Cardiac MRI in Congenital Heart Disease – Our Experience

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Abstract

A significant recent advance that has occurred world over in the continuously evolving field of Magnetic Resonance Imaging (MRI) practice is the introduction of Cardiac applications. Cardiac MRI has moved to the centre stage of clinical management strategy by non-invasively imaging the structure as well as function of the heart. It has a wide range of specific applications such as delineation of morphological anatomy, quantification of flow and pressure across cardiac valve dysfunction, evaluation of myocardial function, assessment of infarcts, mapping coronary arteries and so on. Evaluation of congenital heart disease (CHD) is an important application of Cardiac MRI since the morphological details of chambers, septum, defects and anomalous connections are depicted accurately. Besides, flow information across valves, chambers, outflow tracts and shunts are also provided. This article describes our experience in the use of cardiac MRI in congenital heart disease.

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Introduction

Congenital heart disease is a common clinical entity occurring in 0.8% of live newborns [1]. During infancy and childhood a large number of cases are diagnosed and treated. Notwithstanding this, a large subset of 150,000 adults are undiagnosed and untreated [2,3]. On the MRI front, Cardiac MRI has emerged as an accurate method for the evaluation of structure and function of the heart. Significant advances in MR technology like the emergence of high-field-strength magnets, high-performance gradient hardware and ultrafast pulse sequences have reinforced traditional techniques like cardiac gating and suspension of breathing resulting in a generation of newer sequences that address the issues of anatomical mapping and functional analysis of the heart, well into the realms of cardiac cycle [4,5].

Material and Methods

Patient Population

Twenty-six patients with an age range 1½ to 38 years with CHD underwent MR imaging at our Institute from Jan 2003 to Jul 2004. 10 male and 16 female patients formed the study cohort. 4 patients of tetralogy of fallot (TOF) were scanned before and after surgery. All patients were previously evaluated by echocardiography except the cases of coarctation of aorta. MR imaging was performed using a 1.5 T (Magnetom Symphony with Quantum gradients (maximum gradient amplitude 30 mT/m; slew rate 125 mT/m/sec); Siemens Medical Systems, Erlangen, Germany) with use of an

integrated body coil. Each patient was evaluated in detail by clinical and cardiological examination with echocardiographic confirmation and cardiac catheterization data and operative status.

Patient preparation

For all patients in this study, MR-compatible electrocardiographic leads were placed on the anterior chest wall and attached to the MR imaging unit for electrocardiographic gating. An integrated body array coil was used. The referring physician and anesthetist were consulted for the type of sedation (general anesthetic sedation, deep sedation or conscious sedation) for every case. Sedation techniques included medication by midazolam, ketamine injection or propofol infusion. Scans were performed generally using non breath holding techniques, with few cases undergoing breath-holding scan techniques.

Cardiac MRI Protocol

MR imaging protocol commenced with a localizer using Tru FISP (Fast Imaging with Steady-state Precession) sequence. Axial and Coronal scans of 10mm thickness using Half Fourier Acquired Turbo Spin Echo (HASTE) sequence were obtained with acquisition extending from the level of aortic arch to below the diaphragm. It was useful in demonstrating ventricular and atrial septal defects as well as providing anatomical landmark for deriving oblique sections, subsequently. From the localizer, vertical long axis, short axis and horizontal long axis (4-chamber) images were obtained. Functional analysis was performed using a combination of Cine Trufi (steady-state free precession), Cine Flash (gradient-

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echo) and Cine phase-contrast sequences. Contrast-enhanced MR Angiography (CE MRA) using a 3D gradient echo sequence was performed to evaluate the vascular anomalies associated with the cardiac defects following an IV bolus of MR contrast (Magniscan, Dabur) at a dose of 0.1 to 0.2 mmol/kg body weight delivered with a pressure injector. The average duration of the entire study was about 35 minutes with a range of 20 to 45 minutes depending on whether CE MRA was included in the protocol or not.

Results

Thirty cardiac MRI studies were performed on 26 patients with CHD. There were 10 male and 16 female patients while the age range was 1½ to 38 years. The commonest congenital heart disease in this study evaluated by MRI was tetralogy of Fallot. 13 studies were performed in 9 patients with TOF, including four studies done on cases before and after surgery. The other cases in this study, consisted of 6 cases of atrial septal defect (ASD), 3 cases of ventricular septal defect (VSD), patent ductus arteriosus (PDA) and Coarctation each, 1 case of truncus arteriosus (TA) and single ventricle each. MR images of each patient were analysed for morphological information like chamber and valve anatomy, structure and integrity of septum, alignment and caliber of outflow tracts and atrioventricular connections. The functional information comprised of quantification of flow across valves, outflow tract and defects. Cine imaging provided dynamic information

of the cardiac size, valve morphology, leaflet mobility, wall thickness, chamber size, flow jets, outflow tracts, septum anatomy, defect morphology and aortopulmonary connections.

Discussion

In the last few years, Cardiac MR imaging has been increasingly considered as a valuable modality for evaluation of CHD [5,6]. Cardiac MR imaging has much to offer by not only generating high resolution morphological images, but also provides quantitative information of the severity of regurgitant or stenotic lesions or shunts with peak velocity and flow measurements across them [4]. An overview of the wide range of cardiac MRI techniques that can be used in congenital heart disease is presented as Table 1.

Cardiac MRI has always posed a big challenge to researchers in MR technology, due to the specific exigency of imaging a moving organ. Rising to this challenge, current state of art MRI offers a physiological snapshot of the heart in relation to the cardiac cycle. In practice, there are other important challenges faced in Cardiac MRI [7]. These include picking the optimal combination of imaging techniques from a numerous set of sequences, interference in MR acquisition from

Table 1
Overview of cardiac MR imaging strategies in congenital heart disease

Strategy	Family of sequence	Physical principles	Utility in congenital heart disease
Black Blood Sequence	Spin-echo imaging	Blood exposed to an initial 90° pulse moves out of section prior to receiving 180° pulse, leading to black blood appearance	Ideal for anatomy, is less susceptible to metal, but cannot provide optimal cine information
White Blood Sequence	Gradient-echo imaging	There is no slice-selective refocusing pulse following the initial excitation pulse, with all the excited spins detected. Inflow of “fresh” unsaturated spins results in bright signals	Identifies areas of turbulent flow caused by stenotic or regurgitant valves, septal defects. Useful for evaluating ventricular mass, chamber volumes, and ejection fraction
Cine FLASH Sequence	Segmented k-space gradient-echo imaging	Requires stronger gradient systems with faster switching capabilities	Provides anatomic and physiologic info of simple and complex congenital heart lesions
Cine TRUF1 Sequence	Segmented k-space gradient-echo imaging	Acquisition of multiple lines of k space per R-R interval, which decreases the imaging time	Depicts morphologic and functional abnormalities of congenital heart disease with great clarity and more rapidly than FLASH Images
Cine Phase Sequence Velocity encoded	Modified Phase-contrast	Multiple images in the same MRA section	Determines direction of blood flow, and quantifies blood velocity and blood flow gradients across shunts and valves
Gadolinium enhanced MR angiography	3D spoiled GRE with interpolation	Paramagnetic Chelates shorten spin-lattice relaxation time of blood, giving it a high signal intensity on T1-weighted images. It is not cardiac gated, requires a shorter time of acquisition, and allows 3D images during a breath hold.	3D volume rendering of Gd-enhanced MR angiographic data provides information about stenoses and anatomic relationships of aorta and pulmonary vasculature.
Myocardial tagging Cine imaging	Modified gradient Echo sequence	Saturation bands are placed across the myocardium at the end of diastole, throughout the cardiac cycle	Used to evaluate right ventricular dysfunction of patients with right ventricular outflow obstruction, and to study the left ventricle of patients with transposition of the great arteries

cardiac arrhythmias that result in non conclusive studies, difficulty in conducting breath hold sequences in cyanotic infants and the need for anesthesia and monitoring in critical cases. Notwithstanding these limitations and difficulties, there is an increasing role of Cardiac MRI in the evaluation of structure and function of the heart.

With the increasing use of Cardiac MRI in specialised centres across the world, a general consensus has emerged regarding the indications for MRI in the evaluation of patients with congenital heart disease [8]. They include: a) segmental description of cardiac anomalies; b) evaluation of thoracic aortic anomalies; c) noninvasive detection and quantification of shunts, stenoses and regurgitations; d) evaluation of cono-truncal malformations and complex anomalies; e) identification of pulmonary and systemic venous anomalies; and f) postoperative studies.

Congenital heart diseases have been traditionally classified into the following categories, namely a) potentially cyanotic; b) cyanotic and c) acyanotic. The potentially cyanotic group consists of ASD, VSD and PDA. The cyanotic group comprises of TOF, tricuspid atresia, TA, single ventricle, total anomalous pulmonary venous drainage (TAPVD) and transposition of great vessels (TGV). The acyanotic group includes coarctation. The role of MR imaging in the evaluation of few of these common types of congenital heart lesions is described herein.

a) Potentially cyanotic group

ASD are the most common type of CHD to present in adult life and account for 10% of CHD. ASD are classified into three morphological types: secundum which is the commonest, sinus venosus and primum. The sensitivity and specificity of MRI for the diagnosis of ASD are both > 90% [9]. The defect is best seen in horizontal long axis and oblique sagittal sections, as was evident in this study (Fig 1).

The defect preferably should be identified on MRI in two or more sections reinforced by correlation with right-sided volume loading. The dilated right chambers rotate the heart into the left chest across the midline. An important finding in ASD presenting in adults is pulmonary hypertension which occurs in 40 % cases, mostly due to increased flow. This finding and associations like mitral valve prolapse and regurgitation can also be identified.

VSD is classified into following morphological types: membranous (70-80%), muscular (10%), endocrinal cushion defects (5-10%) and conal (5%). MRI identifies the defect in the above locations and shows increased vascularity when a 2:1 or greater shunt exists. Moreover, Cine gradient echo images can differentiate left to right,

Fig. 1 : Atrial Septal Defect : “Freeze Frame” Contrast enhanced MRA of the of a 34 year old lady with a large atrial septal defect with reconstructed images in coronal plane reveals opacification of right atrium and pulmonary trunk, with unopacified blood from left atrium entering through the defect (black arrow) at left panel. Image obtained 5 seconds later at right panel shows opacification of both atria, with shunting of blood from left to right. Note the large size of the defect in coronal plane represented by dotted lines.

right to left and bi-directional shunts. In this study, on evaluation of the 3 patients with VSD, the defect was correctly identified in horizontal and vertical long axis views, while the dilatation of left atrium and right ventricle was identified on axial images.

PDA is the persistence of the sixth aortic arch. The ductus arteriosus is a fetal connection between origin of the left pulmonary artery and the descending aorta just beyond the origin of the left subclavian artery. Soon after birth it closes due to a decrease in pulmonary arterial pressure as well as contraction of the muscles in the duct wall. In PDA, there is a persistent communication of the ductus arteriosus with blood shunted from the aorta to the left pulmonary artery as shown in this case of PDA that was associated with a single ventricle (Fig 2).

b) Cyanotic group

Tetralogy Of Fallot is the commonest cyanotic congenital heart disease, with a combination of overriding of aorta, VSD, infundibular pulmonary stenosis, and right ventricular hypertrophy. MRI demonstrates all findings in this condition including infundibular stenosis, ventricular septal defect which is either membranous or muscular beneath crista. It also shows a wide range of associated anomalies of pulmonary valve and coronary artery, major aortopulmonary collateral arteries (MAPCA's), right aortic arch with either mirror imaging branching of great vessels or an aberrant left subclavian

Fig. 2 : Single Ventricle : Contrast enhanced MRA of a 15 year old cyanotic girl with single ventricle in coronal plane reveals abnormal pulmonary connections. Pulmonary artery is formed by shunting of blood from aorta through a patent ductus arteriosus (white arrow), while right pulmonary artery originates from the single ventricle (white block arrow). No MAPCA's (major aortopulmonary collateral arteries) are present.

artery. MRI is also excellent for evaluating both palliative and corrective surgical repair that have effectively resulted in improved long term survival [10]. Palliative shunt corrections between the systemic and pulmonary circulations include Blalock-Taussig shunt (described first in 1944) between subclavian artery and pulmonary artery (Fig 3). Definitive repair is usually performed at 5-7 years which consists of pericardial patching the VSD and wide reconstruction of the RV outflow tract to relieve the pulmonary outflow obstruction. During follow up period, MRI provides useful information on adequacy of pulmonary circulation (Fig 3) and regurgitation at reconstructed pulmonary outflow tract which does not have a valvular apparatus [11,12].

Single ventricle is synonymously called as primitive ventricle, common ventricle, double or single inlet left or right ventricle, cor biloculare and triloculare. In this admixture lesion, mixing of venous and arterial blood occurs within the heart and great arteries. In this condition, both atria and their AV valves communicate with a single ventricle [3] (Fig 2). The Fontan surgical procedure and its modifications is the common surgical technique designed to direct total systemic venous return to the lungs [13].

TA presents with a common aorticopulmonary trunk, as a result of a failure of splitting by a spiral septum of the primitive common truncus arteriosus into aorta and pulmonary artery. It is classified into 4 types : Type 1

Fig. 3 : Blocked Blalock-Taussig shunt : Contrast enhanced MRA in a 6 year-old girl with tetralogy of Fallot subject to palliative repair by Blalock-Taussig shunt at 3 years age, presenting with severe pulmonary insufficiency. The study was done to evaluate a blocked shunt for corrective repair. Left panel shows oligemia at left side (white arrow). Note presence of left sided vena cava, a known association of TOF (outline block arrow). Right panel reveals faint opacification of left subclavian artery with features suggestive of a blockage of BT shunt (white arrowhead).

Fig. 4 : Truncus Arteriosus : Panel of Coronal T1 MRI sequences in a 8 yrs cyanotic boy shows a single arterial vessel at the base of the heart without a distinct and separate aorta or pulmonary trunk from which the aortic arch, pulmonary, coronary arteries originate. The pulmonary arteries (white block arrows) originate from the left inferior aspect of the common arterial trunk with a short main stem, classifying it into Type I Truncus Arteriosus.

(pseudopulmonary trunk) describes a common truncal vessel giving off a well formed main pulmonary artery which bifurcates into right and left pulmonary arteries. Type 2 has separate pulmonary arteries arising directly from dorsal surface of the ascending truncal vessel. Type 3 has separate pulmonary arteries arising from lateral surface of the ascending truncal vessel. Type 4 is essentially called pseudotruncus occurring due to pulmonary conal agenesis, rather than failure of truncal septation. In this study, there was one case of Truncus Arteriosus, exhibiting Type 1 / pseudopulmonary trunk features (Fig 4). It is associated with a large, high riding VSD and an ASD can occur in about 20% of cases.

c) Acyanotic group

Coarctation of aorta is characterised by a congenital narrowing of the aorta, occurring just distal to the left

Fig. 5 : Coarctation of Aorta : MRI Images in a 38 year hypertensive female shows coarctation of the aorta (white block arrow) distal to origin of left subclavian artery. Contrast enhanced MRA reveals numerous collaterals involving the intercostals (white arrows), internal mammary and dorsal scapular arteries, diverting blood flow to lower body and lower limb. Delayed phase reveals increased opacification of the distal thoracic aorta (white arrowhead), principally through incoming blood from the intercostals.

subclavian artery, either proximal, adjacent or distal to the site of entry of the ductus arteriosus, or insertion of the ligamentum arteriosum. In infants, it is associated with a VSD in 50% of cases and a PDA in 30%, which supplies blood to the descending aorta beyond the area of narrowing. In adults, it presents with systemic hypertension, aortic dissection, stroke, heart failure or endocarditis. MRI is useful in diagnosis of coarctation, in presurgical planning, and during post surgical follow-up [6]. Preoperatively, MRI offers useful information on location and severity of the narrowed aortic segment. The coarctation segment is best seen on oblique coronal and sagittal images [Fig. 5]. Besides MRI delineates the collateral pathways, that involves subclavian, internal mammary, intercostals, dorsal scapular and epigastric arteries.

MR imaging appears to be superior to echocardiography for determining abnormalities of the pulmonary arteries in patients with cyanotic CHD [14,15]. In our study, MRI proved superior to echocardiography in the delineation of the presence and severity of pulmonary atresia in 7 of 9 patients with TOF. While echocardiography was able to identify pulmonary atresia in these patients, MRI was beneficial in delineating the severity of the same and associated abnormalities of the branch pulmonary arteries, which could not be reliably visualized on echocardiography. This was particularly true in assessing the pulmonary vasculature post-operatively in four patients with TOF who underwent post-operative cardiac MRI. In one of these patients, pulmonary oligoemia due to a blocked Blalock-Taussig shunt could be ably demonstrated on

CE MRA (Fig 3) while echocardiography could not provide any information regarding the pulmonary artery branches. Similar findings have been reported by other workers in the post-operative evaluation of pulmonary arteries [16]. In one patient with TA in our study, MRI and CE MRA were crucial in classifying the type of TA which could not be assessed on echocardiography.

CE MRA was the single most useful sequence in delineating the vascular abnormalities associated with cardiac anomalies including coarctation of the aorta in which echocardiography has a limited role. In 3 patients with PDA in our study, echocardiography had suggested the possibility of the same. However, CE MRA was conclusive in the depiction of the PDA and in assessing its size and location with specific information regarding the site of its pulmonary arterial attachment. Roche et al have suggested that combined cine gradient-recalled echo MRI and MR angiography is a reliable method for imaging pulmonary vascular supply in patients with cyanotic CHD and have suggested that it may preclude the need for conventional catheter angiography in the pre-operative evaluation of these patients [17].

Conclusion

Cardiac MRI is a useful tool in evaluation of congenital heart disease. In practice, its role is not limited merely in the delineation of morphologic appearances of various defects, shunts and anomalies but it offers useful data on functional information. While echocardiography is a widely available, cost-effective and convenient modality for the assessment of cardiac anomalies in a patient with CHD, MRI and CE MRA are valuable tools for the assessment of the associated vascular abnormalities and flow patterns which are crucial in the pre-operative evaluation of these patients. CE MRA provides angiographic information which would otherwise be provided by invasive catheter angiography. In the assessment of CHD, catheter angiography requires the placement of catheters in various systemic and pulmonary vessels to derive optimum information regarding the vascular anomalies and associated shunts. CE MRA is not only non-invasive and free of the hazards of ionizing but also dispenses with the need for iodinated contrast media which are subject to dose-restrictions in the evaluation of infants and young children due to their nephrotoxicity and other potential side effects. Gadolinium-based MR contrast agents do not suffer from these limitations and are safe to use even in the pediatric age group. All these significant advances have encouraged the use of Cardiac MRI not only in presurgical planning, but also during post surgical follow-up of congenital heart diseases.

The advantages of Cardiac MRI have to be weighed

against the disadvantages in terms of limited availability and cost. The need for adequate sedation including general anesthesia has to be factored in while assessing the current role of cardiac MRI in the evaluation of CHD. It is expected that continuing advancements in fast scanning MRI technologies combined with wider availability and cheaper access with further push the frontiers of cardiac MRI applications.

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