Imaging Diagnostic/Prognostic Approaches to Six Cardiomyopathies:
Myocarditis, Dilated, Amyloid, Sarcoid, Hypertrophic, Non-compaction

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Cardiomyopathies

- Diseases of the myocardium that lead to impaired ventricular function and CHF
- Caused by genetic abnormalities, myocyte injury or infiltration
- Abnormal cardiac function results from cellular and/or extracellular pathological alterations
Cardiomyopathies
Morphological Classification

- Dilated
  - Enlarged LV cavity, reduced LV ejection fraction
- Restricted
  - Small or normal LV cavity size
  - Normal, reduced or increased cardiac mass
  - Impaired diastolic with normal or reduced ejection fraction
- Hypertrophic
  - Symmetrically or asymmetrically increased LV and or LV myocardial wall thickness
  - Usually small or normal LV cavity size
- ARVR
  - Regionally or globally impaired RV systolic function

## Cardiomyopathies

### Ethiological Classification

<table>
<thead>
<tr>
<th>Category</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Idiopathic</td>
<td>50%</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>9%</td>
</tr>
<tr>
<td>Ischemic*</td>
<td>7%</td>
</tr>
<tr>
<td>Infiltrative disorders</td>
<td>5%</td>
</tr>
<tr>
<td>Peripartum</td>
<td>4%</td>
</tr>
<tr>
<td>Hypertension</td>
<td>4%</td>
</tr>
<tr>
<td>HIV</td>
<td>4%</td>
</tr>
<tr>
<td>Connective tissue disorders</td>
<td>3%</td>
</tr>
<tr>
<td>Substance abuse</td>
<td>3%</td>
</tr>
<tr>
<td>Chemotherapy-toxic</td>
<td>1%</td>
</tr>
<tr>
<td>Others</td>
<td>10%</td>
</tr>
<tr>
<td>Valvular disease</td>
<td></td>
</tr>
<tr>
<td>Congenital heart disease</td>
<td></td>
</tr>
<tr>
<td>Post-radiation</td>
<td></td>
</tr>
<tr>
<td>Neuromuscular</td>
<td></td>
</tr>
<tr>
<td>Endocrine</td>
<td></td>
</tr>
<tr>
<td>Malignancies</td>
<td></td>
</tr>
</tbody>
</table>

*Defined as global dysfunction secondary to chronic ischemia*

Dilated Cardiomyopathy

DCM encompass a group of diseases with different etiologies that affect the myocardium and are characterized by LV dilatation and reduced LVEF.

- LVEDD > 5.6 cm
- LVEDV > 74ml/m²
- LVEF < 52%

As a group they account for 1/3 cases of HFrEF

Etiology of HFrEF

Probability of CAD
- Low
- Intermediate
- High

Diagnostic Evaluation
- Medical Therapy
- ICA-CTA DSE-CMR ?
- Invasive Coronary Angiography
CTA - Ischemic vs Non-ischemic CM

61 DCM of unknown origin, 139 patients with suspected CAD

Andreini, J Am Coll Cardiol 2007;29:244-50
CMR - Ischemic vs. Non-ischemic CM

CMR - Ischemic vs. Non-ischemic CM

90 patients with DCM (63 with non-obstructed CAD), 15 controls

McCrohon, Circulation 2003;108: 54-59
Acute Myocarditis

Acute inflammatory process that affects the myocardium, often accompanied by pericarditis

Most common viral etiology. Less common toxic, autoimmune?

Global vs. segmental LV dysfunction of varying degree

Late Gadolinium Enhancement if present spares endocardium
$$dv/dt \approx -\text{MVA}/(\text{LA-LV Compliance})$$

**DT and LV Stiffness**

$$y = 231.16e^{-1.7712x}$$

$$r = 0.87$$

$$p < 0.001$$

$$\frac{DT}{\text{msec}} = \frac{11.6 \times C_n \times \sqrt{\Delta p_{\max}}}{\text{MVA}}$$

*Garcia, Am J Physiol 2001;280:H554*
DT and Prognosis

Rihal, Circulation 1994;90:2772
Prognostic Value of Late Gadolinium Enhancement in Nonischemic Cardiomyopathy

Juan Gaztanaga, MD, Vijayapraveena Paruchuri, MD, Elliott Elias, MD, Jonathan Wilner, MD, Shahidul Islam, MPH, Simonette Sawit, MD, Juan Viles-Gonzalez, MD; PhD, Javier Sanz, MD, and Mario J. Garcia, MD*

Am J Cardiol 2016;118:1063e 1068
LV Non-Compaction

Diagnosis of NC is usually made by echocardiography. The likelihood of missing the diagnosis on TTE is 30%.

Echocardiographic diagnosis is made by measuring the ratio of thickness of the non-compacted region to the compacted region measured at the end systole. The ratio of >2 is considered to be diagnostic for non-compaction.

Differential diagnosis: HCM, hypertensive cardiomyopathy

LV Thrombus often present
Restrictive Cardiomyopathies

Restrictive cardiomyopathy (RCM) is the least common of the 3 described cardiomyopathies.

RCM is characterized by **restrictive filling** and 1) **reduced diastolic volume of either or both ventricles** with normal or near-normal systolic function, 2) normal or increased wall thickness, 3) dilated atria.

Amyloidosis most frequent etiology.

A 54 year-old man with CHF and low ECG voltage
## Amyloidosis

**LV wall thickness and Prognosis**

<table>
<thead>
<tr>
<th>Group</th>
<th>Pt. No. (%)</th>
<th>Wall thickness (mm)</th>
<th>Median survival, (years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>34 (26)</td>
<td>&lt;12</td>
<td>2.4</td>
</tr>
<tr>
<td>II</td>
<td>23 (17)</td>
<td>12&lt;&lt;LV&lt;15</td>
<td>1.3</td>
</tr>
<tr>
<td>III</td>
<td>68 (52)</td>
<td>&gt;15</td>
<td>0.4</td>
</tr>
<tr>
<td>IV</td>
<td>7 (5)</td>
<td>Atypical echo</td>
<td>0.5</td>
</tr>
</tbody>
</table>

### TABLE 34.2
CLASSIFICATION OF CARDIAC AMYLOIDOSIS

<table>
<thead>
<tr>
<th></th>
<th>Precursor protein</th>
<th>Cardiac involvement</th>
<th>Other organ involvement</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary (AL)</td>
<td>Immunoglobulin light chain</td>
<td>Common</td>
<td>Kidney, liver, PNS, GI</td>
<td>Chemotherapy</td>
</tr>
<tr>
<td>Senile systemic</td>
<td>Wild-type transthyretin</td>
<td>Common</td>
<td>Lungs, carpal tunnel syndrome</td>
<td>Supportive</td>
</tr>
<tr>
<td>Hereditary systemic</td>
<td>Mutant transthyretin, Apo A-I, Apo A-II, etc.</td>
<td>Common</td>
<td>PNS</td>
<td>Liver transplantation</td>
</tr>
<tr>
<td>Secondary (AA)</td>
<td>Serum amyloid A</td>
<td>Rare</td>
<td>Kidney</td>
<td>Treatment of underlying disease</td>
</tr>
<tr>
<td>Isolated atrial amyloidosis</td>
<td>Atrial natriuretic peptide (ANP)</td>
<td>Common</td>
<td>None</td>
<td>None required</td>
</tr>
</tbody>
</table>

PNS, peripheral nervous system; GI, gastrointestinal.

*Curtin, In Garcia: Non-Invasive Cardiovascular Imaging, LWW 2009*
LV Strain

\[ \text{Strain} = \frac{(L - L_0)}{L_0} \]
\( \varepsilon_l \): Longitudinal Strain

\( \varepsilon_r \): Radial Strain

\( \varepsilon_c \): Circumferential Strain
Relative apical sparing of longitudinal strain using two-dimensional speckle-tracking echocardiography is both sensitive and specific for the diagnosis of cardiac amyloidosis.
CMR in Cardiac Amyloidosis

33 consecutive patients with CHF and restrictive filling

<table>
<thead>
<tr>
<th>Amyloid LGE Pattern</th>
<th>No/Other LGE</th>
<th>All</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiac amyloid by EMB</td>
<td>1.2</td>
<td>3</td>
</tr>
<tr>
<td>Other patients</td>
<td>1</td>
<td>17</td>
</tr>
<tr>
<td>All</td>
<td>1.3</td>
<td>20</td>
</tr>
</tbody>
</table>

Sensitivity (LGE) = 90%; specificity (LGE) = 94%; positive predictive value (LGE) = 92%; negative predictive value (LGE) = 85%.

Abbreviations as in Table 1.

Diffuse Late Gadolinium Enhancement corresponding to Amyloid deposits

- LV Sub-endocardium
- LA and intra-atrial septum

Contraindicated when GFR <30 mL/min

Vogelsberg, J Am Coll Cardiol 2008;51: 1022-30
Role of Cardiac Scintigraphy With $^{99m}$Tc-DPD in the Differentiation of Cardiac Amyloidosis Subtype


TTR Amyloid

AL Amyloid
Sarcoidosis

Sarcoidosis is a multisystem granulomatous disease

About 5% of patients with sarcoidosis demonstrate cardiac involvement clinically

Localized aneurysms, variable LV dysfunction

Late Gadolinium Enhancement detected in 20% patients with normal echocardiogram and ECG

Increased 18F-FDG uptake on PET if active inflammation
Sarcoidosis

GADOLINIUM

$N_{13}$-NH$_4$

$F_{18}$-FDG
2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy

“Disease state characterized by unexplained LV Hypertrophy associated with non dilated ventricular chambers in the absence of another cardiac or systemic disease that itself would be capable of producing the magnitude of hypertrophy evident in a given patient with the caveat that patients who are genotype positive may be phenotypically negative without overt hypertrophy

“Clinically HCM is usually recognized by maximal wall thickness >15mm based on echocardiography, with wall thickness of 13 to 14 mm considered borderline, particularly in the presence of other compelling information (e.g., family history of HCM)"

“In principle, any degree of wall thickness is compatible with the presence of HCM genetic substrate”
Phenotype vs Genotype in HCM

- Sigmoidal HCM 40 - 50%
- Reverse curve HCM 30 - 40%
- Apical HCM ~ 10%
- Neutral HCM ~ 10%

~ 10% Myofilament Gene +
~ 80% Myofilament Gene +
~ 30% Myofilament Gene +
~ 40% Myofilament Gene +

Boss, J Am Coll Cardiol 2009;54:201-11
HCM Vs Trained Heart

“Gray Zone” of LV Wall Thickness (13-15 mm)

- Unusual Patterns of LV Hypertrophy
- LV Cavity < 45mm
- LV Cavity > 55mm
- Left Atrial Enlargement
- Bizarre ECG Patterns
- Abnormal LV Filling
- Female Gender
- Thickness with Deconditioning
- Family History of HCM
- Max. VO₂ > 45 ml/kg/min > 110% predicted

Hypertrophic Cardiomyopathy

29 yo man with exertional CP and dyspnea
LVOT obstruction in HCM

- Nonobstructive (95; 30%)
- Provokable Obstruction
- Rest Obstruction (119; 37%)

- With Exercise (106; 33%)

70%

Maron M et al. Circulation 2006;114:2232-2239
Hypertrophic Cardiomyopathy

**29 yo man with exertional CP and dyspnea**
Diastolic Dysfunction in HCM

A 48 year-old woman with CP and abnormal ECG
A 48 year-old woman with CP and abnormal ECG
Prognostic Value of Quantitative Contrast-Enhanced Cardiovascular Magnetic Resonance for the Evaluation of Sudden Death Risk in Patients With Hypertrophic Cardiomyopathy

A

Freedom from SCD events

Follow up (years)

LGE absent
LGR ≤ 10%
LGR ≤ 15%
LGE ≥ 20%
p=0.008

B

Incidence of SCD events

0 LGE
N=745 (59%)
≤10% LGE
N=381 (59%)
11-19% LGE
N=94 (7%)
≥20% LGE
N=73 (6%)

LGE by % Left Ventricular Mass

Circulation. 2014;130:484-495.
Summary

- Non-invasive imaging indices provide an accurate and complete evaluation in most patients with known or suspected Cardiomyopathies:
  - Differentiation of HFrEF vs. HFrEF
  - Estimation of Cardiac Hemodynamics
  - Determination of Etiology
  - Prognosis and Guidance of Therapy