Dilated Cardiomyopathy

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Governor Chile Chapter of ACC
I have not a financial relationship to disclosure
Dilated cardiomyopathy (DCM) is defined as left ventricular (LV) dilatation and systolic dysfunction in the absence of coronary artery disease or abnormal loading conditions proportionate to the degree of LV impairment (1).

DILATED CARDIOMYOPATHY

• One of the leading causes of heart failure (HF), DCM predominantly affects younger adults and is the most frequent indication for cardiac transplantation.

• DCM is the final common response of myocardium to a number of genetic and environmental insult.

• Historically the standard approach as like all systolic HF
POINT TO REVIEW

• EVALUATION OF ETIOLOGY

• ASSESSMENT OF REMODELING

• EVALUATION FOR AN ICD

• DETECTION OF THE PRE-DCM PHENOTYPE
Point 1
Evaluation of Etiology

• Exclusion of other causes.

• Routine etiology work up.

• The role of genetic in DCM
Exclusion of main causes of LV Dilatation

• Coronary artery disease
• OH consumption
• Chemotherapy treatment
• Persistent tachyarrhythmia
• Peripartium HF
• HIV
• Inflammatory cardiomiopathy
Inflammatory cardiomyopathy and the role of endomyocardial biopsy (EMBx)

- Biopsy findings carry clear treatment implications in DCM patients with suspected giant cell myocarditis, eosinophilic myocarditis, or sarcoidosis, and EMBx is indicated in these patient groups.

- Modern immuno-histochemical methods improve sensitivity compared with the traditional histopathological Dallas criteria.
Randomized study on the efficacy of immunosuppressive therapy in patients with virus-negative inflammatory cardiomyopathy: the TIMIC study

Andrea Frustaci¹,²*, Matteo A. Russo³,⁴, and Cristina Chimenti¹,²,⁴

doi:10.1093/eurheartj/ehp249
• However, conclusive benefit from EMBx-guided treatment is awaited.

• A rational approach to this conflicting guidance is to consider the incremental value of EMBx on an individual case basis
Point 2
Assessment of Remodeling

• The extent of LV dilation and contractile impairment in DCM is a major determinant of adverse outcomes.

• Reversal of these abnormalities, LV reverse remodeling, is a key therapeutic goal.
ADVERSE REMODELING CHARACTERISTICS IN DCM INCLUDE THE EVALUATION OF:

• LV size and systolic function
• Remodeling of other cardiac chambers
• Functional mitral regurgitation
• Myocardial fibrosis
• Ventricular dyssynchrony (?)
(A) Left ventricular cavity dilation (asterisk) with wall thinning. (B) Extensive left ventricular midwall replacement fibrosis (arrows). (C) Myocyte hypertrophy (black arrow), myocyte atrophy (blue arrow), nuclear pleomorphism (arrowheads), and increased interstitial fibrosis (stained with Picrosirius red); magnification ×500.
Mitral regurgitation
Point 3
Evaluation for an ICD

Defibrillator Implantation in Patients with Nonischemic Systolic Heart Failure

In this trial, prophylactic ICD implantation in patients with symptomatic systolic heart failure not caused by coronary artery disease was not associated with a significantly lower long-term rate of death from any cause than was usual clinical care. (Funded by Medtronic and others; DANISH ClinicalTrials.gov number, NCT00542945.)
Point 4: Detection of the pre-DCM phenotype

- Detect pre-symptomatic DCM have a clear rationale.
- Early treatment can retard adverse remodeling, prevent HF symptoms, and increase life expectancy.
GENETIC CAUSES OF DCM

• Molecular genetic analysis has uncovered “causal” mutations for DCM in over 60 genes.

• At present, routine genetic testing is only recommended in familial disease (>2 affected family members), where its diagnostic yield is 30% to 35%.
<table>
<thead>
<tr>
<th>Stage of left ventricular (LV) remodeling</th>
<th>Latent</th>
<th>Established</th>
<th>Advanced</th>
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</thead>
<tbody>
<tr>
<td>LV</td>
<td>Early LV phenotype (e.g. ↓ strain, LV enlargement, diffuse fibrosis)</td>
<td>↑ LV volume and ↓ LVEF</td>
<td>Severeely ↑ LV volume and ↓ LVEF</td>
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<tr>
<td>+/-</td>
<td>+/-</td>
<td>Limited or no replacement fibrosis</td>
<td>Extensive replacement fibrosis</td>
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<tr>
<td>+/- Pathogenic gene mutation</td>
<td>+/- Functional mitral regurgitation</td>
<td>Wall thinning</td>
<td></td>
</tr>
<tr>
<td>+/- Altered biomarkers</td>
<td>+/- LV dyssynchrony</td>
<td>+/- Right ventricular remodeling</td>
<td></td>
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<td></td>
<td>+/- Active myocarditis</td>
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<tr>
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<td>• Refractory to conventional therapies</td>
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**Treatment strategies**

(Consider ICD based on established and novel risk factors for sudden cardiac death, for all three stages)

- Retard remodeling:
  - Neurohormonal blockade
  - Molecular / gene therapy
  - Imaging & biomarker surveillance

- Reverse remodeling:
  - Neurohormonal blockade
  - Cardiac resynchronization
  - Mitral valve interventions
  - Molecular / gene therapy
  - Immunosuppressive / antiviral treatment

- Regenerate:
  - Stem cell therapy
  - ‘Bridge to recovery’ LV assist device

- Replace:
  - Cardiac transplant
  - ‘Destination therapy’ LV assist device

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