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# Correlating computed tomographic angiography of pulmonary circulation with clinical course and disease burden in patients with tetralogy of Fallot and pulmonary atresia

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ABSTRACT

*Purpose:* To determine the type of pulmonary circulation (PC) in patients with tetralogy of Fallot (TOF) and pulmonary atresia (TOF-PA) with the use of computed tomographic angiography (CTA), and describe their clinical courses, corrective surgery and disease burden.

*Methods:* 145 patients (median age 4 years, interquartile range: IQR2-8 years) were analyzed for PC which divided into 5 CTA-types and 11 subtypes based on presence of main pulmonary trunk (MPA) and confluent pulmonary arteries (confluence-PAs), presence of ductus arteriosus or major aortopulmonary collateral arteries (MAPCAs), respectively. Pulmonary arteries (PAs) were assessed by McGoon ratio and arborization. Corrective surgery or palliative management was recorded by type of PC. Disease burden was calculated as the sum of CTA, diagnostic angiography, and palliative management.

*Results:* The most common (N = 77, 53 %) PC was the presence of MPA with confluent-PAs (type-1) which was encountered mostly in TOF patients, followed by the presence of confluent-PAs with atretic MPA (type-2) (N = 47, 32 %) which found mainly in TOF-PA. McGoon ratio in type-1 (2.44 ± 0.84) was significantly larger than type-2 (1.61 ± 0.61) (median difference 0.84, 95 %CI 0.56–1.11, $p < 0.001$). Almost 2/3 of patients in type-1 (71 %) and 1/3 of patients in type-2 (34 %) achieved corrective surgery. There was no significant difference in amount of disease burden among the different PC, with the median value of 3 (IQR1-4).

*Conclusions:* Types of PC allow suggestions for size and arborization of PAs and successful surgical correction with an inverse relationship with the numbers of MAPCAs. There is no significant difference in amount of disease burden among the types of PC.

## 1. Introduction

Tetralogy of Fallot (TOF) is one of the most common cyanotic congenital heart diseases and has a wide spectrum of pathology [1,2]. The most severe form of TOF is with pulmonary atresia (TOF-PA) [3]. Patients with TOF-PA develop major aortopulmonary collateral arteries (MAPCAs) in utero as a consequence of absence of antegrade pulmonary blood flow [3]. Embryology suggests that MAPCAs are congenital systemic arterial collaterals originating from the persistent intersegmental arteries which should normally regress once the connection between the central and intraparenchymal pulmonary arteries (PAs) has been established [3,4]. Approximately 85 % of MAPCAs are found in patients with TOF-PA [5]. MAPCAs are also developed in TOF. Chronic hypoxia and high hemoglobin levels may contribute to the development of

*Abbreviations:* CTA, computed tomographic angiography; DORV, double outlet right ventricle; LPA, left pulmonary artery; MPA, main pulmonary trunk; MAPCAs, major aortopulmonary collateral arteries; MRA, magnetic resonance angiography; PAs, pulmonary arteries; PC, pulmonary circulation; RPA, right pulmonary artery; TOF, tetralogy of Fallot; TOF-PA, tetralogy of Fallot with pulmonary atresia.

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MAPCAs in patients with TOF [6].

Pulmonary circulation in patients with TOF and TOF-PA is derived from PAs and/ or MAPCAs. The native PAs can vary from mild hypoplasia to absence of a branch or all PAs, and from confluent to non-confluent PAs [5,7]. MAPCAs could be an additional source of pulmonary perfusion aside from the native PAs. In some situations, pulmonary circulation is exclusively dependent on MAPCAs [3]. As a result of the anatomical heterogeneity of pulmonary circulation and essentially uniqueness of each patient, Barbero-Marcial's classification based on the anatomy of PAs and MAPCAs, has been proposed to facilitate the risk stratification for patients with pulmonary atresia [8]. The pulmonary circulation is divided into types A, B, and C as follows: in (A) all the bronchopulmonary segments are connected to the central PAs, in (B) some bronchopulmonary segments are supplied by branches of PAs whereas other segments are supplied by MAPCAs, and in (C) all bronchopulmonary segments are supplied exclusively by MPACAs.

Computed tomographic angiography (CTA) has been considered an accurate diagnostic modality for the assessment of MAPCAs [3,9–11]. This non-invasive diagnostic tool aids in reducing the number of catheter angiographies [9] and lowering radiation dose to the patients [10].

## 2. Purpose

Since pulmonary circulation in patients with TOF and pulmonary atresia is extremely complex, the management approach is based on an individual's anatomical and hemodynamic data [12–14]. Our study seeks to determine various types of pulmonary circulation assessed by CTA and to determine the clinical course, rate of achievable definitive surgical repairs, and disease burden to the patients according to each type of pulmonary circulation.

## 3. Materials and methods

### 3.1. Study population

This was a single-center, retrospective study, that included patients with a diagnosis of TOF, TOF-PA, double outlet right ventricle with TOF subtype (DORV-TOF), and DORV with pulmonary atresia subtype (DORV-PA) who had CTA at our tertiary care radiology department between 2014 and 2020. An initial 231 patients were identified. The exclusion criteria were patients with the aforementioned diagnoses but associated with other complex congenital heart disease ($N = 61$), patients who had corrective surgical repairs prior to CTA examination ($N = 24$), and cases with non-diagnostic CTA image quality ($N = 1$). The excluded 61 patients with complex congenital heart disease were as follows: DORV with transposition of the great artery and pulmonary atresia ($N = 23$), single ventricle with pulmonary atresia ($N = 13$), complex congenital heart disease with abnormal situs and pulmonary atresia ($N = 21$), and others (pulmonary atresia with total anomalous pulmonary venous return, pulmonary atresia with ectopic cordis and in conjoined twin) ($N = 4$). After the exclusion of 86 patients, a total of 145 patients were included in the analysis. The study was approved by the Institutional Human Research Ethics Committee.

### 3.2. CTA protocols and image acquisitions

CTA examinations were performed with one of the following three models: between 2006 and 2009, a 64-multidetector CT (MDCT) (Somatom Sensation 64, Siemens Medical Systems, Erlangen, Germany), between 2009 and 2019, a 320-MDCT (Aquilion one, Toshiba Medical Systems, Tochiki-ken, Japan), and from 2019 to 2020, a 256-slice dual energy MDCT (Revolution CT, General Electric Healthcare, Chicago, USA). All CTA studies were performed without electrocardiogram synchronous (ECG). Prospective ECG-triggered CTA was used only if the coronary origins were not sufficiently identified or a coronary anomaly was suspected by echocardiography. The imaging protocols for the 64-MDCT were as follows: tube voltage 80 kV and tube current 120−350 mA based on patient's weight, collimation 64 × 0.625 mm, rotation time 0.33 s, pitch 1.4–1.5 for pediatrics and 1.2–1.3 for adults, and reconstruction thickness/interval 1.0 mm/0.8 mm. The scan protocols for the 320-MDCT were as follows: volume mode, tube voltage 80 kV and tube current 120−350 mA based on patient's weight, collimation 320 × 0.5 mm, rotation time 0.33 s, reconstruction thickness/interval 1.0 mm/0.5 mm. For 256-slice dual energy MDCT, the images were acquired by using tube voltage at 70−80 kV and tube current set at 120−350 mA based on patient's weight, collimation 256 × 0.625 mm, rotation time 0.28 s, reconstruction thickness 0.625 mm continuous. Six patients with a body weight over 40 kg were scanned with a 100 kV. General anesthesia or sedation by an anesthesiologist was required for children under 10 years of age. The coverage of the scan included the subclavian artery to 1-cm below the cardiac apex.

Intravenous non-ionic low-osmolar contrast medium (Ultravist 300 mgI/mL, Bayer Schering Pharma, Berlin, Germany) was administered with a volume of 1.0−1.2 mL/kg body weight and at a flow rate of 1−3 ml/s for pediatrics and of 3−4 ml/s for adults, followed by 20−30 mL of saline solution. A time density analysis was initiated prior to diagnostic scanning and data acquisition. For the 64-and 320-MDCT, two regions of interest were placed at the pulmonary artery and proximal descending aorta. In the case of absence of pulmonary artery, the region of interest was placed only at the proximal descending aorta. For the 256-slice dual energy MDCT, a region of interest was placed at the proximal descending aorta. The software performed a repetitive scan on an axial slice of the preselected aortic or pulmonary artery level. Data collection was initiated when the contrast enhancement achieved the preset threshold contrast medium opacification.

### 3.3. CTA image analysis

Post-processing image reconstruction was performed on an external workstation; Syngo.via for Siemens Medical Solutions for 64-MDCT, Vitrea fx version 3.0.1 for Toshiba Medical Systems for 320-MDCT; and AW server 3.2 (Ext. 3.2) for 256-slice dual energy MDCT. Images were reconstructed in coronal, sagittal, left and right anterior oblique planes, curve multiplanar reformation, maximum intensity projection, and volume-rendered images, and transferred to the Picture Archiving and Communications System (PACS) at the Department of Radiology, using a DICOM Conformance (Synapse version 4.2, FUJIFILM Medical Systems USA's Synapse® PACS System, USA). Analyses were performed using a combination of these images.

We applied Barbero-Marcial's classification [8] and categorized pulmonary circulation into 5 major CTA types based on presence or absence of main pulmonary trunk or native PAs and either confluence or non-confluence between RPA and LPA as follows: type 1 is characterized by presence of the main pulmonary trunk (MPA) and confluence between the right and the left pulmonary arteries (RPA and LPA); type 2, absence of MPA and confluence between the RPA and LPA; type 3, non-confluence between RPA and LPA; type 4, absence of the central PAs and pulmonary blood supply through either the ductus arteriosus or MAPCAs or both; and type 5, unilateral right or unilateral left central PAs (Fig. 1a–e). All types of pulmonary circulation were further subcategorized based on the presence of the ductus arteriosus and MAPCAs. Finally, a total of 11 CTA subtypes of pulmonary circulation were analyzed. Arborization of the PAs was recorded as "complete", when there was normal branching of both lobar and intrapulmonary PAs following all bronchopulmonary segments, and "incomplete" if there was abnormal or incomplete branching of PAs on any side of the lungs. The size of PAs was expressed as a McGoon ratio, calculated by dividing the sum of the diameters of RPA and LPA (measured at the distal part proximal to the bifurcation into lobar branches) by the diameter of the aorta at the level above the diaphragm [15] (Fig. 2).

MAPCAs were analyzed into number and connection with the native PAs. The number of MAPCAs was counted according to the number of

MAPCAs' origins, not all branches of MAPCAs. Dose-length product and date of CTA study were collected from the CTA studies.

### 3.4. Data collection from medical records

Demographic data including gender, age, height, weight, and oxygen saturation was collected. Patients who underwent CTA studies were classified as the "palliative management prior to CTA group", defined as patients who underwent either surgical palliation or interventional palliative management prior to the CTA study, and the "non-palliative management prior to CTA group". Palliative management included: classical or modified Blalock-Taussig shunt, central shunt, Glenn's shunt, vascular angioplasty or stenting, interventional balloon dilatation of the PAs or valves, banding of the PAs, and ligation or coiling of MAPCAs.

The clinical course was recorded as regular follow-up, referred to a local hospital, lost to follow-up, and death. Patients in the regular follow-up group were divided into "achievable with corrective surgery" and "palliative management". Corrective surgical repairs were defined by three surgical modes: 1) total correction by closure of the ventricular septal defect, and augmentation of the right ventricular outflow or pulmonary arteries, 2) Rastelli operation by creating a connection between the right ventricle and pulmonary artery with closure of the ventricular septal defect, and 3) unifocalization [12,16]. The aforementioned information was recorded according to each type of pulmonary circulation.

Disease burden was measured by summation of the 1) number of palliative management visits, 2) number of CTA studies including the initial CTA study, and 3) number of diagnostic catheter angiographies. The cumulative dose-length product from the total CTA studies was recorded.

Materials and methods of the research was summarized in Fig. 3.

### 3.5. Statistical analysis

Statistical analyses were performed using STATA software version 16.0 (Stata Corp, College Drive, Texas, USA). Continuous variables were presented as mean ± standard deviation (SD) or median (interquartile range [IQR], 25th, 75th percentile) and categorical variables were summarized as percentages. Linear regression analysis was used to compare oxygen saturation and size of PAs (McGoon ratio) between patients with and without palliative management prior to CTA study. Statistically significant differences in PAs arborization and number of MAPCAs between pulmonary circulation type 1 and other types were analyzed, using Fisher's exact test. Median regression analysis was used to compare median values of the total number of MAPCAs in each type of pulmonary circulatory. Differences in disease burden among the types of pulmonary circulation were analyzed with Kruskal-Wallis analysis. Radiation exposure among each pulmonary circulation was compared with the use of median regression analysis. A $p$-value of $< 0.05$ was considered statistically significant.

Inter-observer agreement of type of pulmonary circulation and the measurement for the McGoon ratio were assessed using Cohen's Kappa statistic and a Bland-Altman analysis, respectively. Data were displayed as Kappa, and mean ± SD with 95 % confidence interval (CI), respectively.

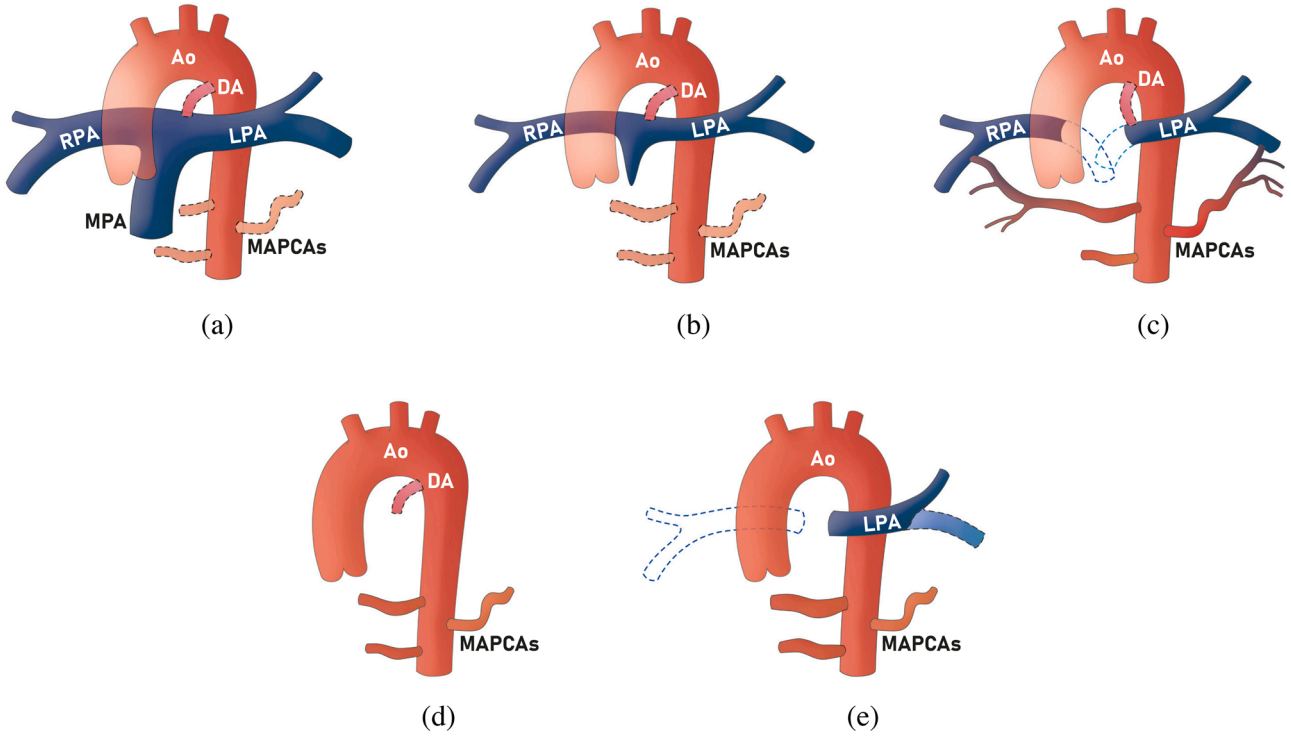


**Fig. 1.** Diagram demonstrates 5 CTA types (11 subtypes) of pulmonary circulation.
(**a**) Type 1-presence of MPA with confluence between RPA and LPA. Subcategorize into 4 subtypes: 1a-abence of both DA and MAPCAs, 1b-presence of DA, 1c-presence of both DA and MAPCAs, and 1d-presence of MAPCAs.
(**b**) Type 2-atretic MPA with confluence between RPA and LPA. Subcategorize into 3 subtypes: 2a-presence of DA, 2b-presence of MAPCAs, and 2c-presence of both DA and MAPCAs.
(**c**) Type 3-non-confluence between RPA and LPA. Subcategorize into 2 subtypes: 3a-presence of MAPCAs, 3b-presence of both DA and MAPCAs.
(**d**) Type 4-absence of central pulmonary arteries.
(**e**) Type 5-unilateral right or unilateral left central pulmonary artery.
Ao = aorta, DA = ductus arteriosus, LPA = left pulmonary artery, MAPCAs = major aortopulmonary collateral arteries, MPA = main pulmonary trunk, RPA = right pulmonary artery.

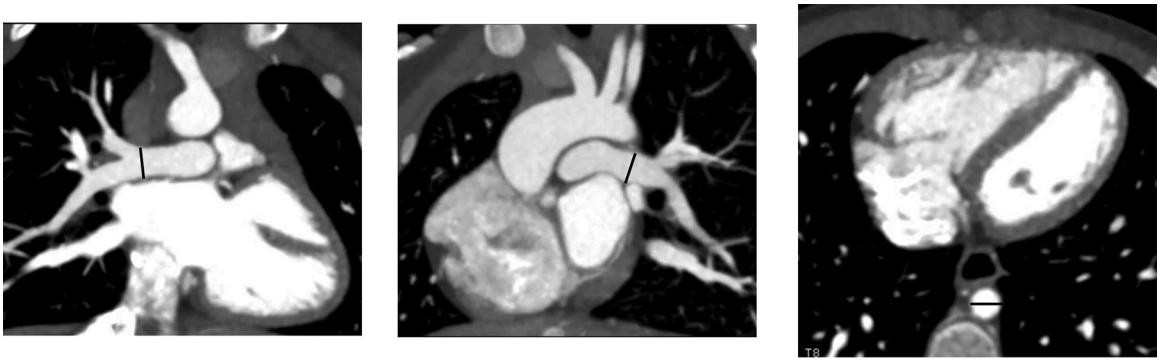


**Fig. 2.** CTA in the right and left oblique views (a-b) and in axial view (c) demonstrating the positions where the right (**a**) and left (**b**) pulmonary arteries, and aorta (**c**) were measured for calculation of the McGoon ratio.

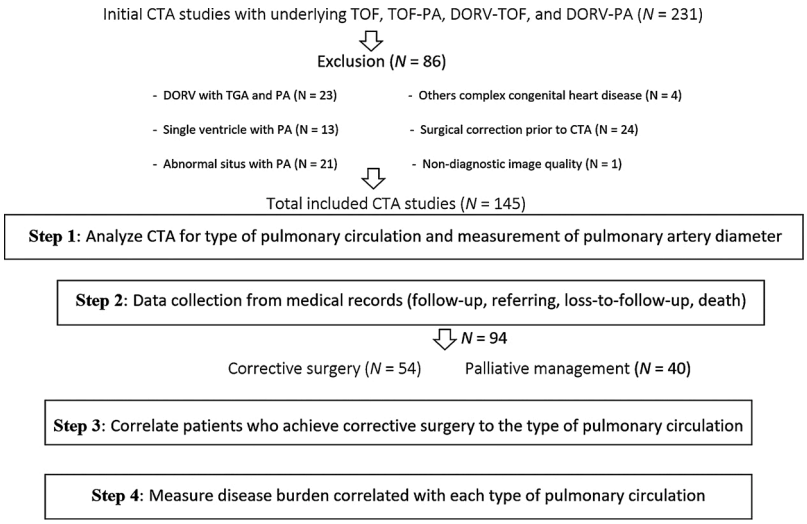


**Fig. 3.** Summary of research methods.

## 4. Results

### 4.1. Demographic data

Demographic data and the clinical diagnosis for each type of pulmonary circulation are shown in Table 1. A majority of patients were pediatric (≤15 years of age) (92 %) with a median age of 3.0 years (IQR 1 month-13 years). The remaining adult patients (7 %) had a median age of 23.0 years (IQR 20-24). There was no significant sex predilection. TOF was diagnosed in 33.8 % of the cases ($N = 49$), TOF-PA in 54.5 % of the cases ($N = 79$), DORV-TOF in 7.6 % of the cases ($N = 11$), and DORV-PA in 4.1 % of the cases ($N = 6$). The total patients with TOF and with pulmonary atresia were $N = 60$ and $N = 85$, respectively. The median dose-length product was 44 mGy*cm (IQR 31-73).

### 4.2. Type of pulmonary circulation, pulmonary arteries and major aortopulmonary collateral arteries

Of 145 patients, 71 (49 %) did not receive palliative management prior to obtaining CTA, while the remaining 74 (51 %) underwent palliative management before CTA. Distribution of the 5 types of pulmonary circulation and oxygen saturation in each circulation type are shown in Table 2. Normal pulmonary circulation (CTA type 1) was encountered in about half of the patients (53 %). Of type I, absence of both ductus arteriosus and MAPCAs occurred in about half (48 %), followed by cases associated with MAPCAs in about 28 %. Confluent PAs with absence of MPA (CTA type 2) was the second most common, accounting for 32 %. About one-third of cases in this group were associated with MAPCAs and without ductus arteriosus. Other types of pulmonary circulation were uncommon, ranging from 3 % to 6 %.

Oxygen saturation data was obtained in 108 (74.5 %) patients, divided into a non-palliative management group (49 patients, 45 %) and a palliative management group (59 patients, 54 %). The median value of oxygen saturation was 82 ± 7.4 %. There was no statistically significant difference in the oxygen saturation between pulmonary circulation type 1 and the other 4 types of cases (who did not receive any palliative management prior to obtaining CTA examination) using linear regression analysis. *P*-values were as follows: comparing type 1 with type 2 (1:2), $P = 0.24$; type 1:3, $P = 0.78$; type 1:4, $P = 0.83$, and type 1:5, $P = 0.10$. There was also no significant difference in oxygen saturation between the non-palliative management group and the palliative management group. The mean difference (95 % CI), and *P*-value were −2.1 % (-4.93 to -0.74), $P = 0.15$. However, the study is limited by a small number of subjects in type 3–5.

Anatomical features of PAs and MAPCAs for each pulmonary circulation are summarized in Table 3. For each individual circulation, linear regression analysis was used to assess differences in pulmonary artery size and numbers of MAPCAs. Type 1 had a significant larger pulmonary artery (McGoon ratio) than type 2, both in the non-palliative management and palliative management prior to CTA groups, with the mean

**Table 1**
Demographic data ($N = 145$).

| | |
|---|---|
| Age at CTA study (years) | 4 (2–8) |
| Sex (female) | 79 (54.5 %) |
| Pediatrics (≤ 15 years) | 134 (92.4 %) |
| Height (cm) | 101.0 ± 29.04 |
| Weight (kg) | 13.3 (10.0–19.5) |
| BSA ($m^2$) | 0.59 (0.49–0.78) |
| Dose-length product (mGy*cm) | 44.0 (31.8–73.6) |

Distribution of diagnosis *N* (%)

| CTA type | TOF | TOF-PA | DORV-TOF | DORV-PA |
|---|---|---|---|---|
| 1 | 46 (59.7 %) | 19 (24.7 %) | 10 (13.0 %) | 2 (2.6 %) |
| 2 | 1 (2.1 %) | 42 (89.4 %) | 0 | 4 (8.5 %) |
| 3 | 1 (14.3 %) | 5 (71.4 %) | 1 (14.3 %) | 0 |
| 4 | 0 | 9 (100.0 %) | 0 | 0 |
| 5 | 1 (20.0 %) | 4 (80.0 %) | 0 | 0 |
| Total | 49 | 79 | 11 | 6 |

Continuous variables presented as mean ± SD and median interquartile range (25th-75th percentile) as appropriate and categorical variables presented as *N* (%).
CTA = computed tomographic angiography, DORV = double outlet right ventricle, TOF = tetralogy of Fallot, PA = pulmonary atresia.

difference (95 % CI), and *P*-value of -1.14 (-1.63,-0.65), $P < 0.001$; and -0.69 (-0.88,-0.29), $P < 0.001$, respectively. Complete arborization of PAs was seen in 104/145 (71 %) patients and typically encountered in type 1 (74/77, 96 %), followed by type 3 (5/7, 71 %). Presence of MAPCAs was found in 86/145 patients (59 %), of which all patients with type 4 and 5 had MAPCAs (100 %). MAPCAs were also commonly encountered in CTA type 3 (6/7 patients, 85 %) and type 2 (37/47, 78 %). Fisher's exact test revealed a significant positive correlation between type of pulmonary circulation and the normal arborization of PAs, as well as the presence or absence of MAPCAs ($P < 0.001$).

Of the 86 MAPCAs, 3 were ligated (three patients in CTA type 1) and excluded from the analysis of whether MAPCAs anastomose to native PAs directly. The median number of MAPCAs was 1 (IQR 0-3). There was a significant difference in the number of MAPCAs as follows: between type 1 and 2, with a median difference of 2 (95 % CI 1.33–2.66), $P < 0.001$, between type 1 and 4-a median difference of 3 (95 % CI 1.73−4.27), $P < 0.001$, and between type 1 and 5-a median difference of 3 (95 % CI 1.34–4.66), $P < 0.001$. There was no statistically significant difference in the number of MAPCAs comparing type 1 with type 3 ($P = 0.17$). In addition, the Kruskal-Wallis test revealed a significant relationship between the type of pulmonary circulation and the total number of MAPCAs ($P < 0.001$).

### 4.3. Clinical course and disease burden

The median follow-up duration was 24 months (IQR 9-40). Clinical courses were as follows: 54 (37.2 %) patients underwent corrective surgery, 40 (27.6 %) patients had been follow-up with palliative management, 31 (21.4 %) patients were referred to other hospitals, 17 (11.7 %) patients were lost to follow-up, and 3 (2.1 %) patients passed away during follow-up (2 died from massive bleeding after corrective surgery and the other died from pulmonary edema after central shunt and LPA plasty).

Patients who have been follow-up (both with corrective surgery and palliative management and mode of surgical correction are summarized in Table 4. Although all patients with type 3 pulmonary circulation achieved surgical correction, the conclusions could not be drawn due to the small number of subjects. Patients with pulmonary circulation type 1 had the best prognosis in terms of successful corrective surgery.

Ninety-four patients were analyzed for disease burden and are summarized in Table 5. Patients with this disease entity had at least one CTA study, with a median of 1 (IQR 1-1). The median number of diagnostic catheter angiography was 0 (IQR 0-1). Patients also had at least one palliative management during follow-up, with a median of 1 (IQR 0-2). There was no statistically significant difference in amount of disease burden among each type of pulmonary circulation by the Kruskal-Wallis test. Median dose-length product of the cumulative CTA studies was 42 mGy*cm (IQR 32-73). There was no statistically significant difference in

**Table 2**
CTA classification of pulmonary circulation and oxygen saturation in each circulatory type ($N = 145$).

| CTA type of pulmonary circulation | Non-palliative *N* (%) | Palliative *N* (%) | Total *N* (%) | Oxygen saturation (%) ($N = 108$) | | |
|---|---|---|---|---|---|---|
| | | | | Non-palliative *N* (%) | Palliative *N* (%) | Total *N* (%) |
| Type 1 Presence of MPA with confluence of RPA and LPA | | | | | | |
| 1a | 17 | 20 | 37 (48.1 %) | | | |
| 1b | 1 | 10 | 11 (14.3 %) | | | |
| 1c | 4 | 3 | 7 (9.1 %) | | | |
| 1d | 10 | 12 | 22 (28.6 %) | $N = 21$ (36.8%) | $N = 36$ (63.2%) | $N = 57$ (52.8%) |
| Subtotal | 32 (41.6 %) | 45 (58.4 %) | **77 (53.1 %)** | 84.8 ± 8.8 | 82.6 ± 6.4 | 83.4 ± 7.3 |
| Type 2 Confluence of RPA and LPA | | | | | | |
| 2a | 3 | 7 | 10 (21.2 %) | | | |
| 2b | 21 | 12 | 33 (70.2 %) | | | |
| 2c | 1 | 3 | 4 (8.5 %) | $N = 17$ (48.6%) | $N = 18$ (51.4%) | $N = 35$ (32.4%) |
| Subtotal | 25 (53.2 %) | 22 (46.8 %) | **47 (32.4 %)** | 81.7 ± 6.8 | 79.4 ± 5.4 | 80.1 ± 6.1 |
| Type 3 Non-confluence of RPA and LPA | | | | | | |
| 3a | 3 | 0 | 3 (42.9 %) | | | |
| 3b | 0 | 4 | 4 (57.1 %) | $N = 2$ (40.0%) | $N = 3$ (60.0%) | $N = 5$ (4.6%) |
| Subtotal | 3 (42.9 %) | 4 (57.1 %) | **7 (4.8 %)** | 86.5 ± 82.1 | 79.3 ± 4.5 | 82.2 ± 5.2 |
| Type 4 Absence of central RPA and LPA | | | | | | |
| Subtotal | 8 (88.9 %) | 1 (11.1 %) | **9 (6.2 %)** | $N = 6$ (100%)<br>84.0 ± 7.5 | –<br>– | $N = 6$ (5.6%)<br>84.0 ± 7.5 |
| Type 5 Unilateral right or unilateral left central pulmonary artery | | | | | | |
| Subtotal | 3 (60.0 %) | 2 (40.0 %) | **5 (3.5 %)** | $N = 3$ (60.0%)<br>76.3 ± 13.2 | $N = 2$ (40.0%)<br>71.5 ± 19.1 | $N = 5$ (4.6%)<br>74.4 ± 13.6 |
| **Total** | 71 (49.0 %) | 74 (51.0 %) | **14 (100 %)** | $N = 49$ (45.4%)<br>83.2 ± 8.1 | $N = 59$ (54.6%)<br>81.1 ± 6.7 | $N = 108$ (74.5%)<br>82.0 ± 7.4 |

Continuous variables presented as mean ± SD and categorical variables presented as *N* (%).
Data of oxygen saturation presented as mean ± SD.
CTA = computed tomographic angiography, LPA = left pulmonary artery, MPA = main pulmonary trunk, RPA = right pulmonary artery.

**Table 3**
CTA findings of pulmonary arteries and major aortopulmonary collateral arteries ($N = 145$).

| CTA type of pulmonary circulation | Pulmonary arteries | | Major aortopulmonary collateral arteries | | |
|---|---|---|---|---|---|
| | Complete arborization of PAs *N* (%) | McGoon ratio (Mean ± SD) | Presence *N* (%) | Total numbers | Anastomose with native PAs *N* (%) |
| Type 1 ($N = 77$) | 74 (96.1 %) | 2.44 ± 0.84<br>Non-Palliative 2.68 ± 1.05<br>Palliative 2.28 ± 0.60 | 29 (37.7 %) | 0 (0–1) | 9 (31.0 %)* |
| Type 2 ($N = 47$) | 22 (46.8 %) | 1.61 ± 0.61<br>Non-Palliative 1.53 ± 0.71<br>Palliative 1.69 ± 0.46 | 37 (78.7 %) | 2 (1–3) | 27 (73.0 %) |
| Type 3 ($N = 7$) | 5 (71.4 %) | 2.02 ± 0.68<br>Non-Palliative 1.79 ± 0.73<br>Palliative 2.19 ± 0.70 | 6 (85.7 %) | 1 (1–3) | 2 (33.3 %) |
| Type 4 ($N = 9$) | N/A | N/A | 9 (100.0 %) | 3 (3–4) | 6 (66.7 %) |
| Type 5 ($N = 5$) | 3 (60.0 %) | N/A | 5 (100.0 %) | 3 (3–6) | 5 (100.0 %) |
| Total ($N = 145$) | 104 (71.7 %) | 2.12 ± 0.85 | 86 (59.3 %) | 1 (0–3) | 49 (57.0 %) |

Continuous variables presented as mean ± SD and median interquartile range (25th-75th percentile) as appropriate and categorical variables presented as *N* (%).
CTA = computed tomographic angiography, N/A = non-assessable, PAs = pulmonary arteries.
* Three patients were excluded from the analysis of whether MAPCAs anastomose with native pulmonary artery due to previous MAPCAs ligation.

**Table 4**
Numbers of patients with corrective surgery and palliative management and mode of surgical correction ($N = 94$).

| CTA type of pulmonary circulation | Corrective surgery *N* (%) | | | | Follow-up with palliative management *N* (%) |
|---|---|---|---|---|---|
| | Total correction | Rastelli operation | Unifocaliza-tion | Total | |
| Type 1 ($N = 53$) | 29 (54.7 %) | 9 (17.0 %) | 0 | 38 (71.7 %) | 15 (28.3 %) |
| Type 2 ($N = 32$) | 0 | 9 (28.1 %) | 2 (6.3 %) | 11 (34.4 %) | 21 (65.6 %) |
| Type 3 ($N = 3$) | 1 (33.3 %) | 2 (66.7 %) | 0 | 3 (100.0 %) | 0 |
| Type 4 ($N = 3$) | 0 | 0 | 1 (33.3 %) | 1 (33.3 %) | 2 (66.7 %) |
| Type 5 ($N = 3$) | 0 | 0 | 1 (33.3 %) | 1 (33.3 %) | 2 (66.7 %) |
| Total ($N = 94$) | 30 (31.9 %) | 20 (21.3 %) | 4 (4.3 %) | 54 (57.4 %) | 40 (42.6 %) |

CTA = computed tomographic angiography.

radiation exposure when compared type I with the type 2 pulmonary circulation (for patients with corrective surgery, $P = 0.458$, and patients with palliative management, $P = 0.090$).

### 4.4. Inter-observer agreement

We randomly selected 36 patients (every 4th patient) to assess inter-observer agreement in measurement of RPA, LPA and descending aorta using Bland-Altman analysis. There were no significant differences between the two observers (mean difference, 95 % CI) with details as follows: at RPA 0.025 (-0.68 to-0.63), at LPA 0.005 (-0.43 to-0.44), and at descending aorta 0.017 (-0.07 to-0.11), respectively (Fig. 4a–c). We observed good inter-observer agreement (94.3 %) in determining CTA type of pulmonary circulation (Kappa 0.82).

**Table 5**
Disease burden to patients according to pulmonary circulation ($N = 94$).

| CTA type | Numbers of patients (*N*) | Numbers of investigations and palliative managements | | | Disease burden | Dose-length product (mGy*cm) |
|---|---|---|---|---|---|---|
| | | CT angiography | Diagnostic angiogram | Palliative management | | |
| Type 1 ($N = 53$) | Correction ($N = 38$) | 1 (1–1) | 0 (0–1) | 1 (0–2) | 2 (1–4) | 45.6 (32.8–66.2) |
| | Palliative ($N = 15$) | 1 (1–1) | 1 (0–1) | 1 (0–2) | 3 (1–4) | 63.2 (37.2–86.8) |
| | Total ($N = 53$) | 1 (1–1) | 0 (0–1) | 1 (0–2) | 2 (1–4) | 48.8 (32.9–73.6) |
| Type 2 ($N = 32$) | Correction ($N = 11$) | 1 (1–1) | 0 (0–1) | 1 (0–2) | 3 (2–4) | 37.7 (29.5–81.0) |
| | Palliative ($N = 21$) | 1 (1–1) | 0 (0–1) | 1 (0–1) | 3 (1–4) | 37.7 (26.7–62.0) |
| | Total ($N = 32$) | 1 (1–1) | 0 (0–1) | 1 (0–2) | 3 (2–3.5) | 37.7 (27.2–69.1) |
| Type 3 ($N = 3$) | Correction ($N = 3$) | 1 (1–1) | 0 (0–1) | 2 (1–2) | 3 (3–3) | 64.5 (32.1–154.9) |
| | Palliative ($N = 0$) | 0 | 0 | 0 | 0 | 0 |
| | Total ($N = 3$) | 1 (1–1) | 0 (0–1) | 1(1–2) | 3 (3–3) | 64.5 (32.1–154.9) |
| Type 4 ($N = 3$) | Correction ($N = 1$) | 1 | 0 | 0 | 1 | 35.0 |
| | Palliative ($N = 2$) | 1 (1–1) | 0.5 (0–1) | 0 | 1.5 (1–2) | 107.8 (33.9–181.6) |
| | Total ($N = 3$) | 1 (1–1) | 0 (0–1) | 0 | 1 (1–2) | 35.0 (33.9, 181.6) |
| Type 5 (= 3) | Correction ($N = 1$) | 2 | 0 | 1 | 3 | 524.6 |
| | Palliative ($N = 2$) | 1 (1–1) | 0 | 1 (0–2) | 2 (1–3) | 48.3 (26.5–70.1) |
| | Total ($N = 3$) | 1 (1–1) | 0 | 1 (0–2) | 3 (1–3) | 70.1 (26.5–524.6) |
| Summaries | | | | | | |
| Surgical correction ($N = 54$) | | 1 (1–1) | 0 (0–1) | 1 (0–2) | 3 (1–4) | 45.6 (32.8–68.0) |
| Palliative management ($N = 40$) | | 1 (1–1) | 0 (0–1) | 1 (0–2) | 2.5 (1.5–3) | 40.0 (27.2–83.8) |
| Total ($N = 94$) | | 1 (1–1) | 0 (0–1) | 1 (0–2) | 3 (1–4) | 42.3 (32.0–73.6) |

Continuous variables presented as median interquartile range (25th-75th percentile) and categorical variables presented as *N*.
CTA = computed tomographic angiography.

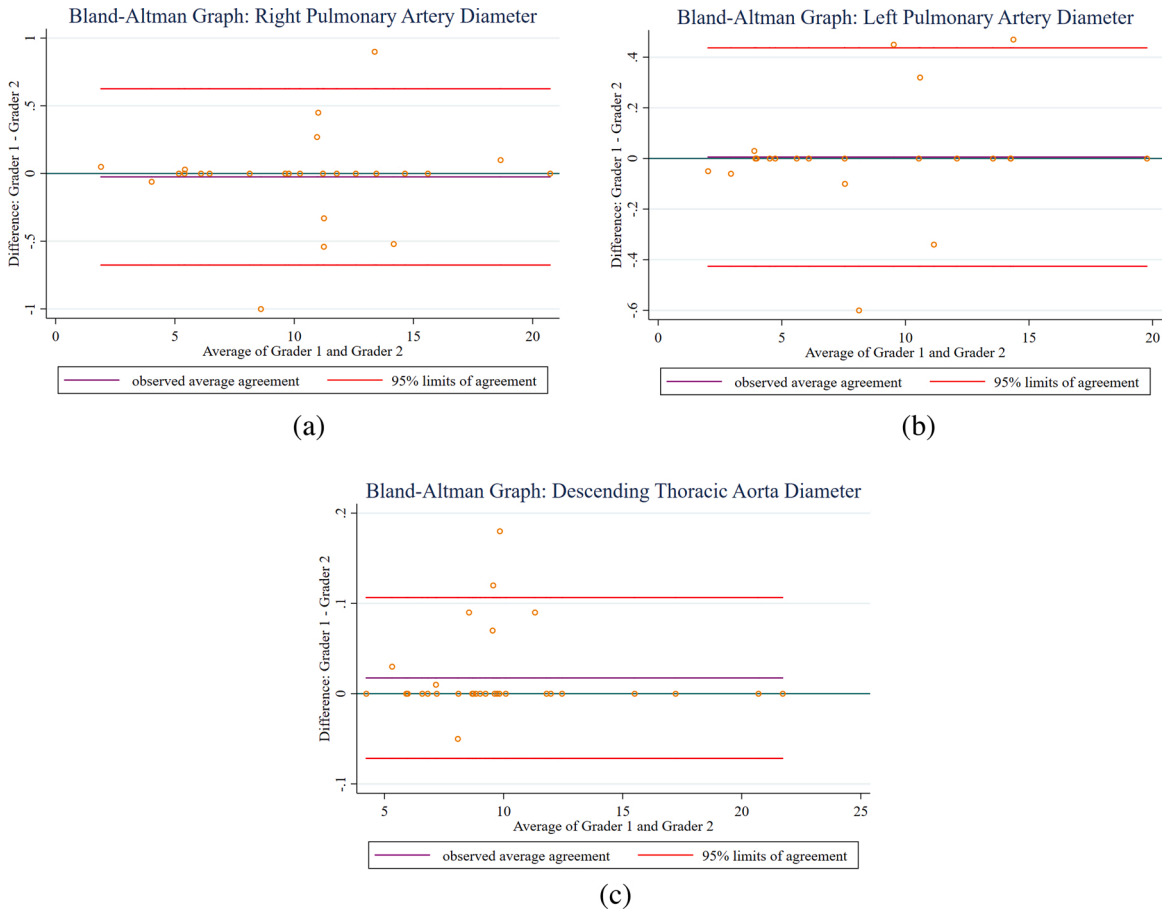


**Fig. 4.** Bland-Altman plots of the inter-observer agreement in the measurements for right pulmonary artery (**a**), left pulmonary artery (**b**), and descending aorta (**c**). The central violet line indicates the mean difference between the two graders. The upper and lower lines represent upper and lower margins of 95 % limits of agreement, respectively.

## 5. Discussion

Pulmonary abnormalities are commonly seen in patients with TOF and pulmonary atresia, leading to impaired pulmonary perfusion, cyanosis and lower oxygen saturation. The mean oxygen saturation was 83 ± 8 %.

### *5.1. CTA type of pulmonary circulation and pulmonary arteries*

The Congenital Heart Surgery Nomenclature and Database Project and the former "Barbero-Marcial's classification" proposed a classification of pulmonary circulation in patients with pulmonary atresia, based on the presence or absence of native PAs and MAPCAs, into three major types as follows: A, B, C- pulmonary circulation derived from only native PAs; from both PAs and MAPCAs; and from only MAPCAs, respectively [8,17]. The "Barbero-Marcial's classification" was subgroup into A1 and A2 based on the normal or under-development of native PAs and non-confluence or stenosis PAs, respectively [8]. However, other important information including complete branching of native PAs and the relation between MAPCAs and native PAs (anastomose with the native PAs) are also fundamental for the management and outcome. Therefore, our study describes pulmonary circulation with the use of CTA in 145 patients, divided into two major disease entities: 1) TOF population (including TOF and DORV-TOF, $N = 60$) and 2) pulmonary atresia population (including TOF-PA and DORV-PA, $N = 85$). The most common type of pulmonary circulation found in our study population and in patients with TOF was type 1-presence of main pulmonary trunk, RPA, and LPA (93 %), and subcategory type 1a-absence of associated ductus arteriosus or MAPCAs. In addition, we also found that nearly all patients with the type 1 of pulmonary circulation had normal pulmonary branching to lungs (96 %) with adequate pulmonary artery diameter, McGoon 2.44 ± 0.8.

In contrast with patients with TOF, the most common type of pulmonary circulation in patients with pulmonary atresia was type 2-absence of main pulmonary trunk with a confluent PAs (54 %). Incomplete arborization of PAs occurred in about half of these patients, and about two-third of them had associated MAPCAs (type 2b) to provide an additional source of pulmonary perfusion. In addition, patients with pulmonary atresia have a significant smaller size of PAs than patients with TOF. Our results are different from a previous study which reported the angiographic classification of the pulmonary circulation in 56 patients with pulmonary atresia, which found that the most common pulmonary circulation was type 1 with the presence of ductus arteriosus [5]. In addition, our study found a higher incidence of incomplete arborization of PAs than the previous study (53 % vs 29 %) [5]. The discrepancy in results may be related to geographic differences in the pattern of pulmonary circulation among the Asian and South American study population and the different numbers of the study population ($N =$ 85 vs 56).

Our results of pulmonary artery circulation are comparable to a previous CTA study of 116 infants and children with pulmonary atresia [18]. We found a higher incidence of the presence of right and left pulmonary arteries than the previous study (84 % vs 65 %). Among patients with the presence of both branch pulmonary arteries, the proportion with a main pulmonary trunk and with a confluence pulmonary artery were similar, approximately 24 % vs 28 %, and 93 % vs 89 %,

respectively. In addition, the incidence of unilateral right or unilateral left pulmonary artery (type 5) was comparable, 4.7 % vs 3.4 %. However, we found a lower incidence of absence of central pulmonary arteries (10.6 % vs 29 %).

Therefore, our study suggests that the CTA type of pulmonary circulation determines the completeness of PAs in terms of complete arborization and size. There is a significant difference in the type of pulmonary circulation, prevalence of complete arborization of PAs, and pulmonary artery dimension between patients with TOF and patients with pulmonary atresia.

### 5.2. Type of pulmonary circulation and major aortopulmonary collateral arteries

With the exception of pulmonary circulation type 3, our study found that the total number of MAPCAs is inversely related to the completeness of the pulmonary vascular bed and the diameter of the pulmonary artery. This data is comparable with a previous study [19]. MAPCAs anastomoses with native PAs were more commonly seen in pulmonary circulation type 2 than type 1, reflecting that this form of MAPCAs is more commonly seen in patients with pulmonary atresia. MAPCAs are essential in type 4 and type 5 where pulmonary perfusion is exclusively dependent on MAPCAs.

A recent study in 9 children with 31 MAPCAs suggested that CTA and catheterization are equivalent in their ability to evaluate pulmonary circulation and MAPCAs. The study also suggested that catheterization should be reserved for patients who require catheter-based intervention or hemodynamic assessment [20]. An additional advantage of three-dimensional CTA over the 2-dimensional projection of cardiac catheterization is that it allows assessment of the superimposed complex anatomy of MAPCAs. We agree with the previous study and our institution routinely uses CTA to evaluate MAPCAs as a baseline anatomical roadmap for treatment planning.

### 5.3. Clinical course and disease burden

Our study confirms that the morphology of both PAs and MAPCAs may suggest successful corrective surgery [13,21]. Cases with confluent native PAs had a better outcome than cases with non-confluent native PAs [13]. Anatomical features of pulmonary circulation type 1 with a proper size and normal branching of PAs are the prerequisite features for the achievement of definitive surgical management (achievable rate of 71 %). Although, there is a lower rate of surgical correction in our study compared with a previously published report, the results are still comparable [21]. In that study, all patients ($N = 5$) with the presence of confluent PAs (Barbero-Marcial classification A1 corresponding with CTA type 2) were able to achieve definitive surgery, while our study found that about one-third of patients ($N = 11$) achieved corrective surgical repair. Our result agrees with the previous study that patients who have no central PAs (CTA type 4) least often achieved definitive surgery. Only one-third of them have undergone unifocalization.

There are a few reports about disease burden in patients with this disease entity. Our study found that the median value of disease burden in patients who achieved surgical repair and with palliative management were 3 (1−4) and 2.5 (1.5−3), respectively. The overall disease burden in patients with TOF and pulmonary atresia was 3 (1−4). Although there was no statistically significant difference in the amount of disease burden among the different types of pulmonary circulation, there was a tendency for the more complex anatomical pulmonary circulations to have a greater number of procedures.

The previous study in 19 newborns with pulmonary atresia reported that the mean radiation dose (Dose-area product) was 169 mGy*cm$^2$ (47–169 mGy*cm$^2$) [22]. Median radiation exposure (Dose-length product) from the current study was 42 mGy*cm (IQR 32-73). There was no statistically significant difference in radiation exposure to patients with different types of pulmonary circulation.

Children with TOF and pulmonary atresia frequently undergo several imaging assessments, either cardiac catheterization or CTA, and catheter-based interventions. Therefore, reduction of radiation exposure is an important issue. To avoid radiation exposure, magnetic resonance imaging and angiography (MRA) is an important reliable modality allowing delineation of MAPCAs [23–25]. The MRA study can be accomplished either with or without gadolinium injection. Free breathing whole heart 3D steady-state free precession is an efficient technique for detection of MAPCAs without the use of gadolinium contrast agent [23]. Gadolinium-enhanced three-dimensional MRA is a non-ECG gating pulse sequence that has been proven for evaluation of great vessels and MAPCAs in children [24]. Roche et al. demonstrated that MRA revealed more minor MAPCAs than angiography [25].

### 5.4. Study limitations

There are several limitations. First, this was a single-center, retrospective study. The data may have been influenced by selection bias, with only survivors referred for CTA to our tertiary imaging center. CTA studies were analyzed in the absence of comparison with catheterized angiographic studies. Second, counting the number of MAPCAs based on the number of origins instead of the number of all branches of MAPCAs could underestimate the total number of MAPCAs. In addition, we did not include the systemic arterial collateral originating from the arterial branches of the aortic arch. Third, our study could underestimate the presence of ductus arteriosus since the median age at the time of the CTA study was 4 years, and the ductus arteriosus may have already closed. We established the presence of ductus arteriosus by the remnant of a closed ductus from CTA. Fourth, the small sample size in pulmonary circulation types 3, 4, and 5 reduced the power of the statistical analysis. In addition, given the limited median follow-up period of 24 months (IQR 9-40), we may have underestimated the number of patients who could achieve corrective surgery. A number of patients who had future planning for corrective surgery at a time point beyond the study period and were considered as “patients with palliative management”. Moreover, our study does not include anesthesia or sedation time in the disease burden. Finally, cumulative radiation dose data from CTA studies were the total of both ECG-gated CTA and non-ECG gated CTA, which may result in a higher radiation dose.

## 6. Conclusions

Most patients with TOF have normal pulmonary circulation while patients with pulmonary atresia have atrectic pulmonary trunk with confluent PAs. Type of pulmonary circulation based on CTA suggests the complete arborization and proper size of PAs as well as successful corrective surgery with an inverse relationship to the presence of MAPCAs. Although morphological information of PAs and MAPCAs is important, other factors should be considered for guidance in the management. Serial diagnostic imaging and palliative management are the sources of disease burden among these patients. As life expectancy increases, the risks of each procedure and cumulative radiation dose should be considered in the long-term follow-up of survivors with TOF and pulmonary atresia.

## CRediT authorship contribution statement

**Suvipaporn Siripornpitak:** Conceptualization, Methodology, Data collection, Formal analysis, Investigation, Writing - original draft, Writing - review & editing, Supervision, **Uracha Kunjaru:** Data collection, Formal analysis, Investigation, Writing - review & editing, **Apichaya Sriprachyakul:** Writing - review & editing, Worakan Promphan: Writing - review & editing, **Poomiporn Katanyuwong:** Writing - review & editing

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All authors have nothing to declare.

## Ethical statement

The study was approved by the institutional Human Research Ethics Committee, Faculty of Medicine, Ramathibodi Hospital, Mahidol University.

All procedures performed in this studies were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

## Declaration of Competing Interest

The authors have no conflict of interest to declare.

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