Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia (ARVC/D)

Author: Teresa Castiello  
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Indications and Purpose of the Scan:

Hypertrophic cardiomyopathy (HCM) is the most common inherited cardiac condition with a prevalence of 1:500 in the general population. It is largely caused by mutations in genes encoding sarcomeric proteins, manifesting in left ventricular hypertrophy (LVH) in absence of secondary causes. The three main LV phenotypes are asymmetric (most common), concentric and apical. Typically, the asymmetric phenotype results in dynamic LV outflow-tract obstruction (LVOT) and define the obstructive variant (HOCM). The concentric phenotype is usually associated to non-inherited form, such as infiltrative disease, long standing hypertension - which are not enclosed in this short paper.

Patient presentation is phenotypically diverse, differing from asymptomatic to heart failure or sudden cardiac death.

**CMR is therefore requested:**

- To confirm the diagnosis suspected with medical history, ECG and other imaging modalities
- To distinguish between inherited and non-inherited, obstructive and non-obstructive
- For initial or subsequent evaluation of left/ right ventricular function (LVEF/ RVEF)
- To assess extension of late gadolinium enhancement (LGE)
- To measure maximum LV hypertrophy, opposed to contralateral wall and myocardial mass
- To evaluate diastolic dysfunction
- To evaluate “burned out” HCM and subsequent HF
- For prognostic evaluation (maximal LVH, HF, LGE)

Description:

Studies are typically performed according to standardized protocol. A typical HCM protocol takes about 40 minutes plus stress, if requested. Studies start with volume and function images, usually using steady-state free precession (SSFP) ECG gated cine imaging. Tissue characterization are the key sequences: late gadolinium enhancement images are most commonly done at the end of the scan. LGE typically identifies area of myocardial fibrosis, scar tissue after alcohol ablation or co-existing pathologies such as myocardial infarction. Additional sequences can be used, depending on clinical question. Stress images can be used to assess microvascular obstruction commonly co-localized with the area of maximal hypertrophy. Other sequences, such as oedema images (T2 STIR), myocardial blood velocities/flow, early gadolinium images, tagging (diastolic dysfunction) and, most recently, T1 and T2 mapping may be performed depending on suspected aetiology and referral request.

**Why CMR (Specific Advantages):**

CMR is very useful in the aetiology work up and can confirm the diagnosis. It can correctly distinguish acquired hypertrophic cardiomyopathy -including athletic heart- from familiar diseases, enabling appropriate requests for family screening. Although echocardiography is largely used as first line imaging modality, CMR is superior for segment visualization and tissue characterization. The higher spatial resolution and the tomographic imaging capability of CMR allow accurate measurements of LV wall thickness and myocardial mass. Long-axis cine bright blood imaging can identify systolic anterior movement of the anterior mitral valve apparatus in obstructive HCM. Tissue characterisation is distinctive to CMR and gives crucial information. Growing evidence show correlation between extension of LGE and poor prognosis. Myocardial fibrosis appears to be an arrhythmic substrate associated with sudden cardiac death (U-shaped correlation) and correlates with systolic and diastolic dysfunction. CMR can also provide information on congestion -for those who developed HF- and extra-cardiac finding. The CMR images are highly accurate and reproducible and do not expose the patient to ionic radiation.
Evidence:

1. European Heart Journal (2014) 35, 2733–2779 doi:10.1093/eurheartj/ehu284 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC). Authors/Task Force members: Perry M. Elliott* (Chairperson) (UK) Aris Anastasakis (Greece), Michael A. Borger (Germany), Martin Borggrefe (Germany), Franco Cecchi (Italy), Philippe Charron (France), Albert Alain Hagege (France), Antoine Lafont (France), Giuseppe Limongelli (Italy), Heiko Mahrholdt (Germany), William J. McKenna (UK), Jens Mogensen (Denmark), Petros Nihoyannopoulos (UK), Stefano Nistri (Italy), Petronella G. Pieper (Netherlands), Burkert Pieske (Austria), Claudio R apezzi (Italy), Frans H. Rutten (Netherlands), Christoph Tillmanns (Germany), Hugh Watkins (UK).


3. The ACCF/AHA and ESC guidelines describes diagnosis and management of cardiomyopathy, including diagnostic test and different images modality.


5. The consensus documents describe the application of CMR in HCM (page 2632).


These two reviews (4,5) describe clinical use, indication, advantages/disadvantages of CMR in HCM patients.

Contraindications:

- Implanted devices that are not MRI compatible are currently considered absolute contraindication in most Centres
- Intravascular clips or metallic prosthesis /object are not safe
- Inability to lie flat is a common contraindication in severe decompensated HF patient
- Claustrophobia, altered mental status prohibit the study
- eGFR <30mL/min unless risk-benefit analysis suggests otherwise
- Severe arrhythmias and inability to hold breath, affect quality images and represent relative contraindication to the study

References:

1. European Heart Journal (2014) 35, 2733–2779 doi:10.1093/eurheartj/ehu284 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC). Authors/Task Force members: Perry M. Elliott* (Chairperson) (UK) Aris Anastasakis (Greece), Michael A. Borger (Germany), Martin Borggrefe (Germany), Franco Cecchi (Italy), Philippe Charron (France), Albert Alain Hagege (France), Antoine Lafont (France), Giuseppe Limongelli (Italy), Heiko Mahrholdt (Germany), William J. McKenna (UK), Jens Mogensen (Denmark), Petros Nihoyannopoulos (UK), Stefano Nistri (Italy), Petronella G. Pieper (Netherlands), Burkert Pieske (Austria), Claudio R apezzi (Italy), Frans H. Rutten (Netherlands), Christoph Tillmanns (Germany), Hugh Watkins (UK).


