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Anomalous Origin of the Right Pulmonary Artery from the Ascending Aorta: Diagnosis by Magnetic Resonance Imaging

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Abstract
Anomalous origin of the right pulmonary artery from the ascending aorta is a rare congenital anomaly. Magnetic resonance imaging (MRI) was performed on three patients with anomalous origin of the right pulmonary artery from the ascending aorta. ECG-gated, T1-weighted, spin-echo MRIs and cine MRIs were obtained. In one patient, postoperative MRI was also obtained. Echocardiography and cardiac catheterization were performed in three patients and angiocardiography was performed in two. MRI clearly showed anomalous origin of the right pulmonary artery from the posterior aspect of the ascending aorta, as well as combined anomalies including patent ductus arteriosus, aortopulmonary window, and interruption of the aortic arch in all three patients. Echocardiography missed this anomaly in all three. We suggest that MRI is an accurate imaging modality in diagnosing anomalous origin of the right pulmonary artery from the ascending aorta, obviating the need to perform angiocardiography.

Key words: Anomalies, congenital—Arteries, abnormalities—Heart, abnormalities—Magnetic resonance imaging

Anomalous origin of the right pulmonary artery from the ascending aorta is a rare congenital anomaly and approximately 80 cases have been reported since 1868. Early diagnosis and operation within the first year of life are mandatory to avoid irreversible pulmonary vascular disease [1–3]. Because clinical findings and chest radiograph are nonspecific, the diagnosis will rest on cardiovascular imaging techniques [4]. Findings of echocardiography [2, 5] and computed tomography (CT) [6] have been described in the literature, but the role of magnetic resonance imaging (MRI) in this anomaly is still to be elucidated. We report three cases of the anomaly in which MRI was utilized. MRI was performed on a 0.5 Tesla scanner (Toshiba Medical Co., Tokyo, Japan). Electrocardiography (ECG)-gated spin echo (SE) T1-weighted images with repetition time (TR) of 416–427 ms and echo delay time (TE) of 20 ms, and cine MRI with TE of 22 ms and flip angle of 20–30° were obtained.

Case Reports

Case 1

A 6-month-old, 7-kg male infant was evaluated for irritability and vomiting of 1-month duration. Auscultation of the heart revealed a grade 3/6 systolic murmur at the left sternal border. The ECG showed right ventricular hypertrophy. The chest radiograph showed a cardiothoracic ratio of 70% and increased pulmonary vascularity. On echocardiography, this anomaly was diagnosed as a large aortopulmonary window. Cardiac catheterization revealed marked pulmonary hypertension (Table 1). Angiocardiography showed an anomalous origin of the right pulmonary artery from the posterior aspect of the ascending aorta, 1.5 cm above the aortic valve, and a patent ductus arteriosus. There was no intracardiac defect.

The axial image (Fig. 1A) and the right anterior oblique sagittal image (Fig. 1B) of ECG-gated, T1-weighted, spin echo MRI clearly showed the anomalous origin of the right pulmonary artery from the posterior aspect of the ascending aorta.

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Table 1. Cardiac catheterization data for three patients with anomalous origin of the right pulmonary artery from the ascending aorta and associated anomalies

	Patient 1		Patient 2		Patient 3	
	Pressure (mmHg)	Oxygen saturation (%)	Pressure (mmHg)	Oxygen saturation (%)	Pressure (mmHg)	Oxygen saturation (%)
MPA	70/23 (41)[a]	65	120/51 (77)	73	123/76 (97)	69
RPA			112/43 (69)	84		
LPA			113/46 (70)	75	149/74 (102)	67
RV	64/−4	63	122/−20	66	145/−3	66
PCW					12/3 (6)	
LV	73/10	91				
AA			115/40 (72)	92	123/76 (97)	88
DA	65/19 (36)	86	53/44 (48)	90		

MPA, main pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery; RV, right ventricle; PCW, pulmonary capillary wedge; LV, left ventricle; AA, ascending aorta; DA, descending aorta
[a] Mean pressures in parentheses

Reimplantation of the right pulmonary artery to the main pulmonary artery, ligation of the patent ductus arteriosus, and lung biopsy specimens of both upper lobes were performed. Pathology revealed mild medial hypertrophy, Health-Edwards grade I, identical in both specimens. The patient was doing well at 1-year follow-up.

Case 2

A 45-day-old, 3.5-kg male infant was admitted for the evaluation of respiratory difficulty and poor phonation since a few days after his birth. His pulse rate was 156/min and respirations were 70/min. The blood pressures of the right and left arms were 90/48 mmHg and 100/41 mmHg, respectively. Those of right and left legs were 64/45 mmHg and 69/43 mmHg, respectively. Auscultation of the heart revealed a grade 3/6 systolic murmur at the left sternal border. The ECG showed right axis deviation and biventricular hypertrophy. The chest radiograph showed a cardiothoracic ratio of 70% and increased pulmonary vascularity. The echocardiographic diagnosis was interruption of the aortic arch type A [7] and patent ductus arteriosus.

The oblique sagittal image (Fig. 2A) of ECG-gated, T1-weighted, spin echo MRI showed interruption of the aortic arch distal to the origin of the left brachiocephalic branches and an anomalous origin of the right pulmonary artery from the posterior aspect of the ascending aorta. The oblique axial image along the plane of the right pulmonary artery (Fig. 2B) showed an anomalous origin of the right pulmonary artery from the ascending aorta and large distal aortopulmonary window [8]. The coronal image (Fig. 2C) also showed a large aortopulmonary window. The MRI diagnosis was Berry syndrome [9, 10].

Cardiac catheterization and angiocardiography confirmed the MRI findings and documented marked pulmonary hypertension in both pulmonary arteries (Table 1).

The patient underwent two-staged corrective surgery. First the interrupted aortic arch was reconstructed by using a 10-mm Goretex tube interposition graft. Ten days later, tunneling of the right pulmonary artery to the main pulmonary artery was performed by using a Dacron patch, and a separate pericardial patch was applied to the vertically incised margin of the anterior wall of the ascending aorta. Postoperative axial MRI (Fig. 2D) showed a patent, reconstructed right pulmonary artery passing anteriorly to the ascending aorta. The patient was doing well at follow-up 13 months after the operation.

Case 3

A 34-year-old man was evaluated for cyanosis. His hemoglobin was 21.4 mg% and hematocrit was 62.9%. Auscultation of the heart showed P2 accentuation without a definite murmur. The ECG showed right ventricular hypertrophy.

The chest radiograph showed a cardiothoracic ratio of 60% and enlarged central pulmonary arteries. On echocardiography, due to an inadequate acoustic window, no cardiac anomaly was detected except right ventricular hypertrophy. Right-sided heart catheterization was performed. The angiographic catheter passed from the main pulmonary artery to the aorta through a patent ductus arteriosus, and severe pulmonary hypertension of the main and left pulmonary arteries was detected (Table 1). The catheter could not enter the right pulmonary artery. Angiocardiography was not performed because of severe pulmonary hypertension. The diagnosis was Eisenmenger syndrome secondary to a large patent ductus arteriosus.

The systolic phase of the axial image along the plane of the right pulmonary artery of a cine MRI (Fig. 3A) showed anomalous origin of the right pulmonary artery from the posterior aspect of the ascending aorta and enlarged central pulmonary arteries. During early diastole (Fig. 3B), there was little decrease of central pulmonary artery sizes compared with the systolic phase. Percentage diameter changes in the main, proximal right, and descending left pulmonary arteries during the cardiac cycle were 4.8%, 18.5%, and 9.5%, respectively.

No surgical intervention was considered due to severe pulmonary hypertension, and the patient was managed medically.

Discussion

Anomalous origin of the right pulmonary artery from the ascending aorta is a rare congenital cardiac abnormality resulting from incomplete leftward migration of the right pulmonary artery [1, 4]. The lesion must be distinguished from other abnormalities in which the origin of right pulmonary artery is atretic but the arterial supply is derived from either a ductus arteriosus or via collaterals between the systemic and pulmonary arteries [1, 2]. Mostly, the anomalous right pulmonary artery arises from the posterior or left posterior aspect of the ascending aorta close to the aortic valve [11]. Early diagnosis and surgical treatment before 12 months of age are imperative, as this malformation leads to irreversible pulmonary vascular disease [1–3].

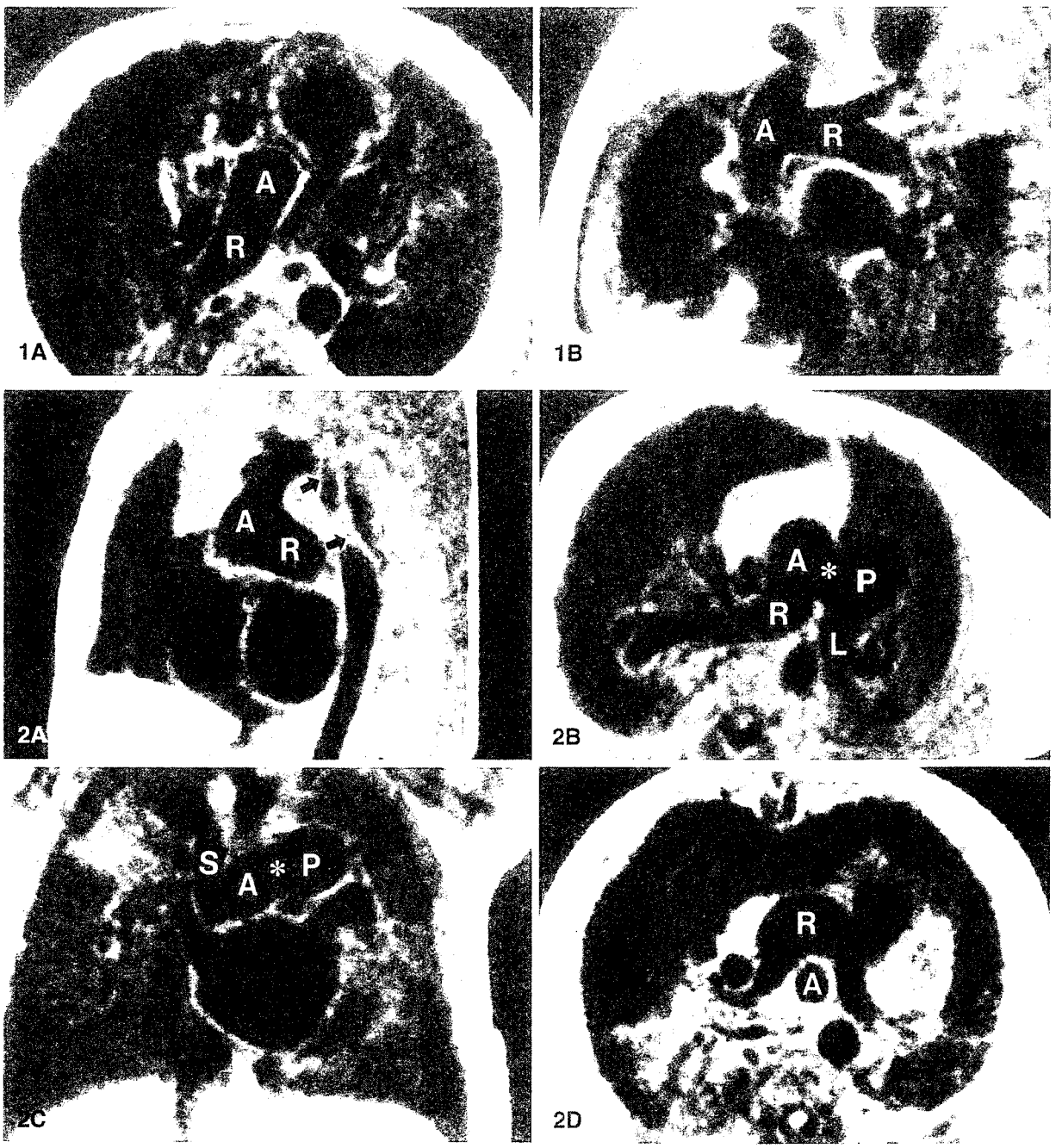

Fig. 1. Case 1. Axial (A) and right anterior oblique sagittal (B) MRIs show the origin of right pulmonary artery (R) from the posterior aspect of the ascending aorta (A).

Fig. 2. Case 2. **A** Oblique sagittal MRI demonstrates interruption of aortic arch (arrows) distal to left bracheocephalic branches as well as the anomalous origin of right pulmonary artery (R) from the posterior aspect of the ascending aorta (A). **B** Oblique axial MRI along the plane of the right pulmonary artery shows a large aortopulmonary window (*) between the ascending aorta (A) and the main pulmonary artery (P) and the anomalous origin of the right pulmonary artery (R) from the ascending aorta (A). L = left pulmonary artery. **C** The coronal MRI also shows the large aortopulmonary window (*) between the ascending aorta (A) and the main pulmonary artery (P). S = superior vena cava. **D** Postoperative axial MRI showing a patent reconstructed right pulmonary artery (R) passing anteriorly to the ascending aorta (A).

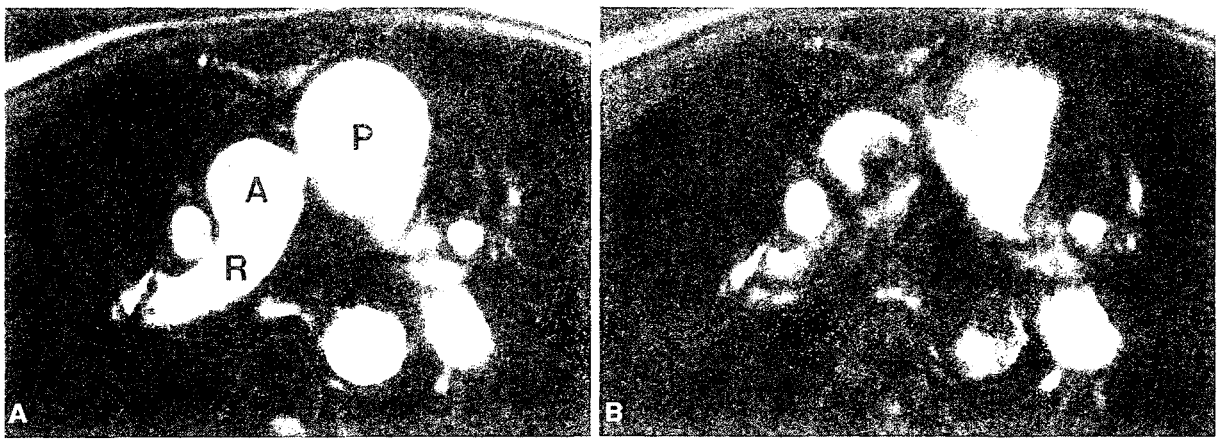

Fig. 3. Case 3. **A** Axial image of a cine MRI along the plane of the right pulmonary artery during systole clearly showing the anomalous origin of the right pulmonary artery (R) from the posterior aspect of the ascending aorta (A). Note that the main pulmonary artery (P) is larger than the ascending aorta (A). **B** During early diastole, there is little decrease of the central pulmonary artery size compared to systole, suggesting pulmonary arterial hypertension.

There are no distinctive clinical features to allow diagnosis of this anomaly, so the diagnosis requires cardiovascular imaging studies [4]. The diagnosis can be made by cross-sectional echocardiography, when a posterior vessel arising from the ascending aorta and supplying the lung can be demonstrated, but the finding can mimic an aortopulmonary window [2], and if the patient has an inadequate acoustic window as in patient 3, echocardiography can miss the anomaly. Echocardiography has the advantage of being safe, noninvasive, and relatively inexpensive to perform, but it suffers from the limitations imposed by body size and habitus and is relatively operator dependent [12]. Cardiac catheterization with angiocardiography is diagnostic and also provides valuable information regarding hemodynamics and pulmonary vascular resistance, but the procedure is invasive and risky in patients with severe pulmonary hypertension.

In our three patients, MRI clearly showed the exact site of the origin of the right pulmonary artery from the ascending aorta, as well as associated complex anomalies, comparable to angiocardiography. In patient 3, cine MRI also provided functional information of pulmonary hypertension by demonstrating the loss of the normal pulsatile systolic caliber increase of the proximal pulmonary arteries [13].

In conclusion, MRI is an accurate and effective imaging modality in diagnosing this rare congenital anomaly, overcoming the limitations of echocardiography and obviating the need for invasive angiocardiography.

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