Imaging of congenital heart disease in adults: choice of modalities

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Received 4 April 2013; accepted after revision 11 June 2013; online publish-ahead-of-print 2 August 2013

Major advances in noninvasive imaging of adult congenital heart disease have been accomplished. These tools play now a key role in comprehensive diagnostic work-up, decision for intervention, evaluation for the suitability of specific therapeutic options, monitoring of interventions and regular follow-up. Besides echocardiography, magnetic resonance (CMR) and computed tomography (CT) have gained particular importance.

The choice of imaging modality has thus become a critical issue. This review summarizes strengths and limitations of the different imaging modalities and how they may be used in a complementary fashion. Echocardiography obviously remains the workhorse of imaging routinely used in all patients. However, in complex disease and after surgery echocardiography alone frequently remains insufficient. CMR is particularly useful in this setting and allows reproducible and accurate quantification of ventricular function and comprehensive assessment of cardiac anatomy, aorta, pulmonary arteries and venous return including complex flow measurements. CT is preferred when CMR is contraindicated, when superior spatial resolution is required or when “metallic” artefacts limit CMR imaging.

In conclusion, the use of currently available imaging modalities in adult congenital heart disease needs to be complementary. Echocardiography remains the basis tool, CMR and CT should be added considering specific open questions and the ability to answer them, availability and economic issues.

Keywords multimodality imaging • adult congenital heart disease • cardiovascular magnetic resonance • echocardiography • computed tomography

Introduction

As the majority of infants born with congenital heart disease (CHD) now survive into adulthood, lifelong follow-up at specialized centres is an integral part of their ongoing care. Adequate management of this group of adult congenital heart disease (ACHD) patients requires advanced imaging to assess the morphology and function of heart and vessels. In recent years, strategies for assessing the anatomy and physiology of CHD have evolved rapidly, with a shift from cardiac catheterization to non-invasive modalities such as echocardiography and more recently, cardiovascular magnetic resonance imaging (CMR) and cardiac computed tomography (CT).1 It is particularly important to determine the most appropriate and cost-effective diagnostic pathways for ACHD patients and the use of any imaging method has to be carefully considered.

Since the majority of cardiac defects are nowadays diagnosed in infancy and childhood, the adult cardiologist is mostly faced with patients known to have CHD. Nevertheless, the diagnosis may be incomplete or sometimes incorrect and has to be re-established in adulthood. Therefore, a comprehensive understanding of cardiac anatomy is required and particularly for describing complex malformations a structured segmental approach is recommended.2

Because of altered anatomy and physiology essential differences exist between paediatric patients with CHD and most adults with previously diagnosed or repaired congenital heart defects. In addition, different imaging modalities are available and measurements cannot be used interchangeable. For example, CT tends to overestimate and echocardiography to underestimate right ventricular (RV) volumes in comparison with the gold standard CMR.3

The aim of this paper was to describe the current armamentarium of imaging tools available to the ACHD cardiologist and to develop a structured approach to imaging in CHD. Specific emphasis is placed on the current role and future potential of advanced imaging modalities. Selected cardiac lesions will be addressed in more detail.

Strengths and limitations of currently available imaging modalities

An overview of strengths and limitations of available imaging modalities is presented in Table 1.

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### Table 1: Overview of strengths and limitations of available imaging modalities in ACHD

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<th>Modality</th>
<th>Strengths</th>
<th>Limitations</th>
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<tr>
<td>Echocardiography</td>
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<tr>
<td>General</td>
<td>First-line non-invasive imaging technique; high temporal resolution; real-time imaging capability; superior for intra-cardiac structures, hemodynamic assessment, and velocity measurement.</td>
<td>Highly user dependent; access to the heart limited by acoustic windows; extracardiac anatomy difficult to visualize.</td>
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<tr>
<td>2-D echo and Doppler</td>
<td>Superior in assessing valve disease and septal defects, estimating gradients, PA pressure, and detecting highly mobile structures such as vegetation.</td>
<td>Limited acoustic window to assess the hole RV and pulmonary arteries/veins, the aorta beyond its proximal ascending part and the arch.</td>
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<tr>
<td>TEE</td>
<td>Superior for imaging posterior cardiac structures, monitoring during interventional and surgical procedures; high sensitivity for the detection of thrombi and vegetations.</td>
<td>Usually requires sedation or—for monitoring of interventions—general anaesthesia; limited access to extracardiac structures.</td>
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<tr>
<td>Stress-echo</td>
<td>Feasible and safe in CHD, enables assessment of perfusion reserve.</td>
<td>Clinical significance in ACHD still unknown.</td>
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<td>Deformation imaging</td>
<td>Two methods: Doppler tissue imaging (DTI) and speckle-tracking echocardiography (STE), evolving technology to assess ventricular function in ACHD patients.</td>
<td>Vendor-specific solutions and therefore lack of industry standardization, DTI is angle dependent.</td>
</tr>
<tr>
<td>3D-TTE/TEE</td>
<td>New way to assess valve morphology and motion and to measure ventricular volumes.</td>
<td>Underestimates ventricular volume and function in comparison with the gold standard.</td>
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<tr>
<td>Intracardiac echo</td>
<td>May be helpful in guiding percutaneous interventional procedures, no need for sedation or oesophageal intubation.</td>
<td>Large shaft size, only monoplane image sections, no defined standard views, possible right heart catheterization complications, high costs.</td>
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<tr>
<td>Cardiac magnetic resonance imaging</td>
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<tr>
<td>General</td>
<td>Unrestricted access/views; coverage of the hole thorax; no ionizing radiation; superior for extracardiac structures, good temporal resolution, good assessment of flow; can be used in pregnancy when clinically indicated (some centres avoid imaging during the first trimester).</td>
<td>Sometimes limited tolerability due to claustrophobia, patient cooperation is required (breath-hold instructions); most implanted cardiac devices are not tested to be MR conditional; susceptibility to artefacts caused by metallic surgical material, loop recorder, coils, or stents can make measurement impossible; time consuming, expertise during scanning needed; expensive; availability may be limited.</td>
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<tr>
<td>SSFP acquisitions</td>
<td>Gold standard for the assessment of biventricular function and quantification of volumes (especially RV ejection fraction in Taf and systemic RV, evaluation of the RVOTO and RV-PA conduits, PAs, aorta, systemic and pulmonary veins; quantification of myocardial mass (LV and RV), highly reproducible measurements.</td>
<td>ECG-gated cine images and flow maps are typically acquired over a breath-hold and not in real time. Thin mobile structures may not be well seen (endocarditis).</td>
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<tr>
<td>Deformation imaging</td>
<td>Different techniques: e.g. Tagging-techniques and feature tracking.</td>
<td>Only a few studies in ACHD patients.</td>
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<tr>
<td>Angiography</td>
<td>Evaluation of pulmonary arteries and the aorta; evaluation of systemic and pulmonary veins (anomalous connection, obstruction, etc.).</td>
<td>Gadolinium-based contrast media rarely cause allergic reaction or nephrogenic systemic fibrosis in patients with severe renal dysfunction.</td>
</tr>
<tr>
<td>Flow (phase-contrast velocity mapping)</td>
<td>Measurements of flow volumes, quantification of pulmonary and aortic regurgitation, quantification of shunts.</td>
<td>Lower temporal resolution than for continuous-wave Doppler velocity measurement, dependent on a homogenous magnetic field; background offset error may occur.</td>
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<tr>
<td>LGE</td>
<td>Detection and quantification of focal myocardial fibrosis/scar; tissue characterization (fibrosis, fat, iron, etc.).</td>
<td>See angiography; no detection of diffuse myocardial fibrosis.</td>
</tr>
<tr>
<td>4-D velocity imaging</td>
<td>Visualization of multidirectional blood flow in a 3-D volume, pathline visualization may be helpful in selected ACHD patients.</td>
<td>Long-acquisition time; large resulting datasets need time consuming post-processing.</td>
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<tr>
<td>T₁ mapping</td>
<td>Quantitative imaging of extracellular volume fraction, detection of diffuse fibrosis.</td>
<td>Still no standardized technique and protocol, prognostic relevance currently unknown.</td>
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<tr>
<td>Computed tomography</td>
<td>Excellent spatial resolution; fast acquisition time; alternative to CMR in patients with pacemaker or implantable cardioverter defibrillator.</td>
<td>Ionizing radiation; inferior temporal resolution compared with CMR; fair tissue characterization and hemodynamic assessment, costly.</td>
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<thead>
<tr>
<th>Modality</th>
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<tr>
<td>ECG-gated cine CT</td>
<td>Ventricular size and function.</td>
<td>Lower temporal resolution than CMR.</td>
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</table>

Emerging techniques are in italic font.

EEG, electrocardiogram; TTE, transthoracic echocardiography; LV, left ventricle; RV, right ventricle; LGE, late gadolinium enhancement; ToF, tetralogy of Fallot; RVOTO, right ventricular outflow obstruction; SSFP, steady-state free precession; PA, pulmonary artery.

**Echocardiography**

Due to its relatively low cost, good safety profile, and wide availability, transthoracic echocardiography (TTE) remains the first-line tool for the regular assessment of ACHD. Two-dimensional TTE together with colour and spectral Doppler indeed allows the accurate and comprehensive assessment of cardiac morphology as well as function in most ACHD patients. This includes basic cardiac anatomy with orientation and position of the heart, venous return, connection of the atria and ventricles and origin of the great arteries, the morphology of the heart chambers, ventricular function and shunt connections, as well as morphology and function of heart valves. Doppler echocardiography provides information on the severity of obstructive lesions, RV/pulmonary artery (PA) pressures from tricuspid regurgitant velocity and allows to semiquantitatively assess valvular regurgitation and shunt lesions.

For certain indications such as evaluation of the aorta, pulmonary venous return, the interatrial septum, or left atrial appendage, transoesophageal echocardiography (TOE) is of particular advantage.

Besides the problems with image quality in patients after surgery or with abnormal position of the heart in the chest, echocardiography has some additional limitations: assessment of ventricular volumes and function remains unsatisfying in patients with systemic and non-systemic right ventricles and univentricular hearts particularly because of the unique geometry. Furthermore, Doppler gradients may sometimes be misleading particularly in RV outflow tract obstruction (RVOTO), coarctation, and stenosis occurring in series. Finally, the assessment of venous return and the great arteries often requires additional imaging modalities.

Real-time three-dimensional (3-D) echocardiography in patients with CHD is increasingly used and represents a sensitive tool to identify left ventricular (LV) and RV dysfunction in this patient cohort.

Evolving echocardiographic technologies such as angle-independent speckle-tracking echocardiography (STE) have been introduced lately in the evaluation of cardiac function in ACHD. Longitudinal ventricular dysfunction may precede systolic dysfunction as assessed by conventional parameters and thus, may be more sensitive in detecting myocardial impairment before it becomes evident with conventional imaging modalities. In fact, STE is increasingly used for the quantification of cardiac function in ACHD and has the potential to predict adverse clinical outcomes. However, the lack of standardization and differences between measurements obtained with different machines limits comparability between different studies and has so far hampered clinical uptake of the technique.

**Cardiac magnetic resonance imaging**

In the last decade, CMR has gained particular importance in the morphological and functional assessment of ACHD, particularly in patients with complex malformations. The unrestricted views, free choice of imaging planes, wide range of imaging sequences, and the lack of ionizing radiation make this technique ideal for the serial assessment of ACHD patients.

To extract optimal clinical information from the scan, however, a thorough understanding of the underlying anatomy and pathophysiology as well as associated or residual lesions prior to a CMR scan is required. In most of the cases, CMR imaging is tailored to address diagnostic questions that have been incompletely addressed by echocardiography.

For data acquisition and analysis, a standard approach should be applied to ensure that measurements are reproducible and accurate between patients. The European Society of Cardiology (ESC) recommends standardized CMR protocols for the use in ACHD patients as published recently.

The standard technique used for cine images is steady-state free precession (SSFP) imaging. It is well suited for imaging ventricular function and is the gold standard for the serial assessment of both right and left ventricular volumes and function.

Beside CMR tagging—as an established technique for measuring regional myocardial function—CMR myocardial feature tracking is a novel but promising method that detects myocardial borders and tracks myocardial motion at the voxel level from standard SSFP cine acquisitions. Recent studies, even in ACHD patients, showed good inter-observer reproducibility for measures of global left and RV strains, suggesting that the approach could have clinical relevance and deserves further study.

CMR imaging has evolved to provide increasingly precise functional analysis of flow dynamics mainly by the use of phase-contrast sequences. In the assessment of an incompetent valve by calculating regurgitant volume and fraction, velocity-encoded CMR is the gold standard. Shunt quantification based on pulmonary and systemic flow estimates can be performed by plane velocity-encoded imaging through the main PA and aorta.

Studies in different ACHD pathologies have shown the potential of four-dimensional (4D) phase-contrast velocity mapping of distorted...
blood flow patterns in the heart and adjacent large vessels, suggesting possible roles of fluid dynamic factors in the initiation or progression of pathology.\(^{37-39}\) The ability to measure multidirectional flows throughout a study volume has provided novel insights into cardiovascular blood flow patterns.

Contrast-enhanced MR angiography is particularly useful in the setting of ACHD. Visualization of anomalous pulmonary or systemic venous return or assessment and illustration of aortic pathology, such as coarctation and associated collaterals, can be performed.

Myocardial late gadolinium enhancement (LGE) detects myocardial scar and has been used to demonstrate fibrosis in different groups of ACHD. The finding of LGE is generally associated with worse functional (New York Heart Association) class, adverse ventricular mechanics, and history of arrhythmias. However, LGE provides only an incomplete estimate of the degree of fibrosis as it relies on a difference in signal intensity that may not exist in the case of interstitial myocardial fibrosis.\(^{40}\) CMR T\(_1\) mapping is a promising modality for the evaluation of diffuse myocardial fibrosis, and pilot studies in ACHD patients have already been conducted.\(^{41}\)

Computed tomography
Multidetector CT provides excellent spatial resolution and rapid acquisition time and therefore, plays an increasing role in the assessment of cardiac and pulmonary structures in ACHD patients.\(^{42-44}\) Besides its role in the assessment of the coronary artery anatomy, CT allows for the assessment of biomedical devices such as stents, valves, and sternal wires. CT scanning of the aorta is a quick and widely available imaging modality. Retrospectively gated cardiac CT angiography studies create the images needed for the assessment of ventricular function and volume. This provides a 4-D volumetric dataset for the analysis, which showed a good correlation but slight overestimation to CMR results.\(^{3}\) Tracheobronchial pathology is often associated in CHD, and can also be nicely evaluated by CT.

Recent developments aim to establish a role for CT in the functional imaging of the heart beyond the mere visualization of anatomy.\(^{45}\) Newer techniques to reduce the radiation dose, such as electrocardiogram (ECG)-controlled dose modulation, ECG-triggered sequential CT, low kV scanning, and interactive reconstruction, allow cardiac imaging at a much lower radiation dose.\(^{46,47}\)

these recent developments, adults with CHD often need repetitive imaging, exposing them to repeated ionizing radiation.

Chest X-ray
Chest X-ray is helpful for serial comparison of heart size, exclusion of cardiac malposition, and evaluation of pulmonary vascularity, thoracic skeleton, as well as peripheral lung fields.\(^{7}\) It has also been shown to provide prognostic information in ACHD patients.\(^{48}\)

In some centres, it is still routinely performed; however, other imaging modalities avoiding radiation exposure are increasingly used and have largely replaced chest radiograph at our and many other centres.

Cardiac catheterization
Invasive diagnostic studies should only be undertaken once other non-invasive imaging modalities have been used exhausted. It is then primarily required for hemodynamic assessment rather than imaging. Specific indications for invasive assessment remain the measurement of pulmonary vascular resistance, the evaluation of coronary arteries, and collateral vessels. Invasive assessment of systolic and end-diastolic ventricular function, pressure gradients, as well as shunts remains reserved for patients in whom non-invasive evaluation leaves uncertainty and the information is critical for clinical decision-making.

All the above-mentioned imaging techniques require special expertise in complex CHD. Imaging by CT and CMR scanning should be physician-supervised.

General recommendations for the choice between echocardiography, CMR, and CT when imaging ACHD in clinical practice
(Adapted from the ESC guidelines on the management of grown-up CHD\(^{1}\)). These recommendations are by and large-based expert opinion due to a lack of randomized and prospective data.
(1) Echocardiography is always the first modality to be used with the attempt to answer all questions required for patient management.

(2) CMR (CT) is indicated as an alternative to echocardiography, if both techniques can provide similar information but echocardiography cannot be obtained with sufficient quality to provide all information essential for patient management.

(3) CMR is indicated as a second method if echocardiography measurements of parameters critical for the clinical decisions (i.e. intervention) are borderline or ambiguous. These indications include

(i) Ventricular volumes and ejection fraction (particularly in regurgitant and shunt lesions).
(ii) Severity of regurgitant lesions.
(iii) Severity of shunt lesions.

(4) CMR or CT is indicated in settings where they are considered superior to echocardiography and should then be regularly used when the information is essential for patient management. For the choice between CMR and CT, several aspects must be considered including radiation exposure, resolution required, sum of information required, presence of pacemaker/ICD, or other implants. These indications include:

(i) Quantification of RV volumes and ejection fraction (tetralogy of Fallot [ToF], systemic RV, and tricuspid regurgitation) [CMR].
(ii) Evaluation of the RV outflow tract and RV-PA conduits [CMR and CT].
(iii) Quantification of pulmonary regurgitation (PR) [CMR].
(iv) Evaluation of pulmonary arteries (stenosis and aneurysms) and the aorta (aneurysm, dissection, and coarctation) [CMR and CT].
(v) Evaluation of systemic and pulmonary veins (anomalous connection, obstruction, etc.) [CMR and CT].
(vi) Collaterals and arteriovenous malformations (CT is superior to CMR).
(vii) Coronary anomalies and coronary artery disease (CT is superior to CMR).
(viii) Evaluation of intra- and extracardiac masses [CMR and CT].
(ix) Quantification of myocardial mass [CMR and CT].
(x) Detection and quantification of myocardial fibrosis/scar (gadolinium late enhancement) [CMR].
(xi) Tissue characterization (fibrosis, fat, iron etc.) [CMR].

Specific considerations in selected lesions generally encountered after repair during childhood

Tetralogy of Fallot

Despite excellent long-term survival and quality of life after repair in the current era, residua and sequela can cause long-term complications in patients with repaired ToF.49,50 Thus, lifelong regular follow-up after surgery is required. Chronic PR is a very common problem and may lead to progressive RV dilatation, increased RV wall stress, and RV dysfunction resulting in a substrate for potentially malignant arrhythmias, and sudden death. As a consequence of RV dysfunction and inter-ventricular interaction, LV dysfunction may also occur and represents an adverse prognostic feature in itself.1,51–53

TTE as the first-line diagnostic technique for follow-up provides comprehensive information on residual RVOTO, the presence and degree of PR, residual ventricular septal defect, RV and LV size, as well as systolic and diastolic functions. Two recent studies found a high degree of correlation between indexed apical RV diastolic area and indexed CMR end-diastolic volume.54,55 In addition, the degree of tricuspid regurgitation, RV pressure (RVP), aortic root size, and aortic regurgitation can be assessed by echocardiography.

Despite its prognostic importance, assessment of PR remains difficult. The treatment for PR is pulmonary valve replacement (PVR), but its optimal timing in asymptomatic patients, which is based on clinical, electrocardiographic, and volumetric measurements remain insufficiently defined.56,57 Although pre-PVR threshold values have been identified for RV dilation beyond that normalization of RV size is unlikely after PVR (Table 2), they do not provide ideal criteria for the timing of intervention. Normalization of RV size does not necessarily translate into normalization of RV function and—more importantly—improvement of exercise capacity indicating that intervention for PR may be required even earlier. In addition, dynamic changes of RV size and function may be more important than single measurements. Thus, regular follow-up is essential.

Some patients present with restrictive RV physiology resulting in less RV dilation despite severe pulmonary valve incompetence and better clinical tolerance of PR. Restrictive RV filling can be identified by the presence of end-diastolic forward flow in the PA during atrial contraction indicating diastolic RV stiffness.58–60 (Figure 2)

Myocardial deformation parameters were found to be useful to detect early myocardial dysfunction in TOF patients.61–63 Nevertheless, echocardiographic assessment of RV volumes and function as well as PR remain limited by the complex RV geometry, and CMR is superior in quantifying both.64 Thus, CMR plays a key role in the diagnostic work-up after repair of ToF and is recommended during the routine follow-up of these patients.21,65–68 It allows for highly reproducible quantification of RV volumes, especially if performed

<table>
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<tr>
<th>Table 2</th>
<th>Pre-PVR threshold values of RV end-diastolic volume (EDVi) and RV end-systolic volume index (ESVi) measured by CMR predicting volume normalization after PVR</th>
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<tbody>
<tr>
<td>N</td>
<td>EDVi (mL/m²)</td>
</tr>
<tr>
<td>Buechel et al.57</td>
<td>20</td>
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<tr>
<td>Therrien et al.56</td>
<td>17</td>
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<tr>
<td>Dave et al.58</td>
<td>39</td>
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<tr>
<td>Oosterhof et al.99</td>
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<tr>
<td>Geva et al.101</td>
<td>64</td>
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<tr>
<td>Lee et al.102</td>
<td>67</td>
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</tbody>
</table>

na, not available.
from datasets in axial orientation using manual blood-myocardial de-
lineation.69 A recent study by Wald et al.70 concluded that indexed PR
time rather than PR fraction may be a more sensitive and accurate
measure of RV volume load and thus, better describe the physio-
logic significance of PR. Using phase-contrast analysis of the main PA
flow or analysis of differential right and left ventricular stroke
volumes, PR can be quantified by providing regurgitant volume and
regurgitant fraction.

CMR should also routinely be used for the evaluation of RVOTO,
the pulmonary arteries to assess their size and shape and exclude
stenosis, the ascending aorta, and the position of great vessels or con-
ducts in relation to the sternum (essential when planning reoperation
with sternotomy). Although the rate of aortic growth seems to be
slow, mild or modest aortic dilatation is common in repaired TOF
patients.71

Finally, LGE allows the detection of fibrosis that has been reported
to be related to the risk of ventricular tachycardia and sudden cardiac
death in this patients.72

Cardiac CT provides information on the extent of conduit calcifi-
cation, postsurgical PA stenosis, coronary arteries, and lung paren-
chyma and has a specific role in the planning for percutaneous
pulmonary valve implantation to assess the degree of calcification
and the proximity of the coronary arteries.

Transposition of the great arteries

Transposition of the great arteries (TGA) is characterized by the
origin of the great arteries from the morphologically inappropriate
ventricles, while there is a normal connection between the atria
and the ventricles. Many current ACHD patients will have had surgi-
cal correction with an atrial switch procedure (Senning or Mustard
operation) to redirect the blood and provide ‘physiological’ repair.

In addition to arrhythmias, systolic dysfunction of the systemic
(subaortic) RV is the most frequent and relevant clinical problem.73
Quantifying RV function (particularly subclinical RV dysfunction)
is challenging; however. Different echocardiography parameters have
been proposed to assess RV function. These include visual qualitative
assessment (‘eyeballing’, preferred in many centres), measuring RV
fractional area change, tricuspid annular plane systolic excursion,
myocardial performance index, rate of systolic RVP increase (dp/dt)
measured across the tricuspid regurgitant jet, as well as more re-
cently strain and strain rate-based measures.6,74–76 In this patient
cohort, reduced biventricular longitudinal strain is related to
subpulmonary ventricular function, and relates to adverse clinical
outcome.12

However, despite these advances in echocardiographic technol-
ygy, CMR assessment of the systemic RV function and volumes
with multislice axial b-SSFP imaging remains the gold standard.19

Tricuspid regurgitation is primarily related to the deteriorating
systemic RV function. Its quantification is frequently difficult by echo-
cardiography and CMR using volumetric measurements may add in
this context.

The intra-atrial tunnels (baffles) can leak with either L–R or R–L
shunt, or can develop obstruction to systemic venous and/or pul-
monary venous drainage. Echocardiography (TTE and TOE) with
the use of Doppler techniques and contrast media may provide this
information, but the evaluation remains frequently incomplete. In
particular, obstruction of the superior vena cava (a frequent site of
baffle stenosis) is difficult to image in adults. CMR spin-echo sequences
in combination with magnetic resonance angiography provide excel-
lent visualization of systemic and pulmonary venous atrial pathways,77
although CT and invasive angiography deliver the highest resolution
(Figure 3).

In the past three decades, repair by the arterial switch operation
has become the method of choice providing not only ‘physiological’
but also ‘anatomical’ repair with the morphologic LV serving as sys-
temic ventricle and avoidance of the ‘surgical trauma’ at the atrial
level with all its consequences. The most common long-term pro-
blems are PA and PA branch stenosis (in part related to PA-stretching
after LeCompte manoeuvre), dilatation of the proximal neo-aorta
accompanied by aortic regurgitation, and less commonly coronary
artery pathology causing myocardial ischaemia.78,79 CMR is ideal
for imaging the aorta, branch pulmonary arteries, and RVOT, the
main sites of concern. Flow measurement can reveal a distributional
flow imbalance between the left and right lungs. Echocardiographic

**Figure 2:** Continuous-wave Doppler (A) and CMR flow (B) profiles in the main PA in a patient with repair of tetralogy of Fallot. Note late diastolic antegrade flow (arrows) as a possible marker for restrictive RV diastolic function.
evaluation focuses on LV global and regional function, evaluation of aortic root dimensions, as well as the presence and severity of AR. Supravalvular pulmonary stenosis (PS) and PA branch stenosis may be difficult to visualize and elevated RVP as indicated by tricuspid regurgitation velocity is then an important indirect sign guiding further evaluation.

CT is an additional adjunct that can be used for non-invasive imaging of coronary arteries, including the ostia, and in the case of suspicion of pulmonary branch stenosis as an alternative to CMR (Figure 4).

In the case of inconclusive CT or CMR imaging of the coronary arteries and the evaluation of RVOTO, cardiac catheterization is indicated. The third type of operation for TGA is the Rastelli procedure, in which the anatomical LV is connected through a VSD patch to the aorta and the RV is connected with a valved conduit to the PA in patients with a suitable VSD. Echocardiography is used among others to assess the function of the conduit between RV and the pulmonary trunk, but Doppler gradients may be unreliable. Therefore, RVP estimation from tricuspid regurgitation velocity is of particular

**Figure 3:** Assessment of intra-atrial tunnels in TGA repaired by Mustard operation. Angiogram (A) and CMR (B) demonstrate narrowing (arrows) of the superior limb of the systemic venous atrium (SVA). SVC, superior vena cava. Stenosis at the isthmus of the pulmonary venous atrium (PVA) between its posterior (PVAp) and anterior segment (PVAAa) revealed by TOE (C and D).

**Figure 4:** CT angiography of TGA after arterial switch and Lecompte-Maneuver. Normal dimension (A) and narrowing (B and C) of the left pulmonary arteries (LPA). Ao, aorta.
importance for the assessment of conduit stenosis. For morphologic assessment, CMR and CT are used.

**Coarctation of the aorta**

Aortic coarctation (CoA) is characterized by a narrowing of the aorta close to the area where the arterial duct inserts (distal to the origin of the left subclavian artery). The condition is considered to be part of a generalized arteriopathy and is commonly associated with other lesions of the aorta, such as bicuspid aortic valve, and hypoplasia of the arch. The majority of adult patients will have undergone surgical repair in childhood and some of them will suffer from aortic complications such as restenosis, aneurysm formation of the repair site, and dissection.81,82

With echocardiography CoA or re-coarctation can be sometimes visualized from a suprasternal view. While high systolic velocities are commonly found on continuous-wave Doppler, this does not represent a reliable sign of significant obstruction. High Doppler velocities that continue throughout the cardiac cycle, resulting in a diastolic ‘run-off’ or ‘diastolic tail’ phenomenon indicate significant coarctation.83,84 (Figure 5)

Echo is not sufficient for the quantification and morphologic characterization of coarctation (and lacks the ability to reliably detect aneurysm formation) as well as re-coarctation, thus requiring additional diagnostic modalities such as CMR and CT, which are the preferred non-invasive techniques to evaluate the anatomy of the entire aorta.85 If possible all patient should have at least one assessment of the aortic anatomy—preferably using CMR—in adulthood followed by subsequent periodic assessments with intervals depending on the individual pathologic findings. Both methods directly detect residual stenosis, restenosis/re-coarctation, aneurysms or significance of collateral vessels, and CMR flow measurements allow for the detection of diastolic prolongation of forward flow—a concept similar to the diastolic run-off on echo.86 CT has the advantage of better resolution whereas CMR is superior by providing collateral flow measurements.87

Nevertheless, echocardiography remains essential in this patient group to assess associated lesions such bicuspid valve etc. and to assess the response of the LV (hypertrophy and function) to increased afterload due to hypertension with and without re-coarctation. CT is particularly helpful in evaluation of patient post-coarctation stenting. CMR multidirectional, 3-D cine velocity acquisitions may help to detect altered patterns of aortic flow, although the clinical significance is still unknown.88

**Univentricular heart and Fontan circulation**

The term univentricular heart encompasses a wide spectrum of conditions in which one dominant functional chamber and nearly always a second hypoplastic accessory ventricle exists.88 For many years now, the Fontan operation in various modifications has become the palliation procedure of choice with direct connection of the superior and inferior vena cava to the pulmonary arteries.

With echocardiography, the morphology of the dominant chamber and basic anatomy of the heart defect as well as the systolic function of the dominant ventricle, the morphology and function of the valves, as well as the presence of a restriction to pulmonary blood flow can generally be assessed. However, in most of the cases, complete evaluation including the venous and arterial anatomy requires CMR and/or CT in addition.

In patients after Fontan palliation, CMR or CT is used to evaluate the patency of cavo-pulmonary pathways (this may require simultaneous injection of contrast agent through the arm and leg veins) and in combination with cardiac catheterization to assess the shunt fenestration flow89 (Figure 6).

CMR is also the method of choice for quantification of ventricular function and relative distribution of blood flow in the left and right lungs. Comprehensive coverage using a contiguous stack of transaxial cines is recommended, this generally being suitable for the identification of any intra-atrial thrombus or stenosis of the cavo-pulmonary connections19 and helps to evaluate hepatic congestion. Myocardial fibrosis as identified by LGE is a common finding and is associated with systolic dysfunction and arrhythmias.90

Frequently, cardiac catheterization is necessary for treatment decision because of missing information on hemodynamic significance of stenosis, PAP, pulmonary vascular resistance, and collateral vessels.

**Figure 5:** (A) Coarctation of the aorta. Continuous-wave Doppler from a suprasternal approach with diastolic prolongation of forward flow (‘diastolic tail’). (B) 3-D reconstruction based on CT angiography shows a severe stenosis (arrow) with collaterals.
Specific considerations in selected lesions frequently encountered undiagnosed in adult life

Like most paediatric patients with CHD, adults with previously undiagnosed anomalies benefit from a sequential, segmental approach for the initial evaluation.2

Atrial septal defect

Atrial septal defect (ASD) is the most common CHD in adults and is often undiagnosed until adulthood, due to a lack of early symptoms and subtlety of physical findings. Depending on the location several types of ASDs can be distinguished and several associated lesions may exist. The secundum ASD is by far the most common type of ASDs, which is localized centrally in the intra-atrial septum. In the presence of unexplained RV volume overload, shunting at the atrial level should always be suspected. Echocardiography remains the key diagnostic technique, providing information of RV volume overload, size, and morphology of defect and exclusion of additional defects. Direct defect visualization should be attempted by TTE (short axis and subcostal view) and should be confirmed with Doppler echocardiography. In case of poor visualization by TTE (especially in the setting of sinus venous defects) or uncertain diagnosis TOE should be employed (Figure 7). TOE also allows the visualization of pulmonary venous return and can be used to rule out anomalous pulmonary venous connection. Three-dimensional TOE has been proposed for appreciating the defect’s shape prior to and during catheter interventional closure.91,92 In addition, good correlation between 3-D TOE measurements and balloon stretched ASD diameters has been reported.93 Nevertheless, 2-D TOE remains more important for rim assessment and balloon measurement is indispensable in clinical practice.

CMR can serve as an excellent supplement particularly for the assessment of RV volume overload in borderline cases and the confirmation of normal pulmonary venous connection.

Congenitally corrected transposition of the great arteries

Congenitally corrected transposition of the great arteries (ccTGA) is a rare condition characterized by atrioventricular (AV) and ventricular-arterial discordance. As a consequence, the morphological RV supports the systemic circulation. Associated lesions are common (e.g. VSD and PS); however, in isolated ccTGA patients are often asymptomatic until adulthood and may remain undiagnosed for a long time.

Deterioration of systemic ventricular function and systemic AV valve regurgitation (tricuspid insufficiency) are well-known late complications.62,94,95

Echocardiography is the mainstay of diagnosis, demonstrates the double discordance, and helps to identify associated anomalies, particularly systemic AV valve abnormalities (Ebstein-like or dysplastic) and regurgitation. (Figure 8)
CMR assessment of RV size and function is challenging due to the complex shape and the multiple thick trabeculations of the systemic right ventricle, and the use of a consistent and reproducible delineation of the cavity is important. CT helps to evaluate the complex mirror-image distribution of coronary arteries. In most of the cases, the morphologic right coronary artery arises from the left posterior sinus and gives rise to a circumflex artery, and the morphologic left main coronary artery arises from the right anterior sinus.

Summary
Non-invasive imaging modalities are widely used to assess ACHD patients, and the need for invasive assessment has been significantly reduced in the last years. The assessment of ACHD has to involve a variety of imaging modalities that should be used in a complementary fashion. Echocardiography remains the workhorse of imaging routinely used in all patients but in complex disease and after-surgery echocardiography is frequently insufficient. CMR is particularly useful in this setting and allows reproducible and accurate quantification of ventricular function and comprehensive assessment of cardiac anatomy, aorta, pulmonary arteries, and venous return including complex flow measurements. CT may be used when CMR is contraindicated, when superior spatial resolution is required or when “metallic” artefacts limit CMR imaging.

The challenge remains to appreciate the different advantages and disadvantages of cardiovascular imaging modalities and to determine the most appropriate and cost-effective diagnostic pathways for individual ACHD patients.

Conflict of interest: none declared.

Funding
None.

References


