

Imaging congenital heart disease in adults

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ABSTRACT. Transthoracic echocardiography is the first-line modality for cardiovascular imaging in adults with congenital heart disease (ACHD). The windows of access that are possible with transthoracic echocardiography are, however, rarely adequate for all regions of interest. The choice of further imaging depends on the clinical questions that remain to be addressed. The strengths of MRI include comprehensive access and coverage, providing imaging of all parts of the right ventricle, the pulmonary arteries, pulmonary veins and aorta. Cine images and velocity maps are acquired in specifically aligned planes, with stacks of cines or dynamic contrast angiography providing more comprehensive coverage. Tissues can be characterised if necessary, and MRI provides relatively accurate measurements of biventricular function and volume flow. These parameters are important in the assessment and follow-up of adults after repairs for tetralogy of Fallot or transposition of the great arteries and after Fontan operations. The superior spatial resolution and rapid acquisition of CT are invaluable in selected situations, including the visualisation of anomalous coronary or aortopulmonary collateral arteries, the assessment of luminal patency after stenting and imaging in patients with pacemakers. Ionising radiation is, however, a concern in younger patients who may need repeated investigation. Adults with relatively complex conditions should ideally be imaged in a specialist ACHD centre, where dedicated echocardiographic and cardiovascular MRI services are a necessary facility. General radiologists should be aware of the nature and pathophysiology of congenital heart disease, and should be alert for previously undiagnosed cases presenting in adulthood, including cases of atrial septal defect, aortic coarctation, patent ductus arteriosus, double-chambered right ventricle and congenitally corrected transposition.

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This article provides an overview of the role of imaging in the assessment of adults with congenital heart disease (ACHD). Congenital malformations of the heart affect at least 1% of newborn infants [1]. Without intervention, the prognosis for more complex disease forms is poor. However, advances in paediatric cardiology and cardiac surgery made over the past few decades have revolutionised patient management, and the majority of patients now survive into adulthood. This has led to new challenges as increasing numbers of congenital heart patients pass into the care of adult cardiac services. The need for expert knowledge to allow the appropriate investigation of highly variable anatomy and pathophysiology, and to manage this patient group, has led to the expansion of the cardiological subspecialty of adult congenital heart disease [2, 3]. Many ACHD patients will have undergone palliative or reparative surgery during early life. These procedures are rarely curative and life-long follow-up is generally required to optimise the quality and span of life. Many ACHD patients develop arrhythmias secondary to intrinsic conduction abnormalities, surgical interventions or residual haemodynamic lesions. It is important for imaging specialists to understand the palliative shunts, surgical repairs and reconstructive procedures used, some

of which entail techniques that have changed greatly over the years [4, 5].

Where should patients with ACHD be imaged?

For those ACHD patients who are likely to need further surgery or other interventions to treat arrhythmia or haemodynamic lesions, it is recommended that imaging is undertaken in the specialist centre where they are being followed up [6]. Examples include patients with re-stenosis or aneurysm formation after repair of aortic coarctation, patients with pulmonary regurgitation after repair of tetralogy of Fallot (ToF), those who have undergone surgery for transposition of the great arteries (TGA) or for a functionally single ventricle, and patients with a variety of other more unusual or complex conditions. Relevant experience is needed to acquire and interpret images in these patients, especially those acquired by echocardiography and cardiovascular MRI, and collaboration between the imaging specialists, cardiologists and surgeons in such centres contributes to improved understanding of the complex and variable conditions they have to deal with. Large imaging data sets that include acquisitions of various types are best reviewed interactively in the context of multidisciplinary discussion of clinical management.

Having said that, general radiologists and cardiac imaging specialists must be able to recognise and appropriately

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investigate previously undiagnosed congenital heart disease that may present in adulthood. At the very least, they must appreciate the nature and potential complications of the main types of congenital heart disease. Besides aortic coarctation as a possible cause of upper limb hypertension, clinicians should be alert to atrial septal defect (ASD) as a possible cause of a dilated right ventricle. Sinus venosus ASD, which may not be directly visible on routine echocardiography, is commonly associated with partial anomalous pulmonary venous drainage. Ventricular septal defect, double-chambered right ventricle (subinfundibular stenosis) and patent ductus arteriosus can all present in adulthood as an audible murmur, through the dilatation of a chamber or as entities on a chest radiograph. Congenitally corrected transposition (atrioventricular and ventriculo-arterial discordance) can also remain undiagnosed until adulthood, and it is important to be able to distinguish a morphologically right ventricle (RV) from a left ventricle (LV) on cross-sectional images.

Issues specific to congenital heart disease

Functional assessment of the right heart represents an important aspect of imaging in ACHD patients [7]. The free wall and apical regions of the RV are normally extensively trabeculated, with relatively little compact myocardium. The pathophysiology of a subpulmonary RV that is volume loaded by pulmonary or tricuspid regurgitation differs markedly from that of a systemic LV with an incompetent valve. This is not only because ventricular form and structure differ, but also because of the relative situations of the ventricles with respect to the high systemic and the low pulmonary resistances. The LV normally delivers most of the total circulatory work and is connected in series, via the high-resistance systemic and low-resistance pulmonary vasculature, with the less powerful RV. Free pulmonary regurgitation, with little or no effective valve action, may be tolerated without symptoms for decades, although right ventricular dysfunction, arrhythmia and premature death can result. Imaging, with measurements of RV function and pulmonary regurgitation, plays a key role in deciding the nature and timing of re-intervention in such patients.

Additional areas to be addressed by imaging in ACHD include the assessment of stenoses of both outflow tracts (the aorta and pulmonary arteries), the quantification of regurgitation and the identification and quantification of any intracardiac or extracardiac shunts. Flow measurement by phase-contrast MRI plays a particular role here, with velocities encoded through planes transecting the ascending aorta and/or main pulmonary artery. Such measurements can be subject to error, which can be minimised by avoiding unnecessarily high velocity encoding ranges and ensuring that the region of measurement is as close as possible to the magnet isocentre. If still problematic, correction by subtraction of a corresponding subsequent "flow" acquisition in a static phantom is recommended [8].

Adequate non-invasive imaging can avoid the need for diagnostic catheterisation in most ACHD patients, and can shorten the procedure time should invasive investigation or intervention be required. The costs of imaging need to be weighed against the potential costs of less-informed management.

In complex malformations, the cardiovascular interconnections should be established and described methodically, which is best done according to sequential segmental analysis [9, 10]. The structures of the atrial and ventricular cavities have typical left- and right-sided forms, as do the upper abdominal organs and the bronchi. "Left" and "right" are used in this context to indicate morphological characteristics rather than position. A patient may thus be said to have a "left-sided right ventricle", identified as morphologically right by virtue of its moderator band, additional coarse trabeculations arising from the septum and, in most cases, a myocardial infundibulum encircling its outflow tract.

Sequential segmental analysis begins by stating the atrial arrangement (*situs*), which can be "usual" (in Latin, atrial *situs solitus*) (i.e. a right atrium on the right and a left atrium on the left) or "inverted" (*situs inversus*), or there can be bilateral duplication of one type of atrium, known as right or left atrial "isomerism". Isomerism is generally associated with accompanying malformations. In almost all cases, the atrial arrangement corresponds to that of the bronchi, and usually to that of the abdominal organs. Thus, the visibility of a long pre-branch segment with passage inferior to the corresponding pulmonary artery of a "left" main bronchus, or of a shorter pre-branch segment with passage superior to the corresponding pulmonary artery of a "right" main bronchus, gives a useful indication of the atrial morphology on the same side, as do the two lobes of a "left" lung and the three lobes of a "right" lung.

Atrioventricular and ventriculo-arterial connections are described as concordant (e.g. right atrium (RA) to RV, or LV to aorta) or discordant (e.g. left atrium (LA) to RV). TGA is synonymous with discordant ventriculo-arterial connections, and the transposed outflow tracts tend to lie almost parallel to one another rather than curving across each other in the usual manner. It is also possible for both inflow or both outflow valves to connect predominantly with one ventricle (a double-inlet or a double-outlet ventricle) or for one of the atrioventricular connections to be absent, as in tricuspid atresia.

Overview of the main non-invasive imaging modalities

The different non-invasive modalities are, to a large extent, complementary (Table 1). More than one modality is likely to be needed to address all of the relevant clinical questions, particularly in more complex cases.

Chest radiography

Postero-anterior with or without lateral chest radiographs provide a valuable, inexpensive and inclusive single-image overview of the heart, mediastinum, pulmonary vessels, lung fields and thoracic skeleton, all of which are potentially relevant in ACHD.

Transthoracic echocardiography

Transthoracic echocardiography (TTE) is the first-line cardiovascular imaging modality in ACHD patients,

Table 1. The relative strengths and limitations of the three main imaging modalities used for adults with congenital heart disease (ACHD)

	Echocardiography	MRI	CT
Methodological strengths	Real time Portable Local velocity measurements M-mode traces and 3D Suitable for operating theatre and during exercise Free of ionising radiation	Versatile Unrestricted access Multislice cine coverage Biventricular function Volumetric flow measurement Tissue characterisation Dynamic angiography Free of ionising radiation	Excellent spatial resolution 3D visualisation and access Short acquisition time Calcium seen well Lumen generally seen within stents
Clinical strengths in ACHD	Applicable in almost all cases Strong for valve disease, jet velocities, septal defects and vegetations	Many applications, notably in aortic coarctation, Fallot's tetralogy, shunts, TGA and complex cases	Visualisation of coronary and collateral arteries Visualisation of conduits and stents Visualisation of associated lung pathology
Methodological limitations	Limited windows, angles and depth of access Not suitable for volumetric flow measurement	Image slices are relatively thick Acquisition is relatively time consuming and expensive Pacemakers are generally regarded as a contraindication	Ionising radiation No flow measurement Limited cine visualisation
Clinical weaknesses in ACHD	Cannot usually access all parts of the RV, pulmonary arteries, pulmonary veins or aorta	Cannot reliably exclude patent foramen ovale or a small ASD, detect mobile vegetations in endocarditis or measure tricuspid regurgitation jet velocity for RV pressure estimation	Relatively limited functional information. Unsuitable for repeated follow-up studies because of radiation

3D, three-dimensional; ASD, atrial septal defect; RV, right ventricle; TGA, transposition of the great arteries.

being particularly suitable for the real-time interrogation of relatively small, thin and mobile intracardiac structures, leaflets, septa, jets and infective vegetations, which may be less well seen on MRI. In the majority of patients, TTE allows the evaluation of cardiac anatomy (*i.e.* orientation and veno-atrial, atrioventricular and ventriculo-arterial connections), the morphology of cardiac structures, ventricular and valvular function, the presence of shunt lesions and haemodynamic function (*e.g.* severity of regurgitation and evaluation of shunts and velocities across obstructions). Nevertheless, the suboptimal penetration of ultrasound poses the chief limitation of TTE, especially in adults after cardiovascular surgery. Moreover, TTE does not offer MRI's repertoire of tissue-contrast options, with or without contrast agent, and lacks MRI's unrestricted fields of view and frame of spatial reference with respect to the axes of the patient's body. Doppler techniques measure only velocities directed to or from the transducer and not those across or through the sector of the image. Nevertheless, Doppler velocity recording has the advantage over MRI of being able to measure the velocities of narrow tricuspid or pulmonary regurgitant jets, which allows the estimation of pressure differences between RV and RA in systole or between the pulmonary artery (PA) and RV in diastole. This is rarely practicable by MRI velocity mapping, mainly owing to the size and shape of voxels relative to the limited coherent cores of such jets.

Transoesophageal echocardiography

Transoesophageal echocardiography (TEE) has the advantage of access to more posterior parts of the heart, particularly for three-dimensional (3D) visualisations of

the valves, and is suitable for intra-operative use. The main disadvantages are its relatively invasive nature, generally requiring sedation or anaesthesia, and its limited field of view. The ability of TEE to evaluate extracardiac anatomy is limited and the alignment of the Doppler beam with unusually orientated flow jets can be challenging. TEE suffers from many of the limitations of TTE and is generally less acceptable than MRI for serial follow-up investigation.

Cardiovascular MRI

This modality is not restricted by body size or poor acoustic windows and is versatile, offering a repertoire of velocity-mapping and tissue-contrast options, without any ionising radiation [11, 12]. The versatility presents challenges, however, and consistency of methods between studies or between centres should not be assumed. MRI is widely regarded as the gold standard for measurements of both right and left ventricular volumes, although these analyses take time and require rigorously consistent methods of acquisition and measurement that may be difficult to maintain in practice [13, 14]. Unlike Doppler ultrasound, MRI velocity mapping allows the measurement of flow through planes transecting a large vessel for the calculation of regurgitant or shunt flow volumes [15]. First-pass adenosine stress myocardial perfusion imaging can be performed by MRI using gadolinium contrast. The freedom from ionising radiation makes MRI an appealing choice for the relatively young patient group with congenital heart disease [16]. This includes patients with TGA treated by arterial switch, which entails re-implantation of the coronary arteries. Late gadolinium enhancement can give information on myocardial fibrosis

in both the RV and LV [17]. MRI offers robust spatial localisation with respect to the axes and dimensions of the patient's body.

Pacemakers and implantable defibrillators are currently regarded as contra-indications for cardiac MRI. Arrhythmias tend to degrade the quality of MRI images, cine images generally being reconstructed from electrocardiogram (ECG)-gated acquisition during a breath-hold.

Computed tomography

CT offers excellent spatial resolution and relatively unrestricted access in much shorter acquisition times than MRI [18, 19]. It is well suited for imaging the epicardial coronary arteries and their relations to adjacent structures or conduits [20]. ECG-gated cine CT allows measurements of biventricular size and function, albeit at a lower temporal resolution than MRI and subject to adequate opacification of each intraventricular blood volume [21, 22]. CT provides an alternative to MRI in patients with a pacemaker or implantable cardioverter-defibrillator (ICD). The main drawback of CT is exposure to ionising radiation and its associated risk of cancer [23–26]. This risk is dose, age and gender dependent, and makes repeat CT examinations or studies in young patients unattractive. Other drawbacks compared with MRI include less versatile tissue contrast and an inferior ability to evaluate cardiovascular physiology, although the 3D spatial resolution provided by CT is unrivalled.

Cardiac catheterisation

Although used less for diagnosis since improved non-invasive approaches have become available, catheterisation has roles in the calculation of pulmonary resistance, assessment of coronary artery disease and for percutaneous interventions using balloons, stents, occlusion devices or implantable valves, notably for placement of a valve in a pre-existing but regurgitant RV to PA conduit.

Specific categories of congenital heart disease

Aortic coarctation

The majority of surgical repairs and catheter interventions for aortic coarctation have satisfactory outcomes, but a mild degree of residual aortic narrowing is common and it is important to screen periodically for re-stenosis or aneurysm formation. Aneurysms may occur after attempted balloon dilatation, or after repairs by a prosthetic patch—an approach rarely, if ever, used now. Aortic coarctation may be associated with abnormal properties of the wall of the aorta. A functionally bicuspid aortic valve is present in at least 50% of cases, and is more prone to progressive stenosis and/or to regurgitation than a normal valve. Echocardiography from a suprasternal window can usually provide a Doppler trace of jet flow through the residual coarctation, enabling an assessment of the presence or absence of diastolic prolongation of forward flow (a “diastolic tail”), which can be a telling sign of significant obstruction [27]. However, two-dimensional

echo rarely provides full visualisation of the repaired region in adults. Because repeated imaging is likely to be needed through the course of a lifetime, MRI is probably the modality of choice for assessment every few years, or more frequently if there is specific cause for concern [28]. An exception is after deployment of a stent, when at least one contrast-CT study provides better assessment of the in-stent lumen and any possible aneurysm formation in the vicinity of the stent [29]. Berry aneurysms of the circle of Willis or other cerebral vessels occur in up to 10% of patients with coarctation that bears the risk of rupture [30]. Rupture of a cerebral aneurysm is associated with high mortality, and hence screening for cerebrovascular aneurysms by an additional MRI study may be advisable if symptoms develop.

Valve disease

Echocardiography remains the first line of investigation in cases of heart valve disease [31, 32], but MRI can contribute to the measurement of regurgitation and to the assessment of myocardial or other pathology associated with this disease. In patients with bicuspid aortic valve, MRI allows the assessment of any ascending aortic ectasia, aneurysm or dissection. Regurgitant or stenotic jets are visible on cine imaging, although their appearances depend on the jet size and characteristics and on the relative location and orientation of the imaging slice. The visible extent of a jet does not necessarily relate to the severity of a lesion. Planimetry of an orifice, or of the cross-section of the jet immediately downstream of the orifice, by cine MRI is feasible in some but not all cases, depending on the structure of the jet and the relative thickness and location of the imaging slice. MRI jet velocity mapping can contribute to the quantification of stenosis, but velocities may be underestimated if the slice is not optimally located or if the coherent core of the jet is too narrow or too fragmented to contain several whole voxels. An appropriately aligned and encoded in-plane jet velocity acquisition may be helpful in locating the region of highest velocity prior to through-plane velocity measurement. For each of these applications, it is important to bear in mind the voxel dimensions, in particular the slice thickness, relative to the shape and orientation of a jet, and the potential for velocity underestimation owing to partial volume averaging. Quantification of regurgitation of the inflow valves by MRI is feasible. In the case of a single-valve lesion, ventricular stroke volume difference is used. Alternatively, regurgitations can be quantified by subtracting the outflow volume, measured by mapping velocities through a plane transecting the relevant great artery, from the corresponding ventricular stroke volume. For identification of tethering, prolapse or failure of coaptation of all parts of the mitral (or tricuspid) leaflets, a contiguous stack of cine images aligned perpendicular to the central part of the line of coaptation is recommended [33, 34].

Marfan syndrome

Measurement of the aortic root, of the ascending aorta and its branches and of any aortic regurgitation are likely to be needed periodically for Marfan syndrome patients.

Hence, MRI is likely to be the modality of choice unless echo windows are considered to be adequate in a particular individual. The LV and mitral valve can be assessed by either echo or MRI. It is important to check for aortic dissection, which may be small and focal [35]. MRI of the lumbosacral spine may be used to identify dural ectasia [36].

Ebstein anomaly and tricuspid regurgitation

The chest radiograph gives an excellent overview of heart size. Marked dilatation of the RA and RV generally reflects severe tricuspid regurgitation in Ebstein anomaly. Echocardiography allows visualisation of the tricuspid valve and any regurgitation. In Ebstein anomaly, the septal leaflet, particularly its more inferior parts, is apically displaced with "atrialisation" of the basal part of the RV. In some severe cases, the tricuspid valve plane can be rotated to lie in an almost horizontal plane beneath a dilated RV outflow tract. Such malformation is even better

visualised by MRI, using stacks of transaxial and short axis cines, and four chamber cines and an oblique coronal (or RV vertical long axis) cine to show the displacement of the inferior insertion of the tricuspid valve (Figure 1). The severity of tricuspid regurgitation can be assessed using through-plane MRI velocity mapping, the velocity encoding range typically set at 250 cm s^{-1} , to depict the cross-section of the regurgitant stream through a plane transecting the jet immediately on the atrial side of the defect. A regurgitant jet cross-section, reflecting the regurgitant defect, of $6 \times 6 \text{ mm}$ or more can be regarded as severe. An ASD resulting from the distension and gaping of a patent foramen ovale is present in about 50% of adult Ebstein patients, and should be sought by echo or by an MRI atrial short-axis cine stack.

Repaired Tetralogy of Fallot

Important issues in the relatively large subgroup of patients who have undergone surgical repair of ToF are

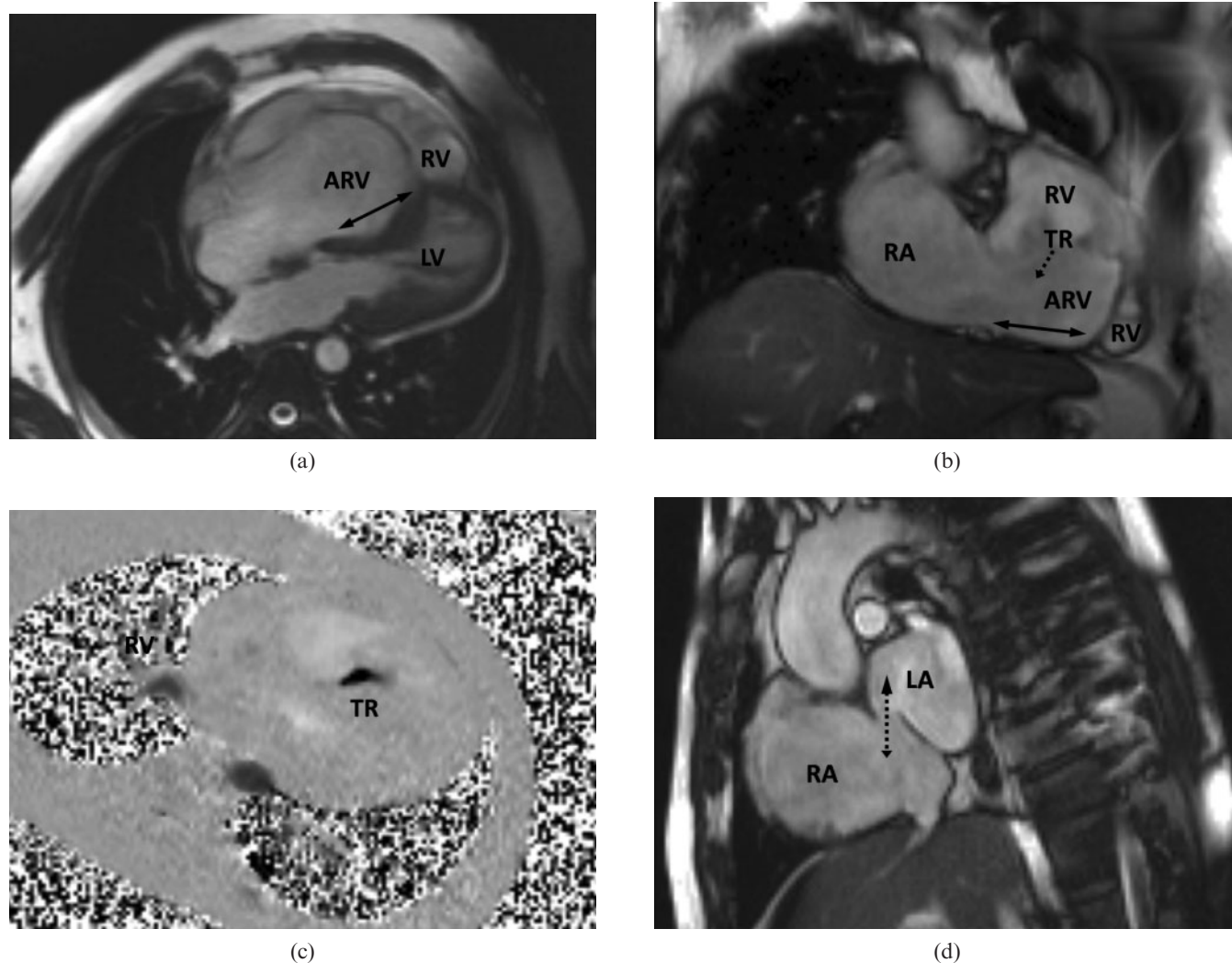


Figure 1. Features of Ebstein anomaly in a 43-year-old who has not had surgery shown by MRI. (a) Four chamber and (b) oblique coronal cines show the atrialised part of the right ventricle (ARV) relative its functional part (RV, right ventricle). The double-headed arrows indicate the extent of apical displacement of the septal and inferior insertions of the tricuspid valve. The dotted arrow indicates the direction of tricuspid regurgitation (TR). (c) Flow (dark) through a velocity mapping plane located to transect the jet (dimensions about $5 \times 14 \text{ mm}$) of severe TR. (d) An atrial short axis cine shows a widely patent foramen ovale with bidirectional flow, mainly from the right atrium (RA) to the left atrium (LA).

the size, regional function and global function of the RV, function of the pulmonary valve, and the patency of the right and left pulmonary arteries [37]. Free or almost free pulmonary regurgitation (PR) is common. Dysfunction of the LV, aortic dilatation, tricuspid regurgitation and a residual ventricular septal defect (VSD) patch leak are less common problems. Echocardiography can usually address most but rarely all of these, which gives an important role to MRI [38, 39]. The pathophysiology of PR differs from that of aortic regurgitation. Free PR, with little or no effective valve function, may be tolerated without symptoms for decades and is typically associated with a regurgitant fraction of ~35–45% [40]. It is often associated with a triphasic pattern of flow in the pulmonary trunk: systolic forward flow, early to mid-diastolic reversal of flow (regurgitation) and then late diastolic forward flow as atrial contraction boosts forward flow through a fully

expanded and therefore conduit-like RV (Figure 2). RV dysfunction, arrhythmia and premature death can result. In most centres, pulmonary valve replacement is considered in such patients, but the timing of surgery remains controversial, particularly if the patient is asymptomatic and bearing in mind that a homograft replacement may only function effectively for 15–20 years or less [41–45]. Once a conduit is in position, however, progressive stenosis or regurgitation may be treatable by percutaneous placement of a stented valve within the relatively rigid tube of the conduit [46, 47]. MRI, with contrast angiography, has a role in the selection of patients for such procedures, as has CT, which allows the visualisation of calcium and the clear delineation of the coronary arteries relative to a previously placed conduit.

Even in the absence of an effective pulmonary valve, the amount of regurgitation depends on factors upstream

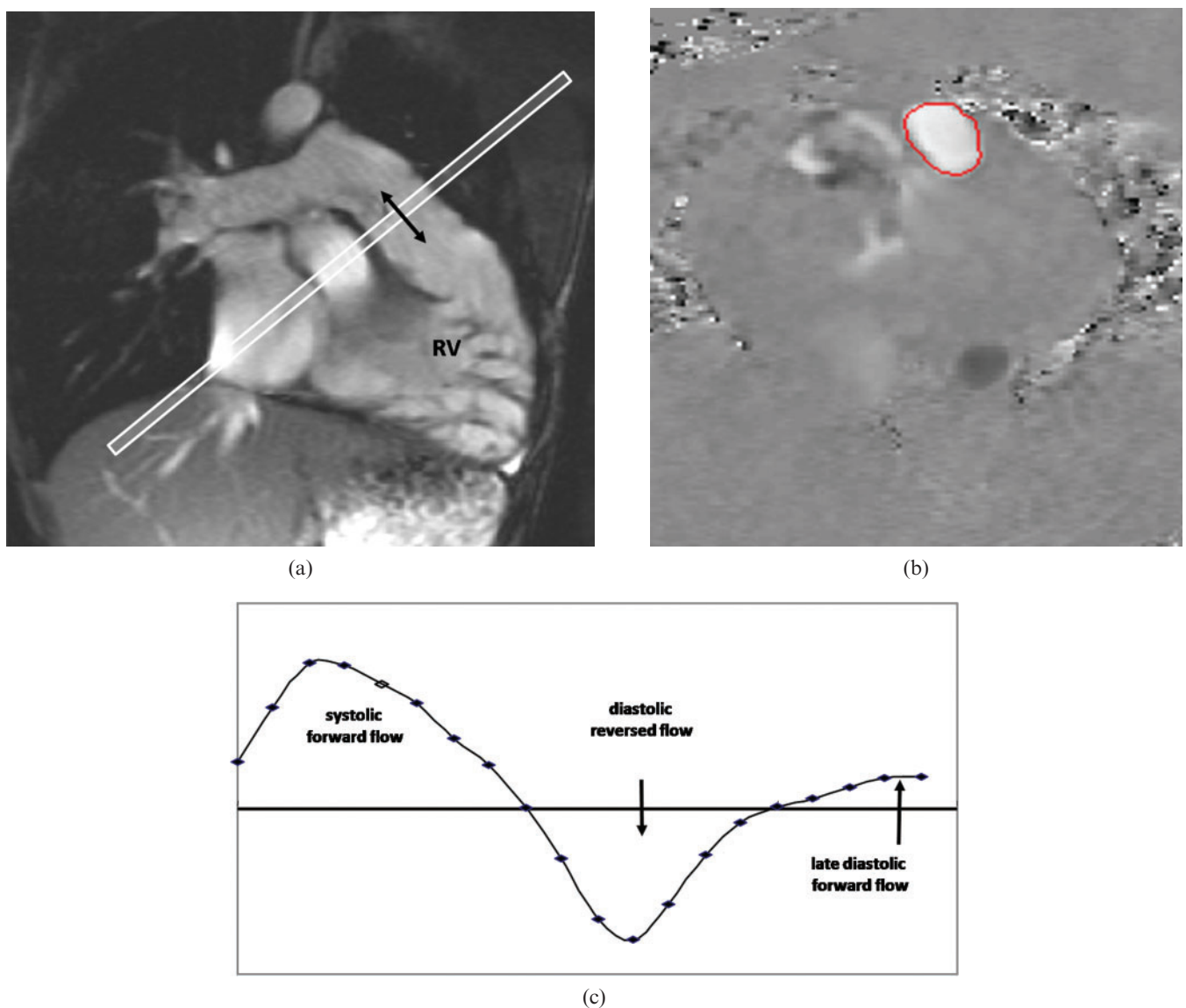


Figure 2. Pulmonary regurgitation measured by MRI through-plane velocity mapping in repaired tetralogy of Fallot. (a) Cine imaging aligned with the right ventricular outflow tract showed no effective pulmonary valve. Mapping of velocities through (b) a plane transecting the main pulmonary artery showed (c) systolic forward flow, diastolic reversed flow and late diastolic forward flow at the time of atrial systole. Absence of effective valve action typically results in a regurgitant fraction of ~35–45%, only occasionally exceeding 50%, for example if exacerbated by branch pulmonary artery stenosis or otherwise increased resistance distal to compliant arterial segments. RV, right ventricle.

and downstream. In occasional cases, the regurgitant fraction can exceed 50% [48]. This may be attributable to an unusually large and compliant RV, to a large and compliant pulmonary trunk and PA branches whose recoil contributes to the regurgitation [49], to branch PA stenosis or elevated peripheral resistance limiting the distal escape of flow or to combinations of these [50]. In summary, the evaluation of repaired ToF requires thorough assessment of the left and right heart, extending to the branch PAs and ascending aorta.

Double-chambered right ventricle or subinfundibular stenosis

This disease entity results from obstructing muscular bands or ridges between the hypertrophied body or sinus of the RV and the non-hypertrophied infundibulum. It is usually associated with a VSD into the higher pressure part of the RV, close to the upper part of the tricuspid valve, and may progress during adulthood. It should be possible to identify double-chambered RV echocardiographically, although limited visualisation may make it hard to distinguish between a jet through

a VSD, the subinfundibular stenosis and possible infundibular or pulmonary valve stenosis. MRI can help to differentiate between these, although flow through the VSD can be hard to detect. The subinfundibular origin of a DCRV jet, directed into the non-hypertrophied and non-obstructive infundibulum, is generally visible in routine basal short-axis cines (Figure 3) [51].

Major aortopulmonary collateral arteries

Contrast-enhanced 3D MRI or CT angiography is valuable for delineation of all sources of pulmonary blood supply prior to surgical or transcatheter procedures in patients with major aortopulmonary collateral arteries (MAPCAs) associated with severe pulmonary stenosis or atresia [52].

Pulmonary hypertension

Doppler echocardiography has the important advantage of being able to measure the velocities, and hence the presumed intercavity pressure difference, of relatively

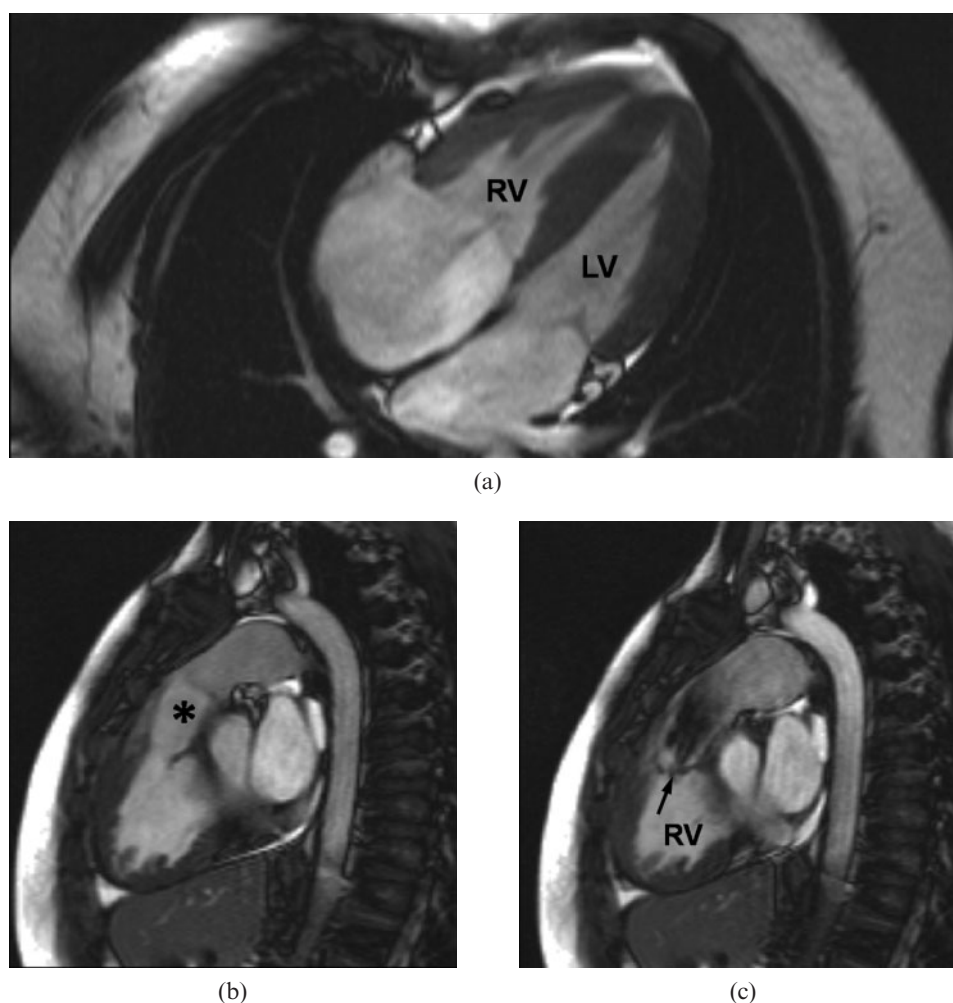


Figure 3. MRI features of a double-chambered right ventricle (RV), or subinfundibular stenosis, in a 16-year-old who has not had surgery. (a) The RV appears hypertrophied in a four-chamber view, whereas (b) the infundibular region (*) and the pulmonary valve above it are unobstructed. (c) The level of obstruction is seen in a systolic short axis image. The jet from the hypertrophied part of the RV to the infundibular cavity is arrowed. LV, Left ventricle.

narrow jets of tricuspid or pulmonary regurgitation. This cannot usually be done reliably by MRI. However, MRI does give good visualisation of RV size and function, of RV hypertrophy, of any flattening of the interventricular septum in systole and of the size and relative (lack of) expansion of the main and branch pulmonary arteries. MRI also allows measurement of aortic and pulmonary artery flow for calculation of indexed cardiac output and any shunting. It also enables good visualisation of any anomalies that might contribute to pulmonary hypertension, such as a patent ductus arteriosus (PDA) or VSD [53]. Contrast-enhanced angiography by CT or MRI may be used for the identification of thrombo-embolic disease or aortopulmonary collateral vessels [54]. The shorter acquisition time of CT is an advantage in patients with limited breath-holding ability.

Shunts: ASD, VSD or PDA

Echocardiography is the modality of choice for visualisation of ASD, VSD or PDA defects, except perhaps for PDA, for which MRI or CT are usually preferable in adults. MRI can also help to clarify the nature of the lesion, the amount of shunting [55], and biventricular size and function, and can detect associated anomalies, notably the possibility of anomalous pulmonary venous drainage [56–58]. Ascending aortic flow measured by MRI represents systemic flow (Q_s) when there is an intracardiac left-to-right shunt. However, when the shunt is from the descending aortic arch through a PDA, increased ascending aortic flow is expected that is equivalent to the amount of pulmonary flow (Q_p). The smaller amount of flow measured in the pulmonary trunk will be equivalent to Q_s [50].

Transposition of the great arteries post surgery

Specialist knowledge is needed for adequate imaging of patients who have TGA. There have been at least three distinct surgical approaches to TGA. Atrial switch operation (Mustard or Senning) was widely used until a decade or so ago. The reconstructed atrial pathways then need to be appropriately imaged, as does the function of the systemic RV [59]. In the past decade, correction by arterial switch has gained favour as it makes the LV the systemic ventricle. However, this procedure, which is usually performed in early infancy, involves re-implantation of the coronaries in the new aortic position and re-connection of the pulmonary trunk to what was an anteriorly located aortic root. This last manoeuvre results in a significant incidence of stenosis of the main, left and right pulmonary arteries, which is best assessed by MRI. A third type of operation for TGA is the Rastelli procedure, which is reserved for patients who also have pulmonary stenosis and a sufficiently large subarterial VSD. After Rastelli operation, assessment of possible stenosis or incompetence of the RV-to-PA conduit, of LV outflow tract (LVOT) flow, of biventricular function and of possible residual shunt are needed. These assessments are probably best achieved by MRI.

Congenitally corrected TGA

Congenitally corrected TGA involves discordant atrio-ventricular and discordant ventriculo-arterial connections (Figure 4). If uncomplicated, the affected patients can occasionally remain symptom free and undiagnosed into adulthood. Associated lesions include dextrocardia, a VSD and (sub)pulmonary stenosis. The subaortic RV is prone to dysfunction and regurgitation of its tricuspid valve [60]. Which ventricle is morphologically “right” and which is “left” can be determined by the presence of multiple coarse trabeculations, including the moderator band, arising from the RV but not the LV side of the septum. Each atrio-ventricular valve and ventriculo-arterial connection should be visualised by appropriately aligned views.

Fontan operations for functionally single ventricle

Fontan operations, generally performed in children with only one effective ventricle, result in a fundamental departure from normal circulatory dynamics [5]. The systemic and pulmonary vascular beds are surgically connected in series with one another downstream of the single effective ventricle, so eliminating shunting at the cost of a critically elevated systemic venous pressure that maintains flow through the lungs. Earlier procedures incorporated the RA between the caval veins and PAs, whereas total cavopulmonary connection, connecting inferior vena cava (IVC) flow to the PAs via a lateral tunnel or extracardiac conduit, has been favoured in recent years [5]. Patients who have had this surgery may become subject to arrhythmias, thrombosis, ascites, protein-losing enteropathy and other complications. Imaging by echocardiography and MRI needs specific expertise and is best undertaken in a specialist ACHD centre. Patency of the cavopulmonary pathways, of ventricular and valvular function, and of possible causes of shunting need to be investigated.

Complex congenital heart disease

MRI allows clarification of anatomy and function, including the presence of anomalous vessels, connections, shunts, stenoses, abdominal situs and possible polysplenia. Comprehensive cardiac, mediastinal and upper abdominal coverage using stacks of contiguous transaxial and coronal cines is recommended, as is dynamic contrast-enhanced MRI angiography. Cine images should also be aligned with each inflow and outflow valve, and with any shunt flow, so that connections can be established. Cardiovascular connections should be described according to sequential segmental analysis, as outlined above.

Coronary artery anomalies

Contrast-enhanced CT is recommended for coronary artery visualisation [61, 62]. The origin and proximal course of the coronary arteries can, in most cases, be visualised by MRI. Image quality depends, however, on meticulous technique, with fat suppression, diaphragm navigator respiratory gating and ECG gating, and suboptimal images are prone to misinterpretation.

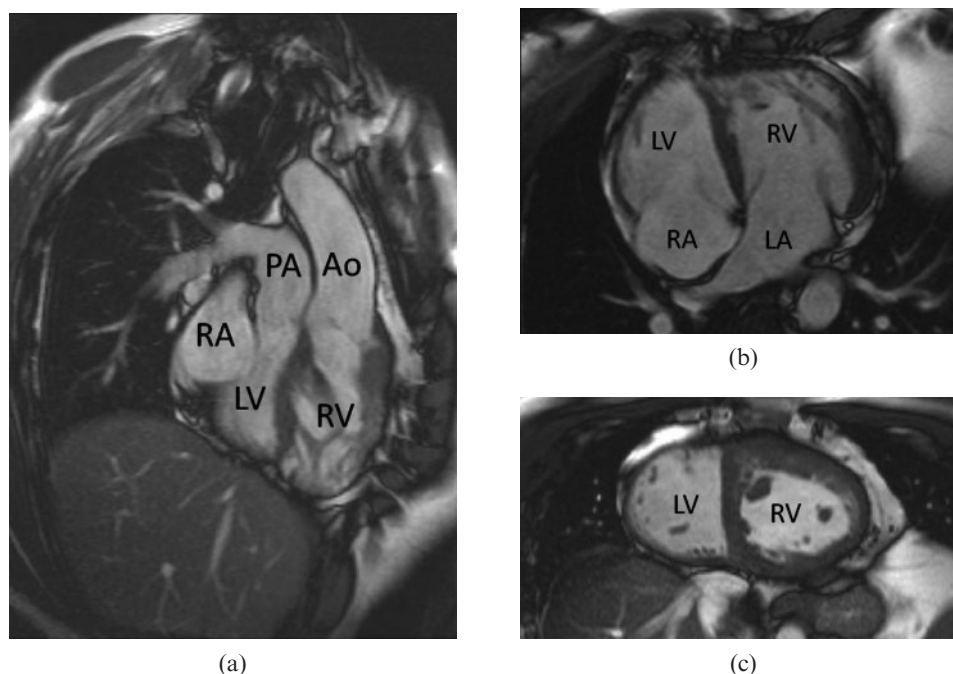


Figure 4. Unoperated “congenitally corrected” transposition of the great arteries shown by MRI. (a,b) Both the atrio-ventricular and the ventriculo-arterial connections are discordant. Note the expected apical displacement of the septal insertion of the tricuspid valve of the right ventricle (RV) relative to that of the mitral valve of the left ventricle (LV), and the hypertrophied muscle of the systemic RV. (c) In the mid short axis image, the left ventricular cavity can be identified as the one on the smoother, less trabeculated, side of the ventricular septum. PA, pulmonary artery; Ao, aorta.

Conclusions

After transthoracic echocardiography, which remains the first-line imaging modality in ACHD, the choice of further imaging depends on the clinical questions that remain to be addressed. Besides tissue characterisation, the strengths of MRI include comprehensive access and coverage, for example by the use of a stack of transaxial cine images or by dynamic contrast-enhanced angiography, and the relatively accurate measurements of biventricular function and volume flow. These are particularly useful in the assessment and follow-up of adults after repairs of ToF, aortic coarctation and TGA, and for imaging those who have undergone Fontan operations or with operated or unoperated complex malformations. In the author’s view, a dedicated MRI service should be regarded as a necessary facility within a centre specialising in the care of ACHD, with CT being preferable in selected situations. Adults with more complex conditions should ideally be both investigated and managed in a specialist ACHD centre. To realise their full potential and to avoid pitfalls, however, echocardiography, MRI and CT of ACHD require appropriate training and experience.

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