Cardiovascular Magnetic Resonance of Cardiomyopathies

Focus:
HCM, DCM and Inflammatory Disease

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Introduction

Cardiac Magnetic Resonance (CMR) has the capability to assess cardiac function, to visualize small myocardial lesions and to differentiate myocardial tissue changes. These tools offer outstanding possibilities in the diagnostic work-up of cardiomyopathies. We will focus on hypertrophic cardiomyopathy and dilated forms including inflammatory disorders. Depending on the issue under investigation a routine CMR examination will take between 30-60 minutes.

Steady state free precession (SSFP) sequences are applied in long axes to evaluate volumes and mass independent of the disease. The free choice of slice is a great advantage for visualizing small atypical localized lesions. For even better reproducibility of ejection-fraction, volumes and mass short axis multi-slice data sets have to be acquired. Within clinical studies the reduced standard deviation allows for smaller sample size. [1]

Furthermore a differentiation of myocardial tissue changes (e.g. assessment of edema, fibrosis and perfusion abnormalities) is possible applying a set of various sequences.[2, 3].

Based on our experience we advise to establish dedicated scan protocols for the assessment of the different diseases.

Dilated Cardiomyopathy

Most frequently the referring clinician asks for assessment of volumes and function. We quantify the dimensions and the function of left ventricle applying a set of SSFP movies (10 mm slice-thickness, no gap). Long axis will be added to assess the apical region. Concurrent mitral regurgitation and left atrial volume can be assessed in the same session.

Myocardial perfusion studies under vasodilator stress revealed impaired perfusion reserve in DCM despite macroscopically normal coronary arteries. [4] However, perfusion studies are not part of our routine protocol so far.

A subgroup of DCM patients feature pathologic patterns of delayed enhancement. [5] While the non-ischemic DCM may or may not feature a hyperintense “central line sign” in the middle of the myocardial wall, ischemic cardiomyopathy after myocardial
infarction is diagnosed based on subendocardial lesions within the coronary territories. With increasing experience it may be possible to elucidate various patterns of delayed enhancement characteristic of certain systemic diseases. [6]
Due to its excellent reproducibility CMR is the ideal tool for follow-up studies to monitor therapy. CMR is more sensitive for therapeutic effects on LV size and function in comparison to echocardiography.[7] Aldosterone antagonists have been shown to reduce fibrosis in animal models of DCM. [8] Human experimental studies may take advantage of CMR to assess fibrosis reduction.

Figure 1:

Dilated cardiomyopathy, delayed enhancement images show a bright signal in the middle layer and subepicardial, typical exclusion of the subendocardial region
Short axis view,
Left: steady state free precession
Right: T1-weighted gradient-echo inversion recovery sequence

Myocarditis
Dilated cardiomyopathy can develop from myocarditis caused by different agents (e.g. viral infection, toxic reaction, myocardial involvement in systemic disorders). As the clinical diagnosis is challenging, CMR is a useful tool to add information and to differentiate between reversible and irreversible lesions. Myocarditis can present with a variable pattern of impaired wall motion, tissue edema and inflammation and focal fibrosis. We suggest a comprehensive protocol including T2-weighted spin-echo imaging to detect myocardial edema [9] [10], T1-weighted spin-echo imaging for early contrast enhancement [11, 12] and inversion recovery gradient echo for delayed enhancement. [13] The multi-sequential approach increases sensitivity and specificity
in diagnosing acute myocarditis. [10] Even if this approach is a workhorse in several experienced centres, multi-centre trials are required.

Figure 2:
Myocarditis, bright spots in the lateral wall indicating fibrosis (delayed enhancement) long axis view, T1-weighted gradient-echo inversion recovery sequence (TI 250 ms)

Cardiac involvement in systemic disease
In several systemic diseases the prognosis is heavily influenced by the cardiac involvement. This presentation is focused on inflammatory diseases, however, CMR is able to visualize myocardial tissue changes even in Fabry’s disease (Moon) or in Amyloidosis. (Maceira, Circ 05)
The detection of myocardial injury was already possible in patients with preserved left ventricular function [14, 15]
In vasculitis like Churg-Strauss-syndrome (CSS) inflammation affects small myocardial vessels resulting in myocarditis with potentially impaired ventricular function. CMR can detect all kinds of reversible and irreversible myocardial damage (i.e. edema, effusion, inflammation, fibrosis) in CSS including delayed enhancement with a striking subendocardial pattern.

General advice for scanning dilative cardiomyopathies
- Full coverage of the left ventricle
- Assessment of long axis
to differentiate
- ischemic from non-ischemic cardiomyopathy: application of contrast-enhanced sequences
Hypertrophic Cardiomyopathy

CMR is superior in comparison to echocardiography in detecting small and uncommon localized hypertrophied regions. The high spatial resolution allows the visualization of small abnormalities including findings affecting the papillary muscle. [16]

The evaluation of the apical region is a challenge for echocardiography. Due to the fact, that the apical forms of HCM are not uncommon, a reliable diagnosis is of high clinical impact. Applying CMR morphological changes of the apical region could be detected, improving the understanding of the disorder. [17]

The localization of the obstruction is possible by detecting the signal void due to turbulent flow by applying gradient echo sequences, usually SSFP. The left-ventricular outflow tract (LVOT) can be reliably measured by quantification of the smallest systolic area of the LVOT. This technique showed a good relation to the clinical outcome after septal artery embolisation (TASH)[18] and it is possible to provide cut-off values to differentiate the degree of obstruction. (Schulz-Menger et al, abstract ACC 2005)

Contrast-enhanced techniques can be used to follow the induced myocardial infarction in the patients after septal artery embolization. [19] (figure 3)

Interestingly, contrast-enhancement can also be detected in patients without interventions. These findings seems to have a relation to the patients risk in retrospective analysis [20], but are also detectable in asymptomatic patients [21].
Figure 3
Scarring in hypertrophic cardiomyopathy after septal artery embolization, bright signal in the septal wall (delayed enhancement)
Short axis view, T1-weighted gradient-echo inversion recovery sequence (TI 250 ms)

General advice for scanning hypertrophic cardiomyopathies
- Full coverage of the left ventricle
- Assessment of long axis
- Visualization of signal void (obstruction)
- Quantification of obstruction
- Application of contrast-enhanced sequences

References
3. Dymarkowski, S., et al., Value of t2-weighted magnetic resonance imaging early after myocardial infarction in dogs: comparison with bis-gadolinium-


