The Role of MR Imaging in Various Cardiomyopathy

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Introduction

Definition and Classification of Cardiomyopathy (CMP)

MR Technique for Assessment of CMP

Clinical Impact of Cardiac MRI

- Dilated CMP
- Hypertrophic CMP
- Restrictive CMP
- Arrhythmogenic right ventricular dysplasia (ARVD)
- Specific CMP
  (Stress-induced CMP, Non-compaction, Diverticulum, Myocarditis)

Conclusion
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- Conclusion
The cardiomyopathy (CMP) include a variety of disease where the primary pathology directly involves the myocardium.

Cardiac MR (CMR) is proving increasingly valuable in the identification and management in these conditions.

We will illustrate the various MR techniques for the evaluation of CMP and characteristic MR findings. This exhibit will discuss the merit and the potential role of CMR in the evaluation of various CMP.
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• Conclusion
The cardiomyopathies (CMP) constitute a group of disease in which dominant feature is direct involvement of the heart muscle itself.

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.

In the WHO/ISFC classification, the cardiomyopathies are classified based on their predominant pathophysiologica features (Table 1).
### Definition and Classification

<table>
<thead>
<tr>
<th>Disorder</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dilated cardiomyopathy</td>
</tr>
<tr>
<td>Hypertrophic cardiomyopathy</td>
</tr>
<tr>
<td>Restrictive cardiomyopathy</td>
</tr>
<tr>
<td>Arrhythmogenic right ventricular cardiomyopathy</td>
</tr>
<tr>
<td>Unclassified cardiomyopathy</td>
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</table>

**Specific cardiomyopathy**

<table>
<thead>
<tr>
<th>Disorder</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ischemic cardiomyopathy</td>
</tr>
<tr>
<td>Valvular cardiomyopathy</td>
</tr>
<tr>
<td>Hypertensive cardiomyopathy</td>
</tr>
<tr>
<td>Inflammatory cardiomyopathy</td>
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<tr>
<td>Metabolic cardiomyopathy</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Disorder</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peripatal cardiomyopathy</td>
</tr>
<tr>
<td>Neuomuscular disorder</td>
</tr>
<tr>
<td>Sensitivity and Toxins</td>
</tr>
<tr>
<td>Muscular dystrophies</td>
</tr>
<tr>
<td>General systemic disease</td>
</tr>
</tbody>
</table>

**Table 1. WHO/ISFC Classification of Cardiomyopathies**
Three basic types of functional impairment have been described (Table 2).

- Dilated CMP
- Hypertrophic CMP
- Restrictive CMP

Most of specific cardiomyopathies are characterized by the dilated CMP pattern.

Ischemic CMP has been used to describe condition in which coronary artery disease causes multiple infarctions, diffuse fibrosis, and left ventricular dysfunction.
<table>
<thead>
<tr>
<th>Functional Classification of the Cardiomyopathies</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Dilated</strong></td>
</tr>
<tr>
<td>Most common form (60%)</td>
</tr>
<tr>
<td>Ventricular dilatation</td>
</tr>
<tr>
<td>Contractile dysfunction</td>
</tr>
<tr>
<td>Often symptoms of congestive heart failure</td>
</tr>
</tbody>
</table>

*Table 2. Functional Classification of the Cardiomyopathies*
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      - (Stress-induced CMP, non-compaction, diverticulum, Myocarditis)
- Conclusion
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.
MR Techniques for Assessment of CMP

- SSFP sequence:
  morphologic and functional information (Table 3).
- VENC:
  evaluation of diastolic and valvular function
- DE-MRI:
  identification of myocardial necrosis and fibrosis
- Myocardial perfusion MR:
  presence or extent of inducible ischemia
- Spin9Echo images (T1-, T2-weighted images):
  identification of signal change of myocardium
- MR spectroscopy: for the evaluation of metabolic state
### MR Techniques for Assessment of CMP

<table>
<thead>
<tr>
<th>Morphology</th>
<th>Dilated</th>
<th>Hypertrophic</th>
<th>Restrictive</th>
</tr>
</thead>
<tbody>
<tr>
<td>LV and RV size</td>
<td>+++</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Hypertrophy</td>
<td>+</td>
<td>+++</td>
<td>+/-</td>
</tr>
<tr>
<td>Atrial dilatation</td>
<td>++</td>
<td>+</td>
<td>++</td>
</tr>
<tr>
<td>Pleural effusion</td>
<td>+</td>
<td>-/+</td>
<td>+</td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td>+</td>
<td>-/+</td>
<td>+</td>
</tr>
<tr>
<td>ventricular thrombus</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>SVC/IVC dilatation</td>
<td>+</td>
<td>-</td>
<td>+++</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Myocardial function</th>
<th></th>
<th></th>
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</thead>
<tbody>
<tr>
<td>Global dysfunction</td>
<td>+++</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Segmental dysfunction</td>
<td>+</td>
<td>+++</td>
<td>+</td>
</tr>
<tr>
<td>Diastolic dysfunction</td>
<td>+</td>
<td>++</td>
<td>+++</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Valvular function</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Mitral regurgitation</td>
<td>+</td>
<td>++</td>
<td>+</td>
</tr>
<tr>
<td>Tricuspid regurgitation</td>
<td>+</td>
<td>-</td>
<td>+</td>
</tr>
</tbody>
</table>

Table 3. Morphologic and Functional Abnormalities of the CMP
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Conclusion
Clinical Impact of Cardiac MRI

• Recently, the use of MRI for the evaluation of CMP is expanding, aided by the administration of paramagnetic contrast agents.

• Cardiac MRI offers the accurate evaluation of morphology and function, and it also offers characterization of various CMP.

• Suspected myocardial ischemia and fibrosis are also diagnosed by using dynamic first-pass and delayed-enhancement MRI.
Table 4. Current indication of cardiac MRI for the evaluation of CMP

<table>
<thead>
<tr>
<th>Indication</th>
<th>Class</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dilated cardiomyopathy:</td>
<td></td>
</tr>
<tr>
<td>Differentiation from dysfunction related coronary artery disease</td>
<td>I</td>
</tr>
<tr>
<td>Hypertrophic cardiomyopathy:</td>
<td></td>
</tr>
<tr>
<td>Apical</td>
<td>I</td>
</tr>
<tr>
<td>Non-apical</td>
<td>II</td>
</tr>
<tr>
<td>Restrictive cardiomyopathy:</td>
<td>II</td>
</tr>
<tr>
<td>Arrhythmogenic RV dysplasia</td>
<td>II</td>
</tr>
<tr>
<td>Non compaction</td>
<td>II</td>
</tr>
</tbody>
</table>

*** Classification

Class I: provides clinically relevant information and is usually appropriate, may be used as first line imaging technique; usually supported by substantial literature

Class II: provides clinically relevant information and is frequently useful; other techniques may provide similar information; supported by limited literature

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Dilated CMP

- Dilatation and impaired contraction of left ventricle
- Cause:
  - idiopathic, familial/genetic, viral, immune, alcoholic/toxic
- Histological hallmark:
  - progressive interstitial fibrosis with a numerical decrease of myocyte
- The main target of MRI
  - Differentiation from an ischemic origin (DE-MRI)
  - Prediction of functional improvement (DE-MRI)
Dilated CMP

• 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. No enhancement type on DE-MRI 39 year old man with chest pain. (A) Cine MR showed globally reduced systolic function (EF = 39.3%). (B) DE-MRI shows no delayed enhancement. (C) MRI permits accurate measurement of cardiac function and LV mass. (D) Follow-up cine MRI after 3 month, wall motion is normalized (EF = 52.9%) on cine MRI.
Figure 2. Enhancement type on DE-MRI (Alcoholic CMP) 36 year old man with dyspnea. (A) 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 (B) and 4 chamber view (C). MR Spectroscopy (D) was performed at septal wall and showed depletion of creatine metabolism. Function was not improved during the follow-up period.
Figure 3. Extensive Enhancement on DE-MRI 65 year old man with dyspnea. Severe global systolic dysfunction (EF = 29.3%) with severe dilated LV (A) (B) DE-MRI view shows extensive delayed enhancement at apico to basal anterior and anteroseptal wall suggesting extensive fibrosis. This finding is hallmark of end-stage of dilated cardiomyopathy.
Dilated vs Ischemic CMP

- DE-MRI
  - HF with CAD:
    - Subendocardial or Transmural
  - HF related DCMP:
    - No enhancement (59%)
    - Subendocardial or transmural (13%)
    - Patchy of longitudinal striae of midwall (28%)


Figure 4. Typical findings of DE-MRI in patient with ischemic CMP
DE-MRI shows strong enhancement along the LAD and RCA vascular territory with transmural extent (75-100%).
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Conclusion
Hypertrophic CMP

- Myocardial hypertrophy with impaired diastolic and systolic function (mainly diastolic dysfunction)
- Narrowing of the LVOT in obstructive cases
- Histology:
  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 5. **Apical hypertrophic CMP** (A) ECG shows giant negative T-wave (arrows) suggesting anterolateral wall ischemia. Cine MRI (B) nicely demonstrates thickening of myocardium (“Spade Ace sign”) at apex and apical segment. DE-MRI on 4 chamber view (C) shows patch enhancement with multiple foci at hypertrophied myocardium (arrows).
Hypertrophic CMP

Figure 6. Hypertrophic CMP (Apical + Diffuse Type)
Cine MRI on short axis (A) and 2 chamber (B) shows diffuse thickening of myocardium. Color mapping (C) using semiquantitative measurement well visualized diffuse wall thickness at apex, apico to basal anterior and septal wall, and apico to mid inferior and lateral wall. Cine MRI on LVOT view shows no significant obstruction at left ventricular outflow tract.
Figure 6. Hypertrophic CMP (Apical + Diffuse Type) Stress (D) and rest (E) MR perfusion images shows reversible subendocardial perfusion defect at apical and mid anterior wall (arrows). DE-MRI (F) reveals subendocardial scarring at apico to mid inferior wall (arrows).
Figure 6. Hypertrophic CMP (Apical + Diffuse Type) Transmitral flow (G) was acquired by VENC technique and showed pseudonormalization pattern (Grade II diastolic dysfunction). MR spectroscopy (H) show markedly decreased creatine metabolism.
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Restrictive CMP

- Restrictive filling and reduced diastolic size of either and both ventricles with normal or near-normal systolic function
- Hallmark of restrictive CMP: Abnormal diastolic dysfunction
- Cause: idiopathic or associated with other disease (amyloidosis, sarcoidosis etc)
- 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)
Restrictive CMP

- 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
Restrictive CMP: Amyloidosis

- A cause of restrictive CMP
- primary or secondary in origin. Infiltration with fibrillar
- MR findings:
  - Variable, but usually increased SI on T1, T2 WI
  - Thickening of Interatrial septum and right atrial wall thickness (myocardial thickness > 6 mm):
    DDx point of symmetric hypertrophic CMP
    *Fattori et al, Am Heart J 1998*

- DE-MRI:
  - Global and subendocardial enhancement (69%):
    Increased LV and RV mass index, lower LV EF (57%)
    *Maceira AM et al Circulation 2005*
Figure 7. Restrictive CMP due to amyloidosis  
Cine MRI (A) shows mild depressed systolic function (EF = 48.2%). T1-weighted (B) and T2-weighted (C) 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 (D) demonstrates global and subendocardial enhancement at right and left ventricle.
Restrictive CMP vs Constrictive CMP

Restrictive CMP

- Increased wall thickness
- Delayed ventricular filling
- Frequently combined MR, TR

Constrictive CMP

- Pericardial thickening (> 4mm)

*Masui et al, Radiology 1992*
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Arrhythmogenic RV dysplasia (ARVD)

- Ventricular tachycardia with LBBB from right ventricle
- Young males, variable symptoms
- The definite diagnosis is challenging
- Histology:
  - Fibro-fatty infiltration with thinning of RV free wall
- The main target of MRI
  - Explicit demonstration of regional thinning and fatty infiltration of right ventricular wall (Spin-echo, SSFP)
  - Demonstration of myocardial fibrosis at right ventricular wall (DE-MRI)
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

Tandri et al, J Am Coll Cardiol 2005
Figure 8. T1-weighted MRI in patient with Arrythmogenic right ventricular dysplasia.

Axial MR image obtained at the midventricular level shows myocardial wall thinning and fatty infiltration (arrows) at the right ventricular free wall and also there is global right ventricular dilation. The interventricular septum shows convexity to the left ventricular side.
Figure 9. 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, D) nicely demonstrates diffuse thinning and dilation with the strong enhanced wall of the right ventricle (arrows) suggesting extensive fibrosis.
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Stress-induced CMP ("Tako-tsubo")

• Definition: acute onset of a cardiovascular event, usually associated with substernal chest pain, initially regarded as ST-segment elevation myocardial infarction/evolving coronary syndrome.

• Systolic dysfunction (ejection fraction 29±9%), predominantly characterized by akinesia/hypokinesia of the mid-to-distal portion of the LV chamber "apical ballooning", with hypercontractile basal LV

• Absence of significant atherosclerotic luminal narrowing in each of the 3 epicardial coronary arteries (0 to < 25%).
Stress-induced CMP (“Tako-tsubo")

- Profound psychological stress immediately preceding and triggering the cardiac events.

- Possible mechanism: catecholamine-mediated cardiotoxicity, in which the distal LV chamber is selectively vulnerable to a form of myocardial stunning.

- DE-MRI: no enhancement except focal infarction at apex (5%)

*Sharkey et al, Circulation 2005*

**Figure 10.** “Tako-tsubo" cardiomyopathy describes the resemblance of the LV angiogram to an octopus trap.
Stress-induced CMP (“Tako-tsubo”)

Figure 11. 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

- Prominent trabeculation and recess
- Non-compact/compact layer > 2.0 on end-systolic phase
- Autosomal dominant inheritance.
- Failure of normal embryonic development of the myocardium from loosely arranged muscle fibers to the mature compacted form of myocardium.
- Microvascular dysfunction and ventricular arrhythmias.
Figure 12. 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: saccular with narrow neck
- Location: apex, basal
- Two types
  - muscular type: saccular with narrow neck, contractility (+), DE-MRI (-).
  - fibrous type: contractility (+), DE-MRI (+).
- Differential diagnosis
  - True Aneurysm: Thin-walled with wide opening contractility (akinetic or dyskinetic), DE-MRI (+).
  - Pseudoaneurysm: saccular with narrow neck, but contractility (akinetic or dyskinetic), DE-MRI (acute stage - ?, chronic stage +).
Figure 13. 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 saccular 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

- Myocardial inflammation caused by
  - viral or postviral autoimmune response (primary)
  - specific pathogen such as bacteria, drug, chemical etc (secondary)

- DE-MRI
  - enhancement predominantly in lateral wall
    (associated with active inflammation)
  - enhancement in 88% of patients with myocarditis
  - follow-up: decreased extent of enhancement

Mahrholdt et al. Circulation 2004
Figure 14. 51 year-old male with ventricular tachycardia due to myocarditis. T1-weighted (A) and T2-weighted (B) MRI show ill-defined low signal area at inferior and inferolateral wall. DE-MRI (C) shows abnormal enhancement at multifocal area (arrows). Enhancement pattern is non-vascular territory and non-subendocardial dominant area suggesting sequaleae of myocarditis.
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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 cardiomyopathy.