Comparison of computed tomography angiography versus cardiac catheterization for preoperative evaluation of major aortopulmonary collateral arteries in pulmonary atresia with ventricular septal defect

Rajesh Krishnamurthy[1], Farahnaz Golriz[2], Benjamin J Toole[3], Athar M Qureshi[4], Matthew A Crystal[5]

[1]Department of Radiology, Nationwide Children's Hospital, Columbus, OH, USA, [2]Department of Radiology, Baylor College of Medicine, Houston, TX, USA, [3]Department of Pediatrics, Emory University, Atlanta, GA, USA, [4]Department of Pediatrics, Baylor College of Medicine, Houston, TX, USA, [5]Department of Pediatrics, Columbia University Medical Center, New York, NY, USA

ABSTRACT

Introduction : Pulmonary atresia with the ventricular septal defect is a rare congenital heart defect with high anatomic variability. The most important management question relates to the sources of pulmonary blood flow. The ability to differentiate between ductal dependence and major aortopulmonary collateral arteries is critical to achieving good outcomes and avoiding life-threatening hypoxia in the postneonatal period. Having accurate information about pulmonary arteries, major aortopulmonary collateral arteries, and sources of blood supply to each pulmonary segment is crucial for choosing the optimal surgical strategy. The purpose of this study is to compare computed tomography angiography (CTA) with cardiac catheterization for anatomic delineation of surgically relevant anatomy in pulmonary atresia with ventricular septal defect with major aortopulmonary collateral arteries.

Materials and Methods : Retrospective review of all children with pulmonary atresia with ventricular septal defect with major aortopulmonary collateral arteries cared for at a large tertiary children's hospital who underwent cardiac catheterization with angiography and CTA close to each other without interval therapy. All studies were performed between 2007 and 2011.

Results : There were 9 patients who met the inclusion criteria. Pulmonary artery anatomy (confluent vs. nonconfluent) was correctly identified in 9 patients by CTA and 8 patients by catheterization. There were no significant differences between CTA and catheterization in the identification of major aortopulmonary collateral arteries (mean = 3.4 collaterals/study via catheterization; mean = 3.1 collaterals/study via CTA; $P = 0.67$). CTA was superior to catheterization in the delineation of segmental pulmonary blood flow ($P = 0.006$).

Conclusion : CTA and catheterization are equivalent in their ability to delineate pulmonary artery anatomy and major aortopulmonary collateral arteries.

Keywords : Catheterization, computed tomography angiography, major aortopulmonary collateral arteries, pulmonary atresia

Access this article online	
Quick Response Code:	**Website:** www.annalspc.com
	DOI: 10.4103/apc.APC_94_19

How to cite this article: Krishnamurthy R, Golriz F, Toole BJ, Qureshi AM, Crystal MA. Comparison of computed tomography angiography versus cardiac catheterization for preoperative evaluation of major aortopulmonary collateral arteries in pulmonary atresia with ventricular septal defect. Ann Pediatr Card 2020;13:117-22.

Address for correspondence: Dr. Matthew A Crystal, New York Presbyterian Morgan Stanley Children's Hospital - Columbia University Irving Medical Center, New York, NY 10032, USA. E-mail: mac2376@cumc.columbia.edu

INTRODUCTION

Pulmonary atresia with the ventricular septal defect is a rare congenital heart defect with high anatomic variability. The most important question regarding management in the newborn period relates to the status of the pulmonary blood flow. Patients with pulmonary atresia with ventricular septal defect and ductal dependent pulmonary blood flow often have well-developed, and confluent branch pulmonary arteries without major aortopulmonary collateral arteries.[1,2] These patients require prostaglandin E1 infusion to preserve ductal patency until they undergo an initial surgical procedure for pulmonary blood flow augmentation, typically with a modified Blalock-Taussig shunt, within the 1st day of life. Patients who do not have ductal dependent pulmonary blood flow derive the blood supply from major aortopulmonary collateral arteries, which arise from the aorta and systemic arteries. The latter group can potentially postpone surgery beyond the newborn period since pulmonary blood flow is relatively stable, although there is a tendency for major aortopulmonary collateral arteries to become stenotic over time. The ability to differentiate between ductal dependence and major aortopulmonary collateral arteries is critical to achieving good outcomes and avoiding life-threatening hypoxia in the postneonatal period.

The end goal of treatment in both groups is to achieve a biventricular repair with a conduit from the right ventricle to the pulmonary artery confluence, while optimizing pulmonary artery growth for the best long-term outcomes.[3-5] This is achieved by normalizing blood flow and pressure in as many pulmonary segments as possible. In patients with major aortopulmonary collateral arteries, bronchopulmonary segments receive their blood supply by either a pulmonary artery or a major aortopulmonary collateral artery (single supply) and sometimes by both (dual supply). When pulmonary segments with dual supply are small or remote to the native pulmonary arteries, they are typically treated by major aortopulmonary collateral artery catheter-based occlusion or surgical ligation, while segments with single supply, or large major aortopulmonary collateral arteries that are dual supply, will be incorporated into the central pulmonary circulation by a process known as unifocalization.[6] Therefore, having accurate information about pulmonary artery confluence, presence and location of major aortopulmonary collateral arteries, and the source of blood supply to each pulmonary segment is crucial for choosing the optimal surgical strategy. Cardiac catheterization is considered the reference standard to delineate the pulmonary artery anatomy and the sources of pulmonary blood flow, but catheterization has intrinsic drawbacks. It is an invasive modality that requires significant experience, especially for selective catheterization of major aortopulmonary collateral arteries in small children. In addition, issues with radiation exposure, femoral vessel injury, and need for general anesthesia are relevant in young infants. Morbidity related to cardiac catheterization is often compounded by the need for repeat studies in the 1st year of life. Recent advances in computed tomography angiography (CTA) that allows for unsedated, free-breathing, high-resolution imaging without pulsation artifact, and significant reduction in radiation exposure have pushed this modality forward as an important tool in the diagnostic armamentarium for congenital heart disease.[7-10] However, data comparing the ability of catheterization versus multidetector CTA to delineate surgical relevant anatomy in patients with pulmonary atresia with ventricular septal defect with major aortopulmonary collateral arteries is limited.[11,12]

The purpose of this pilot study is to compare CTA with cardiac catheterization for anatomic delineation of surgically relevant anatomy in pulmonary atresia with ventricular septal defect with major aortopulmonary collateral arteries.

MATERIALS AND METHODS

The institutional review board approved this study, and the requirement for informed consent was waived.

Patient population

A retrospective review of all children with pulmonary atresia with ventricular septal defect with major aortopulmonary collateral arteries cared for at a large tertiary children's hospital between January 1980 and June 2011 was performed. Among 92 children with the diagnosis of pulmonary atresia with ventricular septal defect with major aortopulmonary collateral arteries, only 9 patients had both a pre-operative cardiac catheterization with angiography and preoperative CTA close to each other without interval therapy. All other patients only had one pre-operative assessment modality. All studies were performed between 2007 and 2011. The mean interval between the two studies was noted.

All patients underwent the following testing: electrocardiogram, complete blood count, electrolytes, urea/creatinine, basic coagulation panel, and a chest radiograph. A recent echocardiogram was not required at the time of the catheterization or CTA, but all patients had a baseline echocardiogram in the neonatal period.

Computed tomography angiography protocol

All 9 CTAs were performed on a 64-detector CT scanner (Lightspeed VCT XT; GE Healthcare) under the direct supervision of an attending pediatric radiologist. Eight studies were performed without sedation or

general anesthesia, and one study required general endotracheal anesthesia. Optiray 320 mg/ml was injected at a dosage of 2–2.5 ml/kg through a peripheral catheter (4 upper extremity and 5 lower extremity) using a power injector at an injection rate of 2–3 cc/s. The scans were performed in a cranial–caudal direction without ECG-synchronization. The following acquisition parameters were used: Collimation: 0.625 mm, slice thickness: 0.625 mm, tube voltage: 80–120 kV, and tube current: 220–350 mA. All CTA images were reconstructed with filtered back projection.

Cardiac catheterization protocol

All patients were brought for cardiac catheterization to delineate pulmonary artery anatomy. In addition, hemodynamic data were also obtained, including ventricular systolic and end-diastolic pressures, aortic pressure, and saturation. An estimate of pulmonary artery pressures was obtained by direct pulmonary artery measurement if access was possible, or by pulmonary vein wedge measurement, if the direct measurement was not possible.

General aorta angiography was performed as an initial roadmap [Figure 1a]. Whenever possible, selective right and left pulmonary artery angiograms were performed to identify the lung segments perfused by each pulmonary artery and to exclude discrete stenoses or tubular hypoplasia [Figure 2a]. Often, a negative "wash-out" pattern can be seen that is due to a stream of unopacified blood from a connecting pulmonary artery or collateral flowing into an area of the opacified pulmonary arterial tree. Balloon occlusion of a collateral vessel while injecting into that pulmonary artery or vice versa was then performed.

Pulmonary vein wedge injection was performed to identify central pulmonary arteries if the initial aortogram and

selective injection of the major aortopulmonary collateral arteries did not identify a pulmonary artery confluence.

Selective hand-injections into major aortopulmonary collateral arteries were performed to delineate the extent of pulmonary arterial tree supplied by each collateral vessel and to determine which type of pulmonary artery connection was present (sometimes enhanced by selective balloon occlusion techniques). Collaterals found to be connected to the central PAs ("dual supply") were considered for coil occlusion to simplify the operative approach in some cases where the vessel was small. In some cases, collaterals with severe proximal obstructions providing single supply to lung segments would be considered for balloon dilation or stent implantation to improve distal flow before unifocalization, but in our patient cohort did not occur. Not all of the above injections were always necessary, but an attempt was made to account for perfusion of each pulmonary segment. The total dose of contrast did not exceed 10 ml/kg.

The distal pulses were monitored if arterial access was used. If pulses were weak or absent, a Heparin infusion was started promptly within 1-h postcath. Good hydration was maintained to reduce the risk of thromboembolic complications or hyperosmolality secondary to a large volume of injected contrast. Children were discharged home on the same day unless access site or other complications required an overnight stay.

Image analysis

All cardiac CT examinations were transferred to a post-processing workstation (Vitrea, Toshiba America Inc.,). One experienced pediatric radiologist reviewed all computed tomography (CT) angiograms. Various image reformatting techniques, including multiplanar reformation, thin-slab maximum intensity projection, and volume rendering were used. Volume rendering with a combined airway-vessel setting was used to study the relationship between the major aortopulmonary
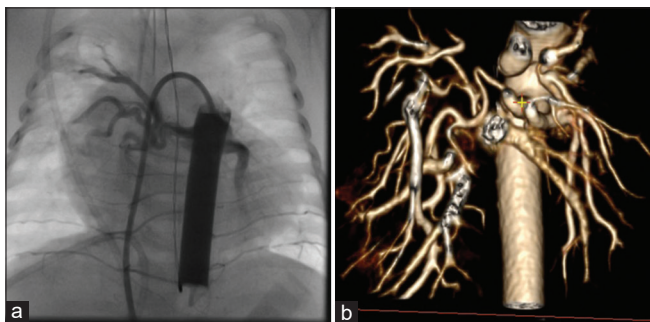
Figure 1: (a) Fluoroscopy during cardiac catheterization demonstrating a large major aortopulmonary collateral artery originating from the descending aorta and supplying pulmonary segments of both the right and left lungs. (b) Three-dimensional reconstruction from computed tomography angiography of the same patient showing several major aortopulmonary collateral arteries originating from descending aorta to partially supply both lungs. Three-dimensional capacity of computed tomography angiography avoids superimposition of major aortopulmonary collateral arteries as seen on two-dimensional catheterization
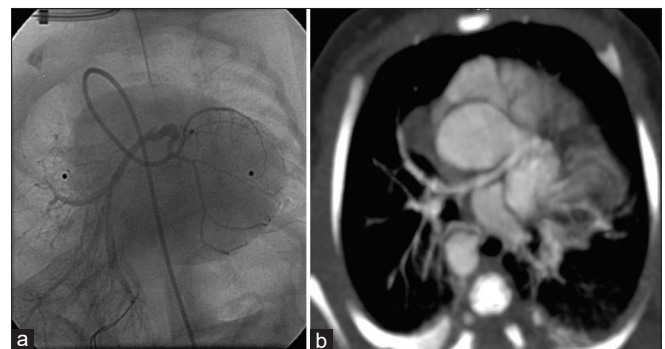
Figure 2: (a) Selective angiography performed during catheterization demonstrating the right pulmonary artery arising from the left coronary artery in a study patient. (b) Axial computed tomography angiography image performed in the same patient demonstrating RPA® originating from the left coronary artery (L)

collateral arteries and the tracheobronchial tree, with the latter serving as a landmark to describe the mediastinal course of the native branch pulmonary arteries and the major aortopulmonary collateral arteries [Figures 1b and 2b].

A single interventional pediatric cardiologist performed a review of all catheterization angiograms. The "native" pulmonary arteries and major aortopulmonary collateral arteries anatomy were summarized. The central PAs were measured immediately proximal to the first lobar branch. The collateral vessels that exclusively perfuse large pulmonary segments ("single supply") were also measured. If central pulmonary arteries were of borderline size or overtly hypoplastic, the NAKATA index was calculated as follows: Nakata index (PA index) = (RPA area [mm^2] + LPA area [mm^2])/BSA (m^2), with an optimal index ≥200.

The following data were obtained from CTA and cardiac catheterization:

Pulmonary artery anatomy: Pulmonary artery status (confluent vs. nonconfluent) as assessed by CT and catheterization, and confirmed at the time of unifocalization surgery.

Major aortopulmonary collateral artery identification: Number of major aortopulmonary collateral arteries, location of major aortopulmonary collateral arteries, size of major aortopulmonary collateral arteries at the origin, and presence of stenosis.

Segmental pulmonary blood flow: Blood supply to 20 study-defined pulmonary segments, along with characterization as a single supply or dual supply.

All nine patients underwent unifocalization surgery. The status of the branch pulmonary arteries was noted at the surgery and used as the reference standard.

Statistical analysis

All calculations were performed with a SAS version 9.3 (SAS Institute, Inc., Cary, North Carolina). A Wilcoxon matched-pair sum rank test was performed to compare CTA and cardiac catheterization for the mean number of major aortopulmonary collateral arteries identified, as well as the number of pulmonary segments that were determined to have single versus dual supply. Two-tailed $P < 0.05$ were considered to indicate statistical significance.

RESULTS

The patient population consisted of 5 males and 4 females. The median age at the time of CTA was 4 days (range: 1–1967 days), and at the time of catheterization was 35 days (range: 5–167 days). The median interval between CTA and catheterization was 24 days (range: 4 days to 5 months). Of the nine patients, three patients underwent catheterization before CTA. The mean weight at the time of the catheterization was 3.8 kg (range 2.8–7) and at the time of surgery was 3.8 kg (3–8.1 kg). Two of the patients had complications at the time of the catheterization. No complications were associated with the CTA. Decreased pedal pulses occurred in one patient (3 kg) who required a heparin infusion overnight, resulting in a normal pulse the following day. In addition, one patient (2.8 kg) received adenosine for intraprocedural supraventricular tachycardia with no hemodynamic compromise and no recurrence.

Pulmonary artery anatomy

Pulmonary artery anatomy (confluent vs. non-confluent) was correctly identified in 9 patients by CTA and 8 patients by catheterization, as confirmed at the time of surgery.

Major aortopulmonary collateral arteries identification

An equal number of major aortopulmonary collateral arteries were identified by catheterization and CTA in 6 patients. In one patient, CTA showed four major aortopulmonary collateral arteries, whereas catheterization indicated 1 major aortopulmonary collateral artery. CTA did not show all major aortopulmonary collateral arteries demonstrated by catheterization in 2 cases [Table 1]. Overall, a total of 31 major aortopulmonary collateral arteries were detected by catheterization, compared with 28 detected on CTA. There were no significant differences between CTA and catheterization in the identification of major aortopulmonary collateral arteries (mean = 3.4 collaterals/study via catheterization; mean = 3.1 collaterals/study via CTA; $P = 0.67$).

Segmental pulmonary blood flow

In 8/9 CTAs, the blood supply to all 20 study-defined pulmonary segments was identified (mean = 19.5 pulmonary segments); however, the blood supply to all 20 pulmonary segments was identified in only 1/9 patients via catheterization (mean = 17.5 pulmonary segments). CTA was superior to catheterization in the delineation of segmental pulmonary blood flow ($P = 0.006$).

DISCUSSION

Surgical management of pulmonary atresia with major aortopulmonary collateral arteries requires accurate preoperative delineation of the pulmonary blood supply. This study showed that CTA with a 64 multi-detector CT (MDCT) scanner and catheterization are equivalent in their ability to delineate pulmonary artery anatomy and the number of major aortopulmonary collateral arteries. In addition, CTA was able to delineate

Table 1: Description of patients and collateral arteries identified

Gender	Age at Cath (days)	Age at CTA	Confluent pulmonary arteries	Number collaterals		Number pulmonary segments w/identified blood supply	
				Cath	CTA	Cath	CTA
Female	7	1 day	Cath=No CTA=No	7	3	18	20
Male	8	5 years	Cath=No CTA=No	4	4	15	16
Female	5	2 months	Cath=Yes CTA=Yes	3	3	20	20
Female	167	1 day	Cath=No CTA=No	1	3	13	20
Male	35	4 days	Cath=No CTA=No	2	2	18	20
Male	98	27 days	Cath=Yes CTA=Yes	4	4	19	20
Male	30	10 days	Cath=Yes CTA=Yes	4	3	18	20
Male	149	2 days	Cath=No CTA=Yes	3	3	18	20
Female	75	2 days	Cath=Yes CTA=Yes	3	3	19	20

CTA: Computed tomography angiography

pulmonary segmental blood supply comparably, and demonstrate important surgical landmarks for major aortopulmonary collateral arteries with respect to the vertebrae, tracheobronchial tree, and esophagus. The three-dimensional (3D) nature of CT data, as compared with the 2D projection data of cardiac catheterization, avoided the superimposition of complex branching vessels. Cardiac catheterization was also limited by the need for augmented balloon occlusion injection techniques to demonstrate competitive flow in the setting of dual supply.

Recent advances in CT scanner technology, including dual-source scanner, wide-detector scanner, high-efficiency detectors, EKG-synchronization, and improved image processing, have consolidated the role of CT in the evaluation of congenital heart disease. These techniques have obviated the need for breath-holding and sedation for most indications even in neonates and infants,[8,9] and can routinely achieve effective radiation dose of <1 mSv, which is 10–15 fold less radiation than cardiac catheterization for the assessment of congenital heart disease.[13] Previous studies have shown that CTA provides excellent anatomic details of the pulmonary arteries and major aortopulmonary collateral arteries with resolution <1 mm with the newest scanners.[14] Yin *et al.* compared the accuracy of CTA using dual-source CT-scan with cardiac catheterization for the assessment of pulmonary blood flow in patients with pulmonary atresia and similar to our results, reported that CTA is 100% accurate for identification of major aortopulmonary collateral arteries.[15] Another study by Meinel *et al.* demonstrated that CT with 16 MDCT scanners could reliably identify the origin and size of the major aortopulmonary collateral arteries, and the supplied lung lobes. According to that study, radiation exposure

during CTA was also significantly lower compared to catheterization.[7]

Cardiac catheterization plays an important role in the management of patients with pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. Almost all of these patients require cardiac catheterizations over time for hemodynamic information and interventions on their major aortopulmonary collateral arteries, pulmonary arteries, and right ventricle to pulmonary artery conduit. Given the invasiveness of catheterization, it would seem prudent to minimize the number of cardiac catheterization procedures a patient has over his/her lifetime. Before the first surgical intervention, hemodynamic information is not absolutely necessary in most patients. Hence, based on our data, we feel that CTA alone can provide accurate pre-surgical data. In our experience, cardiac catheterization can be reserved as an initial procedure/test only in patients who need an intervention (e.g., collateral closure when surgical ligation may not be easily performed based on the major aortopulmonary collateral artery location), in the rare case when hemodynamic data are needed before surgery or where CTA at one's institution provides equivocal data.

This study has some limitations. It is a retrospective study with small sample size. Due to the sensitivity of small children to radiation, comparing CT to cardiac catheterization in all patients with pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries cannot be performed. There was a potential selection bias, with only patients with unanswered questions related to branch pulmonary artery status or major aortopulmonary collateral arteries being referred to catheterization or CT following the first

study and the second study may not be as thorough given the information obtained on the first study. Due to the lack of a reference standard for single versus dual-supply to the pulmonary segments, in cases of discrepancies between CTA and cardiac catheterization, it was not possible to determine the final diagnosis.

CONCLUSION

In summary, this study suggests that CTA and catheterization are equivalent in their ability to delineate pulmonary artery anatomy and major aortopulmonary collateral arteries. Cardiac catheterization should be reserved for patients, in whom a catheter-based intervention or hemodynamic information may be needed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Liao PK, Edwards WD, Julsrud PR, Puga FJ, Danielson GK, Feldt RH. Pulmonary blood supply in patients with pulmonary atresia and ventricular septal defect. J Am Coll Cardiol 1985;6:1343-50.

2. O'Leary P, Edwards W, Julsrud P, O'Leary P, Edwards W, Julsrud PR, *et al.* Pulmonary Atresia and Ventricular Septal Defect. Moss and Adam's Heart Disease in Infants, Children, and Adolescents. Baltimore, MD: Lippincott Williams and Wilkins; 2008. p. 878-88.

3. Gupta A, Odim J, Levi D, Chang RK, Laks H. Staged repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries: Experience with 104 patients. J Thorac Cardiovasc Surg 2003;126:1746-52.

4. Yagihara T, Yamamoto F, Nishigaki K, Matsuki O, Uemura H, Isizaka T, *et al.* Unifocalization for pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. J Thorac Cardiovasc Surg 1996;112:392-402.

5. Malhotra SP, Hanley FL. Surgical management of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals: A protocol-based approach. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2009;12:145-51.

6. Prieto L. Management of tetralogy of fallot with pulmonary atresia. Images Paediatr Cardiol 2005;7:24-42.

7. Meinel FG, Huda W, Schoepf UJ, Rao AG, Cho YJ, Baker GH, *et al.* Diagnostic accuracy of CT angiography in infants with tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries. J Cardiovasc Comput Tomograp 2013;7:367-75.

8. Jadhav SP, Golriz F, Atweh LA, Zhang W, Krishnamurthy R. CT angiography of neonates and infants: Comparison of radiation dose and image quality of target mode prospectively ECG-gated 320-MDCT and ungated helical 64-MDCT. AJR Am J Roentgenol 2015;204:W184-91.

9. Han BK, Overman DM, Grant K, Rosenthal K, Rutten-Ramos S, Cook D, *et al.* Non-sedated, free breathing cardiac CT for evaluation of complex congenital heart disease in neonates. J Cardiovasc Comput Tomogr 2013;7:354-60.

10. Baker TB, McFall RM, Shoham V. Current status and future prospects of clinical psychology: Toward a scientifically principled approach to mental and behavioral health care. Psychol Sci Public Interest 2008;9:67-103.

11. Hayabuchi Y, Inoue M, Watanabe N, Sakata M, Nabo MM, Kitagawa T, *et al.* Assessment of systemic-pulmonary collateral arteries in children with cyanotic congenital heart disease using multidetector-row computed tomography: Comparison with conventional angiography. Int J Cardiol 2010;138:266-71.

12. Greil GF, Schoebinger M, Kuettner A, Schaefer JF, Dammann F, Claussen CD, *et al.* Imaging of aortopulmonary collateral arteries with high-resolution multidetector CT. Pediatr Radiol 2006;36:502-9.

13. Watson TG, Mah E, Schoepf UJ, Watson TG, Mah E, Schoepf UJ, *et al.* Effective radiation dose in computed tomographic angiography of the chest and diagnostic cardiac catheterization in pediatric patients. Pediatr Cardiol 2013;34:518-24.

14. Rigsby CK, McKenney SE, Hill KD, Chelliah A, Einstein AJ, Han BK, *et al.* Radiation dose management for pediatric cardiac computed tomography: A report from the Image Gently 'Have-A-Heart' campaign. Pediatr Radiol 2018;48:5-20.

15. Yin L, Lu B, Han L, Wu RZ, Johnson L, Xu ZY, *et al.* Quantitative analysis of pulmonary artery and pulmonary collaterals in preoperative patients with pulmonary artery atresia using dual-source computed tomography. Eur J Radiol 2011;79:480-5.