

Comparison of contrast enhanced magnetic resonance angiography with invasive cardiac catheterization for evaluation of children with pulmonary atresia

Soha Romeih,^{1,2} Fathia Al-Sheshtawy,¹ Mai Salama,¹ Nico A. Blom,³ Ahmed Abdel-Razek,⁴ Hala Al-Marsafawy,⁵ Abdou Elhendy⁶

¹Department of Cardiology, Tanta University Hospital, Tanta, Egypt; ²Department of Radiology, Academic Medical Center, Amsterdam; ³Department of Pediatric Cardiology, Leiden Medical University Center, Leiden, The Netherlands; ⁴Department of Radiology, Mansoura University Hospital; ⁵Department of Pediatric Cardiology, Mansoura Children's University Hospital, Egypt; ⁶Department of Cardiology, Marshfield Clinic, Marshfield, WI, USA

Abstract

Complete assessment of the source of pulmonary blood supply and delineation of the anatomy of pulmonary arteries are essential for the management and prognostic evaluation of pulmonary atresia (PA) patients. Invasive cardiac catheterization is considered

the gold standard imaging modality to achieve this. We investigated the role of contrast enhanced magnetic resonance angiography (MRA) to evaluate the pulmonary blood supply and the anatomy of the pulmonary arteries and compared this with cardiac catheterization in children with PA. We studied 20 children with PA. Median age was 2.5 years (range 6 months-13 years). All patients were examined with cardiac catheterization and contrast enhanced MRA, and the results of both modalities were compared. There was a complete agreement between both modalities in the detection of the main pulmonary artery morphology and determination of the confluence state of the central pulmonary arteries. There was an 88% agreement for patency of the ductus arteriosus and 66% for patency of the surgically placed shunt. There was a complete agreement between both techniques on determining the presence of collaterals more than 2.5 mm. Twenty-eight collaterals of less than 2.5 mm were detected only by contrast enhanced MRA. There was a strong correlation between both modalities in measuring the pulmonary arteries and collaterals diameter ($P < 0.001$). Contrast enhanced MRA is a safe and accurate non-invasive technique to evaluate the pulmonary artery morphology and the sources of pulmonary blood supply in children with PA.

with ventricular septal defect (PAVSD) and pulmonary atresia with intact ventricular septum (PAIVS).^{2,3} The central pulmonary arteries may be hypoplastic, discontinuous, or entirely absent. The pulmonary vascular bed receives a blood supply from different sources, such as patent ductus arteriosus (PDA), systemic to pulmonary collaterals, or surgically placed shunts.

Complete assessment of the pulmonary blood supply source and accurate delineation of the morphology of the pulmonary arteries are essential for both optimal management and prognostic evaluation of patients with PA.⁴ Traditionally, cardiac catheterization is considered the gold standard method to achieve this. However, cardiac catheterization is invasive and involves radiation exposure and, therefore, serial investigations can be problematic.^{5,6} Echocardiography is of limited value in this group of patients due to the poor visualization of vascular structures outside the mediastinum with both a transthoracic and a trans-esophageal approach.⁶⁻⁸ The use of cardiac magnetic resonance imaging in complex congenital heart diseases is on the increase. The role of contrast enhanced magnetic resonance angiography (MRA) in accurate delineation of complex pulmonary artery anatomy and evaluation of the pulmonary blood supply source in adult patients with PA has been previously reported.⁹⁻¹¹ However, data on the efficacy and safety of the contrast enhanced MRA in children with PA are still limited. Pediatric patients often represent a special challenge for acquisition of arterial phase datasets because the children vary widely in size, circulation time and ability to cooperate. The aim of this study is to evaluate the role of contrast enhanced MRA, as compared to cardiac catheterization, in the evaluation of the anatomy of the central pulmonary

Correspondence: Abdou Elhendy, Marshfield Clinic, 1000 N Oak Ave, Marshfield WI 54449, USA. E-mail: elhendy.abdou@marshfieldclinic.org

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Introduction

Pulmonary atresia (PA) is defined as a complete obstruction between the right ventricle and the pulmonary artery with developmental abnormalities of the pulmonary valve or pulmonary arterial tree.¹ Two anatomical classifications have been identified: pulmonary atre-

arteries and the pulmonary blood supply in children with PA.

Materials and Methods

Patients' characteristics

There were 12 male and 8 females. Median age was 2.5 years (range 6 months-13 years). Eleven patients were younger than 2 years old, 10 patients weighed less than 10 kg. Sixteen patients had PAVSD, 2 patients had PAVS and 2 patients had complex coronary heart disease (CHD) with atretic PA. Five patients had had previous surgical interventions; 4 had modified Blalock-Taussig (MBT) shunts and one patient had left-sided pulmonary unifocalization.

Patients were prospectively recruited from Manasoura Children's University Hospital, Tanta University Hospital, Egypt, from November 2004 till March 2008. The local medical ethics committee approved the study. Informed consent was obtained from all parents before enrolment. Twenty-five eligible children were identified through the database of the pediatric cardiology department. Five patients were not enrolled; 4 had over NYHA class II and one patient's family did not agree for the child to take part. Diagnosis was established by transthoracic echocardiography (Hewlett Packard, Philips Medical Systems, Best, The Netherlands.). All patients underwent cardiac catheterization followed by magnetic resonance imaging (MRI) examination within one week. No clinical events occurred during that interval.

Cardiac catheterization

Cardiac catheterization was performed on a biplane catheter (System Philips Medical Systems, Best, The Netherlands). Contrast angiography was used and directed towards evaluation of the anatomy of the pulmonary arteries and the source of pulmonary blood supply. Evaluation included upper descending aortography, right ventriculography, pulmonary vein wedge angiography, and selective injections in the surgical placed shunt and/or the collaterals. In each patient, the morphology and the size of main pulmonary artery (MPA), left pulmonary artery (LPA), right pulmonary artery (RPA), the collaterals, and the source of the pulmonary blood supply were evaluated.

Magnetic resonance imaging

MRI was performed on 1.5 Tesla MR Scanner (Siemens Magnetom, Siemens Medical Systems, Germany). A cardiac-phased array radiofrequency coil was used in patients weighing over 10 kg. A head or surface coil was used in those with a body weight under 10 kg. Young patients (under the age of 8 years) were

sedated with chloral hydrate (100 mg/kg, maximum dose less than 2 g). Patients were monitored by pulse oximetry, electrocardiogram (ECG), and closed circuit television. MRI examination includes HASTE (half Fourier acquisition single-shot turbo spin echo) sequence that was used to define the cardiac anatomy and to guide contrast enhanced MRA planning.

Half-fourier acquisition single-shot turbo spin-echo

Scan parameters: echo time (TE) 15-26 ms; repetition time (TR) was the R-R cycle length; flip angle (FA) 90°; matrix 256 x 192; field of view (FOV) 200-400 mm; number of signal averages 2; slice thickness 5-7 mm. HASTE sequences were obtained in 3 orthogonal planes: transverse, sagittal and coronal.

Contrast enhanced magnetic resonance angiography

A non ECG-triggered 3-dimensional spoiled gradient echo pulse sequence was used. Scan parameters: TE 1.18-1.7 ms; TR 5.6-6.8 ms; FA 45°; number of signal averages 1; FOV 300-400 mm; matrix 256 x 126; slice thickness 1.2-1.5 mm; acquisition time 18-20 s. MRA scanned in a coronal view from posterior (the spinal canal) to anterior (the ascending aorta). Gadopentetate dimeglumine contrast (0.2 mL/kg) was injected via a peripheral intravenous line either by hand (in patients weighing under 10 kg) or by a power injector at rates ranging from 3-5 mL/s (in patients weighing over 10 kg). The time delay between the start of contrast injection and data acquisition was determined by the *best estimate* method and ranged between 6-8 s. MRA was reconstructed using 3-dimensional reformatting methods: multi-planer reformatting, maximum intensity projection, and 3-dimensional surface shading. In each patient, the morphology and the size of MPA, LPA, RPA, the collaterals and the source of the pulmonary blood supply were evaluated.

Post processing of images

Cardiac catheterization and contrast enhanced MRA images were compared for the qualitative and the quantitative assessment of the anatomy of the pulmonary arteries and the source of pulmonary blood supply. For the qualitative assessment, the morphology of the MPA and the central pulmonary arteries, LPA and RPA was evaluated. The arteries were classified as atretic (luminal discontinuity), hypoplastic (diameter of the pulmonary vessel less than 60% of aorta diameter) or stenotic (discrete narrowing of the artery diameter with the distal part 40% less than the proximal part).¹² The source of the pulmonary blood supply was determined by identification of PDA, surgically placed shunts or collaterals. To facilitate comparison between both modalities, the collaterals

were labeled numerically according to their order of origin from the descending aorta or aortic arch vessels. For the quantitative assessment, the smallest caliber of the pulmonary arteries (MPA, LPA, and RPA) and the collaterals was measured. MPA was measured when it crossed posterior to the ascending aorta. The diameter of the distal portion of LPA and RPA was measured just before the takeoff of the first upper lobe branch. LPA was measured when it crossed the left main stem bronchus to avoid an over-estimated measurement. Imbalance of growth is considered when the difference between distal LPA and RPA is more than 30%. The collaterals smaller than 2.5 mm were considered to be small collaterals and the collaterals larger than 2.5 mm were considered to be major collaterals.¹³

Cardiac catheterization results were analyzed by HA (with 21 years experience with CHD) who had not been informed of the results of the contrast enhanced MRA. The analysis of CMR/MRA was performed by AA and SR (14 and 4 years experience with CMR, respectively) who had not been informed of the angiographic results.

Statistical analysis

All statistical testing and data analysis were performed with SPSS software version 14 (SPSS Inc, Chicago, Ill, USA). Parametric data are expressed as mean \pm SD unless stated otherwise. The agreement between both modalities on the qualitative assessment was analyzed by calculating Kappa. Kappa below 0.4 is a poor concordance, Kappa between 0.4 to 0.75 is a moderate concordance, and Kappa between 0.76 to 1 is a strong concordance.¹⁴ The agreement between both modalities on the quantitative evaluation was analyzed by calculating the mean difference \pm SD, as described by Bland and Altman, and interclass correlation (ICC). ICC below 0.4 is a poor concordance, ICC between 0.4 to 0.75 is a moderate concordance, and ICC between 0.76 and 1 is a strong concordance.¹⁵ $P < 0.05$ was considered statistically significant.

Results

All patients underwent cardiac catheterization with no adverse effect. Also, all patients underwent contrast enhanced MRA with no adverse effect. In particular, none of the patients showed changes in vital signs during examination. Only minor adverse effects have been observed: headache in 2 patients (10%) and nausea in 3 patients (15%). These all resolved spontaneously and no treatment was required.

The morphology of main pulmonary artery, left pulmonary artery, right pulmonary artery and the collaterals

MPA was atretic in 7 patients, hypoplastic in 10 patients and absent in 3 patients.

There was a complete agreement (Kappa = 1) between both modalities in delineation of MPA anatomy. Both modalities were also concordant (Kappa = 1) in defining the central pulmonary arteries as confluent in 15 patients,

non-confluent in 4 patients, and absent in one patient. Kappa was 0.9 for the agreement between both modalities for PDA evaluation. There was a disagreement between both modalities in one of 3 patients with PDA: contrast enhanced MRA was not able to detect the presence of PDA while it was detected by the cardiac catheterization. Kappa was 0.7 for the agreement between both modalities in determining MBT shunt patency. There was a disagreement between both modalities in one of 4 patients with MBT shunt: the MBT shunt was found to be occluded by the cardiac catheteri-

zation while it was totally missed by contrast enhanced MRA. Figure 1 shows the agreement percentage between both modalities for the qualitative assessment.

Fifty-eight collaterals were detected by contrast enhanced MRA. Cardiac catheterization detected only 30 collaterals; therefore, overall Kappa was 0.6. All the 28 collaterals that were detected only by contrast enhanced MRA were smaller than 2.5 mm, *i.e.* small collaterals (Figures 2 and 3).

For the 30 collaterals that were detected by both modalities, there was a complete agreement in visualization of the origin of the collaterals: 19 collaterals were described as direct collaterals (taking origin from the descending aorta), 11 collaterals were described as indirect collaterals (5 collaterals taking origin from the right subclavian artery, 4 collaterals taking origin from the left subclavian artery, and 2 collaterals taking origin from the innominate artery). Kappa was 0.9 for the agreement between both modalities for visualization of the collateral insertions. There was disagreement in visualization of 2 collateral insertions. By cardiac catheterization, the insertion point of the first collateral was diffuse to the entire right lung while by contrast enhanced MRA it was localized in the upper lobe of the right lung. By cardiac catheterization, the insertion point of the second collateral was diffuse to the middle and lower lobe of the right lung while by contrast enhanced MRA it was more localized to the middle lobe.

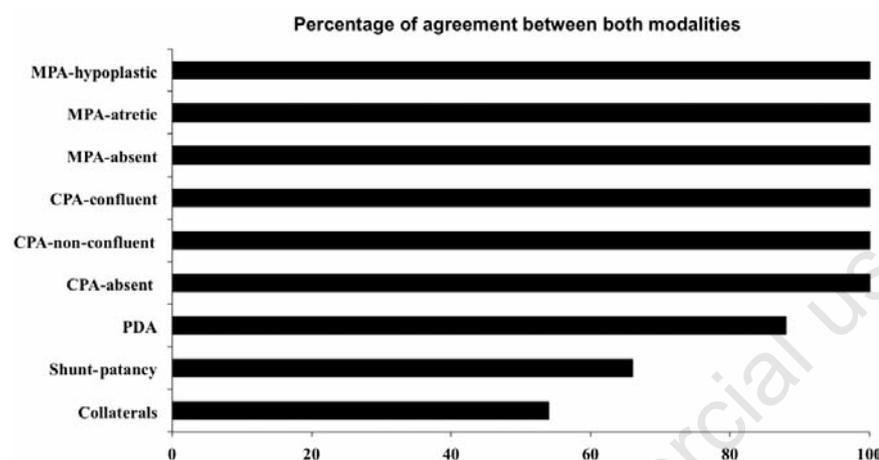


Figure 1. Agreement of the qualitative assessment between cardiac catheterization and contrast enhanced magnetic resonance angiography. MPA, main pulmonary artery; CPA, central pulmonary artery; PDA, patent ductus arteriosus.

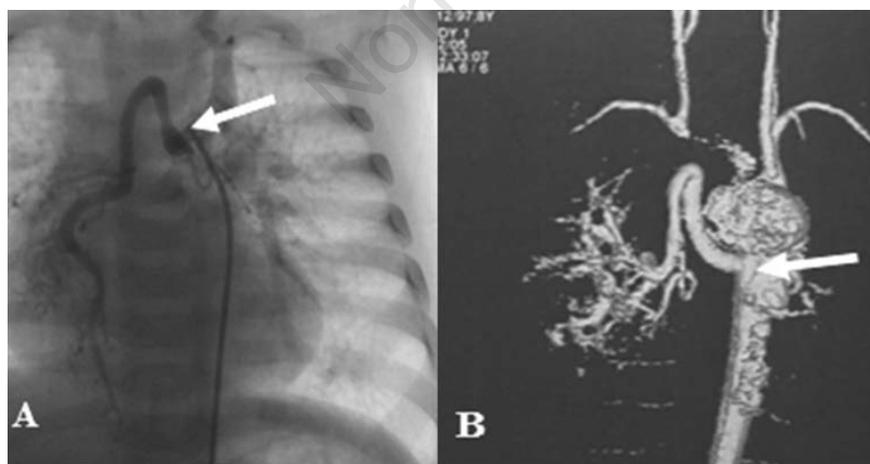


Figure 2. Comparison between both methods in evaluating a systemic to pulmonary artery collateral in a case of pulmonary atresia with ventricular septal defect. (A) Antero-posterior view of selective injection in the collateral demonstrating a direct collateral to the right lung taking origin from the upper part of descending aorta. (B) Antero-posterior view of 3-dimensional surface shading reconstruction of contrast enhanced magnetic resonance angiography demonstrating the same information. (Arrows show the collateral).



Figure 3. Three-dimensional surface shading of contrast enhanced magnetic resonance angiography demonstrating a large indirect collateral taking origin from the left subclavian artery going beneath the aortic arch to supply the right lung (solid arrow), giving small collaterals (<2.5 mm) to the upper lobe of left lung (dashed arrow).

Table 1. Comparison between cardiac catheterization and contrast enhanced magnetic resonance angiography measurement of the pulmonary arteries and the collaterals.

	Invasive catheterization	Contrast enhanced magnetic resonance angiography	ICC %
Mean pulmonary artery	3.0±2.4 mm	2.8±2.5 mm	96
Left pulmonary artery	3.9±1.6 mm	3.6±1.7 mm	80
Right pulmonary artery	4.5±1.6 mm	4.2±1.9 mm	85
Collaterals	1.7±0.7 mm	1.8±0.7 mm	88

ICC, inter-class correlation. Data are presented as mean ± SD

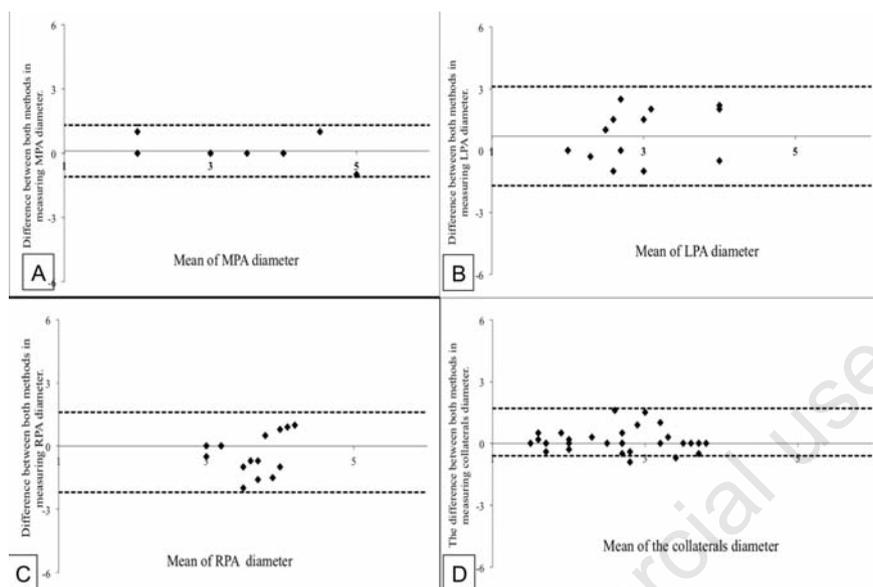


Figure 4. Comparison between cardiac catheterization and contrast enhanced magnetic resonance angiography measurements of the pulmonary artery and branches diameter by the Bland Altman test. (A) Main pulmonary artery, MPA; (B) left pulmonary artery, LPA; (C) right pulmonary artery, RPA; (D) collaterals.

The size of left pulmonary artery, right pulmonary artery and collaterals

Diameters of MPA, LPA, RPA and the collaterals are shown in Table 1. Bland-Altman analysis revealed no significant difference between methods in measuring these diameters (Figure 4).

Discussion

The present study showed that contrast enhanced MRA is a safe and effective, rapid, high-resolution imaging modality for complete assessment of the pulmonary blood supply and accurate delineation of the anatomy of the pulmonary arteries in children with PA. The large field of view and rapid data acquisition in one

breath-hold after a single injection into a peripheral vein mean that a full delineation of the anatomy with the entire course of the collateral can be achieved by using contrast enhanced MRA. There was a strong agreement between contrast enhanced MRA and the cardiac catheterization in measuring the diameter of the pulmonary arteries and the collaterals. There was a complete agreement between both methods in delineation of MPA morphology and determining the confluence state of the central pulmonary arteries. For the patency of PDA and MBT shunt, there was an agreement in all cases except 2. PDA in one patient and MBT shunt in another patient were missed by contrast enhanced MRA, while they were clearly detected by the cardiac catheterization; this was probably due to incorrect timing of the contrast peak due to the jerky movements of the children during the contrast injection, even though they were sedated.

Optimal contrast timing in children is a problematic issue. A *bolus track* technique was considered a reliable approach to ensure the acquisition of arterial phase and highly reproducible contrast enhanced MRA results in the pediatric population.¹⁶ However, bolus track requires greater operator expertise than the *best estimate* method used in our study.

This study demonstrated the ability of contrast enhanced MRA to detect the small collaterals (<2.5 mm) which were not completely detected by invasive cardiac catheterization. Contrast enhanced MRA offers a better spatial resolution and a good signal to noise ratio, so that vessels as small as 0.5 mm with a slow flow and the intra-parenchymal pulmonary vessels can be visualized. Detecting the small collaterals is of great clinical importance to avoid or to identify the possibility of having a bloody surgical field.⁴

Our results show that contrast enhanced MRA was a reliable non-invasive imaging modality to define pulmonary arteries and pulmonary blood supply. These results agree with previous reports that compared contrast enhanced MRA with cardiac catheterization for assessment of the pulmonary artery anatomy and the pulmonary blood supply in Tetralogy of Fallot patients and PA patients.⁹⁻¹¹ However, the median age in these studies was 28 years,⁹ 4.7 years,¹¹ and a range of 3-30 years,¹⁰ respectively. Our patients were much younger and median age was 2.5 years (range 6 months-13 years). Our results suggest that contrast enhanced MRA can be used safely and effectively in children with PA.

Study limitations

The present study had some limitations. The sample size was small, the contrast enhanced MRA sequence was not able to delineate the peripheral branches of the pulmonary arteries beyond the 3rd or 4th generation. Also, the present study did not include the other MRI sequences that may be useful during a comprehensive examination of this group of patients, such as phase velocity cine MRI that can quantify blood flow, or fast gradient echo sequences that can measure ventricular volume, function and mass.

Conclusions

Contrast enhanced MRA is a safe and accurate non-invasive technique to evaluate the pulmonary artery morphology and the sources of pulmonary blood supply in children with PA. Its high spatial resolution and 3-dimensional properties allow the course of collaterals and detection of the small sized collaterals to be better evaluated.

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