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Formación online en actualizaciones en Cardiología

GUÍAS ESC ARRITMIAS VENTRICULARES Y PREVENCIÓN DE MUERTE SÚBITA 2022

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ESC GUIDELINES

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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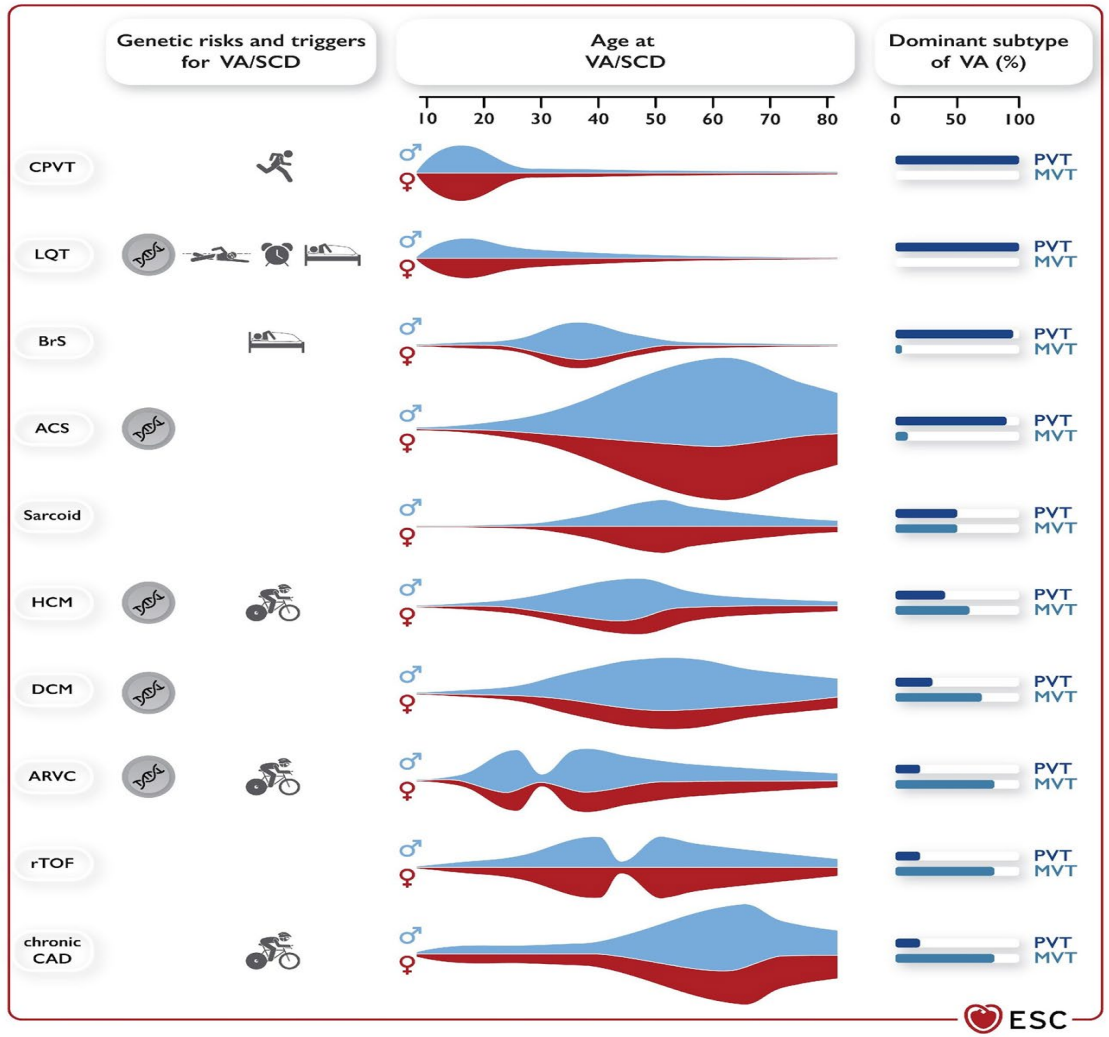

Zeppenfeld K et al. 2022 ESC Guidelines for the managememnt of patients with ventricular arrhytmias and the prevention of sudden death. European Heart Journal (2022) 00,1-130 https://doi.org/10.1093/eurheartj/ehac262

New sections and concepts

New sections and concepts
Provocative diagnostic tests
Genetic testing
Diagnostic evaluation at first presentation with VA in patients without known cardiac disease
Management of patients with electrical storm
Special aspects of device therapy

PREVENCIÓN: Acceso a desfibriladores y educación en SVB desde la infancia

Recommendations	Class[a]	Level[b]
It is recommended that public access defibrillation be available at sites where cardiac arrest is more likely to occur.[c,90–92]	I	B
Prompt CPR by bystanders is recommended at OHCA.[93–95]	I	B
It is recommended to promote community training in basic life support to increase bystander CPR rate and AED use.[93,97,104]	I	B
Mobile phone-based alerting of basic life support-trained bystander volunteers to assist nearby OHCA victims should be considered.[101–103,105]	IIa	B

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Diagnostic test	Indication	Protocols Dose/infusion rate/duration	Positive test	Contraindications	Criteria to stop test, counselling and management	Observation times	Location
Ajmaline	Family history of BrS or SADS. Resuscitated CA without SHD.	1 mg/kg over 5–10 min (maximum dose 100 mg) or 1 mg/kg at 10 mg/min. Record in standard and high precordial leads over 30 min.	BrS type 1 ECG.	Type I BrS ECG, HF. Precaution if evidence of conduction disease (consider temporary pacing wire).	VT/VF, Type 1 BrS ECG, PVCs, QRS widening >150%. If VT/VF, administer iv isoprenaline, iv sodium bicarbonate.	30 min if negative test; 4 h if positive test.	Cath lab or outpatient testing location with full resuscitation equipment.
Flecainide	Same as ajmaline.	2 mg/kg over 10 min (maximum dose 150 mg). Record in standard and high precordial leads over 30 min.	Same as ajmaline.	Same as ajmaline.	Same as ajmaline.	4 h if negative test; 24 h if positive test.	Same as ajmaline.
Epinephrine	CPVT and resuscitated CA with or without SHD when exercise test not feasible. Family history of SADS.	Rest 10 min. Start at 0.025 µg/ kg/ min for 10 min increase sequentially to 0.05, 0.1 and 0.2 µg/ kg/min in 5 min steps.	≥3 beats of PVT or bidirectional VT.	QTc prolongation ≥480 ms.	Systolic blood pressure ≥ 200 mmHg, non-sustained VT or PVT, >10 PVCs /min, T-wave alternans, or patient intolerance. If symptoms persist after discontinuation, iv metoprolol 2.5–5 mg over 1 min.	30 min.	Same as ajmaline.
Acetylcholine	Suspicion of coronary vasospasm.	Intracoronary injection: RCA: 20 and 50 µg. LCA: 20, 50, and 100 µg over 20 s. >3-min intervals between injections. Maximal dose of 50 µg in the RCA and 100 µg in the LCA.	Coronary artery spasm visualized during procedure.	Left main stenosis >50%, 3-vessel disease, 2-vessel disease with total occlusion, NYHA III/IV HF, renal failure, severe bronchial asthma.	Temporary wire for back-up pacing. Risk of cardiogenic shock.	Normal post-procedure observational time.	Cath lab.
Ergonovine	Same as acetylcholine.	Intracoronary stepwise injection: RCA (20–60 mg) LCA (20–60 mg) over a period of 2–5 min.	Same as acetylcholine	Left main stenosis >50%, 3-vessel disease, 2-vessel disease with total occlusion, NYHA III/V HF, renal failure.	Temporary wire should be placed for back-up pacing. Risk of cardiogenic shock.	Same as acetylcholine.	Cath lab.
Adenosine	Exclude latent pre-excitation.	6, 12, 18 mg boluses up to maximum dose 24 mg or until AV block or pre-excitation occurs.	Identification of accessory pathway.	Asthma, sinus node disease, allergy to adenosine.	Side effects: Bronchospasm, bradycardia, asystole, AF, seizure. Antagonist: theophylline.	5 min.	Same as ajmaline.

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Recomendaciones para el E. Genético

Recommendations	Class[a]	Level[b]
Genetic testing is recommended when a condition is diagnosed in a living or deceased individual with a likely genetic basis and a risk of VA and SCD.[56,183]	I	B
When a putative causative variant is first identified, evaluation for pathogenicity is recommended using an internationally accepted framework.[176]	I	C
When a Class IV or Class V variant has been identified in a living or deceased individual with a condition that carries a risk of VA and SCD, genetic testing of first-degree and symptomatic relatives and obligate carriers is recommended.	I	C
It is recommended that genetic testing and counselling on its potential consequences should be undertaken by an expert multidisciplinary team.[179]	I	C
It is recommended that Class III (variants of uncertain significance) and Class IV variants should be evaluated for segregation in families where possible, and the variant re-evaluated periodically.	I	C
It is not recommended to undertake genetic testing in index patients with insufficient evidence of a genetic disease.	III	C

New sections and concepts

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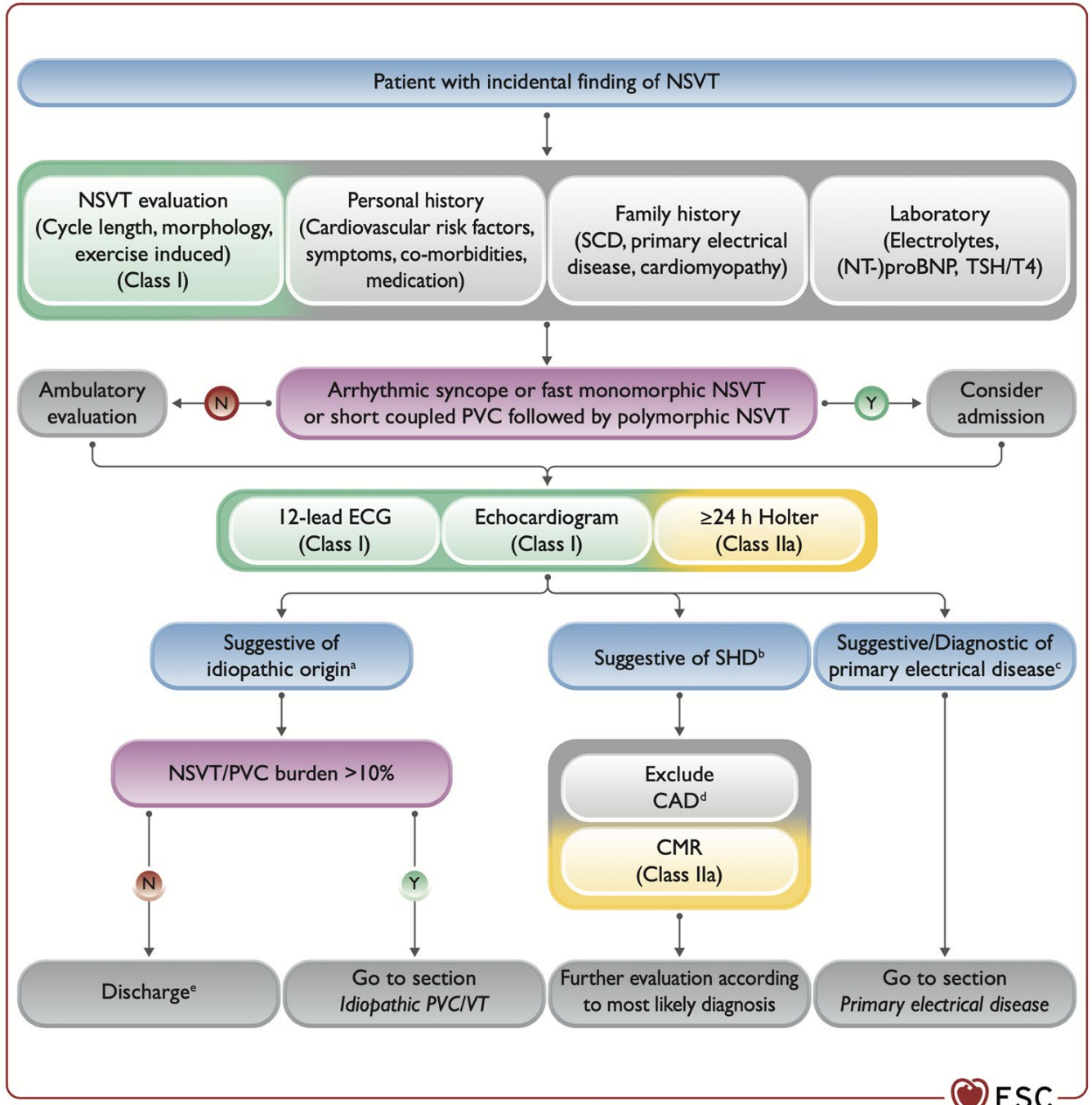
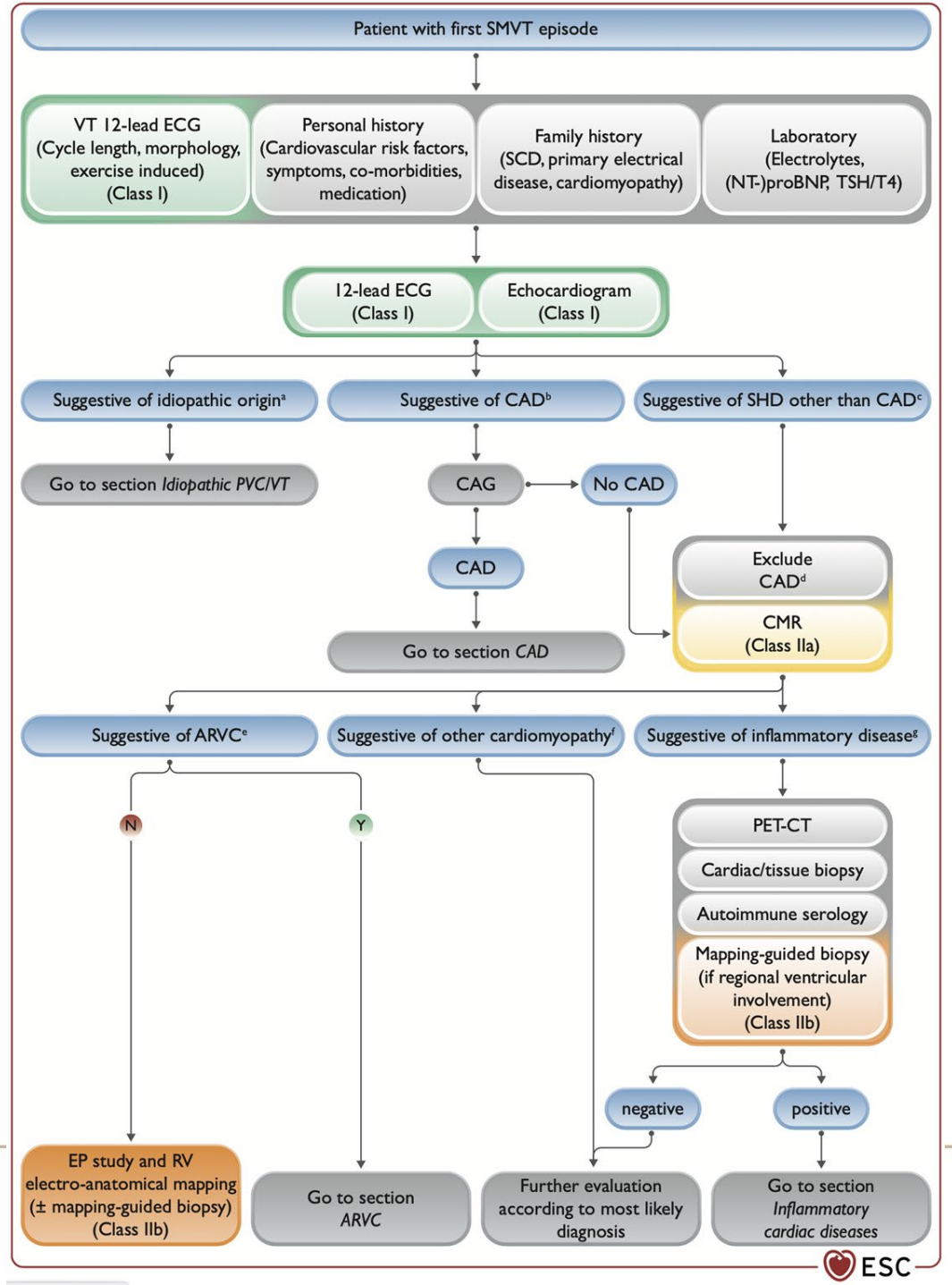
TVNS INCIDENTAL

Recommendations	Class[a]	Level[b]
In patients with newly documented VA (frequent PVCs, NSVT, SMVT), a baseline 12-lead ECG, recording of the VA on 12-lead ECG, whenever possible, and an echocardiogram are recommended as first-line evaluation.	I	C
In patients with newly documented VA (frequent PVCs, NSVT, SMVT) and suspicion of SHD other than CAD after initial evaluation, a CMR should be considered.[194,195]	IIa	B
In patients with an incidental finding of a NSVT, a ≥24 h Holter ECG should be considered.	IIa	C



TVMS

Recommendations	Class[a]	Level[b]
In patients presenting with a first SMVT episode, electrophysiological study, electroanatomical mapping, and mapping-guided biopsies may be considered for aetiological evaluation.[197–199,202]	IIb	C



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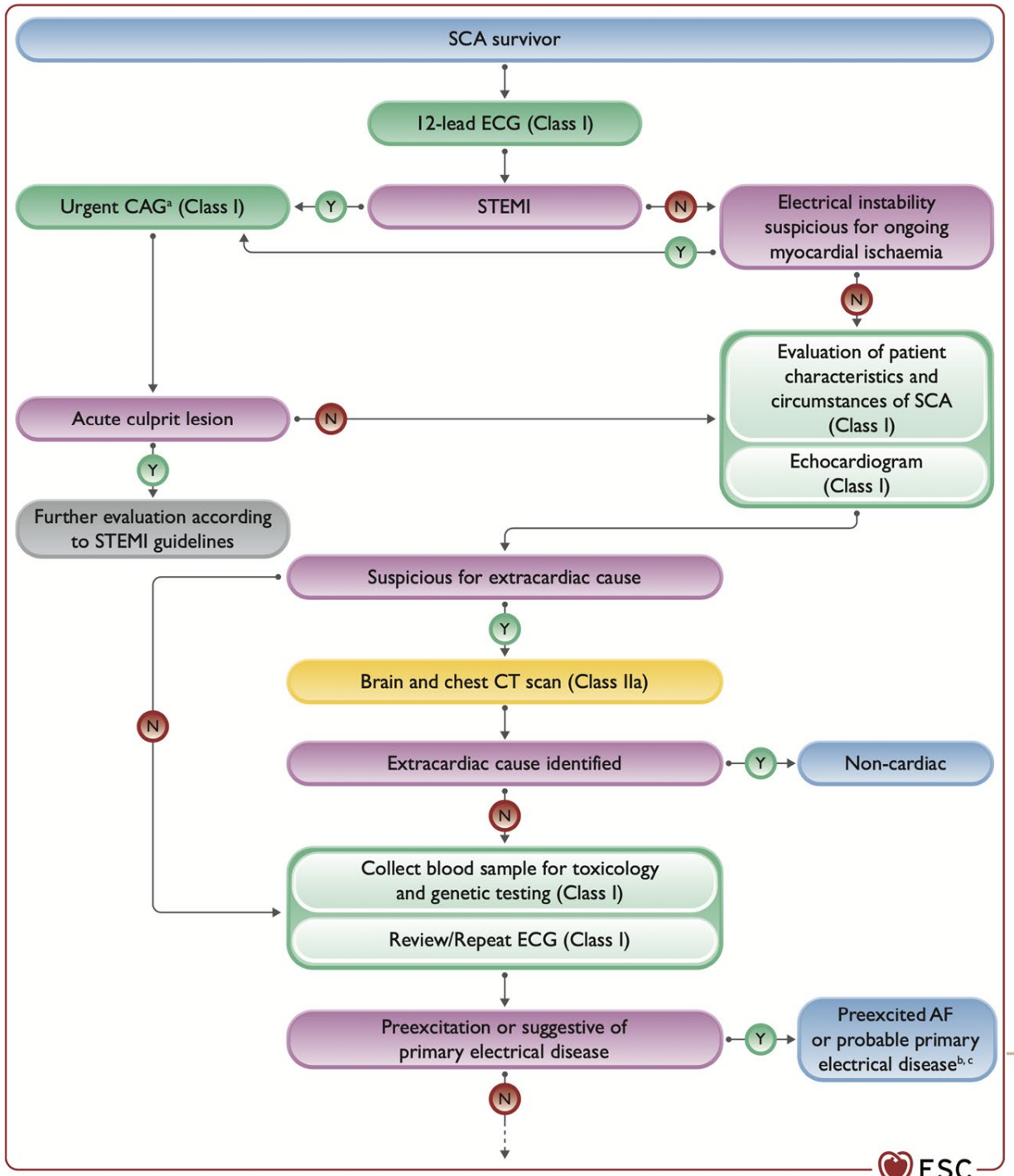

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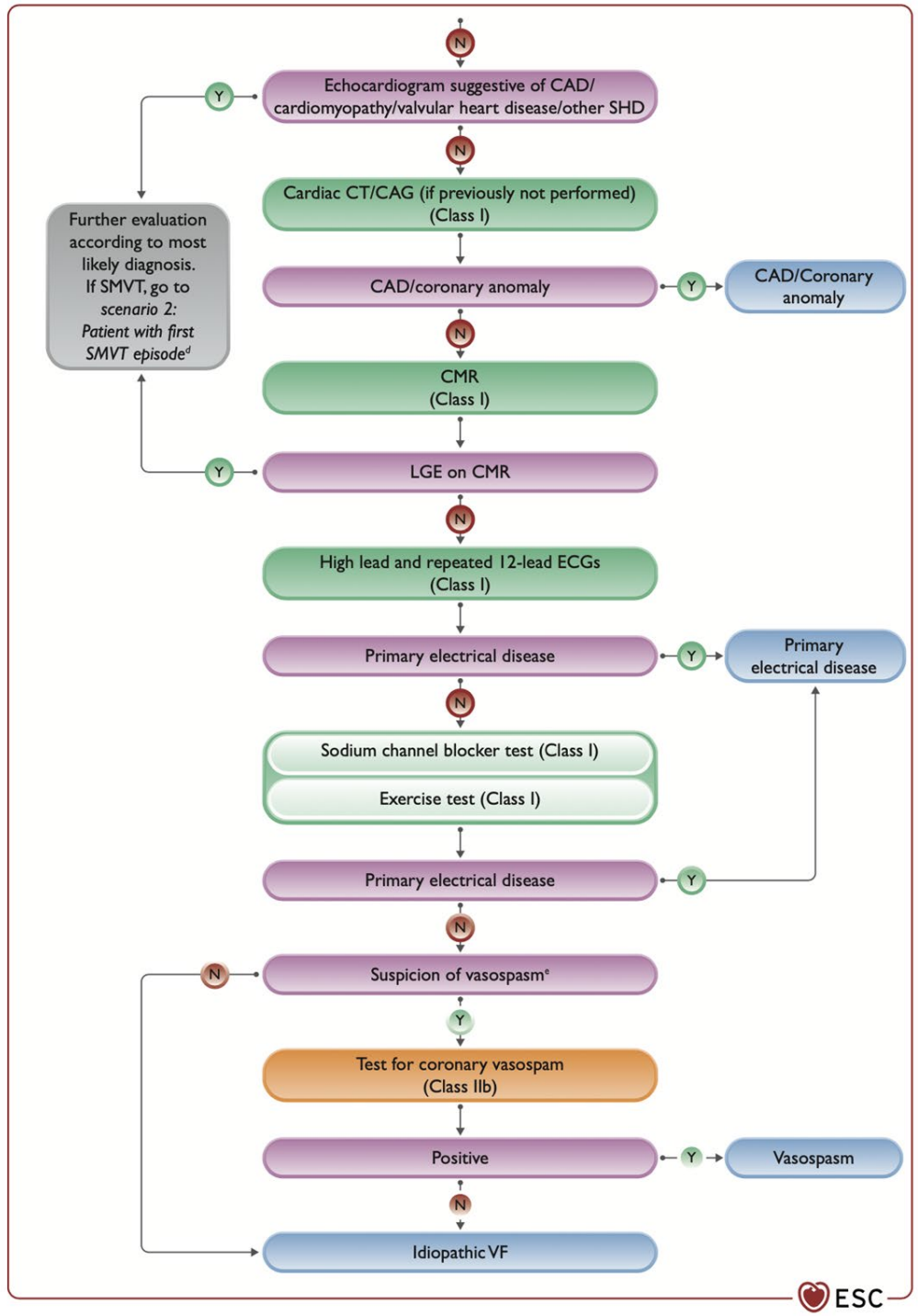

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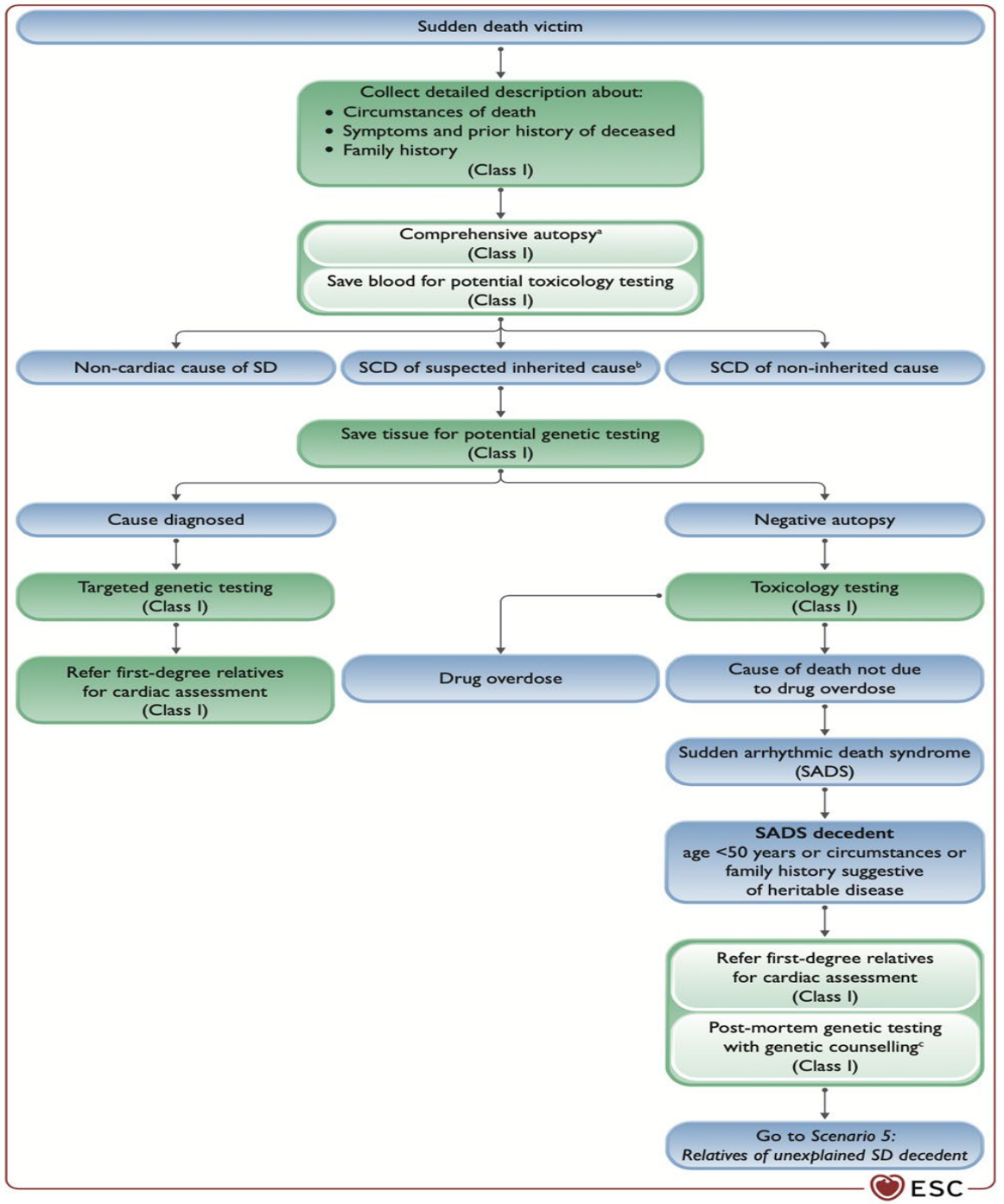
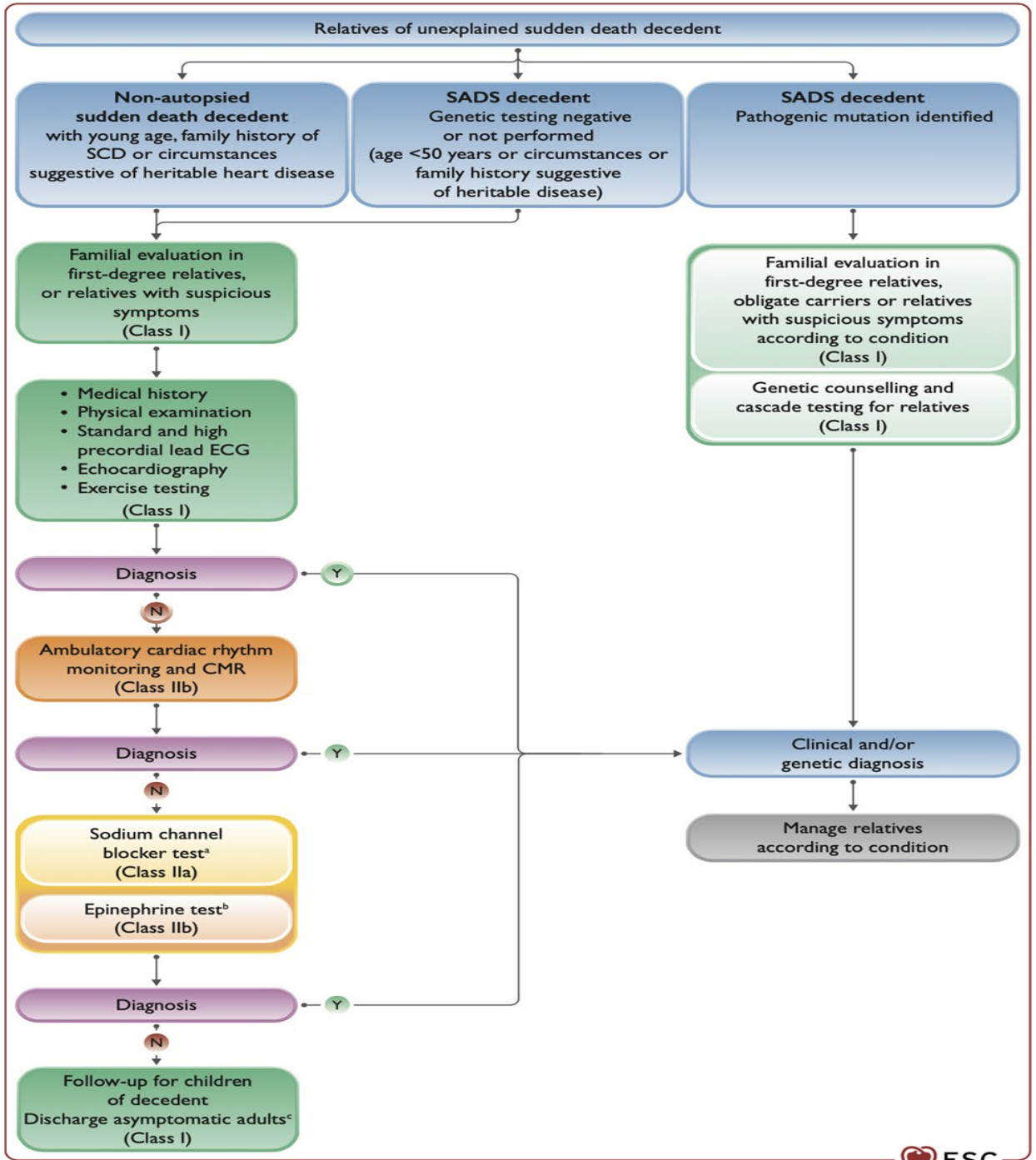
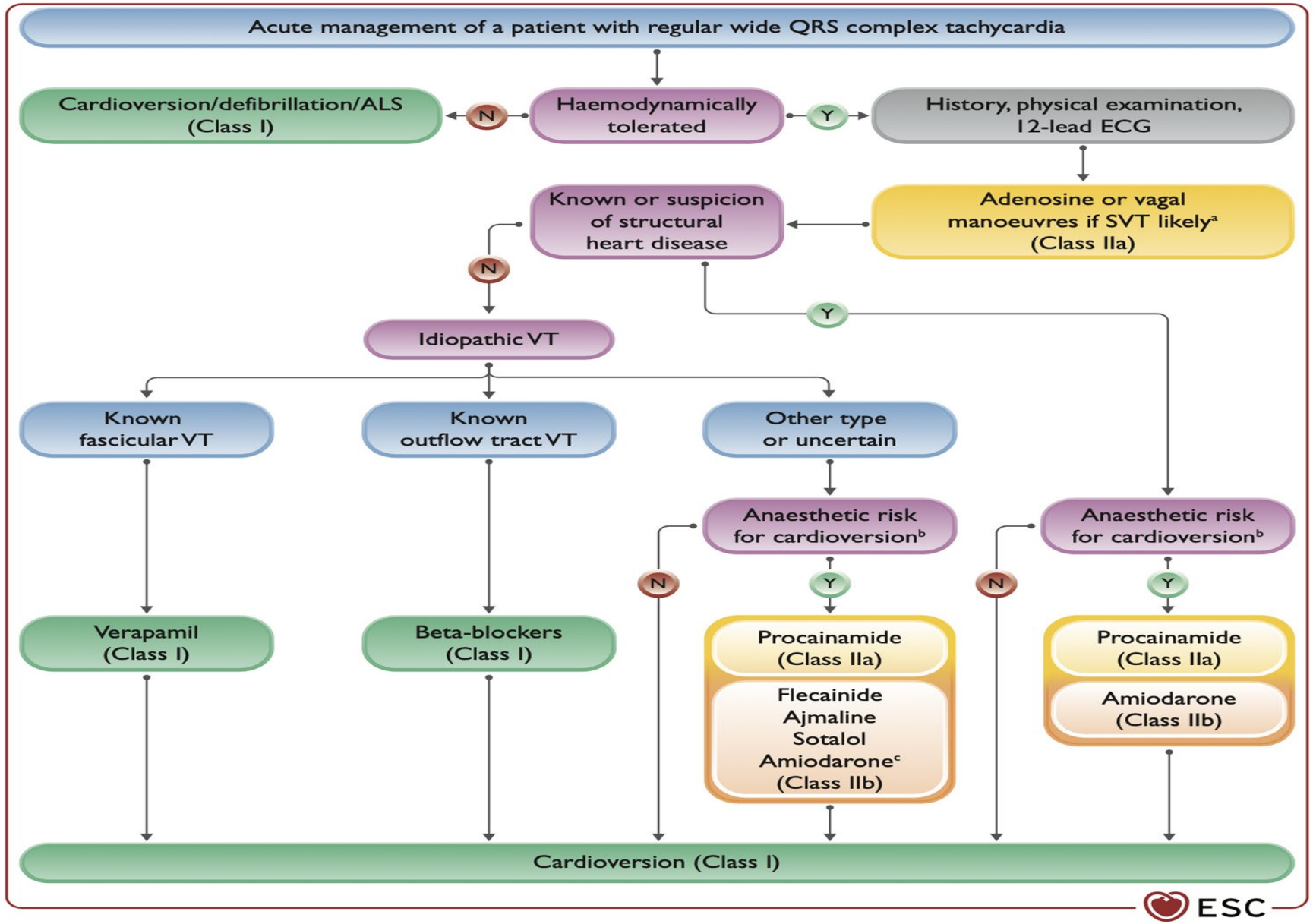
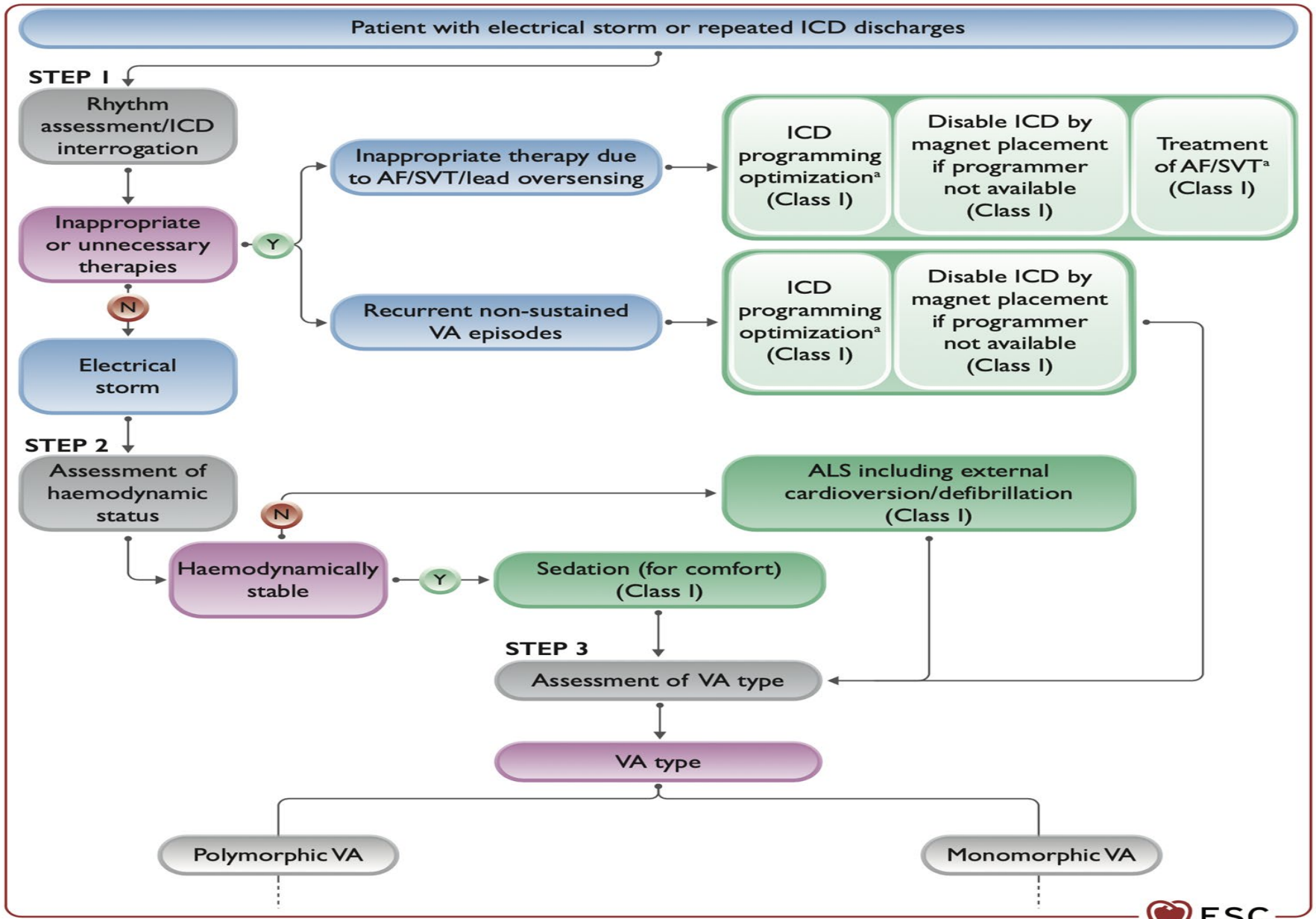
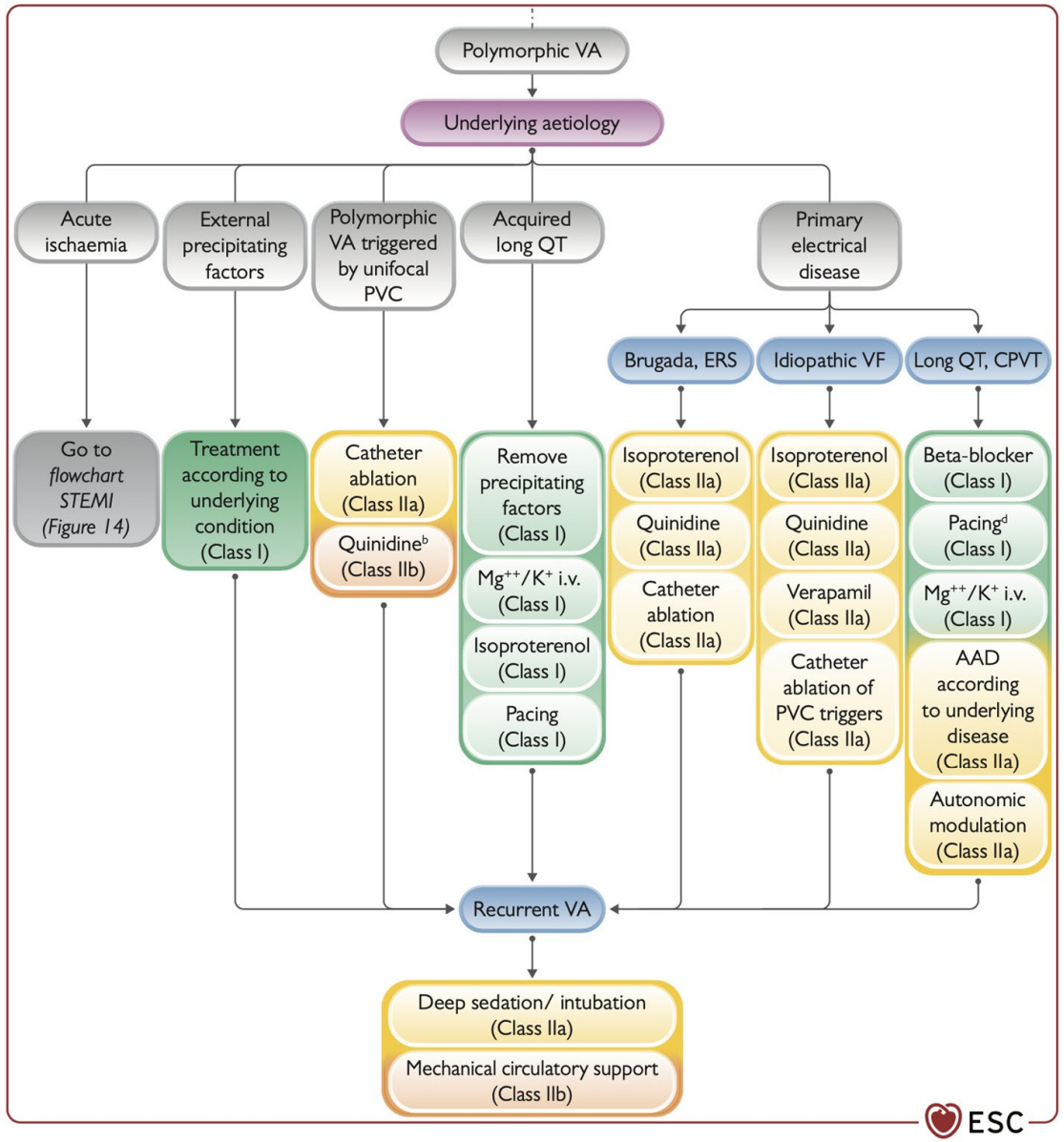


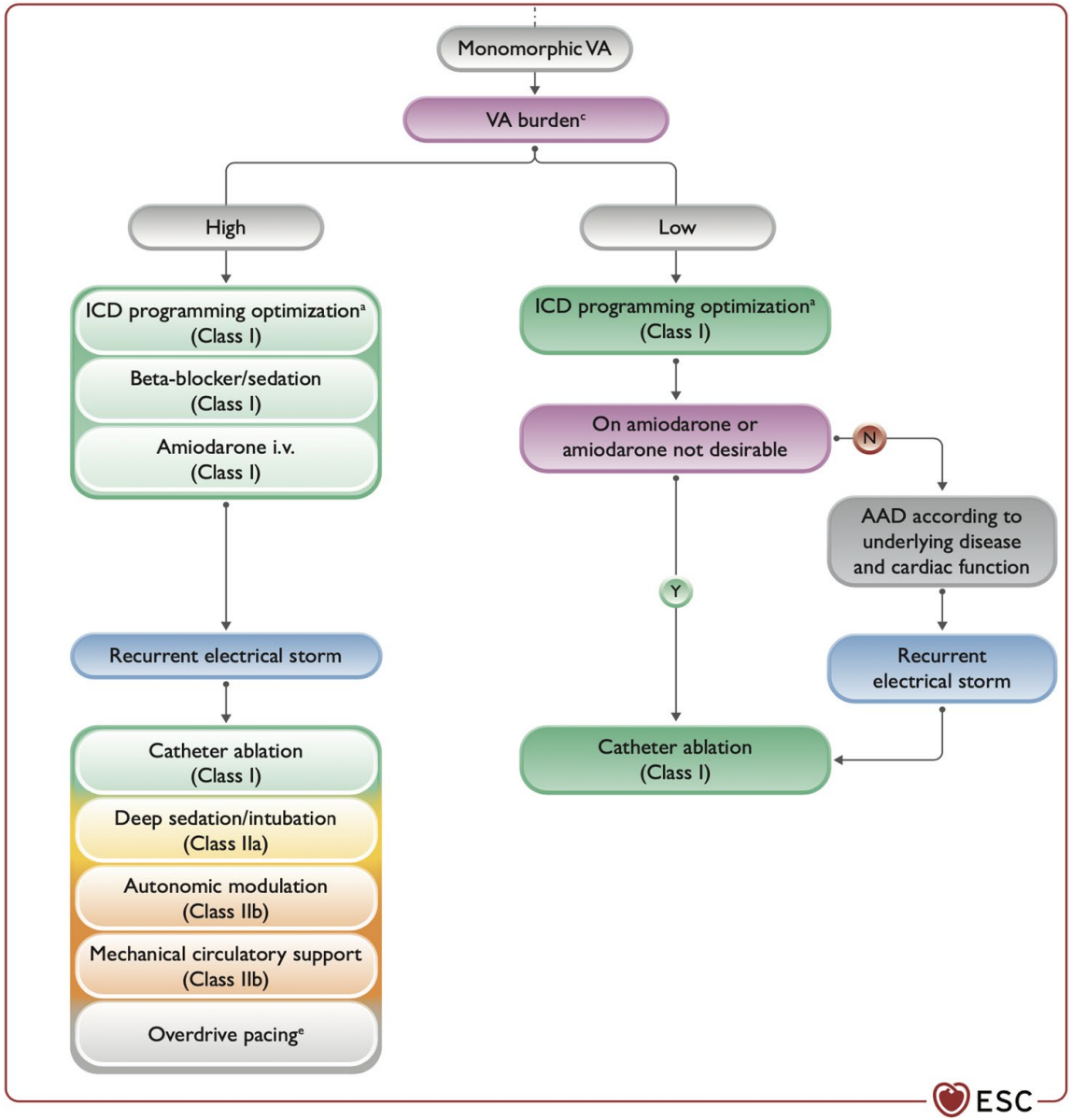
FAMILIARES DE MS




Acute management of a patient with regular wide QRS complex tachycardia
Haemodynamically tolerated
N
Cardioversion/defibrillation/ALS (Class I)
Y
History, physical examination, 12-lead ECG
Adenosine or vagal manoeuvres if SVT likely[a] (Class IIa)
Known or suspicion of structural heart disease
N
Y
Idiopathic VT
Known fascicular VT
Known outflow tract VT
Other type or uncertain
Anaesthetic risk for cardioversion[b]
N
Y
Anaesthetic risk for cardioversion[b]
N
Y
Verapamil (Class I)
Beta-blockers (Class I)
Procainamide (Class IIa)
Flecainide
Ajmaline
Sotalol
Amiodarone[c]
(Class IIb)
Procainamide (Class IIa)
Amiodarone (Class IIb)
Cardioversion (Class I)
ESC


Patient with electrical storm or repeated ICD discharges
STEP 1
Rhythm assessment/ICD interrogation
Inappropriate or unnecessary therapies
Y
N
Electrical storm
Inappropriate therapy due to AF/SVT/lead oversensing
Recurrent non-sustained VA episodes
ICD programming optimization[a] (Class I)
Disable ICD by magnet placement if programmer not available (Class I)
Treatment of AF/SVT[a] (Class I)
ICD programming optimization[a] (Class I)
Disable ICD by magnet placement if programmer not available (Class I)
STEP 2
Assessment of haemodynamic status
N
Haemodynamically stable
Y
Sedation (for comfort) (Class I)
ALS including external cardioversion/defibrillation (Class I)
STEP 3
Assessment of VA type
VA type
Polymorphic VA
Monomorphic VA
ESC


Polymorphic VA
Underlying aetiology
Acute ischaemia
External precipitating factors
Polymorphic VA triggered by unifocal PVC
Acquired long QT
Primary electrical disease
Brugada, ERS
Idiopathic VF
Long QT, CPVT
Go to flowchart STEMI (Figure 14)
Treatment according to underlying condition (Class I)
Catheter ablation (Class IIa)
Quinidine[b] (Class IIb)
Remove precipitating factors (Class I)
Mg++/K+ i.v. (Class I)
Isoproterenol (Class I)
Pacing (Class I)
Isoproterenol (Class IIa)
Quinidine (Class IIa)
Catheter ablation (Class IIa)
Isoproterenol (Class IIa)
Quinidine (Class IIa)
Verapamil (Class IIa)
Catheter ablation of PVC triggers (Class IIa)
Beta-blocker (Class I)
Pacing[d] (Class I)
Mg++/K+ i.v. (Class I)
AAD according to underlying disease (Class IIa)
Autonomic modulation (Class IIa)
Recurrent VA
Deep sedation/ intubation (Class IIa)
Mechanical circulatory support (Class IIb)
ESC


Monomorphic VA
VA burden[c]
High
Low
ICD programming optimization[a] (Class I)
Beta-blocker/sedation (Class I)
Amiodarone i.v. (Class I)
Recurrent electrical storm
Catheter ablation (Class I)
Deep sedation/intubation (Class IIa)
Autonomic modulation (Class IIb)
Mechanical circulatory support (Class IIb)
Overdrive pacing[e]
ICD programming optimization[a] (Class I)
On amiodarone or amiodarone not desirable
N
Y
AAD according to underlying disease and cardiac function
Recurrent electrical storm
Catheter ablation (Class I)
ESC

Esquemas terapéuticos en fase crónica

Recommendations	Class[a]	Level[b]
Optimal medical treatment including ACE-I/ARB/ ARNIs, MRAs, beta-blockers, and SGLT2 inhibitors is indicated in all heart failure patients with reduced EF.[343–347]	I	A

NEW

Recommendation Table 12 — Recommendations for secondary prevention of sudden cardiac death*

Recommendations	Class[a]	Level[b]
ICD implantation is recommended in patients with documented VF or haemodynamically not-tolerated VT in the absence of reversible causes.[349–352]	I	A
In patients with VT/VF, an indication for ICD, and no contraindication for amiodarone, amiodarone may be considered when an ICD is not available, contraindicated for concurrent medical reasons, or declined by the patient.	IIb	C
In patients with SMVT or SPVT/VF triggered by a PVC with similar morphology and an indication for ICD, catheter ablation may be considered when an ICD is not available, contraindicated for concurrent medical reasons, or declined by the patient.	IIb	C

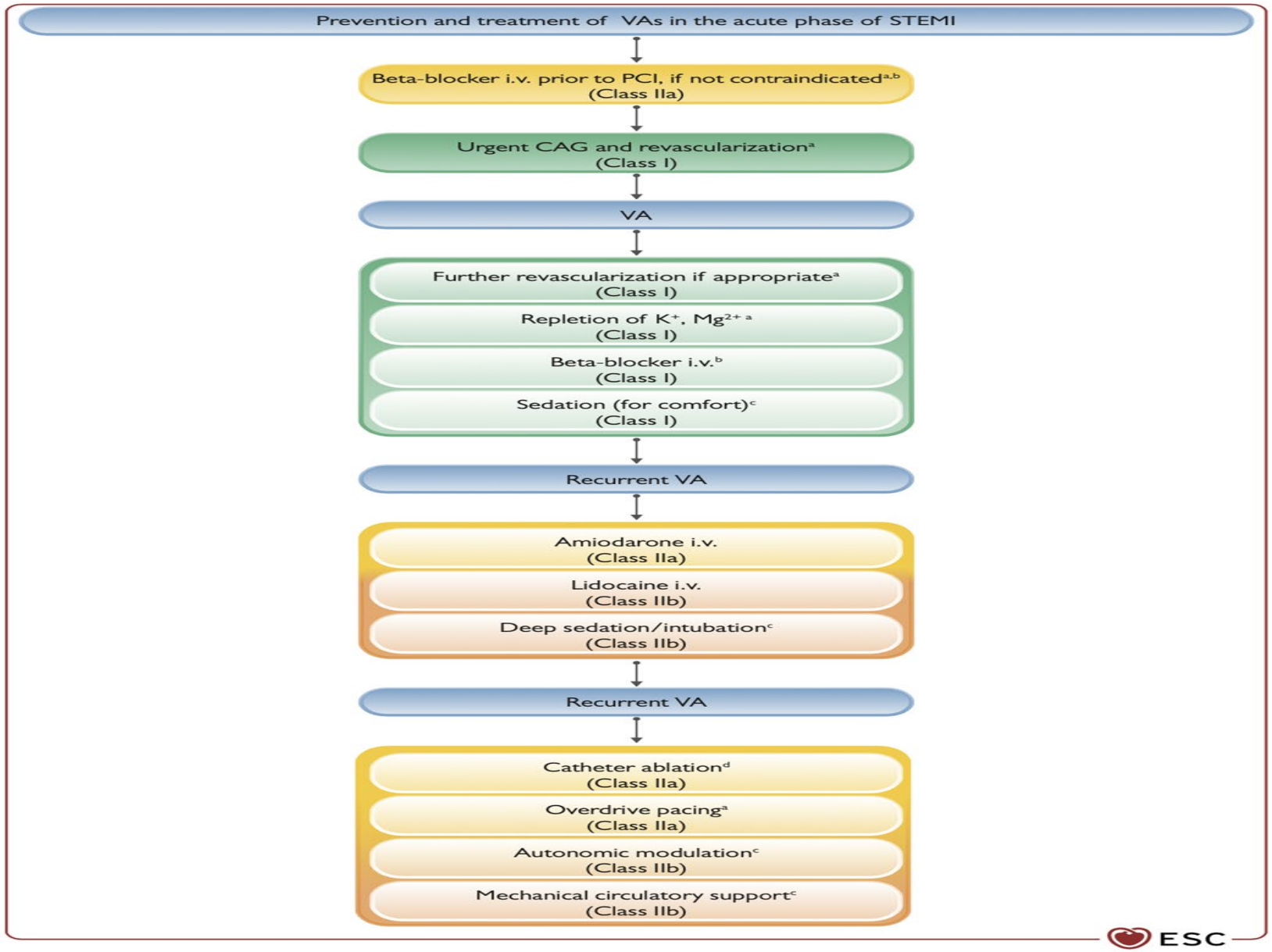
NEW

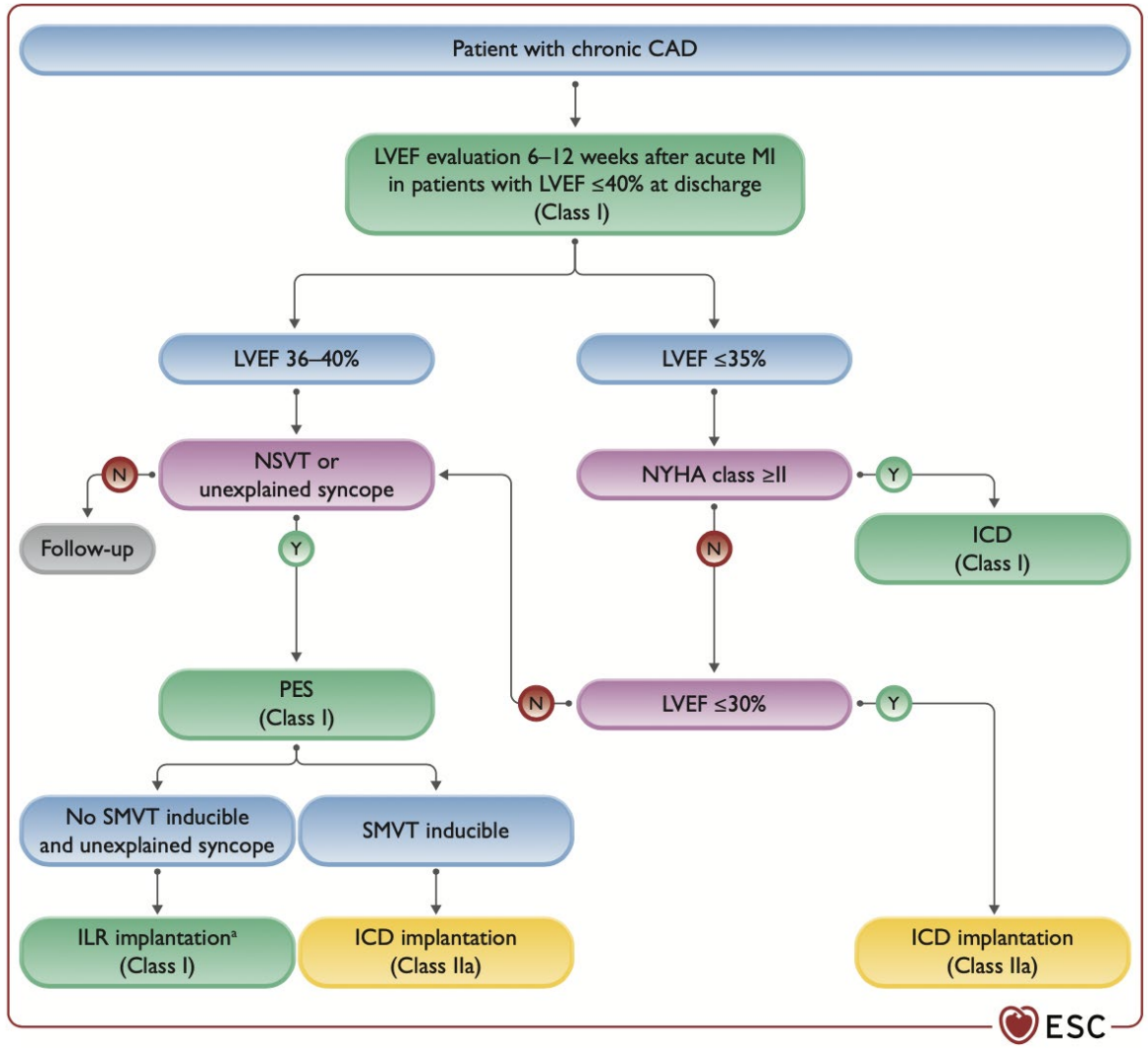
Recommendations	Class[a]	Level[b]
Implantation of a cardioverter defibrillator is only recommended in patients who have an expectation of good quality survival >1 year.	I	C
It is not recommended to implant an ICD in patients with incessant VAs until the VA is controlled.	III	C

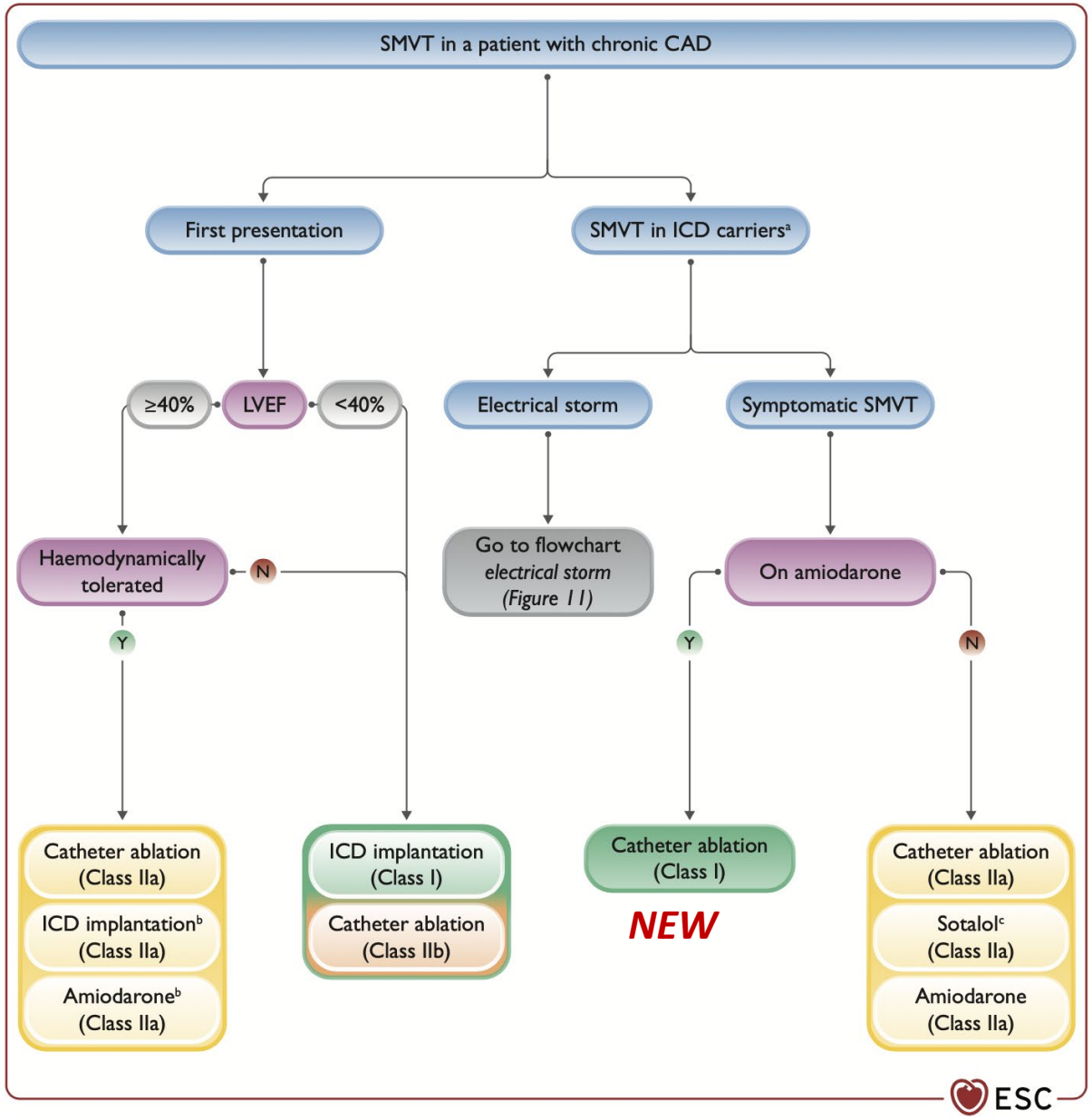
NEW

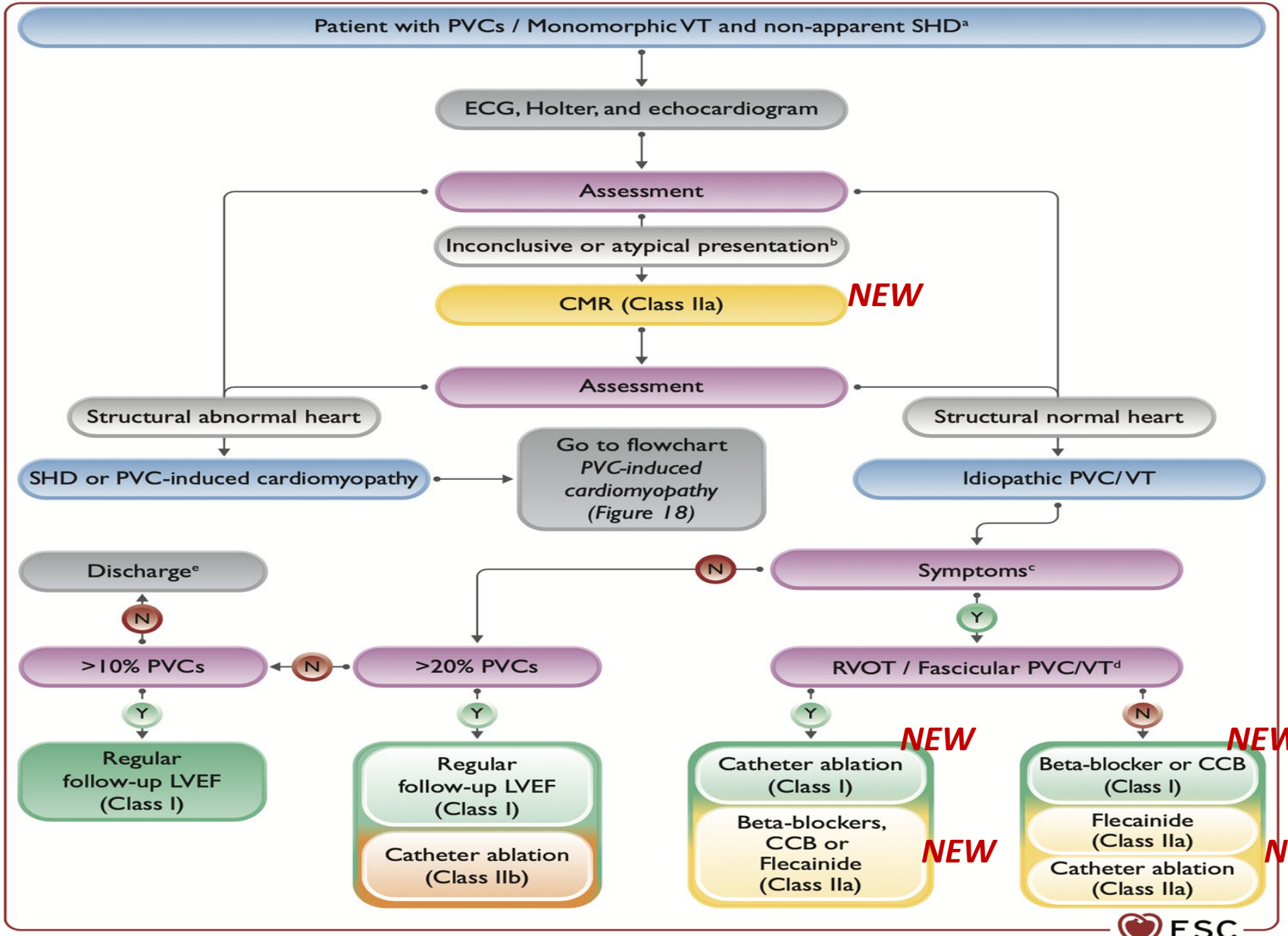
Recommendations	Class[a]	Level[b]
When an ICD is indicated, it is recommended to evaluate whether the patient could benefit from CRT-defibrillator.[367]	I	C

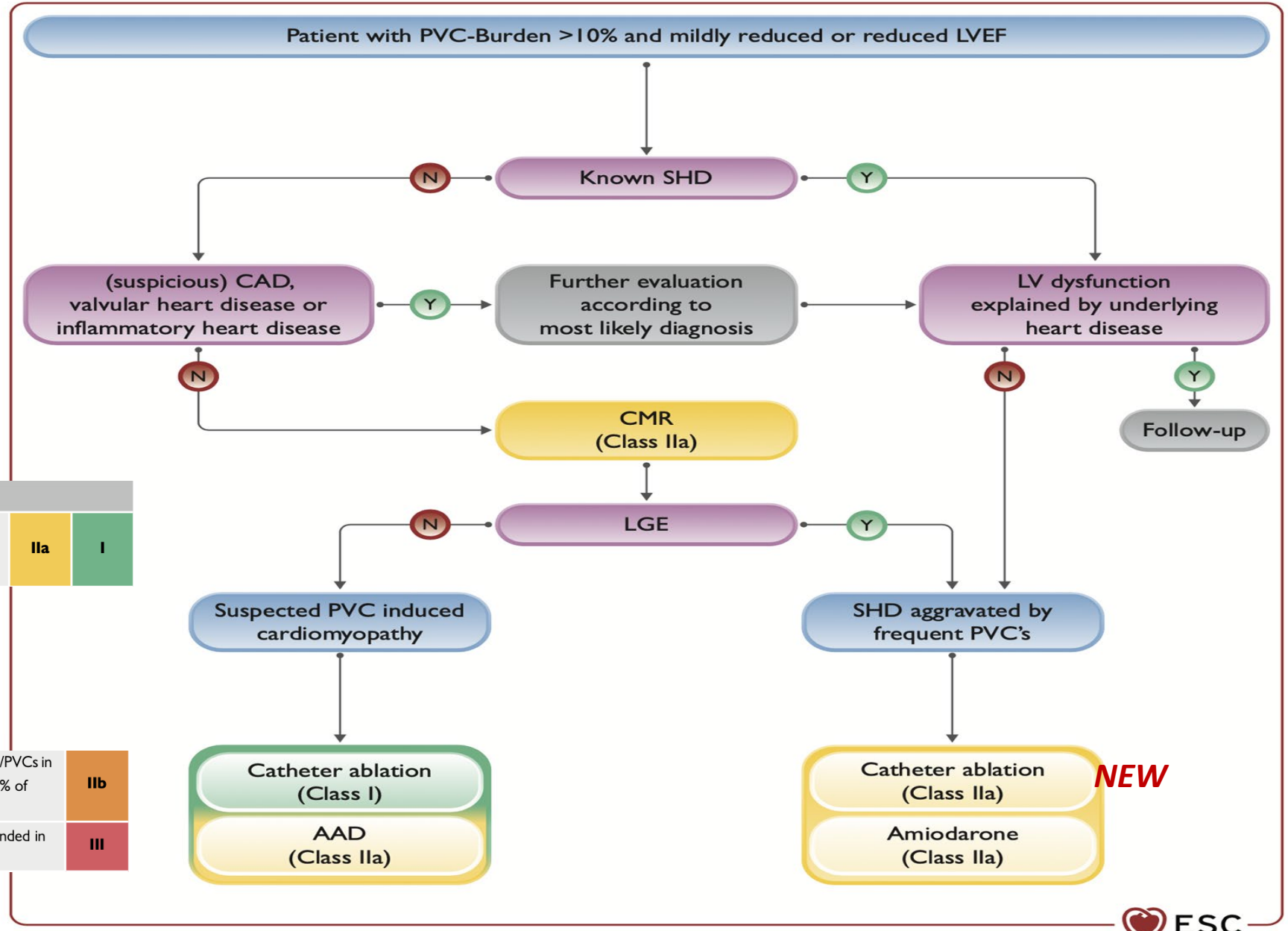
ICD, implantable cardioverter defibrillator; PVC, premature ventricular complex; SMVT, sustained monomorphic VT; SPVT, sustained polymorphic VT; VF, ventricular fibrillation; VT, ventricular tachycardia.
*For primary prevention and specific aspects of secondary prevention, go to Section 7.
[a]Class of recommendation.
[b]Level of evidence.


Prevention and treatment of VAs in the acute phase of STEMI
Beta-blocker i.v. prior to PCI, if not contraindicated[a,b] (Class IIa)
Urgent CAG and revascularization[a] (Class I)
VA
Further revascularization if appropriate[a] (Class I)
Repletion of K^+, Mg^{2+} [a] (Class I)
Beta-blocker i.v.[b] (Class I)
Sedation (for comfort)[c] (Class I)
Recurrent VA
Amiodarone i.v. (Class IIa)
Lidocaine i.v. (Class IIb)
Deep sedation/intubation[c] (Class IIb)
Recurrent VA
Catheter ablation[d] (Class IIa)
Overdrive pacing[a] (Class IIa)
Autonomic modulation[c] (Class IIb)
Mechanical circulatory support[c] (Class IIb)
ESC


Patient with chronic CAD
LVEF evaluation 6–12 weeks after acute MI in patients with LVEF ≤40% at discharge (Class I)
LVEF 36–40%
LVEF ≤35%
NSVT or unexplained syncope
N
Y
Follow-up
NYHA class ≥II
Y
N
ICD (Class I)
PES (Class I)
N
LVEF ≤30%
Y
No SMVT inducible and unexplained syncope
SMVT inducible
ILR implantation[a] (Class I)
ICD implantation (Class IIa)
ICD implantation (Class IIa)
ESC


SMVT in a patient with chronic CAD
First presentation
SMVT in ICD carriers[a]
≥40%
LVEF
<40%
Electrical storm
Symptomatic SMVT
Haemodynamically tolerated
N
Y
Go to flowchart electrical storm (Figure 11)
On amiodarone
Y
N
Catheter ablation (Class IIa)
ICD implantation[b] (Class IIa)
Amiodarone[b] (Class IIa)
ICD implantation (Class I)
Catheter ablation (Class IIb)
Catheter ablation (Class I)
NEW
Catheter ablation (Class IIa)
Sotalol[c] (Class IIa)
Amiodarone (Class IIa)
ESC


Patient with PVCs / Monomorphic VT and non-apparent SHD[a]
ECG, Holter, and echocardiogram
Assessment
Inconclusive or atypical presentation[b]
CMR (Class IIa)
NEW
Assessment
Structural abnormal heart
Structural normal heart
SHD or PVC-induced cardiomyopathy
Go to flowchart PVC-induced cardiomyopathy (Figure 18)
Idiopathic PVC/VT
Discharge[e]
N
Symptoms[c]
N
Y
>10% PVCs
N
>20% PVCs
RVOT / Fascicular PVC/VT[d]
Y
Y
Y
N
Regular follow-up LVEF (Class I)
Regular follow-up LVEF (Class I)
Catheter ablation (Class IIb)
Catheter ablation (Class I)
NEW
Beta-blockers, CCB or Flecainide (Class IIa)
NEW
Beta-blocker or CCB (Class I)
NEW
Flecainide (Class IIa)
Catheter ablation (Class IIa)
NEW
ESC



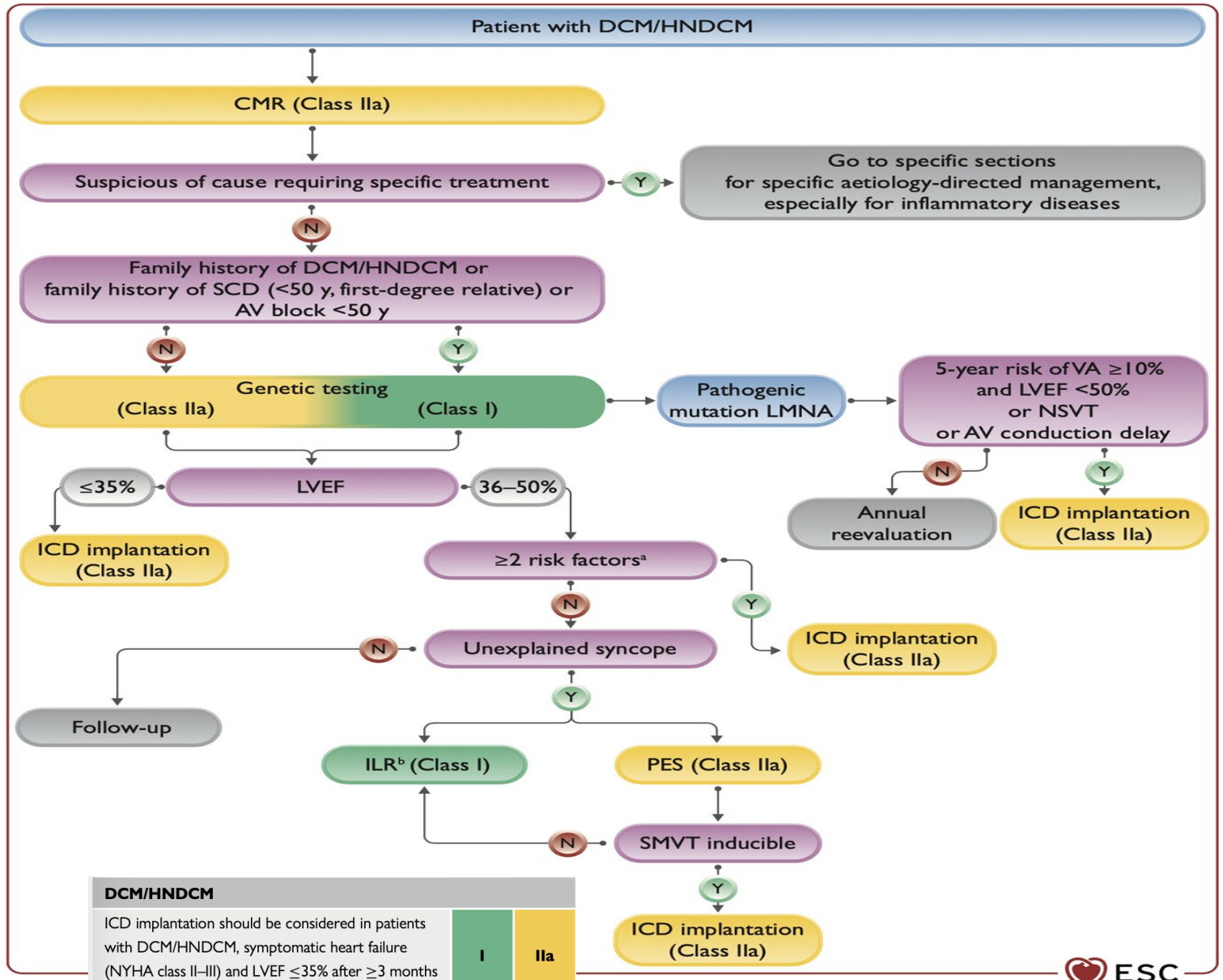
PVC-induced cardiomyopathy		
In patients with a cardiomyopathy suspected to be caused by frequent and predominately monomorphic PVCs, catheter ablation is recommended.	IIa	I

NEW

Catheter ablation may be considered for idiopathic VT/PVCs in asymptomatic patients with repeatedly more than 20% of PVCs per day at follow-up.	IIb
Amiodarone as a first-line treatment is not recommended in patients with idiopathic VTs/PVCs.	III

NEW

DCM/HNDCM	
Genetic testing (including at least *LMNA*, *PLN*, *RBM20*, and *FLNC* genes) is recommended in patients with DCM/HNDCM and AV conduction delay at <50 years, or who have a family history of DCM/HNDCM or SCD in a first-degree relative (at age <50 years).	I
In a first-degree relative of a DCM/HNDCM patient, an ECG, and an echocardiogram are recommended if: • the index patient was diagnosed <50 years of age or has clinical features suggestive of an inherited cause, or • there is a family history of DCM/HNDCM, or premature unexpected SD.	I
CMR with LGE should be considered in DCM/HNDCM patients for assessing the aetiology and the risk of VA/SCD.	IIa
Genetic testing (including at least *LMNA*, *PLN*, *RBM20*, and *FLNC* genes) should be considered for risk stratification in patients with apparently sporadic DCM/HNDCM, who present at young age or with signs suspicious for an inherited aetiology.	IIa
ICD implantation should be considered in DCM/HNDCM patients with an LVEF <50% and ≥2 risk factors (syncope, LGE on CMR, inducible SMVT at PES, pathogenic mutations in LMNA, *PLN*, *FLNC*, and *RBM20* genes).	IIa
ICD implantation should be considered in patients with DCM/HNDCM and haemodynamically tolerated SMVT.	IIa
In a first-degree relative of a patient with apparently sporadic DCM/HNDCM, an ECG, and an echocardiogram may be considered.	IIb
Participation in high-intensity exercise including competitive sports is not recommended for individuals with DCM/HNDCM and a *LMNA* mutation.	III



DCM/HNDCM		
ICD implantation should be considered in patients with DCM/HNDCM, symptomatic heart failure (NYHA class II–III) and LVEF ≤35% after ≥3 months of OMT.	I	IIa
Catheter ablation in specialized centres should be considered in patients with DCM/HNDCM and recurrent, symptomatic SMVT, or ICD shocks for SMVT, in whom AADs are ineffective, contraindicated, or not tolerated.	IIb	IIa

MIOCARDIOPATÍA ARRITMOGÉNICA

Diagnostic evaluation and general recommendations			
In patients with suspected ARVC, CMR is recommended.[676–678]	I	B	NEW
In patients with a suspected or definite diagnosis of ARVC, genetic counselling and testing are recommended.[672,673]	I	B	NEW
Avoidance of high-intensity exercise is recommended in patients with a definite diagnosis of ARVC.[683–685]	I	B	
Avoidance of high-intensity[c] exercise may be considered in carriers of ARVC-related pathogenic mutations and no phenotype.[683,687]	IIb	C	
Beta-blocker therapy may be considered in all patients with a definite diagnosis of ARVC.	IIb	C	

Risk stratification and primary prevention of SCD			
ICD implantation should be considered in patients with definite ARVC and an arrhythmic syncope.[696,701,711–713]	IIa	B	
ICD implantation should be considered in patients with definite ARVC and severe RV or LV systolic dysfunction.[675,691]	IIa	C	
ICD implantation should be considered in symptomatic[d] patients with definite ARVC, moderate right or left ventricular dysfunction, and either NSVT or inducibility of SMVT at PES.[695,696,701,703,705]	IIa	C	NEW
In patients with ARVC and symptoms highly suspicious for VA, PES may be considered for risk stratification.[695,705]	IIb	C	

MIOCARDIOPATÍA ARRITMOGÉNICA

Secondary prevention of SCD and treatment of VAs		
ICD implantation is recommended in ARVC patients with haemodynamically not-tolerated VT or VF.[700]	I	C
In patients with ARVC and non-sustained or sustained VAs, beta-blocker therapy is recommended.	I	C
In patients with ARVC and recurrent, symptomatic SMVT or ICD shocks for SMVT despite beta-blockers, catheter ablation in specialized centres should be considered.[482,709,714]	IIa	C
In ARVC patients with indication for ICDs, a device with the capability of ATP programming for SMVT up to high rates should be considered.[698]	IIa	B
ICD implantation should be considered in ARVC patients with a haemodynamically tolerated SMVT.[692]	IIa	C
In patients with ARVC and recurrent, symptomatic VT despite beta-blockers, AAD treatment should be considered.[709,710]	IIa	C
Management of relatives of a patient with ARVC		
In a first-degree relative of a patient with ARVC, ECG and echocardiogram are recommended.[675]	I	C

NEW

NEW

Avoidance of high-intensity[e] exercise may be considered in carriers of ARVC-related pathogenic mutations and no phenotype.	IIb
Beta-blocker therapy may be considered in all patients with a definite diagnosis of ARVC.	IIb
In patients with ARVC and symptoms highly suspicious for VA, PES may be considered for risk stratification.	IIb

ARVC		
ICD implantation should be considered in patients with definite ARVC and an arrhythmic syncope.	IIb	IIa
ICD implantation should be considered in patients with definite ARVC and severe RV or LV systolic dysfunction.	IIb	IIa

MIOCARDIOPATÍA HIPERTRÓFICA

Recommendation	Class[a]	Level[b]	
Diagnostic evaluation and general recommendations			
CMR with LGE is recommended in HCM patients for diagnostic work-up.[716–718]	I	B	NEW
Genetic counselling and testing are recommended in HCM patients.[721–725]	I	B	NEW
Participation in high-intensity exercise may be considered for asymptomatic adult HCM patients without risk markers.[733]	IIb	C	
Risk stratification and primary prevention of SCD			
It is recommended that the 5-year risk of SCD is assessed at first evaluation and at 1–3-year intervals, or when there is a change in clinical status.	I	C	
ICD implantation should be considered in patients aged 16 years or more with an estimated 5-year risk of SD ≥6%.[c,85,728,729]	IIa	B	
ICD implantation should be considered in HCM patients aged 16 years or more with an intermediate 5-year risk of SCD (≥4 to <6%)[c] and with (a) significant LGE at CMR (usually ≥15% of LV mass); or (b) LVEF < 50%; or (c) abnormal blood pressure response during exercise test[d]; or (d) LV apical aneurysm; or (e) presence of sarcomeric pathogenic mutation.[716,717,722,736–739]	IIa	B	NEW
In children less than 16 years of age with HCM and an estimated 5-year risk of SD ≥6% (based on HCM Risk-Kids score[e]), ICD implantation should be considered.[84,742]	IIa	B	NEW
ICD implantation may be considered in HCM patients aged 16 years or more with an estimated 5-year risk of SCD of ≥4 to <6%.[c,85,728,729]	IIb	B	
ICD implantation may be considered in HCM patients aged 16 years or more with a low estimated 5-year risk of SCD (<4%)[c] and with (a) significant LGE at CMR (usually ≥15% of LV mass); or (b) LVEF < 50%; or (c) LV apical aneurysm.[716,717,722,736–739]	IIb	B	

Secondary prevention of SCD and treatment of VAs			
ICD implantation is recommended in HCM patients with haemodynamically not-tolerated VT or VF.[744–746]	I	B	
In patients with HCM presenting with haemodynamically tolerated SMVT, ICD implantation should be considered.	IIa	C	NEW
In patients with HCM and recurrent, symptomatic VA, or recurrent ICD therapy, AAD treatment should be considered.	IIa	C	NEW
Catheter ablation in specialized centres may be considered in selected patients with HCM and recurrent, symptomatic SMVT or ICD shocks for SMVT, in whom AAD are ineffective, contraindicated, or not tolerated.[753,754]	IIb	C	
Management of relatives of a patient with HCM			
In a first-degree relative of a patient with HCM, ECG and echocardiogram are recommended.	I	C	NEW

MIOCARDIOPATÍA HIPERTRÓFICA

NEW

Participation in high-intensity exercise may be considered for asymptomatic adult HCM patients without risk markers.	**IIb**
ICD implantation may be considered in HCM patients aged 16 years or more with a low estimated 5-year risk of SCD (<4%),[f] and with (a) significant LGE at CMR (usually ≥15% of LV mass); or (b) LVEF < 50%; or (c) LV apical aneurysm.	**IIb**
Catheter ablation in specialized centres may be considered in selected patients with HCM and recurrent, symptomatic SMVT, or ICD shocks for SMVT, in whom AADs are ineffective, contraindicated, or not tolerated.	**IIb**

MIOCARDIOPATÍA NO COMPACTADA Y AMILOIDOSIS

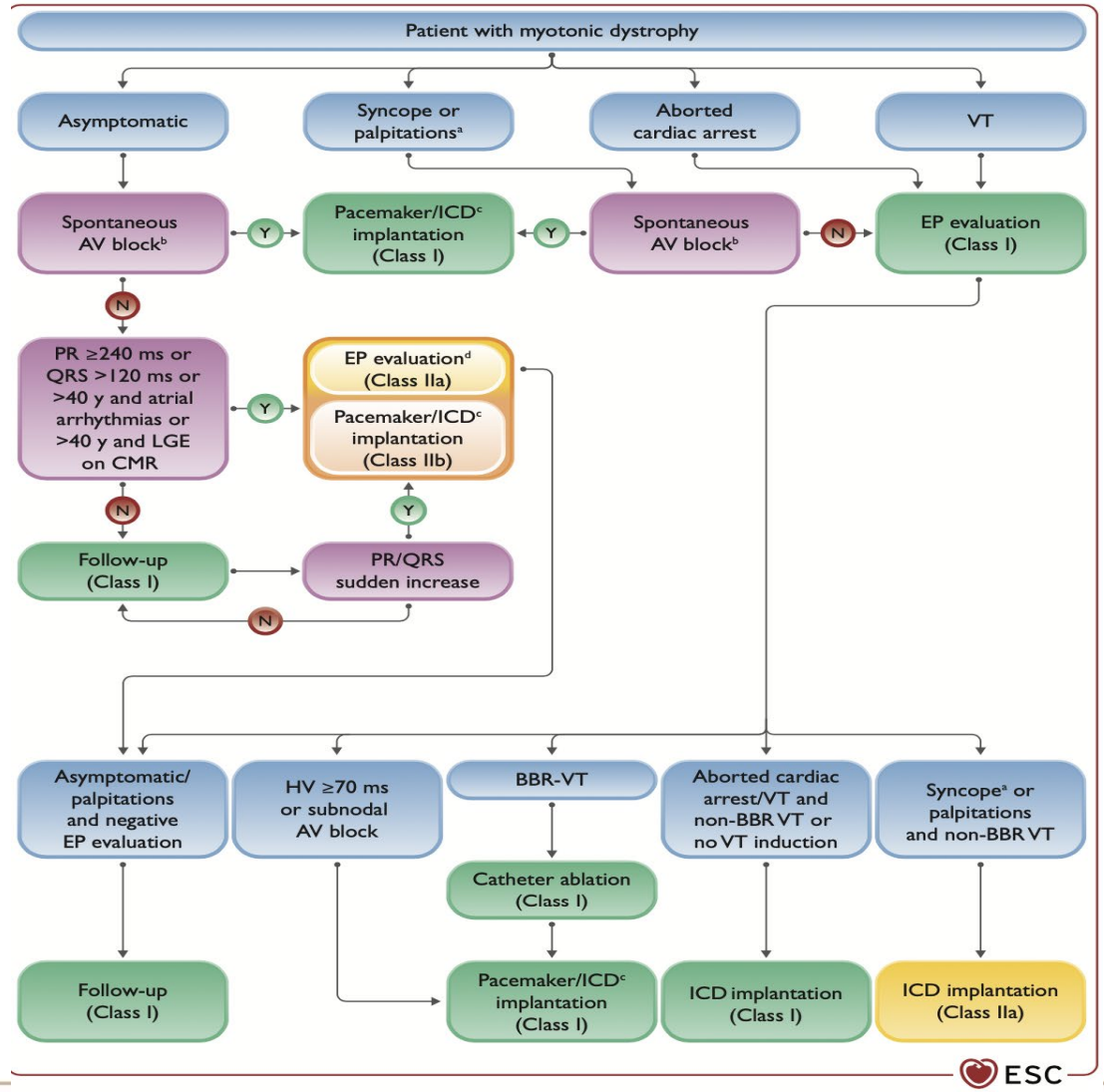
NEW

Recommendations	Class[a]	Level[b]
In patients with a LVNC cardiomyopathy phenotype based on CMR or echocardiography, implantation of an ICD for primary prevention of SCD should be considered to follow DCM/ HNDCM recommendations.	IIa	C

NEW

Recommendations	Class[a]	Level[b]
An ICD should be considered in patients with light-chain amyloidosis or transthyretin-associated cardiac amyloidosis and haemodynamically not-tolerated VT.	IIa	C

DISTROFIA MIOTÓNICA



Recommendations	Class[a]	Level[b]
General recommendations		
Annual follow-up with at least a 12-lead ECG is recommended in patients with muscular dystrophies, even in the concealed phase of the disease.[6,13]	I	C
It is recommended that patients with neuromuscular disorders who have VAs or ventricular dysfunction are treated in the same way for arrhythmia as patients without neuromuscular disorders.[17,765]	I	C
Risk stratification, primary and secondary prevention of SCD		
Invasive electrophysiological evaluation is recommended in patients with myotonic dystrophy and palpitations or syncope suggestive of VA or surviving a CA.[153]	I	C

NEW

DISTROFIA MIOTÓNICA

ICD implantation is recommended in patients with myotonic dystrophy and SMVT or aborted CA not caused by BBR-VT.[766]	I	C
Invasive electrophysiological evaluation should be considered in patients with myotonic dystrophy and a sudden increase in the PR interval or QRS duration.[766,767]	IIa	B
Invasive electrophysiological evaluation should be considered in patients with myotonic dystrophy and a PR interval ≥240 ms or QRS duration ≥120 ms or who are older than 40 years and have supraventricular arrhythmias[c] or who are older than 40 years and have significant LGE on CMR.[c,5,14,16,766]	IIa	B
In myotonic dystrophy patients without AV conduction delay and a syncope highly suspicious for VA, ICD implantation should be considered.[766]	IIa	C
In myotonic dystrophy patients with palpitations highly suspicious for VA and induction of a non-BBR-VT, ICD implantation should be considered.[766]	IIa	C
In patients with limb–girdle type 1B or Emery–Dreifuss muscular dystrophies and indication for pacing, ICD implantation should be considered.[769]	IIa	C
Implantation of an ICD may be considered in patients with Duchenne/Becker muscular dystrophy and significant LGE at CMR.[770,771]	IIb	C
Implantation of an ICD over a permanent pacemaker may be considered in myotonic dystrophy patients with additional risk factors[d] for VAs and SCD.	IIb	C
In myotonic dystrophy patients, serial electrophysiological evaluation of AV conduction and arrhythmia induction is not recommended without arrhythmia suspicion or progression of ECG conduction disorders.[772]	III	C
Management of VA		
In symptomatic patients with BBR-VT, catheter ablation is recommended.[e,153,474,475,477]	I	C
In patients with myotonic dystrophy undergoing ablation for BBR-VT, pacemaker/ICD implantation is recommended.[153]	I	C

NEW

MIOCARDITIS

Recommendations	Class[a]	Level[b]
General recommendations		
In confirmed or clinically suspected acute myocarditis, it is recommended that patients who present with life-threatening VAs are referred to a specialized centre.[786,804]	I	C
Secondary prevention of SCD and treatment of VA		
In patients with haemodynamically not-tolerated SMVT occurring in the chronic phase of myocarditis, an ICD implantation is recommended.[794,805]	I	C
NEW In patients with haemodynamically not-tolerated sustained VT or VF during the acute phase of myocarditis, ICD implantation before hospital discharge should be considered.[788,794,806]	IIa	C
AADs should be considered (preferably amiodarone and beta-blockers) in patients with symptomatic non-sustained or sustained VAs during the acute phase of myocarditis.	IIa	C
NEW In post-myocarditis patients with recurrent, symptomatic VT, AAD treatment should be considered.	IIa	C
NEW Catheter ablation, performed in specialized centres, should be considered in post-myocarditis patients with recurrent, symptomatic SMVT or ICD shocks for SMVT in whom AADs are ineffective, not tolerated, or not desired.[752,801,802]	IIa	C
NEW In patients with haemodynamically tolerated SMVT occurring in the chronic phase of myocarditis, ICD implantation should be considered.	IIa	C

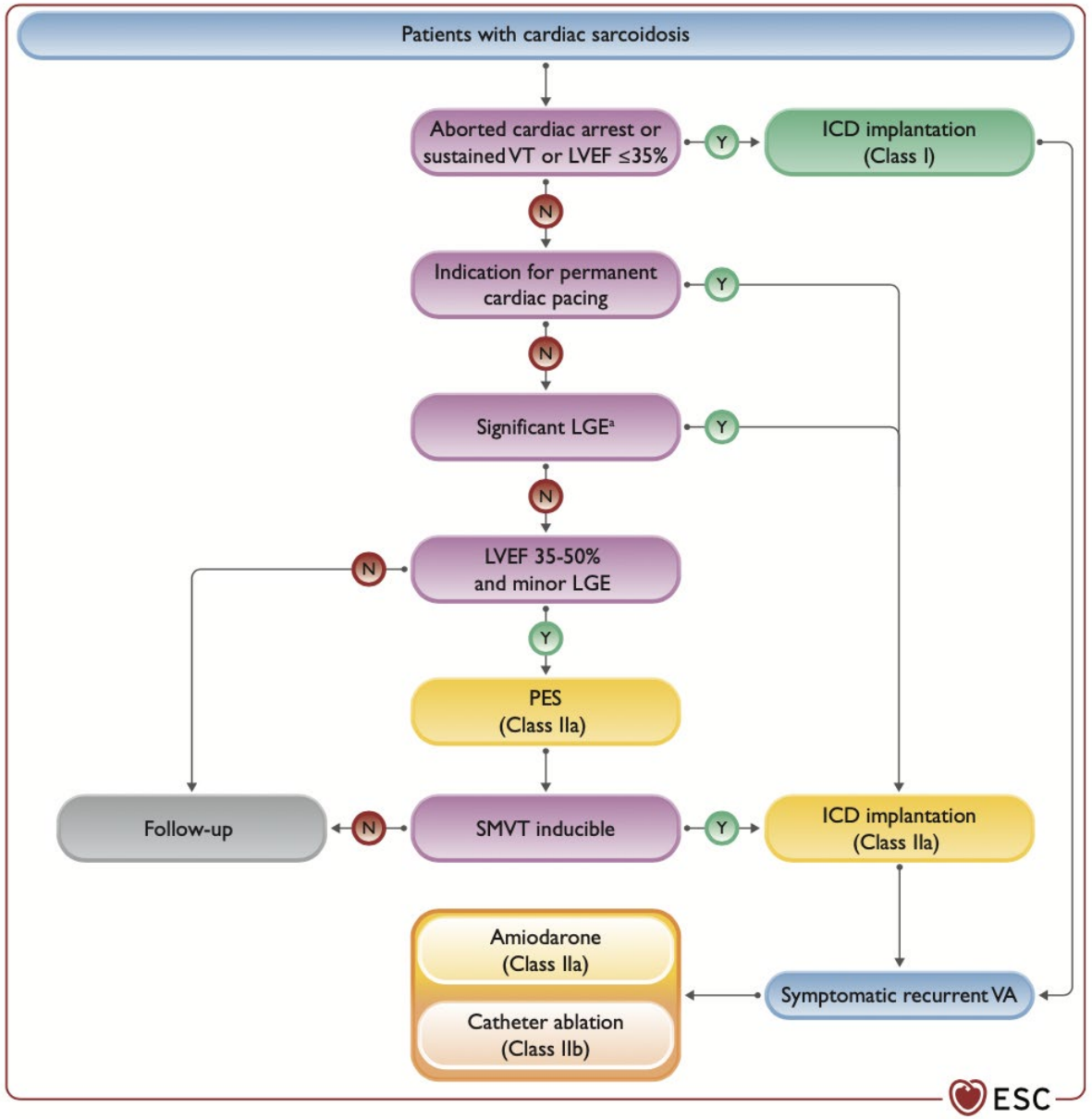
IIa---I

NEW

In patients with haemodynamically well-tolerated SMVT occurring in the chronic phase of myocarditis, preserved LV function and a limited scar amenable to ablation, catheter ablation may be considered as an alternative to ICD therapy, after discussion with the patient and provided that established endpoints have been reached.[c]	IIb	C

SARCOIDOSIS

ICD implantation is recommended in patients with cardiac sarcoidosis who have an LVEF ≤35%.	IIb	I
ICD implantation is recommended in patients with cardiac sarcoidosis who (1) have documented sustained VT, or (2) aborted CA.	IIb	I
In patients with cardiac sarcoidosis who have an indication for permanent cardiac pacing related to high-degree AV block, ICD implantation should be considered, regardless of LVEF.	IIb	IIa



SARCOIDOSIS

Recommendations	Class[a]	Level[b]	
Risk stratification and primary prevention of SCD			
ICD implantation is recommended in patients with cardiac sarcoidosis who have a LVEF ≤35%.[812,828–830,832]	I	B	
In patients with cardiac sarcoidosis who have an indication for permanent cardiac pacing related to high-degree AV block, ICD implantation should be considered, regardless of LVEF.[816]	IIa	C	
In patients with cardiac sarcoidosis who have a LVEF >35% but significant LGE at CMR after resolution of acute inflammation, ICD implantation should be considered.[817–819,821,833,834]	IIa	B	***NEW***
In patients with cardiac sarcoidosis who have a LVEF 35–50% and minor LGE at CMR, after resolution of acute inflammation, PES for risk stratification should be considered.	IIa	C	***NEW***
In patients with cardiac sarcoidosis, LVEF 35–50% and inducible SMVT at PES, ICD implantation should be considered.[823–825]	IIa	C	***NEW***

Secondary prevention of SCD and treatment of VAs			
ICD implantation is recommended in patients with cardiac sarcoidosis who (1) have documented sustained VT, or (2) aborted CA.[812,828–830,832]	I	B	
In patients with cardiac sarcoidosis and recurrent, symptomatic VA, AAD treatment should be considered.	IIa	C	***NEW***
Catheter ablation, in specialized centres, may be considered in cardiac sarcoidosis ICD-recipients with recurrent, symptomatic SMVT or ICD shocks for SMVT, in whom AADs are ineffective, contraindicated, or not tolerated.[839,841,842]	IIb	C	***NEW***

ENFERMEDAD DE CHAGAS

In patients with Chagas' cardiomyopathy and symptomatic VT in whom AADs (amiodarone and beta-blockers) are ineffective or not tolerated, ICD implantation may be considered.	IIa	IIb

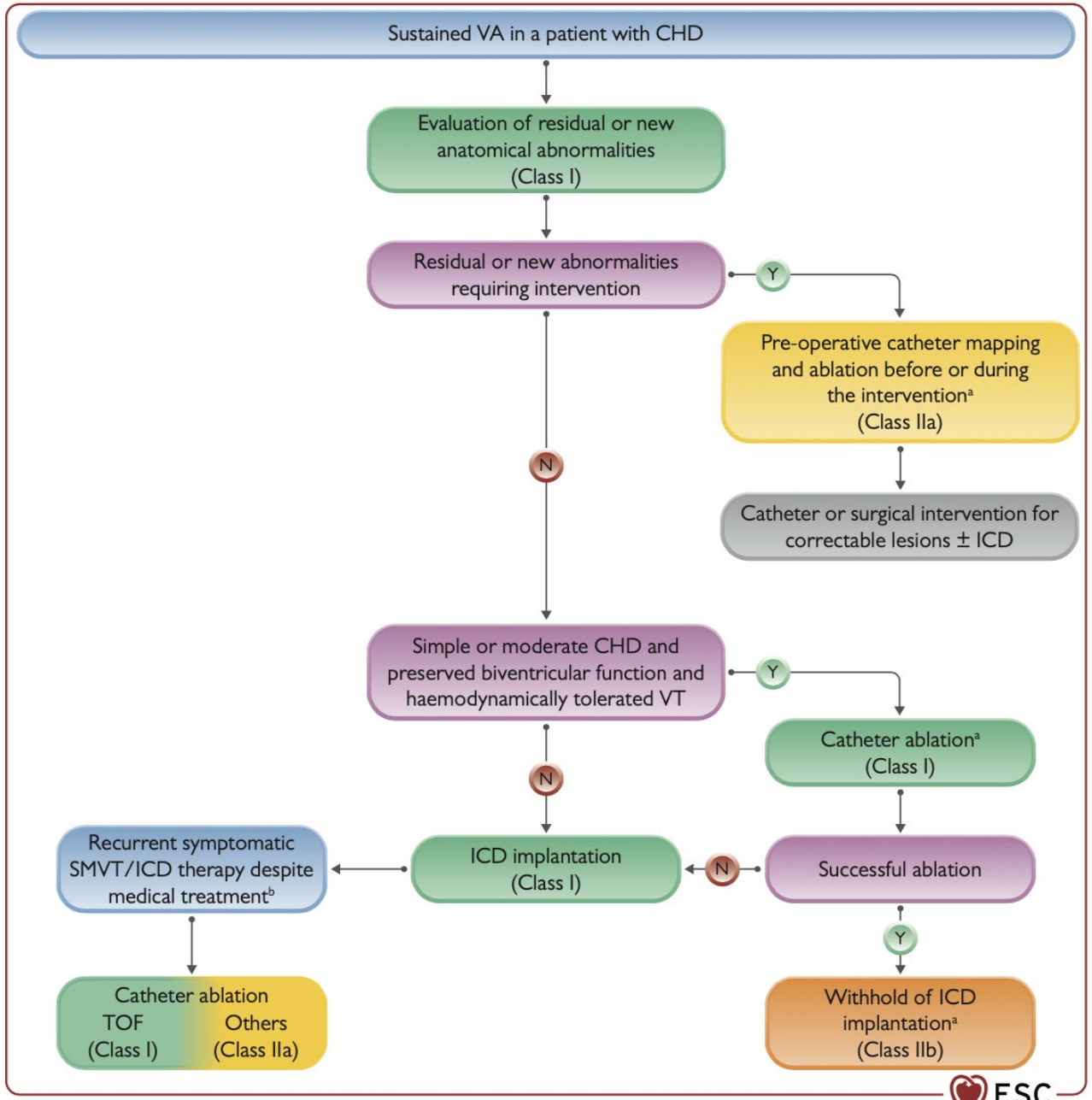
Recommendations	Class[a]	Level[b]	
Amiodarone should be considered to reduce arrhythmia burden in patients with Chagas' cardiomyopathy who present with symptomatic PVCs or VT.[857]	IIa	C	*NEW*
In patients with Chagas' cardiomyopathy and recurrent, symptomatic SMVT or ICD shocks for SMVT in whom AADs are ineffective, contraindicated, or not tolerated, catheter ablation in specialized centres should be considered.[859,860]	IIa	C	*NEW*
In patients with Chagas' cardiomyopathy and symptomatic VT in whom AADs (amiodarone and beta-blockers) are ineffective or not tolerated, ICD implantation may be considered.[851,854–856]	IIb	C	

VALVULOPATÍAS

Recommendations	Class[a]	Level[b]
PES with standby catheter ablation is recommended in patients with aortic valve disease and SMVT to identify and ablate BBR-VT, especially if it occurs following a valve intervention.[871,872,881]	I	C
In patients with valvular heart disease and persistent LV dysfunction after surgical correction, (if possible) it is recommended that ICD implantation for primary prevention follows DCM/HNDCM recommendations.[868]	I	C

CARDIOPATÍAS CONGÉNITAS

CHD		
In patients after repair of TOF without arrhythmia symptoms, but with a combination of other risk factors,[a] electrophysiologic evaluation, including PES, may be considered.	IIa	IIb
In patients with CHD and recurrent, symptomatic SMVT, or ICD shocks for SMVT not manageable by medical therapy or ICD reprogramming, catheter ablation performed in specialized centres should be considered.	I	IIa



CARDIOPATÍAS CONGÉNITAS

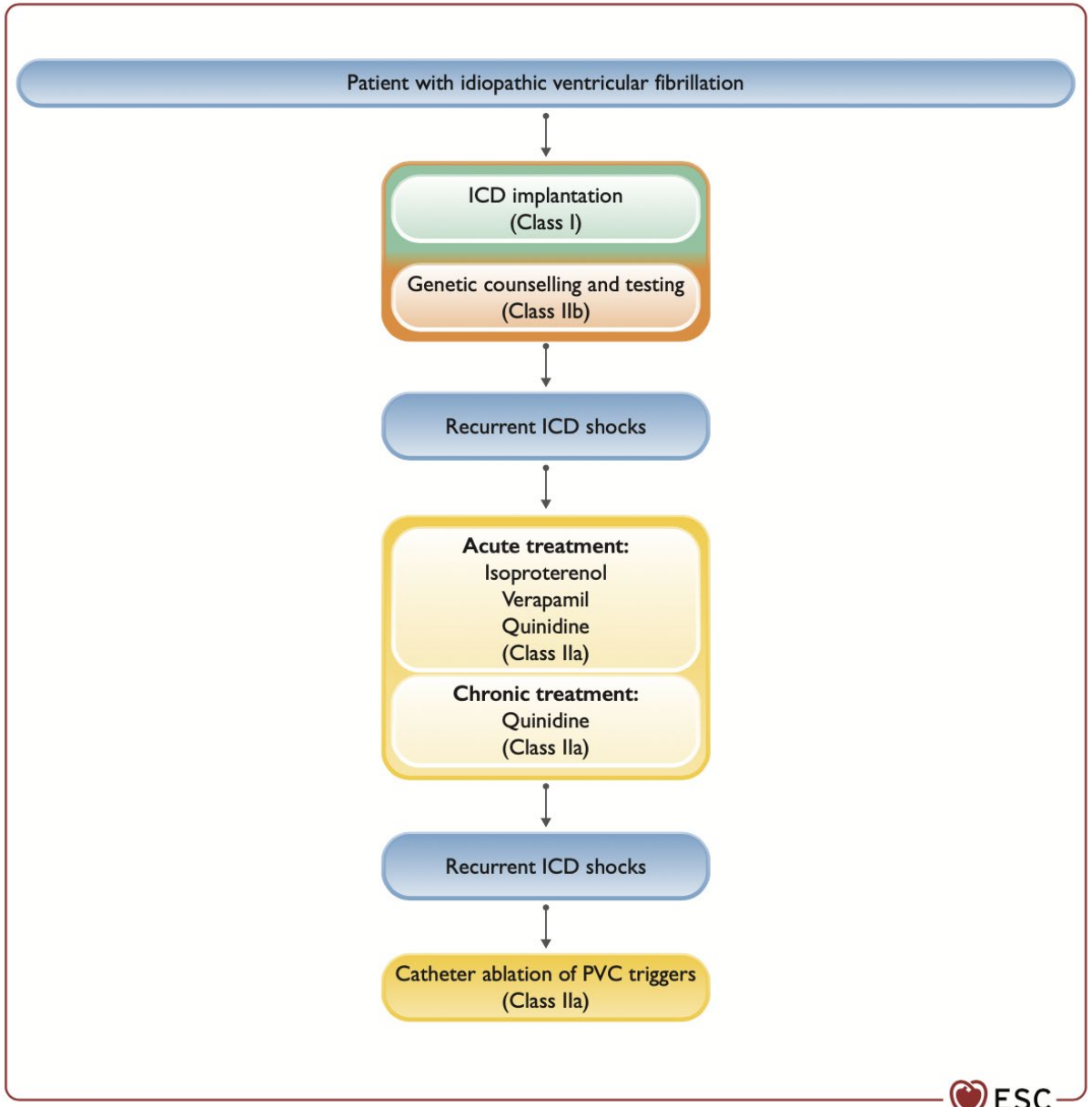
Recommendations	Class[a]	Level[b]	
Risk stratification and primary prevention of SCD			
All CHD patients			
In adults with CHD with biventricular physiology and a left systemic ventricle presenting with symptomatic heart failure (NYHA II/III) and EF ≤35% despite ≥3 months of OMT, ICD	I	C	
In patients with CHD with presumed arrhythmic syncope and with either at least moderate ventricular dysfunction or inducible SMVT on PES, ICD implantation should be considered.[887,889,902]	IIa	C	
In patients with advanced single ventricle or systemic RV dysfunction with additional risk factors,[c] ICD implantation may be considered.[890,891]	IIb	C	
Tetralogy of Fallot			
In patients after repair of TOF with arrhythmia symptoms and NSVT, electrophysiologic evaluation including PES should be considered.[889,903–905]	IIa	C	
In patients after repair of TOF with arrhythmia symptoms and a positive PES, or a combination of other risk factors[d] and a positive PES, ICD implantation should be considered.	IIa	C	
In patients after repair of TOF without arrhythmia symptoms, but with a combination of other risk factors,[d] electrophysiologic evaluation, including PES, may be considered.	IIb	C	
In patients with repaired TOF undergoing surgical or transcutaneous pulmonary valve replacement, pre-operative catheter mapping and transection of VT-related anatomical isthmuses before or during the intervention may be considered.[894]	IIb	C	*NEW*

Recommendations	Class[a]	Level[b]	
Secondary prevention of SCD and treatment of VA			
All CHD patients			
In patients with CHD presenting with sustained VAs, evaluation for residual lesions or new structural abnormalities is recommended.[892,893]	I	B	*NEW*
In patients with CHD with not tolerated VT/ aborted CA due to VF, ICD implantation is indicated after exclusion of reversible causes.[349,350]	I	C	
In patients with CHD and recurrent, symptomatic SMVT or ICD shocks for SMVT not manageable by medical therapy or ICD reprogramming, catheter ablation performed in specialized centres should be considered.[c] [899–901]	IIa	C	
In selected patients with CHD (including atrial baffle repair for transposition of the great arteries, Fontan operation and Ebstein anomaly) presenting with CA, evaluation and treatment of SVT with rapid ventricular conduction should be considered.[890,895]	IIa	C	*NEW*
Tetralogy of Fallot			
In patients with repaired TOF who present with SMVT or recurrent, symptomatic appropriate ICD therapy for SMVT, catheter ablation performed in specialized centres is recommended.[899–901]	I	C	
In patients with repaired TOF with SMVT who are undergoing surgical or transcutaneous pulmonary valve replacement, pre-operative catheter mapping and transection of VT-related anatomical isthmuses before or during the intervention should be considered.[888,893,894]	IIa	C	
In patients with repaired TOF with a preserved biventricular function and symptomatic SMVT, catheter ablation or concomitant surgical ablation performed in specialized centres may be considered as an alternative to ICD therapy.[899,901]	IIb	C	*NEW*

FV IDIOPÁTICA

NEW

Idiopathic VF	
It is recommended that idiopathic VF is diagnosed in a SCA survivor, preferably with documentation of VF, after exclusion of an underlying structural, channelopathic, metabolic, or toxicological aetiology.	I
Isoproterenol infusion, verapamil, or quinidine for acute treatment of an electrical storm or recurrent ICD discharges should be considered in idiopathic VF.	IIa
Quinidine should be considered for chronic therapy to suppress an electrical storm or recurrent ICD discharges in idiopathic VF.	IIa
Clinical testing (history, ECG, and high precordial lead ECG, exercise test, echocardiogram) of first-degree family members of idiopathic VF patients may be considered.	IIb
In idiopathic VF patients, genetic testing of genes related to channelopathy and cardiomyopathy may be considered.	IIb

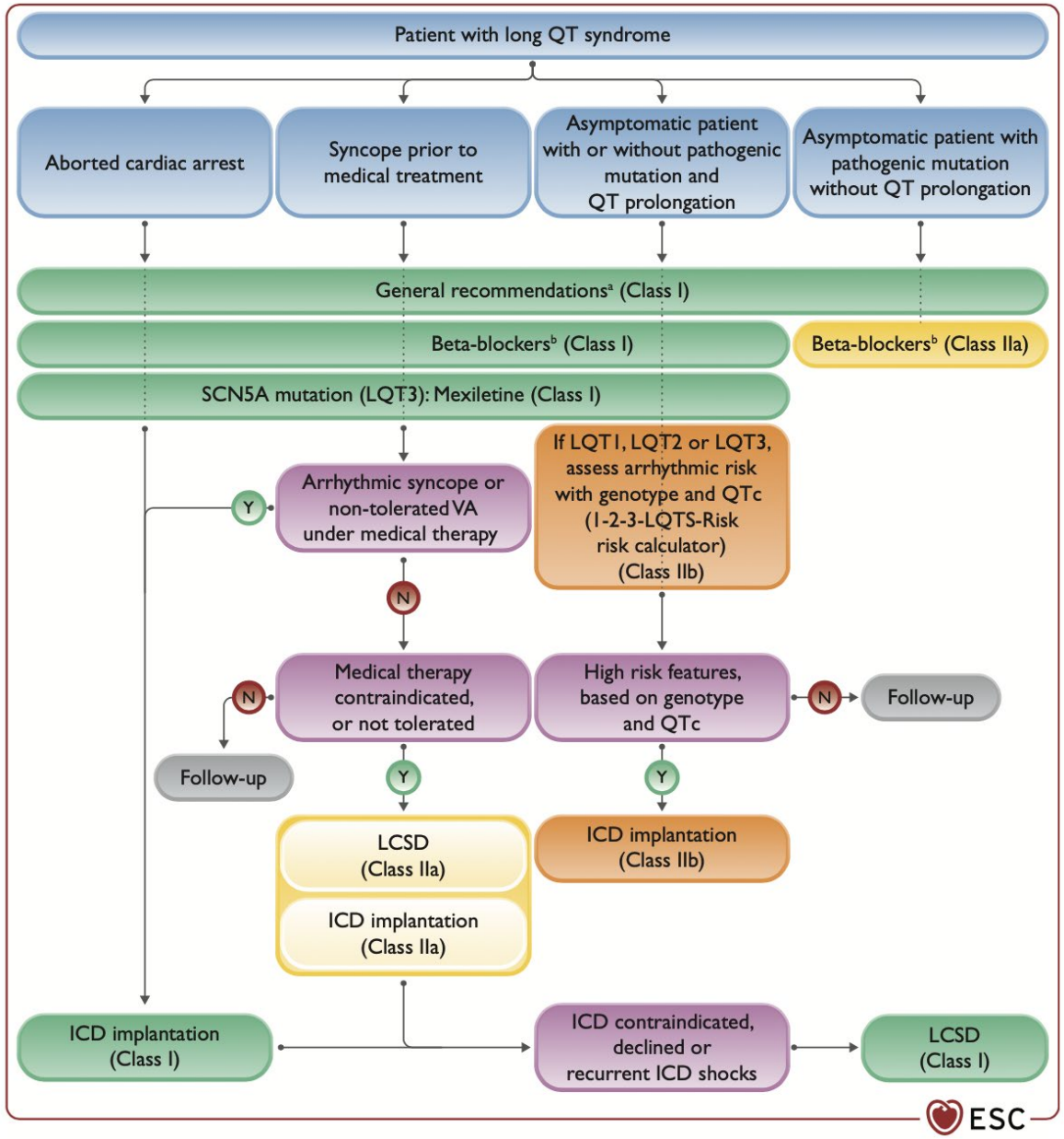


SD. QT LARGO

ICD implantation is recommended in patients with LQTS who are symptomatic[b] while receiving beta-blockers and genotype-specific therapies.	IIa	I

NEW

Long QT syndrome	
In patients with clinically diagnosed LQTS, genetic testing, and genetic counselling are recommended.	I
Beta-blockers, ideally non-selective beta-blockers (nadolol or propranolol), are recommended in LQTS patients with documented QT interval prolongation, to reduce risk of arrhythmic events.	I
Mexiletine is indicated in LQT3 patients with a prolonged QT interval.	I
In LQTS, it should be considered to calculate the arrhythmic risk before initiation of therapy based on the genotype and the duration of QTc interval.	IIa
ICD implantation may be considered in asymptomatic LQTS patients with high-risk profile (according to the 1-2-3 LQTS Risk calculator) in addition to genotype-specific medical therapies (mexiletine in LQT3 patients).	IIb
Routine diagnostic testing with epinephrine challenge is not recommended in LQTS.	III



SD. ANDERSEN-TAWIL TIPO 1: LQT7

Recommendations	Class[a]	Level[b]
Diagnosis		
Genetic testing is recommended in patients with suspected Andersen–Tawil syndrome.[964,967]	I	C
Andersen–Tawil syndrome should be considered in patients without SHD who present with at least two of the following: • Prominent U waves with or without prolongation of the QT interval • Bidirectional and/or polymorphic PVCs/VT • Dysmorphic features • Periodic paralysis • *KCNJ2* pathogenic loss of function mutation.[964,965,967,968,972]	IIa	C
Risk stratification, prevention of SCD and treatment of VA		
ICD implantation is recommended in patients with Andersen–Tawil syndrome after aborted CA or not-tolerated sustained VT.[964,967]	I	C
Beta-blockers and/or flecainide with or without acetazolamide should be considered in patients with Andersen–Tawil syndrome to treat VA.[964,970]	IIa	C
An ILR should be considered in patients with Andersen–Tawil syndrome and unexplained syncope.	IIa	C
ICD implantation may be considered in patients with Andersen–Tawil syndrome who have a history of unexplained syncope or suffer from tolerated sustained VT.[967]	IIb	C

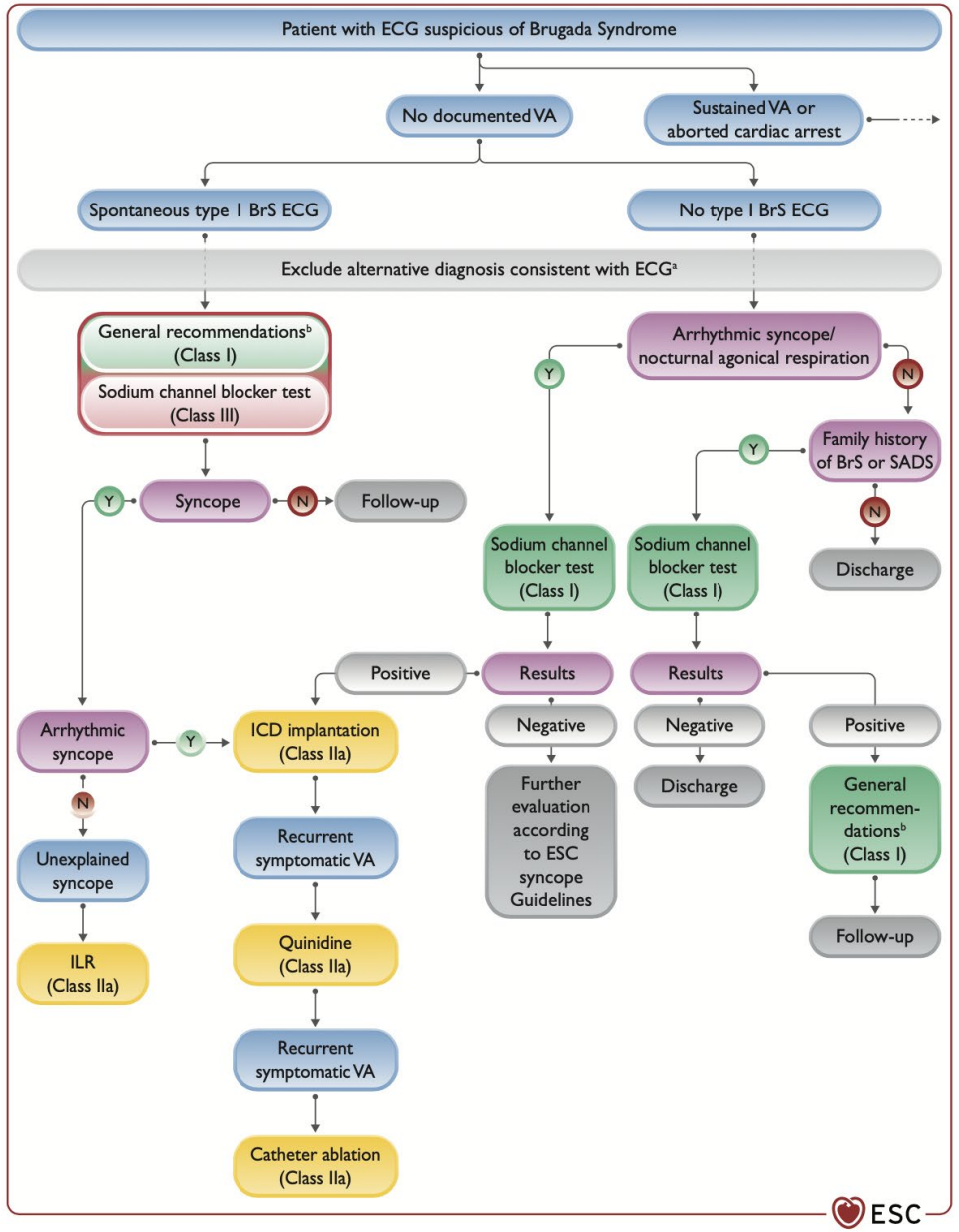
NEW

SD. BRUGADA

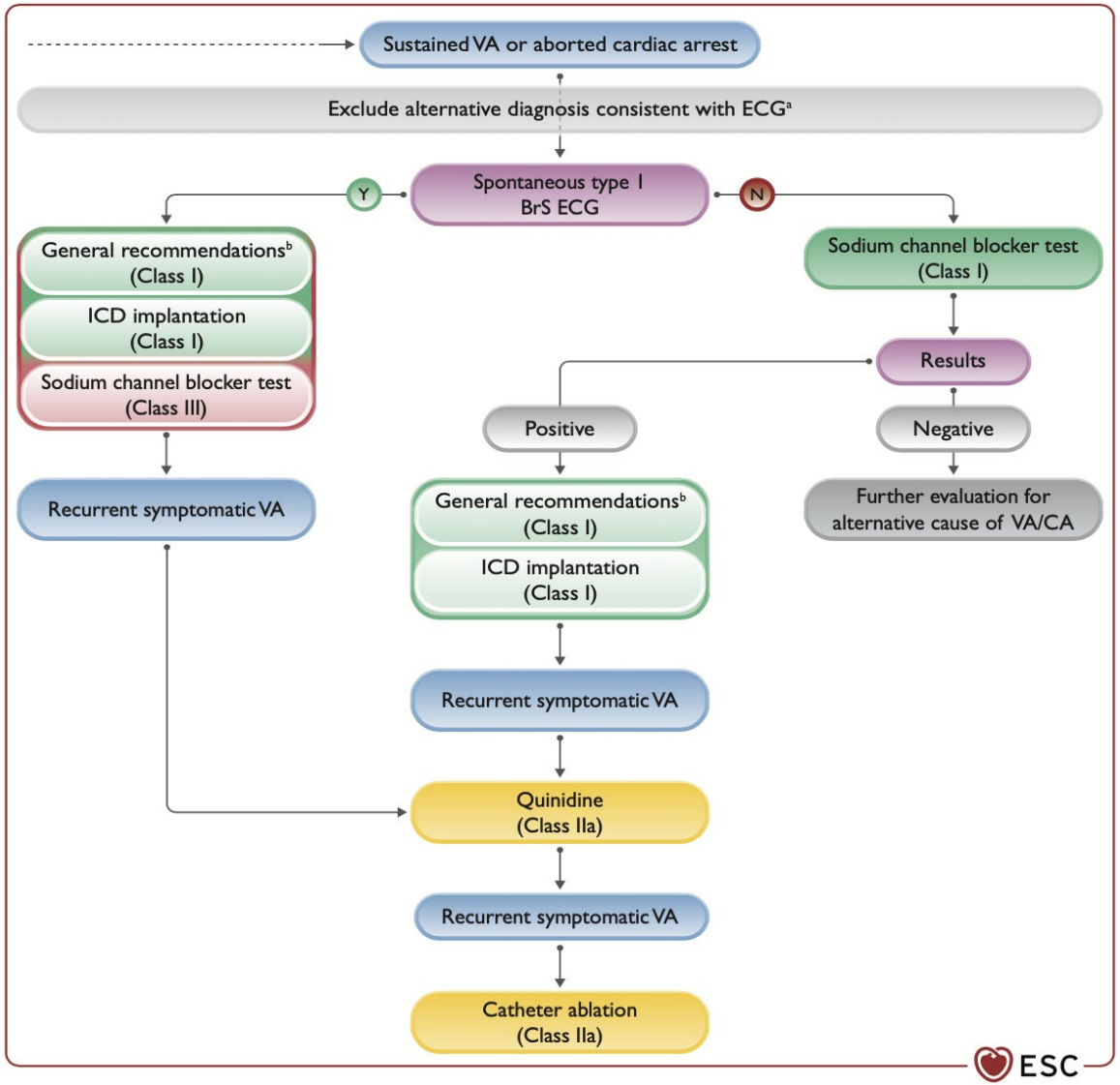
Catheter ablation of triggering PVCs and/or RVOT epicardial substrate should be considered in BrS patients with recurrent appropriate ICD shocks refractory to drug therapy.	IIb	IIa

Brugada syndrome	
Genetic testing for *SCN5A* gene is recommended for probands with BrS.	I
BrS should be considered in patients with no other heart disease and induced type 1 Brugada pattern who have at least one of the following: • Arrhythmic syncope or nocturnal agonal respiration • A family history of BrS • A family history of SD (<45 years old) with a negative autopsy and circumstance suspicious for BrS.	IIa
Implantation of a loop recorder should be considered in BrS patients with an unexplained syncope.	IIa
BrS may be considered as a diagnosis in patients with no other heart disease who exhibit an induced type 1 Brugada ECG.	IIb
PES may be considered in asymptomatic patients with a spontaneous type I BrS ECG.	IIb
Sodium channel blocker test is not recommended in patients with a prior type I Brugada pattern.	III
Catheter ablation in asymptomatic BrS patients is not recommended.	III

NEW



SD. BRUGADA



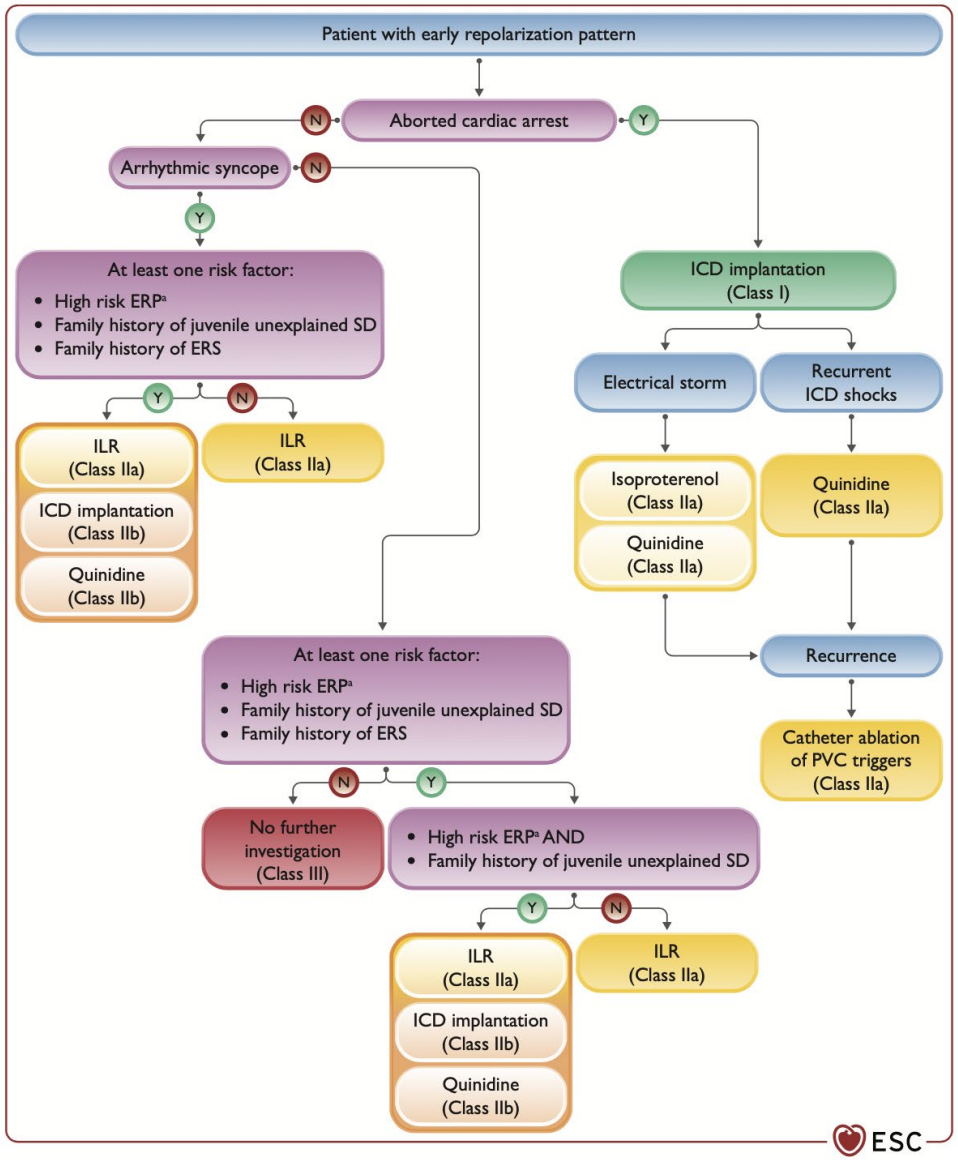
Sodium channel blocker test is not recommended in patients with a prior type I Brugada pattern.	III	C
Catheter ablation in asymptomatic BrS patients is not recommended.	III	C

SD. REPOLARIZACIÓN PRECOZ

Early repolarization syndrome	
It is recommended that the ERP is diagnosed as J-point elevation of ≥1 mm in two adjacent inferior and/or lateral ECG leads.	I
It is recommended that the ERS is diagnosed in a patient resuscitated from unexplained VF/PVT in the presence of ERP.	I
ICD implantation is recommended in patients with a diagnosis of ERS who have survived a CA.	I
In a SCD victim with a negative autopsy and medical chart review, and an ante-mortem ECG demonstrating the ERP, the diagnosis of ERS should be considered.	IIa
First-degree relatives of ERS patients should be considered for clinical evaluation for ERP with additional high-risk features.[j]	IIa
ILR should be considered in individuals with ERP and at least one risk feature[k] or arrhythmic syncope.	IIa
Isoproterenol infusion should be considered for ERS patients with electrical storm.	IIa
Quinidine in addition to an ICD should be considered for recurrent VF in ERS patients.	IIa
PVC ablation should be considered in ERS patients with recurrent VF episodes triggered by a similar PVC non-responsive to medical treatment.	IIa
Genetic testing in ERS patients may be considered.	IIb
ICD implantation or quinidine may be considered in individuals with ERP and arrhythmic syncope and additional risk features.[k]	IIb

NEW

ICD implantation or quinidine may be considered in asymptomatic individuals who demonstrate a high-risk ERP[j] in the presence of a family history of unexplained juvenile SD.	IIb
Clinical evaluation is not recommended routinely in asymptomatic subjects with ERP.	III
ICD implantation is not recommended in asymptomatic patients with an isolated ERP.	III

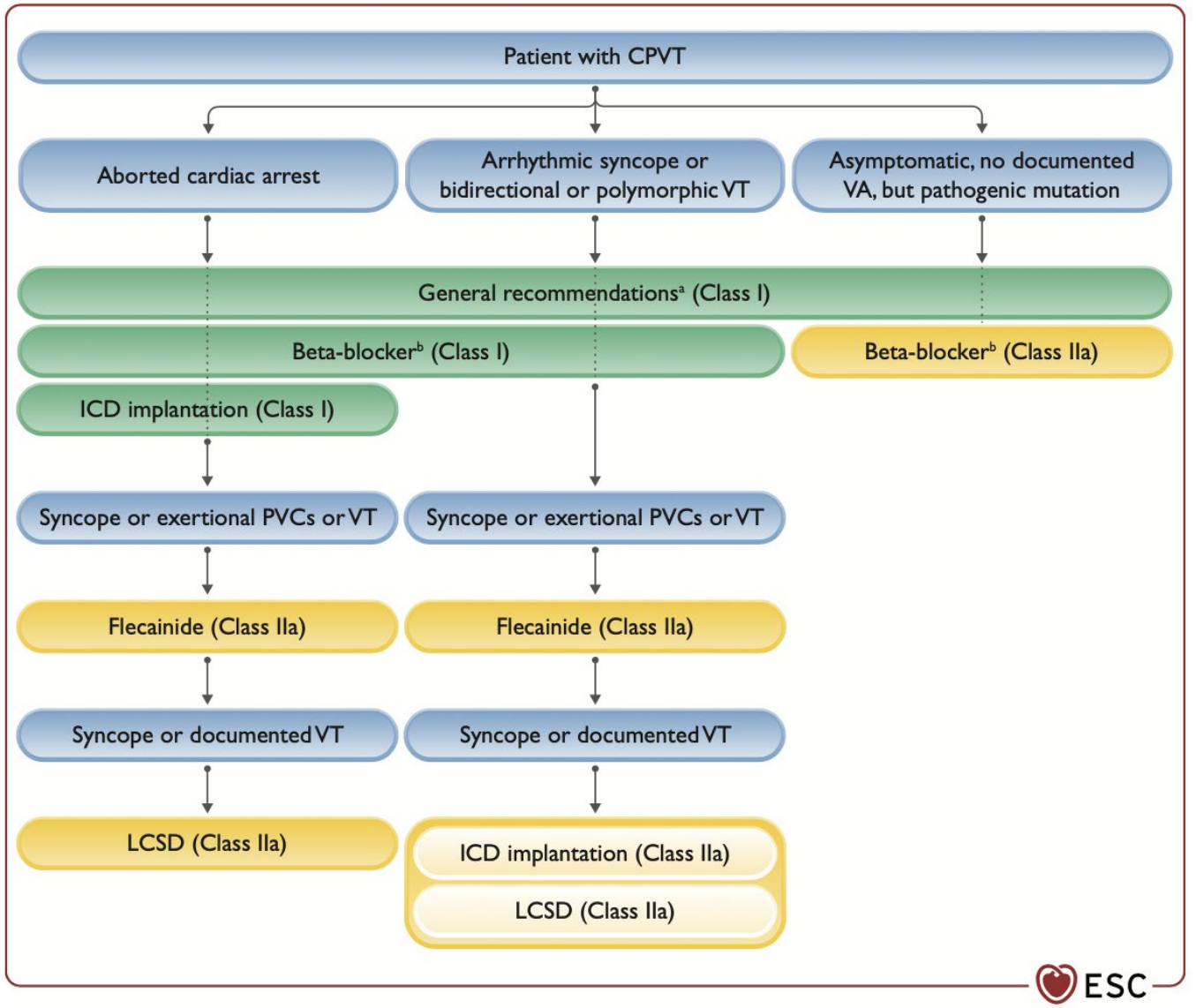


TVPC

ICD implantation should be considered in patients with CPVT who experience arrhythmic syncope and/or documented bidirectional/PVT while on the highest tolerated beta-blocker dose and on flecainide.	I	IIa
LCSD should be considered in patients with diagnosis of CPVT when the combination of beta-blockers and flecainide at therapeutic dosage are either not effective, not tolerated, or contraindicated.	IIb	IIa

NEW

CPVT	
Genetic testing and genetic counselling are indicated in patients with clinical suspicion or clinical diagnosis of CPVT.	I
Beta-blockers, ideally non-selective (nadolol or propranolol) are recommended in all patients with a clinical diagnosis of CPVT.	I
Epinephrine or isoproterenol challenge may be considered for the diagnosis of CPVT when an exercise test is not possible.	IIb



SD QT CORTO

Recommendations	Class[a]	Level[b]	
Diagnosis			
It is recommended that SQTS is diagnosed in the presence of a QTc ≤360 ms and one or more of the following: (a) a pathogenic mutation, (b) a family history of SQTS, (c) survival from a VT/VF episode in the absence of heart disease.[1061,1068]	I	C	
Genetic testing is indicated in patients diagnosed with SQTS.[1063]	I	C	NEW
SQTS should be considered in the presence of a QTc ≤320 ms.[1064–1067,1073,1074]	IIa	C	NEW
SQTS should be considered in the presence of a QTc ≥320 ms and ≤360 ms and arrhythmic syncope.	IIa	C	NEW
SQTS may be considered in the presence of a QTc ≥320 ms and ≤360 ms and a family history of SD at age <40 years.	IIb	C	NEW

Risk stratification, SCD prevention and treatment of VA			
ICD implantation is recommended in patients with a diagnosis of SQTS who: (a) are survivors of an aborted CA and/or (b) have documented spontaneous sustained VT.[1063]	I	C	
ILR should be considered in young SQTS patients.	IIa	C	NEW
ICD implantation should be considered in SQTS patients with arrhythmic syncope.	IIa	C	NEW
Quinidine may be considered in (a) SQTS patients who qualify for an ICD but present a contraindication to the ICD or refuse it, and (b) asymptomatic SQTS patients and a family history of SCD.[1069–1071]	IIb	C	NEW
Isoproterenol may be considered in SQTS patients with an electrical storm.[1075]	IIb	C	NEW
PES is not recommended for SCD risk stratification in SQTS patients.	III	C	

EMBARAZO

Recommendations	Class[a]	Level[b]	
Acute management of VA			
During pregnancy, electrical cardioversion is recommended for sustained VT.[1084]	I	C	
For acute conversion of haemodynamically tolerated SMVT during pregnancy, a beta-blocker, sotalol, flecainide, procainamide, or overdrive ventricular pacing should be considered.	IIa	C	
Long-term management of VA			
If ICD implantation is indicated during pregnancy, implantation is recommended with optimal radiation protection.[1087,1107]	I	C	
Continuation of beta-blockers is recommended during pregnancy and post-partum in women with LQTS or CPVT.[955,1094–1096]	I	C	
Continuation of beta-blockers should be considered during pregnancy in women with ARVC.[1097–1099]	IIa	C	*NEW*
Oral metoprolol, propranolol, or verapamil should be considered for long-term management of idiopathic sustained VT during pregnancy.	IIa	C	*NEW*
Catheter ablation using non-fluoroscopic mapping systems should be considered, preferably after the first trimester, in women with highly symptomatic recurrent SMVT refractory or who are intolerant to AADs.[1105]	IIa	C	*NEW*

DEPORTISTAS

Pre-participation cardiovascular evaluation of competitive athletes should be considered.	I	IIa

Recommendations	Class[a]	Level[b]
In athletes with positive medical history, abnormal physical examination, or ECG alterations, further investigations including echocardiography and/or CMR to confirm (or exclude) an underlying disease are recommended.[1123,1133,1135]	I	C
It is recommended that athletes diagnosed with a cardiovascular disease associated with SCD are managed according to current guidelines for sports eligibility.	I	C
It is recommended that staff at sporting facilities are trained in CPR and in the use of AED.[93,1137]	I	C
Pre-participation cardiovascular evaluation of competitive athletes should be considered.[46,1122,1123,1127]	IIa	C
It should be considered that cardiovascular evaluation of young (<35 years) competitive athletes includes history, physical examination, and 12-lead ECG.[1123,1126,1130,1140]	IIa	C
The cardiovascular risk of middle-aged and elderly individuals should be evaluated before engaging in strenuous sports through established scores such as the SCORE2 risk chart.[46,1141,1142]	IIa	C

NEW

MENSAJES CLAVE GENERALES

- IMPLICACIÓN DE LAS AUTORIDADES PÚBLICAS
- ESTUDIO FAMILIAR Y GENÉTICO + RMN (EQUIPOS MULTIDISCIPLINARES)
- AUTOPSIA EN MS < 50 AÑOS
- CARDIOVERSIÓN PRIMERA ELECCIÓN Y PROCAINAMIDA PRIMER FÁRMACO DE ELECCIÓN
- TORMENTA ARRÍTMICA REFRACTARIA REQUIERE EQUIPO CAPAZ DE ABLACIÓN AVANZADA, SOPORTE CIRCULATORIO MECÁNICO Y MODULACIÓN AUTONÓMICA
- ABLACIÓN EN CASO DE TVMS REFRACTARIA A AMIODARONA EN PACIENTE ISQUÉMICO
- ABLACIÓN PRIMERA ELECCIÓN EN CARDIOMIOPATÍA INDUCIDA POR EXTRASISTOLIA
- EN MCD CONSIDERAR CRM Y GENÉTICA ADEMÁS DE FEVI PARA EL DAI
- CONSIDERAR OPTIMIZACIÓN, SUPERVIVENCIA Y CALIDAD DE VIDA ANTES DE IMPLANTAR DAI
- ESCENARIOS ESPECÍFICOS

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