Cardiovascular MRI of Adult Congenital Heart Disease

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Learning Objectives

• To provide an overview of the basic cardiac Cardiac MR (CMR) sequences and protocols used for the evaluation of Adult Congenital Heart Disease (ACHD)
• To understand the strengths and limitations of CMR for the evaluation of ACHD
• To review the role of CMR in the evaluation of the major ACHD including repaired Tetralogy of Fallot (TOF), Transposition of the great arteries, Coarctation of the aorta, and post Fontan operations

CMR ACHD

• The population of patients with ACHD continues to grow as a result of advances in cardiac surgery and critical care
• Many ACHD patients have undergone palliative or reparative surgery earlier in life
• The operations performed for more complex malformations are rarely curative
• The majority of these patients have residual hemodynamic abnormalities requiring serial imaging, and often further intervention

Advantages of CMR for ACHD

• Unrestricted access to cardiovascular anatomy and function, including the systemic and pulmonary venous connections, the right ventricle (RV) and pulmonary arteries, and the whole aorta, without ionizing radiation.
• Well suited for repeated, life-long follow-up investigation, if needed.
• Versatility, including measurements of biventricular size and function regardless of chamber geometry, measurements of flow volumes, characterization of tissues, and assessment of myocardial function, viability, and perfusion, when required.

CMR Sequences for ACHD

• 2D Steady state free precession (SSFP)
• Black blood imaging- Double inversion recovery
• Phase contrast imaging
• 3D SSFP
• 3D Gadolinium enhanced MRA
• Delayed enhancement
**CMR Sequences: 2D SSFP; Standard CMR sequence for cardiac morphology and function**

Congenitally Corrected Transposition of the Great Arteries

**CMR Sequences: 2D SSFP Short axis Reference standard for ventricular volumes, systolic function and ventricular mass.**

- Highly accurate and reproducible for ventricular volumes and mass in a variety of disease states, including in those with a complex chamber geometry.

**CMR Sequences: 3D Gadolinium enhanced MRA**

Aortic Coarctation
3D Gad MRA
High resolution vascular isotropic imaging

**CMR Sequences: 3D MRA Co A. Volume rendering**

Ao
PA
AVSD
09/0A 2.5.1

**CMR Sequences: Phase contrast Imaging**

Coarctation of the aorta
Sinus venosus ASD

Particularly useful for evaluation of cardiac morphology when 2D SSFP imaging has artifacts such as due to metal.
CMR ACHD. Sequences: 3D Whole heart SSFP

- Isotropic; Multiplanar reformating
- Comprehensive evaluation of intracardiac and extracardiac anatomy including coronary artery anatomy
- Does not require IV gadolinium
- Navigator gated free breathing
- Typically triggered in end diastole

Situs solitus
L transposition
(SLL) congenitally corrected Transposition (CCTGA)

CMR Sequences: Delayed Myocardial enhancement

Post op TOF: Enhancement in the RVOT

Fibrosis in a variety of ACHD is increasingly recognized as a risk factor for adverse events including arrhythmias

General CMR protocol in ACHD

2D Cine Steady State Free Precession (SSFP)
- Short axis plane for the study heart function
- Long and short axes for evaluation of complex anatomy

Black Blood Imaging
- For anatomy and morphology: If cine imaging is unsatisfactory due to susceptibility artifacts

3D SSFP imaging
- For evaluation of extracardiac and intracardiac anatomy including coronary artery anatomy

Phase Contrast Imaging
- Through the plane of the aortic root, main pulmonary artery and other planes as needed

3D Gadolinium Enhanced MRA
- Evaluation of extracardiac vascular anatomy

Late Gadolinium Enhancement (LGE)
- For detection of fibrosis as increasing recognized prognostic marker in ACHD

CMR ACHD. Safety issues and Limitations

- Metallic objects and implantable devices: Pacemakers and AICD.
- Gadolinium chelates (MRA and LGE). Not recommended in severe renal impairment eGFR <30ml/min/1.73sqm. NSF.
- Claustrophobia and patient compliance
- Portability; availability during open heart surgery

CMR ACHD: Repaired Tetralogy of Fallot

Sequential segmental approach to analysis
- Atrial situs.
- Ventricular Morphology
- Ventriculoarterial connection
- Identification of other abnormalities: Abnormal venous connections, septal defects, valve abnormalities.
Post op TOF. Key postoperative issues

- Residual pulmonary regurgitation
- RV dilatation and dysfunction due to pulmonary regurgitation, possibly with associated tricuspid regurgitation
- Residual RVOT obstruction, branch pulmonary artery stenosis or hypoplasia
- Sudden cardiac death
- Sustained VT, AV block, atrial flutter, and/or atrial fibrillation
- VSD patch leaks
- Residual Aortopulmonary collaterals
- Aortic root dilatation

CMR Repaired TOF

? Optimal timing for PVR

? Preoperative thresholds for PVR

CMR Repaired TOF

RV dilatation and dysfunction by CMR predict adverse outcomes such as RHF, arrhythmia, and death

LV systolic dysfunction by CMR independently correlates with clinical status and correlates with RV dysfunction

Geva T et al JACC 2004

CMR Repaired TOF

Pulmonary regurgitation as measured by CMR is closely associated with the degree of RV dilatation

Rebergen SA Circulation 1993

PA EDPFF as measured by CMR correlates with more severe pulmonary regurgitation and poorer exercise performance at mid- to long-term follow-up


CMR Repaired TOF

LV systolic dysfunction: RV dilatation and dysfunction

Markedly enlarged RV post repair: EDV 200 ml/m²

CMR. Repaired TOF

Pulmonary regurgitation as measured by CMR.

50 M. Post TOF repair. Tachypnea & Shortness of breath. Severely dilated RV. EDV 363 ml (183.5 ml/m², z score 6.75). RVOT patch aneurysm. RV EF of 24%
The presence and extent of regional dysfunction including aneurysms at the RVOT adversely affects global RV function, LV systolic function and exercise capacity after TOF repair.

Davleous PA et al. JACC 2002

Delayed enhancement at operative and non-operative sites correlates with adverse outcomes including ventricular dysfunction, exercise intolerance and clinical arrhythmia.


Bilateral branch PA stenosis

RV EDV < 170ml/sqm & RV ESV < 85ml/sqm
Normalize post PVR

Therrien J et al AJC 2005

Even large RV gets smaller, normalization of RV size only if pre op RV EDV < 160ml/sqm

Oosterhof T et al. Circulation 2007

CMR Repaired TOF: Perspective

- CMR evaluation of RV/LV function, regional dysfunction, RVOT obstruction, conduit or PA stenosis and scarring contribute to decision making
- RV EDV of 150-170 ml/sqm a guide to RV volumes that should not be exceeded for PVR, however more factors to be considered...
- In the individual patient MRI data is used in conjunction with clinical features including symptoms, electrophysiological data & exercise testing

CMR Repaired TOF: Perspective for Percutaneous PV replacement

CMR aids appropriate patient selection by:

- 3D assessment of RVOT morphology, size and dynamics preprocedure
- Defining coronary artery anatomy

Schievano et al. JCMR 2007
**CMR ACHD. RV-PA Conduits**

Stenosed conduit between the RV and Pulmonary arteries, Post Truncus repair

**CMR ACHD: Post operative Aortic Coarctation**

- Allows assessment of restenosis or aneurysm formation in the region of coarctation repair, as well as any associated pathology such as stenosis or regurgitation of a bicuspid AoV, aortopathy, or LV hypertrophy
- Collateral flow can be quantified by comparing through-plane measurements of flow immediately proximal to the stenosis and at the level of the diaphragm. A decrease of 10% is expected physiologically, whereas an increase implies collateral flow rejoining the descending thoracic aorta
- The presence of diastolic prolongation of forward flow, or a diastolic tail, is a useful sign of significant (re-)coarctation, and can be demonstrated by plotting a velocity-time curve of jet flow beyond the coarctation

**CMR ACHD: Post operative Aortic Coarctation**

**CMR ACHD. D-Transposition of the great arteries**

**CMR ACHD. Atrial switch**
**D-TGA Atrial switch  
Mustard/Senning**

Key Imaging Issues:
- Systemic RV function
- Pulmonary vein or systemic venous baffle stenosis or leaks.

**CMR ACHD. D-TGA Post atrial Switch**

- SVC baffle obstructed
- Azygos vein bypass to IVS

**D –TGA Post atrial switch**

- Patent PV baffles

**D-TGA Post atrial switch**

- Dilated failing RV : EF 20%

**CMR ACHD. D-TGA Arterial switch**

**D-TGA Post Arterial Switch**

Arterial Switch Complications/Key Imaging issues:
- Main and branch PA stenosis
- Coronary ostial occlusion
- Aortic root dilatation
D-TGA Post arterial switch
Status post arterial switch. RPA stenosis and Aortic root dilatation

CMR ACHD
Post Fontan operation for functional single ventricles

CMR ACHD. Post Fontan
Key Imaging issues
• Patency of the cavo-pulmonary connections; stenosis, leak, thrombus
• Patency and size of the Pulmonary arteries
• Pulmonary veins
• Single ventricle size and function
• Atrio-ventricular valve function
• Outflow tract of the ventricle, neo aorto, aortic valve and arch
• Collateral vessels

CMR ACHD. Post Fontan Procedure
14yr old male .Post Fontan for HLHS

CMR ACHD. Post Fontan Procedure
14yr old male .Post Fontan for HLHS
Right Ventricular Dilatation/ PHTN ? Cause

In RV dilatation, CMR is of great utility:

- Assess RV size, function and RV cardiomyopathy
- Identify intracardiac shunts
- Identify extracardiac shunts
- Assess Qp:Qs

CMR can be considered a non-invasive alternative to cardiac catheterization in this group of patients.

If, as a CMR practitioner, a patient is referred for CMR as "? ARVC", and there is RV dilatation, consider the possibility of a shunt and systematically exclude or search for it.

51-year-old male. PHTN. Dilated RV on Echo.
CMR ACHD. Right Heart enlargement. PHTN. ? Cause

Right heart enlargement Pulmonary HTN ? Cause. Coronary sinus ASD

Qp/Qs 2:6: 1. Marked RV enlargement. RV EF 48%.

General recommendations for CMR in ACHD European Heart Journal. Kilner et al. 2010

- A dedicated CMR service should be regarded as an indispensable facility, complementary to echocardiography, in a centre specializing in the care of ACHD
- Baseline CMR: Many ACHD patients benefit from at least one CMR study. Provides a baseline for future reference and may identify unexpected anomalies, previously overlooked or misinterpreted
- Follow-up CMR: Echocardiography is generally suitable for routine follow-up, but CMR may be indicated if change is suspected. When serial CMR is required intervals of 3 or more years is appropriate in most cases

When to use CMR as an adjunct to echocardiography in clinical practice? European Heart Journal. Kilner et al. 2010

- Echo study is suboptimal unable to provide images and measurements of sufficient quality to inform clinical management.
- When values provided by echo are borderline or ambiguous, CMR should be used to corroborate or amend the echo values before making clinical decisions.

When to use CMR as an adjunct to echocardiography in clinical practice? European Heart Journal. Kilner et al. 2010

Where CMR usually informs management more effectively
- Evaluation of systemic and pulmonary veins
- Quantification of RV/LV volumes and EF, and myocardial mass
- Evaluation of the RVOT, RV–PA conduits, and the branch PAs.
- Quantification of pulmonary regurgitation
- Quantification of shunts by measurements of flow in the ascending aorta and pulmonary trunk.
- Evaluation of the entire aorta
- Aortic-pulmonary collaterals and arterio-venous malformations
- Coronary anomalies and coronary artery disease, including possible assessment of viability and perfusion
- Tissue characterization in the ventricles

CMR ACHD Conclusions

- Ideal imaging tool for ACHD offering a one stop shop for form, function and flow
- Supplements echocardiography and avoids cardiac catheterization in many instances
- Knowledge of surgical procedure, anatomy and natural history of ACHD essential for accurate interpretation