Left Ventricular Non-Compaction Case Studies

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Left Ventricular Noncompaction Cardiomyopathy

Historical Perspective

- 1926 Grant - Malformed heart of a child
- 1975 Dusek - Spongy Myocardium
- 1984 Englbérding – Echo Diagnosis of Myocardial Sinusoids
- 1986 Jenni – Biventricular Sinusoids
- 1990 Chiu Isolated noncompaction

LVNC

- Although heterogeneous it has genetic underpinnings – TAZ gene
- Autosomal dominant
- Sarcomeric genes
- Our knowledge and understanding regarding prevalence and mortality is similar to HCM 50 years ago

<table>
<thead>
<tr>
<th>Year</th>
<th>HCM Prevalence</th>
<th>LVNC Prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>1960</td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td>1980</td>
<td>Common</td>
<td>Common?</td>
</tr>
<tr>
<td>2010</td>
<td>Common</td>
<td>Low?</td>
</tr>
</tbody>
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Left Ventricular Noncompaction Cardiomyopathy

<table>
<thead>
<tr>
<th>Year</th>
<th>HCM Mortality</th>
<th>LVNC Mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>1980</td>
<td>High</td>
<td>High</td>
</tr>
<tr>
<td>2000</td>
<td>Low</td>
<td>Low</td>
</tr>
<tr>
<td>2020</td>
<td>Low?</td>
<td>Low?</td>
</tr>
</tbody>
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**Left Ventricular Noncompaction Cardiomyopathy**

**Nomenclature**
- Persistent Sinusoids
- Spongy myocardium
- Embo myocardium
- Honeycomb LV
- Hypertrabeculations
- Isolated LVNC

**Left Ventricular Noncompaction: A 25-Year Odyssey**

Timothy E. Paterick, MD, JD, FACC, FASE, Matt M. Umland, RDMS, FASE, M., Fuss Jan, MBBS, MD, Khawaja Attili Ammar, MD, Christopher Kramer, RDMS, Bijoy K. Khanderia, MD, FACC, FESC, FASE, FAHA, FACP, James B. Seward, MD, FACC, FASE, and A. Jami Tajik, MD, FACC, FAHA, Milwaukee, Wisconsin, Rochester, Minnesota

**Non-Compaction**

- Normal
- Noncompacted

**Fetal Heart Development**

- Week 3-4 of fetal life
- No coronary circulation
- Compaction
- Coronary arteries
The SPECTRUM of LVNC

- Heterogeneity
- From 12-18 weeks of gestation until the 90s

Presenting Symptoms

- SOB: 66%
- Chest Pain: 15%
- Palpitations: 13%
- Syncope: 9%
- Embolism: 3%

Left Ventricular Noncompaction Cardiomyopathy

**Diagnostic Criteria**

- Bilayered myocardium (C+NC)
- Ratio of NC/C ≥ 2.0
- Large trabecular
- Prominent intertrabecular recesser
- Apical location
Left Ventricular Noncompaction Cardiomyopathy

- This is the major and primary diagnostic imaging criterion
- It is critical to obtain images that are not foreshortened and are perpendicular to the ventricular long-axis view
**Tips & Tricks**

- Meticulous attention must be paid to the short-axis images, and a circular cavity image should be obtained and maintained in all short-axis views.
- Oblique and tangential cuts (noncircular LV cavity appearance) should not be used to measuring noncompacted and compacted layers as this will lead to false diagnosis of LVNC.

**Contrast**

- The use of contrast for opacification of the LV apex and demonstration of the intertrabecular recesses has been beneficial in selected cases as discussed in the literature.

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**CASE**

34 y/o male

**CONTRAST ENHANCEMENT**

_left Ventricular Noncompaction Cardiomyopathy_
Hypertrophic Cardiomyopathy and Apical LVNC
Hypertrophic Cardiomyopathy and Apical LVNC

FATHER……………HCM
MOTHER……………LVNC

26 y/o male
DCM, LVNC
Long QT

M_D+LVNC_O_HAD_E_G_RMBM20, p. Gly1031_S_D-III

26 y/o male
DCM, LVNC
Long QT

Strain of Left Ventricular Segments (Apex, Mid and Base)

<table>
<thead>
<tr>
<th>Strain (%)</th>
<th>LVNC</th>
<th>DCM</th>
<th>Controls</th>
</tr>
</thead>
<tbody>
<tr>
<td>Apex</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mid</td>
<td></td>
<td></td>
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<tr>
<td>Base</td>
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M. Niemann et al: Eur J Heart Fail 2012