Gadolinium-Enhanced 3-Dimensional Magnetic Resonance Angiography of Pulmonary Blood Supply in Patients With Complex Pulmonary Stenosis or Atresia
Comparison With X-Ray Angiography
Tal Geva, MD; Gerald F. Greil, MD; Audrey C. Marshall, MD; Michael Landzberg, MD; Andrew J. Powell, MD

Background—In patients with complex pulmonary stenosis or atresia, a detailed delineation of all sources of pulmonary blood supply is necessary for planning surgical and transcatheter procedures and usually requires diagnostic cardiac catheterization. The goals of this study were to determine whether gadolinium-enhanced 3D magnetic resonance angiography (MRA) can provide a noninvasive alternative to diagnostic catheterization and to compare MRA and x-ray angiography measurements of pulmonary arteries and aortopulmonary collaterals (APCs).

Methods and Results—Thirty-two patients with pulmonary stenosis or atresia (median age: 4.7 years, range: 1 day to 46.9 years) underwent both MRA and cardiac catheterization (median time: 1 month). Diagnoses included tetralogy of Fallot (TOF) with pulmonary atresia (n=13), TOF with pulmonary stenosis (n=4), post-Fontan palliation (n=5), and other complex congenital heart disease (n=10). Compared with catheterization and surgical observations, MRA had a 100% sensitivity and specificity for the diagnosis of main (n=10) and branch pulmonary artery (PA) stenosis or hypoplasia (n=38), as well as absent (n=5) or discontinuous (n=4) branch PAs. All 48 major APCs diagnosed by catheterization were correctly diagnosed by MRA. Three additional APCs were diagnosed by MRA but not by catheterization. The mean difference between MRA and catheterization measurements of 33 pulmonary vessel diameters was 0.5±1.5 mm, with a mean interobserver difference of 0.4±1.5 mm.

Conclusions—Gadolinium-enhanced 3D MRA is a fast and accurate technique for delineation of all sources of pulmonary blood supply in patients with complex pulmonary stenosis and atresia and can be considered a noninvasive alternative to diagnostic catheterization with x-ray angiography. (Circulation. 2002;106:473-478.)

Key Words: heart defects, congenital □ magnetic resonance imaging □ catheterization □ pulmonary heart disease
steno-
sis.16 However, its application to complex PA anomalies such as discontinuous or absent pulmonary arteries and to APCs has not been examined in detail. The present study, therefore, was undertaken to compare MRA with catheterization and x-ray angiography in patients with complex pulmonary stenosis or atresia.

Methods

Patients

All patients referred to the Cardiac MRI Program at Children’s Hospital Boston from March 1, 1998 through November 30, 2001 who fulfilled the following criteria were included in this study: a diagnosis of pulmonary stenosis or atresia; prior Gd-enhanced 3D MRA; and cardiac catheterization with x-ray angiography evaluation of all sources of pulmonary blood supply within 1 year of the MRA without an interim surgical procedure. All MRI studies were clinically indicated. The Children’s Hospital Committee on Clinical Investigations approved review of the medical records and computer databases.

MRI Protocol

MRI studies were performed with a commercially available 1.5 T scanner (either a Signa Horizon HighSpeed with operating systems 5.7/5.8 or Signa Horizon LX EchoSpeed with operating systems 8.1/8.3, GE Medical Systems). A torso or cardiac-phased array radiofrequency coil was used in patients weighing >10 kg, and a head or a surface coil was used in those with a body weight ≤10 kg. General anesthesia was used in patients who were unable to cooperate. The MRA sequence was prescribed from an axial localizing image in either a coronal (n=20) or a sagittal (n=12) orientation and was centered at the level of the mid-thorax. The MRA sequence consisted of a non–ECG-triggered 3D spoiled gradient echo pulse sequence (echo time 1.44±0.26 ms; repetition time 6.4±0.6 ms; flip angle 45°; number of excitations=1 in 28 patients and 0.5 in 4 patients; median field of view 300 mm [range=180 to 420 mm]; matrix 256×128 to 192; median in-plane resolution 1.2×2 mm [range=0.7×0.9 to 1.6×3.3]; median slice thickness 2.2 mm [range=1.2 to 3 mm]; median number of partitions 34 [range=25 to 57]; k-space filling=centric [n=28] or elliptic centric [n=4]; median acquisition time=25 s [range=19 to 44].15,17 Gadopentetate dimeglumine (0.2 mmol/kg; Magnevist, Berlex Laboratories) was injected via a peripheral intravenous cannula either by hand (in patients weighing <10 kg) or by a power injector (Medrad) at rates ranging from 1.5 to 2.5 cc/sec. Patients were instructed to take several deep breaths before image acquisition. The time delay between the start of contrast injection and data acquisition was determined by the “best estimate” method14 and ranged from 6 to 8 seconds in infants to 12 to 14 seconds in adults. Two sequential breath-hold 3D MRA acquisitions were performed 10 to 15 seconds apart. Patients under anesthesia, ventilation was suspended during imaging.

MRA Image Analysis

The MRA studies were reviewed on a commercially available computer workstation (Advantage Windows version 2.0, GE Medical Systems) using a combination of user-defined subvolume maximal intensity projections (MIPs), user-defined multiplanar reformatting, and 3D shaded-surface displays.15–18 The following anatomic variables were recorded in each examination by one of 2 investigators (T.G. or A.J.P.) without knowledge of the catheterization findings: presence of main pulmonary artery atresia, hypoplasia, or stenosis; continuity of the pulmonary arteries; presence of branch pulmonary atresia (defined as luminal discontinuity), hypoplasia (defined as long-segment narrowing), or stenosis (defined as discrete narrowing); identification of APCs and their course; and patency of surgically placed shunts. To facilitate subsequent comparison with findings at catheterization, the APCs were labeled numerically according to their order of origin from the descending aorta and subclavian arteries. The smallest caliber of the pulmonary arteries and APCs was measured by one of the investigators (T.G.) on subvolume MIP images using electronic calipers. To determine interobserver variability, a second set of measurements was recorded by another investigator (G.F.G.) who was unaware of the first set of measurements or the findings at catheterization.

Cardiac Catheterization

Biplane contrast angiography was used and directed toward evaluation of the pulmonary arteries, APCs, and aortopulmonary shunts. In addition to aortography and ventriculography, pulmonary vein wedge angiography and selective injections of aortic arch branches and APCs were performed as needed.6 An investigator (M.L.) who was unaware of the MRA findings reviewed the angiograms and recorded the same anatomic variables as described above for MRA. Another investigator (A.C.M.) measured the caliber of the pulmonary arteries and APCs using catheter size for calibration.

Statistical Analysis

The MRA and catheterization findings regarding the anatomic variables detailed above were recorded on a spreadsheet (Microsoft Excel, version 5.0, Microsoft) and analyzed for discrepancies. When discrepancies were noted, a consensus based on surgical observations and catheterization was used as a reference standard. The agreement between MRA and catheterization measurements and interobserver variability were analyzed by calculating the mean difference (bias) and the standard deviation of the difference as described by Bland and Altman.19

Results

Patients

The demographic characteristics and cardiac diagnoses of the 32 patients who fulfilled inclusion criteria for this study are summarized in Table 1. One patient was included in a previous report.8 There were no complications associated with the MRI studies.

Comparison Between MRA and Catheterization

Table 2 summarizes and compares the MRA and catheterization findings. There was complete agreement between the 2 modalities in the diagnoses of branch PA hypoplasia and

<table>
<thead>
<tr>
<th>Table 1. Demographic Characteristics of 32 Patients With Complex Pulmonary Stenosis or Atresia</th>
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<tbody>
<tr>
<td>Sex, male/female</td>
</tr>
<tr>
<td>Median age at MRI (range)</td>
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<tr>
<td>Median time between MRA and catheterization (range)</td>
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<tr>
<td>Cardiac diagnoses</td>
</tr>
<tr>
<td>Tetralogy of Fallot with pulmonary atresia</td>
</tr>
<tr>
<td>Tetralogy of Fallot with pulmonary stenosis</td>
</tr>
<tr>
<td>Tetralogy of Fallot with dysplastic pulmonary valve</td>
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<tr>
<td>Status after Fontan operation</td>
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<tr>
<td>Status after hypoplastic left heart syndrome palliation</td>
</tr>
<tr>
<td>Isolated congenitally absent branch pulmonary artery</td>
</tr>
<tr>
<td>Status after pulmonary artery band</td>
</tr>
<tr>
<td>Williams syndrome</td>
</tr>
<tr>
<td>Heterotaxy syndrome</td>
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<tr>
<td>Status after arterial switch operation</td>
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<td>Status after coarctation repair</td>
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stenosis, which occurred in 38 vessels in 23 patients. MRA and catheterization findings were also concordant in the determination of PA discontinuity (n=4; Figure 1), absent branch PA (n=5; Figure 2), and the evaluation of surgical aortopulmonary (n=9) and cavopulmonary (n=4) shunt patency. MRA demonstrated long-segment main pulmonary atresia in 3 patients (Figure 3). Catheterization correctly diagnosed main pulmonary atresia in 1 of these patients. However, catheterization misclassified the main pulmonary artery as atretic in 3 other patients in whom a hypoplastic main pulmonary artery was demonstrated by MRA and confirmed at surgery.

A total of 51 APCs were found in 17 patients; 45 collateral vessels arose from the descending aorta and 6 from the subclavian arteries. There was complete agreement between MRA and catheterization in 14 patients with 48 APCs. Catheterization did not delineate 1 APC seen on MRA in 3 patients. The first was a 2.5 mm APC from the proximal descending aorta to the right upper lobe in a 46-year-old woman with tetralogy of Fallot, pulmonary atresia, and multiple APCs who underwent unifocalization of the APCs. The APC was imaged by MRA on both pre- and postoperative scans. The second was a 2 mm APC in a 1-week-old infant with tetralogy of Fallot and pulmonary atresia who underwent surgical repair with unifocalization. The APC was confirmed at surgery. The third was a 2.5 mm APC from the abdominal aorta to the right lower lobe in a 3-month-old infant with tetralogy of Fallot, dysplastic pulmonary valve syndrome, and multiple APCs to the right lung. A subsequent catheterization confirmed the collateral, which was coil occluded (Figure 4).

**Table 2. Comparison of MRA and Catheterization Findings in 32 Patients**

<table>
<thead>
<tr>
<th></th>
<th>MRA/Cath</th>
<th>MRA/Catheterization Discrepancies</th>
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<tbody>
<tr>
<td>MPA atresia</td>
<td>3/4</td>
<td>3 patients*</td>
</tr>
<tr>
<td>MPA hypoplasia or stenosis</td>
<td>10/7</td>
<td>3 patients*</td>
</tr>
<tr>
<td>LPA stenosis or hypoplasia</td>
<td>21/21</td>
<td>None</td>
</tr>
<tr>
<td>RPA stenosis or hypoplasia</td>
<td>17/17</td>
<td>None</td>
</tr>
<tr>
<td>Absent branch pulmonary artery</td>
<td>5/5</td>
<td>None</td>
</tr>
<tr>
<td>Discontinuous branch pulmonary arteries</td>
<td>4/4</td>
<td>None</td>
</tr>
<tr>
<td>Aortopulmonary collaterals</td>
<td>51/48</td>
<td>3 APCs were seen by MRA but not by catheterization (see text)</td>
</tr>
<tr>
<td>Aortopulmonary shunt patency</td>
<td>9/9</td>
<td>None</td>
</tr>
<tr>
<td>Glenn shunt patency</td>
<td>4/4</td>
<td>None</td>
</tr>
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Cath indicates catheterization; LPA, left pulmonary artery; MPA, main pulmonary artery; and RPA, right pulmonary artery.

*Catheterization misclassified the MPA as atretic in 3 patients in whom MRA and surgical observations demonstrated a hypoplastic vessel.

**Figure 1.** Gd-enhanced 3D MRA in a 32-year-old man with tetralogy of Fallot and pulmonary atresia, s/p left Blalock-Taussig shunt in infancy that subsequently occluded. MRA showed discontinuous pulmonary arteries. A, Subvolume MIP image in the oblique coronal plane showing a good-sized right pulmonary artery (RPA) and the distal segment of the main pulmonary artery (MPA). B, Subvolume MIP image in the axial plane showing a 2.5 cm distance (arrow) between the RPA and the hypoplastic peripheral left pulmonary artery (LPA). Ao arch indicates aortic arch; IVC, inferior vena cava; DAO, descending aorta; RUPV, right upper pulmonary vein; and RLPV, right lower pulmonary vein.

**Correlation Between MRA and Catheterization in Measuring Vessel Diameter**

To minimize confounding effects from changes over time, measurements by MRA and catheterization were compared only when the examinations were performed within 1 month of each other. In these 14 patients, the smallest diameter of 10 branch PAs and 33 APCs was measured by 2 blinded investigators. The measurements of vessel diameter ranged from 0.9 mm to 25 mm. Bland-Altman analysis of agreement yielded a mean difference (±SD) of 0.5±1.5 mm (Figure 5).

**Interobserver Variability of MRA Measurements**

Measurements by 2 independent observers of the smallest diameters of 10 branch PAs and 33 APCs on MRA of 14 patients had a mean difference of 0.4±1.5 mm.
Discussion

The results of this study demonstrate that compared with x-ray angiography, Gd-enhanced 3D MRA accurately depicted all sources of pulmonary blood supply in patients with complex pulmonary stenosis or atresia ranging in age from newborns to adults. Previous investigations on MRI evaluation of patients with abnormal pulmonary blood supply demonstrated that conventional 2D techniques such as spin echo and gradient echo cine sequences can reliably image the central pulmonary arteries and large APCs. Moreover, comparison between these MRI techniques and x-ray angiography showed good agreement in the measurements of the central pulmonary arteries. These traditional MRI techniques suffer, however, from several limitations. Although the introduction of faster MRI techniques, such as fast spin echo with double inversion recovery and segmented k-space fast gradient echo sequences, significantly shortened image acquisition time, complete anatomic coverage of the entire thorax may still be lengthy and require multiple breathholds. Consequently, this approach may be difficult for cyanotic patients or those with cardio-respiratory compromise. Furthermore, because APCs may arise from the subclavian arteries and from the abdominal aorta, the required anatomic coverage in these patients is large, extending from the thoracic inlet to the renal arteries. This is illustrated by the patient depicted in Figure 4 in whom major APCs to the right lung originated from the abdominal aorta immediately above the renal arteries, as well as from the descending thoracic aorta. Other limitations of traditional MRI techniques include their difficulty imaging very small blood vessels, vessels with slow flow, tortuous vessels, and the intrapulmonary segments of APCs and pulmonary arteries. These weaknesses are largely overcome by the 3D MRA sequence, which is capable of imaging large anatomic volumes in a single breathhold. Its high signal-to-noise ratio also allows for depiction of blood vessels as small as 0.5 mm, vessels with slow blood flow, and intraparenchymal pulmonary blood vessels.

It is worth noting that although this study demonstrates the advantages of Gd-enhanced 3D MRA over traditional MRI techniques in assessing all sources of pulmonary blood supply, other MRI sequences may also be useful during a comprehensive examination of patients with complex anomalies of the pulmonary arteries. Phase velocity cine MRI can quantify blood flow; segmented k-space fast gradient echo sequences can measure ventricular volumes, function, and mass; and fast spin echo sequences can minimize image artifact caused by endovascular stents or coils. Recently developed MRI techniques such as steady state free precession and parallel processing further enhance image quality and shorten acquisition time.

Technical Considerations

Several limitations of the MRA technique used in this study are noteworthy. Because both pulmonary arteries and veins are enhanced with contrast, the reader must be careful to distinguish them. The experience with this cohort and a previous study on contrast-enhanced 3D MRA evaluation of pulmonary and systemic venous anomalies indicate that pulmonary arteries and veins are easily differentiated using the subvolume MIP method. Another limitation of the MRA sequence used in this study is that the more peripheral branches of the pulmonary arteries, usually those beyond the third or fourth generations, are not routinely delineated. Finally, the present version of the MRA sequence does not allow reliable determination of which lung segments have
dual blood supply (ie, from the native branch pulmonary arteries and from APCs). Newer versions of this sequence, however, as well as recent improvements in MRI hardware and software, hold promise to overcome these limitations.21,22

Clinical Implications
The experience with this cohort has several important clinical implications. It suggests that MRA can accurately identify all sources of pulmonary blood supply noninvasively and is a viable alternative to diagnostic cardiac catheterization. Of particular interest are patients with tetralogy of Fallot and pulmonary atresia or pulmonary stenosis with diminutive pulmonary arteries. These patients traditionally undergo routine preoperative cardiac catheterization to identify all sources of pulmonary blood supply for surgical planning.3 Replacing this catheterization procedure with an MRA examination has several potential advantages, including reduced risks of morbidity and mortality, avoidance of ionizing radiation exposure, preservation of vascular access for future interventional catheterization procedures, and a lower cost.23,24 In patients with pulmonary overcirculation from excessive APCs, coil occlusion may be indicated. In these patients, MRA may provide an anatomic “roadmap” that shortens the duration of the catheterization and reduces the amount of contrast and ionizing radiation exposure.2 This approach has been used in several of our patients with satisfying results.
Altman.19
MRA and x-ray angiography using the method of Bland and Forschungsgemeinschaft (DFG), Bonn, Germany
Dr Greil was supported by grant No. 1751/1 from the Deutsche
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Figure 5. Comparison between branch PA and APC caliber on
MRA and x-ray angiography using the method of Bland and Altman.19

Study Limitations
Because of the retrospective nature of this study, the time
interval between MRA and catheterization was as long as 10
months. However, a maximal time interval of \( \leq 1 \) month was
selected for the comparison of measurements by these tech-
niques. Moreover, none of the discrepancies between MRA
and catheterization could be attributed to the elapsed time
between studies.

Conclusions
The results of this study indicate that Gd-enhanced 3D MRA
is a fast and accurate technique for delineation of all sources
of pulmonary blood supply in patients with complex pulmo-
nary stenosis and atresia and can be considered a noninvasive
alternative to diagnostic x-ray angiography. In conjunction
with information obtained by other pulse sequences, MRI can
provide a comprehensive evaluation of cardiovascular anat-
omy and physiology that facilitates planning for transcatheter
and surgical therapies.

Acknowledgments
Dr Greil was supported by grant No. 1751/1 from the Deutsche
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_Circulation_. 2002;106:473-478; originally published online July 1, 2002; doi: 10.1161/01.CIR.0000023624.33478.18

_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

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