

Imaging the adult with congenital heart disease: a multimodality imaging approach—position paper from the EACVI

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Reviewers: This document was reviewed by members of the 2016–2018 EACVI Scientific Documents Committee: Victoria Delgado, Kristina H. Haugaa, Patrizio Lancellotti, Frank Flachskampf, Nuno Cardim, Bernhard Gerber, Pier Giorgio Masci, Erwan Donal, Alessia Gimelli, Denisa Muraru, and Bernard Cosyns.

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Received 24 June 2018; editorial decision 27 June 2018; accepted 28 June 2018; online publish-ahead-of-print 6 August 2018

Advances in the diagnosis and management of congenital heart disease have led to a marked improvement in the survival of adult with congenital heart disease (ACHD) patients. However, ACHD patients are a heterogeneous population, with a large spectrum of anatomic substrates even within specific lesions. In addition, the nature of previous surgery and other intervention is highly variable rendering each patient unique and residual anatomic and haemodynamic abnormalities are very common. As the ACHD population continues to age, acquired heart disease will also require cardiac imaging assessment. It is increasingly recognized in ACHD community that the diagnostic utility of a multimodality cardiovascular approach is greater than the sum of individual tests. In ACHD patients, diagnostic information can be obtained using a variety of diagnostic tools. The aims of this document are to describe the role of each diagnostic modality in the care of ACHD patients and to provide guidelines for a multimodality approach. The goal should be to provide the most appropriate and cost-effective diagnostic pathway for each individual ACHD patient.

Keywords

ACHD • multimodality imaging • echocardiography • cardiovascular magnetic resonance imaging • cardiac computed tomography

Introduction

The concept of multimodality imaging in congenital heart disease (CHD) has recently gained traction within the congenital imaging

community. With the increasing realization that the diagnostic utility of a multimodality cardiovascular approach is greater than the sum of individual tests, it can be expected that the integration of this concept into clinical practice will accelerate in the next decade.^{1–4} The core

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of the concept is synthesis and integration of image data from two or more modalities by imagers who are clinically expert in the involved modalities. However, this integration is expected to be further enhanced by technologic advances in individual modalities (e.g. higher spatial and temporal resolutions, improved signal-to-noise ratio), better integration of image data from different sources (e.g. fusion imaging), training of a new generation of imagers with expertise in several key modalities, and employing a patient-centred individualized approach towards optimal utilization of potentially expensive technology. Recommendations for imaging approaches to surveillance of adult congenital heart disease (ACHD) patients highlight the potential of a multimodality approach to strike an optimal balance between utilization of tests and diagnostic yield.¹⁻⁷

Imaging in ACHD must be preceded by a thorough clinical evaluation including the patient's history, review of previous surgical procedure(s) or other intervention(s) and physical examination. Knowledge and understanding of congenital cardiac anatomy, terminology, pathophysiology, and surgical and/or interventional procedures are fundamental to optimal ACHD imaging and interpretation. Access to medical records, including operative reports is of the utmost importance. Data obtained by the different imaging modalities must be integrated with functional assessment, particularly cardiopulmonary exercise demonstrated strong prognostic information in ACHD patients.⁸

The choice of imaging modalities should be made by determining the best combination of tests with the least risk to the patient given a specific clinical question. In some cases, it should be influenced by the age of the patient. The best tests for an individual patient will vary, and be influenced by the availability of both advanced imaging technology and skilled personnel available locally.

Echocardiography remains the first imaging modality to use and in many instances will provide all information required for patient management. If echocardiography is of poor quality and cannot therefore provide sufficient information, cardiac magnetic resonance (CMR) will in general be the next choice for further evaluation. CMR is also being increasingly used when echocardiography is of good quality but provides borderline or ambiguous measurements of parameters that are essential for the decision-making (catheter intervention, cardiac surgery) such as right ventricular (RV) volumes and ejection fraction (EF) or quantification of valvular regurgitation and shunt lesions.

There are specific questions where CMR and/or computed tomography (CT) are superior to echocardiography or are the only modalities to provide the answers. If both are suitable, CMR is preferred over CT to avoid iodinated contrast with potential kidney toxicity and radiation given the potential carcinogenic effect in ACHD patients attributable to the cumulative use of ionizing radiation.^{9,10} In case of significant metallic artefacts or if particularly high resolution is desirable, for example, for coronary anomalies and arteriovenous malformations, then gated cardiac CT may be preferred. Examples of when CMR/CT imaging may be helpful are given in *Table 1*. Cardiac catheterization is mainly reserved for assessment of precise haemodynamics (e.g. pulmonary vascular resistance), evaluation of coronary arteries and extracardiac vessels such as aortopulmonary collaterals particularly when an intervention is contemplated, or when non-invasive data remain inconclusive. Nuclear imaging is generally limited to selected patients only, for example, for myocardial stress perfusion

Table 1 CMR/CT may be useful to answer the following

- Quantification of RV size and function (CMR)
- Evaluation of the RV outflow tract and RV-PA conduits (CMR and CT)
- Quantification of pulmonary regurgitation (CMR)
- Evaluation of pulmonary arteries and the aorta (CMR and CT)
- Evaluation of systemic and pulmonary veins (CMR and CT)
- Collaterals and arteriovenous malformations (CT superior to CMR)
- Coronary anomalies and coronary artery disease (CT superior to CMR)
- Quantification of myocardial mass (CMR and CT)
- Intra- and extra-cardiac masses (CMR and CT)
- Myocardial fibrosis/scar (CMR)
- Tissue characterization (fibrosis, fat, iron etc.) (CMR)

imaging or differential pulmonary blood flow quantification when CMR is not available.²

Thus, the choice of imaging modality in clinical practice is based on the different strengths and limitations of the modalities, their availability, and on economic and safety (radiation and biological) aspects. Because of the lack of randomized and prospective data, contemporary recommendations are informed by expert consensus rather than evidence-based.¹⁻⁵

Echocardiography

Echocardiography, with its wide range of approaches (transthoracic, transoesophageal, 3D, contrast, and stress), is generally considered the first-line modality for cardiovascular imaging in ACHD.²⁻⁵ It provides comprehensive assessment of anatomy and physiology and contributes significantly to clinical management many years after surgical or catheter interventional procedures. It is recommended to develop lesion-specific imaging protocols for follow-up studies, to ensure the critical information for clinical decision-making is included in the imaging study, and for longitudinal data comparison. For example, transthoracic echocardiography (TTE) in repaired tetralogy of Fallot (rTOF) patients, may include a more detailed assessment of RV size and function while a patient after repair of coarctation of the aorta may require a more detailed assessment of left ventricular (LV) size and function.

When performing and interpreting echocardiographic studies in ACHD, it is important to systematically assess cardiac anatomy, morphology, function, and haemodynamic relevance of all malformations. Sequential segmental analysis should be used for all ACHD cases to ensure that important pathology is not missed. Intracardiac shunt lesions can be demonstrated by 2D or 3D echocardiography complemented with colour and spectral Doppler measurements for full pathophysiological evaluation (size, direction, and velocities across defect).

3D echocardiography provides en-face visualizations of cardiac valves that are easily interpreted by surgeons to plan valve repair surgery. In addition, 3D echocardiography planimetry are useful for

assessing severity of valve stenosis in presence of multiple haemodynamic abnormalities, sizing valve prosthesis, and live dynamic geometric guidance during the interventional procedures [3D transoesophageal echocardiography (TOE)].¹¹

Measurements of LV volumes and EF by 3D echocardiography are preferred over 2D calculations where echocardiographic windows allow.⁹ In patients with compressed LV due to severe RV dilatation [such as rTOF with severe pulmonary regurgitation, Ebstein's anomaly with severe tricuspid regurgitation (TR) or pulmonary hypertension], non-geometric indices of longitudinal LV velocity measured by Tissue Doppler or strain and strain rate measured using speckle tracking are associated with outcome, in particular global longitudinal strain (GLS).¹² It is important to note that criteria used in the general adult cardiac population for the assessment of diastolic function, such as E/e' ratio, may not be accurate in all congenital patients as reduced e' can be the result of localized surgical scarring at the septum or LV free wall rather than representing impaired global LV diastolic function.¹³

Echocardiographic assessment of RV size and function is more difficult, in part because of the shape of the RV. Indirect measures of RV function such as fractional area change (FAC) and tricuspid annular plane systolic excursion (TAPSE) from a four-chamber view are widely used and have shown prognostic value. Tissue Doppler imaging of myocardial velocities and speckle tracking echocardiography (STE) for myocardial deformation have both been applied in ACHD for regional and global ventricular myocardial deformation assessment,^{14–18} but their clinical application to the RV in ACHD remains limited to specialized centres. In the presence of good windows, 3D evaluation of RV volumes and RV EF is recommended in laboratories with experience in 3D.^{11,12,17}

Suboptimal penetration of ultrasound poses the chief limitation of TTE, especially in adults after cardiovascular surgery. Standard views might be inadequate and atypical views are often required. In patients with limited echocardiographic windows, additional imaging techniques will be required and selected according to the clinical question. Contrast echocardiography is of help for visualization of intracardiac shunt, volume quantification, cardiac masses, and for myocardial perfusion.¹⁹ For further assessment of intracardiac anatomy and valve function, TOE is the next logical diagnostic imaging technique. TOE plays a crucial role in the perioperative assessment of patients with CHD and post-bypass TOE has become routine in most surgical centres treating patients with CHD.²⁰ TOE is also used for monitoring and guiding interventional cardiac catheterization, including device closure of septal defects.²⁰ Intracardiac echocardiography (ICE) has been used in certain centres as an alternative for TOE for guidance of specific interventions in recent years.²¹

Newly developed real-time fusion of dynamic synchronized live echocardiography and fluoroscopy is an emerging technique to facilitate ACHD percutaneous intervention procedures and reduce dosage of radiation and contrast.²² Such new echocardiographic techniques will play an increasing role.

Key points

- Echocardiography is the first line imaging modality in the assessment and follow-up of ACHD patients.

- 2D speckle tracking-derived LV GLS appears to be feasible and reproducible for clinical use and can be included in the clinical follow-up.
- 2D speckle tracking-derived strain of the RV free wall, appears to be feasible and reproducible.
- Standard approaches to diastolic assessment are not always applicable in all ACHD patients.
- TAPSE and FAC for RV function are the suggested echocardiographic parameters for regular clinical follow-up of ACHD patients.
- 3D echocardiography, when feasible, is recommended to assess ventricular volumes, EF, valvular morphology, and better understanding of intracardiac anatomy.
- For the follow-up of RV size and function, when CMR is not feasible, not available or contraindicated, in centres with experience, 3D echo is recommended.
- Dedicated protocols for each specific lesion are strongly recommended.
- Echocardiography studies in ACHD should be supervised and reported by appropriately trained ACHD specialists.

CMR imaging

CMR imaging is already routine and well established in the diagnosis and serial follow-up of ACHD patients.^{1–5,23,24} CMR enables life-long accurate volumetric, functional and flow analysis, which is crucial for optimal management and timing of interventions during follow-up. CMR is accurate with low intra- and inter-observer variability,²⁵ and has become the gold standard for volume and flow assessments in ACHD. Unlike some other imaging modalities, CMR image quality is not affected by body habitus, there is no need for geometric assumptions and it is radiation free.

Ventricular volumes are obtained from manual segmentation of end-systolic and end-diastolic cine images, generally acquired from a short axis stack but a transverse stack can also be used. Volumes from axial and short axis stacks are not equal, and axial approach may allow more accurate volume measurements.²⁶ Flows are assessed using phase contrast velocity encoded imaging. The main advantage of CMR-based flow analysis results from the possibility to interrogate the area of interest in any plane, which is of particular value in patients with RV to pulmonary artery (PA) conduits, which are often too anterior to be assessed with echocardiography. Flow can be assessed perpendicular (through plane) or parallel (in plane) to the flow jet, the latter being particularly useful in the presence of multiple level stenoses where information on the area of maximum flow acceleration is needed. CMR allows accurate quantification of valve regurgitation and is considered gold standard for pulmonary regurgitation assessment. Volumes and flow analysis allows non-invasive measurements of intra- and extra-cardiac shunts [ventricular septal defect (VSD), atrial septal defect (ASD), patent ductus arteriosus, baffle leak after Mustard and Senning procedure, etc.], which is of significant value for patient management. In patients with complex single ventricle circulations such as after Fontan palliation, flow analysis provides accurate non-invasive haemodynamic data, including collateral flow and shunt through a fenestration if present. 3D information

obtained from MR angiography and 3D SSFP images allow accurate assessment of cardiac structures and their anatomic relations. This is particularly useful when planning interventions such as percutaneous pulmonary valve implantation, correction of partial anomalous pulmonary venous drainage, or stenting of aortic coarctation. The obtained images can be used to produce 3D-printed heart models, which are becoming an established technique to plan selected surgical and catheter procedures.

In addition, CMR with late gadolinium enhancement (LGE) is the reference standard for the assessment of focal myocardial fibrosis, while T1 mapping allows quantification of interstitial fibrosis.

Presence of metallic implants or a pacemaker may represent a contra-indication to CMR and careful safety check should be performed prior and after scanning. Guidelines already exist regarding the use of CMR in patients with conventional pacemakers and AICD devices^{27,28}; increasingly 'CMR-compatible' devices are being selected for ACHD patients. Occasionally, severe claustrophobia makes it impossible for an ACHD patient to undergo routine awake CMR, so sedation can be considered if CMR imaging would change the clinical management. Finally, CMR can safely be performed during pregnancy although it is general recommendation to avoid scanning during the first trimester and this should be included in informed consent process.^{28–30} Expert centres also advise against breast feeding for 24 h after receiving gadolinium contrast.

In summary, CMR is an important tool in the multimodality assessment of ACHD patients. It provides a detailed structural diagnosis, an accurate volumetric and flow analysis, and as radiation free is safely used for longitudinal follow-up.

Key points

- A baseline CMR is recommended for many patients at time of transition from Paediatric to ACHD programmes.
- CMR is the gold standard for ventricular volumes, EF, flow quantification, and the assessment of extracardiac anatomy.
- CMR frequency should be determined by the underlying defect and clinical status of the individual patient. Intervals between scans depend on the risk profile, findings at the first CMR study, and the expected rate of change. Intervals of 3 years or more are appropriate in most cases, but earlier re-study may be prompted by the onset or progression of symptoms, or the presence of a lesion liable to rapid progression.
- Additionally, CMR is recommended in the presence of clinical deterioration, non-diagnostic echo findings, and prior to surgical or transcatheter intervention.
- CMR studies in ACHD should be supervised and reported by appropriately trained ACHD specialists.

Cardiac CT

In recent years, the use of Multidetector CT (MDCT) in ACHD has increased exponentially. This is because modern MDCT can cover large anatomic volumes with excellent spatial resolution allowing detailed evaluation of small blood vessels such as coronary arteries,

pulmonary veins, collaterals, arteriovenous malformations and distal PA branches.^{31–33} Acquisition time is rapid (<2 min) allowing patients who are unable to lie still or supine for long periods to be imaged. Pulmonary parenchymal imaging is also provided which is highly relevant for patients with pulmonary hypertension. CT is a true 3D technique, allowing more accurate measurement in all planes of dimensions and integration with cathlab data for procedural planning. Furthermore, CT may complement assessment of mechanical heart valve dysfunction, aortic or pulmonary dissection and allows 3D visualization of abscess formation in endocarditis, and is even practical in the critically ill patient. However, CT exposes the patient to ionizing radiation and iodinated contrast agents and does not provide information on haemodynamics, flow rate, or velocity. CT can be used to acquire ventricular volumes and function but with lower temporal resolution than 2D/3D echocardiography or CMR and at the expense of additional radiation exposure.³⁴ CT volumes correlate with CMR and newer generation CT showed excellent accuracy for LV volumes.³⁵ However, CT is clearly unattractive for serial measurements because of the cumulative radiation dose. For preoperative screening in low risk patients, cardiac CT for coronary artery imaging has a useful role. Specific specialist knowledge of ACHD is required for appropriate planning and interpretation of cardiac CT.

Key points

- CT is superior to CMR in the setting of critically ill, or claustrophobic patients, when the length of a CMR acquisition may not be tolerable or practical.
- Cardiac CT allows concurrent evaluation of lung parenchyma or airways.
- CT is indicated when evaluation of calcification is needed (e.g. within vessels and surgical conduits prior to intervention) or if detailed evaluation for coronary stenosis is sought.
- CT can be considered in the presence of CMR non-compatible implants (e.g. pacing leads, non-MR compatible pacemaker/defibrillator) and in the setting of poor CMR image quality due to metallic artefact.

Chest X-ray

The chest X-ray should be used as part of the periodic follow-up of ACHD patients. Chest X-ray has a small radiation dose (dose from two views is limited to 0.1 mSv). Chest X-ray provides useful diagnostic information on heart and vessel size and shape, pulmonary vasculature, and lungs. Furthermore, cardio-thoracic ratio relates to functional class and predicts survival in ACHD.³⁶

Cardiac catheterization

The use of catheterization has evolved in the past few decades away from solely obtaining diagnostic information towards its main contemporary use for therapeutic interventions in ACHD patients. However, fluoroscopic and angiographic imaging may still have a place where more advanced imaging techniques do not provide sufficiently detailed anatomical information. Angiographic coronary

Table 2 Cardiac imaging goals in selected adult congenital heart diseases

| Specific lesions | Echocardiography | CMR | CT | Cardiac cath and angiography |
|---|--|---|--|--|
| Repaired tetralogy of Fallot | RV size and function, RVOT, branch PAs, residual intracardiac shunts, RV pressure, Tricuspid regurgitation, pulmonary regurgitation, LV size and function, aortic root size, proximal ascending aorta | RV volumes and RV EF, regional RV wall motion, RVOT aneurysms, branch PA anatomy and flow, accurate QP: QS, pulmonary regurgitant fraction, LV size and function, aortic root and ascending aortic size, aortic-to-PA collaterals, origin and proximal course of coronary arteries | In selected cases when echocardiography does not answer the clinical question and CMR is contraindicated or limited by artefacts (stent, leads), Ventricular volumes and EF, detailed branch PA anatomy, aortic root and ascending aortic size, aortic-to-PA collaterals, detailed coronary artery visualization, RV to PA conduit calcification and relation to coronary arteries | When haemodynamic data are important for management, in selected cases when echocardiography is inconclusive and CMR is contraindicated or where cardiac CT is insufficient to exclude coronary artery disease |
| Transposition of the great arteries after atrial switch procedure | Systemic RV size and function, systemic and pulmonary venous pathways (baffle leaks or obstructions), residual intracardiac shunts, tricuspid regurgitation, LV size and function, LVOT obstruction (pulmonary stenosis) | Systemic RV volume and RV EF, systemic and pulmonary venous pathways (baffle leaks or obstructions), residual intracardiac shunts, tricuspid regurgitant fraction, LV size and function, LVOT obstruction (pulmonary stenosis), origins and proximal courses of the coronary arteries | In selected cases where echo does not answer the clinical question and CMR is contraindicated, Ventricular volumes and EF, systemic and pulmonary venous pathways (baffle leaks or obstructions), residual intracardiac shunts, LVOT obstruction (pulmonary stenosis), origins and proximal courses of the coronary arteries | When intra-cavity pressures, pressure gradients are required for further medical or surgical decision-making, combined interventional procedures are needed |
| Transposition of the great arteries after arterial switch procedure | LV size and function, residual intracardiac shunts, main PA and proximal branch PA stenosis, neo-aortic root dilation and regurgitation, myocardial stress perfusion | LV volumes and EF, residual intracardiac shunts, main PA and proximal branch PA stenosis, neo-aortic root dilation and regurgitation, origin and proximal course of coronary arteries, myocardial stress perfusion | In selected cases, LV volumes and EF, residual intracardiac shunts, Main PA and proximal branch PA stenosis, neo-aortic root dilation and regurgitation, detailed coronary arteries anatomy | In case of interventional procedure (PAs balloon angioplasty or stent) or when coronary artery stenosis is strongly suspected clinically and by non-invasive imaging |
| After Rastelli operation | Evaluation RV to PA conduit, main PA and branch PAs, RV size and function, tricuspid regurgitation, RV pressure, residual intracardiac shunt | Evaluation RV to PA conduit, main PA and branch PAS flow quantification, RV volumes and EF, tricuspid regurgitation, multilevel obstruction to identify the most relevant | In selected cases where echo does not answer the clinical question and CMR is contraindicated or limited, RV to PA conduit calcification and proximity to coronary arteries | Before redo surgery or percutaneous pulmonary valve implantation to provide intra-cavity pressures, end-diastolic ventricular pressures, pressure gradients and coronary and pulmonary angiography |
| Atrioventricular and ventriculo-arterial discordance (unoperated) | Systemic RV size and function, residual intracardiac shunts, tricuspid valve morphology and regurgitation, Aortic regurgitation, LV (subpulmonary) size and function, LVOT obstruction (pulmonary stenosis). | Systemic RV volumes and EF, residual intracardiac shunts, tricuspid valve morphology and regurgitation, aortic regurgitation, LV (subpulmonary) volumes and EF, LVOT obstruction (pulmonary stenosis) | In selected cases, Ventricular volumes and EF, Residual intracardiac shunts, tricuspid valve morphology and regurgitation, aortic regurgitation, LVOT obstruction (pulmonary stenosis) | Assessment of end-diastolic ventricular pressure and pulmonary vascular resistance in preoperative patients or in cases where full non-invasive evaluation is inconclusive |
| Aortic coarctation | Aortic valve morphology, function, pressure gradient, | Ascending, transverse and descending thoracic aortic | Particularly useful in the case of prior stents, Ascending, transverse, and | Interventional procedures |

Continued

Table 2 Continued

| Specific lesions | Echocardiography | CMR | CT | Cardiac cath and angiography |
|------------------|--|---|--|---|
| | subaortic anatomy (membrane) and pressure gradient, aortic root, ascending aortic and aortic arch anatomy, LV size, function, diastolic function, and wall thickness | pathology (dilation, multi-level stenosis and aneurysm), Aortic valve morphology, LV volumes, EF, and mass, Aortic collaterals | descending thoracic aortic pathology (dilation, aneurysm), aortic collaterals | |
| Single ventricle | Ventricular size and function, atrioventricular valve regurgitation, Fontan pathway patency, branch PA visualization, LVOT obstruction, aortic regurgitation | Ventricular volumes and EF, atrioventricular valve regurgitant fraction, Fontan pathway patency, Branch PA calibre and flow quantification, LVOT obstruction, aortic regurgitation fraction, pulmonary venous compression, aortic-to-pulmonary and systemic-to-pulmonary venous collaterals | In select cases, ventricular volumes and EF, Fontan pathway patency, branch PA calibre, LVOT obstruction, pulmonary venous compression | Haemodynamic data regarding severity of stenosis, PA pressure, pulmonary vascular resistance and to further delineate thoracic vascular anatomy, particularly of the pulmonary vascular bed and collateral vessels, interventional procedures |

Modified from Babu-Narayan *et al.*²

LV, left ventricle; RV, right ventricle; EF, ejection fraction; LVOT, left ventricle outflow tract; PA: pulmonary artery.

evaluation and baffle leak detection are examples in which non-invasive imaging does not give always-sufficient information. Heart catheterization is still the gold standard for intra-cavity and intravascular pressure measurements. Moreover, right heart catheterization is the only way to confirm pulmonary arterial hypertension and pulmonary vascular disease.⁷ Direct peak-to-peak pressure differences can easily be obtained by catheterization and are less sensitive for measurement errors when compared with non-invasive methods. Cardiac output relies on thermo-dilution or Fick methodology.

In complex cases, optimal calculation of pulmonary vascular resistance is derived from a hybrid approach with CMR providing flow data and simultaneous catheterization providing pressure data. Such combined CMR-Catheter procedures are being used to provide enhanced diagnostic data in complex ACHD patients who may often require multiple cardiac catheterizations.^{37–39} This approach to diagnosis as well as treatment, although nowadays limited to few centres, offers the advantages of a radiation-free or minimal radiation examination, increased soft tissue contrast, theoretically unlimited imaging planes for catheter guidance. Hybrid measurements provide a more accurate and comprehensive haemodynamic assessment than other traditional techniques.^{37–39} While hybrid CMR-catheterization laboratories are well placed to do this simultaneously, ACHD patients can still benefit from separate but contemporaneous flow (CMR) and pressure (catheter) acquisitions from adjacent facilities.

Balloon dilatation and/or stenting of vessels, vessel occlusion, balloon dilatation and/or implantation of valves, and other structural interventions have become increasingly common practice in ACHD and peri-procedure imaging such as TOE, ICE, and CT may be

required to support the interventionalist. CMR-guided cardiac catheterization without fluoroscopy is not yet ready to be widely used in the clinical setting,³⁷ because of the increased overall cost, greater operational complexity, reduced temporal and spatial resolution, restricted access to the patient, limited patient monitoring, and ambient noise during the scan.^{37–39}

Key points

- Cardiac catheterization is recommended for cardiac chamber intra-cavity and vascular pressure measurements.
- Cardiac catheterization is the gold standard for the diagnosis of pulmonary vascular disease.
- Cardiac catheterization is mainly employed when a percutaneous therapeutic procedure is contemplated.

3D printing

Cardiac 3D printing produces a replica of the patient's heart anatomy and is a powerful application that offers several advantages to patient care particularly in the ACHD population. In ACHD patients with complex cardiac anatomy and multiple surgeries, 3D models allow precise understanding of the patient's anatomy and physiology enabling detailed pre-surgical planning, and are useful tools for patient counselling, trainee education, and simulation.^{40,41}

Selected specific lesions

Repaired tetralogy of Fallot

Surgical rTOF includes closure of a VSD, relief of RV outflow (RVOT) tract stenosis using either a pulmonary-valve sparing technique or a more extensive RVOT reconstruction incorporating a transannular patch (Table 2). In case of branch pulmonary stenosis, arterioplasty of the PA branches may also be required. The main goals of imaging in the follow-up of rTOF patients are to identify residual or postoperative lesions, quantify their haemodynamic importance, study the effect on RV and LV function and inform clinical decision-making regarding timing and method of reintervention. Typical residual lesions include pulmonary valve regurgitation (common in contemporary ACHD patients), residual RVOT obstruction (less common in the contemporary adult population unless a conduit was implanted as part of the repair), pulmonary branch stenosis, residual VSD, or ASD. Secondary lesions include progressive RV dilatation and dysfunction, TR, aortic root dilatation, and aortic insufficiency. Progressive RV dilatation and development of RV dysfunction secondary to pulmonary regurgitation is the most commonly observed problem in this patient group. Quantification of RV size and function is considered fundamental in decision-making for timing of pulmonary valve replacement.^{1–5} LV dysfunction must also be considered especially in older population (age >40 years).^{42,43}

Echocardiography

Echocardiography with a specific protocol focusing on the relevant post-operative and residual lesions and their haemodynamic implications is strongly recommended as for all ACHD lesions. Echocardiographic evaluation should include the assessment of residual RVOT obstruction and proximal PA branches. RVOT views from parasternal short- and long axis are the standard echocardiographic views for assessing the RVOT.^{5,44} Pulsed and continuous wave Doppler are used for the assessment of gradients. Peak RVOT gradient >64 mmHg is considered as severe but overestimation of the peak gradient is possible in the context of severe pulmonary valve regurgitation and long-segment stenosis (typically conduit stenosis).⁵ Assessment of PA branch stenosis may be more challenging in adults related to limited acoustic windows and a combination of parasternal and suprasternal windows may be required. The assessment of severity of pulmonary regurgitation is based on parasternal short- and long-axis views and includes assessment of jet width at valve level, diastolic flow reversal from the main and distal PA branches and pulsed Doppler (pressure half-time <100 ms). Generally, echocardiography is reliable in patients with haemodynamically significant pulmonary regurgitation.^{5,44,45}

While the echocardiographic assessment of LV size has been standardized, there is more controversy on the techniques used for the assessment of RV size and function (Figure 1). We recommend including quantitative measures of RV size and function for routine follow-up. For RV dimensions, we recommend using RV 2D measurements from different RV views (short-axis, long-axis, and apical four-chamber views) and measuring RV end-diastolic area from the apical four-chamber view.¹² For RV functional assessment, FAC from the RV-focused view, TAPSE or tissue Doppler traces in the tricuspid annulus should be combined. RV volumetry based on 3D

echocardiography and RV strain measurements could be included, as emerging techniques in the echo evaluation of RV function.^{12,46,47} 3D echocardiographically obtained RV volumes, although well correlated, are typically underestimated when compared with CMR volumes. In laboratories with experience in 3D echocardiography, 3D echo RV volumes should be used in the follow-up of the patients but should not be used interchangeably with CMR.^{47,48} Assessment of LV function is an essential part of the assessment as RV dysfunction is associated with LV dysfunction. LV measurements by 3D echocardiography are accurate and reproducible and should be used when available and feasible.¹² 2D STE-derived GLS appears to be reproducible and feasible for clinical use and myocardial deformation parameters offer incremental prognostic value in patients after TOF repair.^{44,49,50}

RV diastolic function can also be assessed by Doppler echocardiography. Anterograde flow in the main PA during atrial contraction throughout the respiratory cycle,^{51,52} is considered a sign of RV restrictive physiology. Restrictive RV filling pattern and dilated right atrium in the absence of significant TR and/or RVOT obstruction are also signs of RV diastolic dysfunction. Quantification of pulmonary regurgitation and clinical interpretation of RV dilatation consequent to pulmonary regurgitation need to account for the presence of restrictive RV physiology.⁵³

The severity of TR and RV systolic pressure assessment is based on generally accepted practices and is not specific for TOF patients. Especially in patients with residual RVOT, assessment of RV systolic pressure is essential as RVOT gradients may be overestimated in cases of long-segment stenosis or underestimated in cases of distal branch stenosis.

Echocardiographic protocols should specifically look for the presence, size and flow direction of residual atrial, and ventricular communications. Progressive aortic root dilatation and aortic regurgitation have been identified in patients after TOF repair and should be included in the routine follow-up. However, prospective data are reassuring suggesting that although aortic dilatation is very common it is not rapidly progressive.⁵⁴

In patients after arrhythmia ablation or after pacemaker implantation, detailed haemodynamic assessment with appropriate pacemaker optimization should be carried out with intention to achieve optimal cardiac output.

CMR imaging

Cardiovascular magnetic resonance imaging is the clinical reference technique for the serial quantification of pulmonary regurgitation, RV volumes and RV function during follow-up of adults after TOF repair (Figure 2). Contemporary publications show that reverse remodelling happens immediately after surgery with a smaller degree of ongoing biological reverse remodelling.^{55–58} Two recent studies show that the optimal preoperative threshold to achieve mid-to-late RV normalization was an indexed RV end-systolic volume (RVESVi) <80–82 mL/m², respectively.^{55,59} CMR also allows for assessment of more distal PA branches, coronary origins and proximal course, LV size and function, aortic root dimensions, and quantification of aortic regurgitation. Research centres are evaluating the role of CMR-defined myocardial scarring and degree of localized or diffuse fibrosis.^{60,61} Different centres use different CMR protocols, relating to local experience

secondary to patch/baffle dehiscence is not uncommon as prosthetic material might degenerate over time.

Echocardiographic

Echocardiographic evaluation of the conduit, pulmonary trunk, and (if possible) PA branches is essential in the follow-up of Rastelli patients. However, as the valved conduit is located immediately behind the sternum, it is relatively difficult to directly estimate the RVOT pressure gradient. As outflow tract gradients might be under- or overestimated, it is important to qualitatively evaluate subpulmonary RV function and estimate RV systolic pressure based on tricuspid valve regurgitation velocity. *TOE* can give complementary information when assessing for residual intracardiac shunt.

Cardiovascular magnetic resonance imaging

Cardiovascular magnetic resonance imaging enables quantification of subpulmonary RV function. CMR can be especially useful in multilevel RVOT to determine the site of the most haemodynamically significant obstruction e.g. at subvalvar, valvar, or supra-valvar anatomical level.⁴⁵

Cardiac CT

Cardiac CT is relevant in selected circumstances (see above).

Cardiac catheterization

Cardiac catheterization is often utilized before redo surgery to provide crucial information on intra-cavity pressures, end-diastolic ventricular pressures, pressure gradients and coronary and pulmonary angiography, before proceeding to percutaneous pulmonary valve implantation in suitable patients.

Key points

- Echocardiography and CMR are performed regularly in ACHD patients after Rastelli operation for evaluation of postoperative complications (RV-PA conduit dysfunction, LV outflow tract obstruction, aortic valve regurgitation, late VSD patch leak, RV and LV function).
- Cardiac CT and cardiac catheterization may be useful before reintervention, including determining suitability for transcatheter pulmonary valve replacement.

Atrioventricular and ventriculo-arterial discordance (unoperated)

Atrioventricular and ventriculo-arterial (AV-VA) discordance or congenitally corrected TGA (ccTGA) is a rare malformation often associated with dextrocardia/mesocardia or situs inversus, VSD, pulmonary stenosis. The condition and associated outcomes are highly variable. Unoperated, patients can be asymptomatic for decades but the morphologic RV and the tricuspid valve in systemic

position tend to fail over time. The tricuspid valve maybe dysplastic, 'Ebstein-like' and contributes to the TR. The TR leads to RV volume overload and exacerbates RV dysfunction. Severe RV failure or fatal tachyarrhythmia can occur unpredictably. Medical follow-up will aim to identify sub-clinical systemic RV dysfunction.⁸⁴⁻⁸⁸

Careful imaging evaluation of the mechanism and severity of TR and RV function allows contribution to clinical decision-making regarding tricuspid valve repair, tricuspid valve replacement, or PA banding to shift the interventricular septum towards the systemic RV.⁸⁷ Surgery for severe TR in ccTGA patients with preoperative RVEF >40% (from echocardiography, angiography, CMR, or CT) has better outcomes with respect to postoperative RV function.⁸⁸

Echocardiography

Echocardiography should be performed at least annually. The common presence of dextrocardia or mesocardia together with concerns regarding the accuracy of systemic RV function and TR quantification are particularly challenging for the echocardiographer. FAC and systemic RV-GLS are the echocardiographic measures which correlated best with CMR-derived RV EF,⁸⁷ exercise capacity and biomarkers.⁸⁸⁻⁹⁰ A cut-off value of RV-GLS <-16.3% identified a CMR-derived RVEF \geq 45% with a sensitivity of 77.3% and a specificity of 72.7%. Decreased RV-GLS is associated with elevated NT-proBNP.^{87,91} PA pressure can be estimated from mitral regurgitation velocity.

Cardiovascular magnetic resonance imaging

Cardiovascular magnetic resonance imaging is the gold standard to evaluate systemic RV EF, mass and volume,⁸³⁻⁸⁶ and can be used to quantify tricuspid regurgitant fraction and may complement echo to understand the mechanism of TR. CMR should be performed as a baseline evaluation, repeated when there change in clinical status or signs of deterioration on TTE and then regularly in patients with progressive TR. The contemporary gold standard for measurement of RVEF is CMR. Devices are not uncommon in ccTGA due to the high incidence of conduction disturbance and arrhythmia. In those with MR non-compatible pacemakers and defibrillators CMR may not be feasible (unpredictable artefacts or lack of local expertise).

Cardiac CT

Cardiac CT is not routinely recommended. However, when there is a contra-indication for CMR and RVEF is a crucial question then it is feasible to obtain RV volumes from retrospectively gated CT acquisitions at the cost of increased irradiation. The use of CT in this population is usually for preoperative screening for coronary artery disease in older patients (>40 years).

Cardiac catheterization

Cardiac catheterization is usually confined to assessment of end-diastolic ventricular pressure and pulmonary vascular resistance in preoperative patients including those being assessed for heart or heart and lung transplantation or in cases where there is uncertainty despite full non-invasive evaluation.

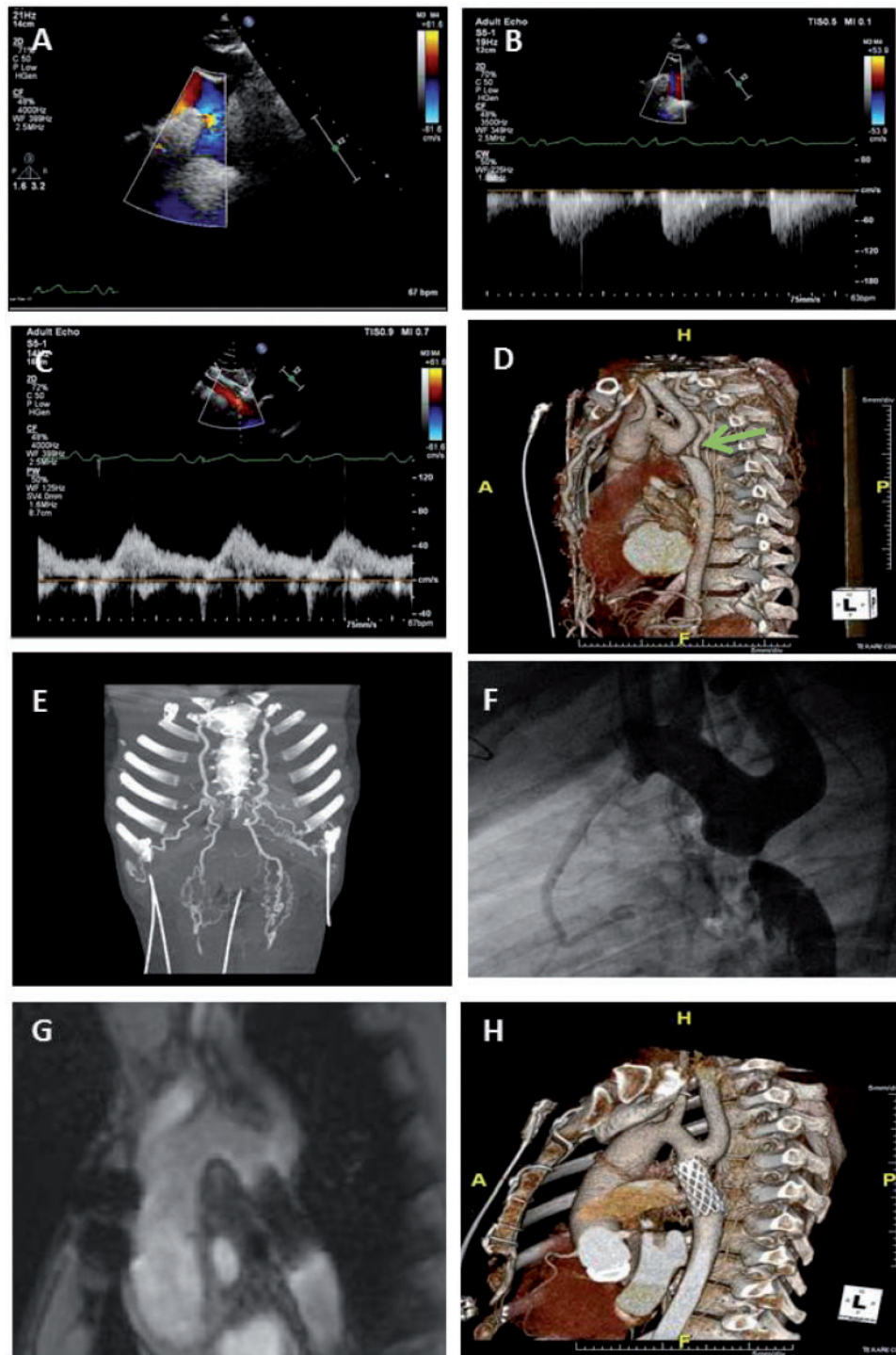


Figure 5 Multimodality imaging evaluation in an adult patient native severe aortic coarctation. (A and B) Echocardiographic suprasternal view with colour Doppler, showing a narrowing of the proximal descending aorta distal to subclavian artery. Corresponding continuous-wave Doppler showing an under-estimated peak gradient across the coarctation with the evidence of diastolic tail. (C) Abdominal aorta with dampened low-velocity signal with continuation flow through diastole. (D) 3D reconstruction at cardiac computed tomography scan showing just distal to the left subclavian artery origin a very severe discrete coarctation of the aorta (green arrow) with probable complete obstruction; the coarctation site is bypassed by numerous collateral arteries. (E) Cardiac computed tomography scan focused on the numerous and very well developed collateral arteries; in addition, there are enlarged intercostal, internal mammary and epigastric arteries, large collaterals around the scapulae, and numerous enlarged vessels throughout the mediastinum. (F) Angiography showing the very severe aortic coarctation with multiple collaterals. (G) Cardiac magnetic resonance imaging showing the stent at the site of previous coarctation. (H) The same stent showed by 3D reconstruction at cardiac computed tomography scan.

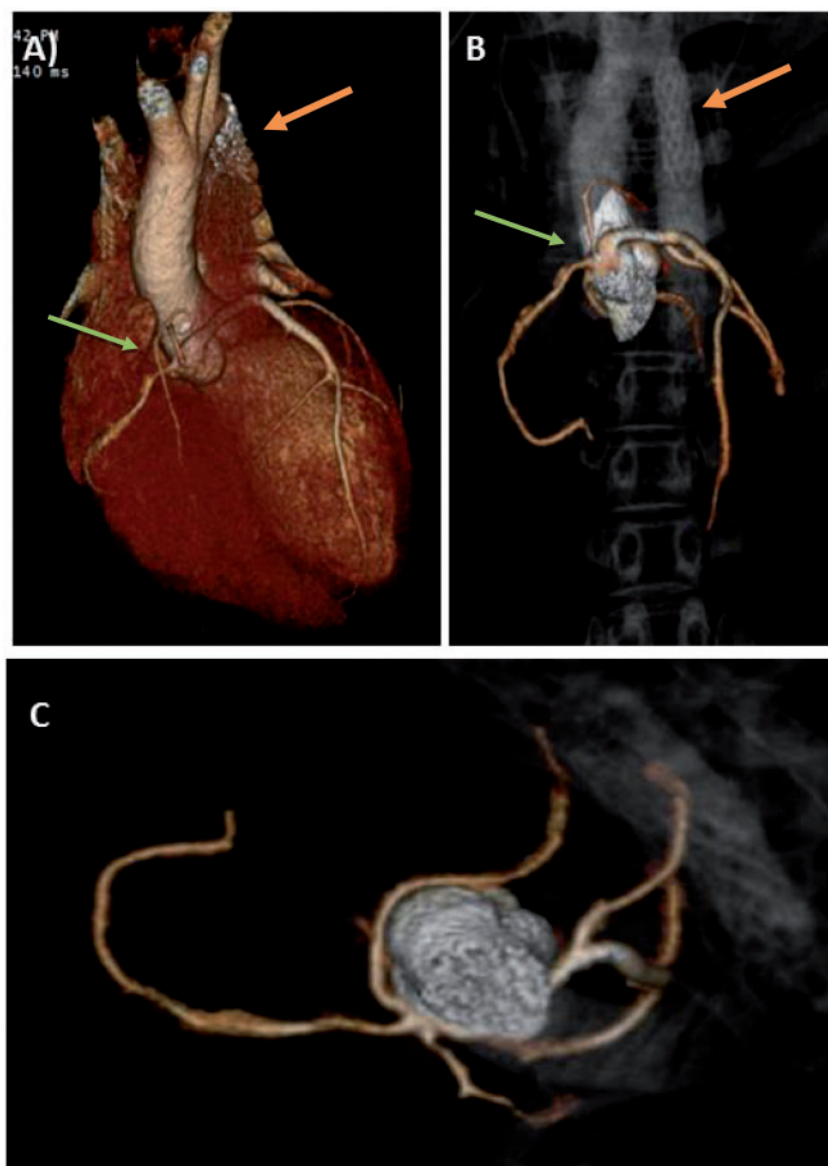


Figure 7 Cardiac computed tomography scan of patient with a rare variant of a single coronary ostium arising from the right sinus of Valsalva (A and B) 3D reconstruction showing a normal ascending aorta with stenting of the proximal part of descending thoracic aorta for a previous coarctation (green arrow); all three coronary arteries originated from a single ostium in the right aortic sinus (orange arrow). (C) Upper view of the single right coronary ostium with the course of coronary arteries. Courtesy of Dr Thomas Semple.

Key points

- Function of the systemic RV and degree of TR should be monitored regularly (annually) with echocardiography.
- CMR should be performed at baseline and be repeated regularly or when there is change in clinical status or echocardiographic findings.
- Cardiac CT is not routinely recommended.
- Cardiac Catheterization should be performed to assess ventricular end-diastolic pressure or pulmonary vascular resistance.

forward flow is achieved relatively passively. In the earlier era, this was achieved with an anastomosis between the right atrial appendage and the pulmonary arteries (atrio-pulmonary Fontan), whereas more recently, a total cavo-pulmonary connection, either using an intra-atrial/lateral tunnel or extracardiac conduit to connect the superior and inferior caval veins into the adjacent PA (usually right PA). The Fontan circulation achieves good early palliation but carries many longer term disadvantages^{3,112} including ventricular dysfunction, atrio-ventricular valve failure (particularly the tricuspid valve), atrial and ventricular arrhythmias, thrombo-embolic complications and high systemic venous pressure which is associated with protein losing enteropathy and hepatic fibrosis as important clinical problems.

Veno-atrial collaterals (systemic venous to left atrium) promotes progressive cyanosis, and collaterals from systemic arterial to the pulmonary arterial system causes volume overload of the single ventricle.³

In tricuspid atresia with TGA, a VSD if present, may become restrictive over time resulting in systemic outflow obstruction.²

Palliation of many forms of univentricular heart (UVH) requires aortic arch reconstruction. The aortic (or neo-aortic) valve can be stenotic or regurgitant because of congenital abnormalities or after surgical repair. The aorta, particularly in patients who underwent surgical repair, is at risk for recurrent obstruction or progressive aneurysmal dilation.^{2,4} Even a minor degree of obstruction after aortic arch reconstruction can lead to worsening diastolic and systolic ventricular function. The benefits of a well-organized multimodality imaging approach to evaluate all the intracardiac and extracardiac complications of operated and unoperated UVH are obvious.

Echocardiography

Subxiphoid, suprasternal, and high-right parasternal views can be used to rule out obstruction of the cavo-pulmonary connections, where a mean gradient of 3 mmHg can reflect clinically important obstruction. Dilated hepatic or innominate veins can raise suspicion of Fontan obstruction. From the subxiphoid views fenestrations can be seen and a mean gradient of 4–8 mmHg are considered reassuring. From subxiphoid views, patch leaks in intracardiac Fontan may be seen as small gaps between the Fontan channel and the atrium.

Aortic outflow obstruction or regurgitation will both lead to an elevation in atrial pressure and, are poorly tolerated in the Fontan patient. The quantification of semilunar valve dysfunction should follow the standard echo-Doppler methodology. Suprasternal and high left parasternal transducer positions are generally best for the visualization of the aortic arch, but interrogation of flow in the abdominal aorta can provide the first clue that aortic arch obstruction is present. In the UVH, the suspicion of residual aortic arch obstruction may be based more on the morphology of the aortic arch rather than on the presence of a significant gradient, which is very often misleading because of the unique haemodynamics of UVH patients.

Ventricular function can be assessed by using biplane Simpson's in case of LV morphology and FAC in case of right or indeterminate ventricular morphology. Myocardial deformation imaging has been shown to identify differences in contraction patterns in UVH compared with normal hearts, as well as changes that occur with different stages of palliation.^{113,114} A recent study in adult patients after Fontan-type repair has shown that atrioventricular valve systolic to diastolic ratio as a non-geometric parameter correlates with ACHD

patient outcome.¹¹⁵ The incorporation of newer quantitative echocardiographic tools into clinical practice may aid in identifying abnormalities in ventricular function as well as the presence of mechanical dyssynchrony which may have potential therapeutic implications for the management of heart failure in this group of patients. 3D *echocardiography* can allow functional assessment of the single ventricle without any geometric assumptions, overcoming the limitations of 2D echo.¹¹⁶ The evaluation of diastolic function is particularly challenging in Fontan patients. However, a prolonged (>28 ms) pulmonary venous atrial reversal relative to atrial forward flow duration into the single ventricle is a marker of elevated filling pressure in the UVH, as well as a progressive prolongation of E wave deceleration time.¹¹⁷

Veno-atrial collaterals or aorticopulmonary arterial collateral connections are difficult to detect by standard 2D echocardiographic imaging, but are often suspected. *Contrast echocardiography* using agitated saline injection in an upper extremity resulting in prompt opacification of the atrium would add supportive evidence for veno-atrial collaterals. However, echocardiography has a very limited capacity for describing course, size and number of collaterals and cannot quantify the amount of shunt.

Unfortunately, acoustic windows are particularly challenging in many adult patients with Fontan in whom the ultrasound penetration is limited by a larger chest as well as by multiple surgical interventions. In most cases, complete evaluation including the venous and arterial anatomy requires multiple imaging modalities.

Cardiovascular magnetic resonance imaging

Cardiovascular magnetic resonance imaging can establish patency of pathways, exclude thrombus, quantify the volume of the dominant ventricle, systemic ventricle EF, quantify atrioventricular valve regurgitation,^{3,118} and image collaterals. CMR is also the method of choice for quantification of relative distribution of blood flow to the left and right lungs. Myocardial fibrosis as identified by LGE is a common finding and is associated with systolic dysfunction and non-sustained ventricular tachycardia¹¹⁹ but prospective studies are pending.

Cardiac catheterization

Cardiac catheterization is frequently necessary to derive haemodynamic data regarding severity of stenosis, PA pressure, pulmonary vascular resistance and may be used to further delineate thoracic vascular anatomy, particularly of the pulmonary vascular bed and collateral vessels. UVH patients will often benefit from a combination of CMR-derived flow measurement with invasive pressure measurements to derive pulmonary vascular resistance.

Fontan-associated liver disease is a vexing condition often associated with poor outcomes and is a well demonstrated precursor of hepatocellular carcinoma.¹²⁰ Surveillance usually includes an abdominal liver ultrasound scan. Ultrasound-derived elastography is a promising non-invasive technique to assess liver stiffness and fibrosis.^{3,112–114}

Lymphatic abnormalities are frequent and multifactorial in the Fontan circulation. Lymphatic anomalies are a critical ingredient in the pathophysiology of plastic bronchitis and protein losing enteropathy.^{120–123} Dynamic contrast lymphangiography using CMR can be helpful to better understand the pathophysiology of pulmonary

lymphatic flow discordance and can guide percutaneous procedures.^{120–123}

Key points

- Echocardiography remains first line imaging for assessment of patency of Fontan pathways, valve function, and systolic and diastolic ventricular function.
- CMR is routine for Fontan surveillance in ACHD and can establish patency of pathways, is sensitive for excluding thrombus, accurately quantifies systolic function (EF), atrioventricular valve regurgitation and allows evaluation of central pulmonary arteries, aortic arch, and veno-venous collaterals.
- CT may be advisable before re-intervention as it may guide the catheterization or surgical approach.

Conclusions

Cardiovascular imaging has evolved from a single modality-based to an integrated multimodality based approach in order to achieve a comprehensive and detailed assessment of individual patient morphology, pathophysiology, and haemodynamics and for guidance of therapy. ACHD patients are a heterogeneous population, with a large spectrum of anatomic substrates even within specific lesions. In addition, the nature of previous surgery and other intervention is highly variable rendering each patient unique. As the ACHD population continues to age, acquired heart disease will also require cardiac imaging assessment. It is, therefore, logical that ACHD patients are amongst those with the most to benefit from the appropriate application of a multimodality imaging approach by appropriately expert imagers in centres with sufficient expertise. It remains important to appreciate the different advantages and disadvantages of different cardiovascular imaging modalities as they evolve. Increasingly, clinicians will benefit from fused multi-modality imaging datasets. The goal should be to provide the most appropriate and cost-effective diagnostic pathway for each individual ACHD patient. In other words, ACHD patients should have the right imaging, at the right place by the right operator at the right time.

Acknowledgement

We gratefully thank Dr Thomas Semple BSc, MBBS, FRCR Radiology department, The Royal Brompton Hospital and National Heart and Lung Institute, Imperial College London, in contributing images to better illustrate this paper.

Conflict of interest: None declared.

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