

When to Order Cardiovascular Magnetic Resonance in Adults with Congenital Heart Disease

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Cardiovascular magnetic resonance (CMR), where available, contributes to the informed management of patients with congenital heart disease. In contrast to echocardiography, CMR becomes easier as patients grow. It is versatile and gives unrestricted access to the heart and intrathoracic vessels, providing functional and structural information. Its relative strengths are discussed, and examples are given of congenital conditions in which it provides clinically important information. CMR can prevent the need for diagnostic catheterization or expedite intervention if indicated, enabling planned, directed procedures. In our practice, CMR is used for serial follow-up, investigation of altered symptoms or signs, planning of transcatheter or surgical interventions, and for baseline assessment after surgery. As CMR becomes more widely available, it will contribute increasingly to the lifelong management of patients with congenital heart disease.

Introduction

The past 50 years have seen major improvements in the prognosis of patients with congenital heart disease (CHD). Consequently, the population of adults with CHD is growing [1•]. An integral part of the ongoing care of these patients includes serial follow-up. This is to investigate and treat the underlying condition, residual lesions after reparative surgery, and any long-term sequelae. Cardiovascular magnetic resonance (CMR) is an important modality in the management of this diverse group, providing information that complements that obtained by transthoracic and transesophageal echocardiography [2,3], computed tomography, or invasive angiography (Table 1).

What Does a Cardiovascular Magnetic Resonance Study Involve?

The CMR study generally lasts 30 to 45 minutes, during which the patient needs to be able to lie still in the magnet, typically in a magnetic field of 1.5 Tesla. In our experience, claustrophobia can be problematic in about 5% of patients. The magnetic gradients cause noise and the patient wears headphones for ear protection and to receive instructions for breath-hold acquisitions. Electrocardiograph (ECG)-gated cardiovascular images are acquired using sequences applied at specific time delays after the R-wave of the ECG, usually through several successive heart cycles, so arrhythmias may degrade image quality. A reasonably comprehensive CMR study entails 20 to 30 breath-hold acquisitions, located with respect to preceding scout images.

Contraindications and Potential Hazards

A permanent pacemaker in a pacemaker-dependent patient is a contraindication to CMR, as is a ferromagnetic cerebral aneurysm clip or a steel fragment lodged in the eye after injury. Most metallic devices and clips implanted in the chest are safe, however, as long as they do not incorporate electrical devices [4,5]. Ferromagnetic implants cause local artifacts, but this does not usually negate the usefulness of the investigation. Various items of personal or hospital equipment may be ferromagnetic, for example jewelry, hair clips, keys, scissors, gas cylinders, wheelchairs, or patient trolleys. They could become potentially lethal missiles if inadvertently brought into the strong magnetic field, so security measures and continual vigilance are needed.

Versatility

Cardiovascular MR gives unrestricted access to the chest in multiple, freely chosen slices. It provides clear images of anatomy throughout the chest. Multiple slices covering the volume of the heart and mediastinum can be acquired in a single breath-hold. Multiple transaxial, coronal, and sagittal planes may all be useful for assessment of anatomy and for placement of subsequent cine acquisitions. Cine imaging, in which blood appears bright with flow-related grada-

Table 1. Comparison of cardiac imaging modalities

Merits	Limitations
Chest radiograph Inexpensive Well-suited for comparison Widely available	All structures in three dimensions are projected to the two-dimensional film with superimposition Ionizing radiation dose
Transthoracic echocardiography Portable, immediate, real-time imaging of structure and flow Safe, convenient, and acceptable for serial study Good for intracardiac structures, stenotic and regurgitant jet velocities, ventricular function, and flow-through proximal coarctation Doppler measurement of jet velocity and time-course of flow Estimation of right ventricular systolic pressure and pulmonary artery pressure Tissue Doppler can contribute information on myocardial function Suitable for stress studies Searches across volumes by real-time sweep of plane	Less good for great vessels and conduits Although in babies and small children acoustic windows are better, they tend to be more limited in larger patients and after surgery Poor views of anterior structures; subpulmonary ventricle and right ventricular outflow tract Operator-dependent
Transesophageal echocardiography Better acoustic access, especially to posterior cardiovascular structures Excellent visualization of intracardiac structures Useful intra-operatively Images parts of thoracic aorta clearly	Generally less good for vessels and conduits outside the heart Semi-invasive, sometimes uncomfortable, requiring sedation Operator-dependent
Cardiovascular magnetic resonance Unrestricted access; entire chest volume can be imaged by multislice acquisitions in any chosen orientation Good for extracardiac vessels Versatile; cross-cuts can hone in on a point of interest Good tissue differentiation using different sequences, with or without gadolinium contrast Functional information from cine imaging and flow velocity mapping Ventricular function, shunts, regurgitant flow volumes, and poststenotic jet velocities can be measured Quantitative assessment of ventricular mass	Expensive, time-consuming, less available, and (for a few patients) claustrophobic Not usually real time Less good for vegetations and valve structure Free choice of imaging parameters giving a certain tension between optimization and standardization Equipment- and operator-dependent Contraindications (eg, pacemakers)
Catheterization and angiography High-resolution images of coronary branches Accurate pulmonary artery pressure measurement and oxygen saturation measurement Transcatheter intervention Projected views of ventricular function and cardiovascular connections	Noninvasive approaches to cardiovascular function and flow can be more informative Invasiveness and radiation limits repeated use
Computed tomography Very high resolution, tomographic imaging Visualization of larger coronary branches and collateral arteries, pulmonary parenchyma, and cardiovascular calcification	Ionizing radiation limits repeated use Static images

tions, depicts movements of the myocardium, valves and flowing blood.

Magnetic resonance imaging (MRI) depends on interaction at tissue level between radio signals passed into the body and the oscillations of hydrogen nuclei in watery and fatty tissues. Control of this resonant interaction by means of applied magnetic field gradients is the key to the versatility of MRI. Protons are effectively played, being energized by pulses of radio signal and tuned and re-tuned in three-dimensional (3D) space by magnetic gradients. A

repertoire of different pulse/gradient sequences allows a variety of image appearances and flow measurements to be achieved, usually without contrast agent [6–8]. Exact sequences available depend on software packages provided by manufacturers, and these continue to be refined and upgraded. For cine imaging, an important development in the past 5 years has been implementation of steady-state free precession (SSFP) imaging, which has been given different names by the three main manufacturers of cardiac-dedicated systems. An advantage of SSFP cine imaging is

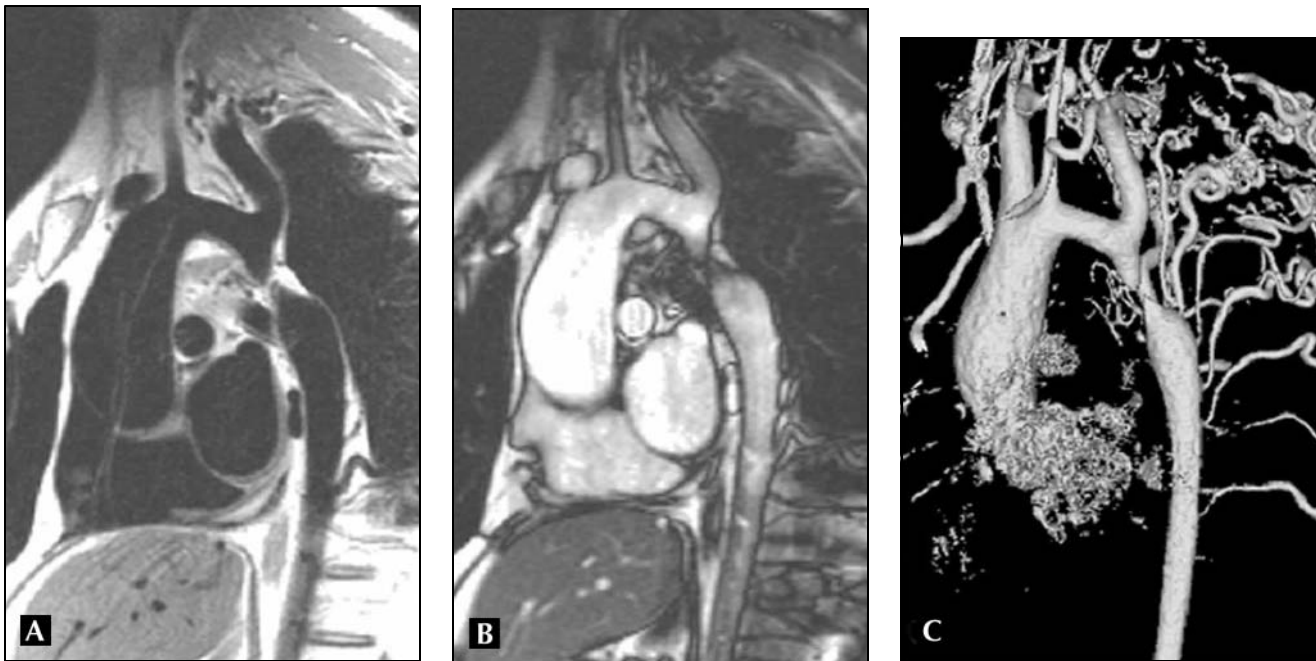


Figure 1. Unoperated aortic coarctation imaged by three different magnetic resonance techniques. The turbo spin echocardiogram image (A) gives no blood signal and differentiated signal from other types of tissue. The steady-state free precession cine image (B) gives bright blood with visualization of flow effects when viewed in cine mode. The three-dimensional (3D) gadolinium contrast angiogram (C) shows the lumen of the aorta and collateral arteries. The 3D dataset can be rotated and viewed from any angle. Although there was no visible orifice through the coarctation on 3D angiography, this patient was successfully treated by transcatheter balloon dilatation and stenting, the size and potential suitability of the stent having been determined from these images. (Courtesy of Drs. R. Mohiaddin and M. Mullen).

the contrast it provides between bright blood and darker myocardium when studying ventricular function. It is also effective at visualizing flow, with clear depiction and if accurately located, of the shear layer that delineates a jet. MR velocity mapping for measurements of jet velocity and volume flow is available on the cardiac dedicated systems [9,10]. It can be accurate and very useful if appropriately implemented, but this requires specific expertise.

Three-dimensional MR angiography (MRA), after venous injection of gadolinium contrast agent, can provide clear views of pulmonary, systemic and collateral artery branches (Fig. 1) [11]. Gadolinium is an inert substance and only rarely causes side effects of nausea or vomiting. Cardiac-gated 3D vascular imaging is also possible without contrast, for example using a 3D version of SSFP imaging. This has been found useful for assessment of anomalous pulmonary veins and anomalous coronary arteries [12].

Used appropriately, CMR can answer a range of functional and anatomic questions, including the location and severity of stenosis (*eg*, aortic coarctation or pulmonary artery stenosis), severity of regurgitation (*eg*, pulmonary) [13], the size and function of heart chambers (the right and left ventricle [RV and LV]) [14,15], and measurement of shunt flow [16]. CMR can provide remarkably comprehensive data, often avoiding the need for diagnostic catheterization, which can then be reserved for visualization of coronary artery branches, measurement of pulmonary artery pressure and resistance, and for patients with pace-

makers. A noninvasive CMR study can be used to plan and direct the transcatheter procedure, shortening diagnostic and therapeutic maneuvers, and minimizing the exposure of both patient and operator to radiation.

Specific Indications for Cardiovascular Magnetic Resonance Imaging in Congenital Heart Disease

Complex congenital heart disease

There are myriad permutations of CHD, and diagnosis may still require clarification in adults. CMR is valuable in this context, illuminating the diagnosis in a patient who has been inconsistently or wrongly diagnosed.

Imaging Extracardiac Disease

Coarctation and aortic disease

The geometry of the aorta is variable in adults with aortic coarctation, especially after different types of repair. CMR can be used to outline the arch anatomy and quantify the severity of native coarctation or re-coarctation (Fig. 1). By MR velocity mapping, a peak velocity of 3 m/sec or more is significant, particularly if associated with diastolic prolongation of forward flow (diastolic "tail"). CMR can visualize flow in the lumen of the aorta, as well as differentiate tissues around it, and is therefore effective for diagnosis of both restenoses and aneurysms,

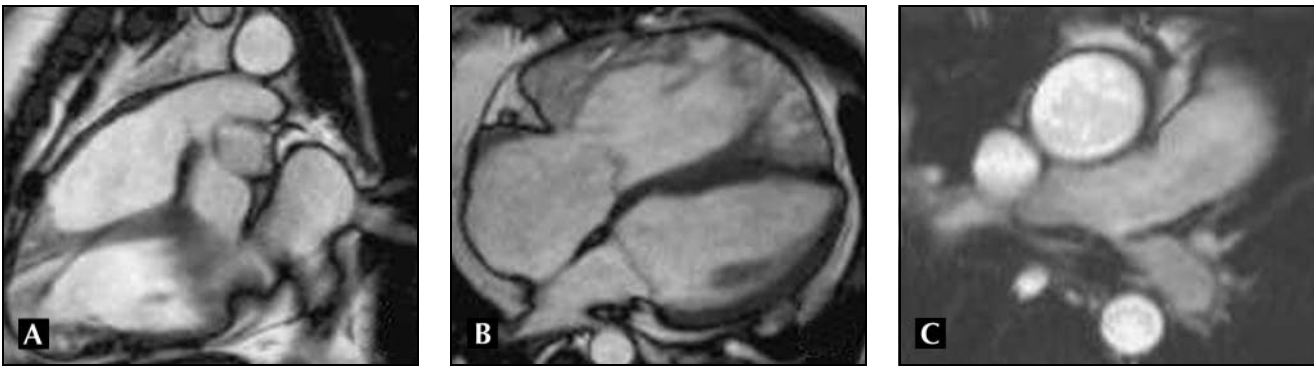


Figure 2. The static image taken from a cine (A) shows the right ventricular outflow tract. There is remnant pulmonary valve only. The four-chamber cine image at end diastole (B) shows function of the left ventricle and the muscular part of the right ventricle. An oblique transaxial slice (C) shows the pulmonary artery bifurcation with acute angulation and stenosis at the origin of the left pulmonary artery.

either of which may complicate repair of coarctation. True or false aneurysms may also complicate balloon interventions. We advocate at least a baseline assessment with CMR in all patients with previous coarctation repair. Patients repaired with Dacron (DuPont, Wilmington, DE) patches, or those with residual hemodynamic lesions (small aneurysm or mild re-coarctation) should be followed up periodically by CMR.

Poststenotic dilatation is common, appearing as fusiform dilatation beyond a stenosed or previously stenosed region, usually distinguishable by its location and smooth contours from more sinister aneurysmal dilatation that may require reoperation or protection with a covered stent. Leakage of blood through a false aneurysm can lead to hemoptysis. In such cases, para-aortic hematoma is generally well visualized by CMR, appearing bright, usually with diffuse edges. Postoperative hematoma is common, however, and sometimes leaves a region of signal adjacent to the aorta, which may only be distinguished from a developing false aneurysm if comparison of images over time is possible. This is an additional factor in support of acquiring baseline postoperative images in adults who undergo repeat surgery for coarctation.

Anomalous vascular branches and collaterals

Magnetic resonance 3D angiography with gadolinium contrast agent can be used to image anomalous pulmonary venous drainage or collateral arteries from the aorta to the lungs [17], or back into to the descending aorta in aortic coarctation (Fig. 1). This may obviate the need for diagnostic catheterization, for example, in patients with tetralogy of Fallot and pulmonary atresia, or expedite a directed percutaneous transcatheter intervention. The degree of collateralization associated with coarctation of the aorta can be visualized, and the 3D angiogram aids the interventionalist in planning the procedure, including selection of an appropriate stent.

Measurement of Right Ventricular Function Repaired tetralogy of Fallot

Magnetic resonance follow-up of repaired tetralogy of Fallot should include measurements of RV and LV volumes, ejection fraction and myocardial mass, cine visualization of the RV outflow tract (RVOT) and left and right pulmonary arteries, measurement of any aortic root dilatation, and measurement of pulmonary regurgitant fraction by through-plane velocity mapping (Fig. 2) [13,18–20,21•]. Interestingly, free (severe) pulmonary regurgitation is only about 40% (diastolic reversed flow expressed as a percentage of forward flow), but may be worsened by the presence of pulmonary artery branch stenosis.

Subpulmonary stenosis, double-chamber right ventricle

The RVOT can be clearly imaged by CMR. Subpulmonary stenosis can be documented and quantified as well as pulmonary regurgitation. An important variant of subpulmonary stenosis, well demonstrated by CMR, is double-chambered RV, where RV obstruction is subinfundibular, within the body of the RV, due to a hypertrophied muscular ridge and fibromuscular bands beneath a nonhypertrophied infundibulum and nonstenotic pulmonary valve [22].

Systemic right ventricle

If the RV is functioning as a systemic ventricle, as in congenitally corrected transposition of the great arteries or after atrial switch procedures, CMR provides a safe means for serial assessment [23].

Ebstein's anomaly

In Ebstein's anomaly, CMR is useful for assessment of suitability for surgery. It provides unrestricted views of the atria and ventricles, the location and function of the displaced tricuspid valve, the size of the functional RV and the adequacy of the pulmonary arteries.

Monitoring of Operated Congenital Heart Disease

Right ventricle to pulmonary artery conduits

Conduits used in the treatment of pulmonary atresia, and in the Rastelli operation for transposition of the great arteries, are rarely visualized adequately by ultrasound in adults, but they can be seen well by CMR. The patency of the conduit and pulmonary arteries, and the severity of any stenosis or regurgitation can be assessed [13,24].

Mustard operation for transposition of the great arteries

Most currently surviving adults born with simple transposition of the great arteries will have had surgery at atrial level during childhood, with removal of the atrial septum and insertion of a baffle (Mustard operation), or infolding of the atrial wall (Senning operation) to redirect blood, leaving the transposed ventriculoarterial connections. Imaging of patients after a Mustard or Senning operation requires assessment of the pulmonary venous flow path (from pulmonary veins to the tricuspid valve) and of superior and inferior vena caval limbs of the systemic venous flow paths, converging on the mitral valve to the left of the baffle [24]. As the hypertrophied RV is delivering systemic pressure in these patients, it is important to assess its function by cine imaging and volume measurements, and to assess any tricuspid regurgitation.

Fontan operations for functionally single ventricle

The Fontan operation aims to eliminate shunting in patients born with only one effective ventricle, routing systemic venous return to the pulmonary arteries without passage through a second ventricle, creating a serial circuit. Pressure must then be elevated in the systemic veins to maintain flow through the lungs, and obstruction of the cavopulmonary flow path can raise systemic venous pressure to an unsustainable level. Fontan connection originally incorporated the right atrium, connected to the pulmonary arteries. Total cavopulmonary connection, with the superior and inferior vena cava connected or channeled directly to the pulmonary arteries, has become more widely used in the past decade. Whichever variant, it is crucial that cavopulmonary flow paths remain unobstructed, and CMR imaging and velocity mapping can be used to identify stenosis, typically at a suture line. Contrast-enhanced 3D angiography is an alternative approach. Thrombus should be looked for in the right atrium and pulmonary arteries, and it is important to assess contractile function of the ventricle, competence of its inflow valve, and patency of the outflow tract.

Assessment of Shunt

Patent ductus arteriosus, and atrial and ventricular septal defects

Shunting can be assessed by measuring the difference between pulmonary trunk flow and aortic flow [16].

Patent ductus arteriosus is identifiable by CMR if sought. Flow through a patent ductus arteriosus is usually directed anteriorly into the top of the pulmonary artery close to the pulmonary artery bifurcation, and is detectable on cine images. Ascending aortic flow will be greater than pulmonary artery flow if duct flow is from the aorta to the pulmonary artery. Although atrial and ventricular septal defects are generally assessed satisfactorily by echocardiography, CMR offers unrestricted access in awkward cases, and enables measurement of shunt flow.

Ischemic heart disease, cardiomyopathies, and intracardiac tumors

Cardiovascular MR is excellent for assessment of regional and global LV function, and for characterization of myocardial tissue following infarction, or in the presence of tumors. Immediately after intravenous injection of gadolinium, perfusion of the tissues can be assessed [25]. A few minutes after injection of gadolinium, lingering of contrast in extracellular fluid allows even small regions of infarcted tissue to be distinguished from viable myocardium [26]. Techniques for coronary angiography by CMR are improving. Although they cannot yet compete with angiography in terms of spatial resolution, they can be valuable in establishing the patency and anatomic distribution of more proximal branches [27].

Limitations

Availability

Cardiovascular MR is not yet widely available for clinical referral, and numbers of adequately trained and experienced staff are limited.

Versatility versus uniformity

The versatility of CMR is a great strength, but also a potential source of confusion. Different MRI systems, or different individuals using the same system, may use different approaches. Given so much choice, uniformity is not easy to maintain.

Expense

Cardiovascular MR is relatively complex and expensive. Variation in underlying anatomy and surgical procedure between patients means that decisions regarding selection of planes and sequences must be made during acquisition. This necessitates experience and skill, which is also true for transesophageal echocardiography. CMR is also time consuming in terms of analysis. Although more reliable than other methods for visualization and measurement of RV function and mass, accurate measurement poses challenges due to difficulty in delineating the base of the RV, extensive trabeculation of the apical region, and possible ambiguity regarding limits of the outflow tract when there is no effective pulmonary valve after repair of tetralogy of

Falot. Measurements are made through manual tracing, which is time consuming.

The cost of imaging and analysis, however, should be weighed against potential costs of inappropriate management, which might entail complicated repeat surgery, more extended hospitalization than necessary, and increased patient morbidity.

Difficulty in unstable or anesthetized patients

Neonates and infants cannot cooperate as required, and need to be anesthetized for CMR. This is possible, but requires MRI-compatible monitoring equipment and appropriate knowledge and experience on the part of the anesthetist. As echocardiography views in the younger patient are often very good, it is rare for a patient under 6 years of age to undergo CMR in our practice. Although CMR can visualize dissection of the aorta, it is likely that computed tomography with contrast will be more rapidly and freely available for the initial diagnosis in the acutely unwell, unstable patient.

Subsequent accessibility of cine and three-dimensional acquisitions

Cine and 3D acquisitions are extremely informative, and should be available for interactive viewing by the clinician responsible for clinical decision-making. Appropriate image storage, networking, and software may be needed for interactive image review. Companies providing dedicated CMR image processing software include Medis (Leiden, the Netherlands) [28•] and CMRtools (Imperial College London, UK) [29•].

Future Applications

Demand and funding for CMR will increase as its capabilities in the assessment of ischemic heart disease are developed and validated. This will lead to more widespread availability of systems that will also be very well suited for assessment of CHD. Acquisition and analysis are likely to become more rapid, automated, and comprehensive in the coming years; for example, more automated measurement of ventricular function from data acquired in a single breath-hold. Functional measurements during exercise may come into clinical use, as may assessments of perfusion of the lungs and LV and RV myocardium. Guidance by CMR of catheters and catheter-mounted interventional devices has already been tried on a research basis, and will come into routine clinical use, as methods and equipment are refined. Over radiographic approaches, MRI guidance has the advantages of freedom from ionizing radiation for both patient and operator, direct flow measurement, varied tissue characterization, and more comprehensive imaging with respect to 3D space.

Conclusions

Cardiovascular MR gives unrestricted access to structures throughout the chest, including the RV, great arteries, and surgical conduits, making important contributions to the diagnosis and follow-up of adults with CHD. In our practice, CMR is used for serial follow-up, if there is a change in symptoms, and in planning percutaneous transcatheter or surgical procedures. As it becomes more widely available and efficient, CMR is likely to contribute increasingly to the informed, lifelong management of patients with CHD.

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Medis (Leiden, The Netherlands) has been supplying software for the viewing and analysis of CMR for several years. It will run on a PC, but only after installation of software by the company.

29. • CMRtools website for image processing software. Accessible at <http://vip.doc.ic.ac.uk/cmrttools/index.php>. Accessed January 2003.

This software is still in development. It allows viewing of still and cine Digital Imaging and Communications in Medicine (DICOM) images on a PC, with ability to measure areas, volumes, and other parameters. An evaluation version, effective for a limited period, can be downloaded for free trial.

Measurements of RV and LV function and pulmonary regurgitant fraction in a large series of patients late after repair of tetralogy of Fallot.