Pre-Operative Evaluation with Magnetic Resonance Imaging in Tetralogy of Fallot and Pulmonary Atresia with Ventricular Septal Defect

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Background: *Preoperative evaluation of patients with pulmonary atresia and ventricular septal defect (PA/VSD) are generally done by echocardiogram and cardiac catheterization. The authors' objective of the present study was to compare the findings of Gadolinium (Gd) enhanced cardiac magnetic resonance angiography (MRA) with cardiac catheterization.*
Material and Method: *Patients who had PA/VSD were prospectively evaluated using cardiac catheterization and cardiac MRA. A branch of the pulmonary arteries was divided into: main pulmonary artery (MPA), left and right branch pulmonary artery (LPA & RPA), major aortopulmonary collateral arteries (MAPCA) and minor collaterals. Each study was interpreted blindly. The agreement of findings was compared using Kappa statistics.*
Results: *There were 43 patients who received both cardiac catheterization and cardiac MRI within a 2 month period. The average age was 13.8 $\pm$ 8.4 (2-30) years old. There was an agreement among measurement of both MPA and LPA & RPA with Kappa statistics of more than 0.8. Gd-enhanced MRA was able to identify more branches of MAPCA when compared to cardiac catheterization.*
Conclusions: *The results of the present study indicate that Gd-enhanced MRA is a feasible, fast and accurate technique for identification of all sources of pulmonary blood supply in patients with complex pulmonary atresia. The present study was a noninvasive alternative to cardiac catheterization. Gd-enhanced MRA can better delineate small (minor) branches of collateral.*

Keywords: *Cardiac magnetic resonance imaging, Tetralogy of fallot, Pulmonary atresia*

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Tetralogy of Fallot (TOF) including pulmonary atresia with ventricular defect (PA/VSD) is one of the most common types of cyanotic heart disease seen in children and adults. Its incidence is approximately 5 to 6% of all congenital heart disease[1-3]. All patients will require open heart surgery to repair the stenotic right ventricular out flow tract (RVOT), pulmonary artery (PA) and closure of ventricular septal defect (VSD). However, in patients with PA/VSD, the existence of confluence of the pulmonary artery (main pulmonary artery that was joined by both the right and left branch pulmonary arteries) is the predetermined factor for staging repair[2,3]. Generally, preoperative evaluations of patients receiving repair of TOF and PA/VSD were done by echocardiogram and cardiac catheterization.

At the Faculty of Medicine, Siriraj Hospital, cardiac magnetic resonance imaging (MRI) has been used to evaluate congenital heart lesions since February 2000[4-6]. MRI gave noninvasive images of

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the cardiovascular system (degree of RVOT obstruction, PA size[7-9], VSD anatomy and coronary artery anomalies) with no limitation of imaging plane. The authors' objective of the present study was to compare findings of Gadolinium (Gd) enhanced magnetic resonance angiography (MRA) with cardiac catheterization for patients under going surgical repair of pulmonary atresia/VSD.

Material and Method

The present study was designed as a prospective study for comparison of results obtained from Gd-enhanced MRA in patients with PA/VSD. The authors enrolled patients with unoperated PA/VSD at the Faculty of Medicine, Siriraj Hospital from 2002 to 2004. This group of patients generally was seen in the Pediatric cardiology clinic (Department of Pediatrics) or adult congenital heart disease clinic (Her Majesty Cardiac Center). Each patient was evaluated using standard left and right heart catheterization. Angiogram was performed to evaluate all branches of pulmonary arteries. Branches of pulmonary artery were divided into (Fig. 1):

1. Main pulmonary artery (main PA)
2. Left and right pulmonary artery (LPA & RPA)
3. Major aortopulmonary collateral arteries (MAPCA) which supplied the major lobe of each lobar segment.
4. Minor collateral arteries which supplied only a small segment of the lung.

Magnetic Resonance Angiography (MRA)[1,9-12] is a reliable and accurate method for noninvasive vascular evaluation. The Contrast-Enhanced MRA (CE MRA) has recently gained more acceptance due to its superior image quality compared to the pervious non-contrast MRA technique (e.g. Time of Flight or Phase Contrast MRA). The success of the technique basically depends on the synchronization of the MR acquisition to the presence of the contrast agent during the first pass. The technique is based on the T1-shortening effect of intravenously injected contrast agent (e.g. gadolinium) and rapid image acquisition using a 3D gradient-echo pulse sequence. Pre-contrast and postcontrast 3D data sets generally are acquired and a subtraction method is utilized to suppress the background tissue (i.e. to enhance the vascular signal).

Cardiac magnetic resonance imaging

All examinations were performed using a 1.5 Tesla whole-body MR scanner (Philips, ACS-NT, maximum gradient performance, 30-mT/m amplitude; slew rate, 150 T/m per sec) with a five-element phase-array cardiac coil. Patients who were younger than eight years old received cardiac MRI under general anesthesia. In the present study, CE- MRA was acquired with a time resolved subtraction technique[12] for four consecutive 3D data sets. A manual fluoroscopic triggering was employed to detect the arrival of the contrast agent. The fluoroscopic triggering was acquired using 2D-thick-slab-gradient-echo pulse sequence on the coronal plane with a frequency of one image per second. A double dose (0.2 mmol/kg body weight) of gadopentetate dimeglumine (Gd-DTPA) (Magnevist; Schering, Berlin, Germany) was injected intravenously at a rate of 2-3 mL/sec with a power

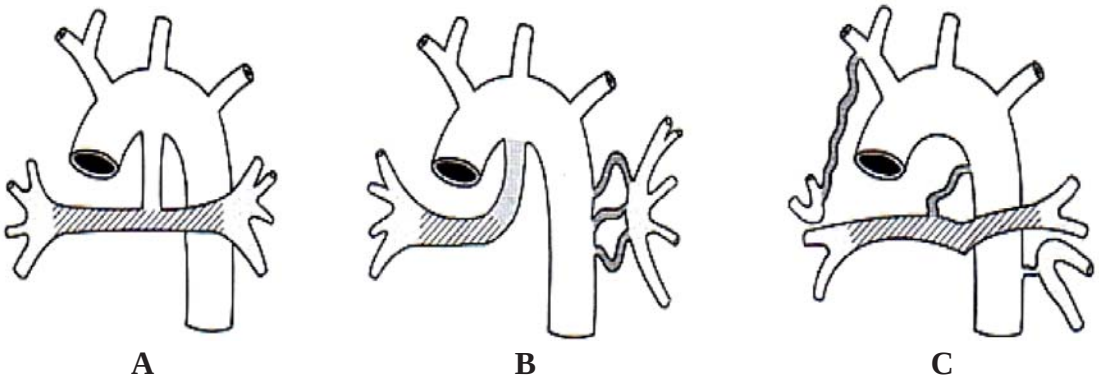

Fig. 1 Different type of pulmonary atresia/VSD
A) Confluent pulmonary artery
B) Large right major aortopulmonary collateral arteries (MAPCA)
C) Right lung with dual blood supply from minor collaterals

injector (Spectris, Medrad) followed by 10-20 mL of saline solution at the same rate. Once the increased signal due to the contrast agent had been detected, the scanner was manually switched to a fast 3D spoiled gradient-echo pulse sequence (TR 5.2 ms, TE 1.49 ms, flip angle 40°, matrix 400 x 260, slice thickness 1.2 mm, field of view 400 x 360 mm, with elliptical centric k-space order and SENSE factor 3) for four consecutive data sets. A breath-hold method was used to suppress motion artifacts during acquisition. The high-resolution scan of ten seconds per data set (as described above) was utilized for the patients who could corporate to hold their breath during the acquisition. However, for the incorporative patients, the spatial resolution and signal to noise was traded for the temporal resolution by acquiring for only four seconds per data set (i.e. matrix 256 x 192, slice thickness 2 mm with the same field of view and SENSE factor 4). The image was enhanced by subtracting the contrast from the precontrast images. The subtraction data sets were then subjected to the thin-slab maximum-intensity projection post-image processing to obtain a clear view of the interested arteries.

Gd-enhance MRA and cardiac catheterization were interpreted by different investigators Measurements of PA size by cardiac catheterization was performed a using sizing catheter as a calibration (10 mm distance between each marker). For MRA the calibration was done using the automated program. The data are presented as mean ± standard deviation. Comparison of the agreement of findings of main PA, branches PA and MAPCAs were performed using Kappa statistics. The degree of correlation was considered from values of Kappa statistics as follows: 0.3-0.499: low, 0.5-0.699: medium, 0.7-0.99: high and 1 is absolute correlation respectively. Measurement of the size of the main pulmonary artery, branches pulmonary arteries were performed using Pearson correlation. A p-value of less than 0.05 was considered statistically significant.

The present study was approved by the ethics committee of the Faculty of Medicine, Siriraj Hospital, and the present study was supported by a grant from the Research Committee, Faculty of Medicine, Siriraj Hospital.

Results

There were 43 patients who received both cardiac catheterization and Gd-enhanced MRA. The average age was 13.8 ± 8.4 (2-30 years old). All patients had both cardiac catheterization and Gd-enhanced MRA within two months. All patients received both procedures without any complications. There were 10 patients who required general anesthesia during the MRI procedure. All confluent pulmonary arteries were identified (Fig. 2). In addition, stenosis of the LPA was also noted opposing the anastomosis site of a Blalock Taussig shunt (Fig. 2) (arrow). Gd-enhance MRA can demonstrate all branches within one picture. Cardiac catheterization procedures require several selective angiograms for demonstration of all the branches.

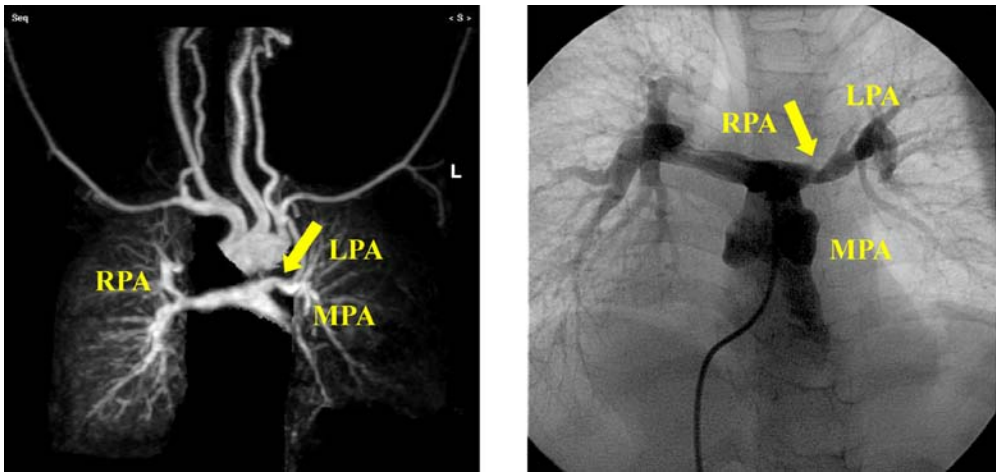

Fig. 2 Pictures demonstrate cardiac MRI (left) and cardiac catheterization (right) of confluent pulmonary artery, both main pulmonary artery (MPA), right and left pulmonary artery (RPA, LPA) are seen
Left pulmonary artery stenosis (arrow) was noted from both cardiac cath and MRI

Measurement of pulmonary artery

There was an agreement among dimensions of both main and branches (right and left) pulmonary artery using Pearson correlation (Table 1). The highest r-value was seen in the right pulmonary artery (r = 0.844). Agreement of cardiac catheterization and Gd-enhanced MRA were performed using Kappa statistics (Table 2). The evaluation of both MPA atresia (absence) and hypoplasia had Kappa statistics of 1 which indicated an absolute agreement between both methods. Other evaluations on branch pulmonary artery or MAPCA had high agreement between both methods.

Comparison of both cardiac MRA and cardiac catheterization (cath) for evaluation of major aortopulmonary collateral (MAPCA) is shown in Table 3. The number of MAPCAs found by both methods is shown in Table 3. MRI was able to identify additional branches of MAPCA when compared to cardiac catheterization. The Kappa statistic was 0.39 which indicated low agreement. Most of the additional branches identified by cardiac MRA were minor collaterals which will require coil embolization.

Discussion

Tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries (MAPCAs) is unique from most other forms of complex congenital heart disease. Its complexity is a function of heterogeneous and frequently severe anomalies of pulmonary blood supply.

In order to obtain an accurate unifocalization for staged repair, an imaging study for each branch of the pulmonary blood supply will need to be performed, traditionally using cardiac catheterization. The major draw back is that it is an invasive procedure with some morbidity, injury to the vascular access site and radiation exposure. Above all, cardiac catheterization may not be able to demonstrate small minor MAPCA if the branches were not engaged or injected directly[3]. Several studies have previously shown that standard MRI techniques, such as spin echo and gradient echo cine MRI, can be used to image the central pulmonary arteries and MAPCA in this patient population[8,13]. These MRI techniques, however, require relatively long scan times. Gadolinium (Gd)-enhanced 3D magnetic resonance imaging (MRI) or angiography (MRA) is a fast imaging technique that has been shown to accurately evaluate major arteries and veins[1,3,7]. MRA has recently been shown to correlate well with catheterization in the evaluation of branch pulmonary artery (PA) stenosis. However, its application to complex PA

Table 1. Comparison of measurement of main pulmonary artery (MPA), right pulmonary artery (RPA) and left pulmonary artery (LPA) by cardiac catheterization (cath) and cardiac MRA (MRA) using Pearson correlation analysis

Branches	Method			
	Measurement (mm)		p-value	r-value
	Cath	MRA		
MPA	19.7 $\pm$ 11.3	20.2 $\pm$ 8.2	0.018	0.756
RPA	16.2 $\pm$ 8.5	17.0 $\pm$ 7.1	<0.001	0.844
LPA	15.0 $\pm$ 9.7	16.7 $\pm$ 6.9	<0.001	0.829

Table 2. Comparison between agreement of reading between both cardiac catherization and cardiac MRA using Kappa statistics. The degree of correlation was considered from values of Kappa statistics as follows: 0.3-0.499: low, 0.5-0.699: medium, 0.7-0.99: high and 1 is absolute correlation respectively (MPA = main pulmonary artery, RPA = right pulmonary artery and LPA = left pulmonary artery)

	Kappa statistics
MPA atresia	1.000
MPA hypoplasia	1.000
MPA stenosis	0.853
RPA hypoplasia	0.859
RPA stenosis	0.858
LPA hypoplasia	0.861
LPA stenosis	0.858
MAPCA	0.801

Table 3. Comparison of both cardiac MRA and cardiac catherization (cath) for evaluation of major aortopulmonary collateral (MAPCA). The column and rows here show the number of MAPCAs found by both methods. MRA was able to identify more branches of MAPCA compared to cardiac catheterization. The Kappa statistics was 0.39

Cath	MRA					
	0	1	2	3	4	5
0	17	0	3	1	0	1
1	3	1	2	4	0	3
2	0	0	0	0	1	3
3	0	0	1	0	0	2
4	0	0	0	0	1	0

anomalies such as discontinuous or absent pulmonary arteries and particularly for delineation of MAPCA has not been examined in detail.

The results of the present study demonstrate that compared with x-ray angiography, Gd-enhanced 3D MRA accurately depicted all sources of pulmonary blood supply in patients with complex pulmonary stenosis or atresia ranging in age from 2 years old to adult age group. The procedure was non-invasive and required no hospitalization. The comparison between these MRA techniques and x-ray angiography showed good agreement in the measurements of the central pulmonary arteries. A previous study has shown a 100% sensitivity and specificity for the diagnosis of main and branch pulmonary artery (PA) stenosis or hypoplasia as well as absent or discontinuous branch PAs[3]. All major MAPCA that were diagnosed by catheterization were correctly diagnosed by MRA. Three additional MAPCAs were diagnosed by MRA but not by catheterization. It appeared that MRA can detect more branches of minor MAPCA than cardiac catheterization. The authors also found a good correlation between measurement of main and branch pulmonary arteries.

Limitation of the study

Cardiac MRA can be performed in children up to 8 years old without general anesthesia since it required sequential breath holding during the study period of which a small child could not comply. In a larger child or adult the procedure is much simpler to perform when compared to cardiac catheterization. In the present study there are a few patients who were younger than 8 years old. In particularly a young infant (less than 1 year old) cardiac catheterization might be the procedure of choice since it did not require general anesthesia during the procedure.

Conclusion

The results of the present study indicate that Gd-enhanced 3D MRA is a feasible, fast and accurate technique for identification of all sources of pulmonary blood supply in patients with tetralogy of Fallot and complex pulmonary atresia. The present study was a noninvasive alternative to cardiac catheterization. Gd enhanced MRA can better delineate small (minor) branches of collateral that were used as a guide for planning of transcatheter coil embolization in addition to routine cardiac catheterization.

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การเปรียบเทียบการประเมินก่อนการผ่าตัดด้วยเทคนิคการตรวจคลื่นแม่เหล็กไฟฟ้าและการตรวจสวนหัวใจในผู้ป่วยโรคหัวใจพิการแต่กำเนิดชนิด Tetralogy of Fallot และ pulmonary atresia

กฤตย์วิกรม ดุรงค์พิศิษฏ์กุล, ไพรัช สายวิรุณพร, จารุพิมพ์ สูงสว่าง, ดวงมณี เลาหประสิทธิพร, อภิชาติ นานา

ภูมิหลัง: โดยทั่วไปแล้วการประเมินก่อนการผ่าตัดในผู้ป่วยที่มี pulmonary atresia และ ventricular septal defeat (PA/VSD) นั้นจะทำโดยใช้การตรวจด้วยการตรวจสวนหัวใจ วัตถุประสงค์ของการศึกษานี้เพื่อทำการเปรียบเทียบการตรวจหลอดเลือดในหัวใจ ด้วยคลื่นสนามแม่เหล็กโดยมีการใช้สาร gadolinium (gadolinium enhanced cardiac magnetic resonance angiography, Gd-enhanced MRA) กับการตรวจสวนหัวใจ

วัสดุและวิธีการ: ผู้ป่วยที่ผ่านการผ่าตัดแก้ไข PV/VSD จะถูกประเมินโดยใช้การตรวจสวนหัวใจและการตรวจหลอดเลือดในหัวใจด้วยคลื่นสนามแม่เหล็ก (cardiac MRA) แขนงของหลอดเลือดแดงใหญ่ในปอดสามารถแบ่งออกได้เป็น: main pulmonary artery (MPA), left and right branch pulmonary artery (LPA & RPA), major aortopulmonary collateral arteries (MAPCA) และ minor collaterals จะทำการแปลผลการศึกษาแต่ละชนิดแบบปิดบังผลของการวินิจฉัยที่เหมือนกันจะถูกนำมาเปรียบเทียบกันโดยใช้หลักการทางสถิติของ Kappa

ผลการศึกษา: มีผู้ป่วยทั้งสิ้นจำนวน 43 รายที่ได้รับการตรวจด้วยการตรวจสวนหัวใจและการตรวจคลื่นแม่เหล็กไฟฟ้าของหัวใจ ในระยะเวลา 2 เดือน อายุเฉลี่ย 13.8 $\pm$ 8.4 ปี (2-30 ปี) มีความเหมือนกันของการตรวจทั้งของ MPA และ LPA & RPA เมื่อทดสอบด้วยหลักการทางสถิติของ Kappa โดยมีค่ามากกว่า 0.8 ยังพบว่า Gd-enhanced MRA สามารถที่จะตรวจพบแขนงของ MAPCA ได้มากกว่าการตรวจด้วยการตรวจสวนหัวใจ

สรุป: ผลจากการศึกษานี้ชี้ให้เห็นว่า Gd-enhanced MRA เป็นเทคนิคที่สามารถทำได้ รวดเร็ว และมีความถูกต้องสำหรับการตรวจหาแหล่งให้เลือดในปอดสำหรับผู้ป่วยที่มี pulmonary atresia ที่ซับซ้อน ซึ่งวิธีนี้เป็นทางเลือกที่ไม่มีการรุกล้ำร่างกายจากการตรวจสวนหัวใจ นอกจากนี้ยังสามารถจำแนก small (minor) branches of collateral ได้ดีกว่าอีกด้วย