Journal of the American Heart Association

ORIGINAL RESEARCH

Angiographic Anatomy of Major Aortopulmonary Collateral Arteries and Association With Early Surgical Outcomes in Tetralogy of Fallot

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BACKGROUND: Due in part to the heterogeneity of the pulmonary circulation in patients with tetralogy of Fallot and major aortopulmonary collateral arteries (MAPCAs), research on this condition has focused on relatively basic anatomic characteristics. We aimed to detail pulmonary artery (PA) and MAPCA anatomy in a large group of infants, assess relationships between anatomy and early surgical outcomes, and consider systems for classifying MAPCAs.

METHODS AND RESULTS: All infants (<1 year of age) undergoing first cardiac surgery for tetralogy of Fallot/MAPCAs from 2001 to 2019 at Stanford University were identified. Preoperative angiograms delineating supply to all 18 pulmonary segments were reviewed for details of each MAPCA and the arborization and size of central PAs. We studied 276 patients with 1068 MAPCAs and the following PA patterns: 152 (55%) incompletely arborizing PAs, 48 (17%) normally arborizing PAs, 45 (16%) absent PAs, and 31 (11%) unilateral MAPCAs. There was extensive anatomic variability, but no difference in early outcomes according to PA arborization or the predominance of PAs or MAPCAs. Patients with low total MAPCA and/or PA cross-sectional area were less likely to undergo complete repair.

CONCLUSIONS: MAPCA anatomy is highly variable and essentially unique for each patient. Though each pulmonary segment can be supplied by a MAPCA, central PA, or both, all anatomic combinations are similarly conducive to a good repair. Total cross-sectional area of central PA and MAPCA material is an important driver of outcome. We elucidate a number of novel associations between anatomic features, but the extreme variability of the pulmonary circulation makes a granular tetralogy of Fallot/MAPCA classification system unrealistic.

Key Words: catheterization ■ DiGeorge syndrome ■ major aortopulmonary collateral arteries ■ pulmonary artery ■ pulmonary atresia ■ tetralogy of Fallot

The complexity of managing patients with tetralogy of Fallot (TOF) and major aortopulmonary collateral arteries (MAPCAs) revolves around the anatomy and function of the pulmonary circulation. Each of the 18 bronchopulmonary segments can be supplied by MAPCAs, central pulmonary artery (PA), or both, with variable location and severity of stenoses.[1] While the spectrum of MAPCA and PA anatomy is generally recognized, there has been no large or comprehensive evaluation of pulmonary vascular anatomy in a clinical cohort with this condition. Because of the challenge of defining and evaluating this variability, the literature on this condition has typically included limited anatomic information related to the pulmonary circulation, such as the presence or absence of intrapericardial PAs, the number of MAPCAs, and/or the number of lung

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For Sources of Funding and Disclosures, see page 13.

Supplementary Material for this article is available at https://www.ahajournals.org/doi/suppl/10.1161/JAHA.120.017981

JAHA is available at: www.ahajournals.org/journal/jaha

CLINICAL PERSPECTIVE

What Is New?

- We provide novel insight into the distribution and constellation of anatomic features in infants with tetralogy of Fallot and major aortopulmonary collateral arteries (MAPCAs).
- The distribution of MAPCAs were essentially unique for each patient, making a universal classification scheme unrealistic.
- A predominance of MAPCAs versus central pulmonary arteries, as measured both by cross-sectional area and by number of lung segments supplied, was not associated with early outcomes.

What Are the Clinical Implications?

- Anatomic associations revealed in this work can guide the preoperative evaluation and surgical approach when managing infants with MAPCAs.
- Using a strategy of early complete unifocalization, a good early surgical result can be achieved for essentially any anatomic variation.
- Less total cross-sectional area of usable native MAPCA and pulmonary artery material portends a worse outcome.

Nonstandard Abbreviations and Acronyms

MAPCAs	major aortopulmonary collateral arteries
TNPAI	total neo-pulmonary artery index

segments connected to the central PAs.[2–11] Although we recently reported outcomes in a large cohort of patients with TOF/MAPCAs,[3] we have not analyzed patients according to more detailed anatomic features of the pulmonary circulation. However, we suspect that there are important anatomic aspects that can be elucidated via detailed characterization of the pulmonary circulation. The aims of this study were to detail the PA and MAPCA anatomy in a large group of infants, assess relationships between anatomic variables and early outcomes, and consider systems for classifying patients with MAPCAs in an effort to improve communication, facilitate understanding of risk factors and outcomes, and define subgroups to enhance collaboration and clinical and translational research.

METHODS

The data that support the findings of this study are available from the corresponding author upon reasonable request. This study was approved by the Stanford University Institutional Review Board (Protocol 56280) with a waiver of consent.

Patients

With approval from the Stanford University Institutional Review Board (Protocol 56280), all patients with TOF/MAPCAs undergoing cardiac surgery at Lucile Packard Children's Hospital between November 2001 and August 2019 were identified. Patients were included in this study if preoperative angiograms were available that clearly and completely delineated the native MAPCA and PA anatomy as well as the blood supply to all 18 bronchopulmonary segments. Patients >1 year of age at catheterization were excluded given that MAPCAs left in a native state for a prolonged period can become atretic, and untreated older children may develop acquired collaterals to the pulmonary circulation, both of which can obscure understanding of the native anatomy.

Characterization of MAPCAs

MAPCAs were characterized angiographically according to number, origin, course, and segmental supply, as well as the location of any connection to the central, branch, or lobar PAs (Table S1).

Number

Although MAPCAs can be quantified in several ways, for this anatomic study a vessel was counted if it had an identifiable origin from the systemic circulation. A vessel with a common trunk before branching was counted as a single MAPCA with branches, whereas if no common trunk was seen, branches arising from the same site were counted as separate MAPCAs. This is different from our surgical definition[1] where each branch requiring unifocalization is counted separately.

Origin

Origins were identified as left or right subclavian artery (SCA) (including branches thereof), transverse aorta, descending thoracic aorta, abdominal aorta (at or below the level of the diaphragm), or coronary artery. The presence of a unilateral ductus arteriosus (PDA) or an anomalous PA from the ascending aorta were also documented.

Course

The course of a MAPCA was characterized based on its insertion into the lung parenchyma: leftward (into the left PA), rightward (into the right PA), bilateral (branches to right and left PA), or midline (into the intrapericardial PA).

Nature of supply

Each MAPCA was classified as (1) single-supply (MAPCA serves as the only source of blood flow to the lung segment[s]), with no connection to a central PA, (2) isolated supply to central PAs (the only MAPCA connecting to the central PAs; (3) dual-supply (1 of ≥2 MAPCAs connecting to the central PAs); or (4) mixed-supply (≥1 single-supply and ≥1 dual-supply branches), as demonstrated in Figures 1 and 2.

Unilateral PDA

A unilateral PDA was defined as a patent or involuted vessel arising from the typical location of a PDA (ie, the isthmus or the innominate artery, depending on arch laterality and branching) and supplying a normally arborizing PA, with no MAPCAs to that lung and no intrapericardial PA (Figure 1). Based on our understanding that a PDA and single-supply collaterals should not supply the same lung, and on the observation that the recurrent laryngeal nerve was not anatomically associated with any such vessel on intraoperative inspection, a vessel arising from the same general location as a PDA but supplying only part of 1 or both PAs was considered a MAPCA (Figure 1).

Angiogram Review

Angiograms were reviewed offline by 2 investigators (G.T.A., S.B.P.) masked to clinical details. MAPCAs and PA characteristics were recorded, along with aortic arch sidedness and branching, and coronary anatomy. When present, the proximal intrapericardial PAs were measured in the anteroposterior projection and the average of left and right PAs reported. Because intrapulmonary PAs are often larger than intrapericardial PAs in TOF/MAPCAs (Figure 2), PA diameters were also measured at the first lobar bifurcation in the anteroposterior projection and used to calculate a modified Nakata index, with the assumption of circular cross-section.[12] Single-supply MAPCAs were measured distal to the likely unifocalization site, beyond any central, lobar, and segmental stenoses (Figure 2). The sum of the cross-sectional areas of single-supply MAPCAs and single-supply branches from mixed-supply MAPCAs (Figures 1 and 2) was defined as the MAPCA index. If a single supply MAPCA branch arose proximal

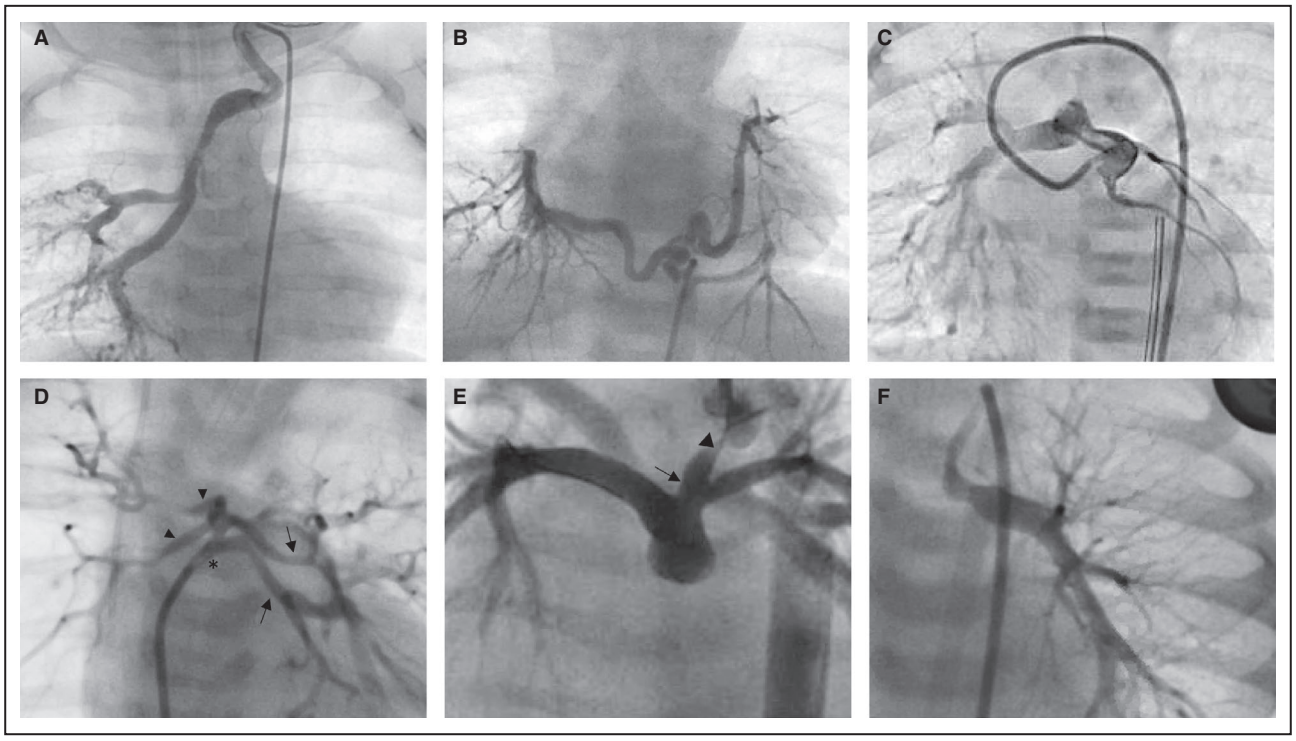

Figure 1. Examples of major aortopulmonary collateral arteries (MAPCA) and a ductus arteriosus in 6 different patients. **A**, Single-supply MAPCA arising from the left subclavian artery and coursing rightward to provide sole supply to a portion of the right lower pulmonary artery (PA). **B**, Abdominal aorta MAPCA giving 2 dual-supply branches, 1 to the right intermediate and 1 to the left PA. **C**, Coronary MAPCA arising from a dilated left main coronary, inserting in the intrapericardial main PA and supplying a normally arborizing right PA. **D**, Mixed-supply MAPCA with a common trunk (*) and multiple branches; 2 (arrow) connect to a central left PA, and 2 (arrowheads) are single-supply to portions of the right PA. **E**, MAPCA with origin (arrowhead) and insertion (arrow) consistent with a ductus arteriosus, but supplying incompletely arborizing intrapericardial PAs, with 10 segments supplied by single-supply MAPCAs. **F**, Prostaglandin-sensitive ductus arteriosus supplying a normally arborizing left PA.

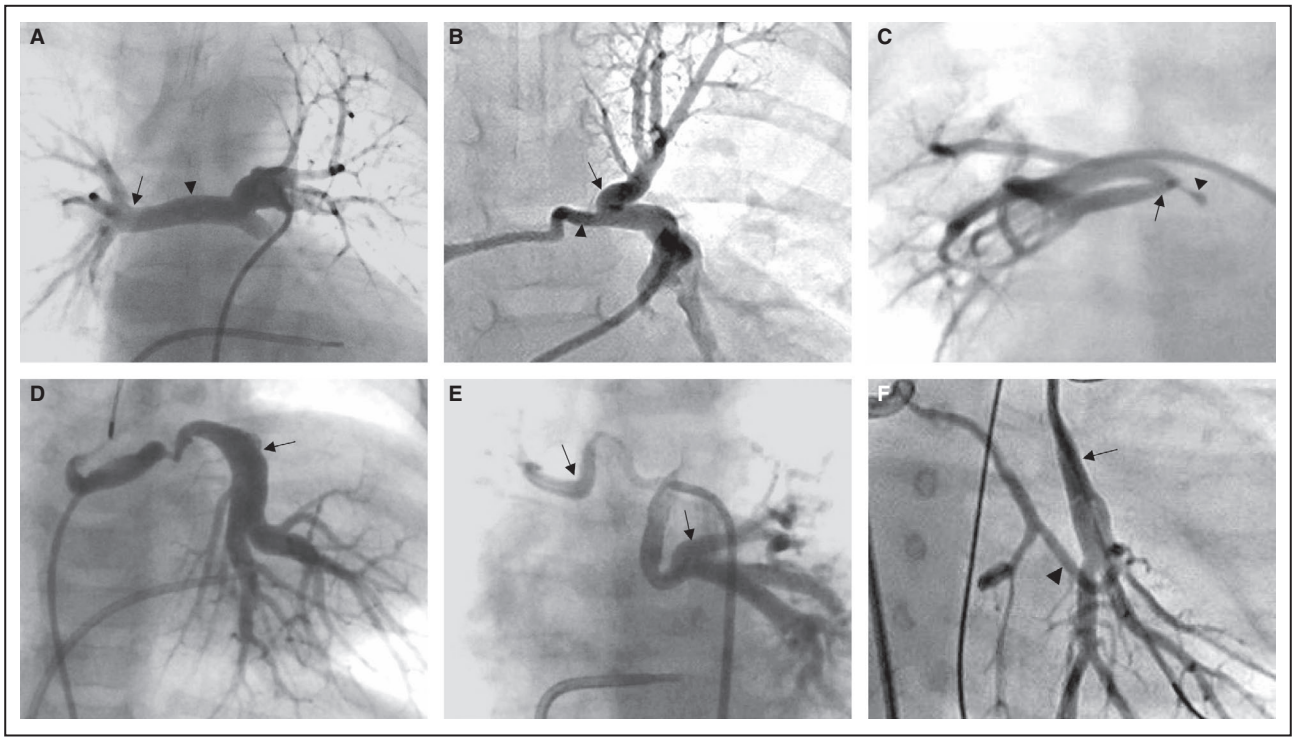

Figure 2. Examples of major aortopulmonary collateral arteries (MAPCA) and central pulmonary arteries (PAs) and location of measurements in 6 different patients.
A, Dual-supply MAPCA connecting at the left hilum and supplying large incompletely arborizing central PAs with intrapericardial (arrowhead) and intrapulmonary (arrow) portions similar in size. **B** and **C**, Dual-supply MAPCAs feeding an incompletely arborizing intrapulmonary PA (arrow) with relatively hypoplastic intrapericardial PAs (arrowhead). **D** and **E**, Single-supply MAPCA with 2 branches, with arrows at the sites of measurement, past all stenoses at the site of anticipated unifocalization. **F**, Dual-supply MAPCA with a small connection to the central PA (arrowhead), which will require unifocalization and be included in the MAPCA index (measured at black arrow).

to a small dual-supply connection to the PAs, it was measured separately and included in the MAPCA index (Figure 2F). The modified Nakata and MAPCA indices were summed to give a total neo-pulmonary artery index (TNPAI), similar to prior work.[13] Patients in whom all measurements could not be made confidently were excluded because of inadequate angiography.

Surgical Management

Our management approach for TOF/MAPCAs has been detailed previously.[1,3] Complete repair refers to unifocalization, ventricular septal defect closure, and placement of a right ventricle (RV) to PA conduit, which are usually performed during the same operation (single-stage repair), and less often in stages. Unifocalization to a central shunt is performed if an intraoperative flow study shows that RV pressure would be too high with ventricular septal defect closure.[13] For patients with hypoplastic normally arborizing central PAs and all dual-supply MAPCAs, an aortopulmonary window is performed early in life as the first surgery.[1,14] Further details about our surgical approach are in Data S1.

Surgical Outcomes

The first surgery was classified as single-stage complete repair, aortopulmonary window, or other palliation (unilateral or bilateral unifocalization to a central shunt, or unilateral PA banding). For patients who did not undergo single-stage complete repair, each subsequent surgery was reviewed, and classified as complete repair (multi-stage) or ongoing palliation (unrepaired). Follow-up was conducted by a combination of medical record review and contact with the primary physician for all patients who did not undergo complete repair at our institution, and patients' current status classified as "unrepaired" or "death without repair". The ratio of right ventricle to aortic pressure (RV:Ao) at time of complete repair was taken in the operating room via intracardiac lines following chest closure, which is routinely recorded in the operative and intensive care unit records.

Patient Grouping and Analysis

One aim of this study was to determine whether an anatomically-based classification system could be derived for patients with MAPCAs. To provide the most detailed analysis possible, we classified patients with

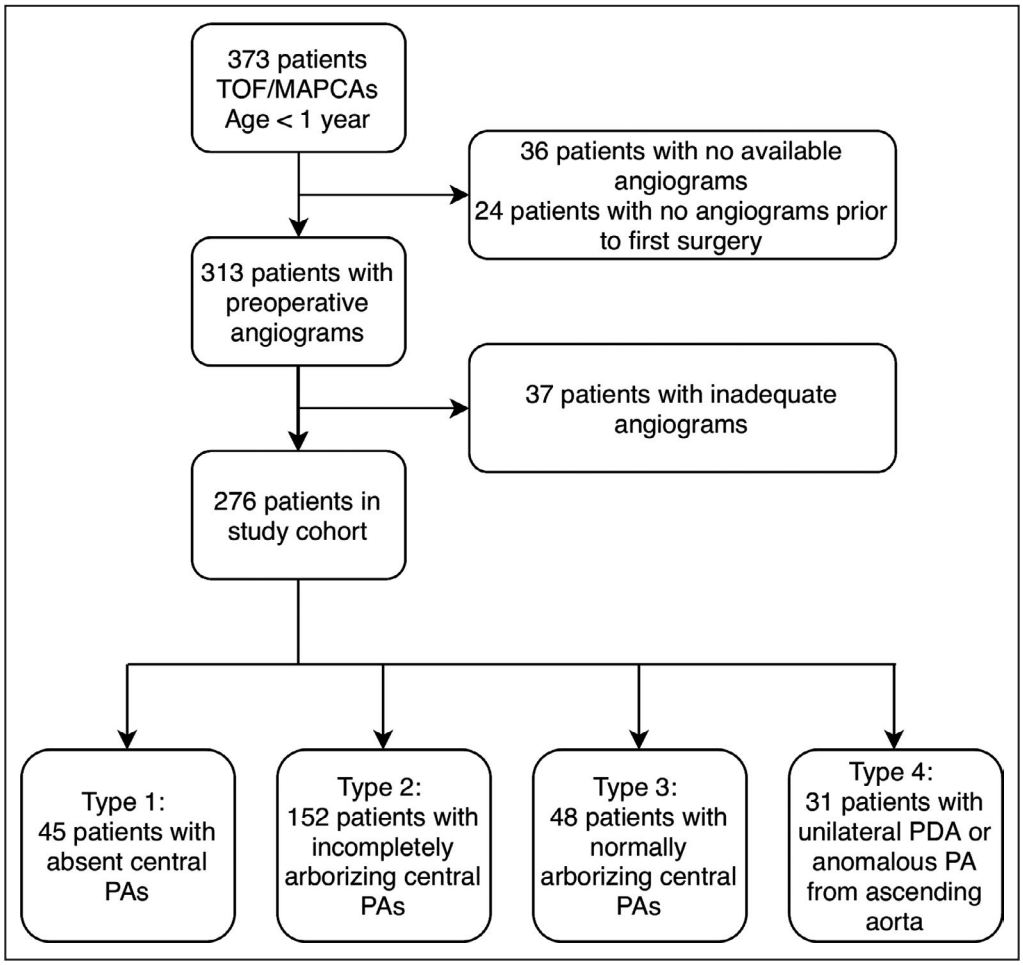

Figure 3. Study flowchart.
Diagram of the exclusion and inclusion criteria and breakdown of included patients by pulmonary artery arborization pattern. PAs indicates pulmonary arteries; PDA, ductus arteriosus; and TOF/MAPCAs, tetralogy of Fallot with major aortopulmonary collateral arteries.

a scheme similar to that proposed by Soquet et al,[15] using features described above plus the number of segments supplied by each MAPCA. Every MAPCA was named according to origin and nature of supply, and patients were organized based on these characteristics, as described in Data S1.

In addition, for the purposes of analysis, patients were grouped according to several criteria: (1) the number of bronchopulmonary segments fed by single-supply MAPCAs (0–3, 4–8, 9–13, and 14–18 segments), (2) the ratio of MAPCA index to TNPAI (0–0.25, 0.25–0.5, 0.5–0.75, and 0.75–1), and (3) the TNPAI (<100, 100–150, 150–200, and >200 mm^2/m^2). Also, based on our management protocol, each patient was assigned to 1 of 4 groups according to central PA arborization and MAPCA supply, similar to the system proposed by Barbero-Marcial[16]:

Type 1: Absent intrapericardial PAs (all single-supply MAPCAs).

Type 2: Incompletely arborizing central PAs (combination of single, dual, and mixed-supply MAPCAs).

Type 3: Normally arborizing central PAs (all dual-supply MAPCAs).

Type 4: Unilateral PDA or anomalous PA arising from the ascending aorta, with MAPCAs to the contralateral PA (with or without an intrapericardial PA segment).

Statistical Analysis

Data were presented as number (%) or median (25th, 75th percentiles). Comparison of categorical variables between groups was performed using Fisher exact test. For numerical variables, Wilcoxon rank sum test or the Kruskal–Wallis test was used. All analyses were performed with R version 3.6.1 and SPSS version 25.0.

RESULTS

Study Cohort

A total of 373 patients <1 year of age with TOF/MAPCAs underwent surgery at our center during the study period. As summarized in Figure 3, 97 of these patients were excluded and the remaining 276 comprised the study cohort. Forty (14%) of the angiograms were performed at outside institutions.

Characteristics of MAPCAs

In these 276 patients, 1068 MAPCAs were identified: 564 (53%) single-supply, 87 (8%) isolated-supply to central PAs, 386 (36%) dual-supply to central PAs, and 31 (3%) mixed-supply. Details are summarized in Table 1. MAPCAs arising from the abdominal aorta were almost always dual-supply (23 of 24), while all 15 MAPCAs from the coronary arteries were dual-supply, and in 6 cases (40%) were the only supply to the central PAs. Nearly all coronary MAPCAs (14 of 15) coursed midline to the intrapericardial PAs. There were 8 MAPCAs with an origin and course suspicious for a PDA that did not supply a normally arborizing unilateral PA and were not associated with the recurrent laryngeal nerve. Other than these 8 MAPCAs and the 14 coronary MAPCAs that coursed to the intrapericardial PAs, all MAPCAs connected at the hilum or more distally in the PA tree. Twenty-six patients had a PDA and 5 had an anomalous PA from the ascending aorta providing full and sole supply to a normally arborizing left (n=29) or right (n=2, 1 PDA, 1 anomalous PA) branch PA.

Patient Details and Anatomy

Demographic, clinical, anatomic, and surgical details of the overall cohort and the 4 anatomic groups are summarized in Table 2.

Anatomy

Patients had 1 to 10 MAPCAs (median 4), with descending thoracic aortic MAPCAs in 99% of patients. The next most common site of origin was the SCA (38%; unilateral in most cases), while MAPCAs from other sites were observed in 5% to 7% of patients each. A right aortic arch was present in 129 (47%) patients and 44 (16%) had an anomalous SCA. Patients with a chromosome 22q11 deletion (n=110, 41%) were more likely to have a right arch, anomalous SCA, and MAPCAs from an SCA, and less likely to have coronary MAPCAs. There were no other differences in MAPCA origins according to 22q11 status or arch sidedness or branching (Table 3 and Table S2). There were several associations between arch sidedness, branching, and SCA origin of MAPCAs; for example, MAPCAs tended to arise from the SCA contralateral to the arch, particularly when anomalous (Table S3). Overall, 31% of SCAs (73 of 232) originating normally from the innominate artery (contralateral to the arch), 12% (33 of 276) of SCAs ipsilateral to the arch, and 41% (18 of 44) of aberrant SCAs had a MAPCA (P<0.001). Coronary anomalies were found in 21% of patients (Table 4).

Intrapericardial PAs were present in 217 (79%) patients. The most common PA pattern was incompletely arborizing central PAs (Type 2; n=152 [55%]), followed

Table 1. Features of MAPCAs Overall and by Type for the Entire Cohort

	MAPCA Type				
	Total MAPCAs (n=1068)	Single-Supply (n=564, 53%)	Isolated Supply to Central PA (n=87, 8%)	Dual-Supply to Central PA (n=386, 36%)	Mixed-Supply (n=31, 3%)
Origin					
Descending thoracic aorta	868 (81%)	487 (86%)	75 (87%)	278 (72%)	28 (90%)
Right subclavian artery	74 (7%)	31 (5%)	3 (3%)	40 (10%)	0 (0%)
Left subclavian artery	71 (7%)	35 (6%)	2 (2%)	31 (8%)	3 (10%)
Coronary artery	15 (1%)	0 (0%)	6 (7%)	9 (2%)	0 (0%)
Transverse aortic arch	16 (1%)	10 (2%)	1 (1%)	5 (1%)	0 (0%)
Abdominal aorta	24 (2%)	1 (<1%)	0 (0%)	23 (6%)	0 (0%)
Course					
Rightward	560 (52%)	309 (55%)	46 (53%)	198 (51%)	7 (23%)
Leftward	384 (36%)	204 (36%)	23 (26%)	151 (39%)	6 (19%)
Bilateral	102 (10%)	51 (9%)	7 (8%)	26 (7%)	18 (58%)
Midline	22 (2%)	0 (0%)	11 (14%)	11 (3%)	0 (0%)
Bronchopulmonary segments supplied	7 (3, 16)	3 (1, 5)	7 (8, 11)	18 (14, 18)	15 (14, 17)

Data presented as n (%) or median (Q1, Q3). MAPCA indicates major aortopulmonary collateral artery; and PA, pulmonary artery.

Table 2. Demographic and Anatomic Features of Patients Overall and by Type of PA Arborization

	Total (n=276)	Type 1: Absent Central PA (n=45)	Type 2: Incompletely Arborizing PA (n=152)	Type 3: Normally Arborizing PA (n=48)	Type 4: PDA or Anomalous PA (n=31)	*P* Value
Demographic features						
Female sex	141 (51%)	22 (49%)	72 (47%)	27 (56%)	20 (65%)	0.30
Age, mo	3.7 (1.0, 6.0)	4.7 (2.5, 6.4)	4.6 (1.7, 6.2)	1.6 (0.1, 4.1)	1.3 (0.2, 4.4)	<0.001
Weight, kg	5.2 (3.7, 6.5)	5.6 (4.3, 6.6)	5.3 (4.0, 6.5)	4.2 (3.5, 6.2)	4.8 (3.6, 5.7)	0.13
Genetic features						
Chromosome 22q11 deletion (n=271)	110 (41%)	25 (56%)	59 (40%)	18 (38%)	8 (27%)	0.085
Alagille syndrome	7 (3%)	1 (2%)	5 (3%)	1 (2%)	0 (0%)	0.94
Anatomic/physiologic features						
Intrapericardial PAs present	217 (79%)	0 (0%)	152 (100%)	48 (100%)	17 (55%)	<0.001
Right aortic arch	129 (47%)	20 (44%)	72 (47%)	25 (52%)	12 (39%)	0.69
Abnormal arch branching	44 (16%)	10 (22%)	24 (16%)	7 (15%)	3 (10%)	0.55
Pulmonary valve stenosis	33 (12%)	0 (0%)	6 (4%)	23 (48%)	4 (13%)	<0.001
Estimated Qp:Qs (n=230)	1.4 (1.0, 2.1)	1.7 (1.0, 2.3)	1.4 (0.9, 2.1)	1.1 (0.9, 1.7)	1.6 (1.3, 3.4)	0.041
No. of MAPCAs	4 (3, 5)	4 (3, 4)	4 (3, 5)	4 (3, 5)	3 (2, 3.5)	<0.001
MAPCA origins						
Thoracic aortic MAPCAs	274 (99%)	45 (100%)	151 (99%)	47 (98%)	31 (100%)	0.70
Thoracic aortic MAPCAs only	153 (55%)	32 (71%)	87 (57%)	17 (35%)	17 (55%)	0.006
Subclavian MAPCAs	105 (38%)	10 (22%)	57 (38%)	28 (58%)	10 (32%)	0.004
Both RSCA and LSCA MAPCAs	26 (9%)	1 (2%)	6 (4%)	19 (40%)	0 (0%)	<0.001
Abdominal aortic MAPCAs	20 (7%)	0 (0%)	2 (1%)	15 (31%)	3 (10%)	<0.001
Coronary MAPCAs	15 (5%)	0 (0%)	5 (3%)	4 (8%)	6 (19%)	0.002
Transverse aortic MAPCAs	15 (5%)	3 (7%)	9 (6%)	2 (4%)	1 (3%)	0.93
PA and MAPCA measurements						
Intrapericardial PA diameter, mm	2.1 (0.8, 3.2)	0 (0, 0)	2.6 (1.8, 3.9)	2.6 (2.1, 3.2)	0.6 (0, 1.4)	<0.001
Intrapericardial PA index, mm^2/m^2	29 (4, 64)	0 (0, 0)	40 (18, 89)	44 (29, 70)	4 (0, 22)	<0.001
Modified nakata, mm^2/m^2	56 (16, 97)	0 (0, 0)	60 (27, 105)	63 (43, 82)	118 (72, 162)	<0.001
MAPCA index, mm^2/m^2	71 (33, 129)	154 (119, 207)	80 (49, 122)	0 (0, 39)	35 (20, 63)	<0.001
TNPAI, mm^2/m^2	148 (99, 195)	154 (119, 207)	165 (111, 204)	80 (51, 116)	165 (121, 225)	<0.001
First surgery						
Complete repair	182 (66%)	34 (76%)	109 (72%)	20 (42%)	19 (61%)	<0.001
Aortopulmonary window	35 (13%)	0 (0%)	10 (7%)	25 (52%)	0 (0%)	
Other palliation	59 (21%)	11 (24%)	33 (22%)	3 (6%)	12 (39%)	
Current status						
Single-stage complete repair	182 (66%)	34 (76%)	109 (72%)	20 (42%)	19 (61%)	<0.001
Multi-stage complete repair	67 (24%)	7 (16%)	27 (18%)	25 (52%)	8 (26%)	
Unrepaired	12 (4%)	1 (2%)	6 (4%)	3 (6%)	2 (6%)	
Death without repair	15 (5%)	3 (7%)	10 (7%)	0 (0%)	2 (6%)	
Complete repair (ever)	249 (90%)	41 (91%)	136 (89%)	45 (94%)	27 (87%)	0.76
RV:Ao at complete repair	0.33 (0.28, 0.40)	0.31 (0.27, 0.36)	0.33 (0.28, 0.40)	0.31 (0.28, 0.38)	0.34 (0.25, 0.40)	0.40

Data presented as n (%) or median (Q1, Q3). LSCA indicates left subclavian artery; MAPCA, major aortopulmonary collateral artery; PA, pulmonary artery; PDA, ductus arteriosus; Qp:Qs, ratio of pulmonary to systemic blood flow; RV:Ao, ratio of right ventricle to aortic pressure; and TNPAI, total neo-pulmonary artery index.

by normally arborizing PAs (Type 3; n=48 [17%]), and absent central PAs (Type 1; n=45 [16%]), with a PDA or anomalous PA from the ascending aorta and contralateral MAPCAs (Type 4) the least common pattern (n=31, 11%). Among patients with intrapericardial PAs, a median of 12 (9, 17) pulmonary segments were associated with the central PAs. There was remarkable variability in the number, origin, and segmental supply of MAPCAs, and in PA arborization, but some associations were apparent. Patients that were Type 1 were

Table 3. Features of the Pulmonary Circulation According to Chromosome 22q11 Status

	Total (n=271)	Chromosome 22q11 Deletion		*P* Value
		No (n=161)	Yes (n=110)	
Thoracic aorta MAPCAs	269 (99%)	160 (99%)	109 (99%)	>0.99
Thoracic aorta MAPCAs only	151 (56%)	93 (58%)	58 (53%)	0.46
Subclavian MAPCAs	103 (38%)	53 (33%)	50 (45%)	0.042
Abdominal MAPCAs	20 (7%)	12 (7%)	8 (7%)	>0.99
Coronary MAPCAs	14 (5%)	12 (7%)	2 (2%)	0.050
Transverse arch MAPCAs	15 (6%)	10 (6%)	5 (5%)	0.60
RSCA and LSCA MAPCAs	25 (9%)	15 (9%)	10 (9%)	>0.99
Intrapericardial PAs	213 (79%)	132 (82%)	81 (74%)	0.13
Right aortic arch	127 (47%)	59 (37%)	68 (62%)	<0.001
Abnormal arch branching	44 (16%)	9 (6%)	35 (32%)	<0.001
Pulmonary stenosis	33 (12%)	23 (14%)	10 (9%)	0.26
Modified Nakata, mm^2/m^2	56 (16, 99)	60 (22, 111)	44 (13, 90)	0.089
MAPCA index, mm^2/m^2	72 (32, 129)	65 (29, 119)	86 (39, 146)	0.040
TNPAI, mm^2/m^2	149 (100, 197)	150 (94, 193)	149 (108, 200)	0.66
Complete repair	245 (90%)	143 (89%)	102 (93%)	0.30
RV:Ao at complete repair	0.33 (0.28, 0.40)	0.31 (0.26, 0.37)	0.35 (0.30, 0.41)	<0.001

Only includes patients with known presence or absence of a chromosome 22q11 deletion. Data presented as n (%) or median (Q1, Q3). LSCA indicates left subclavian artery; MAPCA, major aortopulmonary collateral artery; PA, pulmonary artery; RSCA, right subclavian artery; RV:Ao, ratio of right ventricle to aortic pressure; and TNPAI, total neo-pulmonary artery index.

more likely to have exclusively descending thoracic aorta MAPCAs than other types (*P*=0.006), while Type 3 were more likely to have abdominal aorta (*P*<0.001) and bilateral SCA MAPCAs (*P*=0.004).

Pulmonary valve stenosis occurred in about half (n=23, 48%) of patients with Type 3, and rarely in Type 4 (n=4, 13%) or Type 2 (n=6, 4%). In 3 of the 4 patients with Type 4 with pulmonary stenosis, the nearly atretic RV outflow tract supplied a hypoplastic normally arborizing single PA. Thus, of the 33 instances of pulmonary stenosis, 26 (79%) were associated with uni- or bilateral hypoplastic normally arborizing PAs. In the patients with Type 2 with pulmonary stenosis, a majority of bronchopulmonary segments (from 12 to 17) connected to the central PAs.

A MAPCA naming system based on origin, supply type, and number of bronchopulmonary segments supplied is detailed in Data S1. When the names of each MAPCA in a given patient were strung together, there were 222 unique codes among the 276 patients, of which 187 (84%) occurred only once. This scheme did not account for additional factors such as the specific segments supplied, the number or severity of PA stenoses, or the location and size of connections of dual-supply MAPCAs to the PAs. Given this heterogeneity, additional attempts to group patients using this methodology were not pursued.

Table 4. Coronary Artery Anomalies

Coronary Artery Pattern and Anomalies (34 Unknown, n=242)	
Normal pattern and supply	192 (79%)
LAD from the right	11 (4.5%)
Dual LAD	4 (1.7%)
Anomalous right from left main	7 (2.9%)
Anomalous left main from right	8 (3.3%)
Coronary artery fistula	5 (2.1%)
MAPCA	15 (6.2%)

LAD indicates left anterior descending; and MAPCA, major aortopulmonary collateral artery.

MAPCA and PA indices

As documented in Table 2 and Figure 4, the TNPAI was significantly lower in patients with Type 3 (*P*<0.001). Within patients with Type 3, those with pulmonary atresia and pulmonary stenosis had similar TNPAI (86.0 [62.3, 125.8] versus 70.2 [46.2, 98.3], *P*=0.066) and intrapericardial PA diameter (2.6 [2.4, 3.2] versus 2.5 [2.1, 3.3]; *P*=0.71).

Patients with and without a chromosome 22q11 deletion had similar TNPAI, but on average those with a deletion had more cross-sectional area contributed by single-supply MAPCAs (Table 3). Patients with Alagille syndrome had lower TNPAI than those without (89.3 [70.5, 131.0] versus 149.1 [100.4, 197.5] mm^2/m^2; *P*=0.043).

Outcomes

Selected surgical outcomes are summarized in Tables 4 through 7. No patients who underwent complete repair

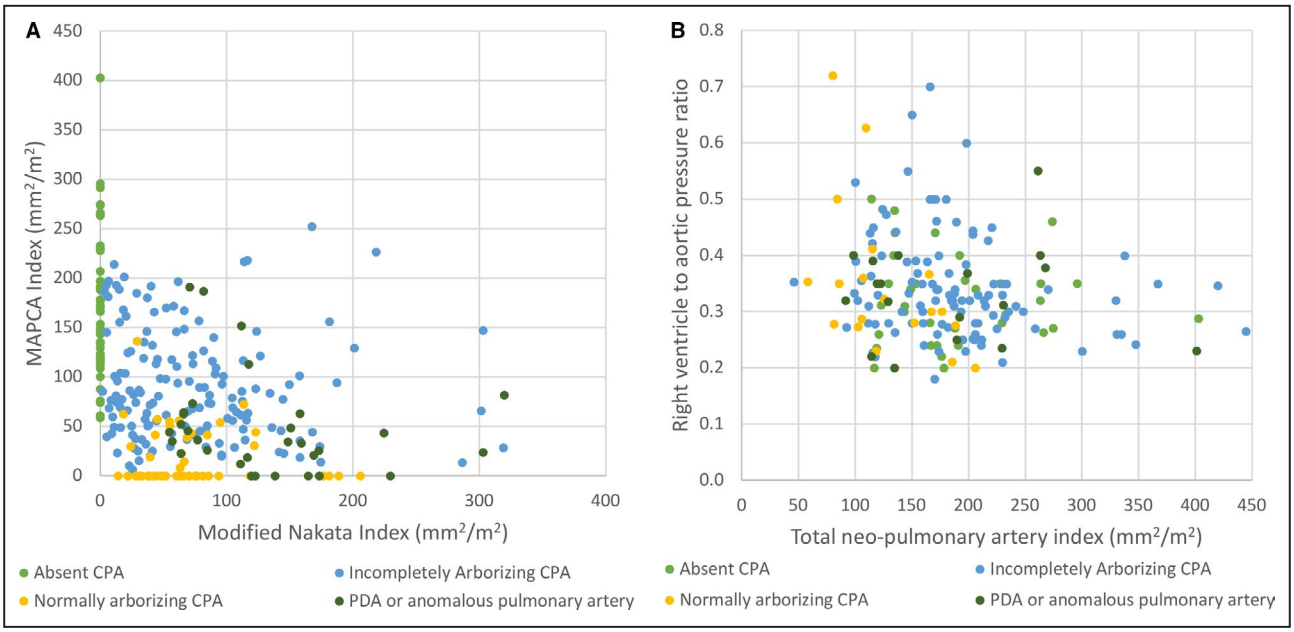

Figure 4. Relationship of pulmonary artery indices and postoperative right ventricle pressure according to pulmonary artery arborization pattern.
Scatterplots showing relationships of (**A**) major aortopulmonary collateral artery index, the modified Nakata index, and patient central pulmonary artery (CPA) arborization pattern (in all patients), and (**B**) postoperative right ventricle to aortic pressure ratio, the total neopulmonary artery index, and the central pulmonary artery arborization pattern (in patients who underwent complete repair). PDA indicates ductus arteriosus; and MAPCAs, major aortopulmonary collateral arteries.

had the ventricular septal defect opened or the RV-PA connection taken down. Patients with Type 3 and Type 4 were more likely to require a staged approach (P<0.001), but ultimately there was no significant difference in the proportion of patients reaching complete repair or in the post-repair RV:Ao between anatomic groups or according to the number of bronchopulmonary segments supplied by single-supply MAPCAs (Table 3 and Figure 4).

Patients with a larger TNPAI were more likely to undergo single-stage complete repair (Table 6 and Figure 5). A TNPAI <100 mm²/m² was associated with remaining unrepaired or experiencing death before complete repair, though 55 (77%) of these patients did achieve complete repair with an RV:Ao of 0.35 (0.28, 0.40). Among patients who achieved complete repair, there was no association between TNPAI and the postoperative RV:Ao. A summary figure with selected

Table 5. Surgical Outcomes Overall and According to the Number of Single-Supply Pulmonary Segments

Outcome	Total (n=276)	No. of Single-Supply Pulmonary Segments				*P* Value
		0–3 (n=98)	4–8 (n=64)	9–13 (n=64)	14–18 (n=50)	
First surgery						
Complete repair	182 (66%)	53 (54%)	51 (80%)	41 (64%)	37 (74%)	<0.001
Aortopulmonary window	35 (13%)	30 (31%)	3 (5%)	2 (3%)	0 (0%)	
Other palliation	59 (21%)	15 (15%)	10 (16%)	21 (33%)	13 (26%)	
Current status						
Single-stage complete repair	182 (66%)	53 (54%)	51 (80%)	41 (64%)	37 (74%)	0.020
Multi-stage complete repair	67 (24%)	36 (37%)	8 (12%)	14 (22%)	9 (18%)	
Unrepaired	12 (4%)	5 (5%)	1 (2%)	5 (8%)	1 (2%)	
Death without repair	15 (5%)	4 (4%)	4 (6%)	4 (6%)	3 (6%)	
Complete repair (ever)	249 (90%)	89 (91%)	59 (92%)	55 (86%)	46 (92%)	0.66
RV:Ao at complete repair	0.33 (0.28, 0.40)	0.32 (0.28, 0.39)	0.32 (0.28, 0.38)	0.35 (0.30, 0.43)	0.33 (0.27, 0.38)	0.31

Data presented as n (%) or median (Q1, Q3). RV:Ao indicates ratio of right ventricle to aortic pressure.

Table 6. Surgical Outcomes Overall and According to TNPAI Grouping

Outcome	Total (n=276)	TNPAI (mm^2/m^2)				*P* Value
		0–100 (n=71)	100–150 (n=70)	150–200 (n=72)	>200 (n=63)	
First surgery						
Complete repair	182 (66%)	11 (15%)	50 (71%)	63 (88%)	58 (92%)	<0.001
Aortopulmonary window	35 (13%)	31 (44%)	3 (4%)	1 (1%)	0 (0%)	
Other palliation	59 (21%)	29 (41%)	17 (24%)	8 (11%)	5 (8%)	
Current status						
Single-stage complete repair	182 (66%)	11 (15%)	50 (71%)	63 (88%)	58 (92%)	<0.001
Multi-stage complete repair	67 (24%)	44 (62%)	14 (20%)	5 (7%)	4 (6%)	
Unrepaired	12 (4%)	8 (11%)	1 (1%)	2 (3%)	1 (2%)	
Death without repair	15 (5%)	8 (11%)	5 (7%)	2 (3%)	0 (0%)	
Complete repair (ever)	249 (90%)	55 (77%)	64 (91%)	68 (94%)	62 (98%)	<0.001
RV:Ao at complete repair	0.33 (0.28, 0.40)	0.35 (0.28, 0.40)	0.35 (0.29, 0.42)	0.33 (0.28, 0.38)	0.31 (0.26, 0.35)	0.053

Data presented as n (%) or median (Q1, Q3). RV:Ao indicates ratio of right ventricle to aortic pressure; and TNPAI, total neo-pulmonary artery index.

central PA angiograms and surgical outcomes is shown in Figure S1.

DISCUSSION

It is challenging to assess and evaluate all of the features of the pulmonary circulation in TOF/MAPCAs in an integrated but straightforward and coherent fashion. This difficulty has potential implications for both treatment and investigation of outcomes and risk factors. In this report, which is the most comprehensive evaluation of pulmonary vascular anatomy in patients with TOF/MAPCAs, we described the highly variable anatomy of >1000 MAPCAs and central PA configurations in 276 infants, providing novel insight into the distribution and constellation of anatomic features in this population. We also attempted to characterize the variations in the pulmonary circulation, but the data revealed that the extensive variability of MAPCA anatomy and supply confounds useful systematic classification. As vexing as that may be, this study also confirmed our empirical impression that if the surgical principles of using all available raw material (all MAPCAs and the central PAs) are adhered to, a good early surgical outcome can be achieved for essentially any anatomic variation.

Anatomy

In this experience, there were minor associations between MAPCA number, origin, supply, arch anatomy, and genetic status, adding to prior smaller reports.[17–20] The presence of a unilateral PDA or anomalous PA from the ascending aorta, for example, should prompt careful evaluation for coronary MAPCAs. Abdominal aorta MAPCAs and MAPCAs from both SCAs were typically, although not always, associated with a normally arborizing PA tree, and awareness of this association may

Table 7. Surgical Outcomes Overall and According to the Ratio of the MAPCA Index to the TNPAI

Outcome	Total (n=276)	MAPCA Index/TNPAI				*P* Value
		0–0.25 (n=65)	0.25–0.5 (n=69)	0.5–0.75 (n=54)	0.75–1 (n=88)	
First surgery						
Complete repair	182 (66%)	30 (46%)	53 (77%)	39 (72%)	60 (68%)	<0.001
Aortopulmonary window	35 (13%)	23 (35%)	5 (7%)	4 (7%)	3 (3%)	
Other palliation	59 (21%)	12 (18%)	11 (16%)	11 (20%)	25 (28%)	
Current status						
Single-stage complete repair	182 (66%)	30 (46%)	53 (77%)	39 (72%)	60 (68%)	0.003
Multi-stage complete repair	67 (24%)	29 (45%)	12 (17%)	8 (15%)	18 (20%)	
Unrepaired	12 (4%)	3 (5%)	3 (4%)	3 (6%)	3 (3%)	
Death without repair	15 (5%)	3 (5%)	1 (1%)	4 (7%)	7 (8%)	
Complete repair (ever)	249 (90%)	59 (91%)	65 (94%)	47 (87%)	78 (89%)	0.52
RV:Ao at complete repair	0.33 (0.28, 0.40)	0.30 (0.26, 0.38)	0.33 (0.30, 0.41)	0.33 (0.28, 0.39)	0.34 (0.28, 0.40)	0.55

Data presented as n (%) or median (Q1, Q3). MAPCA indicates ratio of the major aortopulmonary collateral arteries; RV:Ao, ratio of right ventricle to aortic pressure; and TNPAI, total neopulmonary artery index.

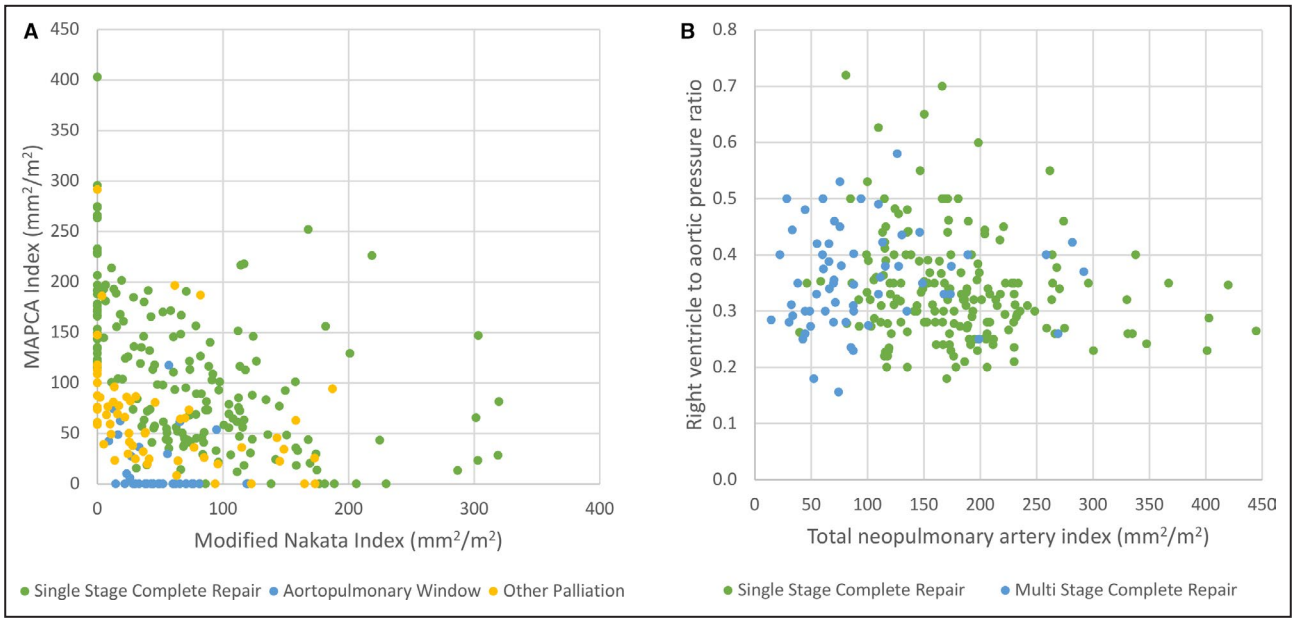

Figure 5. Relationship of pulmonary artery indices and postoperative right ventricle pressure according to surgical approach.
Scatterplots showing relationships of (**A**) major aortopulmonary artery index, modified Nakata index, and the first surgical procedure performed (in all patients), and (**B**) postoperative right ventricle to aortic pressure ratio, the total neopulmonary artery index, and single vs. multi-stage complete repair (in patients who underwent complete repair). In (**A**) patients with the lowest total neopulmonary artery index are closest to the left-lower corner and tended to be managed with a palliative procedure. However, of those who achieved complete repair (**B**), the right ventricle to aortic pressure was similar regardless of initial total neopulmonary artery index. MAPCA indicates major aortopulmonary collateral arteries.

be helpful during the evaluation of these patients. While understanding the associations found in this study are important for case planning, they do not appear to provide extensive insight into other anatomic features or outcomes.

Patients with normal or near-normally arborizing PAs tended to have more MAPCAs but a lower TNPAI compared with those with absent intrapericardial PAs. One possible explanation for this seemingly paradoxical phenomenon is that competitive flow from multiple sources could adversely affect the development of distal vasculature. This observation also suggests that the MAPCAs in this subgroup may have been acquired in fetal life, rather than having developed from early embryologic foregut vessels. The normal arborization of the central PA system, and the occasional patency of the pulmonary valve, also supports this hypothesis of a fetal process occurring after embryogenesis is completed.

Contrary to prior reports,[21,22] there were no significant differences in PA size or anatomy between patients with and without a chromosome 22q11 deletion. The few patients with Alagille in this cohort had markedly less PA/MAPCA cross-sectional area, which we believe is a driver of relatively poor outcome.[1,3] There was a high prevalence of coronary artery anomalies, including MAPCAs and coronaries crossing the RV outflow tract, and therefore preoperative coronary angiography is required.

Outcomes

Patients with large central PAs, large MAPCAs, or both (ie, a large TNPAI) had the highest likelihood of single-stage complete repair and tended to have lower early RV:Ao. Even among patients with the lowest TNPAI (<100 mm²/m²), a majority (>75%) eventually underwent repair with acceptable RV pressure. Few patients with a high TNPAI had a high post-repair RV pressure or did not undergo single-stage repair. Though estimating the TNPAI may help frame expectations about the likelihood of 1-stage repair, we do not use quantitative preoperative estimates of post-reconstruction PA size in surgical decision-making.

Despite the anatomic variability between patients with the different PA arborization patterns (Types 1–4), outcomes did not differ. Nevertheless, we believe that grouping patients based on PA arborization pattern is useful, since it has bearing on the timing and type of surgery.[1,3,23] Patients with Type 3, for example, undergo an elective aortopulmonary window early in life,[14,23] and patients with Type 4 often undergo neonatal intervention to control pulmonary blood flow to the lung without MAPCAs,[1] which helps account for the significant difference in first surgery between patients groups.

There is ongoing debate about the relative effectiveness of unifocalization[3,24,25] versus PA rehabilitative

approaches[10] (or a combination thereof[2,4,26,27]) for patients with incompletely arborizing PAs. The crux of management decisions at certain centers relies on the perceived dominance of central PAs or single-supply MAPCAs in an individual patient. We did not find MAPCA or central PA dominance, based on the number of lung segments supplied by single-supply MAPCAs or by the MAPCA index:TNPAI ratio, to be associated with differences in the rate of complete repair or in the immediate RV:Ao. Though there remains a sentiment that central PAs are a superior platform for repair,[15] the findings of this study suggest that PA segments supplied by single-supply MAPCAs are just as conducive to an overall good immediate repair as PA segments associated with an arborizing central PA. However, absent central PAs were associated with a higher incidence of surgical reintervention for elevated PA pressures,[28] and it is possible that certain PA arborization patterns will fare more poorly over time. Analysis of longer-term outcomes according to patient anatomic groupings described in this article is a subject of ongoing study. Overall, longer-term outcomes are generally favorable.[3]

It is our impression that once a MAPCA reaches the lung parenchyma, lung segments supplied by MAPCAs are indistinguishable from segments arborizing from central PAs, in terms of both quality and distribution. The use of a particular segment in the PA reconstruction does not appear to be related directly to the native course that blood takes to supply that segment. It is necessary that the physiology and anatomy of each lung segment is understood before surgery, such that the surgeon can plan the reconstruction and ensure unobstructed supply to all segments. Complex anatomies are more time intensive and technically challenging but following our approach the RV:Ao generally did not differ.

Although we and others have found worse long-term survival in patients with a chromosome 22q11 deletion than those without,[3,4,29,30] in this study there was no difference in the proportion of patients who achieved complete repair and only marginal differences in the post-repair RV:Ao. These immediate outcome data coupled with the anatomic findings in this study suggest that differences in long-term mortality in patients with a chromosome 22q11 deletion may be unrelated to PA size or anatomy.

Classification

Current classification systems place TOF/MAPCAs as a subset of TOF or ventricular septal defect with pulmonary atresia and/or by presence or absence of intrapericardial PAs.[31] Others have proposed different groupings,[4,15,16,32] but these have not been widely adopted. A system similar to the primary grouping in this study was proposed in 1990 by Barbero-Marcial et al.[16]

Our data suggests that in TOF/MAPCAs essentially every arborization pattern of the PAs, every variation of intrapericardial versus intrapulmonary PA size, and every combination of MAPCA origin and distribution exist on a spectrum, and that none of these variations are strongly associated with early surgical outcomes when using our management strategy and surgical techniques. Although we hoped this study would allow us to discern useful anatomic patterns for classifying patients, we conclude that attempts at a nomenclature system for TOF/MAPCAs will be either too simple or too complicated to be meaningfully tied to clinical outcomes. We continue to group patients (Type 1–4) primarily to facilitate communication about basic PA anatomy and the expected surgical management. Understanding all details, including MAPCA and central PA size, connections, stenoses, and other relationships in each individual case is necessary for the surgeon to achieve a good outcome.

Limitations

Limitations of this study include the single institution retrospective design. The majority of patients were referred from outside our local catchment area, and it is possible that selection bias may limit the generalizability of our findings to the overall population of patients with TOF/MAPCAs. There are significant limitations to using PA indices in patients with TOF/MAPCAs.[4,13] In a minority of cases, there was a degree of subjectivity between what was considered in the modified Nakata Index versus the MAPCA index, but all important cross-sectional PA area was included in the TNPAI, and the fact that these measurements had no influence on surgical decisions increases their power as a retrospective tool. The high repair rate and generally low RV:Ao in this study limits the ability to detect outcome differences between anatomic subtypes, which might be revealed in a larger cohort. There are other anatomic features, particularly the quality of the distal vasculature, that we could not systemically characterize. Therefore, though there are no statistically differences in early outcomes in the majority of analyses, it is possible that other anatomic features may be related to early outcomes in a more complex manner than this analysis could detect. Furthermore, this study was focused on characterization of pulmonary vascular supply, and was limited to early outcomes. Assessment of longer-term outcomes according to detailed PA anatomy is the subject of ongoing study.

CONCLUSIONS

In TOF/MAPCAs, the distribution of MAPCAs is highly variable and essentially unique for each patient. Though each pulmonary segment can be supplied

by a MAPCA, central PA, or both, in our analysis the type of supply was not associated with early surgical outcome, and all anatomic combinations were conducive to a good repair. Of the variables we studied, total cross-sectional area of central PA and MAPCA material (TNPAI) was an important driver of outcome, with those having a paucity of total material being the most challenging patients to manage. We highlighted a number of novel associations between anatomic features as well as genetic associations, but ultimately the extreme variability of the pulmonary circulation makes a granular classification scheme unrealistic. Nevertheless, grouping patients according to PA arborization pattern has clinical utility with regard to the preoperative evaluation and surgical management.

ARTICLE INFORMATION

Received June 9, 2020; accepted October 26, 2020.

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Sources of Funding

None.

Disclosures

None.

Supplementary Material

Data S1
Tables S1–S3
Figure S1

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Supplemental Material

Data S1.

Supplemental Methods

Surgical Approach:

Our programmatic management algorithm for TOF/MAPCAs aims for early complete unifocalization and intracardiac repair incorporating all lung segments, with augmentation of PA branches (whether arborizing from the central PAs or supplied by MAPCAs) down to the segmental level. The majority of patients (i.e. those with incompletely arborizing or absent central PAs) are scheduled for elective surgery at 4-6 months of age. Unifocalization is individualized based on the anatomy of the PAs and the MAPCAs, and the angiograms are displayed in the operating room as a roadmap. Distal branches are probed to assess diameter, orientation, and the presence of stenoses, and MAPCAs are dissected out to the segmental level to maximize vessel length. To minimize the likelihood of recurrent stenosis, native tissue is utilized for all anastomoses and PA reconstructions when possible. Patch augmentations are performed with PA homograft, and circumferential patches are avoided in order to preserve the growth of native tissue. Anastomoses between MAPCAs and central PAs are performed with extended side-to-side connections, typically to the posterior aspect of the augmented central PAs. These anastomoses extend into the lung parenchyma where collateral vessels generally are no longer distinguishable from normally arborizing PAs[1].

MAPCA naming system

Methodology:

As described in the manuscript, each MAPCA has an origin (subclavian artery (SCA), transverse arch (TVA), descending thoracic aorta (DTA), abdominal aorta (ABA), or coronary artery (CA)) and supply type (single-supply (SS), isolated supply to central PAs (CSS), dual-supply (DS), or mixed-supply(MS)), and can fully or partially supply 1 to 18 bronchopulmonary segments. Additionally, there are unilateral patent ductus arteriosus (PDA) and anomalous pulmonary arteries arising from the ascending aorta (HT).

Each MAPCA was assigned names based on identifiers of origin, supply type, and segmental supply. There were two names assigned – one based only on MAPCA type and segmental supply, and one based on all three: origin, supply type, and segmental supply. As examples, a single-supply MAPCA arising from the descending thoracic aorta supplying 8 segments was named SS-8 (Type-#Segments) or DTA-SS-8 (Origin-Type-#Segments). A dual-supply MAPCA from the descending thoracic aorta that connects to a normally arborizing PA system was named DS-18 or DTA-DS-18.

Each patient was then coded by stringing together the MAPCA names. For each patient, MAPCAs were listed in order of largest number of segments supplied. For example, a patient with 3 MAPCAs (DTA-SS-5, DTA-SS-7, and DTA-CSS-6) would be named “DTA-SS-7,DTA-CSS-6,DTA-SS-5”. If the number of segments in two MAPCAs were identical, the order was alphabetical.

Results:

A total of 1,068 MAPCAs, 26 PDAs, and 5 HTs were included. By the more basic system (Type-#Segments), there were 57 different MAPCA names, and by Origin-Type-#Segments there were 113 different MAPCA names. By Type-#Segments, out of 276 patients there were 176 different patient codes, 124 (70%) of which occurred only once. By Origin-Type-#Segments, out of 276 patients there were 222 patient codes, 187 (84%) of which occurred only once.

Patient naming by Origin-Type-#Segments is shown below:

# of Patients	Patient Type by MAPCA Origin-Type-#Segments
7	DTA-DS-18,DTA-DS-18
5	DTA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18
4	ABA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18,RSCA-DS-18
4	DTA-DS-18,DTA-DS-18,DTA-DS-18
4	DTA-DS-18,DTA-DS-18,RSCA-DS-18
3	DTA-CSS-13,DTA-SS-3,DTA-SS-2
3	DTA-SS-10,DTA-SS-8
3	DTA-SS-10,PDA-8
3	DTA-SS-8,DTA-SS-7,DTA-SS-3
3	DTA-SS-8,DTA-SS-8,DTA-SS-2
2	DTA-CSS-10,DTA-SS-4,DTA-SS-4
2	DTA-CSS-10,PDA-8
2	DTA-CSS-11,DTA-SS-3,DTA-SS-2,DTA-SS-2
2	DTA-CSS-12,DTA-SS-3,DTA-SS-2,DTA-SS-1
2	DTA-CSS-12,DTA-SS-5,DTA-SS-1
2	DTA-CSS-15,DTA-SS-2,DTA-SS-1
2	DTA-CSS-16,DTA-SS-2
2	DTA-CSS-7,DTA-SS-4,DTA-SS-3,DTA-SS-3,DTA-SS-1
2	DTA-CSS-9,DTA-SS-3,DTA-SS-3,DTA-SS-2,DTA-SS-1
2	DTA-DS-10,DTA-DS-10,DTA-SS-7,DTA-SS-1
2	DTA-DS-12,DTA-DS-12,DTA-DS-12,DTA-SS-4,DTA-SS-2
2	DTA-DS-15,DTA-DS-15,DTA-SS-2,DTA-SS-1
2	DTA-DS-15,DTA-DS-15,DTA-SS-3
2	DTA-DS-16,DTA-DS-16,DTA-DS-16,DTA-SS-1,DTA-SS-1
2	DTA-DS-16,DTA-DS-16,DTA-SS-2
2	DTA-DS-17,DTA-DS-17,DTA-DS-17,DTA-SS-1
2	DTA-DS-17,DTA-DS-17,DTA-SS-1
2	DTA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18,RSCA-DS-18
2	DTA-DS-18,DTA-DS-18,LSCA-DS-18,RSCA-DS-18
2	DTA-MS-17,DTA-SS-1
2	DTA-SS-14,DTA-SS-2,DTA-SS-1,DTA-SS-1
2	DTA-SS-8,DTA-SS-6,DTA-SS-3,DTA-SS-1
2	DTA-SS-9,DTA-SS-8,DTA-SS-1
2	DTA-SS-9,PDA-8,DTA-SS-1
2	PDA-8,DTA-SS-6,DTA-SS-4
1	ABA-DS-10,DTA-DS-10,DTA-DS-10,DTA-DS-10,RSCA-DS-10,PDA-8
1	ABA-DS-10,DTA-DS-10,DTA-DS-10,RSCA-DS-10,PDA-8
1	ABA-DS-10,DTA-DS-10,RSCA-DS-10,TVA-DS-10,HT-8
1	ABA-DS-18,ABA-DS-18,CA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18,RSCA-DS-18

1	ABA-DS-18,ABA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18,RSCA-DS-18,RSCA-DS-18
1	ABA-DS-18,ABA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18,RSCA-DS-18
1	ABA-DS-18,ABA-DS-18,DTA-DS-18,DTA-DS-18,RSCA-DS-18
1	ABA-DS-18,CA-DS-18,LSCA-DS-18,RSCA-DS-18
1	ABA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18
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1	ABA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,RSCA-DS-18
1	ABA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18,RSCA-DS-18,RSCA-DS-18,TVA-DS-18
1	ABA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18
1	ABA-DS-18,DTA-DS-18,LSCA-DS-18,LSCA-DS-18,LSCA-DS-18,RSCA-DS-18
1	ABA-DS-8,DTA-DS-8,DTA-DS-8,DTA-DS-8,DTA-SS-8,RSCA-DS-8,RSCA-SS-2
1	CA-CSS-8,DTA-SS-5,DTA-SS-2,DTA-SS-2,DTA-SS-1
1	CA-CSS-8,PDA-8,DTA-SS-1,RSCA-SS-1
1	CA-DS-10,DTA-DS-10,DTA-DS-10,RSCA-DS-10,DTA-SS-4,RSCA-SS-4
1	CA-DS-18,DTA-DS-18,DTA-DS-18
1	CA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18
1	CA-DS-9,DTA-DS-9,DTA-SS-6,DTA-SS-3
1	DTA-CSS-10,DTA-SS-3,RSCA-SS-3,DTA-SS-2
1	DTA-CSS-10,DTA-SS-8
1	DTA-CSS-11,DTA-SS-3,DTA-SS-2,DTA-SS-1,DTA-SS-1
1	DTA-CSS-11,DTA-SS-3,RSCA-SS-2,DTA-SS-1,DTA-SS-1
1	DTA-CSS-11,DTA-SS-4,DTA-SS-2,DTA-SS-1
1	DTA-CSS-11,DTA-SS-4,DTA-SS-3
1	DTA-CSS-11,DTA-SS-4,LSCA-SS-2,DTA-SS-1
1	DTA-CSS-11,DTA-SS-5,DTA-SS-2
1	DTA-CSS-11,DTA-SS-7
1	DTA-CSS-11,RSCA-SS-4,DTA-SS-3
1	DTA-CSS-13,TVA-SS-3,DTA-SS-2
1	DTA-CSS-14,DTA-SS-1,DTA-SS-1,DTA-SS-1,RSCA-SS-1
1	DTA-CSS-14,DTA-SS-3,DTA-SS-1
1	DTA-CSS-15,RSCA-SS-2,RSCA-SS-1
1	DTA-CSS-17,DTA-SS-1
1	DTA-CSS-7,DTA-SS-3,LSCA-SS-3,DTA-SS-2,DTA-SS-2,DTA-SS-1
1	DTA-CSS-6,DTA-SS-4,DTA-SS-4,DTA-SS-3,DTA-SS-1
1	DTA-CSS-6,DTA-SS-5,DTA-SS-4,DTA-SS-2,DTA-SS-1
1	DTA-CSS-6,DTA-SS-5,LSCA-SS-3,DTA-SS-2,DTA-SS-2
1	DTA-CSS-7 DTA-SS-5,DTA-SS-4,DTA-SS-1, RSCA-SS-1
1	DTA-CSS-7,DTA-SS-4,DTA-SS-3,DTA-SS-2,RSCA-SS-2
1	DTA-CSS-7,DTA-SS-5,DTA-SS-4,LSCA-SS-2
1	DTA-CSS-7,DTA-SS-5,RSCA-SS-5,ABA-SS-1
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1	DTA-CSS-7,DTA-SS-6,DTA-SS-3,DTA-SS-2
1	DTA-CSS-7,DTA-SS-6,DTA-SS-3,TVA-SS-2
1	DTA-CSS-7,DTA-SS-7,DTA-SS-4
1	DTA-CSS-7,LSCA-SS-6,DTA-SS-2,DTA-SS-2,DTA-SS-1
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1	DTA-DS-10,DTA-DS-10,DTA-DS-10,DTA-SS-4,DTA-SS-2,LSCA-SS-2
1	DTA-DS-10,DTA-DS-10,DTA-DS-10,PDA-8

1	DTA-DS-10,DTA-DS-10,DTA-SS-3,DTA-SS-2,LSCA-SS-2,DTA-SS-1
1	DTA-DS-11,DTA-DS-11,LSCA-SS-4,DTA-SS-3,DTA-SS-1
1	DTA-DS-12,DTA-DS-12,DTA-SS-3,DTA-SS-3
1	DTA-DS-12,DTA-DS-12,DTA-SS-4,DTA-SS-2
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1	DTA-DS-13,DTA-DS-13,RSCA-SS-4,DTA-SS-1
1	DTA-DS-13,LSCA-DS-13,DTA-SS-3,DTA-SS-2
1	DTA-DS-14,DTA-DS-14,TVA-SS-3,DTA-SS-1
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1	DTA-CSS-18
1	DTA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18,RSCA-DS-18
1	DTA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18,LSCA-DS-18,RSCA-DS-18
1	DTA-DS-18,DTA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18,RSCA-DS-18
1	DTA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18
1	DTA-DS-18,DTA-DS-18,DTA-DS-18,LSCA-DS-18,RSCA-DS-18,TVA-DS-18
1	DTA-DS-18,DTA-DS-18,DTA-DS-18,RSCA-DS-18
1	DTA-DS-7,DTA-DS-7,DTA-SS-6,RSCA-SS-2,DTA-SS-1,DTA-SS-1
1	DTA-DS-8,DTA-DS-8,DTA-SS-5,DTA-SS-2,DTA-SS-2,DTA-SS-1
1	DTA-DS-8,DTA-DS-8,LSCA-DS-8,DTA-SS-2,DTA-SS-1,DTA-SS-1,DTA-SS-1,DTA-SS-1
1	DTA-DS-9,DTA-DS-9,DTA-SS-9
1	DTA-DS-9,DTA-DS-9,PDA-8,LSCA-SS-1
1	DTA-DS-9,RSCA-DS-9,DTA-SS-5,DTA-SS-4
1	DTA-MS-12,DTA-DS-10,DTA-SS-4,DTA-SS-2
1	DTA-MS-14,DTA-DS-13,LSCA-SS-4
1	DTA-MS-14,DTA-DS-9,DTA-SS-3,DTA-SS-1
1	DTA-MS-15,DTA-DS-11,LSCA-SS-2,LSCA-SS-1
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1	DTA-MS-15,DTA-DS-14,DTA-SS-1,DTA-SS-1
1	DTA-MS-15,DTA-DS-14,TVA-DS-14,DTA-SS-2,DTA-SS-1
1	DTA-MS-15,DTA-MS-14
1	DTA-MS-16,DTA-DS-14,DTA-DS-14,LSCA-SS-2
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1	DTA-MS-9,DTA-SS-9
1	DTA-SS-10,DTA-SS-4,LSCA-SS-3,DTA-SS-1
1	DTA-SS-10,DTA-SS-5,DTA-SS-1,TVA-SS-1
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1	DTA-SS-13,DTA-SS-2,DTA-SS-2,DTA-SS-1
1	DTA-SS-4,DTA-SS-3,DTA-SS-3,DTA-SS-2,DTA-SS-2,TVA-SS-2,CA-CSS-1,DTA-SS-1
1	DTA-SS-5,DTA-SS-3,DTA-SS-3,DTA-SS-3,DTA-SS-2,DTA-SS-1
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1	DTA-SS-5,DTA-SS-5,DTA-SS-5,DTA-SS-3

1	DTA-SS-5,LSCA-CSS-5,DTA-SS-4,DTA-SS-3,DTA-SS-1
1	DTA-SS-6,DTA-SS-5,DTA-SS-3,DTA-SS-2,DTA-SS-2
1	DTA-SS-6,DTA-SS-5,TVA-SS-3,DTA-SS-2,DTA-SS-2
1	DTA-SS-6,DTA-SS-6,DTA-SS-3,DTA-SS-2,DTA-SS-1
1	DTA-SS-7,DTA-CSS-4,DTA-SS-3,LSCA-SS-2,DTA-SS-1,LSCA-SS-1
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1	DTA-SS-7,DTA-SS-5,LSCA-SS-3,RSCA-SS-3
1	DTA-SS-7,DTA-SS-6,DTA-SS-4,DTA-SS-3
1	DTA-SS-7,DTA-SS-7,DTA-SS-2,DTA-SS-2
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1	DTA-SS-7,LSCA-SS-6,DTA-CSS-5
1	DTA-SS-8,CA-DS-7,TVA-DS-7,LSCA-SS-2,RSCA-SS-1
1	DTA-SS-8,DTA-CSS-7,DTA-SS-2,DTA-SS-1
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1	DTA-SS-8,DTA-SS-5,DTA-CSS-4,LSCA-SS-1
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1	HT-10,DTA-SS-3,RSCA-SS-2,DTA-SS-1,DTA-SS-1,RSCA-SS-1
1	HT-8,CA-CSS-5,DTA-SS-3,DTA-SS-2
1	HT-8,DTA-SS-4,DTA-SS-3,DTA-SS-2,DTA-SS-1
1	HT-8,DTA-SS-7,DTA-SS-3
1	LSCA-CSS-7,DTA-SS-5,DTA-SS-3,DTA-SS-3
1	LSCA-MS-15,DTA-DS-14,DTA-DS-14,LSCA-SS-2,DTA-SS-1
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1	LSCA-MS-18,RSCA-DS-15
1	LSCA-SS-10,DTA-SS-8
1	LSCA-SS-9,DTA-SS-8,DTA-SS-1
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1	PDA-8,CA-DS-4,DTA-DS-4,DTA-SS-3,DTA-SS-2,DTA-SS-1
1	PDA-8,DTA-CSS-6,DTA-SS-4
1	PDA-8,DTA-CSS-7,RSCA-SS-2,DTA-SS-1
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1	PDA-8,DTA-SS-4,RSCA-SS-4,CA-CSS-2
1	PDA-8,DTA-SS-5,DTA-SS-4,DTA-SS-1
1	PDA-8,DTA-SS-5,RSCA-CSS-3,DTA-SS-2
1	PDA-8,DTA-SS-7,DTA-SS-3
1	PDA-8,RSCA-SS-5,DTA-SS-3,DTA-SS-1,RSCA-SS-1
1	RSCA-CSS-10,TVA-SS-6,DTA-SS-2
1	RSCA-CSS-12,DTA-SS-6
1	RSCA-CSS-7,LSCA-SS-5,DTA-SS-4,DTA-SS-2
1	TVA-SS-9,DTA-CSS-4,DTA-SS-4,DTA-SS-1

Table S1. Classification of MAPCAs and unilateral branch PAs.

MAPCA Number	Number of separate MAPCA origins from the systemic circulation
MAPCA Origin	Left or right subclavian artery (including branches thereof) Transverse aortic arch Descending thoracic aorta Abdominal aorta Coronary artery
MAPCA Course	Leftward (into the left PA at the hilum or lobar or segmental branches) Rightward (into the right PA at the hilum or lobar or segmental branches) Midline (into the intrapericardial PA)
Nature of MAPCA supply	Single supply (no connection to the central PAs) Isolated supply to central PA (only MAPCA connecting to the central PAs) Dual supply (1 of at least 2 MAPCAs connecting to the central PAs) Mixed supply (1 or more single-supply and 1 or more dual-supply branches)
MAPCA segmental supply	Specific lung segments (out of 18) supplied by each MAPCA, either in isolation (single-supply) or via the central PAs (dual-supply)
Unilateral PDA	Arising from a typical PDA location and supplying a normally arborizing unilateral PA, with no MAPCAs to that lung and no intrapericardial PA
Anomalous branch PA	Arising from the ascending aorta, supplying a normally arborizing branch PA, with no MAPCAs to that lung

MAPCA: Major aortopulmonary collateral artery; PA: Pulmonary artery; PDA: Ductus arteriosus

Table S2. Relationship of MAPCA origin to aortic arch sidedness and branching.

	Thoracic aorta MAPCAs	Thoracic aorta MAPCAs only	Subclavian MAPCAs	Abdominal MAPCAs	Coronary MAPCAs	Transverse arch MAPCAs	RSCA & LSCA MAPCAs
All Patients (n=276)	274 (99%)	153 (55%)	105 (38%)	20 (7%)	15 (5%)	15 (5%)	26 (9%)
Arch sidedness							
Left (n=147, 53%)	146 (53%)	78 (51%)	61 (58%)	12 (60%)	8 (53%)	10 (67%)	17 (65%)
Right (n=129, 47%)	128 (47%)	75 (49%)	44 (42%)	8 (40%)	7 (47%)	5 (33%)	9 (35%)
p-value	>0.99	0.47	0.22	0.64	>0.99	0.43	0.22
Arch branching							
Normal (n=232, 84%)	231 (84%)	130 (85%)	85 (81%)	17 (85%)	12 (80%)	14 (93%)	21 (81%)
Abnormal (n=44, 16%)	43 (16%)	23 (15%)	20 (19%)	3 (15%)	3 (20%)	1 (7%)	5 (19%)
p-value	0.29	0.74	0.31	>0.99	0.71	0.48	0.58

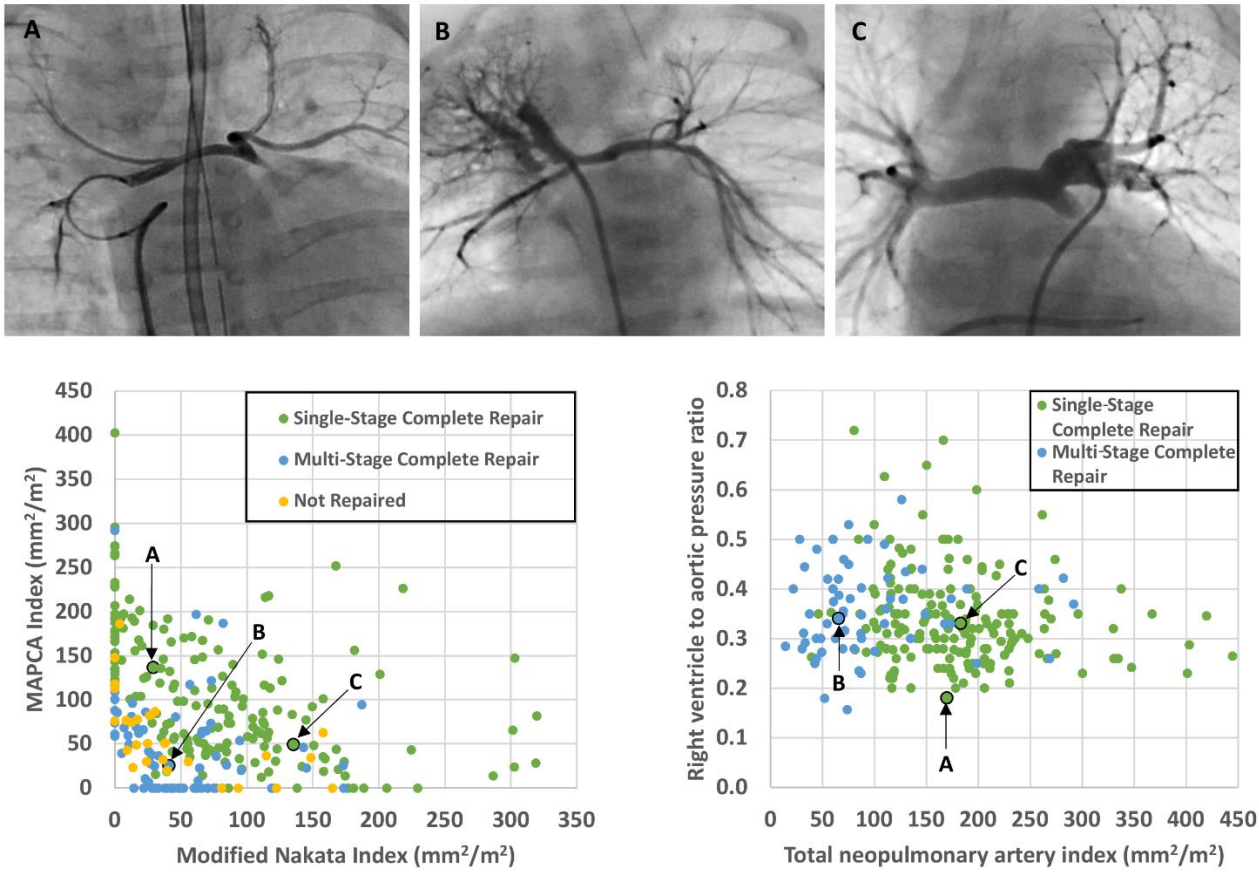
Data presented as n (%). MAPCA: Major aortopulmonary collateral artery, RSCA: Right subclavian artery, LSCA: Left subclavian artery.

Table S3. Relationship between arch sidedness and branching and the presence of MAPCAs from the subclavian arteries.

	Right arch (n=129)	Left arch (n=147)	p-value	Aberrant left SCA (n=35)	Aberrant right SCA (n=9)	p-value
Left SCA MAPCA only	30 (23%)	8 (5%)	<0.001	10 (29%)	1 (11%)	0.22
Right SCA MAPCA only	5 (4%)	36 (24%)	<0.001	1 (3%)	3 (33%)	0.008
Bilateral SCA MAPCAs	9 (7%)	17 (12%)	0.27	4 (11%)	1 (11%)	0.90
No SCA MAPCAs	85 (66%)	86 (59%)	0.26	20 (57%)	4 (44%)	0.34

Data presented as n (%). SCA=subclavian artery; MAPCA=Major aortopulmonary collateral artery

Figure S1. Summary figure.



Left scatterplot: 249 of 276 (90%) patients underwent complete repair, either in 1 (green) or ≥2 (blue) surgeries, and unrepaired (yellow) patients clustered to the left-lower corner, with a paucity of both cross-sectional central PA (modified Nakata index) and MAPCA (MAPCA-index) material. Right scatterplot: Of 249 repaired patients, 182 (73%) underwent repair in 1-stage, and patients that required ≥2 surgeries clustered to the left (blue), with less total cross-sectional PA and MAPCA (TNPAI) material. Angiograms of 3 patients with incompletely arborizing central PAs are shown, with their corresponding positions on the scatterplots noted. Patients A and B had hypoplastic central PAs (low modified Nakata Index). Patient A had a large MAPCA index and underwent single-stage repair, whereas patient B had hypoplastic single-supply MAPCAs and was first palliated with a unifocalization to a central shunt.