Role of Spin Echo and Cine Magnetic Resonance Imaging in Presurgical Planning of Heterotaxy Syndrome

Comparison With Echocardiography and Catheterization

Tal Geva, MD; G. Wesley Vick III, MD, PhD; Richard E. Wendt, PhD; Roxann Rokey, MD

Background Patients with heterotaxy syndrome frequently have complex congenital cardiac and noncardiac malformations requiring detailed diagnostic evaluation by noninvasive as well as invasive imaging modalities for management planning. Recent advances in magnetic resonance imaging (MRI) techniques allow detailed delineation of cardiovascular anatomy and blood flow in young infants with rapid heart rates. The present study was undertaken to prospectively evaluate the role of MRI in the presurgical evaluation of patients with heterotaxy syndrome.

Methods and Results Between January 1 and December 31, 1992, 14 consecutive patients with heterotaxy syndrome and complex congenital heart disease were enrolled in a prospective protocol. After evaluation by echocardiography and cardiac catheterization, a tentative management plan was recorded. Subsequently, a MRI study was performed and surgical planning was reevaluated. MRI was found to be comparable to echocardiography in terms of length of examination and sedation requirements. Surgical planning was altered in four patients because MRI provided additional data not evident on echocardiography and catheterization. Comparison of diagnostic yield between echocardiography, catheterization, and MRI showed that MRI is superior to echocardiography and often to catheterization in delineation of systemic and pulmonary venous anatomy and their relation to mediastinal structures. When the anatomic and hemodynamic data obtained by echocardiography and MRI were considered together, cardiac catheterization data were necessary only to determination of pulmonary vascular resistance before Fontan operation.

Conclusions MRI provides excellent anatomic and functional information that in some patients was not available by echocardiography or catheterization. Combined with echocardiography, MRI provides the high-quality diagnostic information necessary for management planning in most patients with heterotaxy syndrome. Cardiac catheterization is indicated when determination of pulmonary vascular resistance is necessary for decision making or when an interventional procedure is indicated. (Circulation. 1994;90:348-356.)

Key Words • heterotaxy syndrome • imaging • congenital heart disease • echocardiography • catheterization

Visceral heterotaxy, a syndrome characterized by incomplete or inappropriate lateralization of the abdominal and thoracic organs, is frequently associated with complex congenital heart disease.1-3 Affecting approximately 0.8% of patients with congenital heart disease,4 patients with heterotaxy syndrome in the past were of interest mostly to pathologists. In recent years, however, improved results of palliative and corrective surgical techniques5-7 as well as improved survival of infants and young children with complex congenital heart disease who underwent orthotopic heart transplantation8,9 have renewed interest in surgical therapy in patients with heterotaxy syndrome. Planning surgical intervention in these patients requires accurate and detailed delineation of complex cardiovascular anatomy and hemodynamics. Traditionally, echocardiography4 and cineangiography10 have been the imaging modalities most widely used in the preoperative evaluation of patients with heterotaxy syndrome. Although in the majority of young patients echocardiography allows detailed anatomic and hemodynamic evaluations, this technique is highly dependent on operator skills and expertise with complex congenital heart disease. Furthermore, imaging of certain mediastinal and thoracic structures by ultrasonography may be hampered by limited windows and/or presence of adjacent lung tissue. Simultaneous imaging and delineation of anatomic relations between cardiac, vascular, and tracheobronchial structures is not routinely obtained by echocardiography. Cardiac catheterization is limited by its invasive nature, small but not negligible risks of morbidity and mortality, and ability to delineate intracardiac and vascular structures only but not adjacent organs.

In recent years, magnetic resonance imaging (MRI) has been effectively used in pediatric cardiac imaging. MRI allows depiction of intracardiac as well as mediastinal and thoracic anatomy even in young patients with rapid heart rates.11-13 Development of the technique of cine MRI has facilitated delineation and characteriza-
tion of blood flow,\textsuperscript{14} enhancing the potential usefulness of MRI in patients with congenital heart disease. Cardiac MRI became a clinical diagnostic tool in our institution in 1986. It has been our impression that in patients with heterotaxy syndrome, MRI provided additional clinically useful information that was not readily available by other diagnostic modalities. The present study was undertaken to prospectively investigate the potential role of combined spin echo and cine MRI in the preoperative evaluation of patients with heterotaxy syndrome and to compare its potential benefits with those of transthoracic echocardiography and cardiac catheterization.

Methods

Patients

Between January 1 and December 31, 1992, 14 consecutive patients who presented to Texas Children's Hospital for evaluation of congenital heart disease and were subsequently diagnosed with heterotaxy syndrome were prospectively enrolled in the present study. Each patient underwent a detailed clinical evaluation, 15-lead ECG, chest radiogram, complete two-dimensional and Doppler echocardiographic examination, and cardiac catheterization and cineangiography. Based on the results of these tests, the diagnosis of heterotaxy syndrome was established according to the criteria set forth by Van Mierop et al.\textsuperscript{1,2} Macartney et al.\textsuperscript{3} and Van Praagh et al.\textsuperscript{4} including (1) abnormal symmetry of the abdominal visceral organs (eg, midline liver, midline or right-sided stomach), (2) abnormal symmetry of the bronchi, (3) situs discordance between various organ systems and between the various segments of the heart, (4) presence of typical constellation of congenital cardiac malformations such as the coexistence of systemic and/or pulmonary venous anomalies, malformations of the atroventricular canal and conotruncal malformations, and (5) evidence of splenic abnormality (eg, asplenia, polysplenia, or a single right-sided spleen). The status of the spleen was established by imaging (ultrasound and/or radionuclide liver-spleen scan) and the results of a peripheral blood smear for Howell-Jolly bodies. Using published criteria,\textsuperscript{4,5} attempts were made to diagnose specifically the situs of the abdominal viscera, main stem bronchi, and major cardiac segments.

For each patient, the anatomic and hemodynamic diagnoses as well as a tentative management plan were recorded by the attending cardiologist and the study coordinator. Subsequently, the patients underwent an MRI study, and the anatomic and hemodynamic diagnoses were recorded and presented to the attending cardiologist. The management plan was then reevaluated, and revisions were recorded. When applicable, surgical procedures and operative and/or pathologic findings were documented. In each patient, the echocardiographic, angiographic, and MRI studies were performed within 1 month of each other. The time required to perform each examination was recorded.

Imaging Examinations

MRI

MRI examinations were performed on a 1.0- or 1.5-T imaging magnet (Siemens Magnetom, Siemens Medical Systems). Spin echo acquisitions were electrocardiographically synchronized\textsuperscript{16} with a repetition time (TR) equal to the RR cycle length and an echo delay time (TE) of 15 to 26 milliseconds. Cine MRI studies were also electrocardiographically synchronized and acquired using multiphasic gradient echo images of a specific anatomic locations with a TE of 10 milliseconds and a TR of 50 milliseconds. The number of cardiac cycle phases obtained during a cine gradient echo acquisition was limited by the RR interval. Velocity compensa-

sation of the gradient echo sequences was accomplished with a gradient moment nulling technique. Image matrix size was 128×128 or 256×256 pixels, and slice thickness ranged from 3 to 8 mm for both studies. Cine and spin echo images were obtained in identical slice position to allow comparison between anatomic images (spin echo) and flow-sensitive images. The MRIs were acquired in transverse, coronal, or sagittal planes and in oblique planes parallel or perpendicular to the axes of cardiovascular structures. Infants with body weight <15 kg were studied in a head coil. Imaging time required for these evaluations varied from 45 to 75 minutes.

Images were subsequently digitally transferred from the MRI scanner to a Macintosh Quadra microcomputer (Apple Computer). Images were archived to an optical disk and processed with customized software (NIH IMAGE, Wayne Rasband, National Institutes of Health).

Young, uncooperative patients were sedated with oral chloralhydrate (100 mg/kg; maximal dose, ≤2 g). Parents were instructed to keep their child awake and to avoid feeding them for 5 to 6 hours before the study. Sedation was successful in all patients that it allowed completion of the study without need for repeat imaging. Sedated patients were monitored by pulse oximetry, ECG, and closed-circuit television. Older patients were prepared for their study by the cardiologist performing the examination. We found that allowing parents to stay with the child in the examination room greatly facilitated the child's cooperation and successful completion of the test.

Echocardiography

A complete two-dimensional and Doppler examination was performed from the subxiphoid, apical, parasternal, and suprasternal approaches. Tomographic views were performed in multiple planes with special attention given to providing the best possible demonstration of the anatomic features of heterotaxy syndrome. Evaluations were performed with 7.5-, 5.0-, or 3.5-MHz phased-array (Acuson Corp) or annular-array (Vingmed CFM 750) transducers. All echocardiographic examinations were supervised by a staff echocardiographer. Studies were recorded on ½-in super-VHS videocassette tapes. Sedation with oral chloralhydrate (75 to 100 mg/kg; maximal dose, 1 g) was used when necessary.

Cardiac Catheterization and Cineangiography

A comprehensive cardiac catheterization was performed in each patient as described by Nihill.\textsuperscript{17} The angiographic examination was specifically directed toward evaluation of the anatomic features of heterotaxy syndrome. Aortography, ventriculography, pulmonary arterial, and/or pulmonary vein wedge angiography were performed in biplane anteroposterior, lateral, and axial views depending on the lesions to be evaluated. In the majority of patients, the pulmonary-to-systemic flow and vascular resistance ratios as well as pulmonary arteriolar resistance were determined.

Data Analysis

All echocardiographic, cineangiographic, spin echo, and cine MRI studies were independently assessed by two observers (T.G. and G.W.V.) to determine if structures of interest could be identified. The observers knew the patients' names but were unaware of the other observer's diagnoses. For each patient, 13 anatomic variables were analyzed and recorded (total of 182 variables examined). The anatomic variables analyzed were abdominal situs, cardiac segments,\textsuperscript{18} ventriculoarterial alignment, superior vena cava, inferior vena cava, coronary sinus, pulmonary veins, atrial septum, atrioventricular canal, ventricles, conus, pulmonary arteries, aortic arch, and arch vessels. Interobserver agreement was expressed as the ratio between concordant diagnoses reached by the two observers and the total number of anatomic variables exam-
TABLE 1. Anatomic Data for 14 Patients With Heterotaxy Syndrome Based on Magnetic Resonance Imaging Findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age, mo</th>
<th>Sex</th>
<th>Spleen</th>
<th>Bronch</th>
<th>Abd. Situs</th>
<th>Cardiac Segments*</th>
<th>VA Alignment</th>
<th>SVC</th>
<th>IVC</th>
<th>Pulmonary Veins</th>
<th>Atrial Septum</th>
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<tbody>
<tr>
<td>1</td>
<td>24</td>
<td>F</td>
<td>A</td>
<td>S</td>
<td>S</td>
<td>[S, D, A]</td>
<td>DORV</td>
<td>Bil.</td>
<td>Nil</td>
<td>TAPVR</td>
<td>CA*</td>
</tr>
<tr>
<td>2</td>
<td>22</td>
<td>M</td>
<td>A</td>
<td>S</td>
<td>S</td>
<td>[S, D, D]</td>
<td>TGA</td>
<td>Bil.</td>
<td>Nil</td>
<td>Nil</td>
<td>CA</td>
</tr>
<tr>
<td>3</td>
<td>7</td>
<td>F</td>
<td>P</td>
<td>S</td>
<td>S</td>
<td>[S, D, S]</td>
<td>Conc.</td>
<td>Bil.</td>
<td>Int-Az</td>
<td>Nil</td>
<td>CA</td>
</tr>
<tr>
<td>4</td>
<td>8</td>
<td>M</td>
<td>Bil. Rt</td>
<td>I</td>
<td>[S, D, D]</td>
<td>DORV</td>
<td>Nil</td>
<td>Nil</td>
<td>TAPVR</td>
<td>CA</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>3</td>
<td>M</td>
<td>Bil. Lt</td>
<td>I</td>
<td>[S, D, I]</td>
<td>TGA</td>
<td>Bil.</td>
<td>Int-Az</td>
<td>PAPVR</td>
<td>CA*</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>5</td>
<td>M</td>
<td>RS</td>
<td>S</td>
<td>[S, D, S]</td>
<td>DORV</td>
<td>Nil</td>
<td>Nil</td>
<td>2* ASD</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>5</td>
<td>M</td>
<td>A</td>
<td>I</td>
<td>I</td>
<td>[I, L, L]</td>
<td>DORV</td>
<td>LSVC</td>
<td>Nil</td>
<td>TAPVR</td>
<td>CA</td>
</tr>
<tr>
<td>8</td>
<td>120</td>
<td>F</td>
<td>M</td>
<td>Bil. Lt</td>
<td>S</td>
<td>[A, L, L]</td>
<td>DORV</td>
<td>RSVC</td>
<td>Int-Az</td>
<td>PAPVR</td>
<td>CA*</td>
</tr>
<tr>
<td>9</td>
<td>14</td>
<td>M</td>
<td>P</td>
<td>I</td>
<td>I</td>
<td>[I, D, D]</td>
<td>DORV</td>
<td>Bil.</td>
<td>Int-Az</td>
<td>Nil</td>
<td>CA</td>
</tr>
<tr>
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<td>8</td>
<td>M</td>
<td>Bil. Rt</td>
<td>S</td>
<td>[S, D, D]</td>
<td>DORV</td>
<td>Nil</td>
<td>Nil</td>
<td>TAPVR</td>
<td>CA</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>3</td>
<td>F</td>
<td>A</td>
<td>I</td>
<td>[I, D, D]</td>
<td>DORV</td>
<td>LSVC</td>
<td>Nil</td>
<td>PAPVR</td>
<td>CA</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>18</td>
<td>M</td>
<td>P</td>
<td>S</td>
<td>[S, D, S]</td>
<td>DORV</td>
<td>Conc.</td>
<td>Int-Az</td>
<td>PAPVR</td>
<td>Nil</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>41</td>
<td>F</td>
<td>P</td>
<td>S</td>
<td>[S, D, S]</td>
<td>DORV</td>
<td>TGA</td>
<td>Bil.</td>
<td>Int-Az</td>
<td>PAPVR</td>
<td>Nil</td>
</tr>
<tr>
<td>14</td>
<td>9</td>
<td>F</td>
<td>A</td>
<td>Bil. Rt</td>
<td>S</td>
<td>[S, L, S]</td>
<td>DORV</td>
<td>TGA</td>
<td>Nil</td>
<td>PAPVR</td>
<td>CA*</td>
</tr>
</tbody>
</table>

Abd. indicates abdominal; VA, ventriculoarterial; SVC, superior vena cava; IVC, inferior vena cava; AV, atrioventricular; F, female; M, male; A, asplenia; P, polysplenia; RS, right-sided single spleen; S, solitus; Bil., bilateral; Rt, right; Lt, left; I, inversus; DORV, double-outlet right ventricle; TGA, transposition of the great arteries; Conc., concordance; Nil., normal; LSVC, left superior vena cava; R SVC, right superior vena cava; Int-Az, interrupted inferior vena cava with azygous continuation; TAPVR, totally anomalous pulmonary avenous return; PAPVR, partially anomalous pulmonary venous return; CA*, common atrium with malattachment of septum primum; CA, common atrium; CAVC, common atrioventricular canal defect; R, right unbalanced; ICAVC, incomplete atrioventricular canal defect; MV atr., mitral valve atresia; MS, mitral stenosis; Hypop, hypoplastic; LV, left ventricle; RV, right ventricle; P atr., pulmonary atresia; PS, pulmonary outflow tract stenosis.

Comparison of interobserver agreement between echocardiography, catheterization, and MRI was performed using $\chi^2$ analysis.

To compare the diagnostic accuracy of echocardiography, catheterization, and MRI, the prospectively recorded diagnoses (13 anatomic variables per patient) were compared with the final diagnoses, which were based on a retrospective review of all imaging and diagnostic tests (echocardiography, catheterization, and MRI) as well as surgical and autopsy records. Multiple contingency tables were constructed, and $\chi^2$ analysis was used to determine statistical significance. Student's $t$ test was used to compare the mean values of continuous variables. A value of $P \leq 0.05$ was considered significant.

Results

Fourteen patients ranging in age from 3 months to 10 years (median, 8 months) enrolled in this study. There were 9 boys and 5 girls. Patient weights ranged from 3.9 to 18.6 kg (median, 6.5 kg). Asplenia was diagnosed in 7 patients, polysplenia in 6, and a single right-sided spleen in 1 patient.

Anatomic Data

The anatomic findings based on MRI are summarized in Table 1. Although a large liver in a relatively midline position was present in 12 of 14 patients, based on the position of the stomach and liver lobation, MRI allowed determination of the predominant situs of the abdominal viscera in all patients (Table 1, Fig 1). Heart position was equally divided between levocardia and dextrocardia (7 patients each). The anatomy of the main stem bronchi and their relations to the branch pulmonary arteries were clearly demonstrated by MRI (Fig 2). Bilaterally symmetric bronchi were seen in only 5 of 14 patients. Among patients with asplenia, bilaterally right bronchial morphology was seen in 2 of 7 patients (Fig 2B). Bilaterally left bronchial morphology was demonstrated in 2 of 6 patients with polysplenia. The remaining patients had either bronchial situs solitus ($n$=6) (Fig 2C) or inversus ($n$=3). The abdominal and bronchial situs were discordant in all patients in whom the bronchi were asymmetric. Atrial situs, however, was discordant relative to abdominal situs in 2 patients (Table 1). Strictly defined, atrial isomerism was not observed in this series, and atrial situs could be determined in all except 1 patient (Table 1). Discordance between at least two of the three main segments of the heart (atria, ventricles, and great arteries) was found in 12 of 14 patients.

Systemic venous anomalies were found in 13 of 14 patients. Of those, 10 patients were found to have bilateral superior vena cavae. In 6 patients, the inferior vena cava was interrupted at the hepatic segment and the supracardinal segment was continuous with either the azygous or hemiazygous vein (Fig 3). Polysplenia was present in all patients with an interrupted inferior vena cava. Anomalous drainage of the pulmonary veins was diagnosed in 9 of 14 patients. Totally anomalous pulmonary venous return was present in 4 patients, subdiaphragmatic in 1 patient and supracardiac to the innominate vein in 3 patients. Partial anomalous pulmonary venous drainage with malattachment of septum primum was demonstrated in 5 patients (Fig 4). A common atrioventricular valve was found in 11 patients with either a partial or complete common atrioventricular canal defect. In 9 patients, the atrioventricular canal defect was markedly unbalanced, associated with either
hypoplasia (n=8) or absence (n=1) of the left ventricle. Asplenia was present in 6 of those 9 patients (Table 1).

Anomalies of the conotruncus were found in 12 of 14 patients—most often, a bilateral conus with either pulmonary outflow tract obstruction (n=7) or atresia (n=4). One patient had a hypoplastic subpulmonary conus with infundibular and valvar stenosis and an aortic valve–mitral valve fibrous contiguity (eg, tetralogy of Fallot). Pulmonary arterial hypoplasia with or without discrete stenosis was noted in 11 patients (Table 1). Interestingly, aortic arch sidedness was equally split between a right and a left aortic arch.

**Effect of MRI on Surgical Planning**

Surgical planning was altered in 4 patients after review of the spin echo and cine MRI studies. In 3 of those patients, MRI provided essential surgically relevant information regarding complex anomalous pulmonary venous return. In 1 patient with asplenia, visceroatrial situs inversus, a common atrium, an unroofed coronary sinus, and supracardiac totally anomalous pulmonary venous connection to the left innominate vein, the exact anatomy of the pulmonary venous confluence and its relation to neighboring mediastinal structures were determined only after an MRI study. In 2 additional patients, MRI reversed the previous diagnosis of totally anomalous pulmonary venous connection to the right atrium. Spin echo and cine MRI sequences accurately delineated the connections of each individual pulmonary vein to the back wall of the atria and the relations between the orifices of the pulmonary veins and the markedly malaligned and deficient septum primum. In the fourth patient who had asplenia, segmental combination [S, L, L], complete common atroventricular canal, transposed aorta, infundibular and valvar pulmonary atresia, and discontinuous pulmonary arteries, the left pulmonary artery could not be identified with certainty by echocardiography and cardiac catheterization. This patient became increasingly cyanotic despite a patent modified right Blalock-Taussig shunt. Spin echo and cine MRI sequences documented a 5-mm distal left pulmonary artery, and the patient subsequently underwent placement of a modified left Blalock-Taussig shunt with significant improvement in arterial oxygen saturation.
Comparison of Imaging Modalities

Table 2 summarizes the comparison among echocardiography, cardiac catheterization and angiography, and MRI. Cardiac catheterization is superior to the two noninvasive diagnostic modalities only when measurements of pulmonary blood flow and pulmonary vascular resistance are clinically important for management decision making. Echocardiography and MRI were comparable in terms of being noninvasive, requiring relatively light oral sedation in young uncooperative patients, and being associated with no appreciable complications. The average length of examination (including sedation time) also was comparable for this group of young patients with highly complex anatomy, being about 1 hour (Table 2). There were no major complications associated with cardiac catheterization in this study. Minor complications included transient femoral artery occlusion in 2 patients, which resolved after intravenous thrombolytic therapy with return of distal pulses and good perfusion. Another patient had bilateral femoral venous occlusion. This complication later proved to be important because the patient underwent orthotopic heart transplantation and had significant problems with vascular access necessary for endomyocardial biopsies.

Comparison of diagnostic accuracy between the imaging modalities showed that MRI was superior to echocardiography and cineangiography in delineation of pulmonary venous anatomy. Complete accurate delineation of all pulmonary veins and their exact connections was achieved in all patients by MRI, but not achieved in 5 of 12 patients (42%) by echocardiography (P = .007, echo versus MRI) and in 3 of 12 patients (25%) by catheterization (P = .047, catheterization versus MRI). The difference in diagnostic accuracy of pulmonary venous anatomy between echocardiography and catheterization was insignificant (P = .38). Simultaneous imaging of both vascular and nonvascular mediastinal structures was possible only by MRI, as was imaging of the trachea, bronchi, and the spatial relations between mediastinal structures. MRI also provided information regarding the anatomy of the abdominal organs, including splenic status and location of the liver and stomach. Delineation of the abdominal aorta, inferior vena cava, and azygous vein was equally accurate for the three imaging modalities (P = .11).

The diagnostic agreement between the two observers was 96.1% (175 of 182 diagnoses) for MRI, 86.3% (145 of 168 diagnoses) for echocardiography, and 88.1% (148 of 168 diagnoses) for catheterization. The differences between MRI and echocardiography and between MRI
in these patients because we were particularly interested in the effect of this diagnostic modality on surgical planning. Based on our experience with 14 consecutive patients, MRI provided surgically relevant information that affected operative planning in four patients (29%).

Role of MRI in the Initial Evaluation of Heterotaxy Syndrome

The presence of cyanosis, a heart murmur, or other abnormal findings on physical examination together with situs abnormalities on the chest and abdominal radiogram frequently provides the first clue to the presence of heterotaxy syndrome. An echocardiogram is the preferred initial diagnostic modality (Fig 6) because it is readily available to cardiologists, and a systematic examination by experts will delineate the cardiac anatomy and hemodynamics.4 Other advantages of echocardiography include real-time imaging, portability, and cost, and it is ideal for follow-up. However, echocardiography alone did not provide all relevant information necessary for surgical planning in all patients with heterotaxy syndrome. Additional diagnostic information was necessary mostly in patients with complex systemic and pulmonary venous anomalies. We found that the accurate relations of the venous structures to the atria and neighboring mediastinal structures were often difficult to delineate by echocardiography alone, particularly when a complete surgical repair or an orthotopic heart transplantation was planned.

Based on this study, we found that MRI is complementary to echocardiography in the anatomic and hemodynamic evaluation of these patients. MRI provided good visualization of both intracardiac and extracardiac structures. The major advantage of MRI is in the delineation of systemic and pulmonary venous structures as well as arterial structures and their relations to the heart and neighboring mediastinal structures. Our results concur with those of Niwa et al.19 However, the diagnostic rate of pulmonary stenosis or atresia was low in their study. We found that cine MRI provided

Discussion

The results of this study show that spin echo and cine MRI have an important role in the diagnostic evaluation of patients with heterotaxy syndrome. We chose a prospective approach to evaluate the usefulness of MRI and catheterization were statistically significant ($P = .001$ and $P = .005$, respectively), whereas the difference between echocardiography and catheterization was not ($P = .6$).

![Fig 3. Parasagittal magnetic resonance image in a patient with polysplenia and interruption of the inferior vena cava between the renal and hepatic segment. The inferior vena cava is continuous with the azygous vein (Az. V.) which drains into the right superior vena cava (SVC). HV indicates hepatic vein; RA, right atrium.](image)

![Fig 4. Magnetic resonance image in the transverse plane in a patient with polysplenia. The atrial septum (arrow) attaches to the left of the right pulmonary vein orifices (right lower pulmonary vein [RLPV] is seen), resulting in partial anomalous pulmonary venous drainage of the right pulmonary veins to the right atrium (RA). The relations between the pulmonary veins and the atrial septum are clearly defined. LA indicates left atrium; LLPV, left lower pulmonary vein.](image)
TABLE 2. Comparison of Imaging Modalities Based on Experience With 14 Consecutive Patients With Heterotaxy Syndrome

<table>
<thead>
<tr>
<th></th>
<th>Echocardiography</th>
<th>Catheterization</th>
<th>Magnetic Resonance Imaging</th>
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</thead>
<tbody>
<tr>
<td>Sedation</td>
<td>Only young uncooperative patients (oral chloralhydrate)</td>
<td>Yes</td>
<td>Only young uncooperative patients (oral chloralhydrate)</td>
</tr>
<tr>
<td>Average length, min</td>
<td>60 (range, 45 to 75)</td>
<td>150 (range, 120 to 210)</td>
<td>60 (range, 45 to 65)</td>
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<tr>
<td>Complications</td>
<td>None</td>
<td>Minor*</td>
<td>None</td>
</tr>
<tr>
<td>Real-time imaging</td>
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<td>No</td>
<td>No</td>
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<tr>
<td>Portability</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Imaging of intracardiac structures</td>
<td>Excellent</td>
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</tr>
<tr>
<td>Imaging of extracardiac anatomy</td>
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<tr>
<td>Systemic veins</td>
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<td>Excellent</td>
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<tr>
<td>Pulmonary veins</td>
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<td>Pulmonary arteries</td>
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<td>Excellent</td>
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<tr>
<td>Aortopulmonary collaterals</td>
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<td>Excellent</td>
<td>Excellent</td>
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<tr>
<td>Imaging of nonvascular thoracic structures</td>
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<td>Poor</td>
<td>Excellent</td>
</tr>
<tr>
<td>Ability to obtain pulmonary vascular resistance and flow</td>
<td>Poor</td>
<td>Excellent (only if direct access possible)</td>
<td>Poor</td>
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*See text for details.

important hemodynamic information regarding the presence and degree of semilunar and atrioventricular valve regurgitation and obstruction. Cine MRI clearly identified patients with pulmonary outflow stenosis and atresia. Cine MRI also proved crucial in differentiating vascular from nonvascular structures. Newer cine MRI techniques (not used in this study) can also measure flow velocity across areas of obstruction and estimate pressure gradients as well as shunts.20 MRI also provided anatomic information that is difficult to obtained by echocardiography or cineangiography. This included delineation of abdominal anatomy (including splenic status), the anatomy of the trachea and main stem bronchi, and their relations to the pulmonary arteries (Fig 2) and simultaneous imaging of both atrial appendages (Fig 5). These data allowed a more complete anatomic assessment of these patients with complex cardiovascular lesions (Fig 6).

Together with echocardiography, MRI provided all necessary anatomic data to allow surgical planning in our patients. MRI is comparable to echocardiography in terms of need for sedation and length of study (Table 2). In many institutions, however, MRI is not readily accessible to pediatric cardiologists, and its use for repeat follow-up studies is limited. We found that involvement of a pediatric cardiologist experienced in imaging of complex congenital heart disease is essential to the success of the examination.
Role of Cardiac Catheterization

Based on the present study, delineation of cardiac anatomy by cineangiography in patients with heterotaxy syndrome is seldom necessary as part of the initial evaluation (Fig 6). Our data suggest that cardiac catheterization proved important mostly in two situations: first, to determine pulmonary vascular resistance before a modified Fontan operation, a bidirectional cavopulmonary anastomosis (bidirectional Glenn), or orthotopic heart transplantation when the possibility of pulmonary vascular obstructive disease is present; and, second, when an interventional procedure such as balloon dilation or stent placement are indicated.

Cineangiography is currently the technique of choice to delineate tertiary and quaternary intraparenchymal pulmonary arterial branches and distal coronary arterial anatomy. However, information about distal pulmonary artery and coronary artery anatomy did not influence the management of the patients in this series. In selected patients, such as those with pulmonary atresia and intact ventricular septum, information about coronary arterial anatomy obtained by cardiac catheterization and coronary angiography is important for clinical decision making. In the majority of infants with heterotaxy syndrome, coronary artery branching abnormalities can be delineated by echocardiography. Based on this series, we believe that in view of its invasive nature, small but definitive risk of morbidity, and cost, routine cardiac catheterization is not indicated if the anatomic and hemodynamic data necessary to manage the patient can be obtained noninvasively by echocardiography and MRI.

Study Limitations

The study was conducted prospectively, and each diagnostic test was reviewed independently by two investigators, but the patient's names were not hidden from the observers. This might have introduced a potential bias toward unity of diagnoses by each observer. We do not believe that this limitation influenced the overall results of this study.

Conclusions

Spin echo and cine MRI have an important role in the preoperative diagnostic evaluation of patients with heterotaxy syndrome and complex congenital heart disease. MRI provided excellent anatomic and functional information that in some patients was not available by echocardiography or cardiac catheterization. In contrast to previous recommendation, we found that routine cardiac catheterization is not indicated unless determination of pulmonary vascular resistance was necessary or an interventional procedure was indicated. Combined with echocardiography, MRI provided the high-quality diagnostic information necessary for surgical planning in most patients with heterotaxy syndrome.

Acknowledgments

This study was supported in part by Grant-in-Aid 93G-1214 from the American Heart Association, Texas Affiliate, Inc (Dr Geva), and by National Institutes of Health Physician Scientist Award K11-HL-02009 (Dr Rokey).

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Role of spin echo and cine magnetic resonance imaging in presurgical planning of heterotaxy syndrome. Comparison with echocardiography and catheterization.

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Circulation. 1994;90:348-356
doi: 10.1161/01.CIR.90.1.348

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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