Left Ventricular Noncompaction Syndrome: A 25 Year Odyssey
Timothy E Paterick, MD, JD
Matt Umland, R.D.C.S.

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Left Ventricular Noncompaction: A 25-Year Odyssey
Timothy E. Paterick, MD, FACC, FASE, Matt M. Umland, RDCS, FASE, Fuad Jan, MBBS, MD, Khawaja Afzal Ammar, MD, Christopher Kramer, RDCS, Bijoy K. Khandheria, MD, FACC, FESC, FASE, FAHA, FACP, James B. Seward, MD, FACC, FASE, and A. Jamil Tajik, MD, FACC, FAHA, Milwaukee, Wisconsin; Rochester, Minnesota

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Left Ventricular Noncompaction Cardiomyopathy

- Historical Perspective
  - 1926 Grant - Malformed heart of child
  - 1975 Dusek - Spongy Myocardium
  - 1984 Englberding – Echo Diagnosis
  - 1986 Jenni – Biventricular Sinusoids
  - 1990 Chen – Isolated noncompaction

- Nomenclature
  - Persistent Sinusoids
  - Spongy myocardium
  - Embryomyocardium
  - Honeycomb LV
  - Hypertrabeculations
  - Isolated LVNC

- Genetics
  - Autosomal dominant
  - Sarcomeric genes
  - Taz

- Clinical Spectrum
  - Asymptomatic ➔ See D
  - Heart Failure ➔ Tx
  - Arrhythmias
  - Thromboembolic Complications

- Fetal Heart Development
  - Week 3-4 of fetal life
  - No coronary circulation
  - Compaction
  - Coronary arteries

Genetic/embryology
The SPECTRUM of LVNC

- Heterogeneity
- From 12-18 weeks of gestation until the 90s
- Asymptomatic to cardiac transplantation

Left Ventricular Noncompaction

- A distinct Cardiomyopathy
- Characteristic morphologic features best identified by Echo
- Abnormal embryogenesis
- Knowledge and understanding progressing similar to HCM
- 50 years ago

LVNC

- Although heterogeneous it has genetic underpinnings — the TAZ gene so far
- Most echocardiography laboratories do not make the diagnosis
- Our knowledge and understanding regarding prevalence and mortality is similar to HCM 50 years ago
Left Ventricular Noncompaction Cardiomyopathy

<table>
<thead>
<tr>
<th></th>
<th>HCM</th>
<th>LVNC</th>
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</thead>
<tbody>
<tr>
<td>Prevalence</td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td>Diagnosis</td>
<td>4:1:1</td>
<td>C+NC</td>
</tr>
<tr>
<td>Mortality</td>
<td>High</td>
<td>Low</td>
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</tbody>
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Nomenclature

- This inconsistent nomenclature has been an impediment to a definition and understanding of LVNC.
- The various names used to describe the histological profile of LVNC have prevented a standardized nomenclature and impeded the use of diagnostic criteria that would facilitate understanding and promote research.
- The limitations must be overcome to advance our understanding.

Recognition

- What tools do we have to advance our knowledge and understanding?
- The histology, gross specimen, M-mode, 2-D, and 3-D images all give a characterization to the entity called LVNC.
- The echocardiography laboratory is the learning environment to allow advanced understanding and characterization of LVNC.

Left Ventricular Noncompaction Cardiomyopathy

Diagnostic Criteria (Echo):

- Bilayered myocardium (C+NC)
- Ratio of NC/C ≥ 2.0
- Large trabecular myocardium
- 4 or more trabeculae
- Prominent intertrabecular recesses
- Apical location

Left Ventricular Noncompaction Cardiomyopathy

A B

Left Ventricular Noncompaction Cardiomyopathy

A B

Left Ventricular Noncompaction Cardiomyopathy

A B
Left Ventricular Noncompaction Cardiomyopathy

Diagnostic Criteria: 2 D

M-Mode: Compacted & Noncompacted

Patient #1
LV apical obliteration with non-compacted myocardium

Color flow visualized in the **sinusoidal** recesses
Left Ventricular Noncompaction Cardiomyopathy

Embolic events

Advances in Imaging: 3D imaging
Left Ventricular Noncompaction Cardiomyopathy

MRI allows for a multimodality imaging approach

Left Ventricular Noncompaction Cardiomyopathy
**Left Ventricular Noncompaction Cardiomyopathy**

Echocardiographic quantification of regional deformation helps to distinguish isolated left cardiomyopathy

Markus Niemann, Dan Liu, Kai Hu, Maja Cikes, Meinrad Beer, Sebastian Harmann, Philipp Daniel Gaudron, Hanns Hillenbrand, Wolfram Voelker, Georg Erdl, and Frank Weidemann

Methods and results: We investigated 15 patients with LVNC (9 males; 42 ± 9 years), 15 age- and gender-matched DCM patients, and 15 healthy controls.

**Conclusion:** A special regional deformation pattern (preserved deformation in basal segments of LVNC) seems to be of major diagnostic help for the definite differential diagnosis of LVNC and DCM.

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

M. Niemann et al: Eur J Heart Fail 2012

**Reproducibility of Echocardiographic Diagnosis of Left Ventricular Noncompaction**

Susan F. Saleeb, MD, Renee Margossian, MD, Carolyn T. Spencer, MD, Mark E. Alexander, MD, Leslie B. Smoot, MD, Adam L. Dorfman, MD, Lisa Bergersen, MD, MPH, Kimberlee Gaureau, ScD, Gerald R. Marx, MD and Steven D. Colan, MD, Boston, Massachusetts

Results: A total of 104 patients with LVNC were included in the study, 52 with no congenital heart disease.

Agreement in measuring an NC/C ratio ≤ 2 versus >2 was 79% (NCongHD) and 73% (CongHD). Agreement with the original reader in diagnosing LVNC was 67%.

**Conclusion:** The reproducibility of making measurements to diagnose LVNC by accepted criteria is poor. Heart transplantation and death are associated with significant ventricular dysfunction and not with increased trabeculations or NC/C ratios.

**Contrast enhancement of sinusoidal recesses and the best definition of compacted myocardium**

Compacted myocardium

LV apex

Precise measurement is critical
Fetal Heart Development

- Genetic/embryology
- Week 3-4 of fetal life
- No coronary circulation
- Week 5-8 of fetal life
- Coronary arteries

Non-Compaction

2 mo New Born

80 year old man

Presenting Symptoms

- SOB
- Chest Pain
- Palpitations
- Syncope
- Embolism

Ratio of non-compacted to compacted myocardium ≥ 2 at end diastole (our laboratory)

Intratrabecular recesses: low-scale color Doppler or contrast

Echo is diagnostic test of choice!

Heart 2002;88:iv29

J Am Soc Echocardiogr 2004;17:87-90

What is Important?

- Measurement precision is critical

AWARENESS

Screening

- All first degree relatives is important
- Echo is the first step
FDR with LVNC

- Then consider stress testing and Holter monitoring in those that have LVNC

Familial Occurrence

- 75% Screened for Familial Occurrence
- 30% had Noncompaction
- 55% had Noncompaction or Dilated Cardiomyopathy

Monitoring and Treatment

- Heart failure
  - Diastolic / Systolic
- Arrhythmias
- Thromboembolic events

Left Ventricular Noncompaction Cardiomyopathy

- Clinical manifestations:
  - Heart failure
  - Diastolic / Systolic
  - Arrhythmias
  - Thromboembolic events
  - LVNC
  - Increased risk of thromboembolic events

- Echocardiographic manifestations:
  - LVNC
  - Left ventricular dilatation
  - Pericardial effusion

Left Ventricular Noncompaction Cardiomyopathy

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