Left ventricle non compaction

Dr. Heba El Deeb
Lecturer Of Cardiology
Cairo University

Termiology

• Isolated left ventricular non copaction
  - Spongy myocardium
  - Persistent myocardial sinusoids
  - Hypertabeculation

• LVNC in association with other congenital abnormalities
Outline

• Definition
• Embryology
• Pathophysiology
• Association with other disease
• Isolated LV non compaction
• Epidemiology
• Pathology
• Clinical features
• Diagnosis
• Prognosis
• Management

Definition

• Congenital heart disease

• Myocardial wall distortion
  - Prominent trabeculae
  - Deep intertabecular recesses

• Continuity between LV cavity and recesses

• AHA (2006)--- classified as primary genetic cardiomyopathy

• WHO&ESC(2008)---- unclassified cardiomyopathy

_Circ Heart Fail 2008; 1:72–75
Eur Heart J 2008; 29:270–276_
Embryology

• **Myocardial architecture passes through four distinct steps:**
  - Early heart tube
  - Emergence of trabeculations
  - Trabecular remodelling
  - Development of the multilayered spiral system

    - Epicardium – Endocardium
    - Base – Apex
    - Septum – Free wall in the LV

Types

• 1\textsuperscript{st} described in association with other congenital abnormalities
  - Obstruction of LVOT/RVOT (pulmonary atresia with intact ventricular septum)
  - Ebstein anomaly
  - Complex cyanotic congenital heart disease
  - Anomalous coronary arteries
  - VSD, ASD
• Associated with other disease
  - Neuromuscular disorder
  - Metabolic disease
  - Genetic syndrome
    * Barth syndrome
    * Charcot-Marie-Tooth
    * Nail-patella
• Isolated LV non compaction (Chin et al 1990)

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**Epidemiology**

• True prevalence: unknown.

• Echocardiographic laboratory at a tertiary care centre: 0.014: 1.3%.
  - In a population-based, retrospective cohort study of 1st cardiomyopathies in children 9.2:9.5%.

• Heart failure clinic, only a minority of this HF population (3%) had isolated LVNC.

• Men >> female (56-82%)

Eur Heart J 2011; 32: 1446–1456
EurHeartJ.2006;28:1855–1860
cir 2003;108: 2672–2678
Genetics

- A genetically heterogeneous disorder
- Sporadic and familial form
- Familial recurrence (18% adult and 50% pediatric)
- Autosomal dominant inheritance > X-linked inheritance

<table>
<thead>
<tr>
<th>TABLE 1: Gene Mutations Reported in Human Left Ventricular Noncompaction</th>
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<tbody>
<tr>
<td>Gene</td>
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<tr>
<td>---</td>
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<tr>
<td>α-Dystrobrevin (DTNA)</td>
</tr>
<tr>
<td>4.5 (TAZ)</td>
</tr>
<tr>
<td>LIM domain–binding protein (LDB3, Cypher/ZASP)</td>
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<tr>
<td>Lamin A/C</td>
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<tr>
<td>Sarcomere proteins</td>
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<tr>
<td>β-Myosin heavy chain (MYH7)</td>
</tr>
<tr>
<td>α-Cardiac actin (ACTC)</td>
</tr>
<tr>
<td>Cardiac troponin T (TNNT2)</td>
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Pathology

- Histologic examination of the prominent trabeculations
  - Fibrosis
  - Endocardial fibroelastosis
  - Necrotic myocytes
**Clinical picture**

- Asymptomatic (during family screening)
- Advanced cases
  - Heart failure >50%
    * Dyspnea
    * Non specific chest pain
- Thromboembolism (5-38%)
  * CVA/ TIA
- Arrhythmias
  * AF/A flutter
  * VT (2-62%)
  * SCD

- ECG : non specific:
  * BBB (LBBB, RBBB, IVCD)
  * Voltage signs of LV hypertrophy
  * Repolarization
Imaging for diagnosis

Echocardiography

Table I  Diagnostic criteria for left ventricular non-compaction

<table>
<thead>
<tr>
<th>Echocardiography</th>
<th>Chin et al.11</th>
</tr>
</thead>
<tbody>
<tr>
<td>Two-layered structure of the myocardium (epicardial compacted, endocardial non-compacted layer)</td>
<td></td>
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<tr>
<td>Determination of the X-to-Y ratio (≤0.5)</td>
<td></td>
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<tr>
<td>X—Distance between the epicardial surface and through of inter trabecular recess</td>
<td></td>
</tr>
<tr>
<td>Y—Distance between epicardial surface and peak of trabeculation</td>
<td></td>
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<tr>
<td>Acquisition of the images: parasternal short-axis view, measurements of the X-to-Y ratio at end-diastole</td>
<td></td>
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</tbody>
</table>

Circulation 1990;82:507-513
Jenni et al. 13

Thickened myocardium with a two-layered structure consisting of a thin compacted epicardial layer/band (C) and a much thicker, non-compacted endocardial layer (N) or trabecular meshwork with deep endomyocardial spaces; N/C ratio > 2.0

Predominant location of the pathology: mid-lateral, mid-inferior, and apex

Colour Doppler evidence of deep intertrabecular recesses filled with blood from the left ventricular cavity

Absence of coexisting cardiac abnormalities (in the presence of isolated LVNC)

Acquisition of the images: short-axis views, measurements of the N/C ratio at end-systole

Stöllberger et al. 15,21

More than three trabeculations protruding from the left ventricular wall, located apically to the papillary muscles and visible in one image plane

Trabeculations with the same echogenicity as the myocardium and synchronous movement with ventricular contractions

Perfusion of the intertrabecular spaces from the left ventricular cavity

Ratio of non-compacted to non-compacted segment > 2.0 at end-diastole (this criterion was introduced later)

Acquisition of the images: apical four chamber view; angulation of the transducer and acquisition of pictures in atypical views to obtain the technically best picture quality for differentiation between false chords/aberrant bands and trabeculations

Diagnostic criteria have changed during the last years
Contrast echocardiography

<table>
<thead>
<tr>
<th>Location</th>
<th>Pediatric Patients</th>
<th>Adult Patients</th>
<th>Sengupta</th>
</tr>
</thead>
<tbody>
<tr>
<td>LV apex</td>
<td>100%</td>
<td>94%</td>
<td>100%</td>
</tr>
<tr>
<td>Inferior wall</td>
<td>70%</td>
<td>94%</td>
<td>95%</td>
</tr>
<tr>
<td>Lateral wall</td>
<td>41%</td>
<td>100%</td>
<td>100%</td>
</tr>
<tr>
<td>Basal segments of the LV</td>
<td>&lt; 20%</td>
<td>&lt; 27%</td>
<td></td>
</tr>
</tbody>
</table>

LV - left ventricle.

Eur Heart J 2001;32:1446-1456
Accuracy of combined Echocardigraphic criteria

- Kohli et al. There was a poor correlation between the three echocardiographic definitions:
- 199 patients referred to HF clinic, Compared to 60 normal control
- 47 patients (24%) fulfilled any echocardiographic definitions
  * Chin et al 19%
  * Jenni et al 15%
  * Stollberger et al 13%
  * Combined 7% (poor correlation)
- 5 control (8%) satisfied one or more diagnostic criteria for LVNC; interestingly, four of these individuals were black

Eur heart j 2008;29:89-95

Speckle tracking echocardiography

A) Apical torsion/twist mechanics in a normal heart: apical rotation is above and basal rotation is below the baseline. (B) Abnormal apical torsion/twist mechanics both the apical and basal rotations are above the baseline

Circulation 76; 2012:1556-1560
Magnetic resonance imaging

Petersen et al.²⁶
Ratio between the non-compacted and compacted layer > 2.3
Measurement: at end-diastole

Jacquier et al.²⁷
Trabeculated left ventricular mass > 20% of the global left ventricular mass
Measurement: left ventricular trabeculation and global/compacted LV mass were defined at end-diastole

sensitivity and specificity, and positive and negative predictions of 86, 99, 75, and 99%, respectively.

Eur Heart J 2010;31:1098–1104

Magnetic resonance imaging

• Areas of trabecular and Subendocardial delayed contrast enhancement can be seen and are related to the presence of subendocardial and trabecular fibrosis and fibroelastosis
  - An abnormal contractile mechanism
  - Abnormal microvasculature
  - Inadequate microvasculature in the presence of increased myocardial mass
    (Normal epicardial coronary arteries)
• The prevalence and extent of myocardial fibrosis correlated with the severity of LV systolic dysfunction.

Eur J Heart Fail 2011; 13:170–176
MULTI SLICE CT

- Delineate the characteristic two layered myocardium of the left ventricle with prominent myocardial trabeculations
- Diagnosis based on the same morphologic findings reported for cardiac MRI
- Evaluate the coronary arteries noninvasively and the presence of coronary anomalies.
- However, use of ionizing radiation and lower capability of characterizing myocardial tissue and depicting fibrosis compared with MRI
- Can be used for patients with defibrillators or resynchronizing devices which are contraindications to MRI

*Eur J Heart Fail* 2011; 13:170–176

Zuccarino et al AJR:204, May 2015
Differential diagnosis

• Apical hypertrophic cardiomyopathy
• Dilated cardiomyopathy
• Hypertensive cardiomyopathy
• Endocardial fibroelastosis
• Abnormal chords, false tendons
• Apical thrombus
• Tumours (fibroma)
• Cardiac metastasis

Prognosis

• Early reports: 60% SCD or cardiac transplantation (6 years after the diagnosis)

• Recent reports: 97% mean free survival from death and cardiac transplantation

• Average 3.5 years from onset of diagnosis to symptoms

• Mortality did not differ significantly between patients with isolated LVNC and control patients with DCM (3-year survival of 85 vs. 83%).

Circulation 76; 2012:1556-1560
Management

- Screening for 1st degree relatives
- Neurological & musculoskeletal evaluation
- Standard heart failure treatment (Carvidolol)
- Anticoagulation: (LV EF% <40%), AF, thromboembolic history (INR 2.0–3.0)
- ICD implanted: syncope, symptomatic ventricular arrhythmias or LVEF< 35%
- Cardiac resynchronization therapy
- Heart transplantation
Thank you