



2023 ESC Guidelines for Management of Cardiomyopathies: Key Points

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The following are key points to remember from the 2023 European Society of Cardiology (ESC) guidelines for the management of cardiomyopathies:

1. The overarching aim of this guideline is to provide a guide to the diagnostic approach to cardiomyopathies, highlight general evaluation and management issues, and make the reader aware of the relevant evidence base for the recommendations.
2. Evidence suggests that cardiomyopathies are more common than previously thought and they typically require nuanced management that may differ from the conventional approach to patients with arrhythmia or heart failure (HF).
3. Discerning etiology of the cardiomyopathy is fundamental to the management of patients with heart muscle disease, and careful and systematic description of the morphological and functional phenotype is a crucial first step in the diagnostic pathway.
4. Patients with cardiomyopathy may seek medical attention due to symptom onset (HF or arrhythmia related), incidental abnormal findings, or as a result of family screening following the diagnosis in a relative.
5. Multimodality imaging to characterize the cardiac phenotype (morphology and function)—including tissue characterization for nonischemic myocardial scar detection—is necessary, in combination with a detailed personal and family history, clinical examination, electrocardiography, and laboratory investigations. However, imaging results should always be interpreted in the overall clinical context, including genetic testing results, rather than in isolation.
 - Tissue characterization by cardiac magnetic resonance (CMR) is of value in diagnosis, monitoring of disease progression, and risk stratification in each of the main cardiomyopathy phenotypes.
 - 3,3-diphosphono-1,2-propanodicarboxylic acid (DPD)/pyrophosphate (PYP)/hydroxymethylene diphosphonate (HMDP) bone-tracer scintigraphy or single-photon emission computed tomography (SPECT) represent the gold standard for the diagnosis of ATTR-related cardiac amyloidosis.
6. The aim of this multiparametric and systemic approach is to generate a phenotype-based etiological diagnosis, interpreting available data with a cardiomyopathy-oriented mindset that combines cardiological assessment with noncardiac parameters. A multidisciplinary approach to patient care and appropriate transition of care from pediatric to adult cardiomyopathy services is needed.
7. Genetic testing should be performed in patients with cardiomyopathy and may influence risk stratification and management. Genetic counseling, including pre- and post-test counseling, and psychological support are an essential aspect of

- the multidisciplinary care of patients with cardiomyopathy and their relatives.
8. Pediatric cardiomyopathies largely represent part of the same clinical spectrum as those seen in older adolescents and adults, but infant-onset (in the first year of life) cardiomyopathies are often associated with severe phenotypes and a high rate of HF-related morbidity and mortality. Beyond the first year of life, genetic causes of childhood-onset cardiomyopathies are similar to those in adults.
 9. Symptom management, identification, and prevention of disease-related complications (including sudden cardiac death [SCD], HF, and stroke) are the cornerstone of management of all cardiomyopathies. Cardiac myosin inhibitors (mavacamten) should be considered in patients with hypertrophic cardiomyopathy (HCM) and left ventricular outflow tract obstruction (LVOTO) who remain symptomatic despite optimal medical therapy. Validated SCD risk-prediction tools (HCM Risk-SCD and HCM Risk-Kids) are the first step in sudden death prevention in patients with HCM.
 10. Pharmacological treatment of dilated cardiomyopathy (DCM) patients does not differ from those recommended in chronic HF. SCD risk of DCM and nondilated left ventricular cardiomyopathy (NDLVC) patients varies depending on the underlying cause and genetic subtype. CMR findings play an important role in guiding implantable cardioverter-defibrillator (ICD) implantation for patients with DCM and NDLVC. Of note, in DCM and NDLVC patients, ICD should be considered for certain genetic forms even if LVEF is $>35\%$.
 11. It is of importance to define etiology for a tailored management in patients with syndromic and metabolic cardiomyopathies (i.e., enzyme replacement therapy/chaperone in lysosomal storage disease; tafamidis in ATTRwt, etc.).
 12. Pregnancy and the post-partum period are associated with increased cardiovascular risk in women with known cardiomyopathy. A multidisciplinary team should evaluate the patient with cardiomyopathy to assess the risk associated with pregnancy. Beta-blocker therapy on arrhythmic indication can safely be continued during pregnancy; safety data should be checked before initiation of new drugs in pregnancy.
 13. Healthy adults of all ages and individuals with known cardiac disease should exercise with moderate intensity, totaling ≥ 150 minutes per week.
 14. All patients with cardiomyopathy should have an individualized risk assessment for exercise prescription. Evaluation should be guided by three principles:
 - Preventing life-threatening arrhythmias during exercise,

- Symptom management to allow sports, and
- Preventing sports-induced progression of the arrhythmogenic condition.

15. Patients with high-risk genotypes or associated factors for arrhythmic or HF complications or severe LVOTO should be referred for specialized investigations before undergoing elective noncardiac surgery. Identification and management of risk factors and concomitant diseases is recommended as an integral part of the management of cardiomyopathy patients.

Clinical Topics: Arrhythmias and Clinical EP, Heart Failure and Cardiomyopathies, Noninvasive Imaging, Prevention, Sports and Exercise Cardiology, Implantable Devices, SCD/Ventricular Arrhythmias, Atrial Fibrillation/Supraventricular Arrhythmias, Acute Heart Failure, Sports and Exercise and Imaging

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