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MR Imaging of Non-visualized Pulmonary Arteries at Angiography in Patients with Congenital Heart Disease

The aim of this study was to evaluate whether MR could depict pulmonary arterial anatomy in more detail than routine angiography in patients with congenital interruption or acquired occlusion of the left pulmonary artery or pulmonary atresia. This study included 10 patients with tetralogy of Fallot (n=6) or pulmonary atresia with ventricular septal defect (n=3) or aorticopulmonary window (n=1) diagnosed by cardiac angiography and MR. Surgical confirmation was made in seven patients. Interruption of the proximal left pulmonary artery, diagnosed at the time of evaluation, was found in seven patients and acquired obstruction of the hilar pulmonary artery (PA) was found in two at cardiac angiography. In the remaining one patient with pulmonary atresia and an occluded palliative shunt, the central PA was not visualized at angiography. MR showed 3-6 mm-sized hilar PAs in five and a central PA in a patient with pulmonary atresia. In 4 of 6 (67%) surgically-proven patients with congenital or acquired left PA obstruction, the status of the PA distal to the obstruction was correctly diagnosed with MR. In conclusion, MR is an effective modality in depicting sizable PAs when routine angiography fails to visualize the PA anatomy.

Key Words: *Heart defects, congenital; Magnetic resonance imaging, heart; Heart catheterization; Tetralogy of Fallot*

Yeon Hyeon Choe, Jae Kon Ko*, Heung Jae Lee†, I-Seok Kang†, Pyo Won Park‡, Young Tak Lee§

Department of Radiology, Department of Pediatrics†, Departments of Thoracic and Cardiovascular Surgery‡, Samsung Medical Center, College of Medicine, Sungkyunkwan University, Seoul, Korea
*Department of Pediatrics, Asan Medical Center, College of Medicine, University of Ulsan, Seoul, Korea
§Departments of Thoracic and Cardiovascular Surgery, Sejong General Hospital, Puchon, Kyunggi, Korea

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Address for correspondence
Yeon Hyeon Choe, M.D.
Department of Radiology, Samsung Medical Center, College of Medicine, Sungkyunkwan University, 50 Irwon-dong, Kangnam-gu, Seoul 135-230, Korea
Tel: +82.2-3410-2509, Fax: +82.2-3410-2559
E-mail: yhchoe@smc.samsung.co.kr

INTRODUCTION

Preoperative evaluation of the status of pulmonary arteries is important in patients with tetralogy of Fallot with or without pulmonary atresia and in those with truncus arteriosus and unilateral absence of a pulmonary artery (1). The pulmonary arteries can be evaluated by right ventriculography, aortography with or without balloon occlusion, or pulmonary vein wedge angiography (2). However, the central and hilar pulmonary arteries may sometimes not be visualized by these procedures, especially when they are not confluent. Moreover, the angiographic procedures, especially the pulmonary vein wedge angiography, are invasive techniques that may result in complications.

Magnetic resonance (MR) imaging has been utilized in visualization of central pulmonary arteries in patients with right ventricular outflow tract obstruction (3-10). MR imaging has also been used for anatomic depiction of unilateral pulmonary arterial obstruction (5, 9, 10). However, MR has been used to evaluate hilar pulmonary arteries only in a limited number of cases (11-13). We undertook this study to evaluate whether MR imaging can depict the hilar and central pulmonary arteries that were not visualized by cardiac angiography.

MATERIALS AND METHODS

The study population consisted of 10 patients (seven males and three females) in whom MR imaging was performed because of non-visualization of the central or hilar pulmonary arteries at angiography. The patients were selected randomly among the patients who had interruption of a pulmonary artery or pulmonary atresia without visualization of hilar and central pulmonary arteries at angiography. The underlying cardiovascular lesion included tetralogy of Fallot (n=6), pulmonary atresia with ventricular septal defect (n=3), and pulmonary atresia with aorticopulmonary window (n=1). The age of the

patients ranged from 1 month to 34 years (mean, 9.2 years). Three patients underwent modified Blalock-Taussig shunt operation and one patient underwent surgery for total correction of tetralogy of Fallot before angiography and MR imaging. We reviewed the echocardiograms, angiograms, MR images, and clinical records.

Echocardiography with color Doppler interrogation was performed by using 2.5-, 3.5-, and 5-MHz transducers (Acuson 128, Acuson Co., California, U.S.A.).

Right ventriculography was performed in nine patients and selective pulmonary arteriography one. In all the patients, thoracic aortography with or without selective injection in the collateral arteries was performed to visualize the collateral arteries and the central or hilar pulmonary arteries. The pulmonary vein wedge angiography was performed in one patient. Low osmolar ionic contrast medium (Hexabrix 320: sodium and meglumine ioxaglic acid, Guerbet, Cedex, France) was used for angiography and the total amount of contrast medium used was less than 3-4 mL/kg in each patient.

MR imaging was performed on a 0.5-T scanner (MRT 50A, Toshiba Medical Co., Tokyo, Japan). Chloral hydrate (50-100 mg/kg) was given orally in young patients for sedation. An intravenously injected sedative (pentobarbital; initial dose, 2 mg/kg; maximal dose, 6-7 mg/kg) was used in three children older than 3 years. In all the patients, we obtained electrocardiographically-gated T1-weighted oblique coronal images of each pulmonary artery in addition to the orthogonal transverse and coronal images. In one patient, cine MR imaging was performed. Average time taken for MR imaging was about one hour (40-70 minutes).

RESULTS

Echocardiography failed to visualize the distal segments of the pulmonary arteries in nine patients with unilateral pulmonary arterial interruption or occlusion. In one patient with pulmonary atresia and occluded modified Blalock-Taussig shunt, small central pulmonary arteries of both sides were seen at echocardiography.

Pulmonary arterial segments that were not visualized by angiography with right ventricle or main pulmonary arterial injection or by aortography were the left main pulmonary artery in seven, the hilar segment of the left pulmonary artery in two (both of them with previous left modified Blalock-Taussig shunt) and the entire pulmonary arterial tree in one (patient with previous right modified Blalock-Taussig shunt) (Table 1). Pulmonary vein wedge angiography that was attempted in a 3-month-old infant failed to visualize the pulmonary artery because of incomplete wedging of the catheter in the pulmonary vein. Due to the absence of the patency of the foramen ovale, pulmonary vein wedge angiography could not be performed in a 13-year-old girl.

Proximal interruption of the pulmonary artery

In the patients in whom the left main pulmonary

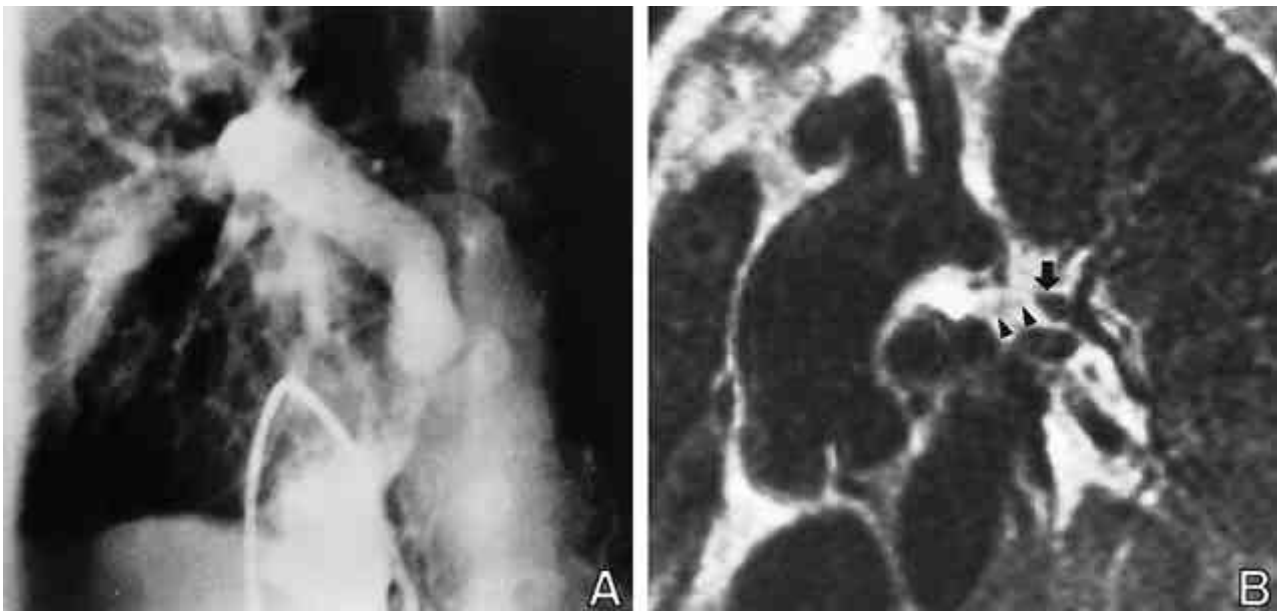

Fig. 1. Proximal interruption of the left pulmonary artery in a patient with tetralogy of Fallot. A: Right ventriculogram shows absent left pulmonary atrery. B: T1-weighted MR image in an oblique coronal plane shows the atretic segment (arrowheads) and patent distal lumen (arrow) measuring 5 mm.

Table 1. Angiographic, MR and surgical findings in 10 patients with pulmonary arterial obstruction

Case No.	Age/Sex	Diagnosis	Pulmonary Arteriography*	Pulmonary vein wedge angiography	MR	Surgery (Name of operation)
1	3m/M	Tetralogy of Fallot; left aortic arch	Non-visualization of LPA	Performed twice. Poorly visible LPA	Hilar LPA, 3 mm-sized	Hilar LPA, 3 mm-sized; (MBT)
2	8y8m/M	Tetralogy of Fallot; left aortic arch; Down syndrome	Non-visualization of LPA	Not done	Hilar LPA, 5 mm-sized	Not done
3	5y1m/M	Aorticopulmonary; window; left aortic arch; pulmonary atresia	Non-visualization of LPA	Not done	No LPA	No LPA
4	8y11m/M	Tetralogy of Fallot; left aortic arch; S/P left MBT	Non-visualization distal LPA; occluded shunt	Not done	No distal LPA; Proximal LPA, 8mm-sized	Hilar LPA, 3 mm-sized; Obstruction of proximal LPA and shunt (angioplasty of LPA)
5	3y4m/F	Tetralogy of Fallot; closing PDA; right aortic arch	Non-visualization of LPA	Not done	Hilar LPA, 4 mm-sized; demonstration with cine MR	Hilar LPA, 3 mm-sized; (total correction with angioplasty of LPA)
6	34y6m/F	S/P Total correction of Tetralogy of Fallot; left aortic arch	Non-visualization of LPA	Not done	No LPA	Absence of LPA; (RV aneurysmectomy)
7	8m/M	Univentricular heart; pulmonary atresia; DORV; right PDA; right aortic arch;	Non-visualization of LPA	Not done	No LPA	Hilar LPA, 3 mm-sized; Left arterial ligament (MBT)
8	13y/F	Pulmonary atresia with VSD; occluded right MBT; left aortic arch	Non-visualization of central pulmonary artery	Not done (no PFO)	Central pulmonary artery was depicted; RPA=6 mm, LPA=5 mm	LPA, 5 mm-sized; transpleural collateral vessels (MBT)
9	17y/M	Tetralogy of Fallot; severe LPA stenosis; right aortic arch S/P left MBT	Non-visualization of distal LPA	Not done	Clearly shown distal LPA, 6 mm-sized	Not done
10	1m/M	Pulmonary atresia with VSD; left PDA; left aortic arch	Non-visualization of LPA	Not done	Clearly shown LPA, 3 mm-sized	Not done

DORV, double-outlet right ventricle; LPA, left pulmonary artery; MBT, modified Blalock-Taussig shunt; PDA, patent ductus arteriosus; PFO, patent foramen ovale; RV, right ventricle; S/P, status post; VSD, ventricular septal defect.
*Right ventriculography or selective pulmonary arteriography.

artery was not visualized by angiography (n=7), MR imaging clearly demonstrated the patent pulmonary arteries and the intervening atretic segment in four patients (case 1, 2, 5, 10) (Fig. 1). MR failed to visualize the hilar left pulmonary artery in the remaining three patients (case 3, 6, 7).

A small pulmonary artery was mistaken for the bronchus in one patient at MR imaging (Fig. 2). In a case with small hilar pulmonary artery, cine MR imaging was helpful in differentiating between the small left pulmonary artery and the bronchus because blood vessels appeared white while the airway was black (Fig. 3). MR imaging in the oblique coronal plane along the left pulmonary artery with the guide of axial images was helpful in the assessment of the length of the atretic segment and the size of the patent hilar pulmonary artery. In all five patients with unilateral pulmonary arterial obstruction and patent distal pulmonary arteries, the distal pul-

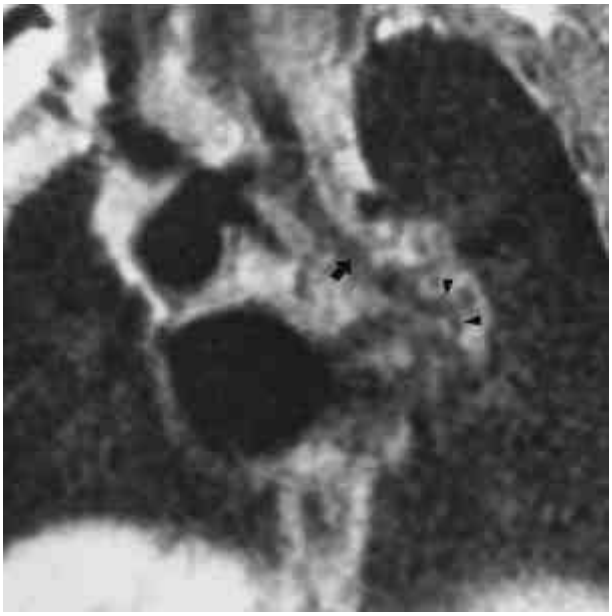

Fig. 2. MR image in oblique coronal plane shows a signal void structure (arrow) in the left hilum. It was not possible to determine whether it represented the left main bronchus or the left pulmonary artery. A slightly tortuous structure in the more peripheral region (arrowheads) seemed to be a left lower lobar pulmonary artery.

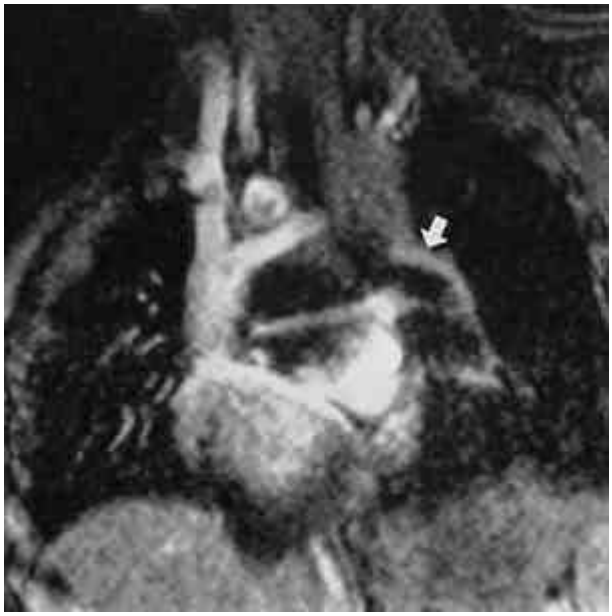

Fig. 3. Cine MR image using gradient echo technique in oblique coronal plane clearly shows a patent pulmonary artery (solid arrow) distal to the interrupted segment in a patient with tetralogy of Fallot.

monary arteries were more clearly shown on the oblique coronal sections than on the axial sections.

Obstruction of the distal main pulmonary artery

In the patients group in whom the hilar pulmonary artery was not visualized by angiography (n=2), MR imaging demonstrated the hilar pulmonary artery measuring 6 mm in one patient (case 9). In the patient (case 4) in whom both angiography and MR imaging failed to demonstrate the hilar pulmonary artery, it measured 3 mm at surgery.

The central pulmonary artery in a patient with pulmonary atresia

In one patient with pulmonary atresia, the confluent central pulmonary artery had been connected to the right modified Blalock-Taussig shunt. The shunt was occluded and the pulmonary artery was invisible on angiograms. On MR images, a 5 mm-sized central pulmonary artery was seen along with rich transpleural collateral vessels.

Correlation between MR and surgical findings

Surgical confirmation of the pulmonary arterial tree was obtained in seven patients including one patient with pulmonary atresia with ventricular septal defect in whom only left pulmonary arterial status was confirmed at surgery. Among them, concordance between MR imaging and surgical findings was found in five patients. Of three patients with discordant MR imaging and surgical findings, two patients showed false negative results at MR imaging. These two patients were found to have 3 mm-sized left hilar pulmonary arteries at surgery. MR findings correlated well with surgical findings in 67% (4 of 6) of the patients with unilateral pulmonary arterial interruption or severe obstruction of distal pulmonary arteries.

Among the five patients with interruption of left pulmonary artery, three patients had 3 mm-sized hilar left pulmonary arteries and left ductal ligaments at surgery while two patients had neither hilar pulmonary arteries nor remnants of ductal structures at surgery.

DISCUSSION

Pulmonary arterial occlusion may be congenital or acquired. Congenital absence of a pulmonary artery is rarely present except in pulmonary agenesis (14). Absence of the mediastinal portion of a pulmonary artery is also termed proximal interruption of the pulmonary artery. The pulmonary artery at the level of hilum can

be identified anatomically by using various imaging modalities (15). When there is distal ductal origin of one pulmonary artery, the absent pulmonary artery and the ductus tend to involve the opposite side of the aortic arch (16).

In patients with pulmonary atresia with ventricular septal defect, the presence of the central pulmonary artery of appropriate size is important in the planning of surgical treatment. The central pulmonary artery is frequently visualized with aortography or selective systemic arterial injection. However, the bronchial arteries have poor communication with the pulmonary arteries and small central arteries can not be filled with contrast media. As in case 8 of our study, even the larger central pulmonary artery could not be visualized by angiography because there was inadequate collateral arterial communication with the artery.

Acquired occlusion of the pulmonary arteries in patients with congenital heart disease is most commonly due to previous surgical procedures such as pulmonary arterial banding, surgical angioplasty, or palliative shunt procedures (6, 11).

When the aortography or selective arteriography fails to visualize the central or hilar pulmonary artery, pulmonary vein wedge angiography can be performed (2, 16). Although this can visualize the arteries that are not opacified by arteriography, it may be complicated by damages to the lung and is often difficult to perform. Computed tomography (CT), spin echo and gradient echo cine MR imaging have been used to evaluate patients with absence of the unilateral pulmonary artery or pulmonary atresia (4-13, 17). Presence or absence of the hilar pulmonary artery of proper size has been evaluated by using these modalities (4, 10-13, 17). MR imaging is considered to be better than CT in the evaluation of the pulmonary arteries because it has multiplanar imaging capability. In previous reports, MR imaging and CT have rarely shown hilar pulmonary arteries of 4-10 mm in diameter in patients with congenital interruption of a pulmonary artery (4, 11, 12, 17). Our cases revealed 3-5 mm hilar pulmonary arteries and interrupted segment of the mediastinal pulmonary artery on MR images.

Oblique coronal plane in the long axis of the mediastinal pulmonary artery was more advantageous than transverse planes in depicting the length of a pulmonary artery. Cine MR imaging in oblique coronal plane is helpful in differentiating between the small bronchus and pulmonary artery because of native contrast between the flowing blood and airway. The distal pulmonary arteries less than 3 mm in diameter may not be visualized at MR imaging with a 0.5 T scanner as in this study.

In a patient with pulmonary atresia, MR imaging was a good imaging modality in depicting the central pulmonary artery which was not visualized on angiography. Occluded modified Blalock-Taussig shunt and transpleural collateral vessels were also well demonstrated on MR images in this case.

The limitations of this study include the small number of patients studied. In most cases except one, pulmonary vein wedge angiography was not performed to visualize the distal pulmonary arterial tree. Although MR imaging was effective for visualization of hilar and central pulmonary arteries in most cases, there were two false negative cases with small hilar arteries. The expense and time taken for MR imaging of these patients can be a hindrance to the use of MR imaging in clinical situations. However, we suggest that MR imaging can be utilized for the patients before cardiac angiography obviating angiography in some cases.

In conclusion, MR imaging is helpful in depicting hilar and central pulmonary arteries in patients with congenital or acquired interruption of the pulmonary artery or pulmonary atresia.

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