure. Clinical outcomes of interest (death, closure of ventricular septal defect (VSD), and subsequent transcatheter/operative interventions) were recorded along with their timing.

Statistical analysis

The primary aim of the study was descriptive. Standard descriptive statistics were calculated and expressed as mean $\pm$ standard deviation, median (range and interquartile range (IQR)), and percent (count) as appropriate. The primary objective of the study was descriptive. All analyses were performed using STATA MP v.13 (Statacorp, College Station, TX USA).

Results

Study Population

Between 1/1/2001 and 1/1/2016, 76 children with PA MAPCA's were treated at our center (Figure 1). Of these, eight underwent their initial therapy prior to 2001, and 3 died or were lost to follow-up in infancy prior to intervention. Of the remaining 65 children, 89% ($n=57$) underwent operative intervention in infancy (median age: 41 days, IQR: 7–91 days) and were excluded from analysis.

A total of 8 children had not undergone an intervention by 1 year of age. Median age at presentation to our institution was 5.9 years (range: 146 days to 13.8 years) (Table 1). None were born in the United States (3 born in El Salvador, 2 each from People's Republic of China and Kuwait, with 1 born in Ghana). Four of eight were female. None were diagnosed with unifying genetic syndromes, though two had significant dysmorphic features in addition to their cardiac disease (severe bronchomalacia in Case 1 and developmental delay, severe scoliosis, concomitant pulmonary hypoplasia, and dysmorphic ears and facial features in Case 7). There was evidence of growth failure (height and weight below the 5th percentile for sex and age) in 38% of the subjects. Six of eight subjects had tetralogy of Fallot with pulmonary atresia. The cardiac diagnoses for the other two cases were unbalanced complete common atrioventricular canal unbalanced to the right with PA (Case 6) and double inlet left

ventricle, PA {S,D,D} (Case 7). Median saturation was 81% (range: 76–86%) with median hemoglobin of 16.5 g/dl (range: 12.0–22.4).

On initial cardiac catheterization, the median ratio of pulmonary to systemic blood flow (Qp:Qs) was 2.2:1 (range: of 0.4–2.9:1). Pulmonary artery anatomy was variable. Three subjects did not have confluent central pulmonary arteries. Of those with confluent pulmonary arteries, all had smaller than normal pulmonary arteries (Nakata index $<300\text{mm}^2/\text{m}^2$), but they were distributed between those with diminutive confluent arteries (Figure 2A) and relatively large diameter arteries (Figure 2B). The median Nakata index was $113\text{mm}^2/\text{m}^2$ (range: 30–260 mm^2/m^2).

Intervention and Outcomes

The decision was made to not pursue operative interventions in 2 cases (Table 2). One (case #1) had a cardiac arrest during overseas transport prior to evaluation. She reportedly had severe airway hypoplasia/malacia. Subsequently, hypoxic ischemic encephalopathy and cystic lung disease resulting in chronic respiratory failure developed. During initial hospitalization, the patient underwent placement of tracheostomy and gastrostomy tube and subsequent resection of a necrotic left lower lobe. She was re-evaluated at 18 months of age, and catheterization demonstrated that there were no confluent pulmonary arteries, loss of 6 pulmonary segments after lobectomy, and severe pulmonary vascular disease in 3 of the remaining segments. She returned to Kuwait and has not been seen at our institution since discharge. The second case (#7), an 8 year old girl with double inlet left ventricle, PA with RV to aorta {S,D,D} presented after immigrating from El Salvador. Her catheterization demonstrated that she did not have confluent pulmonary arteries, that there was no perfusion to her right lung, and that the Qp:Qs was 0.4:1 and severe polycythemia (hemoglobin of 22.4 g/dl). She had 2 major collaterals one of which provided restricted pulmonary blood flow to much of her left lung. Her data was reviewed at our institution's surgical conference, but with single ventricle physiology, absent pulmonary arteries, and limited pulmonary vascular tree, she was not considered a candidate for operative intervention. Stent angioplasty of the collateral to her left lung is being considered.

In addition, one subject (Case 8) is awaiting what is expected to be a complete repair, with delay due to parental preference.

In the remaining 5 cases, operative intervention was performed. Three patients (case #3, 4, and 5) underwent a single stage operative correction with placement of an RV to PA conduit and VSD closure. In all three cases, the decision to close the VSD was made intra-operatively by assessing RV pressure during discontinuation of cardiopulmonary bypass. In two cases with no confluent pulmonary arteries a multi-stage approach was used. In case #2 there were no confluent PA's and 4 MAPCA supplying the lung fields, over the course of a week collaterals were unifocalized with femoral vein homograft via thoractomies in separate operations followed quickly by reconstruction of the RVOT with a conduit and closure of the VSD. Connections between the aorta and MAPCA were left intact until coil occluded 2 months after the initial operation. In case #6, the degree of RV hypoplasia was judged prohibitive to support a biventricular circulation. A central shunt was placed from ascending aorta to the stump of the main PA. He has had significant improvement in his PA size (initial

Nakata index was $67 \text{ mm}^2/\text{m}^2$ and after 1 year had increased to $96 \text{ mm}^2/\text{m}^2$) and is being followed expectantly with hopes that with sufficient PA growth single ventricle palliation can be performed.

Median follow-up after operation was 4.8 years (range: 270 days–7.2 years) representing 18.9 years of total post-operative follow-up. There were no deaths in the hospitalization for initial operation. One patient experienced low cardiac output and a cardiac arrest attributed to RV failure, which was treated with veno-arterial extracorporeal membrane oxygenation (VA-ECMO), transcatheter creation of an atrial communication, and eventually operative fenestration of the VSD patch. This patient recovered and continues to do well. The only death in the series occurred in case #5. After her initial operation, transthoracic echocardiography demonstrated very elevated RV pressure and RV systolic dysfunction. Catheterization four months after her initial operation demonstrated suprasystemic RV pressure. Angiography demonstrated loss of antegrade perfusion to the much of the right lung (which was still supplied by a MAPCA) and severe pulmonary vascular disease to the segments attached to the RV (all of the left lung and several segments of the right lung). She was taken back to the operating room for an attempt at unifocalization of the right lung collateral but was unstable after being positioned for the operation and during the initial dissection. She had a cardiac arrest, and VA-ECMO was initiated. She expired eight days later. These two episodes represent the only episodes of mechanical circulator support ($n=2$, 33% of operations 95% CI: 4–77%) and repeat operation ($n=2$, 0.1 per person-years) in the series. In contrast, 16 cardiac catheterizations were performed (0.7 per person-year) following operative intervention. Interventions were performed in 11 of these catheterizations, almost all of which (9/11) included balloon or stent angioplasty of unifocalized collaterals or pulmonary artery stenosis. Other interventions were coil occlusion of collaterals (3 cases), transcatheter creation of an atrial communication ($n=1$), and transcatheter pulmonary valve replacement ($n=1$). Ultimately, the other three remaining cases all had VSD closure and subsystemic RV pressure (range: 40–75% systemic arterial pressures). There were no attributable adverse events during or after these catheterizations. Nakata index increased in all cases after initial operation. The pattern of Nakata index over serial angiographic assessments is shown in Figure 3.

Discussion

This retrospective case series demonstrates that palliation/repair of some children with PA MAPCA's is possible. There was low perioperative mortality for the initial operation. However, there was significant morbidity and frequent re-intervention was undertaken to maintain tolerable right ventricular pressure. The small size of the study population limits statistical inference, but it is clear that not all patients are equally suitable to treatment (including a minority in whom operative correction was not attempted). Those that have survived intervention did not have severe pulmonary vascular disease and had confluent pulmonary arteries. Ventricular hypoplasia, pulmonary vascular disease, loss of lung segments, and absent intra-pericardial pulmonary arteries represent challenges for surgical palliation.

An increasing proportion of patients with congenital heart disease presenting for treatment in the United States are diagnosed prenatally[26], and by extension likely that this is also true in other industrialized countries. Proponents hope that this allows for efficient referral of fetuses with complicated congenital heart disease to centers of excellence and allows for appropriate stabilization and treatment of these patients in the most stable condition at the optimal time. However, these resources are not universally available. In regions without widespread specialty pediatrics, prenatal and even antenatal diagnosis is not a given. In patients with PA MAPCA's, studies of natural history predating surgical intervention demonstrate that a subset of patients can survive without intervention to a more advanced age[1]. The relative risk of this population relative to infants with similar anatomy is not well understood. These patients are at increased risk of poor nutrition and congestive heart failure, pulmonary vascular disease, and the sequelae of prolonged cyanosis. Alternatively, having survived without medical attention, may represent a protective. The truth of course is more nuanced, and what is clear even in this limited series is that the population at risk is diverse in terms of presenting anatomy and severity of illness.

Though lumped into a single diagnosis, PA MAPCA's includes a heterogeneous set of anatomic and physiologic characteristics. Most commonly the condition is associated with an anterior malalignment VSD (tetralogy of Fallot with pulmonary atresia), but variants with other intracardiac anatomy occur. Several of these factors have previously been described as having significantly worse prognosis for infant operations and subsequent survival, including single ventricle physiology,[27] pulmonary vein anomalies[28], and 22q11.2 microdeletion syndrome [29]. There is controversy over whether absent or extremely diminutive central PA's increase risk or not[11, 13, 15, 30, 31]. Given that the ultimate goal is to reduce RV afterload and RV pressure, loss of segmental pulmonary artery branches and development of pulmonary vascular disease globally or in segments would seem to be risk factors for adverse outcome, but these to our knowledge have not been addressed in previous studies. In this series, the interventions for subjects with absent or diminutive pulmonary arteries were numerous, but there is insufficient sample size to investigate this stringently.

Surgical technique must be adapted to the potentially heterogeneous presenting anatomy. The pulmonary arteries range from absent to almost normal and though collateral vessels tend to occur in similar locations and patterns, determining how to reconstruct the pulmonary arterial tree must be adapted to the patient, which is demonstrated in the current series. As described previously[32], unifocalization using femoral vein homograft enabled the surgeon in case 2 to connect a total of 4 collaterals into a pulmonary tree to receive a conduit. In other cases, mobilizing the collaterals was more feasible and direct unifocalization was performed. Finally, in some cases redundant collaterals or collaterals that were judged to be too remote for unifocalization were ligated or occluded with coils.

Repeat operations were not common, though we acknowledge that operative replacement of smaller conduits is inevitable. Serial transcatheter interventions were performed in all patients who underwent operation and unifocalization with the goal of maintaining sub-systemic RV pressure. The vast majority underwent balloon or stent angioplasty of the connection of collaterals to the pulmonary arteries. There is insufficient data in this series to make any conclusions about longitudinal growth in the pulmonary arteries after intervention,

but subsystemic RV pressure was achievable in all of the cases in which repair was performed, and qualitatively there is improvement in the size of the PA's over time. Nakata index plateaued in the cases with longest follow-up but this was due to increasing patient size relative to PA diameter, rather than decrease in PA size. This is a limitation of the Nakata index as a longitudinal measure of PA growth. There were no complications associated with these catheterization procedures in our series, but with appropriate humility we acknowledge that angioplasty in stenotic pulmonary arteries (especially unifocalized MAPCA's) is always associated with risk of vascular injury, which can result in stenosis or occlusion of the vessel, frank bleeding, or pseudoaneurysm formation[2, 33, 34]. With this regimen, it is notable that even in this relatively high risk group, all of the patients have been able to have their VSD closed and 3 of 4 have been able to maintain subsystemic RV pressure with serial transcatheter intervention with very little repeat open heart surgery (one in four patients).

Ventricular hypoplasia that results in a ventricle that cannot support biventricular circulation is problematic. The neo-portal Fontan circulation depends (even more than serial circulation) on limited resistance for pulmonary blood flow. The results of surgical palliation for this disease is dismal even if intervention occurs with conventional timing[27]. In our series, one such patient had loss of ½ of her pulmonary segments prior to presentation and was clearly not a candidate for palliative surgery. The other had diminutive pulmonary arteries, but has demonstrated significant growth after central shunt placement. It remains to be seen whether this growth will be sufficient to perform the next step in staged palliation. These patients remain a significant challenge, without obvious solution.

None of the cases in this series were treated with pulmonary vasodilator pharmacotherapy. The BREATHE-5 study demonstrated improved hemodynamics and exercise capacity in adults with Eisenmenger syndrome.[35, 36] Whether this finding was applicable to patients with heterogeneous pulmonary vascular disease (some lung segments without disease because they are protected by proximal stenosis while others suffer from vascular disease) was not addressed in this study. Recent case series have demonstrated improvement in symptoms and hemodynamics in patients with PA MAPCA's and pulmonary hypertension treated with either sildenafil[37, 38] or bosentan[37, 39, 40]. It remains to be seen whether this benefit will be reproducible in larger series and whether it is generalizable to all patients with peripheral/segmental pulmonary artery stenosis and RV hypertension.

There are several significant limitations to this study. It was a retrospective analysis describing the outcomes of the contemporary approach at our center. It is not possible to extrapolate how these results would change in other settings or with different approaches. Two of the cases reside outside the United States which limits their follow-up. The small sample size limits the precision of our estimates of event rates (reflected in wide confidence intervals) and makes adjustment for confounding variables and statistical inference prohibitive.

Conclusion

Despite these limitations we conclude that this series demonstrates that operative correction of PA MAPCA's can be performed later in life. The combination of a successful initial operation and persistent transcatheter interventions can result in a serial circulation with tolerable hemodynamics, but there is significant risk of morbidity and mortality.

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Figure 1.
Study population

Figure 2. Representative angiograms of pulmonary arteries

A) Hand injection angiogram through retrograde end-hole catheters demonstrating diminutive confluent pulmonary arteries with a Nakata index of $42 \text{ mm}^2/\text{m}^2$.

B) Power injection angiogram through a retrograde NIH catheter demonstrating relatively large confluent pulmonary arteries (Nakata index of $260 \text{ mm}^2/\text{m}^2$).

Figure 3. Nakata index over serial angiographic assessment

The Nakata index of cross-sectional pulmonary artery area (normalized to body surface area) was measured in subjects at each catheterization. It is plotted against time relative to the first operative intervention.

Table 1

Study population

n	8
Female sex n (%)	50% (4)
Country of origin n (%)	
El Salvador	3
Ghana	1
Kuwait	2
People's Republic of China	2
Age at presentation	5.9 years (range 146 days–12 years)
Height	93.7 cm (range: 59–145)
Height percentile for age and sex	9.7% (range: 0–66%)
Below 5 th percentile for height	38% (3)
Weight	15.1 kg (range: 6.2–21.3)
Weight percentile for age and sex	13.9% (range: 0–54)
Below 5 th percentile for weight	38% (3)
Body mass index	14.2 (range: 13.4–17.5)
Saturation	81% (range: 76–86)
Hemoglobin	16.5 g/dl (range: 12–22.4)
Hematocrit	51% (range: 38–62)
Qp:Qs	2.2 (range: 0.4–2.9)
Nakata index (n=5)	113 mm ² /m ² (range: 30–260)
No confluent PA	25% (2)
Diminutive PA (Nakata index <90 mm ² /m ²)	38% (3)
Small PA (Nakata index: 90–200 mm ² /m ²)	13% (1)
Intermediate PA (Nakata index 200–300 mm ² /m ²)	25% (2)
Normal PA (Nakata index ≥300 mm ² /m ²)	0% (0)
Pulmonary vascular disease	63% (5)

Abbreviations: Qp:Qs ratio of pulmonary to systemic blood flow, PA pulmonary artery

Table 2

Summary of individual cases

Case	Presentation	Height %	Weight %	Hgb (g/dl)	O2 Sat	Qp:Qs	PA anatomy	Nakata index (mm ² /m ²)	PVD	Interventions	Follow-up
1	3 month old girl with TET PA MAPCA from Kuwait	<1	<1	12	81	2.5	No confluent PA's 3 MAPCA Loss of 6 segments	n/a	Severe 3 segments	No interventions	20 months of age n/a
2	4 month old boy with TET PA MAPCA from Kuwait, initial intervention delayed because of diminutive PA's and restrictive collateral connections	1.4	<1	14	85	2.1	Confluent PA's 4 MAPCA	30	None	<p>1</p> <p>23 months old: 3-stage operative correction with i. left unifocalization, ii. right unifocalization, and iii. RV to PA conduit with VSD closure, and creation of an ASD</p> <p>2</p> <p>25 months old: Stent angioplasty of unifocalized collateral to RUL and coil embolization of the residual collateral connections</p> <p>3</p> <p>5 years old: Balloon angioplasty of multiple vessels at anastomosis of unifocalized collaterals</p> <p>At end of last catheterization: RV pressure is 75% of systemic pressure and mild PVD in left lung</p>	5 years of age 3 years post-op
3	4 year old boy with TET PA MAPCA immigrated	9.7	46	17.3	81	1.9	Confluent PA's 3 MAPCA (2 with dual supply)	211	Mild 3 segments	<p>1</p> <p>4 years of age: Operative repair with 16 mm aortic homograft RV</p>	13 years of age 9 years post-op

Case	Presentation	Height %	Weight %	Hgb (g/dl)	O2 Sat	Qp:Qs	PA anatomy	Nakata index (mm ² /m ²)	PVD	Interventions	Follow-up
										<p>to PA conduit and VSD closure (with pre- and post-operative coil embolization of collaterals)</p> <p>2 9 years of age: Balloon and stent angioplasty of RV to PA conduit</p> <p>3 9 years of age: Subsequent catheterization to deploy 18mm Melody transcatheter pulmonary valve</p> <p>At end of last catheterization: Mild gradient across transcatheter pulmonary valve, RV pressure is 40% systemic, no VSD, and no PVD</p>	
4	14 month old girl with TET PA MAPCA diagnosed in Ghana	<1	1.4	16.1	81	2.0	Confluent PA's 4 MAPCA (2 with dual supply)	42	Mild 1 segment	<p>1 14 months: pre-op closure of dual supply collaterals with immediate operative repair with 12 mm homograft RV to PA conduit; VSD closure; ECMO on POD#2 attributed to RV failure; Transcatheter creation of atrial communication with cutting balloon and coil occlusion redundant collaterals; Operative fenestration of</p> <p>8 years of age 7 years post-op</p>	

Case	Presentation	Height %	Weight %	Hgb (g/dl)	O2 Sat	Qp:Qs	PA anatomy	Nakata index (mm ² /m ²)	PVD	Interventions	Follow-up
										<p>VSD patch on POD 15</p> <p>2</p> <p>Multiple rounds of balloon angioplasty (19 months, and 2, 3, 4 and 7 years of age)</p> <p>At last catheterization: mild conduit gradient, VSD open but no significant shunt, RV pressure is 66% systemic.</p>	
5	12 year old girl with TET PA MAPCA recently immigrated from El Salvador	30	6.7	16.9	86	2.9	Confluent PA's 5 MAPCA (1 dual supply)	158	Mild 5 segments	<p>1</p> <p>12 years: pre-op closure of dual supply</p> <p>2</p> <p>Evidence of RV failure and elevated RV pressure, diagnostic catheterization demonstrated severe PVD to the left lung and that most of the right lung was not receiving antegrade pulmonary blood flow</p> <p>3</p> <p>13 years: attempt at unifocalization through right thoracotomy. Positioning patient for operation and initial dissection resulted in hypotension. Operation aborted but patient arrested and was placed on VA-ECMO. Expired 8 days later</p>	Died at 13 years of age 1 year post-op

Case	Presentation	Height %	Weight %	Hgb (g/dl)	O2 Sat	Qp:Qs	PA anatomy	Nakata index (mm ² /m ²)	PVD	Interventions	Follow-up
6	3 year old boy with CCAVC unbalanced to the right and PA (S,D,D) with MAPCA born in the People's Republic of China and recently adopted	<1	66	19.8	80	1.6	Confluent PA's 3 MAPCA (all sole supply)	67	None	1 3 years: Division of MPA, and central shunt from ascending aorta to MPA At subsequent catheterization (4 years) improved PA growth (Nakata index of 96 mm ² /m ² , no PVD, and transpulmonary gradient of 8 mmHg with PVRi of 2.1 WU/m ²)	4 years of age 1 year post-op
7	8 year old girl with DILV PA (S,D,D) with MAPCA and multiple dysmorphic features (severe scoliosis, developmental delay, and ear canal malformation) born in El Salvador and recently immigrated	42	21	22.4	76	0.4	No confluent PA's 2 MAPCA Loss of perfusion to all 10 segments of the right lung	n/a	None	Consensus after discussion was that there was not an operative solution. Discussion as to whether stent angioplasty of the collateral supplying 80% of left lung.	8 years of age n/a
8	7 year old boy with TET PA MAPCA born in the People's Republic of China and recently adopted	66	31	15.7	85	2.3	Confluent PA's 2 MAPCA (both sole supply)	260	Severe 2 segments	Planning on single stage operation in Spring 2016	7 years n/a

Abbreviations: CCAVC: complete common atrioventricular canal, PA: pulmonary arteries, PVD: pulmonary vascular disease, PVRi: pulmonary vascular resistance indexed to body surface area, RV: right ventricle, TET PA MAPCA: tetralogy of Fallot pulmonary atresia MAPCA's, VA-ECMO: veno-arterial extracorporeal membrane oxygenation