Magnetic Resonance Imaging of Anomalous Pulmonary Venous Connections

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Purpose: We evaluated the capability of MR in the diagnosis of anomalous pulmonary venous connection (APVC).

Materials and Methods: The patient group consisted of 11 total APVC and 8 partial APVC diagnosed with MR. Echocardiography was performed in all cases, cardiac angiography in 12 cases and operation in 12 cases. We compared MR findings with those of operation, echocardiography and cardiac angiography.

Results: In surgically proven 12 cases, diagnostic accuracy of preoperative MR, echocardiography and cardiac angiography was 100%, 67% and 63%, respectively. In the remaining cases, MR findings well correlated with those of echocardiography or cardiac angiography. Stenosis of common pulmonary vein or superior vena cava was identified in 4 cases. In one patient, MR depicted associated cor triatriatum clearly.

Conclusion: MR is an effective modality in depicting anomalous pulmonary venous connections.

Index Words: Pulmonary veins, MR Heart, MR

Recently magnetic resonance (MR) imaging has been effectively applied to the evaluation of a variety of congenital heart disease (1-6). In most cardiovascular anomalies, angiography has been considered as the "gold standard" (2) and echocardiography has provided real-time tomographic images of the heart. But in the evaluation of the anomalies of pulmonary veins, echocardiography and cardiac angiography have limitations such as poor far-field resolution and small field of view of precordial echocardiography and insufficient definition of overlapping structures on the levophase images of angiography, especially when anomalous pulmonary venous connection (APVC) is not suspected (4, 7-10). In addition, cardiac catheterization and angiography have a risk of cardiac arrest and even death in severely cyanotic patients with total anomalous pulmonary venous connection. The capability of MR for demonstrating normal and abnormal pulmonary veins has been revealed by several studies (2, 4, 5, 11-15). But little informations are available on the MR findings of anomalous pulmonary venous connections in various imaging planes. And the accuracy of MR in the diagnosis of APVC should be further elucidated because the number of case materials in the previous studies was small (11, 15). The purposes of this study were (a) to evaluate the accuracy of MR in the diagnosis of APVC in comparison with that of echocardiography and cardiac angiography, and (b) to report MR findings of APVC in various imaging planes.

SUBJECTS and METHODS

Patients with echocardiographic or angiographic diagnosis of APVC were selected and referred to the radiology department for MR imaging. And patients with MR diagnosis of APVC without echocardiographic or angiographic one of APVC before MR imaging were also included in this study.

The patient group consisted of 19 patients (10 males and 9 females aged from 12 days to 23 years: mean 5.2 years) with APVC. Among them, 11 patients had total anomalous pulmonary venous connection (TAPVC) and 8 patients had partial anomalous pulmonary venous connection (PAPVC).
According to the classification of Darling et al. (16), patients with TAPVC were classified into three groups. There were 6 cases with TAPVC at the supracardiac level (3 to the left innominate vein, one to the left superior vena cava, one to the right superior vena cava and one to the azygos vein), 4 at the cardiac level (3 to the coronary sinus, one to the right atrium) and one at the mixed levels (right upper and middle pulmonary veins to right superior vena cava and left common pulmonary vein and right lower pulmonary vein to right atrium, respectively). Among the 11 patients, both preoperative and postoperative imaging were performed in two cases and postoperative imaging only in one case.

The group with PAPVC consisted of 4 right upper and lower pulmonary veins to the right atrium, 1 right upper and middle pulmonary veins to the right atrium, 2 right upper pulmonary vein to the right superior vena cava, 1 left upper pulmonary vein to the left innominate vein associated with cor triatriatum.

Stenosis of common pulmonary vein was found in 3 TAPVC and stenosis with aneurysm of superior vena cava in one TAPVC.

Echocardiography was performed in all patients and cardiac angiography was performed in 6 patients with TAPVC and in 6 patients with PAPVC. APVC was confirmed operatively in 12 patients. In one patient with pulmonary atresia receiving staged operation, operatively confirmed PAPVC was demonstrated on MR imaging after palliative surgery.

**MR Imaging Technique**

MR imaging was performed on a 0.5T superconductive system (MRT 50A: Toshiba Medical Co., Tokyo, Japan) with a body coil. Electrocardiogram (ECG)-gated multislice spin echo (SE) technique with echo time of 20–30 msec was used. The interpulse interval (TR) was determined by the R–R interval of ECG of each patient. Respiratory gating was not applied because of negligible effects of respiratory motion in a small number of patients. Chloral hydrate (75–100 mg/kg) was given orally or through rectum in young patients for sedation. In all patients, T1-weighted transaxial, coronal or oblique coronal images were obtained with a 4–6mm slice thickness. In some patients, sagittal images were obtained additionally. In 4 patients, cine MR images along the longitudinal plane of a common pulmonary vein or an anomalous vein were obtained by field echo technique (echo time=22 msec, flip angle=20) with a 7–8mm slice thickness.

**Analysis of Echocardiography and Cardiac Angiography**

Echocardiography was performed before MR imaging in each case. In 12 cases, both precordial echocardiography and cardiac angiography were performed before MR imaging. Echocardiography was done with a color doppler equipment (Acuson 128, Acuson Co., California, U. S. A.) using either a 2.5- or 3.5 MHz transducer by an experienced pediatric cardiologist (H. J. L.). Cardiac angiography and catheterization were performed by J. K. K. In 11 patients with presumed echocardiographic diagnosis of APVC, selective pulmonary arteriography was performed. In cases of APVC neglected by echocardiography, right side ventriculography was performed. In one case with TAPVC, retrograde vertical venography was performed. Ang

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**Fig. 1.** Total anomalous pulmonary venous connection to the left innominate vein in right isomerism, univentricular atrioventricular connection and pulmonary stenosis.

a. Axial section shows the right lower pulmonary vein (open arrow) draining into the common pulmonary vein behind the left atrium. Note the small left atrium, secundum atrial septal defect, single ventricle and stagnant flow within the pulmonary vein.

b. The vertical vein (open arrow) is seen on the coronal section. Note the vein connecting common pulmonary vein with left atrial roof (arrow).

**Fig. 2.** Partial anomalous venous connections at cardiac level (right side total anomalous pulmonary venous connection to the right atrium). Right lower pulmonary vein enters right atrium. Note the inferior sinus venous atrial septal defect(*) and atelectatic changes in both lungs.
Fig. 3. Total anomalous pulmonary venous connection at mixed levels.
a. Axial image shows the right upper pulmonary vein draining into the superior vena cava (arrow).
b. Axial section at the atrial level shows the left pulmonary vein draining into the right atrium.
c. Coronal image shows the right upper and middle pulmonary veins (arrows) draining into the superior vena cava.

Table 1. Sites of Anomalous Pulmonary Venous Connections and Diagnosis with MR Imaging, Precordial Echocardiography and Cardiac Angiography Correlated with Surgical Confirmation in 19 Patients

<table>
<thead>
<tr>
<th>Pt No.</th>
<th>Age/Sex</th>
<th>Sites of APVC/Associated Defects</th>
<th>Accurate Diagnosis with Surgical Confirm.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3y/F</td>
<td>TAPVC to coronary sinus/sec. ASD</td>
<td>Yes / Yes / Yes / ND</td>
</tr>
<tr>
<td>2</td>
<td>7m/F</td>
<td>TAPVC to coronary sinus/two sec. ASD's</td>
<td>Yes / Yes / ND / Yes</td>
</tr>
<tr>
<td>3</td>
<td>9m/M</td>
<td>TAPVC to coronary sinus/sec. ASD, PDA</td>
<td>Yes / Yes / ND / Yes</td>
</tr>
<tr>
<td>4</td>
<td>5y11m/M</td>
<td>TAPVC to RA/UVH, rt isom., prim. and sec. ASD, CPV sten.</td>
<td>Yes / Yes / Yes / Yes</td>
</tr>
<tr>
<td>5*</td>
<td>1y1m/M</td>
<td>TAPVC to LIV/rt isom., CAVSD, DORV, PS</td>
<td>Yes / No / No / Yes</td>
</tr>
<tr>
<td>6*</td>
<td>8m/M</td>
<td>TAPVC to RA and R SVC/rt isom., PS, UVH</td>
<td>Yes / No / No / Yes</td>
</tr>
<tr>
<td>7</td>
<td>5m/M</td>
<td>TAPVC to aneurysmal L SVC/dextrocardia, DORV, rt isom., PDA, sec. ASD, single AVV, sten. of L SVC, VSD</td>
<td>Yes / Yes / Yes / ND</td>
</tr>
<tr>
<td>9**</td>
<td>6y/M</td>
<td>CPV to RA anast. in TAPVC to azygous vein, BCPS, PS, rt isom., sec. ASD, TA, VSD</td>
<td>Yes / Yes / ND / ND</td>
</tr>
<tr>
<td>10</td>
<td>12d/F</td>
<td>TAPVC to LIV/PDA, sec. ASD, sten. of vertical vein</td>
<td>Yes / Yes / ND / ND</td>
</tr>
<tr>
<td>11</td>
<td>1m/F</td>
<td>TAPVC to RSVC/bil. SVC, CAVSD, rt isom., rt aortic arch, SA, UVH, sten. of retrobronchial CPV</td>
<td>Yes / Yes / ND / ND</td>
</tr>
<tr>
<td>12</td>
<td>1y5m/M</td>
<td>PAPVC (RU, RM) to RA/sec. ASD</td>
<td>Yes / Yes / Yes / ND</td>
</tr>
<tr>
<td>13</td>
<td>2m/M</td>
<td>PAPVC (LU) to LIV/cor triat., small L SVC to coronary sinus</td>
<td>Yes / Yes / Yes / ND</td>
</tr>
<tr>
<td>14</td>
<td>5m/M</td>
<td>PAPVC (RU, RL) to RA/sec. and sin. ven. ASD, VSD</td>
<td>Yes / Yes / Yes / Yes</td>
</tr>
<tr>
<td>15</td>
<td>23y/F</td>
<td>PAPVC (RU) to RSVC/bil. SVC, straight back syndrome</td>
<td>Yes / Yes / Yes / ND</td>
</tr>
<tr>
<td>16</td>
<td>4y3m/F</td>
<td>PAPVC (RU, RL) to RA, sin. ven. ASD, it deviation of prim. IAS</td>
<td>Yes / No / Yes / ND</td>
</tr>
<tr>
<td>17</td>
<td>4m/F</td>
<td>PAPVC (RU, RL) to RA, sec. ASD</td>
<td>Yes / Yes / ND / Yes</td>
</tr>
<tr>
<td>18**</td>
<td>14y/F</td>
<td>PAPVC (RU) to RSVC, PA with VSD, rt unifocalization op.</td>
<td>Yes / No / No / Yes</td>
</tr>
<tr>
<td>19</td>
<td>1y8m/M</td>
<td>PAPVC (RU, RM) to RA, sin. ven. ASD</td>
<td>Yes / Yes / ND / ND</td>
</tr>
</tbody>
</table>

—MR imaging was performed before and after the surgical operation in two cases (*) and postoperatively only in two (**). anast. = anastomosis, Angio = cardiac angiography, ASD = atrial septal defect, AVV = atrioventricular valve, BCPS = bidirectional cavopulmonary shunt, bil. = bilateral, CAVSD = complete atroventricular septal defect, confirm. = confirmation, CPV = common pulmonary vein, DORV = double outlet right ventricle, Echo = echocardiography, IAS = interatrial septum, isom. = isomerism, LA = left atrium, LIV = left innominate vein, L SVC = left superior vena cava, LU = left upper, Lt = left, op. = operation, ND = not done, PA = pulmonary atresia, PAPVC = partial anomalous pulmonary venous connection, PDA = patent ductus arteriosus, prim. = primum, PS = pulmonary stenosis, RA = right atrium, RL = right lower, RM = right middle, R SVC = right superior vena cava, RU = right upper, rt = right, SA = single atrium, sec. = secundum, sin. = sinus, ste. = stenosis, ven. = venous, TA = tricuspid atresia, triat. = triatriatum, TAPVC = total anomalous pulmonary venous connection, UVH = univentricular heart, VSD = ventricular septal defect.
iographic images insufficient for the definite diagnosis of APVC or visualization of venous anatomy were regarded as 'nondiagnostic'.

**Analysis of MR Findings**

In 17 cases, MR imaging was performed for the evaluation of diagnostic accuracy of MR. Some of echocardiographic or angiographic findings such as existence of APVC and associated defects were informed to the radiologists(Y. H. C., H. S. K., J. E. K.) during MR imaging. The MR diagnostic criteria of TAPVC were no demonstrable pulmonary venous connection to the left atrium and identification of the common pulmonary vein or pulmonary venous connection to the right atrium. The MR diagnostic criteria of PAPVC were absence of one or more normal individual pulmonary venous connections and identification of anomalous pulmonary venous connection sites.

MR was regarded as 'diagnostic' only if there was a clear evidence of anomalous pulmonary venous connection with above criteria independently of the informations of echocardiography and cardiac angiography.

**RESULTS**

Table is the summary of patients' data. All the surgically proven 12 cases were accurately diagnosed with MR imaging (Figs. 1, 2). In remaining patients without operative confirmation, MR imaging findings well correlated with those of cardiac angiography of echocardiography. Pulmonary venous anomalies were not suspected at echocardiography in 5 cases (2 PAPVC, 3 TAPVC's). And in one case, an initial echocardiography failed to diagnose the TAPVC to the right atrium and stenosis of the common pulmonary vein, which was detected on the next examination. The levophase angiographic images were not enough for the diagnosis and determination of pulmonary venous drainage sites in two cases of TAPVC with small pulmonary arteries(Table 2). And in one case with TAPVC, a selective pulmonary venography was performed only in the right upper pulmonary vein resulting in a suspicion of right-sided PAPVC, while MR accurately detected the TAPVC at mixed levels(Fig. 3). In three postoperative patients with TAPVC, MR showed patent anastomosis between the common pulmonary vein and atrium.

MR depicted stenotic sites of the common pulmonary vein in 3 cases(Fig. 3). Intraluminal high signal intensity suggested slow flow of the common pulmonary vein and the stenosis of common pulmonary vein in case(Fig. 4). Compression of common pulmonary vein between the anatomic structures seemed to result in stenosis of common pulmonary vein in two cases. Instead of lying anterior to the pulmonary artery, the common pulmonary vein passed between the left bronchus and pulmonary artery in one case and passed posterior to the right bronchus in one case with right aortic arch. The common pulmonary veins below the stenotic segment were engorged and signal intensity of lungs increased on T1 weighted images suggesting pulmonary edema. Stenosis of the left superior vena cava and aneurysmal change above the stenotic segment were also demonstrable on MR images in one case.

In one PAPVC to the left superior vena cava, MR depicted associated cor triatriatum and a vestigial segment of the left superior vena cava draining into the coronary sinus as well on coronal section(Fig. 5).

In the case with a common pulmonary vein, pulmonary venous convergence into the common pulmonary

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**Table 2. Diagnostic Accuracy of Preoperative MR, Cardiac Angiography and Precordial Echocardiography in 12 Patients with Anomalous Pulmonary Venous Connections Correlated with Surgical Findings**

<table>
<thead>
<tr>
<th>Disease</th>
<th>MR</th>
<th>Cardiac Angiography</th>
<th>Precordial Echocardiography</th>
</tr>
</thead>
<tbody>
<tr>
<td>PAPVC</td>
<td>5/5(100%)</td>
<td>4/4(100%)</td>
<td>4/5(80%)</td>
</tr>
<tr>
<td>TAPVC</td>
<td>7/7(100%)</td>
<td>1/4(25%)</td>
<td>4/7(57%)</td>
</tr>
<tr>
<td>Total</td>
<td>12/12(100%)</td>
<td>5/8(63%)</td>
<td>8/12(67%)</td>
</tr>
</tbody>
</table>

PAPVC = partial anomalous pulmonary venous connection, TAPVC = total anomalous pulmonary venous connection.
vein was well depicted on the coronal sections parallel to the vascular axis with about 10°-30° obliquity to the table-top plane. Coronal images also provided informations about the atriobronchial situs in the right isomerism. Combined axial, coronal and sagittal imaging clarify the vascular anatomy and orientation of anomalous veins.

In two cases, partial volume averaging effect of MR sections resulted in the erroneous interpretation of the individual pulmonary venous draining sites. In one TAPVC, the left upper pulmonary vein entered the left atrium on the coronal sections, and in another one PAPVC, the right upper pulmonary artery simulated right the upper pulmonary vein draining into the right superior vena cava on the oblique sagittal sections. Cor triatriatum was associated in one case with left upper PAPVC to the left innominate vein and was correctly diagnosed by all modalities. In one case with TAPVC to the left innominate vein, markedly bulged fossa ovalis was misdiagnosed as combining with cor triatriatum preoperatively.

**DISCUSSION**

In the diagnosis of congenital heart disease, two-dimensional echocardiography plays a primary role, and in selected cases cardiac catheterization and angiography confirm the diagnosis. The combination of echocardiography and cardiac angiography still remain as a gold standard in the diagnosis of congenital heart disease. But this may not always be true in the cases with APVC. Poor far-field resolution, overlying lung and bronchial tissue and small field of view hinder proper precordial echocardiographic evaluation of pulmonary veins (8, 11, 17). Cardiac catheterization and angiographic study highly depend on the informations obtained by echocardiography. In cases with or without correct echocardiographic informations about the pulmonary veins, poor levophase images of pulmonary veins and insufficient selective pulmonary venograms make the diagnosis difficult (11, 18). But this study indicates a high accuracy of MR imaging in the identification of the APVC.

According to the several article (7, 19, 20), the detection of the TAPVC has been successful by using two-dimensional echocardiography. Two-dimensional echocardiography with the Doppler flow analysis may be useful for the detection of TAPVC (19, 21, 22). But in this study, precordial echocardiography appears to have limitations because among the surgically-proven cases, only 67% of TAPVC was correctly diagnosed at echocardiography. While transesophageal cross-sectional echocardiography or color flow mapping using a pediatric probe can successfully document the sites of pulmonary venous drainage (8), it is a semi-invasive procedure and has a limitation of scanning planes.

Angiographic demonstration of TAPVC is limited when the pulmonary blood-flow is decreased. The poor levophase images of pulmonary arterial flow makes it difficult to determine the draining site of pulmonary veins correctly. With the lack of echocardiographic informations about the anomalous pulmonary venous connection due to the inherent limitations of echocardiography, the angiographic evaluation of patients often results in insufficient studies. In this study, the lesions not suspected at echocardiography (two TAPVC's) have been also overlooked at the cardiac angiography. On the contrary, MR imaging depicted all cases of TAPVC correctly.

In one case with TAPVC at cardiac level, stenosis of common pulmonary vein which had been initially overlooked at echocardiography, was correctly diagnosed at angiography and MR.

The typical findings of TAPVC on the axial MR images are 1) venous structures posterior to the left atrium (so the authors coined "the retroatrial vein sign") maybe specific for TAPVC) (2), 2) small left atrium and ventricle (23), 3) atrial septal defect (in most cases), and 4) a vertical vein (in some cases). The MR images in the coronal plane parallel to the table top or with 10°-30° obliquity along the plane of the common pulmonary vein are compatible with anteroposterior angiographic views and make the anatomical appreciation of TAPVC easy. The high signal intensity of the pulmonary vein in this study and a report (24) indicates stenosis of pulmonary vein clearly. Cine MR imaging is
also helpful in the evaluation of the stenosis of the common pulmonary vein.

In a certain type of congenital heart disease, TAPVC is found frequently as in this study. In the group of asplenia syndrome, according to one series, 88% had anomalies of pulmonary venous connection, especially TAPVC(5). Therefore, MR images of this patient group should be scrutinized for the detection of the TAPVC.

Most patients with PAPVC are asymptomatic and sometimes incidentally found on the chest CT scans(10, 25, 26). About 50% of the cases with PAPVC have atrial anomalies of asplenia syndrome, according to one series, 88% had also occasionally found on the chest CT scans(10, 25, 26). The superior sinus venous defects(10, 25, 26), septal defects(10). The superior sinus venous defects are well depicted on the 4-chamber view of right upper pulmonary venography. The data of catheterization in PAPVC are from the partial volume average artifacts between the pulmonary arteries and the superior vena cava.(5) Therefore, MR images of this patient is found frequently as in this study. In the group of PAPVC, separate drainage sites of individual pulmonary veins are well depicted on the sagittal images. Some misinterpretation of drainage site of PAPVC of upper pulmonary veins may result from the partial volume average artifacts between the contiguous pulmonary arteries and the superior vena cava or between the left atrial wall and pulmonary veins.

Cor triatriatum has been reported to be depicted better on MR than echocardiography and cardiac angiography(11). In this study, all modalities correctly diagnosed cor triatriatum.

In conclusion, MR imaging is effective in depicting anomalous pulmonary venous connections. MR has a superior diagnostic accuracy compared with echocardiography and cardiac angiography in the diagnosis of anomalous pulmonary venous connections.

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폐정맥 환류이상의 자기공명영상 진단

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목적: 폐정맥 환류이상에 대한 자기공명영상(MR)의 진단능력과 그소견을 알아보고자 하였다.

대상 및 방법: MR에서 폐정맥 환류이상으로 진단된 소아 및 성인 19예(평균연령 5.2세)에 대하여 전예에서 시행한 심초음파검사, 12예에서 시행한 심혈관 조영술, 12예에서 시행한 수술소견을 MR소견과 비교하였다.

결과: 수술로 확인한 12예에서 수술전 진단정확률은 MR이 100%, 심초음파검사가 67%, 심혈관조영술이 63%였으며, 수술후도 7예에서 MR소견과 심초음파 검사 또는 심혈관조영술소견이 일치되었고 폐정맥의 협착(3예)과 상대정맥의 협착(1예), 그리고 1예의 심심방실도 MR로 확인하였다.

결론: MR이 폐정맥 환류이상의 진단에 효과적인 진단방법임을 알 수 있었다.
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(1) 일정:
① 6월 19일(일) - Breast
② 9월 11일(일) - Nuclear Medicine
③ 11월 6일(일) - Head and neck

(2) 문의: 연세의대 신촌세브란스병원 방사선과 사무실 (361-5837)

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