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RUSSIAN GUIDELINES FOR SUDDEN CARDIAC DEATH RISK ASSESSMENT AND PREVENTION (SECOND EDITION) – 2018. POCKET VERSION

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Cardiovascular mortality in Russia is one of the highest in the world reaching 614 deaths per 100,000 annually. The main causes of death from cardiovascular diseases are the progression of congestive heart failure (about half of all cases) and sudden cardiac death (the other half). Thus, we can assume that the incidence of sudden cardiac death in 2016 was no less than 300,000. In the abbreviated version of the National Recommendations in English, the principles of decision-making algorithms in various clinical situations are used.

Keywords: *cardiovascular diseases, sudden cardiac death, risk, treatment, prevention.*

Glossary of Abbreviations

AAA – antiarrhythmic agents
AAP – accessory atrioventricular pathway
ACE inhibitors – angiotensin-converting enzyme inhibitors
ACM – alcoholic cardiomyopathy
ACS – acute coronary syndrome
AED – automatic external defibrillator
AF – atrial fibrillation
AH – arterial hypertension
ALS – advanced life support
ALV – artificial lung ventilation
AMI – acute myocardial infarction
ARVD – arrhythmogenic right ventricular dysplasia
AV – atrioventricular
AVNRT – atrioventricular nodal reentrant tachycardia
AVRT – atrioventricular reciprocating tachycardia
BLS – basic life support
BP – blood pressure
CAD – coronary artery disease
cAMP – cyclic adenosine monophosphate
CVDs – cardiovascular diseases
CHADS_2 – clinical prediction rules for estimating the risk of stroke in patients with atrial fibrillation \ atrial flutter
CHD – congenital heart defects
CI – confidence interval
CKD – chronic kidney disease
CMP – cardiomyopathy
CPR – cardiopulmonary resuscitation
CPVT – catecholaminergic polymorphic ventricular tachycardia
CRT – cardiac resynchronization therapy
CRT-D – cardiac resynchronization therapy defibrillator
CTD – connective tissue dysplasia
DCM – dilated cardiomyopathy
DM – diabetes mellitus
ECG – electrocardiogram
Echo – echocardiography
EPS – electrophysiology study
ERS – early repolarization syndrome
$etCO_2$ – end-tidal carbon dioxide
HCM – hypertrophic cardiomyopathy
HM-ECG – HolterECGmonitoring
HR – heart rate
HRV – heart rate variability
ICD – implantable cardioverter-defibrillator
ICD-10 – International Statistical Classification of Diseases and Related Health Problems 10th Revision
IE – infective endocarditis
LBBB – left bundle branch block LV – left ventricle
LVEF – left ventricular ejection fraction
LVH – left ventricular hypertrophy
LVOT – left ventricular outflow tract
LQTS – long QT syndrome
MVP – mitral valve prolapse
NSVT – non-sustained ventricular tachycardia
NSTEMI – non-ST segment elevation myocardial infarction
OSAHS – obstructive sleep apnea\hypopnea syndrome
OR – odds ratio
PaCO2 – carbon dioxide tension in arterial blood
PAP – positive airway pressure
PTH – parathyroid hormone
PM – pacemaker
PUFAs – polyunsaturated fatty acids
RR – relative risk
QT – QT interval
QTc – corrected QT interval
RV – right ventricle
RVOT – right ventricular outflow tract
PVC – premature ventricular contractions
SaO2 – arterial oxygen saturation
SCD – sudden cardiac death
SDNN – standard deviation of NN intervals
SIDS – sudden infant death syndrome
SSS – sick sinus syndrome
STEMI – ST segment elevation myocardial infarction
TGA – transposition of the great arteries
VA – ventricular arrhythmia
VF – ventricular fibrillation
VRD – ventricular rhythm disorder
VT – ventricular tachycardia
WPW syndrome – Wolff-Parkinson-White syndrome

1. Introduction

Cardiovascular mortality in Russia is one of the highest in the world reaching 614 deaths per 100,000 annually. The main causes of death from cardiovascular diseases are the progression of congestive heart failure (about half of all cases) and sudden cardiac death (the other half). Thus, we can assume that the incidence of sudden cardiac death in our country in 2016 was no less than 300,000.

2. Mechanisms and causes of SCD. Terms and definitions

Definition of SCD. Sudden cardiac death is a non-violent death occurring instantly or within an hour of the onset of acute changes in the patient's clinical status.

3. Classes of recommendations and levels of evidence

Levels of evidence for a statement can be classified as:

- *The highest* (Level A) – data derived from a large number of randomized controlled trials and/or meta-analyses.
- *Moderate* (Level B) – data derived from a limited number (at least one) of randomized controlled trials and/or well-designed controlled trials without randomization.
- *The lowest* (Level C) – statement is only based on individual case reports data and/or expert opinions.

Table III.1. Classes of recommendation

Classes of recommendation	Definition
Class I	Evidence and/or general agreement that a given treatment or procedure is beneficial, useful, effective
Class II	Conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of the given treatment or procedure.
Class IIa	Weight of evidence/opinion is in favor of usefulness/efficacy.
Class IIb	Usefulness/efficacy is less well established by evidence/opinion.
Class III	Evidence or general agreement that the given treatment or procedure is not useful/effective, and in some cases may be harmful.

5. SCD risk factors and risk stratification in clinical practice

Table V.1. Clinical features that characterize the increase in the probability of cardiac and non-cardiac causes of syncope

Clinical features associated with cardiac causes of syncope
Elderly age (> 60 y.o.)
Male sex
Presence of coronary artery disease, structural heart disease, previous arrhythmias or decreased ventricular function
Sudden loss of consciousness without a prodromal period or with a short prodromal period, for example, sudden onset of palpitation
Syncope during exercise
Syncope when supine
Small number of syncopal events (1 or 2)
Abnormalities in the cardiac examination
Family history of hereditary heart diseases or premature sudden cardiac death (before the age of 50)
Presence of congenital heart defects
Clinical features associated with non-cardiac causes of syncope
Young age
Absence of heart disease
Syncope while or after standing
Changes while standing/sitting from a supine position
Prodrome with such symptoms as nausea, vomiting, feeling of heat
Presence of specific triggers: dehydration, pain, stress, medical manipulations
Situational triggers: cough, laughing, micturition, swallow, defecation

Table V.2. Risk factors for short-term and long-term adverse outcomes in syncopal episodes

Risk factors for short-term adverse outcomes	***Risk factors for long-term adverse outcomes***
Evaluation at the outpatient stage or in the emergency department	
Male sex	Male sex
Elderly age (> 60 y.o.)	Elderly age (> 60 y.o.)
Absence of a prodrome	Absence of nausea/vomiting before a syncopal episode
Palpitation preceding loss of consciousness	Ventricular arrhythmias
Syncope during exercise	Cancer
Cerebrovascular diseases	Structural heart disease
Family history of sudden cardiac death	Heart failure
Traumatism	Cerebrovascular diseases
	Diabetes mellitus
	High risk by CHADS2 score
Examination results	
Symptoms of bleeding	ECG changes
Signs of disability	Reduction in glomerular filtration rate
Pathologic ECG changes	
High troponin levels	

Table V.4. Risk factors and probabilities of sudden cardiac death (adapted from Bigger JT, 1984)

	1-year risk of SCD, %
Moderate-risk group	
Previous AMI or LVEF<40%	5
Previous AMI + LVEF<40% or Previous AMI + prequent PVCs, or LVEF<40% + PVCs	10
Previous AMI + LVEF<40% + PVCs	15
High-risk group	
SCD survivor	30 – 50
VT with syncope	30 – 50
VT with minimal symptoms	20 – 30

Note. *AMI – acute myocardial infarction; LVEF – left ventricular ejection fraction; PVCs – premature ventricular contractions; SCD – sudden cardiac death; VT – ventricular tachycardia.*

Table V.5. Risk of sudden cardiac death in patients with cardiac rhythm and conduction abnormalities (adapted from Fogoros R, 2006)

High risk	
1.	Ventricular fibrillation
2.	Ventricular tachycardia
3.	III degree AV block with low rate escape rhythm
4.	WPW syndrome with antegrade conduction in AAP and atrial fibrillation
Moderate risk	
1.	Ventricular ectopy in presence of structural heart disease
2.	II degree AV block
3.	III degree AV block with adequate rate escape rhythm
4.	Atrial fibrillation
Low risk	
1.	Atrial ectopy
2.	Ventricular ectopy in absence of structural heart disease
3.	Supraventricular tachycardia
4.	I degree AV block

Note. *AAP – accessory atrioventricular pathways; WPW – Wolff–Parkinson–White; AV – atrioventricular.*

6. The research methods required for the SCD risk stratification

Table *VI.1.* Diagnostic tests that may be required for SCD risk stratification

Test	Comments	Class of recommen-dations	Level of evidence
Electrocardiography:			
12-lead surface ECG	Allows to discover congenital anomalies associated with high risk of SCD (e.g. long QT syndrome, short QT syndrome, Brugada syndrome, ARVD), and to identify other ECG criteria (e.g. signs of electrolyte abnormalities, conduction blocks in the His–Purkinje system, LVH signs)	I	C
	12-lead surface ECG should be administered to all patients during VRD examination	I	A
Holter monitoring	Is indicated in patients with symptoms of arrhythmia to determine if they are caused by life-threatening arrhythmias	I	B
	Is indicated in patients with PVCs on the ECG in absence of any other symptoms	I	B

Stress tests	Are recommended for adult patients with CAD risk factors and symptoms that may be associated with arrhythmias	I	B
	Are indicated in patients with previously verified or suspected VRD during exercise, including catecholaminergic polymorphic ventricular tachycardia	I	B
	May be considered in middle-aged and elderly patients with PVCs on the ECG in absence of any other symptoms	IIb	C
	ECG-stress test should be considered in order to assess the effectiveness of drug treatment or catheter ablation in patients with diagnosed VRD induced by exertion	IIa	C
Implantable loop recorders	Implantable loop recorders are indicated in patients with mild symptoms that may be associated with arrhythmias, for example, in case of syncope when standard 12-lead ECG cannot establish a causal relationship between the symptoms and heart rhythm abnormalities	I	B
Echocardiography	Is indicated in patients with suspected structural heart disease	I	B
	Is indicated in patients with high risk of SCD and cardiomyopathies (DCM, HCM, ARVD), postinfarction cardiosclerosis and family history of diseases associated with high risk of SCD	I	B
	Stress echocardiography is recommended to detect silent myocardial ischemia in patients with VRD, moderate risk of CAD, treated with glycosides; patients with LVH; patients with ST segment depression >1 mm at rest; patients with WPW syndrome or LBBB.	I	B
	Pharmacologic stress echocardiography is recommended to identify silent myocardial ischemia in patients with moderate risk of CAD who are not able to perform ECG-stress test	I	B
Genetic counseling and genetic testing (DNA testing)	Is aimed at identification and/or clarification of a hereditary disease diagnosis Is recommended for all patients and their family members with hereditary diseases, and should include discussion of risks and benefits of genetic testing	I	C
Biomarkers	BNP serum level determination in patients with structural heart disease can provide additional information of SCD prognosis	IIa	B
Magnetic resonance imaging (MRI)	Should be considered in patients with VRD, when Echo is not able to precisely evaluate LV and RV function and/or identify their structural abnormalities	IIa	B
	Should be considered to confirm the diagnosis of ARVD or non-compaction cardiomyopathy	IIa	B
Magnetic resonance imaging (MRI) with late gadolinium enhancement	Should be considered in patients with HCM to detect the presence and severity of intramyocardial fibrosis as a predictor of SCD	IIa	B
	May be considered for the detection of inflammatory or scar lesions of the myocardium in myocarditis and CAD as possible arrhythmogenic substrates	IIb	C
CT of the heart and coronary CT angiography	May be considered for the identification of prognostically unfavorable congenital anomalies of coronary arteries with ectopic origin from aortic sinus	IIb	C
	CT of the heart may be considered as an alternative to MRI in patients with contraindications for the latter or its inaccessibility	IIb	C
Coronary angiography	Is indicated in patients with CAD and life-threatening VRD (high and medium risk of SCD, see table V.2), as well as in cardiac arrest survivors	I	C
Intracardiac electrophysiology study (EPS)	Intracardiac EPS is recommended in patients with CAD and previous MI who have the following symptoms: palpitations, presyncope and syncope	I	B
	Intracardiac EPS is recommended in patients with structural heart disease and the following symptoms: palpitations, presyncope, syncope and bradyarrhythmia (including non-invasive methods data)	I	C

Notes: *ARVD – arrhythmogenic right ventricular dysplasia; LVH – left ventricular hypertrophy; PVCs – premature ventricular contractions, VA – ventricular arrhythmia; DCM – dilated cardiomyopathy; HCM − hypertrophic cardiomyopathy; LBBB – left bundle branch block, ICD – implantable cardioverter defibrillator; Echo – echocardiography.*

9. SCD risk stratification and prevention in patients with different comorbidities

IX.1. SCD risk stratification and prevention in patients with CAD

IX.1. A. SCD risk stratification and prevention in patients with postinfarction cardiosclerosis and left ventricular systolic dysfunction

Table IX.1.1 SCD risk stratification in patients with postinfarction cardiosclerosis and left ventricular systolic dysfunction

1. Is there a verified episode of cardiac arrest due to VF/VT?	
Yes	No
See p. 2	
2. Is there angina and/or signs of CAD destabilization*?	
Yes	No
Coronary angiography, consider revascularization	See section on recommendations for SCD prevention
3. Are there registered non-sustained ventricular arrhythmias**?	
Yes	No
Holter monitoring, consider coronary angiography, intracardiac EPS	See p. 4
4. Are there any clinical and instrumental signs of chronic left ventricular aneurysm?	
Yes	No
Consider cardiac surgery	See p. 5
5. LVEF is less than 40%	
Yes	No
See section on recommendations for SCD prevention	See section on SCD risk stratification and prevention in patients with chronic CAD and normal left ventricular systolic function

*Note * – CAD destabilization includes unstable angina (according to the defined of National guidelines for management of ACS without persistent ST-segment elevation), stable angina Class III–IV refractory to the adequate antianginal therapy, angina after myocardial revascularization procedures (PCI, CABG).*
*** – Non-sustained ventricular arrhythmias include non-sustained ventricular tachycardia (NSVT) and PVCs.*

Recommendations for SCD prevention

Class I

Adequate medical therapy of CAD and CHF that includes mandatory administration (if there are no contraindications and side effects) of the following drug classes:

- β-adrenergic blocking agents (A);
- ACE inhibitors (A);
- acetylsalicylic acid (A);
- statins (A);
- eplerenone after MI with reduced LVEF in combination with left ventricular failure or diabetes mellitus (B).

Restoration of coronary blood flow with surgical or interventional methods if possible (B).

For secondary SCD prevention, ICD placement is recommended in patients who survived VF or hemodynamically unstable VT episodes (major risk factors) and who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (A).

For primary SCD prevention, ICD placement is recommended in patients with left ventricular dysfunction due to prior MI (no less than 40 days after MI; a major SCD risk factor) with LVEF<40%, NYHA Class I–III, good functional status, who receive continuous optimal medical therapy and who are expected to survive for > 1year (A).

For primary SCD prevention, ICD placement is recommended for patients with following major risk factors of SCD: left ventricular dysfunction due to prior MI (no less than 40 days after MI), LVEF<40%, NYHA Class I–III, non-sustained VT (based on ECG, Holter monitoring) or sustained VT and/or VF (induced on intracardiac EPS), with good functional status, who receive continuous optimal medical therapy and who are expected to survive for > 1year (A).

Class IIa

Radiofrequency ablation (RFA) should be considered in patients with hemodynamically stable ventricular tachycardia (a major SCD risk factor) and LVEF > 40% (B).

RFA for VT should be considered in patients managed with ICD and AAA with frequent (more than 2 times a year) appropriate ICD shocks (C).

Class IIb

1. Amiodarone in combination with β-adrenergic blocking agents in patients with symptomatic VA (a major SCD risk factor) may be considered when β-adrenergic blocking agents are not effective (B).
2. Amiodarone may be considered in patients with VA (a major SCD risk factor), who are intolerant and/or refuse ICD implantation (C).
3. Sotalol may be considered in patients with symptomatic VA (a major SCD risk factor) when β-adrenergic blocking agents are not effective (C).
4. Surgical treatment of chronic cardiac aneurysm (C).
5. PUFAs (B).

Class III

1. AAA administration is not mandatory in patients with asymptomatic PVCs or non-sustained VT (a major SCD risk factor) (B).
2. Class IC antiarrhythmics (aa agents) are contraindicated (B).

IX.1.B. SCD risk stratification and prevention in patients with chronic CAD and normal left ventricular systolic function

Risk stratification

Table IX.1.2. SCD risk stratification in patients with chronic CAD and normal left ventricular systolic function

1. Is there transient or permanent myocardial ischemia and/or recurrent acute coronary episodes?	
Yes	No
Coronary angiography in order to choose revascularization method	See p. 2
2. Are there registered sustained/non-sustained ventricular arrhythmias?	
Yes	No
Coronary angiography in order to choose revascularization method	See section on recommendations for chronic CAD diagnosis and treatment

Recommendations for SCD prevention

Class I

1. Adequate medical therapy of CAD and CHF that includes mandatory administration (if there are no contraindications and side effects) of the following drug classes:

- β-adrenergic blocking agents (A);
- ACE inhibitors (A);
- acetylsalicylic acid (A);
- statins (A);
- PUFAs (B).

2. For secondary SCD prevention, restoration of coronary blood flow is recommended in patients who survived VT or hemodynamically unstable VT (major risk factors), since acute myocardial ischemia usually provokes VT (B).
3. For secondary SCD prevention, ICD implantation is recommended in patients who survived VF or hemodynamically unstable VT episodes (major risk factors), when coronary revascularization is not possible, and for those who receive continuous optimal medical therapy, have good functional status* and who are expected to survive for > 1year (A).

Class IIa

1. Administration of amiodarone in combination with β-adrenergic blocking agents should be considered to reduce severity of symptoms caused by recurrent hemodynamically stable VT (major SCD risk factors) in patients with LV dysfunction due to acute myocardial infarction, who are intolerant or refuse ICD implantation (C).

2. Surgical and/or interventional restoration of coronary blood flow for primary SCD prevention should be considered in patients with chronic CAD and hemodynamically significant stenosis of the coronary arteries (C).

3. ICD implantation should be considered for the treatment of recurrent sustained VT in patients with a history of previous MI (a major SCD risk factor) with normal or close to normal systolic ventricular function, who receive continuous optimal medical therapy, have good functional status* and who are expected to survive for > 1year (C).

Class IIb

1. Radiofrequency catheter ablation or amiodarone administration may be considered as an alternative to ICD implantation in patients with moderate left ventricular dysfunction (LVEF>40%) and recurrent hemodynamically stable VT (a major SCD risk factor) (B).

Class III

1. Antiarrhythmic agents are not recommended as a preventive measure to reduce mortality in patients with non-sustained asymptomatic VA (a major SCD risk factor) (B).

* You can read about functional status assessment methods at http://www.chcr.brown.edu/pcoc/ functi. html.

IX.2. SCD risk stratification and prevention in patients with chronic heart failure

Table IX.2.1. SCD risk stratification in patients with CHF

1. Is there evidence of ischemic etiology of CHF?	
Yes	No
Coronary angiography, consider revascularization	See p. 2
2. Is there history of cardiac arrest?	
Yes	No
See section on recommendations for SCD prevention – Class I, paragraph 1.	Prevention measures depend on: NYHA Class LVEF VA presence ventricular dyssynchrony presence (See section on recommendations for SCD prevention)
3. Are there registered any sustained/non-sustained ventricular arrhythmias?	
Yes	No
Holter monitoring, consider intracardiac EPS	See section on recommendations for SCD prevention

Recommendations for SCD prevention

Class I

1. Adequate medical treatment of CHF according to current National guidelines on the CHF treatment includes mandatory administration (in absence of contraindications and side effects) of β-adrenergic blocking agents (A), ACE inhibitors (A), aldosterone antagonists (A).

2. For secondary SCD prevention, ICD implantation is recommended in patients who survived VF or hemodynamically unstable VT episodes that were not due to reversible causes (major risk factors), and who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (A).

3. For primary SCD prevention, ICD implantation is recommended in patients with left ventricular dysfunction due to prior MI (no less than 40 days after MI; a major SCD risk factor) with LVEF<40%, NYHA Class I–III, and who receive continuous optimal medical therapy, have good functional status

and who are expected to survive for > 1year (A).

4. For primary SCD prevention, ICD placement is recommended in patients with non-ischemic heart diseases with LVEF<35%, NYHA Class I–III (a major SCD risk factor), who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (A).

5. Concomitant therapy with amiodarone or sotalol alone or in combination with β-blockers is recommended in patients with ICDs who receive CHF treatment, to reduce symptoms of ventricular tachycardia (both sustained and non-sustained) (C).

6. Amiodarone is indicated for treatment of hemodynamically significant VT and non-sustained VT if cardioversion and/or correction of the arrhythmia causes cannot effectively eliminate or prevent its early recurrence (B).

Class IIa

1. For primary SCD prevention, ICD combined with biventricular pacemaker implantation (CRT) should be considered in patients with NYHA Class III–IV (a major risk factor), who receive continuous optimal medical therapy, in sinus rhythm with a QRS duration >120 ms and who are expected to survive for > 1year (A).

2. ICD placement should be considered in patients with recurrent hemodynamically stable VT (a major risk factor), normal or close to normal LVEF, who receive optimal CHF treatment with good functional status and who are expected to survive for > 1year (C).

3. Biventricular pacemaker insertion without ICD function is appropriate to prevent SCD in patients with NYHA Class III–IV, LVEF<35% (major risk factors), with a QRS duration >160 ms (or at least 120 ms if other signs of asynchronous ventricular contraction are present), who receive continuous optimal medical therapy and who are expected to survive for > 1year (B).

Class IIb

1. Amiodarone, sotalol and/or β-adrenergic blocking agents may be considered in patients with major and minor SCD risk factors, who receive optimal CHF treatment, and when ICD implantation is impossible (B).

2. For primary SCD prevention, ICD implantation may be considered in patients with non-ischemic heart diseases with LVEF 30–35% (major risk factor), NYHA Class I, who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (B).

Class III

1. Class IC antiarrhythmic agents administration for VA treatment (a major SCD risk factor) is not recommended in patients with CHF (A).

2. ICD implantation is not indicated in patients with refractory CHF who are not expected to achieve the compensation of clinical manifestations and are not expected to survive for >1year (A).

SCD is the causes of about 50% deaths in patients with CHF.

Table IX.2.1. Use of ICD for primary SCD prevention in patients with CHF

NYHA Class	LVEF, %					
	<30		31 – 35		36 – 40	
	CHF etiology					
	Ischemic	Non-ischemic	Ischemic	Non-ischemic	Ischemic	Non-ischemic
Class I	I (not within 40 days of MI)	IIb	I (NSVT+)	IIb	I (NSVT+)	III
Class II	I (not within 40 days of MI)	I	I (not within 40 days of MI)	I	I (NSVT+)	III
Class III	I (not within 40 days of MI)	I	I (not within 40 days of MI)	I	I (NSVT+)	III
Class IV	III	III	III	III	III	III

Notes: *Roman numerals in the table show the indication class for ICD placement. LVEF – left ventricular ejection fraction, CHF – congestive heart failure, NSVT – non-sustained ventricular tachycardia, MI –myocardial infarction.*

IX.3. SCD risk stratification and prevention in patients with bradyasystolic arrhythmias and conduction disorders

Term “Bradyarrhythmia” includes a wide range of diseases with pathogenesis involving reduction of cardiac output by reducing the heart rate due to sinus node dysfunction and/or abnormalities in action potential propagation along the cardiac conduction system. SCD due to bradyarrhythmias occurs in 15% of all cases. However, it is important to realize that the coexistence of bradyarrhythmias and LV systolic dysfunction in the same patient suggests high or moderate risk of VT.

IX.3.A. SCD risk stratification and prevention in patients with sinus node dysfunction

Risk stratification

Sinus node dysfunction leading to severe bradycardia or sinus pauses can manifest with syncope, presyncope, dizziness, hypotension, symptoms of heart failure progression, angina pectoris. SCD in patients with SSS is more likely in presence of LV systolic dysfunction. Presence of such risk factors as syncope, structural heart disease and long-term symptomatic asystolic pauses during Holter monitoring correlate with poor prognosis, including SCD in particular. At the same time, it should be mentioned that the main thing is not the duration of the pause, but its symptomatic significance. (Table V.5).

SCD prevention

Permanent atrial and/or dual chamber pacing in patients with SSS improves symptoms and quality of life, reduces incidence of atrial fibrillation and frequency of its episodes. Long-term results of permanent pacing and its effects on survival and SCD incidence at present remain unknown.

IX.3.B. SCD risk stratification and prevention in patients with AV- and intraventricular conduction abnormalities

Recommendations for SCD prevention

Permanent dual chamber pacing in accordance with the National guidelines on cardiac pacing in patients with AV conduction abnormalities improves symptoms and quality of life and reduces morbidity. Current data on the long-term effects of permanent pacing on the survival and SCD occurrence at present are contradictory.

IX.4. SCD in patients with cardiomyopathies

IX.4.A. SCD risk stratification and prevention in patients with dilated cardiomyopathy

Recommendations on genetic testing

Class I

1. For all patients with DCM careful study of the family history for at least 3 generations is recommended (A).
2. Cardiologic screening of first-degree relatives is recommended: history taking, physical examination, ECG, Echo, Holter monitoring (in case of proband’s death) (A).
3. If first-degree relatives have signs of DCM, regular cardiac examinations should start from early childhood (from 10–12 years in patients with laminopathy) every 12–36 months up to 10 years, every 12–24 months up to 20 years and then every 2–5 years up to 50–60 years; cardiologic screening of proband descendants of each subsequent generation is recommended (B).
4. If the signs of DCM in relatives are not found and genetic testing is impossible, regular cardiac examination is recommended, starting from early childhood and every 12–36 months up to 10 years, every 12–24 months up to 20 years and then every 2–5 years up to 50–60 years. (B).
5. If a causative mutation is detected in a proband with DCM in the result of genetic testing, genetic screening of first-degree relatives is recommended (B).
6. If there are no causative mutations in relatives, further examination should be stopped (C).
7. If there are no causative mutations in relatives, genetic testing of descendants should not be performed (C).

Class IIa

1. In case of presence of causative mutations in relatives of asymptomatic carriers of causative mutations, a regular cardiologic examination should be considered every year from 10 to 20 years and then every 1–3 years with the use of additional examination methods if necessary (C).

Risk stratification

Approaches to SCD risk stratification in patients with dilated cardiomyopathy do not differ from those used for risk stratification in patients with non-ischemic CHF (see Table IX.2.1). In DCM, an extremely high risk of SCD is associated with mutations in the LMNA gene.

Recommendations for SCD prevention

Class I

1. Adequate medical treatment of CHF according to current National guidelines on the CHF treatment includes mandatory administration (in absence of contraindications and side effects) of β-adrenergic blocking agents (A), ACE inhibitors (A), aldosterone antagonists (A).

2. For secondary SCD prevention, ICD implantation is recommended in patients who survived VF or hemodynamically unstable VT episodes that were not due to reversible causes (major risk factors), and who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (A).

3. For primary SCD prevention, ICD implantation is recommended in patients with non-ischemic heart diseases with LVEF<35%, NYHA Class II–III, and who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (A).

4. Catheter ablation of right bundle branch is indicated in patients with bundle branch re-entry ventricular tachycardia (a major risk factor), confirmed with intracardiac EPS (C).

5. Concomitant therapy with amiodarone or sotalol alone or in combination with β-blockers is recommended in patients with ICDs, who receive DCM treatment, to reduce symptoms of ventricular tachycardia (both sustained and non-sustained) (C).

6. Amiodarone is indicated for treatment of hemodynamically significant VT and non-sustained VT if cardioversion and/or correction of the arrhythmia causes cannot effectively eliminate or prevent its early recurrence (B).

Class IIa

1. For primary SCD prevention, ICD combined with biventricular pacemaker (CRT) implantation should be considered in patients with DCM and NYHA Class III–IV (a major risk factor), who receive continuous optimal medical therapy, in sinus rhythm with a QRS duration >120 ms and who are expected to survive for > 1year (B).

2. ICD implantation should be considered in patients with recurrent hemodynamically stable VT (a major risk factor) and slightly reduced LVEF, who receive optimal DCM treatment with good functional status and who are expected to survive for > 1year (C).

3. Biventricular pacemaker insertion without ICD function is appropriate to prevent SCD in patients with DCM and NYHA Class III–IV, LVEF<35% (major risk factor), with a QRS duration >160 ms (or at least 120 ms if other signs of asynchronous ventricular contraction are present), who receive continuous optimal medical therapy and who are expected to survive for > 1year (B).

Class IIb

1. Amiodarone and/or β-adrenergic blocking agents may be considered in patients with major and minor SCD risk factors, who receive optimal CHF treatment, and when ICD implantation is impossible (B).

2. For primary SCD prevention, ICD implantation may be considered in patients with DCM and LVEF 30–35% (major risk factor), NYHA Class I, who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (B).

Class III

1. Class IC antiarrhythmic agents administration for VA treatment (a major SCD risk factor) is not recommended in patients with CHF (A).

2. ICD implantation is not indicated in patients with refractory CHF who are not expected to achieve the compensation of clinical manifestations and are not expected to survive for >1year (A).

IX.4.B. SCD risk stratification and prevention in patients with hypertrophic cardiomyopathy

SCD risk stratification

Table IX.4.1. SCD risk factors in patients with hypertrophic cardiomyopathy

Main (major) SCD risk factors	SCD development is possible in some individuals
Cardiac arrest due to VF or VT Spontaneous sustained ventricular tachycardia Family history of SCD Unexplained syncope Left ventricle walls thickness ≥ 30mm Spontaneous non-sustained ventricular tachycardia	Atrial fibrillation Myocardial ischemia Presence of high risk gene mutations Intensive (competitive) physical activity

Recommendations for SCD prevention

Table IX.4.2. Primary SCD prevention in patients with HCM

Riskfactor	Comments
Age	• Some risk factors are more significant in young patients, especially NSVT, extreme LVH, and unexplained syncope.
Non-sustained ventricular tachycardia	• NSVT (defined as an episode of ventricular tachycardia with a heart rate of at least 120 beats per minute, lasting for at least three beats and persisting less than 30 seconds) develops in 20–30% of patients during Holter monitoring and is an independent SCD risk factor. • SCD prevention measures include administration of β-blockers (I, B), amiodarone (IIb, C), ICD-therapy if indicated (I, B).
Maximal left ventricle walls thickness	• The severity and prevalence of LVH measured by transthoracic echocardiography is associated with the risk of SCD. • Several studies have shown the highest risk of SCD in patients with a left ventricle wall thickness ≥30 mm.
Family cases of SCD in young age	• Although the definitions differ, family cases of SCD are usually considered clinically significant if one or more of the first-degree relatives died suddenly at the age of <40 years, with HCM diagnosis or without it; or when SCD developed in a first-degree relative with a confirmed diagnosis of HCM at any age.
Syncope	• Syncope are often found in patients with HCM, but their causes can be diverse. • Non-neurocardiogenic syncope with no etiological explanation after the examination are considered a SCD risk factor. • Syncope occurring within 6 months from the examination have the highest predictive value regarding SCD. • SCD prevention measures if indicated: septal myectomy (I, C), alcohol ablation (IIb, B), ICD therapy (I, B), dual chamber pacemaker insertion (IIb, B)
Left ventricular outflow tract obstruction	• A large number of studies have shown a significant association between LVOT obstruction and SCD. • SCD prevention measures include administration of β-blockers (I, B), septal myectomy (I, C), alcohol ablation (IIb, B), dual chamber pacemaker insertion (IIb, B)
Blood pressure response during exercise	• Approximately one-third of adult patients with HCM have an abnormal response of systolic blood pressure during exercise, characterized by progressive hypotension. • An abnormal response of systolic blood pressure to the exercise is defined as the inability to increase the pressure by at least 20 mmHg from the rest level at the peak of physical activity or a pressure drop of > 20 mm Hg of the peak pressure. • An abnormal response to systolic blood pressure to the exercise is associated with an increased risk of SCD in patients younger than 40 years.

Class I

1. ICD implantation is indicated in patients with HCM and risk of sudden cardiac death at 5 years of ≥4% (calculated by the HCM Risk-SCD model) and such major SCD risk factors as sustained VT or VF, who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (B).

2. Beta-adrenergic blocking agents are recommended in the symptomatic adult patients with obstructive or non-obstructive HCM, but they should be used with caution in patients with sinus bradycardia or AV conduction disorders (B).

3. Septal myectomy is indicated in patients with severe, refractory to medical therapy symptoms* and LVOT obstruction (C)**.

* Signs and symptoms include angina pectoris CCS Class III–IV, syncope, presyncope, dizziness, hypotension that are refractory to optimal medical therapy. LVOT pressure gradient at rest or during exercise should not exceed 50 mmHg.

** Surgery should be performed only by experienced surgeons (who have performed at least 20 procedures or who are practicing at a center, where at least 50 of such procedures are performed annually).

Class IIa

1. ICD implantation should be considered for primary and secondary prevention of SCD in patients with HCM who have at least one major risk factor (see Table VII.4.1): cardiac arrest, spontaneous sustained VT, family history of SCD, unexplained syncope, LV walls thickness ≥ 30mm, BP abnormalities during stress testing, spontaneous non-sustained ventricular tachycardia, who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (B).

2. Ethanol embolization should be considered in adult HCM patients with LVOT obstruction (a major risk factor in these patients) if signs and symptoms are refractory to medical therapy and there are contraindications for myotomy/ myectomy due to serious concurrent medical conditions and/or advanced age (B).

3. Amiodarone should be considered the drug of choice in patients with HCM and history of persistent VT and/or VF (major risk factors), when ICD placement is contraindicated (C).

4. Expanded myoectomy should be considered in patients with obstructive HCM and resistance to drug therapy (C).

Class IIb

1. ICD implantation may be considered in patients with HCM and risk of sudden cardiac death at 5 years of≤4% (calculated by the HCM Risk-SCD model) only if there are clinical manifestations that have proven prognostic significance (B).

2. Amiodarone use may be considered for the primary prevention of SCD in patients with HCM who have one or more major SCD risk factors, when ICD implantation is impossible (C).

3. Permanent dual chamber pacing with a short AV delay may be indicated in patients with obstructive HCM with severe signs and symptoms that are refractory to medical therapy and who are not candidates for septal rablation procedure if LVOT systolic gradient falls by 25% or more during preliminary dual-chamber pacing with optimal AV delay (B).

4. Sotalol use experience in patients with an HCM is limited, but it may be considered in certain clinical situations, particularly in patients with ICDs (C).

Class III

1. ICD implantation is not recommended in patients with HCM and no major SCD risk factors (C).

2. ICD implantation is not recommended in patients with HCM and risk of sudden cardiac death at 5 years of≤4% (calculated by the HCM Risk-SCD model) (B).

3. ICD implantation is not recommended for patients with HCM and a positive genotype (a possible risk factor) without clinical signs and symptoms (C).

4. Ethanol embolization is not indicated in patients with severe septal hypertrophy (>30 mm) due to uncertain effectiveness of the procedure in these patients (C).

5. Ethanol embolization is not indicated in asymptomatic patients, patients with medically controlled symptoms or in patients with a planned cardiac surgery when myectomy may be performed as a part or a stage of this surgery (C).

6. Nitrates, nifedipine and high doses of diuretics are potentially dangerous in patients with obstructive HCM (C).

7. Cardiac glycosides use in patients with HCM in absence of AF is potentially dangerous (B).

IX.4.1.C. SCD risk stratification and prevention in patients with arrhythmogenic right ventricular dysplasia

Currently, «arrhythmogenic right ventricular dysplasia / cardiomyopathy» is defined as the pathology of the heart muscle, often familial one, characterized by a structural and functional anomaly of the right ventricle, associated with the replacement of the myocardium with fat or fibrous tissue.

Choice of management tactics

Patients with ARVD can be divided into five groups.

1. Patients with a limited form of dysplasia in combination with asymptomatic or low-symptomatic PVCs, without a family history of ARVD. In these cases, systemic antiarrhythmic therapy, with the exception of β-blockers, is not indicated. Undoubtedly, these patients, like all patients with ARVD, should avoid intense physical activity and exercise.

2. In presence of local forms of ARVD in combination with symptomatic PVCs, paroxysms of sustained or non-sustained ectopic tachycardia, antiarrhythmic therapy should be indicated. In case of inefficiency or impossibility of drug treatment, it is efficient to perform catheter ablation of arrhythmogenic foci.

3. To prevent episodes of stable, hemodynamically significant VT without significant right ventricular myocardial dysfunction, an antiarrhythmic drug should be selected with help of an intracardiac EPS. If the recurrence of the tachycardia persists, catheter ablation or ICD implantation is indicated.

4. Patients with paroxysms of sustained VT in presence of severe myocardial dysfunction, as well as SCD survivors, require ICD implantation in combination with antiarrhythmic therapy and concomitant CHF treatment.

5. In rare cases of ARVD with prevalence of myocardial dysfunction (e.g. Uhl disease) or biventricular pathology without VT or VF episodes, correction of CHF is indicated. ICD implantation may be considered, but there are currently no reliable data on its effectiveness in this group of patients.

IX.5. SCD in patients with WPW syndrome

SCD risk stratification

Table IX.5.1. SCD risk stratification in patients with WPW syndrome

Recommendations for SCD prevention

1. Is there evidence of pre-excitation syndrome on ECG?	
Yes	No
See p. 2	
2. Is there symptomatic tachycardia or syncope history?	
Yes	No
Intracardiac EPS and RFA of the AAP	See p. 3
3. Is there family history of WPW syndrome and SCD?	
Yes	No
Intracardiac EPS and RFA of the AAP	See p. 4
4. Is there a structural heart disease?	
Yes	No
Intracardiac EPS and RFA of the AAP	See p. 5
5. Is ventricular pre-excitation asymptomatic?	
See section on recommendations for SCD prevention	

Class I

1. In patients with evidence of ventricular pre-excitation on the ECG, history of cardiac arrest, unexplained syncope (major risk factors) or symptomatic tachycardia RFA of accessory atrioventricular pathway is indicated (B).

2. RFA is indicated in patients with atrial fibrillation (or another atrial tachycardia), accompanied by high-frequency activation of ventricular myocardium (RR interval ≤ 260 ms with anterograde conduction via accessory atrioventricular pathway – a major SCD risk factor) (B).

3. In patients with WPW syndrome and major risk factors who prefer medication therapy to RFA, Class I antiarrhythmic agents or amiodarone are the drugs of choice (C).

4. RFA of accessory atrioventricular pathway is indicated in patients of high risk occupations (aircraft pilots, public transport drivers, athletes) who are diagnosed with WPW syndrome/pattern regardless of the presence of symptoms and the anterograde effective refractory period (ERP) length even if major SCD risk factors are absent (B).

5. RFA of accessory atrioventricular pathway is indicated in patients with WPW pattern and anterograde ERP of accessory AV conduction pathway ≤270 ms (a major risk factor) (B).

Class IIa

1. Regular cardiologic checkups should be considered in patients with ECG ventricular pre-excitation signs in absence of major risk factors (history of symptomatic tachycardia, syncope, family history of SCD, structural heart disease, anterograde ERP in accessory atrioventricular pathway >270 ms) (C).

2. Antiarrhythmic agents should not be considered in patients with ECG ventricular pre-excitation signs in absence of major risk factors (history of symptomatic tachycardia, syncope, family history of SCD, structural heart disease, anterograde effective refractory period in accessory AV pathway >270 ms) (C).

Class IIb

1. RFA may be considered in patients with WPW pattern with anterograde ERP in accessory atrioventricular pathway >270 ms (C).

Class III

1. Digoxin, β-blockers, verapamil and ATP are contraindicated in patients with WPW syndrome/pattern (C).

IX.6. SCD in atrial fibrillation

Recommendations for SCD prevention

Class IIa

1. According to the current National Guidelines, adequate medical therapy of patients with CAD and postinfarction cardiosclerosis should include the use (if not contraindicated) of β-blockers, ACE inhibitors, acetylsalicylic acid, statins and other agents that reduce the risk of SCD in patients with these diseases.

2. According to the current National Guidelines, adequate medical therapy of CHF should include the use (if not contraindicated) of β-blockers, ACE inhibitors, aldosterone antagonists and other agents that reduce the risk of SCD in patients with CHF.

Class IIb

1. According to the current National Guidelines, adequate medical therapy of diseases accompanied by severe left ventricular hypertrophy (hypertrophic cardiomyopathy, arterial hypertension, etc.) may include the use (if not contraindicated) of β-blockers and other agents that reduce the risk of SCD in patients with these diseases.

2. In presence of arrhythmic syndromes with the possibility of SCD, in particular WPW syndrome with life-threatening tachyarrhythmia, RFA may be considered.

3. In presence of hemodynamically significant bradyarrhythmias, as well as symptomatic bradycardia (≤40/min) and pauses ≥3 seconds, permanent PM implantation may be considered.

IX.7.A. SCD in patients with congenital heart defects

Recommendations for SCD prevention

Class I

1. ICD implantation is indicated in patients with congenital heart defects (CHD) who have survived a cardiac arrest (a major SCD risk factor), if the etiology of the disease has been discovered during the examination and other reversible etiological factors have been excluded (B). ICD implantation is indicated in patients who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (B).

2. Invasive hemodynamic assessment and intracardiac EPS are indicated in patients with CHD and spontaneous sustained VT. The recommended treatment approaches are catheter ablation and surgical management of VT. If the above mentioned methods are ineffective, ICD implantation is indicated (C).

Class IIa

1. Invasive intracardiac hemodynamic assessment and intracardiac EPS should be considered in patients with CHD who have major SCD risk factors – history of unexplained syncope and impaired left ventricular contractile function.

2. In absence of determined and potentially correctable cause of a cardiac arrest (a major risk factor) ICD implantation should be considered in patients who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (B).

Class IIb

1. Intracardiac EPS may be considered in patients with CHD and PVC couplets or NSVT for sustained VT risk assessment (C).

Class III

1. Preventive antiarrhythmic therapy is not indicated in patients with asymptomatic CHD and isolated PVCs (C).

IX.7.B. SCD in patients with acquired heart defects

Recommendations for SCD prevention

Class I

1. Clinical assessment and treatment of patients with valvular heart disease and VA should be based on current Guidelines for the diagnosis and treatment of heart defects and the identification of major and minor SCD risk factors.

Class IIa

1. Positive effects of mitral valve repair or replacement on SCD prevention in patients with MPV, severe mitral regurgitation with hemodynamically significant VA (a major risk factor) are not proven.

IX.8. SCD in patients with metabolic and inflammatory diseases

IX.8.A. SCD risk stratification and prevention in patients with myocarditis and infective endocarditis

Myocarditis Recommendations

Class I

1. Treatment in specialized centers capable of monitoring hemodynamics, cardiac catheterization, endomyocardial biopsy and use of mechanical circulatory support devices for cardiopulmonary support, and the possibility of providing specialized types of care for arrhythmias is recommended in patients with life-threatening clinical manifestations of sustained VT and myocarditis probability (C).

2. In patients with acute myocarditis and development of severe bradycardia and/or conduction disorders, accompanied by clinical manifestations, temporary pacemaker placement is indicated (C).

Class IIa

1. AAA should be considered in patients with non-sustained and sustained VT in the acute phase of myocarditis (C).

2. ICD implantation should be considered in patients with sustained hemodynamically significant VT who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year after the acute phase of myocarditis (C).

3. ICD or permanent pacemaker implantation should be considered in patients with inflammatory diseases of the heart after acute condition treatment (C).

4. Wearable external defibrillator should be considered as a transitional therapy before complete recovery or permanent ICD implantation in patients after an inflammatory myocardial disease with persistent severe LV dysfunction and / or electrical instability of the ventricular myocardium (C).

Class IIb

1. ICD implantation may be considered in earlier terms in patients with giant cell myocarditis or sarcoidosis in presence of sustained VT with hemodynamic disturbances or history of cardiac arrest, given the unfavorable prognosis for these conditions, and have good functional status and are expected to survive for > 1year (C).

2. Additional indicators of adverse prognosis and increased risk of SCD in inflammatory myocardial diseases are the signs of myocardial infiltration according to immunohistochemical data and / or pathological focal fibrosis on CMR (cardiac magnetic resonance) after myocarditis (C).

Infective endocarditis

Recommendations

Class I

1. Surgical correction of acute severe aortic regurgitation associated with VT is recommended in absence of contraindications (C).

2. Surgical treatment of acute endocarditis complicated by abscess of aorta or aortic valve, associated with AV block is recommended in absence of contraindications (C).

IX.8. B. SCD risk stratification and prevention in patients with end stage renal disease

Recommendations for SCD prevention

Class I

1. SCD prevention measures in patients with end stage renal disease include major risk factors identification (history of VA, systolic dysfunction, syncope, cardiac arrest) and modification of minor risk factors (hypertension, dyslipidemia, hyperglycemia), and also the risk factors associated with chronic kidney disease and dialysis (treatment of nephrogenic anemia, hyperparathyroidism, vitamin D deficiency, adequate dialysis, avoidance of the dialysis fluid with low potassium and calcium content) (C).

Class IIa

1. For secondary prevention of SCD in patients on hemodialysis or continuous ambulatory peritoneal dialysis, angiotensin II receptor blockers (C) and Class III antiarrhythmic drugs should be considered (C).

2. For primary prevention of SCD in patients on hemodialysis or continuous ambulatory peritoneal dialysis, ACE inhibitors should be considered (B).

3. In patients with CKD and major SCD risk factors (life-threatening arrhythmias and left ventricular systolic dysfunction) ICD implantation is superior to medical therapy. However, in patients on dialysis beneficial effect of ICD implantation on survival rate has not been proven. The decision on ICD

placement should be individual and based on the patient's condition and life expectancy (C). Regular hemodialysis treatment or continuous ambulatory peritoneal dialysis should not be considered as a decisive argument against ICD implantation.

Class IIb

1. For primary prevention of SCD in patients with CAD on hemodialysis or continuous ambulatory peritoneal dialysis, cardioselective β-blockers may be considered (C).

2. For primary prevention of SCD in patients on hemodialysis or continuous ambulatory peritoneal dialysis without signs of coronary arteries involvement, nicorandil may be considered (C).

IX.9. SCD in patients with pericardial diseases

Recommendations for SCD prevention

Class I

1. SCD risk stratification and prevention in patients with pericardial diseases is based on the detection of major and minor SCD risk factors. SCD prevention includes ICD implantation in patients with major SCD risk factors, who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (C).

IX.10. SCD in patients with COPD

Recommendations for SCD prevention

Class I

1. SCD prevention specifics in patients with COPD are based on the detection of major and minor SCD risk factors. It includes ICD implantation in patients with major SCD risk factors, who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (B).

2. When β-blockers are indicated, preference should be given to β-1-selective blockers (A).

Class IIa

1. In patients with CHF, bisoprolol is the drug of choice that does not reduce FEV1 (a major risk factor) and quality of life (B).

2. Patients with stable COPD who are treated with theophylline and long-acting β-2 agonists don't have increased risk of SCD (B).

Class IIb

1. Inhaled corticosteroids reduce the risk of SCD in patients with COPD (B).

2. Elderly patients (over 65 years) with COPD have a lower risk of SCD when treated with long-acting inhaled β-2 agonists than with long-acting inhaled anticholinergic agents (B).

3. Tiotropium inhalation powder does not increase the risk of SCD in patients with COPD (B).

Class III

1. High doses of β-2 agonists should be avoided in patients with unstable angina (A).

2. Use of 14-membered macrolides (erythromycin, clarithromycin) may lead to interval QT prolongation and increased risk of ventricular arrhythmias (a SCD major risk factor) (B).

3. Ipratropium bromide inhalation in patients with COPD is associated with increased risk of SCD (B).

4. Patients with COPD and CHF, who are treated with short-acting inhaled β-2 agonists, have a higher risk of SCD in comparison with those who do not receive these medications (A).

5. While considering possibility of CABG in COPD patients with FEV1 < 60% it should be taken into account that in these patients risk of death in the postoperative period is significantly higher (B).

IX.11. SCD in patients with neuromuscular diseases

Hereditary neuromuscular diseases (myotonic muscular dystrophy, Kearns-Sayre syndrome, Erb's muscular dystrophy, Emery–Dreifuss muscular dystrophy and other myopathies) may predispose to the development of atrial arrhythmias, conduction abnormalities, AV block, monomorphic or polymorphic VT and SCD.

Recommendations for SCD prevention

1. Accurate genetic testing (I, A).
2. Cardiovascular system investigations

A. Registration of the surface ECG (I, C) (once in three months).

B. Echocardiography (I, B) (once in three months):

– Increased attention should be paid to the echocardiographic signs of latent heart failure:

- increase in the left ventricle end-diastolic diameter during the last three months;
- increase in the LV mass during the last three months;
- decrease in the LVEF during the last three months;
- presence of left ventricular dyskinesia;
- the distance from the highest point of the opening of the mitral valve leaflets to the interventricular septum to the LV end-diastolic diameter ratio is >16;
- increase in the left ventricle pre-ejection period to the left ventricle ejection period ratio within the last three months.

If all six of the above mentioned signs are identified in the patient, development of manifest heart failure should be expected within the next six months. It should be noted that during the first two months of the patient's loss of ability to move independently, there is a decrease in the mass of his left ventricle, as 20% decrease of its EED.

C. Holter ECG monitoring (I, B).

3. Performance of biopsy:
   - Myocardial biopsy;
   - Skeletal muscle biopsy (I, A).
4. Adequate treatment, including the administration of the following drug groups (in absence of contraindications and unacceptable side effects):
   - ACE inhibitors (I, A);
   - ARBs (I, B);
   - β-blockers (I, A).

PM placement (IIa, B) should be considered in patients with such hereditary neuromuscular diseases as Duchenne, Becker, Emery-Dreifuss, and Rottauf-Mortier-Beier muscular dystrophy, Mabry syndrome, Werdnig-Hoffmann disease, Davidenkov's myodystrophy, Pompe and von Gierke disease, Kearns-Sayre syndrome, Rossolimo-Curschmann-Steinert-Butten disease in the presence of such SCD risk factor as AV block (including I degree AV block), which is often the main manifestation of cardiac pathology in patients of this group.

ICD implantation should be considered for primary prevention of SCD in patients with LVEF<35% (IIa, B).

IX.12. SCD in patients with channelopathies and early repolarization syndrome

IX.12.A. Long QT syndrome

SCD risk stratification

Table IX.12.1. Risk stratification in patients with congenital LQTS

Risk of cardiac events by the age of 40	QTc at rest	Genotype	Gender
High (>50%)	>500 ms	LQT1 LQT2 LQT3	male/female male/female male
Medium (30–49%)	> or < 500 ms < 500 ms	LQT3 LQT3 LQT2	female male female
Low (<30%)	< 500 ms	LQT2 LQT3	male male/female

Recommendations for genetic testing

Class I

1. Genetic testing is indicated for congenital long QT syndrome diagnosis in all children and adolescents with QT prolongation at rest (QTc> 460 ms in females and QTc> 440 ms in males) with major SCD risk factors (history of unexplained syncope, cardiac arrest history, family history of SCD), and in children with epilepsy without specific treatment effect (B).

Class IIa

1. Genetic testing should be considered in all patients with high probability of LQTS based on the history and ECG phenotype of disease (based on the 12 lead ECG at rest and/or during exercise stress test or catecholamine infusion) (C).

2. Genetic testing should be considered in all asymptomatic patients with QTc> 480 ms (children) or QTc> 500 ms (adults) on 12-lead ECG in absence of diseases or conditions that may cause QT interval prolongation (such as electrolyte imbalance, myocardial hypertrophy, bundle branch block, etc.) (C).

3. If genetic testing of a proband led to identification of a mutation responsible for LQTS, the first and second degree relatives, regardless of the clinical phenotype, should have selective genetic testing performed (C).

Class IIb

1. If QT interval prolongation is caused by specific drugs, genetic testing provides an opportunity to identify carrier status mutations responsible for congenital forms of LQTS (B).

2. If it is impossible to perform genetic testing for all the known mutations, a selective testing of genes responsible for LQT1-3 (KCNQ1, KCNH2, SCN5A) may be considered (C).

3. 12-lead ECG at rest may be considered for the first and second degree relatives of patients with acquired LQTS (C).

Recommendations for SCD prevention

Class I

1. Lifestyle modification is recommended in patients with a diagnosis of LQTS (confirmed clinically and/or by molecular genetic testing) (B).

2. In patients with a diagnosis of LQTS (confirmed clinically and/or by molecular genetic testing) avoidance of QT-prolonging drugs is recommended (B).

3. ICD implantation is recommended for primary SCD prevention in patients diagnosed with long QT syndrome (LQT3) confirmed by molecular genetic testing (B).

4. ICD implantation is recommended for secondary SCD prevention in patients diagnosed with long QT syndrome (LQT1, LQT2, LQT5 and LQT6) confirmed by molecular genetic testing, particularly with history of cardiac arrest (a major SCD risk factor) (B).

Class IIa

1. In patients with a diagnosis of LQTS (LQT1 and LQT5) confirmed by molecular genetic testing, β-blockers should be considered (B).
2. In patients with a diagnosis of LQTS (LQT2 and LQT6) confirmed by molecular genetic testing, potassium supplements should be considered (B).
3. In patients with a diagnosis of LQTS (LQT3) confirmed by molecular genetic testing, sodium channel blockers should be considered (B).
4. ICD implantation with the use of β-blockers should be considered in patients with a clinical diagnosis of long QT syndrome and SCD major risk factors (syncope and/or ventricular arrhythmias) (B).
5. Left cardiac sympathetic denervation and β-blocker therapy should be considered in patients with a clinical diagnosis of long QT syndrome and SCD major risk factors (cardiac arrest history, torsade de pointes) (B).

Class IIb

1. Calcium channel blockers and Class IB antiarrhythmic agents may be considered in patients with a diagnosis of LQTS (LQT2 and LQT6) confirmed by molecular genetic testing (B).
2. Preventive use of β-blockers may be considered in carriers of a causative LQTS mutation (B).

Class III

1. In patients with a diagnosis of LQTS (LQT3) confirmed by molecular genetic testing, β-blockers and nicorandil are contraindicated (C).

IX.12. B. Short QT syndrome

Table IX.12.B. SQTS diagnostic criteria

Diagnostic criteria	Score
QTc interval < 370ms	1
QTc interval < 350ms	2
QTc interval < 330ms	3
J-point – T-peak interval < 120ms	1
Clinical history	
History of sudden cardiac arrest	2
Documented VT or VF	2
Unexplained syncope	1
Atrial fibrillation	1
Family history	
First- or second-degree relative with high-probability SQTS	2
First- or second-degree relative with autopsy-negative sudden cardiac death	1
Sudden infant death syndrome	1
Genotype http://www.sciencedirect.com/science/article/pii/S0735109710047212–tblfn5	
Previously described mutation positive	2
Mutation of undetermined significance in KCNH2, KCNQ1, KCNJ2	1
SQTS probability assessment	
High-probability SQTS	> 4 points
Intermediate-probability SQTS	3
Low-probability SQTS	1 – 2

Recommendations for genetic testing

Class IIa

1. Genetic testing should be considered to confirm the diagnosis in patients with low or intermediate probability of SQTS (C).

Class IIb

1. If a pathogenic mutation has been detected in a proband, genetic testing of the patient's relatives may be considered (C).

Recommendations for SCD prevention

Class I

1. ICD implantation is recommended for secondary prevention of SCD in patients with the diagnosis of short QT syndrome and major SCD risk factors (history of cardiac arrest, syncope, hemodynamically significant sustained ventricular arrhythmias induced by intracardiac EPS) (C).

Class IIa

1. ICD implantation should be considered in patients with high probability of SQTS (B).

2. Quinidine should be considered to effectively reduce risk of SCD in patients with short QT syndrome, if ICD implantation is impossible (C).

IX.12.C. Brugada syndrome

Recommendations for genetic testing

Class I

1. Genetic testing for congenital Brugada syndrome is recommended in all children and adolescents with specific ECG changes, who have major SCD risk factors, including syncope, history of cardiac arrest, ventricular arrhythmias, and family history of SCD (B).

Class IIb

1. Identification of the mutation type in SCN5A gene may provide additional information about risk of SCD (B).

2. If a pathogenic mutation has been detected in a proband, genetic testing of the patient's relatives may be considered (C).

Class III

1. Genetic testing is not indicated in asymptomatic patients with type 2 or 3 Brugada-like ECG changes.

Recommendations for SCD prevention

Class I

1. ICD implantation is recommended in patients with a diagnosis of Brugada syndrome confirmed clinically and/or based on molecular genetic analysis who have a history of cardiac arrest (a major SCD risk factor) (C).

2. In patients with a diagnosis of Brugada syndrome and implanted ICD with frequent appropriate shocks, guinidine and/or RFA is indicated (B).

Class IIa

1. ICD implantation should be considered in patients with Brugada syndrome, spontaneous ST segment elevation in leads V1–V3, who have history of syncope (a major SCD risk factor) and verified causative mutations in SCN5A gene (C).

2. Pharmacological stress tests with sodium channel blockers (flecainide, novocainamide) should be considered in patients with suspicion of Brugada syndrome without ST segment elevation in the right precordial leads (C).

3. ICD implantation should be considered in patients with Brugada syndrome and good functional status who are expected to survive for > 1year and who survived verified VT that did not cause cardiac arrest (C).

Class IIb

1. Intracardiac EPS may be considered for SCD risk stratification in patients with Brugada syndrome with spontaneous ST elevation, without SCN5A mutation and any clinical signs or symptoms (C).

Class III

1. Class IC (e.g. flecainide and propafenone) and Class IA (e.g. procainamide, disopyramide) antiarrhythmic agents are contraindicated in patients with Brugada syndrome.

IX.12. D. Catecholaminergic polymorphic ventricular tachycardia

Recommendations for genetic testing

Class I

1. Genetic screening for CPVT is recommended in all children and adolescents with the following major SCD risk factors: polymorphic VT induced by physical or severe emotional stress, syncope, cardiac arrest, family history of SCD (B).

2. Genetic testing is recommended in patients with high probability of CPVT diagnosis based on clinical history, family history, ECG phenotype of the disease, exercise or pharmacologic (catecholamine) stress test results (C).

Class IIb

1. If a pathogenic mutation in proband is identified, genetic testing of the first- and second-degree relatives may be considered.

Recommendations for SCD prevention

Class I

1. Lifestyle changes (avoidance of any strenuous exercise, competitive sports, emotional distress) are recommended in patients diagnosed with CPVT (confirmed clinically and/or by molecular genetic testing) (B).

2. Beta-blockers are recommended in all patients with the clinical diagnose of CPVT (C).

3. ICD implantation and beta-blocker administration are indicated for secondary prevention of SCD in patients with CPVT (confirmed clinically and/or by molecular genetic testing) who survived cardiac arrest (a major SCD risk factor) (C).

Class IIa

1. ICD implantation should be considered for secondary prevention of SCD in patients with CPVT (confirmed clinically and/or by molecular genetic testing) who survived cardiac arrest (a major SCD risk factor) while on beta-blocker therapy (C).

2. Left cardiac sympathetic denervation should be considered in patients with a clinical diagnosis of CPVT and frequent ICD shocks while on beta-blocker therapy (B).

Class IIb

1. Beta-blockers may be considered in carriers of a pathogenic CPVT mutation (confirmed during childhood or adulthood) without clinical manifestations of CPVT (C).

IX.12. E. SCD in patients with early repolarization

Recommendations for SCD prevention

Class I

1. ICD implantation is recommended in patients with ER ECG pattern who have survived cardiac arrest or sustained VT (major SCD risk factors) (B).

2. In patients with isolated ER ECG pattern dynamic observation without treatment administration is recommended (B).

Class IIa

1. Presence of J waves in the inferior leads (II, III, avF) can be a major risk factor for VF (or even SCD), and a factor that increases susceptibility to fatal arrhythmias during myocardial ischemia (B).

2. The treatment of choice in case of VA (a major SCD risk factor) or an electrical storm in patients with ER ECG pattern in inferior leads is increase of the heart rate by means of temporary cardiac pacing or isoproterenol infusion (B).

3. Prolonged quinidine administration should be considered for SCD prevention if ER ECG pattern in the inferior leads is present (B).

IX.13. SCD in patients with obstructive sleep apnea syndrome

Table IX.13.1. SCD risk stratification in patients with OSAHS

Note:*AHI – Apnea-hypopnea index, OSAHS – obstructive sleep apnea/hypopnea syndrome, SCD – sudden cardiac death.*

1. OSAHS diagnosis is verified	
Yes	No
See p. 2	SCD risk stratification and prevention based on general principles
2. The Apnea–hypopnea index (AHI) >15	
Yes	No
CPAP therapy or non-invasive ventilation choice	Weight reduction, correction of ENT disorders, elimination of OSAHS risk factors
Has 24h Holter monitoring revealed any rhythm and conduction disturbances during the night or daytime sleep?	
Yes	No
See recommendations for SCD prevention in patients with OSAHS	SCD risk stratification and prevention should be based on the detection of major and minor SCD risk factors and general principles of SCD prevention

IX.14. SCD in patients with ventricular arrhythmias of structurally normal heart

SCD risk stratification in patients with VA of structurally normal heart

Table IX.14.1. SCD risk stratification and prevention algorithm (clinical data)
Table IX.14.2. SCD risk stratification and prevention algorithm (clinical history)

Table IX.14.3. SCD risk stratification and prevention algorithm (ECG and Holter monitoring data)

1. History of SCD episode	
Yes	No
Consider ICD implantation	See p. 2
1.1. SCD episode due to reversible causes	
Yes	No \ unknown
Elimination of the underlying causes of cardiac arrest, observation	Consider ICD implantation
1.2. The cause of cardiac arrest was a monomorphic VT (according to ECG data during resuscitation)	
Yes	No \ unknown
Consider RFA	See p. 2
2. VT with dizziness, syncope	
Yes	No
Consider EPS, RFA	See p. 3
3. VA with shortness of breath during exercise	
Yes	No
Consider antiarrhythmic agents, EPS, RFA	See p. 4
4. VT induction or increase in PVCs rate during physical exercise or in the recovery period	
Yes	No
Antiarrhythmic agents (beta-blockers, sotalol). Consider EPS, RFA	See p. 5

4.1. Suppression of VA during exercise	
Yes	No
Class 1C antiarrhythmic drugs / observation / consider EPS, RFA	See p. 5
5. Clinical effectiveness of antiarrhythmic treatment	
Yes	No
Prospective observation	See the recommendations

Table IX.14.4. SCD risk stratification and prevention algorithm (instrumental investigations data)

1. History of syncope of unknown origin	
Yes	No
Multi day Holter monitoring	
Remote ECG monitoring	
Tilt table test	
Long-tern insertable cardiac monitor (REVEAL)	
Consider EPS	See p. 2
2. Family history of syncope	
Yes	No
Genetic counseling, cardiac screening of family members, including children	See p. 3
3. VA in relatives	
Yes	No
Genetic counseling, cardiac screening of first-degree, including children	See p. 4
4. Proarrhythmic effects of antiarrhythmic drugs	
Yes	No
Antiarrhythmic therapy adjustment, consider EPS. Genetic counseling	See the recommendations

Table IX.14.5. SCD risk stratification and prevention algorithm (intracardiac EPS data)

1. Paroxysms of sustained VT	
Yes	No
Consider EPS, RFA (if not effective – ICD)	See p. 4
2. VT cycle length <360ms	
Yes	No
Consider EPS for VF induction (except fascicular tachycardia). If VF is not induced – see p. 1	See p. 1
3. The ventricular ectopic QRS interval duration is >160ms	
Yes	No
Consider EPS for polymorphic VT / VF induction (except VT from right and non-coronary aortic sinus of Valsalva and LVOT).	See p. 1
4. Number of PVCs is > 20.000 per 24 hours (according to several Holter monitoring data)	
Yes	No
Consider antiarrhythmic agents, EPS, RFA	See p. 5
5. Detection of ventricular late potentials	
Yes	No
Consider ECG stress tests, EPS	See p. 6
6. A transient change in QTc duration beyond the standard values according to Holter monitoring data	
Yes	No
Antiarrhythmic therapy adjustment, genetic counseling	See p. 7
7. QT dispersion is >100 ms	

Yes	No
Consider ECG stress tests, EPS	See p. 8
8. Presence of epsilon waves, including transient ones, according to Holter monitoring	
Yes	No
Genetic counseling for ARVD onset	See p. 9
9. Presence of J waves, including transient ones, according to Holter monitoring	
Yes	No
Genetic counseling	See p. 10
10. Presence of notches of the R-wave in the ventricular ectopic QRS interval	
Yes	No
Consider ECG stress tests, EPS	See p. 11
11. Microvolt T-wave alternans according to Holter monitoring	
Yes	No
Consider ECG stress tests, EPS	See p. 12
12. Transient AV or intraventricular conduction abnormalities	
Yes	No
Consider ECG stress tests, EPS, immunological test for latent myocarditis	See p. 13
13. Number of polytopic PVCs is > 10.000	
Yes	No
Consider ECG stress tests, EPS, immunological test for latent myocarditis	See p. 14
14. Combination of atrial and ventricular arrhythmias	
Yes	No
Immunological test for latent myocarditis	Prospective observation

SCD prevention in patients with VA with structurally normal heart

1. MRI: fibrotic zones / thinning in the ventricular myocardium	
Yes	No
Differential diagnosis between ARVD onset and latent myocarditis	See p. 2
2. MRI: intramyocardial fat infiltration	
Yes	No
Analysis of ARVD diagnostic criteria	See p. 3
3. MRI: epicardial fat infiltration	
Yes	No
Immunological test for latent myocarditis	See p. 4
4. MRI: intramyocardial contrast accumulation in the early phase	
Yes	No
Immunological test for latent myocarditis	See p. 5
5. MRI: intramyocardial contrast accumulation in the delayed phase	
Yes	No
Differential diagnosis between ARVD onset and latent myocarditis	See p. 6
6. MRI: RVOT/LVOT enlargement	
Yes	No
Differential diagnosis between ARVD onset and latent myocarditis	See p. 7
7. Scintigraphy: areas of mosaic hypoperfusion of in the LV myocardium	
Yes	No
Immunological test for latent myocarditis	Prospective observation

Class I

1. Pacing-induced VF using standardized ventricular stimulation protocol	
Yes	No
Consider ICD implantation	See p. 2
2. Pacing-induced polymorphic VT using standardized ventricular stimulation protocol	
Yes	No
Consider ICD implantation	See p. 3
3. Pacing-induced monomorphic VT with VT cycle length <360ms using standardized ventricular stimulation protocol	
Yes	No
Consider RFA / ICD implantation	Consider RFA / antiarrhythmic therapy

1. RFA is indicated in patients without structural heart disease with paroxysmal or continuous recurrent ventricular tachycardia originating from RVOT/LVOT and ineffectiveness of the antiarrhythmic agents (B).

2. RFA is indicated in all patients with fascicular left ventricular tachycardia(A).

3. Preventive antiarrhythmic therapy is indicated in patients with structurally normal heart and daily index of arrhythmia load of more than 20% (C).

4. If the «minimal changes» in RV/LV that fit arrhythmogenic variant of latent myocarditis criteria are identified as a cause of VA, etiotropic and pathogenetic treatment of the underlying disease, regardless of its duration, is indicated (C).

5. If the «minimal changes» in RV (rarely in LV) that fit arrhythmogenic variant of latent myocarditis criteria are identified as a cause of VA, contrast enhanced MRI of the myocardium and diagnostic criteria analysis are indicated for ARVD onset exclusion (C).

6. ICD implantation is indicated in patients with ventricular arrhythmias without structural heart disease who have survived an SCD episode without reversible causes (B).

7. ICD implantation is indicated in patients without structural heart disease with sustained VA which requires medical or electrical cardioversion when preventive use of antiarrhythmic agents is ineffective and RFA has failed (C).

Class IIa

1. RFA should be considered in patients without structural heart disease with sustained or continuous recurrent ventricular tachycardia as alternative to successful antiarrhythmic treatment upon patient's request (C).

2. Preventive use of antiarrhythmic agents (β-blockers or sotalol) should be considered in patients without structural heart disease and with daily index of arrhythmia load of more than 20% provoked by physical exercise and registered on Holter monitoring mainly during daytime (C).

3. Preventive use of Class IC antiarrhythmic agents should be considered in patients without structural heart disease and with daily index of arrhythmia load of more than 20% provoked by sinus bradycardia and suppressed by physical exercise and registered on Holter monitoring mainly during the night (C).

4. Intracardiac EPS should be considered in patients with VA without structural heart disease and with history of palpitations, episodes of dizziness and syncope, if VT is expected to be the cause (B).

Class IIb

1. RFA may be considered in patients without structural heart disease with sustained unifocal VA originated from RVOT/LVOT with daily index of arrhythmia load of more than 20% as an alternative to successful antiarrhythmic treatment upon patient's request (C).

2. Adjunctive preventive therapy with omega-3 polyunsaturated fatty acids may be considered in patients with VA without structural heart disease (C).

Class III

1. Preventive use of antiarrhythmic agents is not indicated in patients without structural heart disease and with daily index of arrhythmia load less than 20% (C).

2. Preventive use of antiarrhythmic agents is not indicated in patients without structural heart disease and with VA manifestation (clinical history less than 3 months) (C).

3. RFA is not indicated in patients without structural heart disease with sustained unifocal VA originated from RVOT/LVOT with daily index of arrhythmia load less than 20% (C).

4. RFA is not indicated in patients without structural heart disease and with VA manifestation onset (clinical history less than 6 months) (C).

5. RFA is not indicated in patients with «minimal changes» in RV/ LV that fit the arrhythmogenic variant of latent myocarditis criteria before the pathogenetic and etiologic treatment of the underlying disease, in case of arrhythmia history is less than a year, with multifocal VA (C).

SCD prevention in patients with electrolyte disturbances and structurally normal heart

Class I

1. Administration of potassium and magnesium supplements is indicated for treatment and prevention of ventricular arrhythmias in patients with structurally normal heart treated with thiazide diuretics (B).

2. Administration of potassium and magnesium supplements is indicated for treatment and prevention of ventricular arrhythmias in patients with structurally normal heart after cardiopulmonary bypass surgeries with mandatory correction of blood pH levels (B).

Class IIa

1. In patients with confirmed life-threatening VA and structurally normal heart maintenance of serum potassium level in the range of 4.5 – 5.5 mmol/L should be considered (C).

2. Administration of potassium and magnesium supplements should be considered for treatment and prevention of ventricular arrhythmias in patients with structurally normal heart with cardiac glycoside overdose (B).

Class IIb

1. Administration of potassium and magnesium supplements may be considered for treatment and prevention of ventricular arrhythmias in patients with structurally normal heart and acute or chronic alcohol or narcotic intoxication, anorexia (C).

Class III

1. Administration of potassium and magnesium supplements is not indicated in patients with acute kidney injury and chronic kidney disease (B).

IX.17. SCD risk stratification and prevention in specific population groups

IX.17.A SCD in athletes

Recommendations for SCD prevention

Class I

1. Careful history taking is recommended in athletes in order to identify the latent pathology of CVS, rhythm disturbances, syncope or family history of SCD (C).

2. If ECG changes that suggest the presence of structural heart disease are detected, Echocardiography and / or cardiac MRI are recommended. (C).

Class IIa

1. In young athletes a thorough physical examination and registration of the ECG at rest should be

considered before the competitions for screening purposes (C).

2. In middle-aged people who participate in high-intensity strength training, a physical examination with assessment of complaints and clinical history, ECG registration and determination of SCORE risk should be considered for screening purposes (C).

3. Personnel, participating in the organization of sports competitions should be taught the skills of cardiopulmonary resuscitation and the correct use of automatic external defibrillators (C).

IX.17.B. SCD risk stratification and prevention in elderly patients

Recommendations for SCD stratification and prevention

Class I

1. Management of elderly patients with VA, as a rule, should not differ from younger patients. It implies that the SCD risk stratification and prevention in elderly patients is based on the detection of major and minor risk factors. SCD prevention includes ICD or pacemaker implantation in patients who receive continuous optimal medical therapy, have good functional status and who are expected to survive for > 1year (C).

2. All the elderly patients should be advised to stop smoking and alcohol abuse(B).

3. Beta-blockers (A). Nebivolol has the largest evidence base among patients older>70 years (B).

4. ACE inhibitors (ramipril, enalapril, perindopril, trandolapril) in elderly patients with LV systolic dysfunction effectively prevent SCD (B).

5. ACE inhibitors (perindopril in elderly people under 70 and ramipril – with no age limit) in patients with normal LVEF. The possibilities of other ACE inhibitors in this category of patients have not been studied yet (B).

6. ARBs may be used as an alternative to ACE inhibitors for prevention of SCD in elderly patients with ACE inhibitors intolerance.

7. With proper selection of patients (initially normal levels of potassium, blood creatinine), the addition of spironolactone / eplerenone to ACE inhibitors and β-blockers effectively reduces the risk of SCD in elderly patients with NYHA Class III-IV with systolic LV myocardial dysfunction. Moreover, eplerenone significantly improves prognosis also in patients with NYHA Class II (A).

8. The use of acetylsalicylic acid in doses of 75–100 mg is indicated in all patients with CAD, regardless of age or in patients with a high cardiovascular risk younger than 70 (A).

9. Statins are indicated in all patients with CAD or high cardiovascular risk regardless of age (B).

10. In absence of contraindications for β-blockers and high risk of SCD, amiodarone administration is indicated in addition to β-blocker to prevent arrhythmic death in patients with ventricular arrhythmias (B).

11. Dosage and choice of antiarrhythmic agents should be adjusted to reflect pharmacokinetics changes in elderly patients (C).

Class IIa

1. Administration of omega-3-PUFAs is safe. However, their effectiveness in the prevention of SCD in patients older than 70 requires additional confirmation in clinical studies (C).

2. It is possible to use ARBs as starting agents for the RAAS blockade (without prior administration of ACE inhibitors) (C).

Class III

1. ICD implantation in patients who are expected to survive for < 1year due to severity of the primary disease or comorbidities is not recommended (C).

2. Class I antiarrhythmic agents should not be administered in elderly patients with structural heart disease (A).

3. ARBs should not be combined with ACE inhibitors (B).

НАЦИОНАЛЬНЫЕ РЕКОМЕНДАЦИИ ПО ОПРЕДЕЛЕНИЮ РИСКА И ПРОФИЛАКТИКЕ ВНЕЗАПНОЙ СЕРДЕЧНОЙ СМЕРТИ (2-издание) – 2018. КАРМАННЫЙ ВАРИАНТ

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Смертность от сердечно-сосудистых заболеваний в Российской Федерации является одной из наиболее высоких в мире и составляет 614 на 100 000 жителей в год. Основные причины смерти от сердечно-сосудистых заболеваний – прогрессирование хронической сердечной недостаточности (50% всех летальных исходов) и внезапная сердечная смерть (50%). Число внезапных сердечных смертей в 2016 г. составило не менее 300 000 человек. В представленном сокращенном варианте Национальных Рекомендаций на английском языке использованы принципы алгоритмирования принятия решений при различных клинических ситуациях.

Ключевые слова: *болезни системы кровообращения, внезапная сердечная смерть, риск, лечение, профилактика.*

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Сурмач, Марина Юрьевна.

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Учебно-методическое пособие подготовлено в соответствии с образовательным стандартом высшего образования специальности 1-79 01 01 «Лечебное дело», утвержденным и введенным в действие постановлением Министерства образования Республики Беларусь от 30.08.2013 № 88, типовым учебным планом специальности 1-79 01 01 «Лечебное дело» (регистрационный № L 79-1-001/тип.), утвержденным первым заместителем Министра образования Республики Беларусь 30.05.2013, типовой программой «Типовая учебная программа по учебной дисциплине «Общественное здоровье и здравоохранение» для специальностей 1-79 01 01 «Лечебное дело», 1-79 01 02 «Педиатрия». Содержит материал для подготовки и использования во время практических занятий по предмету «Общественное здоровье и здравоохранение».