2023 ESC Guidelines for the management of cardiomyopathies: the Ten Commandments

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The 2023 European Society of Cardiology Guidelines for the Management of Cardiomyopathies[1] represent the first international clinical practice guideline covering the full range of cardiomyopathies throughout the lifecourse. As this is a new guideline (except for hypertrophic cardiomyopathy, for which a focussed update has been provided), most of the 158 recommendations are new. Choosing the 10 most important aspects is therefore no easy task! Here, however, are 10 key themes explored in this new guideline.

1) **The patient pathway** – Running as a thread throughout the guideline is the concept of the patient pathway that requires a "cardiomyopathy mindset" to consider the possibility of a cardiomyopathy as the underlying cause of common clinical presentations (e.g. heart failure, arrhythmia, incidental findings, family history) and the importance of a systematic, multiparametric approach for its diagnosis and evaluation.
2) **A multidisciplinary approach** to evaluation and management, with close collaboration between expert cardiomyopathy centres and local services is recommended (Class I, LOE C).
3) **A diagnostic workflow** that incorporates the findings of several cardiac and extracardiac traits to arrive at a specific aetiological diagnosis, which in many cases will be genetic, is recommended (Class I, LOE C). This allows aetiology-driven management recommendations.
4) **Phenotypic description of cardiomyopathy subtypes** based on the identification of key morphological and functional myocardial traits, including the presence of myocardial scar on cardiac MRI, at clinical presentation. The resulting phenotype-based classification consists of hypertrophic cardiomyopathy (HCM), dilated cardiomyopathy (DCM), non-dilated left ventricular cardiomyopathy (NDLVC), arrhythmogenic right ventricular cardiomyopathy (ARVC) and restrictive cardiomyopathy (RCM). Importantly, these should not be considered diagnoses in their own right; rather, a careful and consistent description of the phenotype is only a crucial first step in the diagnostic pathway.
5) **Cardiac magnetic resonance (CMR) imaging** has a central role in the diagnosis of cardiomyopathies in patients (Class I, LOE B) and relatives (Class IIa, LOE B) and has important prognostic and therapeutic implications (Class IIa, LOE C).
6) **Genetic testing**, accompanied by genetic counselling, is a key aspect of the clinical workup in patients with cardiomyopathy (Class I, LOE B). For probands, it allows confirmation of an aetiological diagnosis and, in some cases, can have prognostic

implications and guide therapy and reproductive advice and management. For relatives of all ages, cascade genetic testing allows individuals who do not carry the familial variant to be discharged from clinical follow-up.

7) **Cardiac myosin inhibitors** (mavacamten) are recommended as second-line therapy for adults with symptomatic obstructive HCM (Class IIa, LOE A).
8) **Sudden death risk stratification** – a central aspect of the management of all cardiomyopathies, individualised risk assessment using validated risk prediction scores/algorithms is recommended (Class I/IIa, LOE B). These should be used as aids to shared decision-making.
9) **Genotype-based sudden death risk prediction** is particularly important in DCM/NDLVC, where identification of high-risk genotypes and accompanying clinical features informs ICD implantation decision-making (Class IIa, LOE B).
10) **Individualised exercise prescription** and regular low-moderate intensity exercise are recommended in all patients with cardiomyopathy (Class I, LOE C); more nuanced recommendations for different cardiomyopathy phenotypes, including gene carriers, are also made.

References

1. Arbelo E, Protonotarios A, Gimeno JR*, et al.* 2023 ESC Guidelines for the management of cardiomyopathies. *Eur Heart J* 2023;**44**:3503-3626. doi: 10.1093/eurheartj/ehad194