

Introduction

The cardiomyopathies include a variety of disease where the primary pathology directly involves the myocardium. Although ischemic CMP is the most common cause of heart failure, ischemic CMP is not appropriate term because the primary pathology is in the coronary arteries and not the heart muscle. Cardiac MR (CMR) is proving increasingly valuable in the identification and management in these conditions. This exhibit will discuss the merit and the potential role of CMR in the evaluation of various non-ischemic cardiomyopathies.

MR Techniques for Assessment of Cardiomyopathy

The diagnosis of CMP is established by exclusion of other cardiovascular etiologies and an accurate characterization of the phenotype. Treatment is guided by the stage and hemodynamic relevance of the disease and long-term follow-up after therapy is needed. Thus, imaging techniques are important in the diagnosis and therapy of cardiomyopathies.

- SSFP sequence: morphologic and functional information VENC:
- VENC: evaluation of diastolic and valvular function
- DE-MRI: identification of myocardial necrosis and fibrosis
- Myocardial perfusion MR: presence or extent of inducible ischemia
- Spin-Echo images (T1-, T2-weighted images): identification of signal change of myocardium
- MR spectroscopy: for the evaluation of metabolic state

Clinical Impact of Cardiac MRI

Dilated Cardiomyopathy

DCM is characterized by progressive dilatation of the LV or biventricular enlargement with loss of contractile function

- The main target of MRI
 - Differentiation from an ischemic origin (DE-MRI)
 - Prediction of functional improvement (DE-MRI)
- Advantage of CMR
 - Morphology and function: clearly delineated
 - Superior depiction of dilatation of the RV
 - Delay enhancement MRI (DE-MRI)
 - * no enhancement in a majority
 - * only mid-myocardium in a non-coronary pattern in some patients: prognosis is poor
 - * The degree of enhancement: correlates with the severity of functional abnormality.

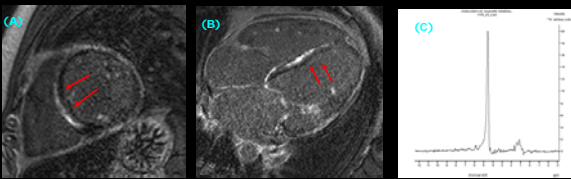


Figure 1. Enhancement type on DE-MRI (Alcoholic CMP) 36 year old man with dyspnea. Cine MR showed globally reduced systolic function (EF = 10.6%). DE-MRI view shows delayed enhancement at mid and epicardial area of septal wall with non-coronary pattern, on short axis view (A) and 4 chamber view (B). MR Spectroscopy (C) was performed at septal wall and showed depletion of creatine metabolism. Function was not improved during the follow-up period.

- DE-MRI for Ischemic CMP vs non-ischemic CMP

- * HF with CAD: Subendocardial or Transmural
- * HF related DCM:
 - no enhancement (59%)
 - subendocardial or transmural (13%)
 - Patchy of longitudinal striation of midwall (28%)

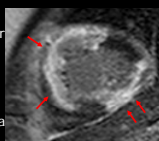


Figure 2. Typical findings of DE-MRI in patient with ischemic CMP DE-MRI shows strong enhancement along the LAD and RCA vascular territory.

Hypertrophic Cardiomyopathy

HCM is characterized by Myocardial hypertrophy with impaired diastolic and systolic function (mainly diastolic dysfunction) due to myocardial disarray as well as patches of myocardial scarring

- The main target of MRI
 - To determine phenotypes such as apical form (Cine MR using SSFP sequences)
 - To assess regional myocardial hypertrophy (Cine MR using SSFP sequences)
- Advantage of CMR
 - Precise definition of the site and extent of hypertrophy, especially LV apex (apical HCM)
 - Accurate assessment of flow dynamics of LV outflow tract
 - Demonstration of myocardial scarring and fibrosis: predominantly in the middle third of the ventricular wall

**** The extent of hyperenhancement on DE-MRI may have prognostic implications for the risk of progressive ventricular dilation and sudden death**

- Evaluation of post-surgical change
- Monitoring and quantification after septal ablation

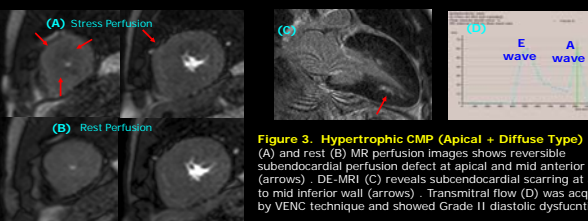


Figure 3. Hypertrophic CMP (Apical + Diffuse Type) Stress (A) and rest (B) MR perfusion images shows reversible subendocardial perfusion defect at apical and mid anterior wall (arrows). DE-MRI (C) reveals subendocardial scarring at apico to mid inferior wall (arrows). Transmittal flow (D) was acquired by VENC technique and showed Grade II diastolic dysfunction.

Restrictive Cardiomyopathy

DCM is characterized by Restrictive filling and reduced diastolic size of either and both ventricles with normal or near-normal systolic function

- The main target of MRI
 - To determine phenotypes such as myocardial infiltrative disease (Spin-echo Images, DE-MRI)
 - To differentiate from constrictive pericarditis (Cine MR using SSFP sequences)
- Advantage of CMR
 - Clearly depict the anatomic and functional abnormalities
 - Define myocardial infiltrative disease such as amyloidosis on the basis of typical findings on DE-MRI
 - Visualization of pericardial thickness
 - Objective monitoring and quantification after treatment

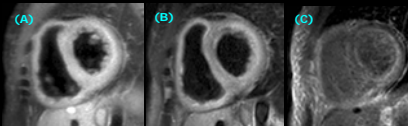


Figure 4. Restrictive CMP due to amyloidosis T1-weighted (A) and T2-weighted (B) MR image shows diffuse high signal intensity at entire myocardium of right and left ventricle. Also, thickness of right ventricle is increased up to 10 mm. DE-MRI (C) demonstrates global and subendocardial enhancement at right and left ventricle.

Arrhythmogenic RV dysplasia (ARVD)

- Advantage of CMR
 - Regional thinning and wall-motion abnormality of right ventricle: clearly delineated
 - Detailed differentiation between myocardium, epicardial fat, trabeculae and myocardial fatty infiltration
 - Delay enhancement MRI (DE MRI): noninvasive detection of myocardial fibrotic changes

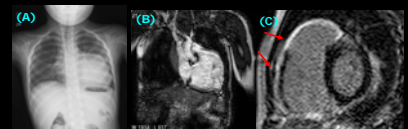


Figure 5. A 19-month-old patient with abnormal cardiac border. Chest PA (A) shows abnormal enlarged left heart border. Cine MRI (B) reveals global right ventricular dilatation with hypokinesia. DE-MRI (C) nicely demonstrates diffuse thinning and dilatation with the strong enhanced wall of the right ventricle (arrows) suggesting extensive fibrosis.

Stress-Induced Cardiomyopathy

- Advantage of CMR
 - accurate evaluation of hypokinesia at apico to mid entire wall with hypercontractile basal entire wall
 - Identification of myocardial injury

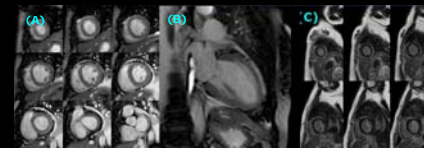


Figure 6. A 45-year-old female with Stress-induced Cardiomyopathy. She had experienced psychological stress, and also suffered from severe rhabdomyolysis with ARF. Cine MRI on short axis view (A) and 2 chamber view shows hypokinesia at apico to mid entire wall, but contractility at basal level is well preserved. Note that there is no enhancement on DE-MRI (C). Conventional coronary angiography revealed no significant steno-occlusive lesion.

Non-compaction

Non-compaction is characterized by prominent trabeculation and recess and noncompact/compact layer > 2.0 on end-systolic phase.

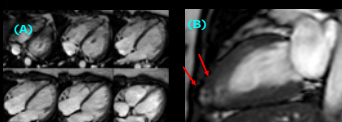


Figure 7. A 35-year-old female with LV non-compaction. Cine MRI on 4 chamber view (A) and 2 chamber view (B) shows prominent trabeculation and recess at apex, but myocardial contractility is well preserved. End-systolic image is nicely demonstrated non-compaction layer at apex (arrows).

Diverticulum

- Morphology: sacular with narrow neck, Location: apex or basal
- Two types
 - muscular type: sacular with narrow neck, contractility (+), DE-MRI (-).
 - fibrous type: contractility (+), DE-MRI (+).

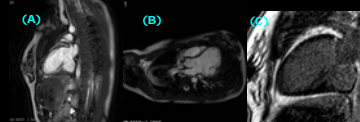


Figure 8. A 5-year-old boy with LV diverticulum. On routine echocardiography, LV aneurysm was suspected. Cine MRI (A,B,C) shows focal outpouching lesion at basal inferior wall with narrow neck and sacular shape. Note this lesion is normal contracted during cardiac cycle. There is no enhancement (D) at LV diverticulum on DE-MRI (arrows) suggesting muscular type.

Myocarditis

- DE-MRI
 - enhancement predominantly in lateral wall (associated with active inflammation)
 - enhancement in 88% of patients with myocarditis
 - follow-up: decreased extent of enhancement
- T2WI: High signal intensity on involve myocardium

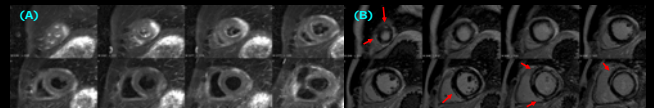


Figure 8. A 33 year-old women with acute chest pain, T2 E1 MRI (A) showed multifocal high signal intensity at apex, apical lateral wall, mid inferoseptal and inferior wall, and basal anteroseptal, anterolateral wall, and inferoseptal wall with non-coronary pattern. DE-MRI (B) also showed hyperenhancement on corresponding area.

Conclusion

The understanding of various cardiomyopathies and knowledge of characteristic MR findings is provided more valuable information for the accurate diagnosis and proper management. With the advances of MRI technology and, it will more increase the role MRI for the assessment of various cardiomyopathies.