

Left ventricle non compaction

Dr. Heba El Deeb
Lecturer Of Cardiology
Cairo University

Terminology

- Isolated left ventricular non compaction
 - 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
- Mangement

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

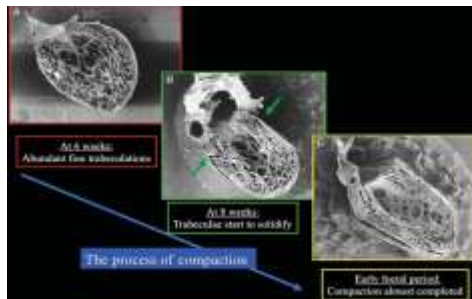
- **Myocardial architecture passes through four distinct steps:**

- 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

- 1st 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)

Epidemiology

- True prevalence: unknown.
- Ehco laboratory at a tertiary care centre: 0.014: 1.3%.
 - In a population-based, retrospective cohort study of 1^{ry} 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

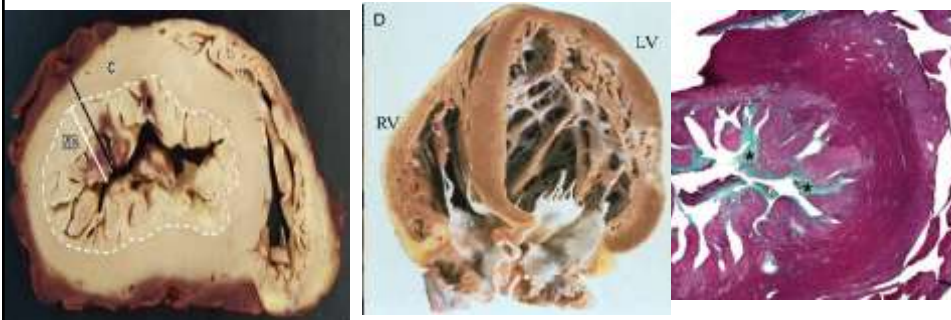
- A genetically heterogeneous disorder
- Sporadic and familial form
- Familial recurrence (18% adult and 50% pediatric)
- Autosomal dominant inheritance > X-linked inheritance

TABLE 1: Gene Mutations Reported in Human Left Ventricular Noncompaction

Gene	Disease	Location	Reference	Age Group
α -Dystrobrevin (<i>DTNA</i>)	LVNC with CHD, muscular dystrophy in human	18q12	[12]	Child
G4.5 (<i>TAZ</i>)	LVNC, DCM, Barth syndrome	Xq28	[12, 14, 15]	Child
LIM domain-binding protein (<i>LDB3</i> , <i>Cypher/ZASP</i>)	LVNC, DCM	10q22-q23.2	[13, 16]	Child, adult
Lamin A/C	LVNC, DCM, muscular dystrophy in human	1q22	[17]	Child, adult
Sarcomere proteins				
β -Myosin heavy chain (<i>MYH7</i>)	HCM, DCM, LVNC	14q11.2-q13	[18]	Adult
α -Cardiac actin (<i>ACTC</i>)	HCM, DCM, LVNC	15q11-q14	[18]	Adult
Cardiac troponin T (<i>TNNI2</i>)	HCM, DCM, LVNC	1q32	[18]	Adult

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 1 Diagnostic criteria for left ventricular non-compaction

Echocardiography

Chin et al.¹¹

Two-layered structure of the myocardium (epicardial compacted, endocardial non-compacted layer)

Determination of the X-to-Y ratio (≤ 0.5)

X—Distance between the epicardial surface and through of intertrabecular recess

Y—Distance between epicardial surface and peak of trabeculation

Acquisition of the images: parasternal short-axis view, measurements of the X-to-Y ratio at end-diastole

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

Heart 2001;86:666-671

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

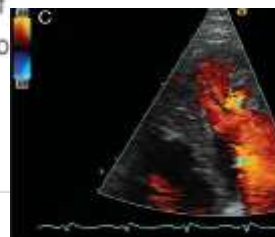
Trabeculations with the same echogenicity as the myocardium and synchronous movement with ventricular contractions

Perfusion of the intertrabecular spaces from the left ventricular cavity

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

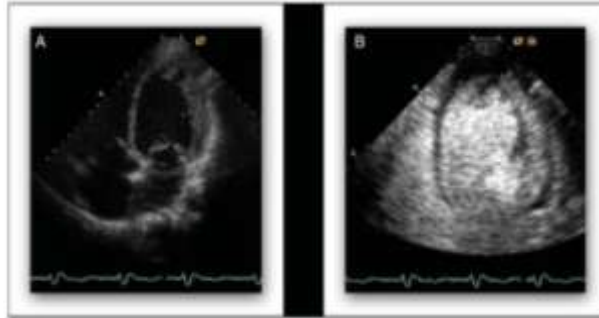
Acquisition of the images: apical four chamber view; angulation of the transducer and acquisition of pictures in atypical views to obtain the technically best picture quality for differentiation between false chords/aberrant bands and trabeculations

Diagnostic criteria have changed during the last years



Am J Cardiol 2002;90:899-902

Contrast echocardiography



Eur Heart J 2001;32:1446-1456

Table 2 - Location of trabeculations in patients diagnosed with noncompaction cardiomyopathy

	Pediatric patients Ichida ²⁸	Adult patients Oechslin ⁷	Sengupta ¹¹
LV apex	100%	94%	100%
Inferior wall	70%	94%	95%
Lateral wall	41%	100%	100%
Basal segments of the LV		< 20%	< 27%

LV - left ventricle.

Eur Heart J 2001;32:1446-1456

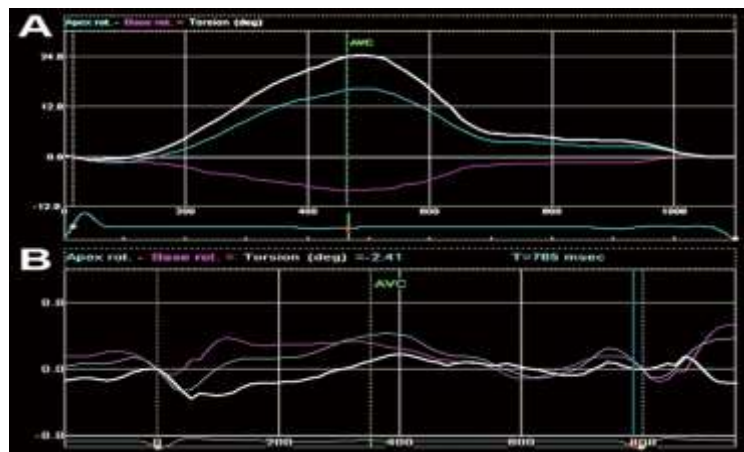
Accuracy of combined Echocardiographic criteria

- 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

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

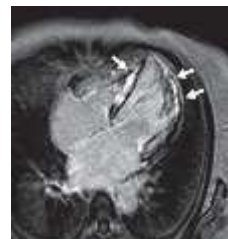
sensitivity and specificity, and positive and negative predictions of 86, 99, 75, and 99%, respectively.

J Am Coll Cardiol 2005;46:101–105

Eur Heart J 2010;31:1098–1104

Magnetic resonance imaging

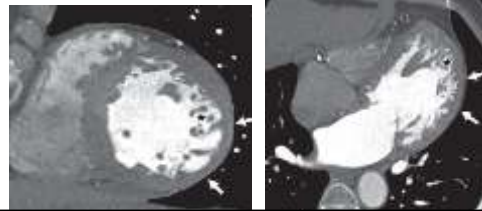
- Areas of trabecular and Subendocardial delayed contrast enhancement can be seen and are related to the presence of subendocardial and trabecular fibrosis and fibroelastosis
 - An abnormal contractile mechanism
 - Abnormal microvasculature
 - Inadequate microvasculature in the presence of increased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



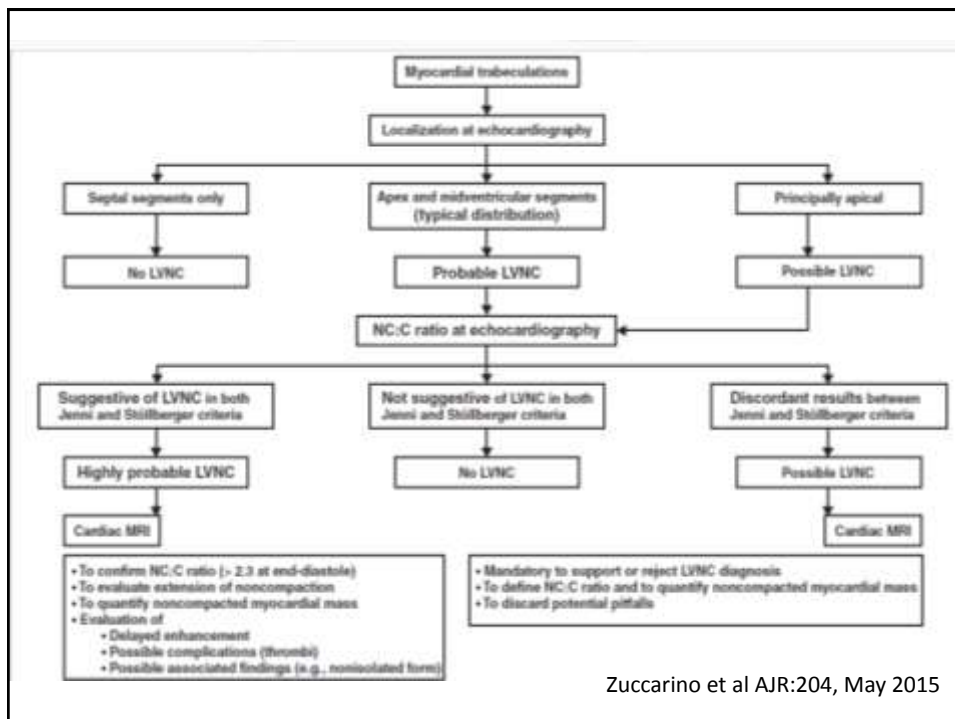
Eur J Heart Fail 2011; 13:170–176

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



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%).

Mayo Clin Proc 1997;72(1):26-31
Eur Heart J 2005;26(2):187-192
Circulation 76; 2012:1556-1560

Table 4 Predictors of death or heart transplantationClinical predictors^{30,51,59}

- Age at initial presentation
- Functional capacity, NYHA class III–IV
- Sustained ventricular arrhythmias

Echocardiographic parameters^{24,30,51,57,63}

- Ratio of non-compacted to compacted layers
- Number of affected segments
- LV end-diastolic diameter
- Abnormal lateral mitral tissue Doppler Ea velocity

Eur Heart J 2005;26(2):187-192

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

Heart 2001;86:E4

Thank you