MRI of Cardiomyopathies: Scan protocols, imaging findings and key features

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Authors: M.-Y. Ng¹, S. Kumar¹, C. K. Liew¹, A. Jones², R. W. Bury¹;
¹Blackpool/UK, ²Manchester/UK
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Learning objectives

1. Describe the classification of the cardiomyopathies according to the European Society of Cardiologists (ESC)
2. Present and explain MRI protocols to best optimise imaging of the different cardiomyopathies
3. Describe and illustrate the MR imaging features of each cardiomyopathy
4. Highlight key features which are helpful for the cardiologist and establishing a diagnosis

Background

There are two different classifications for cardiomyopathies. One from the American Heart Association and the other from the European Society of Cardiology (ESC). For the purposes of this poster, we have opted to use the European Heart Association classification.

Cardiomyopathy is defined by the European Society of Cardiology as a myocardial disorder in which the heart muscle is structurally and functionally abnormal, in the absence of coronary artery disease, hypertension, valvular disease and congenital heart disease sufficient to cause the observed myocardial abnormality. Based on the ESC classification, there are 5 main groups of cardiomyopathies (Fig 1). They are:

1. Hypertrophic cardiomyopathy
2. Dilated cardiomyopathy
3. Arrhythmogenic right ventricular cardiomyopathy
4. Restrictive cardiomyopathy
5. Unclassified cardiomyopathy.

Unclassified cardiomyopathies includes Tako-Tsubo cardiomyopathy and left ventricular non-compaction cardiomyopathy.

Understanding cardiomyopathies is important as they are relatively common requests for Cardiac MRI. It is important to understand the imaging features characteristic of each cardiomyopathy and the features which are useful to a cardiologist for ultimate management.
Images for this section:

**Fig. 1:** Classification based on the European Society of Cardiology.
1. **Hypertrophic Cardiomyopathy:**
   - Relatively common condition affecting 1 in 500 people
   - Autosomal dominant inheritance.
   - Characterised by hypertrophy of the left ventricular wall particularly the septum (Fig 2) but there is variation in the areas of the left ventricle affected by the hypertrophy.

   **Important features to interrogate the images for are:**

   1. Left ventricular hypertrophy (Left ventricular wall thickness # 15mm)
   2. Late gadolinium enhancement (also known as delayed enhancement) characteristically seen in the mid wall of the hypertrophied segments (Fig 3). This is seen in up to 80% of patients with left ventricular hypertrophy.
   3. Systolic anterior motion which is the movement of the anterior mitral leaflet anteriorly during systole. This is thought to be caused by a Venturi effect. This finding confirms obstruction.

   **Additional imaging sequences:**

   1. Long axis 3 chamber phase contrast images to look for aliasing artefact in the ascending aorta at 250cm/s (Fig 4).
   2. Three-chamber left ventricular outflow tract cine to look for systolic anterior motion.

   **Note:**

   Fabry’s Disease (Fig 5 & 6) is a mimicker of hypertrophic cardiomyopathy and should be kept in mind especially if the patient is male as this is an X-linked recessive disorder. However, Fabry’s Disease does occur in female carriers also but less frequently. Fabry’s disease is an important diagnosis to consider as it is potentially a treatable cause of hypertrophic cardiomyopathy with enzyme replacement therapy.

2. **Dilated Cardiomyopathy:**
   - Very common cardiomyopathy of which 50% are idiopathic.
   - Causes include coronary artery disease, infective myocarditis, toxins (eg. alcohol, chemotherapy), autoimmune conditions (eg. scleroderma,
systemic lupus erythematosus, sarcoidosis) and endocrine conditions (eg. thyrotoxicosis).3.

Imaging features:

1. Dilatation of the left ventricle (LV) or biventricular enlargement (Fig 7). (Note: Lack of right ventricular dilatation and dysfunction does not exclude the diagnosis of dilated cardiomyopathy)
2. Reduced systolic function but normal diastolic function.
3. Global hypokinesia of the left ventricle

Useful tip:

Look for ischaemia/ coronary artery disease as this is a treatable cause for dilated cardiomyopathy. Cardiac MRI plays an important and useful role here. On the delayed gadolinium enhancement images, ischaemia would show subendocardial enhancement in a vascular territory (Fig 8) whereas myocarditis or an idiopathic cause would show mid wall enhancement (Fig 9) or no enhancement at all.

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC):

• Uncommon cardiomyopathy
• Predominantly inherited in an autosomal dominant disease with incomplete penetrance.3
• Characterised by fatty or fibro-fatty infiltration of the right ventricle but this has been shown to also extend into the left ventricle in some cases.
• It is a well known cause of sudden cardiac death in young people.

In 2010, the criteria for diagnosing ARVC was updated. We have picked out the relevant criteria pertinent to Cardiac MRI.

Major Imaging Criteria for Cardiac MRI (Fig 10 & 11):

• Regional RV akinesia, dyskinesia, or dyssynchronous RV contraction

AND

• Ratio of RV end-diastolic volume to BSA #110 mL/m² (male) or #100 mL/m² (female)
• RV ejection fraction #40%
• Minor Imaging Criteria for Cardiac MRI:
• Same as above except ratio of RV end-diastolic volume to BSA #100 to
  <110mL/m² (male) or #90 to <100 mL/m² (female)
• RV ejection fraction >40% to #45%

**Restrictive Cardiomyopathy:**

• Least common of the four main cardiomyopathies. This is characterised by rigid walls and the heart is restricted from stretching and filling with blood properly.
• Most common cause of restrictive cardiomyopathy outside the tropics is amyloidosis (Fig 12 & 13).
• Other causes include idiopathic, familial, sarcoidosis, haemochromatosis, eosinophilia and endomyocardial fibrosis (Fig 14).
• Important purpose of cardiac MRI is to help differentiate between restrictive cardiomyopathy and constrictive pericarditis.

**Key Imaging Features:**

1. Normal systolic function but impaired diastolic function (ie. normal ejection fraction but decreased end diastolic volume)
2. Dilated atria but normal sized ventricles (Fig 12 & 13)
3. Biventricular thickening

<table>
<thead>
<tr>
<th>Restrictive Cardiomyopathy</th>
<th>Constrictive Pericarditis</th>
</tr>
</thead>
<tbody>
<tr>
<td>No indentation of the ventricular cavities</td>
<td>Indentation of the ventricular cavities or tubular appearance to the ventricular cavities</td>
</tr>
<tr>
<td>No pericardial thickening</td>
<td>Pericardial thickening</td>
</tr>
<tr>
<td>Prominent atrial enlargement</td>
<td>Less prominent atrial enlargement</td>
</tr>
<tr>
<td>Myocardial thickening</td>
<td>Normal myocardial thickness</td>
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</tbody>
</table>

**Restrictive Cardiomyopathy vs Constrictive Pericarditis**
Additional Imaging Sequences:

1. Free breathing short axis bright blood cine to look for paradoxical movement of interventricular septum which is seen in constrictive pericarditis.

Amyloidosis:

The most common cause of amyloidosis is primary amyloidosis. The typical appearances of amyloidosis is of a restrictive cardiomyopathy with diffuse subendocardial enhancement on late gadolinium enhancement images (Fig 13).

Unclassified Cardiomyopathies:

There are many unclassified cardiomyopathies. In this poster we will demonstrate two types which are Tako-tsubo Cardiomyopathy and peripartum cardiomyopathy.

Tako-Tsubo Cardiomyopathy:

- Commonly seen in women >50yrs old.
- Aetiology is not fully understood
- Commonly, there is a recent history of emotional or physical stress.
- Characterised by acute but rapidly reversible LV systolic dysfunction typically giving the appearance of a Tako-Tsubo (Japanese for octopus trap) (Fig 15 & 16).
- No atherosclerotic coronary artery disease

Imaging Features:

1. Apical hypokinesia (Fig 15)
2. Myocardial oedema in the acute phase
3. Lack of late gadolinium enhancement helps differentiate this from other cardiomyopathies

Left Ventricular Non-compaction Cardiomyopathy:

- Characterised by prominent left ventricular trabeculae and deep intertrabecular recesses
- Involves predominantly the apical region of the left ventricle
- Aetiology is an arrest in the compaction of the subendocardial myocardium
• Associated with other cardiac abnormalities such as anomalous coronary arteries, and cyanotic congenital heart disease.

**Imaging features:**

1. Prominent trabeculae particularly in the left ventricular apex
2. Non-compacta to compacta ratio >2.3

**Peripartum Cardiomyopathy:**

• Rare and potentially fatal
• Occurs in the last month of pregnancy up to 6 months after delivery
• Usually occurs in the postpartum period with about 45% within the first week and 75% within the first month
• Symptoms of heart failure
• Diagnosis of exclusion/ other causes of heart disease ruled out
• ~50% of patients achieve complete or almost complete recovery

**Imaging features:**

1. No specific imaging features
2. Left ventricular dysfunction (Fig 18 & 19)

**Useful Sequences:**

1. Early perfusion images to look for thrombus which is a common complication of peripartum cardiomyopathy

**Images for this section:**
Fig. 1: Classification based on the European Society of Cardiology.
**Fig. 2:** Hypertrophic obstructive cardiomyopathy. Left ventricular outflow tract (LVOT) view showing focal septal thickening, turbulent flow in the aorta and systolic anterior motion of the mitral valve.

![Image of LVOT view showing focal septal thickening, turbulent flow in the aorta and systolic anterior motion of the mitral valve.]

**Fig. 3:** Hypertrophic cardiomyopathy. Delayed enhancement short axis view showing enhancement at the RV insertion point of the antero-septal wall (arrow).
**Fig. 4:** Same patient as figure 2. Phase contrast image at 250cm/sec shows aliasing artefact (arrow) which confirms clinically significant LVOT obstruction. Aliasing artefact disappears on phase contrast image at 350cm/sec.
**Fig. 5:** 4 chamber cine of a patient with known Fabry's Disease. There is a moderate pericardial effusion with no evidence of constriction. Borderline left atrial enlargement.
Fig. 6: Fabry’s Disease. Delayed enhancement short axis image showing enhancement in the anterior, lateral and inferior walls. Enhancement of the inferolateral wall is characteristic of Fabry’s disease.
Fig. 7: Dilated Cardiomyopathy due to ischaemia. 4 chamber view showing severe LV and RV dilatation and poor contractility of both ventricles. The atria are both dilated.
Fig. 8: Dilated cardiomyopathy due to ischaemia. Delayed gadolinium enhancement sequence showing high signal in the LAD and circumflex artery territories with subendocardial involvement typical of myocardial infarction.
**Fig. 9:** Dilated cardiomyopathy in another patient showing subtle midwall enhancement (arrows). This is characteristic of myocarditis or an idiopathic cause.
**Fig. 10:** Arrhythmogenic right ventricular cardiomyopathy (ARVC) on RVOT view which demonstrates hypokinesia and dilatation of the right ventricle which are common features of ARVC.
Fig. 11: Arrhythmogenic Right Ventricular Cardiomyopathy on 4 chamber view showing a dilated right ventricle with hypokinesia.
**Fig. 12:** 4 chamber showing restrictive cardiomyopathy due to amyloidosis. There is diastolic dysfunction with dilated atria bilaterally and normal sized ventricles.
**Fig. 13:** Delayed enhancement images in the same patient in figure 12 with amyloidosis. There is characteristic diffuse subendocardial enhancement. The atria are also grossly dilated which is the consequence of the restrictive cardiomyopathy caused by amyloidosis.
**Fig. 14:** Eosinophilic Cardiomyopathy leading to endomycocardial fibrosis on 4 chamber view cine. There is loss of the normal trabecula pattern and the right ventricular apex is filled out. There is impairment of the systolic function particularly of the right ventricle.
**Fig. 15:** Tako-Tsubo Cardiomyopathy on 2 chamber view. There is good basal contraction but the left ventricular apex is akinetic creating the appearance of a Tako-Tsubo. Echo images subsequently showed improvement in the left ventricular function and restoration of the left ventricular apex contractility.
**Fig. 16:** Tako-Tsubo Cardiomyopathy on echocardiography showing resolution of the apical ballooning and normalisation of the LV function.
**Fig. 17:** Left ventricular non-compaction cardiomyopathy. This 4 chamber view demonstrates prominent left ventricular trabeculae particularly towards the apex. Non compacta to compacta ratio was 2.4. Mildly impaired left ventricular systolic function.
Fig. 18: Peripartum Cardiomyopathy 4 chamber view showing dilated ventricles and reduced function in a 25yr old female patient who initially presented with shortness of breath 3 weeks after delivery. Subsequent echo images showed improvement in ventricular function.
Fig. 19: Peripartum Cardiomyopathy Follow-up echo showing resolution of the LV impairment.
Conclusion

Despite the large number of cardiomyopathies, cardiac MRI has proven to be a very useful tool at identifying the type of cardiomyopathy as well as pointing to the likely aetiology which can have important implications on patient management. As Cardiac MRI becomes more widely available this is likely to be used more frequently and allow earlier detection of cardiomyopathies than was previously available.

Personal Information

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