

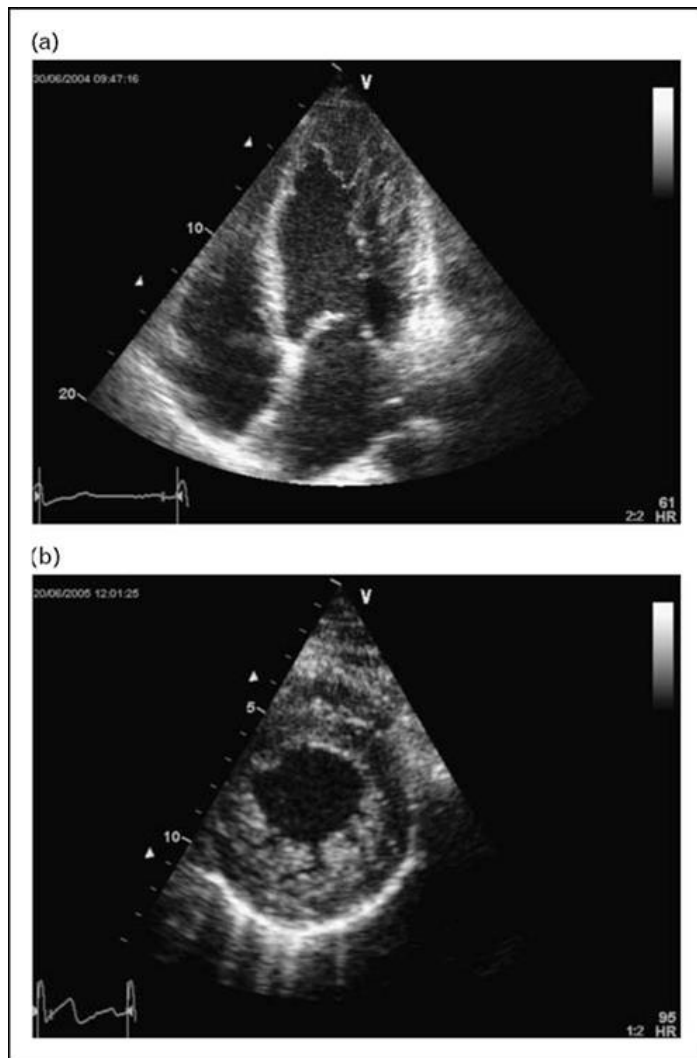


Figure 1. Apical echocardiographic view of a noncompacted left ventricle with extended and prominent trabeculations in the apical region and in the lateral wall. Two-layered appearance of the myocardium in a short axis view of the left ventricle.

Pantazis AA, Elliott PM. Left ventricular noncompaction. *Curr Opin Cardiol.* 2009;24:209-13.

Table 1. EHRA/HRS/APHRS/LAHS 2022 Consensus Statement on genetic testing.
Genes implicated in left ventricular noncompaction in adults

Gene	Locus	Syndrome	Protein (functional effect)	Frequency	ClinGen classification
<i>MHY7</i>	14q11.2	LVNC, DCM or HCM	Beta myosin heavy chain	10–15%	NA/major gene
<i>MYBPC3</i>	11p11.2	LVNC, DCM or HCM	Myosin binding protein C	5–15%	NA/major gene
<i>TTN</i>	2q31.2	LVNC, DCM	Titin	5–10%	NA/major gene
<i>ACTC1</i>	15q11.14	LVNC, DCM or HCM	Cardiac alpha-actin	1–5%	NA/rare gene
<i>RYR2</i>	1q43	LVNC, DCM	Ryanodine receptor type 2	1–2%	NA/rare gene
<i>PRDM16</i>	1p36	LVNC	PR domain zinc finger protein 16	1–2%	NA/rare gene
<i>LBD3</i>	11p15.1	LVNC, DCM	LIM domain binding 3	1–2%	NA/rare gene
<i>TBX5</i>	12q24.1	LVNC, Holt-Oram syndrome	T-box transcription factor 5	1–2%	NA/rare gene
<i>NKX2-5</i>	5q35.1	LVNC, DCM, conduction disease	Homeobox protein Nkx2-5	1–2%	NA/rare gene
<i>HCN4</i>	15q24.1	LVNC, conduction disease	Hyperpolarization-activated cyclic nucleotide-gated K ⁺ channel 4	1–2%	NA/rare gene
<i>TAZ</i>	Xp28	LVNC, Barth syndrome	Tafazzin	1–2%	NA/rare gene

DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; LVNC, left ventricular noncompaction.

Table 2. Spectrum of left ventricular noncompaction

Monogenic syndromes
 Chromosomal abnormalities
 Congenital heart disease
 Cardiomyopathies
 Muscular dystrophies
 Hematologic disorders (Sickle cell anaemia)
 Polycystic kidney
 Chronic renal failure
 Athletes
 Individual variability

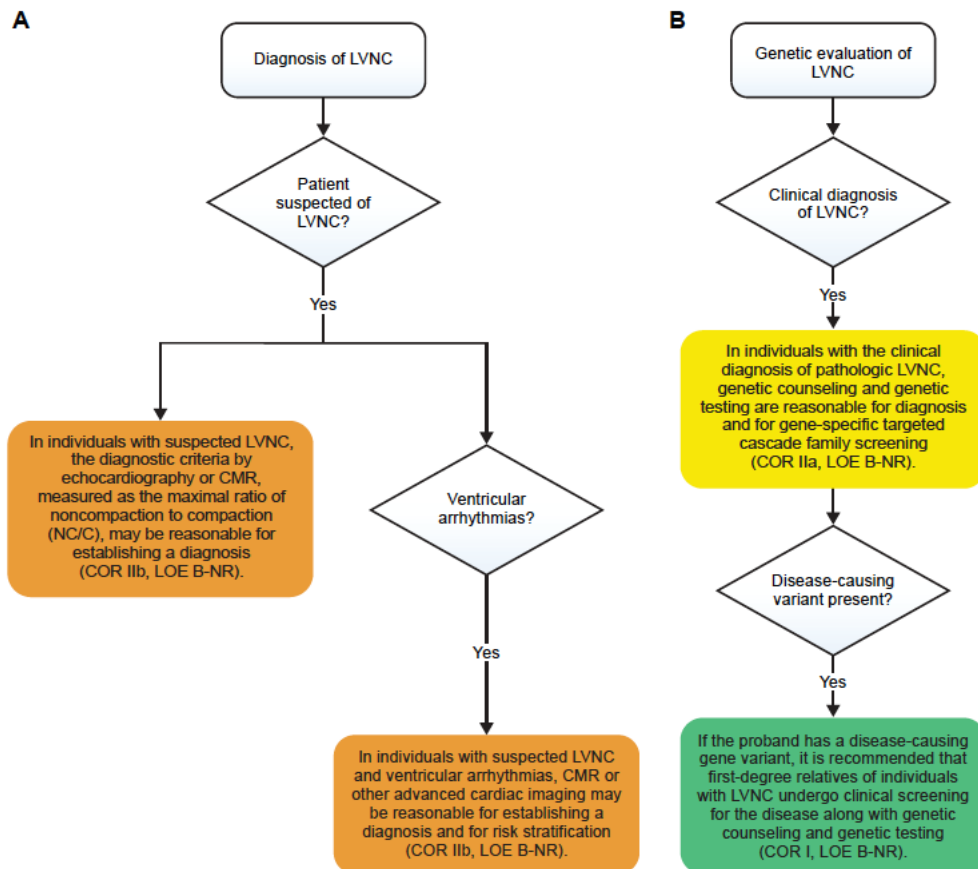


Figure 2. Diagnosis and risk stratification of LVNC (A) and family and genetic evaluation of LVNC (B).

COR=Class of Recommendation; LOE=Level of Evidence; LVNC = left ventricular noncompaction; CMR = cardiac magnetic resonance imaging; NC/C = maximum noncompaction to compaction ratio.

Towbin JA, et al. 2019 HRS expert consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy. *Heart Rhythm*. 2019.

**Table 3. EHRA/HRS/APHRS/LAHRs 2022 Expert Consensus Statement.
Arrhythmogenic Cardiomyopathy**

Impact of genetic testing for the index case

Disease	Diagnostic	Prognostic	Therapeutic
LVNC	+	+	–

Recommendations

May do

LVNC cardiomyopathy genetic testing for patients with an established clinical diagnosis of LVNC based on examination of the patient's clinical history, family history, and electrocardiographic/echocardiographic/MRI phenotype.

Genetic testing for patients with a clinical diagnosis of LVNC cardiomyopathy associated with other cardiac or noncardiac syndromic features.

Variant specific genetic testing for family members and appropriate relatives following the identification of the disease-causative variant.

Do not do

Genetic testing should not be performed in isolated (incidental) LVNC with normal LV function, no associated syndromic features and no family history.

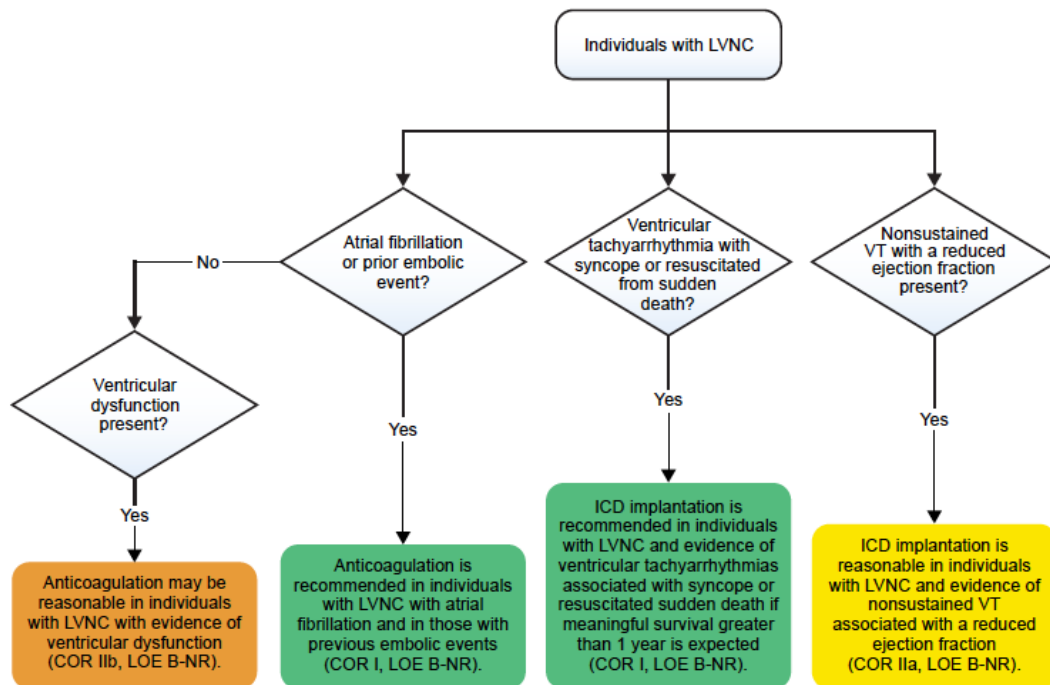


Figure 3. LVNC treatment recommendations. Anticoagulation refers to vitamin K antagonists and direct oral anticoagulants. Children are often administered aspirin.

COR = Class of Recommendation; ICD = implantable cardioverter defibrillator; LOE = Level of Evidence; LVNC = left ventricular noncompaction; VT = ventricular tachycardia.

Towbin JA, et al. 2019 HRS expert consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy. *Heart Rhythm*. 2019.

Table 4. ESC 2022 GL on VA and SCD. Recommendations for implantable cardioverter defibrillator implantation in left ventricular non-compaction

In patients with a LVNC cardiomyopathy
phenotype based on CMR or echocardiography,
implantation of an ICD for primary prevention of
SCD should be considered to follow DCM/
HNDCM recommendations. IIa C

CMR, cardiac magnetic resonance; DCM, dilated cardiomyopathy; HNDCM, hypokinetic non-dilated cardiomyopathy; ICD, implantable cardioverter defibrillator; LVNC, left ventricular non-compaction; SCD, sudden cardiac death.

Table 5. ESC 2020 GL on Sports. Recommendations for exercise in individuals with left ventricular non-compaction cardiomyopathy

Recommendations for diagnosis

A diagnosis of LVNC in athletic individuals should be considered if they fulfil
imaging criteria, in association with cardiac symptoms, family history of
LVNC or cardiomyopathy, LV systolic (EF<50%) or diastolic (E'<9 cm/s) dysfunction,
a thin compacted epicardial layer (<5 mm in end-diastole on
CMR, or <8 mm in systole on echocardiography), or abnormal 12-lead ECG. IIa B

Exercise recommendations

Participation in high-intensity exercise and all
competitive sports, with the exception where syncope may cause serious harm
or death, for asymptomatic individuals with LVNC and LVEF>_50% and absence
of frequent and/or complex VAs. IIb C

Participation in recreational exercise programmes of low to moderate intensity,
in individuals with LVEF 40_49% in the absence of syncope and frequent
or complex VAs on ambulatory Holter monitoring or exercise testing. IIb C

Participation in high- or very high-intensity exercise including competitive sports,
for individuals who are gene positive for LVNC but phenotype negative (with the
exception of lamin A/C or filamin C carriers). IIb C

Participation in high-intensity exercise or competitive sports is not recommended
in individuals with any of the following: symptoms, LVEF<40% and/or
frequent and/or complex VAs on ambulatory Holter monitoring or exercise testing. III C

Follow-up and further considerations

Annual assessment for risk stratification for individuals with LVNC
and genotypepositive/phenotype-negative individuals who exercise
on a regular basis. I C

CMR = cardiac magnetic resonance; ECG = electrocardiogram; EF = ejection fraction; LV = left ventricular;
LVEF = left ventricular ejection fraction; LVNC = left ventricular non-compaction; VA = ventricular arrhythmia.