

NON-DILATED LEFT VENTRICULAR CARDIOMYOPATHY

Definition

The non-dilated LV cardiomyopathy phenotype is defined by the presence of non-ischaemic LV scarring or fatty replacement in the absence of LV dilatation, with or without global or regional wall motion abnormalities, or isolated global LV hypokinesia without scarring (as assessed by the presence of LGE on CMR) that is unexplained solely by abnormal loading conditions (hypertension, valve disease) or CAD (Figure 1).¹ Thus it denotes conditions that may have variably been described as having DCM (but without LV dilatation), arrhythmogenic cardiomyopathy, left dominant ARVC, or arrhythmogenic DCM but often without fulfilling diagnostic criteria for ARVC.²

Diagnosis

A diagnostic approach is presented in Figure 2. Specific characteristics, especially electrocardiographic, are discussed in the Chapter on Classification and Diagnosis. Ambulatory ECG monitoring is recommended annually to reveal supraventricular and ventricular arrhythmias or bradycardias due to AV conduction block in ***non-dilated LV cardiomyopathy*** (Table 1). In patients at high risk of developing conduction disease and/or arrhythmias (including laminopathies, neuromuscular disease, PLN, and FLNC-truncating variants), Holter monitoring may be considered more frequently.² Male sex, baseline NSVT, LVEF < 45%, septal LGE, ring-like LGE, PVs/LPVs in high-risk genes, and myocardial inflammation are critical predictors of arrhythmic events in patients with non-dilated LV cardiomyopathy.³

Genetic Testing

The genes most commonly implicated in NDLVC are DSP, FLNC (truncating variants), DES, LMNA, or PLN, but there is substantial overlap with the genetic background of both DCM and ARVC. A detailed description can be found in Appendix 2. Desmoplakin variants, in particular, cause a unique form of cardiomyopathy with a high prevalence of LV fibrosis and myocardial inflammatory episodes.⁴

Therapy

General recommendations can be found in the Chapter on Epidemiology Diagnosis and Principles of Therapy. Recommendations for medical therapy, ICD implantation, and exercise are the same as in DCM.

References

1. Pinto YM, et al. Proposal for a revised definition of dilated cardiomyopathy, hypokinetic non-dilated cardiomyopathy, and its implications for clinical practice: a position statement of the ESC working group on myocardial and pericardial diseases. *Eur Heart J*. 2016;37:1850-1858. doi: 10.1093/eurheartj/ehv727
2. Arbelo E, et al. 2023 ESC Guidelines for the management of cardiomyopathies. *Eur Heart J*. 2023. doi: 10.1093/eurheartj/ehad194
3. Peretto G, et al. Major arrhythmias in non-dilated left ventricular cardiomyopathy: a novel prediction score. *Eur Heart J*. 2026;47:94-106. doi: 10.1093/eurheartj/ehaf477
4. Smith ED, et al. Desmoplakin Cardiomyopathy, a Fibrotic and Inflammatory Form of Cardiomyopathy Distinct From Typical Dilated or Arrhythmogenic Right Ventricular Cardiomyopathy. *Circulation*. 2020;141:1872-1884. doi: 10.1161/CIRCULATIONAHA.119.044934