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Original Article

A comparison of magnetic resonance angiography with conventional angiography in the diagnosis of tetralogy of Fallot

Renata Junqueira Moll Bernardes,[1] Edson Marchiori,[2] Paulo Manuel de Barros Bernardes,[3] Maria Beatriz Albano Monzo Gonzaga,[4] Luiz Carlos Simões[1]

[1]Department of Pediatric Cardiology, Laranjeiras National Institute of Cardiology, [2]Department of Radiology, Clementino Fraga Filho University Hospital, Federal University of Rio de Janeiro, [3]Department of Radiology, and [4]Department of Anesthesiology, Copa D'Or Hospital, Rio de Janeiro, Brazil

Abstract *Aims:* Our purpose was to assess the value of magnetic resonance angiography as a non-invasive alternative to catheterization in the evaluation of patients with tetralogy of Fallot, including those with pulmonary atresia. *Methods and results:* We evaluated prospectively, using magnetic resonance angiography, 30 patients, aged from 1 to 18 years, 15 with tetralogy of Fallot and pulmonary stenosis, and 15 with pulmonary atresia. The studies obtained using magnetic resonance provided adequate visualization of the aorta, and provided excellent imaging of the pulmonary trunk and its right and left branches. Compared with catheterization, magnetic resonance had 100 percent sensitivity, specificity and accuracy for defining the presence or absence of the pulmonary arteries. Magnetic resonance also had 93.9 percent sensitivity, 98.2 percent specificity, and 96.7 percent accuracy for detection of stenosis or hypoplasia of the pulmonary arteries. We detected 25 major aortopulmonary collateral arteries with magnetic resonance, but only 22 with conventional angiography. There was complete agreement between the two methods in detecting patency of the arterial duct in 6 patients, and of Blalock-Taussig shunts in 12 patients. *Conclusion:* Magnetic resonance angiography is a useful tool in the evaluation of patients with tetralogy of Fallot. It can be considered a non-invasive alternative to cardiac catheterization in the evaluation of the pulmonary vascular anatomy.

Keywords: Heart defects; congenital; cyanotic heart disease; tomographic imaging; pulmonary atresia

Tetralogy of Fallot is the most common cyanotic congenital cardiac malformation. The variant with pulmonary atresia is an extreme form of the anomaly, and occurs in one-fifth of the cases.[1] Although echocardiography with Doppler interrogation provides an adequate diagnosis of the intracardiac anatomy of these patients, cine-angiography is generally used to complement the echocardiographic study, since it allows a more accurate evaluation of the pulmonary vasculature and coronary arteries.[2–5]

Correspondence to: Renata Junqueira Moll Bernardes MD, Rua Barão de Jaguaripe, 191/101, Ipanema 22421-000 Rio de Janeiro RJ, Brazil. Tel: +21 2247 9508; Fax: +21 2537 8117; E-mail: paulombbernardes@terra.com.br

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The accurate assessment of the anatomy of the pulmonary vasculature is extremely important prior to operative intervention, particularly when unifocalization is being considered, since there frequently are pulmonary arterial abnormalities, and major aortopulmonary collateral arteries may be present, mainly in patients with co-existing pulmonary atresia.[1,2,4,6] Residual pulmonary arterial abnormalities are often detected after palliative and corrective surgeries.[1]

Cine-angiography has been considered the gold standard in the assessment of the pulmonary vasculature. It is an invasive technique, and the patient is exposed to ionizing radiation and contrast medium containing iodine.[2] On the other hand, magnetic resonance imaging is being intensively evaluated as a useful adjunct in patients with congenital cardiac disease,

as it is a noninvasive method that provides both functional and anatomic information.[7–13]

Although some previous studies have shown the utility of magnetic resonance angiography in the evaluation of vascular structures in congenital heart disease, these studies have included only a small number of patients with tetralogy of Fallot with pulmonary stenosis or atresia.[11,13,14] Many centres routinely submit these patients to catheterization better to define the pulmonary arteries and other sources of pulmonary blood supply before operative interventions.[1] Patients with tetralogy of Fallot and pulmonary atresia are not infrequently submitted to repeated catheterizations, better to delineate complex pulmonary vascular anatomy, including aortopulmonary collateral arteries, to follow the growth of the pulmonary arteries after palliative shunts, or to detect residual lesions after corrective surgeries.

In our study, we evaluated prospectively 30 patients using black-blood and gadolinium-enhanced angiographic magnetic resonance techniques. The results were compared to conventional angiography. Our purpose was to assess the value of magnetic resonance angiography as a non-invasive alternative to catheterization in the evaluation of these patients, and to discuss the indications for magnetic resonance angiography in patients with tetralogy of Fallot and pulmonary stenosis or pulmonary atresia.

Materials and methods

Design

The study was undertaken prospectively between January, 2002, and July, 2004, involving 30 patients with tetralogy of Fallot, half having pulmonary stenosis and the other half with pulmonary atresia. All patients were evaluated with black-blood and gadolinium-enhanced angiographic magnetic resonance imaging techniques. The results were compared to conventional angiography.

Population

Of the patients, half had the classic form of tetralogy of Fallot, with pulmonary stenosis, and the other half had the extreme variant, with pulmonary atresia, with 14 patients being female (46.7 percent) and 16 male (53.3 percent). Their ages ranged from 12 months to 18 years old, with a median age of 6 years. Their weights ranged from 8.6 to 59 kilograms, with a median of 15.8 kilograms.

The clinical findings at the time of the magnetic resonance examination were dyspnoea during effort in 19 patients, different degrees of cyanosis in16 patients and polycythemia in 9 patients. Nine patients were asymptomatic.

Surgical procedures had already been performed in 22 patients. Of these, 14 patients had been submitted only to palliative procedures, including construction of Blalock-Taussig shunts, unifocalization of pulmonary arteries, or construction of central shunts. In 4 patients, there had been only corrective surgeries, while 4 patients were initially submitted to palliative shunts that were removed during corrective procedures.

Our study complies with the Declaration of Helsinki, and approval was obtained from the institutional review board. Informed consent was obtained from each patient, or the parents, before the study.

Methodology for magnetic resonance imaging

Magnetic resonance imaging studies were performed with a 1,5 Tesla scanner (Signa, "Horizon"; General Electric Medical Systems, Milwaukee, Wisconsin, USA). General anaesthesia with sevoflurane was used in patients less than 10 years old. Ventilation was maintained with oxygen and a laryngeal mask. Monitoring was performed with a cardiac monitor, pulse oximeter, capnograph and noninvasive blood pressure monitor. Patients aged 10 years and older were studied without sedation with voluntary apnoea.

The choice of coil was based on the size of the patient, using a neurovascular phased-array coil for the smaller patients, and a torso phased-array coil for the larger ones. Localizing images were obtained with half-Fourier acquisition single-shot turbo spin-echo sequences, using a field of view of 36, and 12 slices, each of 8 millimetres thickness. T1-weighted spin-echo "black-blood" images were obtained in the axial plane, with an echo time of 30 milliseconds, repetition time from 2 to 4 heartbeats, field of view 21 to 28 centimetres, slice thickness of 3 millimetres, and a matrix of 256 by 128, or 256 by 160. A bolus timing sequence for T1-weighted two-dimensional gradient-echo, at 1 slice per second at the level of the aortic arch, was performed using 1 millilitre of gadolinium followed by a saline flush. The contrast transit time thus obtained was used to calculate the delay between the injection of the contrast medium and the start of the angiographic sequence. The delay equals the summation of the transit time plus the duration of injection of the contrast medium divided by 2, and minus half the duration of the angiographic sequence. A dose of 0.2 millimoles per kilogram of gadolinium was injected for the angiographies using the technique of hand injection. Two sequential breath-hold three-dimensional gradient-echo acquisitions were performed in succession without a time gap, using a turbo fast-field echo with linear filling of the K-space, the shortest repetition time and echo time, a flip angle of 30°, bandwidth of 62.5 kilohertz, field of view from

24 to 26 centimetres, slice thickness of 2.6 zero-filled to 1.3 millimetres, and an acquisition matrix of 320–256 by 128. In patients under anaesthesia, ventilation was suspended during acquisition of the images. Muscular relaxation was obtained with 0.2 to 0.3 milligrams per kilogram of atracurium to allow apnoea in these patients. The mean time to acquire the angiographic sequence was 20 seconds. Images were transferred to, and analyzed on, a separate workstation using multiplanar reformatting, maximal intensity projections, and volume rendering algorithms.

Methodology for catheterization

The time interval between the magnetic resonance studies and catheterization varied from 6 days to 12 months, with a mean of 4.0 months. We excluded from further study any patients submitted to any type of palliative or corrective cardiovascular surgeries during the interval between the studies. The angiographic studies included, whenever possible, right ventricular angiography, ascending and descending aortic angiography, and pulmonary angiography. Selective injection into the brachiocephalic arteries, coronary arteries, aortopulmonary collateral arteries, and also pulmonary venous wedge angiography were used when necessary to delineate as completely as possible the pulmonary arterial supply.

Analysis of images

Interpretation of magnetic resonance images was done by two independent observers, one radiologist and one paediatric cardiologist, who were unaware of the catheterization and clinical data. Any differences noted were resolved by consensus. The angiograms were interpreted by two paediatric cardiologists who were unaware of the magnetic resonance findings. All observers used the same criterions and reference values defined for the study and described below. We compared the results of the consensus interpretation of the findings from magnetic resonance investigation and catheterization.

Our reference values were based on normal values published by Snider and colleagues.[15] Structures were regarded as dilated or hypoplastic if they were greater than or less than two standard deviations, respectively. The presence of local stenosis in a vessel was defined as a reduction in diameter greater than 40 percent. Although the data published by Snider and colleagues[15] is based on echocardiography, little data exist on normative reference values for cardiac and vascular structures in children obtained using magnetic resonance imaging, except in small populations.[16] These data were compared to other echocardiographic and angiographic studies with good correlation.[14,17]

Statistics

We compared the results obtained with both methods, calculating sensitivity, specificity and accuracy of magnetic resonance angiography using conventional angiography as the gold standard.

Results

Analysis of the results included assessment of the aorta, the pulmonary trunk and its right and left branches, and identification of patency of the arterial duct, aortopulmonary collateral arteries, and surgically created shunts.

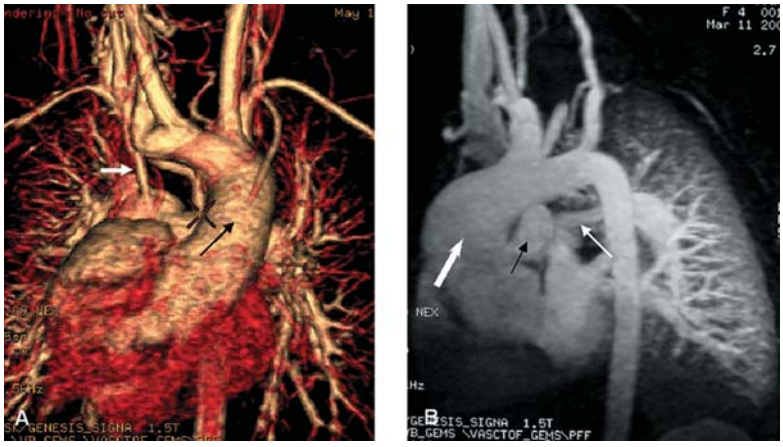

Figure 1.
Magnetic resonance angiography of the aorta. A: Dilated ascending aorta (black arrow), left-sided aortic arch and right-sided Blalock-Taussig shunt (white arrow). B: Dilated ascending aorta (broad white arrow) and left-sided aortic arch. Dilated right pulmonary artery (black arrow) and hypoplastic left pulmonary artery (narrow white arrow).

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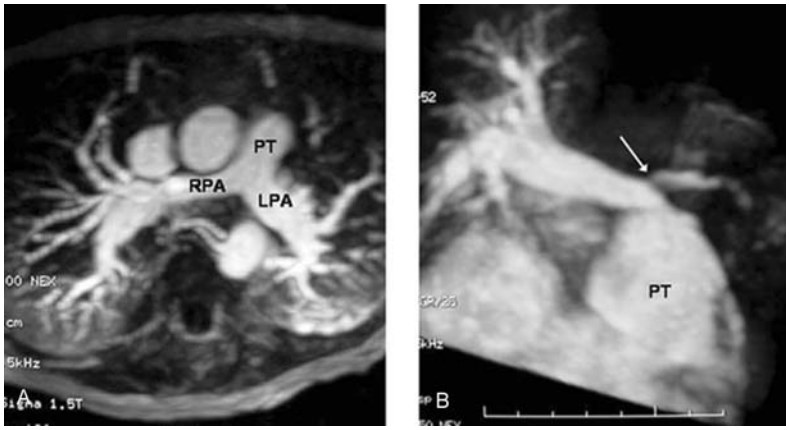

Figure 2.
Magnetic resonance angiography of the thorax. A: Narrow pulmonary trunk (PT) and normal right (RPA) and left (LPA) pulmonary arteries. B: Dilated pulmonary trunk (PT) and stenosis at the origin of the left pulmonary artery (arrow).

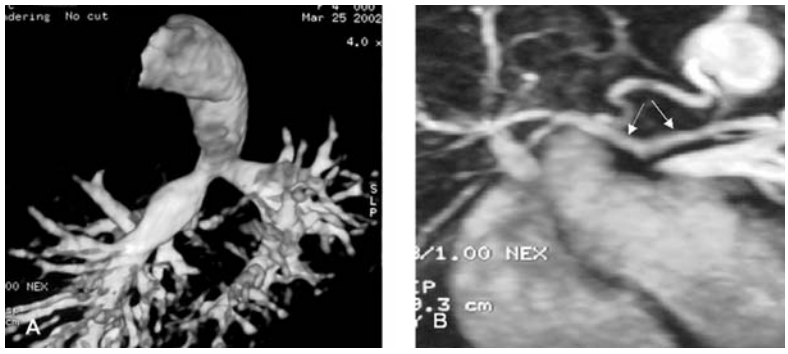

Figure 3.
Magnetic resonance angiography of the thorax. A: Stenosis in the origin of the right and left pulmonary arteries. B: Gadolinium-enhanced magnetic resonance angiography of the pulmonary arteries, showing hypoplasia of the right and left pulmonary arteries (arrows).

Aorta

The aorta was adequately visualized in all of the cases by contrast-enhanced magnetic resonance studies (Fig. 1). It was dilated in 21 patients (70 percent), and normal in nine (30 percent). The aortic arch was situated on the left side in 23 patients (76.7 percent), and on the right side in seven patients (23.3 percent).

Cine-angiography confirmed aortic dilation in 21 cases, normality in nine cases, and the location of the aortic arch in all 30 cases. There was complete agreement between the two methods regarding aortic assessment, with 100 percent sensitivity, specificity and accuracy for resonance imaging compared to catheterization.

Pulmonary arteries

The pulmonary arteries were assessed for normality, presence of stenosis, hypoplasia, dilation, or absence of the vessel. Magnetic resonance angiography allowed excellent visualization of the pulmonary trunk and its branches. We identified 86 vessels with magnetic resonance in the 30 patients, including 26 pulmonary trunks, 30 right, and 30 left pulmonary arteries. In 4 patients, (4.5 percent), the pulmonary trunk was judged to be absent. Of the pulmonary arteries, 39 were judged to be normal in size (43.3 percent), 20 hypoplastic (22.2 percent), and 15 dilated (16.7 percent). In 9 arteries, stenosis was seen at the origin (10 percent), while 3 had stenosis with downstream dilation (3.3 percent – Figs. 2 and 3).

There was agreement between magnetic resonance angiography and conventional angiography as regards 85 of the 90 pulmonary arteries (94.4 percent), but disagreement for 5 of the arteries (5.6 percent). In one case, the pulmonary trunk was judged normal by magnetic resonance angiography, shown to be hypoplastic by cine-angiography. In another case, the pulmonary trunk was again deemed normal by magnetic resonance angiography, with supravalvar stenosis being detected by angiography. In the case of two right pulmonary arteries, considered

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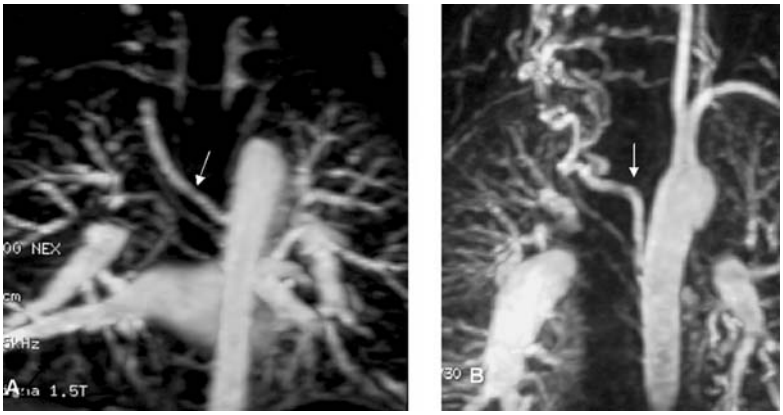

Figure 4.
A: A major aortopulmonary collateral artery identified by magnetic resonance angiography (arrow). This collateral vessel was not identified by conventional angiography. B: Magnetic resonance angiography showing an aortopulmonary collateral artery (arrow) originating in the descending thoracic aorta and directed to the right lung.

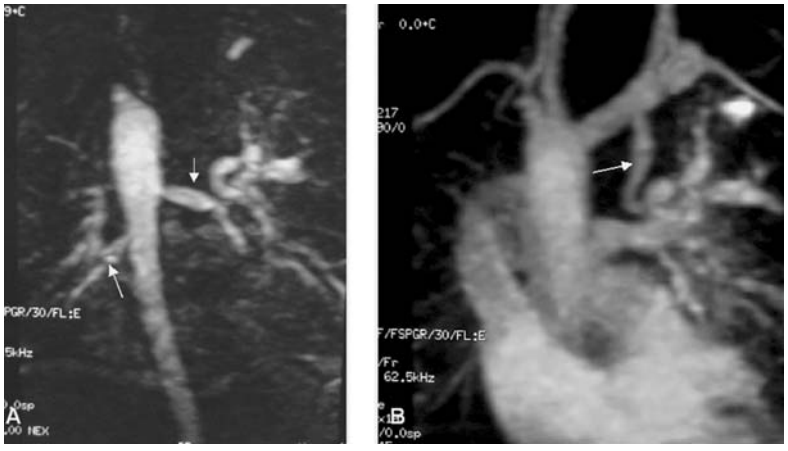

Figure 5.
Three aortopulmonary collateral arteries identified by magnetic resonance angiography. A: Two collateral arteries (arrows) originate in the descending thoracic aorta. B: A third collateral artery (arrow) originates in the brachiocephalic artery. This collateral vessel was not detected by conventional angiography, but was confirmed during surgery.

hypoplastic by magnetic resonance angiography, cine-angiography revealed normal findings in one patient, and proximal stenosis in the other. In the last discordant case, the left pulmonary artery was regarded as hypoplastic by magnetic resonance angiography, but proximal stenosis was revealed by cine-angiography.

When compared to catheterization, therefore, magnetic resonance angiography had 100 percent sensitivity, specificity and accuracy for defining the presence or absence of the pulmonary arteries. It had 93.9 percent sensitivity, 98.2 percent specificity, and 96.7 percent accuracy for the diagnosis of pulmonary arterial stenosis or hypoplasia.

Aortopulmonary collateral arteries

In half of our patients, magnetic resonance angiography detected aortopulmonary collateral arteries originating from the aorta and its branches. The number of collateral arteries varied from 1 to 3 per patient. In all, we detected a total of 25 collateral arteries. Of these, 24 arose from the descending aorta (Figs. 4 and 5A), and one from the brachiocephalic artery (Fig. 5B). Of the 15 patients with pulmonary atresia, 14 (93.3 percent) had aortopulmonary collateral arteries, in addition to one patient with tetralogy of Fallot and pulmonary stenosis (6.7 percent).

Cine-angiography identified a total of 22 collateral vessels, all of them also identified by magnetic resonance angiography. In one case, a third collateral artery was identified solely by magnetic resonance angiography, and this was confirmed during surgery (Fig. 5B). In another case, two vessels were identified by magnetic resonance, but only one of them was detected by cine-angiography. In a third

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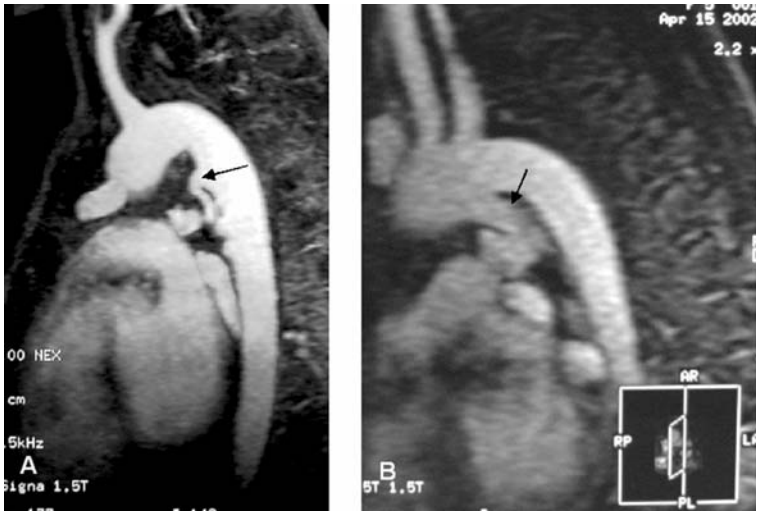

Figure 6.
Patency of the arterial duct detected by magnetic resonance angiography in two patients (arrows).

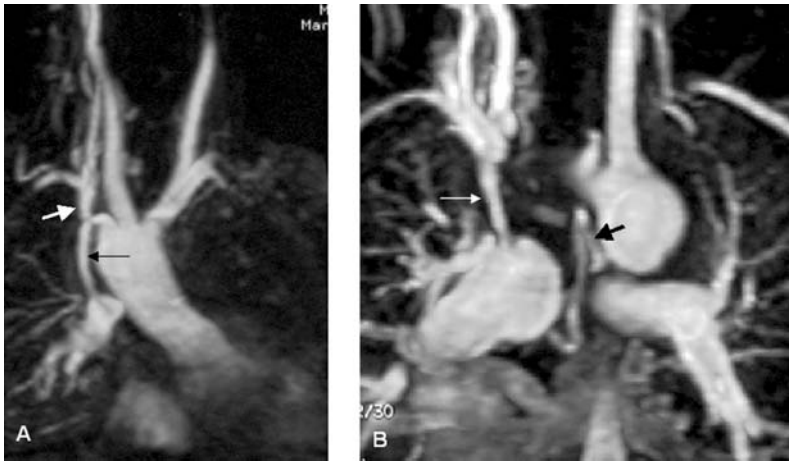

Figure 7.
A: Magnetic resonance angiography detected a patent right-sided modified Blalock-Taussig shunt (black arrow). The right subclavian artery is being pulled down by the graft (white arrow). B: Magnetic resonance angiography shows a right-sided modified Blalock-Taussig shunt (white arrow) and an aortopulmonary collateral artery (black arrow).

case, a collateral vessel was visualized by magnetic resonance, but not by cine-angiography (Fig. 4A).

Accordingly, all of the collateral arteries (100 percent) identified by cine-angiography were also identified by magnetic resonance. Magnetic resonance angiography additionally identified 3 collateral arteries that were not detected by conventional angiography, and one of them was confirmed during surgery.

Arterial duct

A patent arterial duct was detected with magnetic resonance angiography in six patients (20 percent – Fig. 6). Patency of the duct was confirmed by conventional angiography in all these cases. Compared to conventional angiography, magnetic resonance achieved total sensitivity, specificity and accuracy for identification of patency of the arterial duct.

Aortopulmonary shunts

We detected 12 patent Blalock-Taussig shunts with magnetic resonance angiography (Fig. 7), four on the left side and eight on the right side. These findings proved to be in complete agreement with those obtained using cine-angiography. Of the Blalock-Taussig shunts, 2 were shown to be occluded with both methods, this being confirmed in one during surgery. A central shunt was not visualized by either of the methods. When compared to catheterization, therefore, resonance imaging was 100 percent sensitive, specific, and accurate in the identification of aortopulmonary shunts.

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Discussion

The adequate delineation of the aorta with magnetic resonance angiography was in agreement with studies that found excellent results with use of contrast enhanced magnetic resonance angiography for the assessment of the aorta and its branches.[18] It was possible to delineate the aorta and the pulmonary vasculature almost simultaneously, using two sequential breath-hold three-dimensional magnetic resonance angiography acquisitions. The possibility to study the aorta without increasing the duration of the exam is important, because aortic dilation is often found in these patients,[19] and also because the position of the aortic arch should be determined, particularly before palliative procedures.

Magnetic resonance angiography provided an excellent delineation of the pulmonary trunk and its right and left branches. Some studies report a good correlation between conventional magnetic resonance imaging and angiographic measurements of the pulmonary arteries in patients with pulmonary stenosis or atresia, although sometimes magnetic resonance underestimated the size of the pulmonary arteries.[4,6,9,20]

The excellent delineation of the pulmonary arteries is in agreement with other studies, which achieved 92.7 to 100 percent sensitivity, specificity and accuracy for magnetic resonance angiography when compared to conventional angiography for the diagnosis of stenotic, hypoplastic, and discontinuous pulmonary arteries.[11,13,14,21]

Geva et al.,[13] for example, studied retrospectively 32 patients with diverse congenital cardiac malformations, including 13 patients with tetralogy of Fallot and pulmonary atresia, and 4 patients with tetralogy of Fallot and pulmonary stenosis. In their hands, magnetic resonance was 100 percent sensitive and specific for the diagnosis of stenosis or hypoplasia of the pulmonary trunk and its branches, as well as for the identification of absent and discontinuous branches of the pulmonary trunk.

Roche et al.[14] had studied 11 patients with tetralogy of Fallot and pulmonary atresia. In their study, magnetic resonance imaging depicted four pulmonary trunks, and a total of 17 right or left pulmonary arteries, showing confluence of the pulmonary arteries in five of the six patients in which this was seen at conventional angiography, giving 100 percent sensitivity, 83 percent specificity, and 91 percent accuracy.

Kondo et al.[11] evaluated prospectively 73 patients with various congenital cardiovascular defects, including 12 patients with tetralogy of Fallot. In these patients, resonance imaging was 92.7 percent sensitive, 96.2 percent specific, and 95.2 percent accurate in detecting stenoses in the right or left pulmonary arteries.

It is the aorto-pulmonary arteries that achieve major importance when tetralogy of Fallot is associated with pulmonary atresia. When considering the visualization of these vessels, our results were in accord with other studies that have reported excellent identification using magnetic resonance angiography.[13,14,21] Geva et al.[13] identified 48 aortopulmonary collateral arteries with magnetic resonance angiography and cine-angiography. In their series, 3 additional collateral arteries were diagnosed by magnetic resonance, but not by catheterization. Roche et al.,[14] using magnetic resonance angiography, reported 80 percent sensitivity, 100 percent specificity, and 87 percent accuracy in identification of 36 collateral arteries. Prasad et al.[21] also reported good identification of collateral arteries in 16 adults using magnetic resonance angiography.

Although only a small number of patients had patent arterial ducts, all were detected by magnetic resonance angiography. Holmqvist et al.[4] detected a large duct in one patient, from 14 studied, that had been described as an aortopulmonary collateral artery by magnetic resonance. Some studies have suggested that surgical shunts can be identified using conventional techniques for magnetic resonance imaging, such as spin-echo sequences.[4,10] The delineation of these structures, however, is not always possible with this technique and it often requires acquisitions in multiple planes.[4,6,9,10] In our study, we found magnetic resonance angiography to be very useful when assessing aorto-pulmonary shunts. Our findings again endorse previous studies, with Geva et al.[13] reporting concordance between magnetic resonance angiography and catheterization findings in the evaluation of 9 aortopulmonary and 4 cavopulmonary shunts. Prasad et al.,[21] using contrast-enhanced magnetic resonance angiography, identified all 9 surgical shunts which were patent in the 16 patients they studied.

Conventional magnetic resonance techniques does suffer from some limitations, including difficulty in imaging very small blood vessels, tortuous vessels, and vessels with slow flow. These limitations can be overcome by using contrast enhanced angiographic techniques. The high signal-to-noise ratio provided with this technique allied to fast scan times, with imaging during a single breath-hold reducing respiratory artefacts, allows excellent depiction of blood vessels. Three dimensional acquisition of the images allows multiple post-processing techniques, and facilitates analysis of the data. In this respect, the most frequently used techniques are multiple angle reformatting, maximum intensity projection, and shaded surface reconstruction.

Our study was limited in that the time interval between the two examinations did not permit direct comparison of measurements between the two

methods. Even so, there was a good agreement of the findings. Conventional angiography, therefore, must still be considered the gold standard for evaluation of the anatomic arrangement of the pulmonary vasculature, but it is an invasive method that uses ionizing radiation, contrast medium containing iodine, and is associated with significant complications. Repeated catheterizations are often performed in these patients to assess growth of the pulmonary arteries after palliative surgical procedures, and to define the possibility and optimal time for corrective surgery, or even to assess residual pulmonary stenosis after corrective surgeries. Our study indicates that magnetic resonance angiography is a very useful method with which to evaluate patients with tetralogy of Fallot with or without pulmonary atresia, and it can substitute for conventional angiography in the assessment of pulmonary vascular anatomy in diverse situations, thus avoiding repetitive catheterizations and preserving vascular access for future interventional procedures.

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