Corso di Aggiornamento AINV2024

La disautonomia nella pratica clinica: diagnosi e strategie terapeutiche

TREIA- MC

Aula Didattica Multimediale

Comune di Treia

4 ottobre 2024

Treia (MC)

Le aritmie e la sincope cardiogena

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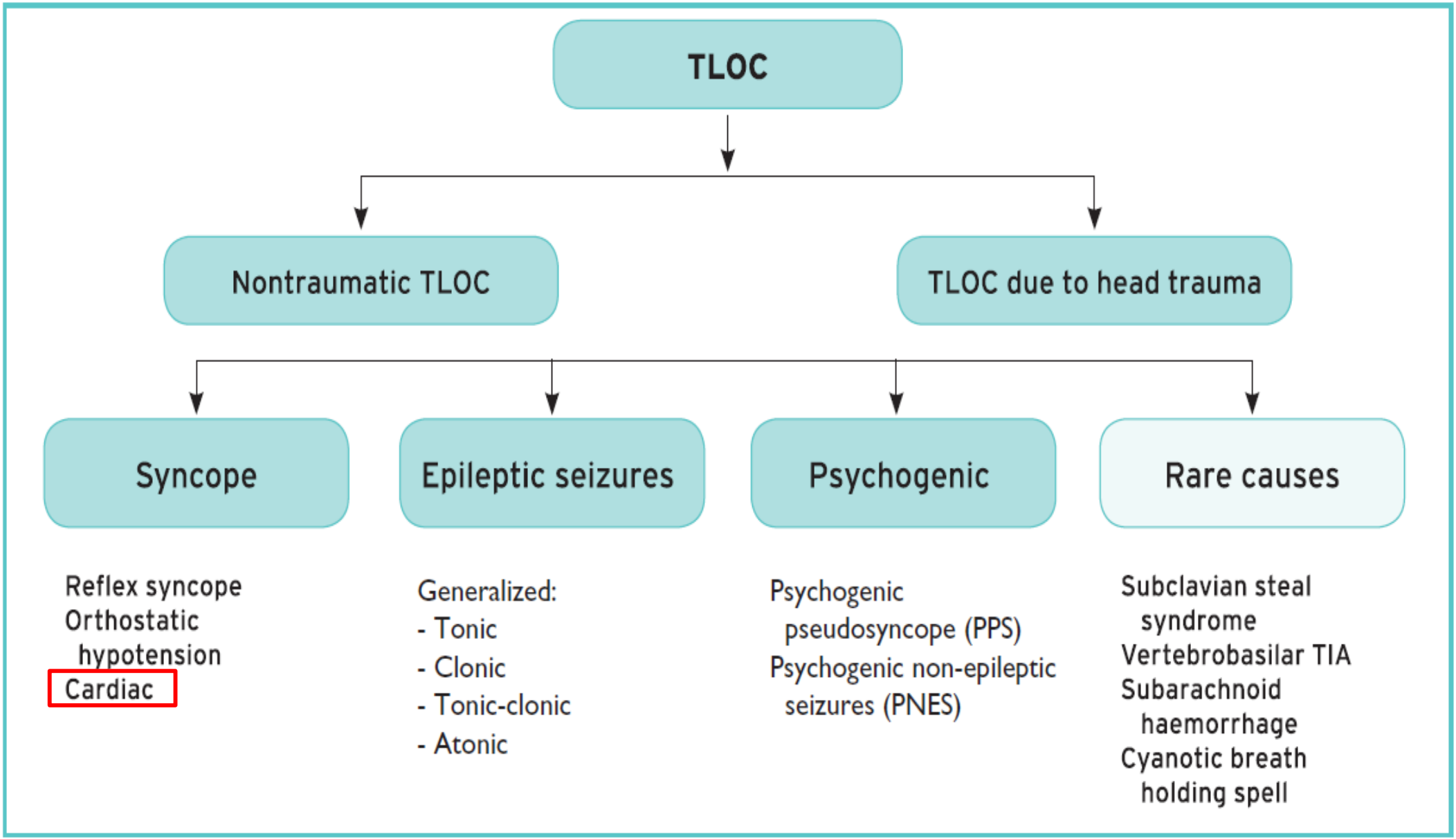
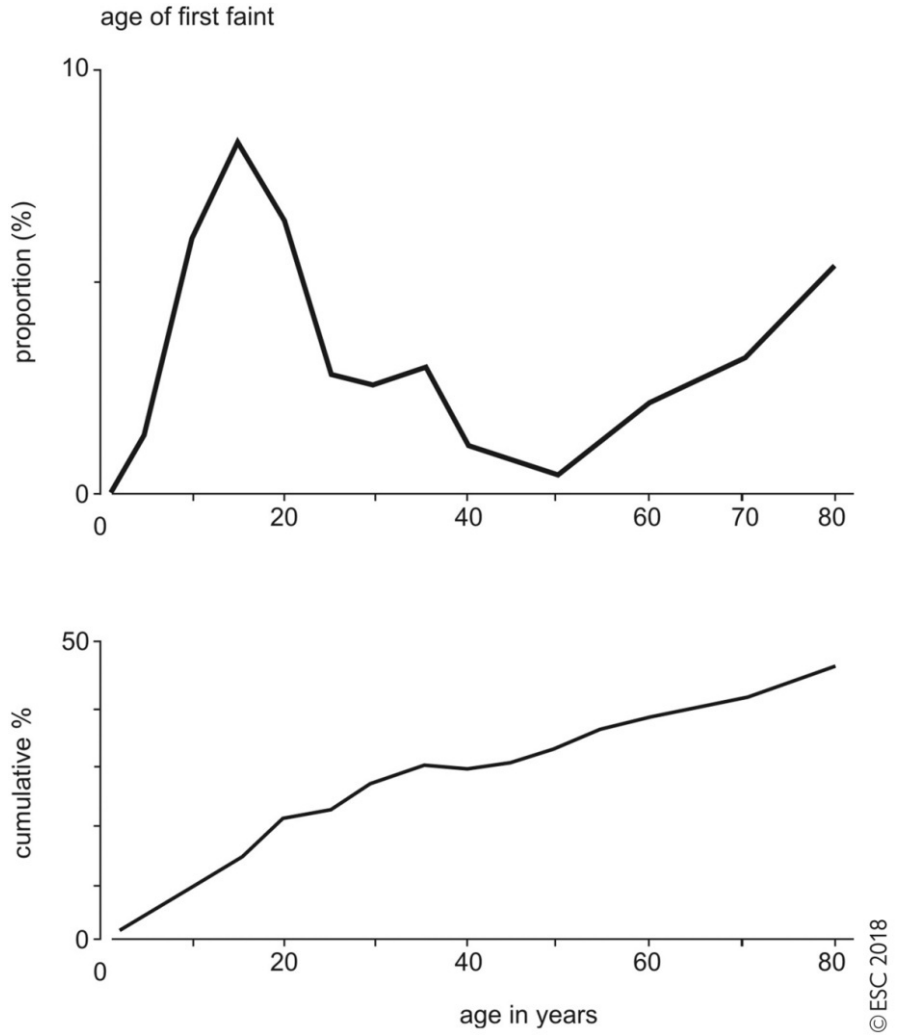
SINCOPE: DEFINIZIONE

- Transitoria perdita di coscienza dovuta ad ipoperfusione cerebrale a rapida insorgenza, di breve durata a risoluzione spontanea e completa.


TLOC
Nontraumatic TLOC
TLOC due to head trauma
Syncope
Epileptic seizures
Psychogenic
Rare causes
Reflex syncope
Orthostatic hypotension
Cardiac
Generalized:
- Tonic
- Clonic
- Tonic-clonic
- Atonic
Psychogenic pseudosyncope (PPS)
Psychogenic non-epileptic seizures (PNES)
Subclavian steal syndrome
Vertebrobasilar TIA
Subarachnoid haemorrhage
Cyanotic breath holding spell

Distribution of age and cumulative incidence of first episodes of syncope in the general population



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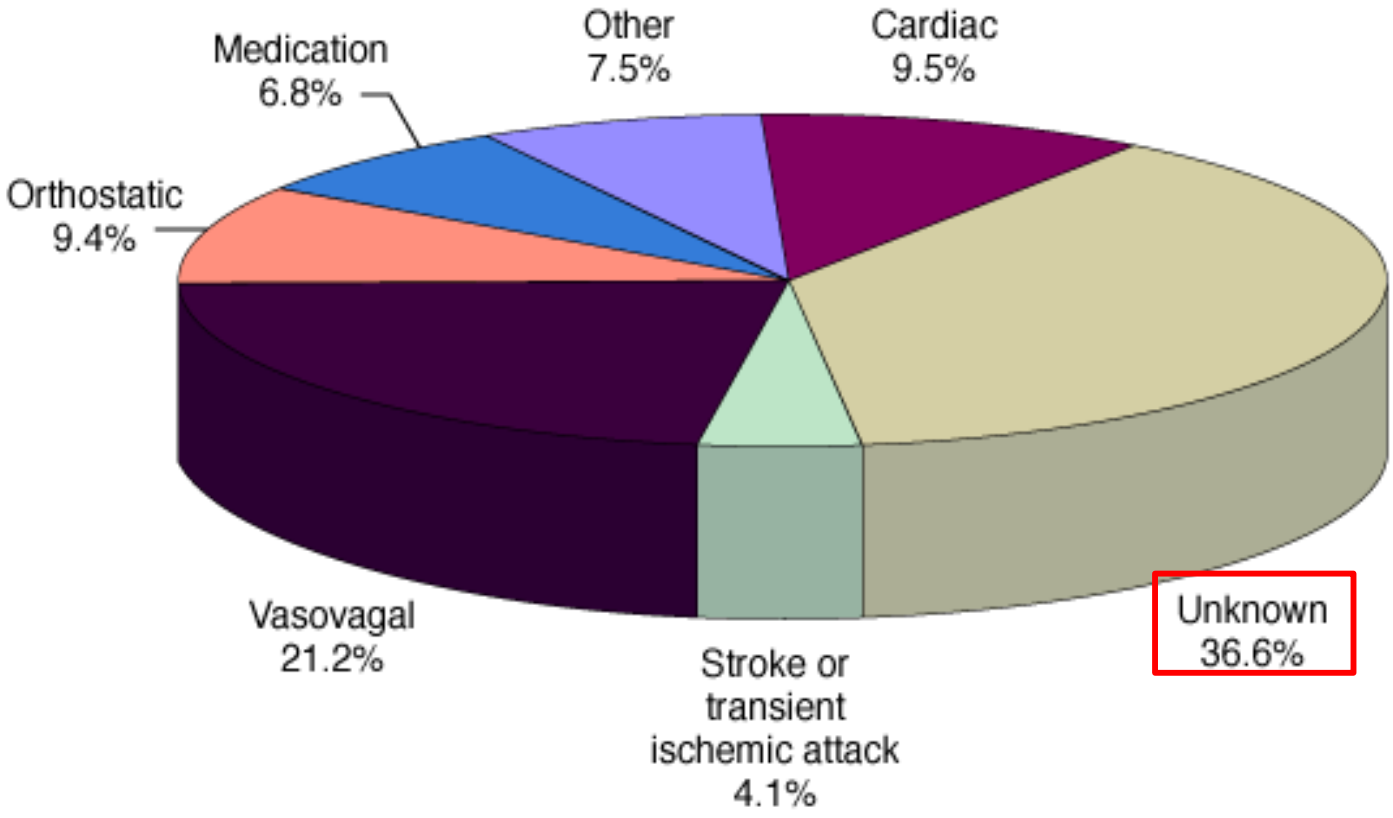
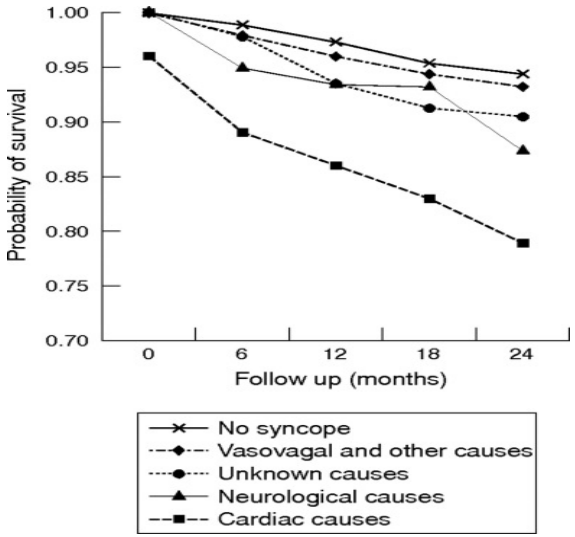
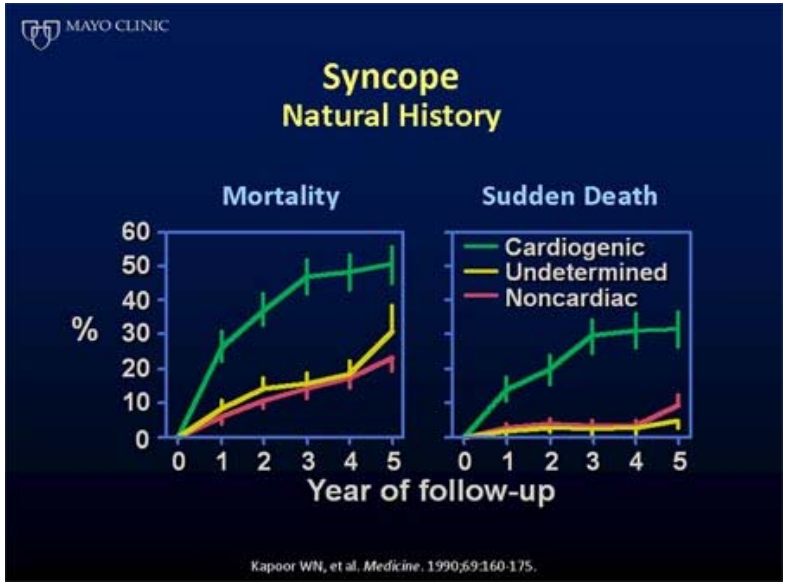
Causes of Syncope



Morbidity and Mortality

- Most cases benign.
- **Syncope of cardiac origin** has the highest morbidity and mortality.
  - **1 year mortality of 18-33%**
- Recurrence in the elderly population is 30%
- Syncope of unknown origin.
  - 1 year mortality of 6-12%.





SINCOPE CARDIACA

Arrhythmia as primary cause:

Bradycardia:

- sinus node dysfunction (including bradycardia/tachycardia syndrome)
- atrioventricular conduction system disease

Tachycardia:

- supraventricular
- ventricular

Structural cardiac:

aortic stenosis,
acute myocardial infarction/ischaemia,
hypertrophic cardiomyopathy,
cardiac masses (atrial myxoma, tumours, etc.),
pericardial disease/tamponade,
congenital anomalies of coronary arteries,
prosthetic valve dysfunction

Cardiopulmonary and great vessels:

pulmonary embolus,
acute aortic dissection,
pulmonary hypertension

ESC
European Society of Cardiology
European Heart Journal (2022) **00**, 1–130
https://doi.org/10.1093/eurheartj/ehac262

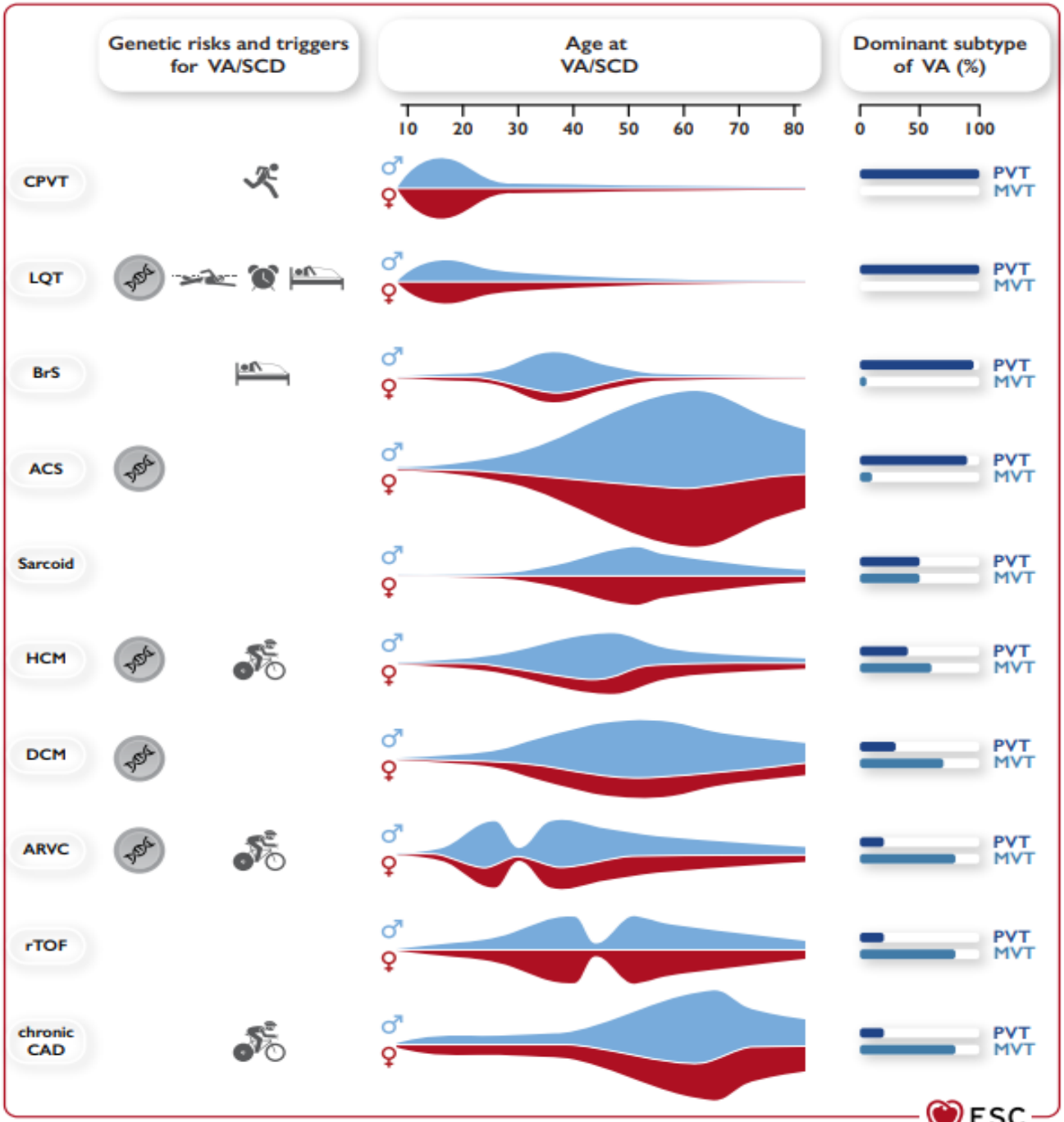
ESC GUIDELINES

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)

Genetic risk for VA/SCD, typical triggers for VA/SCD, age at presentation with VA/SCD, sex predominance, and typical VA (PVT/VF vs. MVT) in different diseases associated with VA/SCD



ANAMNESI

low-risk features (that suggest a benign condition) in patients with syncope at initial evaluation

Low-risk
• Associated with prodrome typical of reflex syncope (e.g. light-headedness, feeling of warmth, sweating, nausea, vomiting)[36,49] • After sudden unexpected unpleasant sight, sound, smell, or pain[36,49,50] • After prolonged standing or crowded, hot places[36] • During a meal or postprandial[51] • Triggered by cough, defaecation, or micturition[52] • With head rotation or pressure on carotid sinus (e.g. tumour, shaving, tight collars)[53] • Standing from supine/sitting position[54]
PAST MEDICAL HISTORY
Low-risk
• Long history (years) of recurrent syncope with low-risk features with the same characteristics of the current episode[58] • Absence of structural heart disease[27, 58]
PHYSICAL EXAMINATION
Low-risk
• Normal examination

High-risk features (that suggest a serious condition) inpatients with syncope at initial evaluation

High-risk
Major
• New onset of chest discomfort, breathlessness, abdominal pain, or headache[26, 44, 55] • Syncope during exertion or when supine[36] • Sudden onset palpitation immediately followed by syncope[36]
Minor (high-risk only if associated with structural heart disease or abnormal ECG):
• No warning symptoms or short (<10 s) prodrome[36, 38, 49, 56] • Family history of SCD at young age[57] • Syncope in the sitting position[54]
PAST MEDICAL HISTORY
High-risk
Major
• Severe structural or coronary artery disease (heart failure, low LVEF or previous myocardial infarction)[26, 27, 35, 55, 59]

ECG

Low-risk	
• Normal ECG[26, 35, 36, 55]	
High-risk	
Major	**Minor** (high-risk only if history consistent with arrhythmic syncope)
• ECG changes consistent with acute ischaemia • Mobitz II second- and third-degree AV block • Slow AF (<40 b.p.m.) • Persistent sinus bradycardia (<40 b.p.m.), or repetitive sinoatrial block or sinus pauses >3 seconds in awake state and in absence of physical training • Bundle branch block, intraventricular conduction disturbance, ventricular hypertrophy, or Q waves consistent with ischaemic heart disease or cardiomyopathy[44, 56] • Sustained and non-sustained VT • Dysfunction of an implantable cardiac device (pacemaker or ICD) • Type 1 Brugada pattern • ST-segment elevation with type 1 morphology in leads V1–V3 (Brugada pattern) • QTc >460 ms in repeated 12-lead ECGs indicating LQTS[46]	• Mobitz I second-degree AV block and 1°degree AV block with markedly prolonged PR interval • Asymptomatic inappropriate mild sinus bradycardia (40–50 b.p.m.), or slow AF (40–50 b.p.m.)[56] • Paroxysmal SVT or atrial fibrillation[50] • Pre-excited QRS complex • Short QTc interval (≤340 ms)[46] • Atypical Brugada patterns[46] • Negative T waves in right precordial leads, epsilon waves suggestive of ARVC[46]

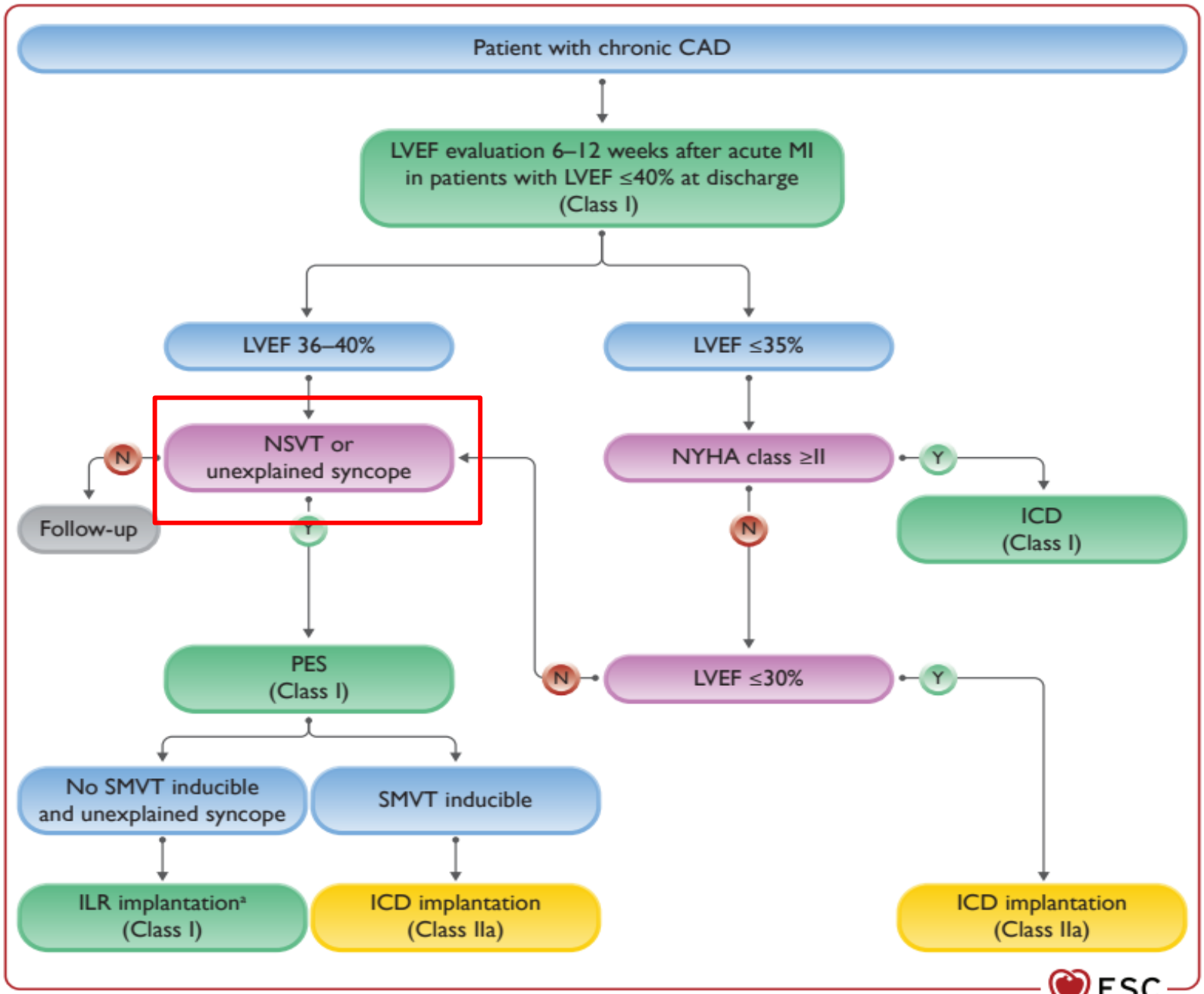
Electrophysiological study

Recommendations	Class[a]	Level[b]
Indications		
In patients with syncope and previous myocardial infarction, or other scar-related conditions, EPS is indicated when syncope remains unexplained after non-invasive evaluation.[218]	I	B
In patients with syncope and bifascicular BBB, EPS should be considered when syncope remains unexplained after non-invasive evaluation.[188,214–217,221]	IIa	B
In patients with syncope and asymptomatic sinus bradycardia, EPS may be considered in a few instances when non-invasive tests (e.g. ECG monitoring) have failed to show a correlation between syncope and bradycardia.[210–212]	IIb	B
In patients with syncope preceded by sudden and brief palpitations, EPS may be considered when syncope remains unexplained after non-invasive evaluation.	IIb	C
EPS-guided therapy		
In patients with unexplained syncope and bifascicular BBB, a pacemaker is indicated in the presence of either a baseline H-V interval of ≥70 ms, second- or third-degree His-Purkinje block during incremental atrial pacing, or with pharmacological challenge.[188,214–217,221]	I	B
In patients with unexplained syncope and previous myocardial infarction, or other scar-related conditions, it is recommended that induction of sustained monomorphic VT is managed according to the current ESC Guidelines for VA.[46]	I	B
In patients without structural heart disease with syncope preceded by sudden and brief palpitations, it is recommended that the induction of rapid SVT or VT, which reproduce hypotensive or spontaneous symptoms, is managed with appropriate therapy according to the current ESC Guidelines.[46,222]	I	C
In patients with syncope and asymptomatic sinus bradycardia, a pacemaker should be considered if a prolonged corrected SNRT is present.[210–212]	IIa	B

Additional advice and clinical perspectives

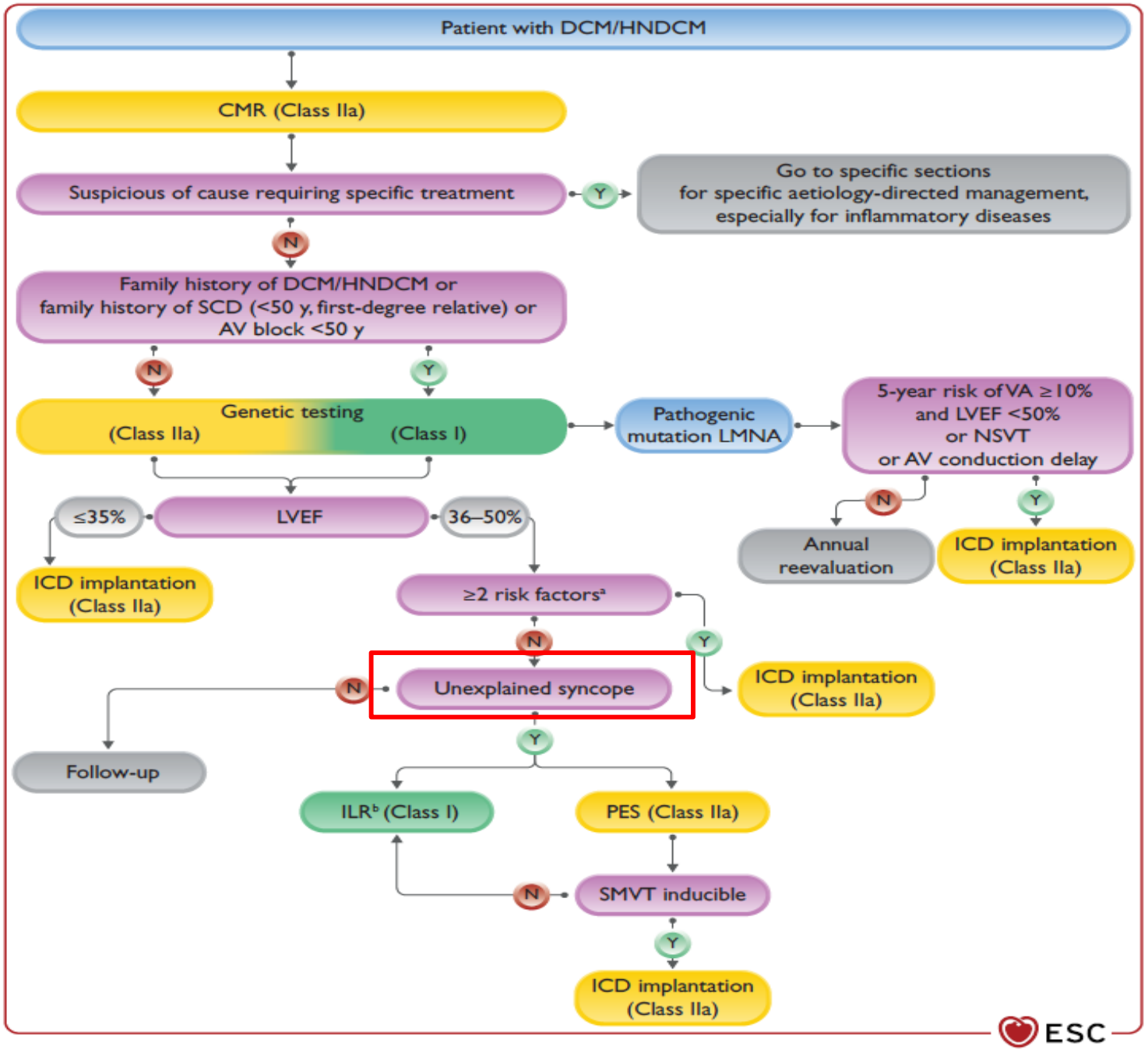
- In general, whereas a positive EPS predicts the cause of syncope, a negative study is unable to exclude an arrhythmic syncope and further evaluation is warranted.
- The induction of polymorphic VT or VF in patients with ischaemic cardiomyopathy or DCM cannot be considered a diagnostic finding of the cause of syncope.
- EPS is generally not useful in patients with syncope, normal ECG, no heart disease, and no palpitations.

Algorithm for risk stratification and primary prevention of sudden cardiac death in patients with chronic coronary artery disease and reduced ejection fraction



ESC 2022

Algorithm for risk stratification and primary prevention of sudden cardiac death in patients with dilated cardiomyopathy/hypokinetic nondilated cardiomyopathy



CARDIOMIOPATIA IPERTROFICA

HCM Risk-SCD Calculator

Age	**Years**	*Age at evaluation*
Maximum LV wall thick-ness	**mm**	*Transthoracic Echocardiographic measurement*
Left atrial size	**mm**	*Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation*
Max LVOT gradient	**mmHg**	*The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernouilli equation: Gradient= $4V^2$, where V is the peak aortic outflow velocity*
Family History of SCD	○ No ○ Yes	*History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).*
Non-sustained VT	○ No ○ Yes	*3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.*
Unexplained syncope	○ No ○ Yes	*History of unexplained syncope at or prior to evaluation.*

Risk of SCD at 5 years (%):	
ESC recommendation:	

HCM Risk-SCD should not be used in:

- ***Paediatric patients (<16 years)***
- ***Elite/competitive athletes***
- ***HCM associated with metabolic diseases (e.g. Anderson-Fabry disease), and syndromes (e.g. Noonan syndrome).***
- ***Patients with a previous history of aborted SCD or sustained ventricular arrhythmia who should be treated with an ICD for secondary prevention.***

Caution should be exercised when assessing the SCD in patients following invasive reduction in left ventricular outflow tract obstruction with myectomy or alcohol septal ablation.

Pending further studies, HCM-RISK should be used cautiously in patients with a maximum left ventricular wall thickness ≥35 mm.

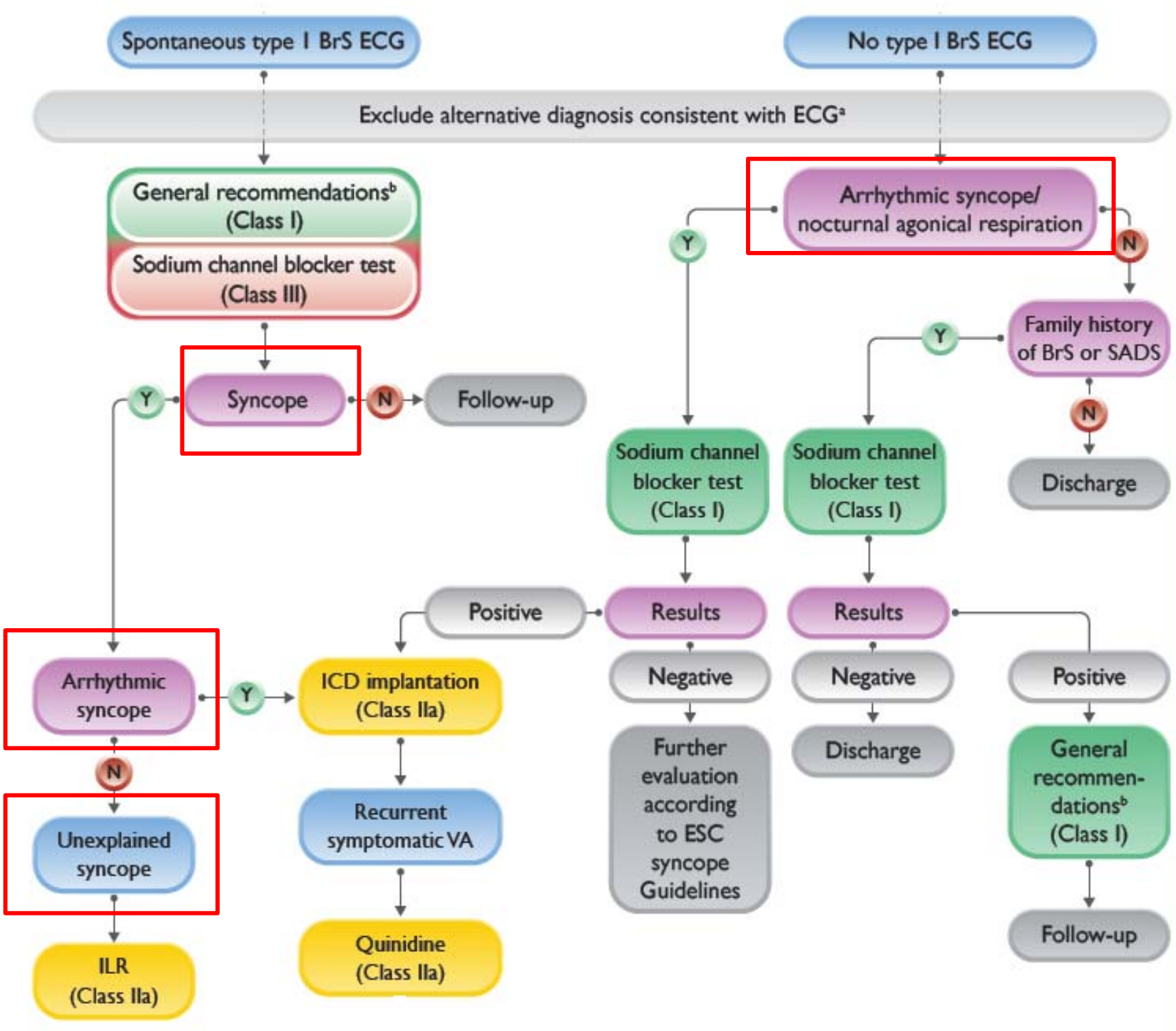
CARDIOMIOPATIA IPERTROFICA

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
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
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

Risk stratification and primary prevention of SCD in arrhythmogenic right ventricular cardiomyopathy

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
In patients with ARVC and symptoms highly suspicious for VA, PES may be considered for risk stratification.[695,705]	IIb	C

Algorithm for the management of patients with Brugada pattern electrocardiogram



Risk stratification, prevention of SCD and treatment of VA

ICD implantation is recommended in patients with BrS who: (a) Are survivors of an aborted CA and/or (b) Have documented spontaneous sustained VT.[980,990–992]	I	C
ICD implantation should be considered in patients with type 1 Brugada pattern and an arrhythmic syncope.[990,992,996]	IIa	C
Implantation of a loop recorder should be considered in BrS patients with an unexplained syncope.[997,999]	IIa	C
Quinidine should be considered in patients with BrS who qualify for an ICD but have a contraindication, decline, or have recurrent ICD shocks.[922,1006,1007]	IIa	C
Isoproterenol infusion should be considered in BrS patients suffering electrical storm.[1008]	IIa	C
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.[1010–1015]	IIa	C
PES may be considered in asymptomatic patients with a spontaneous type I BrS ECG.[155]	IIb	B
ICD implantation may be considered in selected asymptomatic BrS patients with inducible VF during PES using up to 2 extra stimuli.[155]	IIb	C
Catheter ablation in asymptomatic BrS patients is not recommended.	III	C

CONCLUSIONI

La sincope associata ad una malattia cardiaca (comprese le canalopatie) aumenta il rischio di mortalità totale e morte cardiaca improvvisa

Nei pazienti con patologie cardiache a rischio di SCD, è importante definire la natura della sincope (neuromediata o aritmica) in conseguenza del successivo iter terapeutico

Non sempre è possibile determinare la vera causa della sincope (unexplained syncope)
In questa difficile situazione, sarà importante decidere come trattare il paziente:

- attenta valutazione delle circostanze della sincope
- una valutazione completa del paziente che includa una valutazione sia cardiaca che non cardiaca
- in caso di dubbio una discussione chiara con il paziente che spiega sia il rischio della sincope in termini di SCD ma anche il rischio del trattamento se viene proposto un impianto di ICD.