The Role of MR Imaging in Determination of Atrial Situs in Congenital Heart Disease with Situs Ambiguus

Su Mi Park, M.D., Yong Kook Hong, M.D., Je Whan Won, M.D.
Hyang Mee Lee, M.D., Kyu Ok Choe, M.D., Jae Young Choi, M.D. 1, Jong Kyun Lee, M.D. 2
Jun Hi Sul, M.D. 2, Seung Kyu Lee, M.D. 2, Yong Whan Park, M.D. 3, Bum Koo Cho, M.D. 2

Purpose: to assess the role of MR imaging in determining of the atrial situs in complicated congenital heart disease with situs ambiguus.

Method and Materials: In order to classify the situs, the morphology of atrial appendages, on bronchial length ratio, the superior-inferior relation of the pulmonary artery (PA) and main bronchi on each side, and splenic abnormality were evaluated by MR imaging in 22 patients (12 boys and 10 girls), and the results were compared.

Results: In all patients, the superior-inferior relation of the PA and main bronchi tended to lateralize, and in one, bronchial length ratio was not consistent with the relation between the PA and bronchus. Bronchial and atrial situs, as determined by appendage morphology, were consistent in ten of 13 right isomerism patients, and in only three of nine of these with left isomerism. All 13 right isomerism patients, classified by the relation of the PA and main bronchi, showed asplenia, whereas eight of nine of these with left isomerism had polysplenia.

Conclusion: In the assessment of atrial situs by MR imaging, the positional relation of a bronchus and the PA, bronchial length ratio, and splenic abnormality are constant and reliable. The accuracy of classification of situs on the basis of atrial appendage morphology is, however, limited.

Index Words: Heart, abnormalities
Heart, MR
Magnetic resonance (MR), in infants and children

Situs ambiguous is the anatomically uncertain or indeterminate type of visceral situs. Atrial isomerism is a subset of situs ambiguous and is a condition in which the right-sided and left-sided atria are morphologically similar (1, 2). It is not isomerism of the atrial appendage that produces problems, but the associated cardiac defects associated with atrial isomerism that present difficulties in clinical management and prognosis (3).

For logical sequential segmental analysis of a complicated congenital anomaly, a description of cardiac situs is, however, essential (4, 5).

With regard to anatomic description in situs ambiguous of cardiac anomaly, there are two schools of thought. Van Praagh (6) believes that there are only two types of atrial situs: solitus and inversus. Situs solitus is the normal visceral atrial situs; the morphologically right atrium (RA) is right-sided, and the morphologically left atrium (LA) is left-sided. Situs inversus is the mirror image or inverted type of visceral atrial situs; one atrial appendage is morphologically right and the other, morphologically left. Van Praagh confined the meaning of situs ambiguous atrialis to an undiagnosed case, on the other hand, on the basis of its atrial morphology. Van Mierop and Wiglesworth (2) and many others (7-11), advocate atrial isomerism, in which both atria have similar internal and external configuration and appendage morphology, and are considered either bilaterally RA or LA. Atrial situs is most usefully determined by the morphology of the atrial appendages (8, 12, 13).

In clinical study, angiography (14, 15), chest radi-
Su Mi Park, et al: The Role of MR Imaging in Determination of Atrial Situs in Congenital Heart Disease with Situs Ambiguus

Acography(11, 16, 17), ultrasonography(9, 10, 18) radioisotope scanning(19) or computed tomography(18, 20) have been the modalities used to determine cardiac situs, but each method has inherent limitations due to interpretation or risk due to its invasiveness. Because of its wide field of view, good tissue contrast and depiction of cardiovascular structure without contrast media injection, MR imaging is now being applied to the assessment of cardiac disease(21). It can clearly delineate cardiovascular structure, major airways, and upper abdominal structure, and, moreover, can superbly visualize major arteries and veins(22, 23) which are poorly visualized by echo or may sometimes be difficult to visualize even by angiography. The purpose of this study is to evaluate by MR imaging the morphology of the atrial appendage, the ratio of bilateral bronchial length, the superior-inferior relation of the pulmonary artery and main bronchi on each side, the atrium receiving blood from the IVC and also splenic abnormality, and to assess the ability of MRI to identify each anatomic landmark in order to determine cardiac situs.

**Materials and Methods**

Twenty-two patients with asplenia or polysplenia syndrome combined with congenital heart disease were the subjects of this study. They were twelve boys and ten girls and their age ranged from 2 months to 13 years. At the same time, to determine the accuracy with which cardiac situs was recognized, the authors evaluated 16 adult patients with situs solitus hypertrophic cardiomyopathy. The morphology of both atrial appendages was observed on MR by two board-certified cardiopulmonary radiologists and a chief resident, working independently without any clinical information. The criteria for discriminating right and left atrial appendages were as follows. If the appendage was triangular-shaped and had a broad base, with or without prominence at the base due to crista terminalis shadow, it was classified as right; if its shape was long and finger-like, with a narrow base, it was classified as left. The ratio of bronchial length was calculated by measuring and dividing the length of the right and left main bronchi from the carinal angle to the take off point of the upper lobar bronchi. The superior-inferior relation of the main bronchi and pulmonary arteries(PAs) on each side was observed, a main bronchus being defined as one before bifurcation to an intermediate or lower lobar bronchi. The superior-inferior relation of the main bronchi and pulmonary arteries(PAs) on each side was observed, a main bronchus being defined as one before bifurcation to an intermediate or lower lobar bronchi. The name 'epiarterial bronchus' was given to one superior to the PA, and 'hyparterial', if the reverse relation was seen. The presence of a suprahepatic segment of IVC, a seg-
Fig. 2. Pulmonary artery-bronchus relation. Bilateral epiparterial (A) and hyparterial (B) bronchi are clearly demonstrated (arrows: main bronchi, arrowheads: pulmonary arteries).

Fig. 3. Three-year-old male patient who shows discrepancy between bronchial length ratio and pulmonary artery-bronchus relation. The bronchial length ratio of right to left is 2, but pulmonary artery-bronchus relation was bilateral hyparterial bronchi. There is total anomalous pulmonary venous return through vertical vein (arrowhead). (large arrows: pulmonary arteries, small arrows: bronchi)

ment of vessel between the superior dome of the liver and draining atrium, as well as the side and appendage morphology of draining atria, were the criteria on which the status of the liver, spleen and stomach was determined. When there was disagreement, the matter of right or left sidedness was decided by consensus between the three radiologists.

MR images were obtained with a 1.5 Tesla system (Signa General Electric, Milwaukee, Wis.), (with a head coil when children were small enough), using the ECG gated multislice spin echo technique (TR=1 or 2 R–R interval according to heart rate, and TE=20–30msec), T1 weighted images with 3–5mm slice thickness were obtained in the axial, transverse and sagittal planes.

Results

Adult patients with situs solitus

All the right atrial appendages were recognized as being triangular-shaped and with a broad base, and in all cases, the left appendages were also identified. In six patients, however the left appendages were not clearly defined; eleven had a narrow base and were tubular-shaped, but four had a broad base and were triangular-shaped.

All left bronchi were longer than those on the right, with all length ratios greater than 1.9 (mean...
Su Mi Park, et al: The Role of MR Imaging in Determination of Atrial Situs in Congenital Heart Disease with Situs Ambiguus

Fig. 4. Left isomerism with total anomalous hepatic venous connection. Hepatic veins (small arrows) drain respectively to each side atria with interruption of inferior vena cava and hemiazygous continuation (large arrow).

2.5 ± 0.38). All right main bronchi were epiarterial in relation to the pulmonary artery, whereas the left bronchi were hyparterial.

Pediatric situs ambiguus patients
Atrial appendage
With regard to 24 of 44 appendages (55%), agreement as to right or left sidedness was independently reached by two radiologists; the 20 appendages representing disagreement were reviewed, and three radiologists reached a consensus. The result was atrial isomerism in 16 cases (right isomerism 13, left isomerism 3), situs solitus in four, and indeterminate in two.

Bronchial length ratio and PA-Br relation
The PA-Br relation was bilaterally hyparterial in nine cases, and bilaterally epiarterial in 13; there was no interobserver variation. Bronchial length ratios were less than 1.5 in all except one case, in which the right to left length ratio was 2:1, as in situs inversus. The PA-Br relation in this patient was bilaterally epiarterial bronchi.

The atrium received blood from the IVC or hepatic vein
In patients without interruption of the IVC, drainage was to the bottom of a right-sided atrium in two cases, to a left-sided atrium in ten, and to the middle of a common atrium in one. In nine patients in whom the IVC was interrupted, blood

Fig. 5. Left isomerism with polysplenia. Axial MR image shows multiple coalescent nodules (arrows) in retrogastric area.
Fig. 6. Four-month-old female patient who shows discrepancy between atrial morphology and pulmonary artery-bronchus relation. Left sided atrial appendage (white arrow) shows narrow base and tubular shape, but right sided atrial appendage shows relatively broad base and triangular shape. Bilateral bronchi (arrowheads) are hyparterial.

Table 1. Comparison of Bronchial Situs and Situs Determined by Atrial Appendage Morphology in MR Imaging.

<table>
<thead>
<tr>
<th>Atrial appendage</th>
<th>Bronchial situs*</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Rt Isomerism</td>
<td>Lt Isomerism</td>
</tr>
<tr>
<td>Rt isomerism</td>
<td>10</td>
<td>3</td>
</tr>
<tr>
<td>Lt isomerism</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Situs solitus</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Indeterminate</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*according to the superior-inferior relation of pulmonary arteries and main bronchi

Favor the liver drained via the hepatic vein directly to the atrium. The branches of hepatic vein formed a confluence and then drained to a right-sided (n=4), left-sided (n=2) or middle of common atrium (n=2). In the remaining one case, the confluence of two hepatic veins as seen; these, respectively, drained to the atrium on each side.

Abdominal organs
The spleen was absent in 14 patients and multiple in eight. In these latter cases, it was seen as multiple coalescent nodules in the retrogastric area. The liver was symmetric in 13 patients, rather right-sided in five and rather left-sided in four, while the stomach was right-sided in nine, left-sided in four, and not detected in nine.

Comparison of bronchial situs with shape of atrial appendages
Because bronchial situs showed no interobserver variation, and predominantly tended to show laterization, the author considered this situs to be the most reliable method of situs determination, and on the basis of atrial appendage or splenic abnormality, bronchial and other situs were compared. As in cases determined according to bronchial length ratio, a bilateral epiarterial relationship between bronchi and pulmonary arteries was seen in situs inversus. Bronchial and atrial situs were consistent in 10 of 13 cases of right isomerism but in only three of nine of left isomerism (Table 1); a total of 13 of 22 cases thus showed consistency. Compared to the result in cases of right isomerism, that in left isomerism proved to be particularly inaccurate when determined by the method of atrial appendage morphology.
The Role of MR Imaging in Determination of Atrial Situs in Congenital Heart Disease with Situs Ambiguus

Comparison between bronchial and abdominal situs
When bronchial and abdominal situs were compared, all 13 right isomerism patients showed asplenia, while in eight of nine with left isomerism, polysplenia was seen(Table 2). In right isomerism, the incidence of abnormal systemic and pulmonary venous return was seven cases of TAPVR, and in left isomerism, nine cases of interruption of IVC and four of PAPVR(Table 3).

Discussion
Visceral heterotaxy syndrome is failure or incomplete expression of lateralization of the thoracic and abdominal organs(1). Splenic anomaly is not always accompanied by symmetry of the thoracic organs (24), and when such isomerism of the tracheobronchial branching pattern is present, this is not always associated with symmetry of the atria(13). Ninety-six percent of patients with visceral heterotaxy syndrome showed atrial isomerism(13); in 18%, splenic anomaly was different from cardiac situs; three of 22 left isomerism patients had a single or bilobed spleen, while all with right isomerism were asplenic(5). Cardiac situs may thus be determined according to the nature of the corresponding atrial anatomy. In large autopsy (7) and small clinical series (16), atrial and thoracic situs always corresponds. Caruso and Becker(8) reported abdominal heterotaxy with asplenia; in three cases, tracheobronchial tree and atrial anatomy were discordant. Overall, less than 1% of cases showed discordance between bronchial and atrial situs(13), and this is why cardiac situs is most reliably determined according to atrial situs. For the identification of atrial anatomy, the morphology of the atrial appendage is the prime feature.

Atrial type has no bearing on clinical significance ; rather venous connections have a major impact on clinical profile(3, 25). The type of atrium can be reliably distinguished by the shape of its appendage, which represents the true atrium in the sense of embryologic development, while the major part of the definitive atrium is formed by secondary uptake of either sinus venosus tissue or pulmonary vein(26). Sharma et al (12) observed 1842 heart specimens, and in all of which able to determine through study of the atrial appendage whether the atrium was right- or left-sided. This did not mean, however, that the atrial appendages showed features typical of appendages in situs solitus, but that their salient features enabled right or left sidedness to be distinguished.

In patients with visceral heterotaxy syndrome, cardiac anomaly is usually complicated, and for safety reasons, the amount of contrast media is limited; in cardiac angiography, the rule is not to opacify the atrial appendage for the purpose of situs determination. MRI, on the other hand, can delineate the atrial appendage without additional effort. In seven of 17 cases of right isomerism, Wang et al(27) obtained MR images which were adequate for the identification of atrial situs. In normal atria, left appendages are narrower and more hooked, and so were not clearly defined and sometimes appeared triangular-shaped due to the partial volume averaging effect. Moreover, Sharma et al(12) stated that in congenitally malformed heart, appendages in left isomerism were, compared to right isomerism, hypoplastic or virtually absent (12/190 cases), or that their direction was downward(181/190). In isomerism, the left atrial appendage therefore tended to be more atypical, and this seems to be why only one third of left isomerism

| Table 3. Summary of Systemic Venous Return and Cardiac Anomalies Associated to Right and Left Isomerism Observed at MR Imaging. |
|---------------------------------|-----------------|
|                                | Right Isomerism | Left Isomerism |
|                                | (n=13)          | (n=9)          |
| SVC                            | 5               | 8              |
| BSVC                           | 4               | 1              |
| LVSC                           | 4               | —              |
| IVC                            | —               | 9              |
| Hepatic vein                   | —               | —              |
| Normal                         | 8               | —              |
| Partial AHVC                   | 4               | —              |
| Total AHVC                     | 1               | 9              |
| Cardiac                        | —               | —              |
| AV canal                       | 13              | 7              |
| Single ventricle               | 6               | 3              |
| DORV                           | 7               | 5              |
| PS                             | 8               | 4              |
| Pulmonary atresia              | 4               | —              |

IVC; inferior vena cava, SVC; superior vena cava,LSVC; left superior vena cava, RSVC; right superior vena cava, BSVC; bilateral SVC, HV ; hepatic vein TAPVR; total anomalous pulmonary venous return PAPVR; partial anomalous pulmonary venous return
cases were classified as left isomerism on the basis of atrial appendage morphology. The distribution pattern of pectinate muscle is quite helpful for the classification of isomerism, but MR is not able to resolve this fine structure. The presence of terminal crest or the broadness of the junction with the venous atrium were not helpful in this study, because in MR images, they could be influenced by atrial dilatation.

The spleen is absent in 82% of patients with right isomerism(5), but clinical recognition of the absence of multiplicity of the spleen is difficult. The absence can be recognized by the presence of Howell-Jolly bodies in the blood(28) and the failure of scintigraphy(19) or ultrasonography(13, 23) to demonstrate the spleen. Howell-Jolly bodies may, however, appear in patients with normal spleen, are seen rarely in those with polysplenia, or may increase where there is sensitivity to drugs(20). Spleen scintigraphy is difficult to interpret because of radionuclide uptake by the liver. Multiple spleen was seen in 70% of patients with left isomerism, and in less than 1% of those with lateralized atria (24). Ultrasonography can depict multiple spleen but this is unfamiliar to cardiologists, and if located at an unusual site, its acoustic window may be limited by a gas-filled bowel. Abdominal arteriography, CT and MRI are the only reliable diagnostic method, and MRI, in particular, is an excellent modality for demonstrating the presence, shape and position of the spleen.

Right-left bronchial length ratio was less than 1.5 in cases of isomerism(11, 16, 17), and to determine whether this was right or left, bronchial length was compared to that of patients of corresponding age (16), or to tracheal width(17). Bronchial length ratio and the relative positions of a bronchus and its respective PA can be assessed from a good quality frontal image obtained using the high KV technique. If the PA is enlarged, its position can be recognized by bronchial compression, and lateral projection may be useful for evaluation of relative positions of the bronchus and its corresponding pulmonary artery(11). In cases of isomerism, the right and left PA are located either anterior or posterior to the tracheobronchial tree, as seen on lateral view, a bronchus with a posterior stripe due to interface with adjacent lung parenchyma(lateral view) is the right main stem. A great advantage of angiography, CT, and MRI is that even if PAs are hypoplastic, these modalities offer clear visualization of both these and the bronchi, and their positional relationship. In this study, all patients showed either bilaterally hyaeterial or epiarterial bronchi, without any interobserver variation, and in MR imaging, the pulmonary arterial and bronchial relationship is therefore the most constant and reliable indicator of atrial situs. Moreover, identification of bronchial situs, especially the superior and inferior relation of the pulmonary arteries and bronchi, is thought to be sufficient to determine cardiac situs.

Lung situs is the most difficult to recognize clinically; a minor fissure is visible in only half the normal population(29). CT or MRI can identify either the middle lobe or lingular bronchial system, providing further confirmation of main bronchial typing; a superiorly oriented p-wave axis was noticed in 80% of patients with left isomerism.

It is not the point of this study to decide whether the suprahepatic segment of the IVC is the most important landmark in visceral heterotaxy syndrome for determining atrial situs. Our study showed, however, that this segment was not constantly recognized, and on the basis of MRI, the seven cases in which it was visualized were classified by atrial morphology and asplenic shadow as either left isomerism and polysplenia(n=2) or right isomerism and asplenia(n=5).

Both CT and MRI are excellent for the demonstration of major extracardiac arteries and veins, and of abdominal viscera, but the latter best determines not only cardiac situs but also associated complicated cardiac anomaly.

The incidence of systemic and pulmonary venous return and other associated cardiovascular anomalies in this study were not different from the incidence seen in previous investigations(5, 10, 25).

In conclusion, the superior-inferior relation of pulmonary arteries and main bronchi showed no interobserver variation, and lateralization was the predominating tendency. The author thus considered this situs to be the most reliable method of situs determination by MR imaging. The superior-inferior relation of pulmonary arteries and bronchi was compared with situs on the basis of main bronchial length ratio, atrial appendage or splenic abnormality; in the assessment of atrial situs by MRI, bronchial length ratio and splenic abnormality are relatively reliable. The accuracy of classification of situs on the basis of atrial appendage morphology is, however, less reliable, particularly in patients with left isomerism. MRI is therefore useful for the assessment of atrial situs according to bronchial situs, as well as, for assessment of associated cardiovascular abnormality.
References

2. Van Mierop LHS, Wiglesworth FW. Isomerism of the cardiac atria in the asplenic syndrome. Lab Invest 1962; 11:1303-1315
8. Caruso G and Becker AE. How to determine atrial situs? Consideration initiated by 3 cases of absent spleen with a discordant anatomy between bronchia and atria. Br Heart J 1979; 41:559-567
중복위를 가진 선천성 심질환의 심방 위치 결정에 있어서
MR영상의 역할

목적: 중복위를 가진 복합심장기형에서 심방 위치(situs)를 결정하는데 있어 영상의 역할을 평가하고자 하였다.

방법: 22명의 중복위를 가진 환자를 대상으로 MR을 통하여 심방 부속기의 모양, 양측 주기관지 길이의 비, 기관지-폐혈관의 위치 관계, 비장 이상을 통하여 심방 위치를 분류하여, 각 방법에 의한 결과를 비교하였다.

결과: 주기관지와 폐동맥과의 상하 위치는 13명에서 우이성체, 9명에서 좌이성체의 모양을 보였다. 주기관지-폐동맥 위치 관계에 따른 심장 위치를 기준으로 할 때 양측 주기관지 길이의 비는 1명에서 일치하지 않았고, 심방 부속기의 모양은 13명의 우이성체 중 10명, 9명의 좌이성체 중 3명에서만 일치하였다. 모든 우이성체는 비장이 없었으며, 9명의 좌이성체 중 8명에서 다비장증(polysplenia)이 있었다.

결론: MR영상으로 중복위 환자에서 심장 위치를 결정하는데 있어 주기관지-폐동맥의 위치 관계, 양측 주기관지 길이의 비, 비장 이상 등은 일치성이 높아 임상적으로 유용하지만 심방 부속기의 모양을 이용한 심장 위치 결정은 한계성이 있다.
‘98년도 제41회 의사전문의 자격시험 안내

1. 응시원서 교부 및 접수
가) 응시원서 교부
① 교부기간: 1997년 11월 3일(월)~11월 8일(토)
② 교부장소: 대한의사협회 (서울 용산구 이촌1동 302-75) ☎ (02) 794-2482
나) 응시원서 접수
① 접수기간: 1997년 11월 7일(금)~14일(금)
② 접수장소: 본 학회 사무국(서울시 서초구 양재동 121-8 ☎ (02) 578-8003)
다) 수험표 교부
① 교부기간: 1997년 12월 26일(금)~27(토)
② 교부장소: 본 학회 사무국

2. 구비서류(응시원서 접수시 아래 구비서류를 일괄 제출하여야 합니다)
가) 응시원서 및 수험표 (의협소정양식) .................................................. 각 1부
나) 사진 (3개월 이내에 촬영한 동일원판의 탈모정면 상반신
반명함판 3.5cm x 4cm, 제출서류 부착수량 제외) ............................................. 2매
라) 응시료 (원서 교부시 의협에 납부) ............................................................. 90,000원
⑤ 수험료 (원서 접수시 학회에 납부) ............................................................. 200,000원
⑥ 전문의제도개선사업비( ) ................................................................. 10,000원
⑦ 업회비 (원서 접수시 학회에 납부) ........................................................ 100,000원
⑧ 의협회비 (원서 접수시 미납자에 한하여 학회에 납부)
마) 합격자 명부 (의협 소정양식) ..................................................................... 2부
바) 수련과정 이수 또는 예정증명서 (의협 소정양식) ........................................... 2부
(인턴과 레지던트 수련병원이 다른 경우 각각 분리 작성하여 각 2부씩)
사) 해외수련자인 경우 수련과정 이수증명서 사본
(주재국 우리나라 공관장의 확인을 필할 것) .................................................. 2부
아) 외국의 전문의 자격증을 취득한 경우에 서는 그 자격증 사본
(주재국 우리나라 공관장의 확인을 필할 것) .................................................. 2부
자) 의사면허증 사본 (규격 A4 용지) ................................................................. 2부
차) 의과수련확인서 (96년도 수련병원 설대조사를 근거로 의과수련과정을 통보하였음)
카) 전공의 기록부 ......................................................................................... 1부
타) 논문별책 (원저 제1저자 1부, 공저 2부) .................................................. 3부

3. 시험일정
가) 1차 시험
① 일시: 1998년 1월 8일(목) 10:00~13:00
② 장소: 서울대학교 소아병원 제1, 2강당
③ 발표: 1998년 1월 14일(수) 15:00~대한의사협회
나) 2차 시험
① 일시: 1998년 1월 15일(목)~16(금)
② 장소: 서울대학교 소아병원 제1, 2강당 (방안 및 컴퓨터사인펜 지참)
서울대학교 소아병원 1, 2강의실 (방안 및 컴퓨터사인펜 지참)
구술시험 - 1월 16일(금) 08:00~20:00 추후 발표
③ 발표: 1998년 2월 5일(목) 15:00~대한의사협회