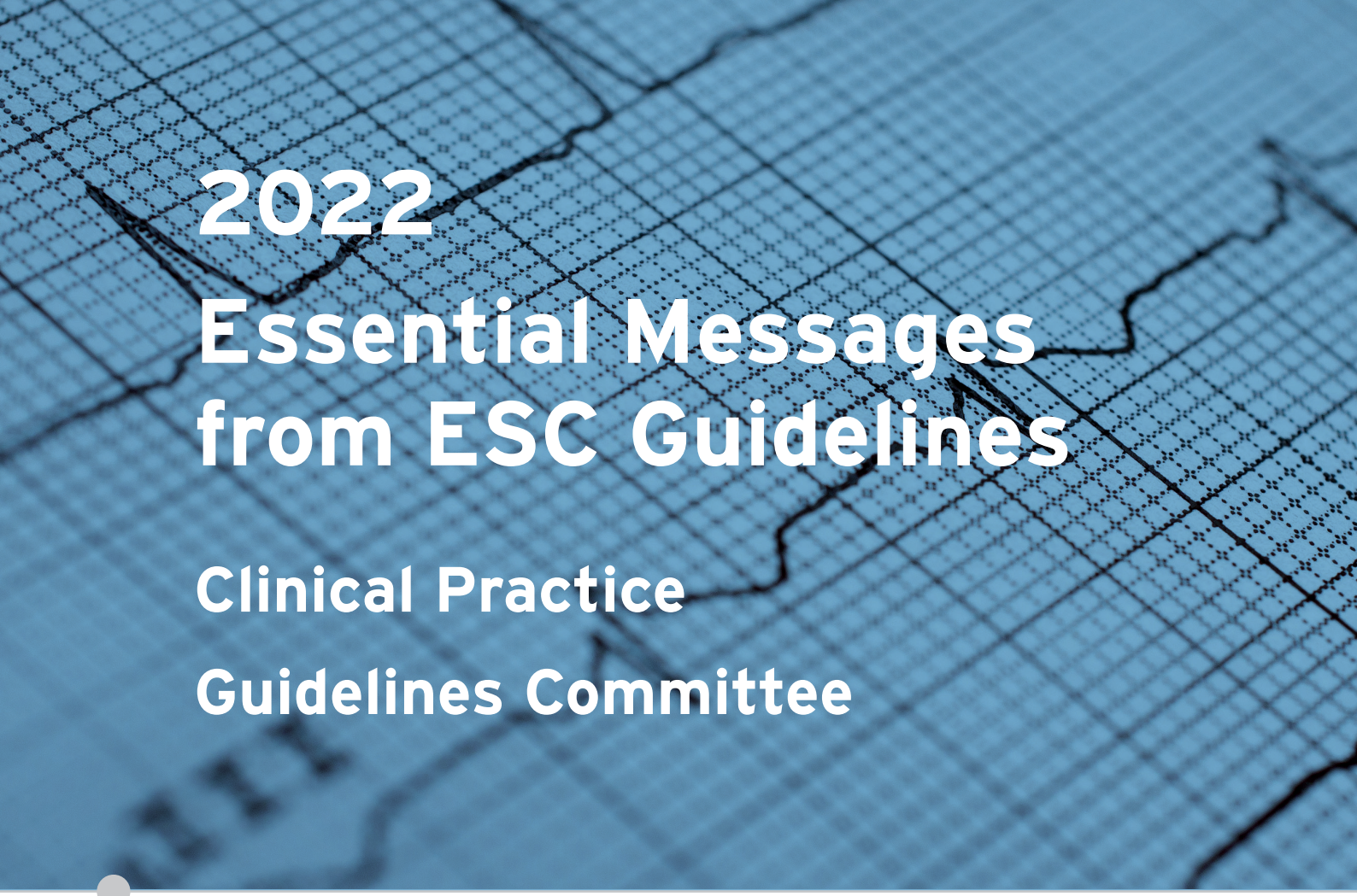

2022

Essential Messages from ESC Guidelines

Clinical Practice Guidelines Committee

VA and SCD

Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

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Essential Messages

2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Developed by the Task Force for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death of the European Society of Cardiology (ESC). Endorsed by the Association for European Paediatric and Congenital Cardiology (AEPC).

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Working Groups: *Cardiac Cellular Electrophysiology, Myocardial and Pericardial Diseases.*

Patient Forum

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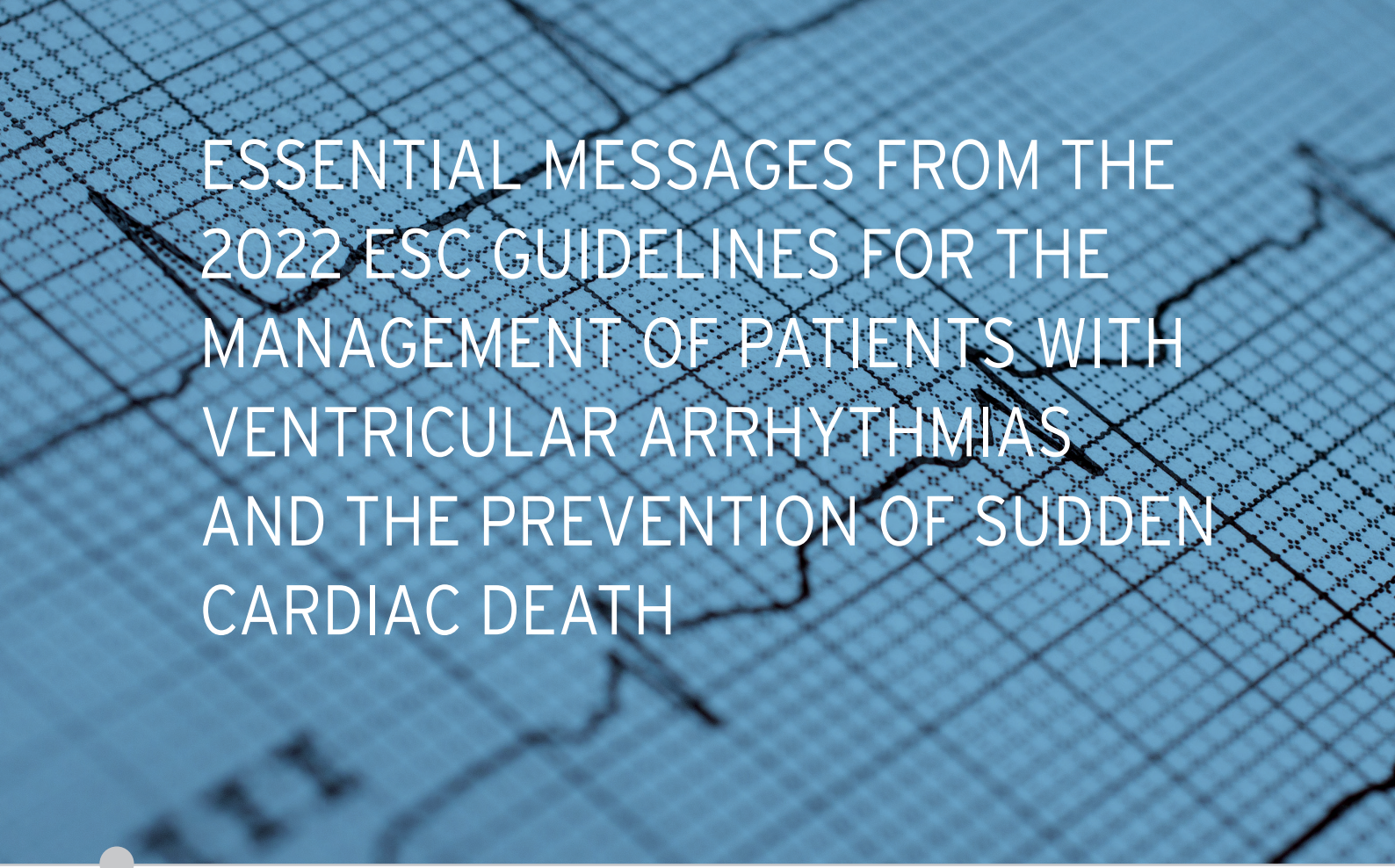

ESSENTIAL MESSAGES FROM THE 2022 ESC GUIDELINES FOR THE MANAGEMENT OF PATIENTS WITH VENTRICULAR ARRHYTHMIAS AND THE PREVENTION OF SUDDEN CARDIAC DEATH

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- Section 2 - Gaps in evidence

European Heart Journal; 2022 - doi: 10.1093/eurheartj/ehac262

Key messages

General aspects

- Increased availability of public access defibrillators and community training in basic life support are key elements to improve survival of out-of-hospital cardiac arrest victims.
- Risk calculators for SCD and VA implemented in clinical practice need to meet agreed high standards for the development, external validation, and reporting of prediction models.
- Patients with genetic cardiomyopathies and arrhythmia syndromes require genetic testing as a routine part of their care.
- Genetic testing and counselling require access to an expert multidisciplinary team.
- A systematic workup of cardiac arrest survivors requires a multimodal approach.
- A comprehensive autopsy is recommended in all cases of sudden death under 50 years and is desirable in all sudden death victims.
- Clinical and genetic evaluation of SADS decedents and their families leads to a diagnosis of genetic heart disease in a substantial proportion of families.
- An electrical storm refractory to drug treatment requires the availability of advanced catheter ablation techniques, mechanical circulatory support, and autonomic modulation.
- When considering ICD therapy benefit, competing risk factors for non-arrhythmic death, and the patient's wishes and quality of life need to be taken into account.

Structural heart disease

- Catheter ablation is recommended in CAD patients with recurrent symptomatic SMVT despite chronic amiodarone therapy.
- Catheter ablation is the first-line treatment for PVC-induced cardiomyopathy.
- In HNDCM/DCM patients, indication for primary prevention ICD implantation should not be restricted to a LVEF $\leq$ 35%. Additional risk factors (e.g. CMR and genetics) are important to consider.
- Patients with a LMNA mutation require specific risk stratification for SCD.
- ARVC patients have a high rate of appropriate ICD interventions which may not necessarily be classified as lifesaving.

- A validated risk calculator (HCM Risk-Kids score) is useful to assess the risk for SCD in HCM patients younger than 16 years.
- Myotonic dystrophy patients with palpitations suspected of arrhythmia, syncope, or aborted sudden death need to be evaluated by invasive electrophysiological study.
- In patients with repaired Tetralogy of Fallot and monomorphic VT, catheter ablation is the preferred treatment.

Primary electrical disease

- Nadolol or propranolol are the preferred beta-blockers in LQTS and CPVT patients.
- In asymptomatic LQTS patients the arrhythmic risk (1-2-3 risk calculator) may be useful to calculate.
- A type 1 Brugada ECG pattern provoked by sodium channel blocker test in the absence of other findings does not diagnose the BrS.
- SCD risk stratification in asymptomatic BrS patients with a spontaneous type 1 pattern remains controversial.
- Routine catheter ablation is not recommended in asymptomatic BrS patients.
- The diagnosis of idiopathic VF requires exclusion of an underlying structural, channelopathic, or metabolic aetiology.
- ERP can be a benign finding and is distinct from ERS.
- Left cardiac denervation plays an important role in the management of CPVT and LQTS patients.

Gaps in evidence

General aspects

- Accurate screening tests to detect heart conditions associated with SCD in asymptomatic individuals in the general population are required.
- In patients with SHD, the optimal time interval between repeated non-invasive and invasive prognostic tests, in case of a negative test, is unknown.
- Improved assessment of genetic variants of uncertain significance and likely pathogenic variants is needed.
- The utility of polygenic risk scores in patients at risk of SCD requires investigation.

Structural heart disease – general

- The long-term safety and efficacy of S-ICDs is unknown.
- The role of primary preventive ICD therapy in patients with SHD and mildly reduced or preserved ejection fraction has not been systematically studied.
- The optimal techniques to undertake VT substrate mapping and ablation in SHD remain to be determined.
- The role of ICD implantation in end-stage heart failure patients supported by current-generation, continuous-flow LVADs is unclear.

Idiopathic PVCs/VT

- The beneficial role of catheter ablation or antiarrhythmic drug treatment in patients with asymptomatic, frequent PVCs and preserved cardiac function needs to be determined.

Coronary artery disease

- It is unknown which patients with chronic CAD and severely impaired LVEF are at low risk for SCD.
- The role of LGE-CMR for risk stratification for SCD in chronic CAD is unclear.
- RCTs are needed to determine the role of ICDs after successful VT ablation in chronic CAD with mildly reduced or preserved LVEF.

Gaps in evidence

Cardiomyopathies

- It is unknown if PVC-induced cardiomyopathy is a diagnosis on its own or if an underlying predisposition is needed.
- The predictive value of LGE-CMR findings (e.g. pattern and amount of LGE) for individual risk stratification for SCD in patients with cardiac sarcoidosis, HCM, and DCM/HNDC is unclear.
- Studies are needed to determine the role of PES in patients with cardiac sarcoidosis and DCM/HNDC, who have a mildly reduced or preserved cardiac function and LGE on CMR.
- Prospective data on the association between intensity and duration of exercise and manifestation and severity of the phenotype in healthy ARVC mutation carriers are lacking.
- The beneficial role of ICDs after successful ablation in ARVC patients who present with haemodynamically tolerated VT needs to be studied.
- Data on clinical outcome, predictors for arrhythmic events, and indication for treatment, including ICD therapy, in patients with biventricular and left-dominant arrhythmogenic cardiomyopathy are required.

Valvular heart disease

- There is a lack of knowledge to identify patients with MVP at risk for VA and SCD.

Congenital heart disease

- There is a lack of knowledge regarding the absolute risk for VA and SCD in CHD that have undergone repair with contemporary surgical approaches.

Primary electrical disease

- Robust evidence is required to support the prophylactic use of the ICD on top of medical therapy with beta-blockers and gene-specific therapy in LQTS patients.
- More data are required to determine the role of LCSD and ICD in high-risk LQTS patients who do not tolerate medical therapy.
- Improved diagnostic and risk stratification tools for asymptomatic Brugada patients and suspected Early Repolarization syndrome are needed.

Gaps in evidence

- The role of endo-epicardial mapping to identify localized structural alterations potentially related to IVF and targeted catheter ablation needs to be further studied.
- Long-term data are required on the efficacy of the ICD versus no ICD in CPVT survivors of cardiac arrest.
- It is poorly understood why women are at low risk of sport-related SCD.

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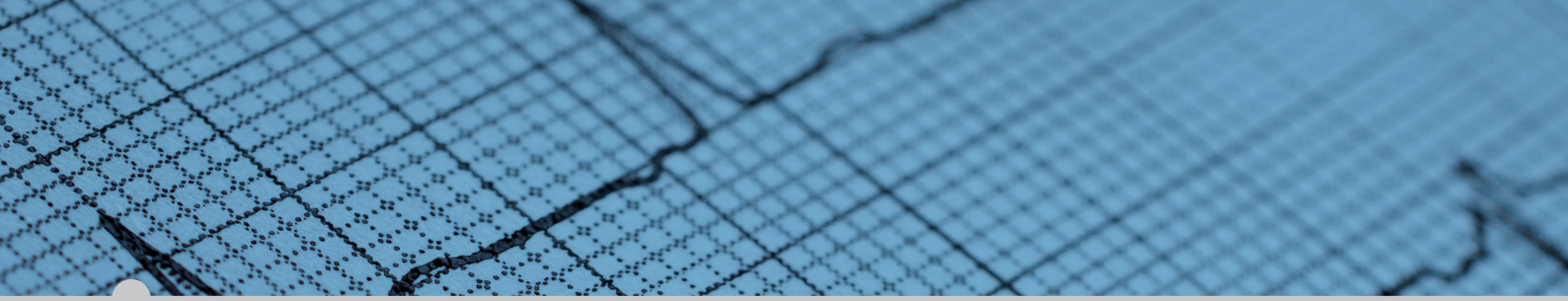

The material was adapted from the 2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death (European Heart Journal; 2022 - doi: 10.1093/eurheartj/ehac262).

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