2018 ESC Guidelines for the diagnosis and management of syncope

2018 ESC Guidelines on Syncope – Michele Brignole & Angel Moya
European Heart Journal 2018;39:1883–1948
Doi:10.1093/eurheartj/ehy037

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2018 ESC Guidelines for the diagnosis and management of syncope

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The Task Force for the diagnosis and management of syncope of the European Society of Cardiology (ESC).

Developed with a special contribution of European Heart Rhythm Association (EHRA).

Endorsed by the following societies:

European Society of Emergency Medicine (EuSEM).
European Federation of Internal Medicine (EFIM).
European Union Geriatric Medicine Society (EUGMS).
European Neurological Society (ENS).
European Federation of Autonomic Societies (EFAS).

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New 2018

Web Practical Instructions

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- ESC checklists of historical clues
- Instruction on how to perform and interpret tests
- Explanatory videos, ECG tracings and figures (total 42)
- ESC information sheets for patients affected by reflex syncope and for patients affected by psychogenic pseudosyncope
- Advice for driving and working

"We have the knowledge, we need to teach it"

Classes of recommendations

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Classes of recommendations	Definition	Suggested wording to use
Class I	Evidence and/or general agreement that a given treatment or procedure is beneficial, useful, effective.	Is recommended/ is indicated.
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 favour of usefulness/efficacy.*	Should be considered.
Class IIb	*Usefulness/efficacy is less well established by evidence/opinion.*	May be considered.
Class III	Evidence or general agreement that the given treatment or procedure is not useful/effective, and in some cases may be harmful.	Is not recommended.

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Level of evidence

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Level of evidence A	Data derived from multiple randomized clinical trials or meta-analyses.
Level of evidence B	Data derived from a single randomized clinical trial or large non-randomized studies.
Level of evidence C	Consensus of opinion of the experts and/or small studies, retrospective studies, registries.

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NEW / REVISED CLINICAL SETTINGS AND TESTS:

- Tilt testing: concepts of *hypotensive susceptibility*
- Increased role of prolonged ECG monitoring
- Video recording in suspected syncope
- "Syncope without prodrome, normal ECG and normal heart" (adenosine sensitive syncope)
- Neurological causes: "ictal asystole"

(OUT-PATIENT) SYNCOPE MANAGEMENT UNIT:

- Structure: staff, equipment, and procedures
- Tests and assessments
- Access and referrals
- Role of the Clinical Nurse Specialist
- Outcome and quality indicators

2018 NEW/REVISED CONCEPTS in management of syncope

NEW / REVISED INDICATIONS FOR TREATMENT:

- *Reflex syncope:* algorithms for selection of appropriate therapy based on age, severity of syncope and clinical forms
- *Reflex syncope:* algorithms for selection of best candidates for pacemaker therapy
- *Patients at risk of SCD:* definition of unexplained syncope and indication for ICD
- *Implantable loop recorder* as alternative to ICD, in selected cases

MANAGEMENT IN EMERGENCY DEPARTMENT:

- List of low-risk and high-risk features
- Risk stratification flowchart
- Management in *ED Observation Unit* and/or fast-track to *Syncope Unit*
- Restricted admission criteria
- Limited usefulness of risk stratification scores

What is new in 2018 syncope guidelines ? (1)

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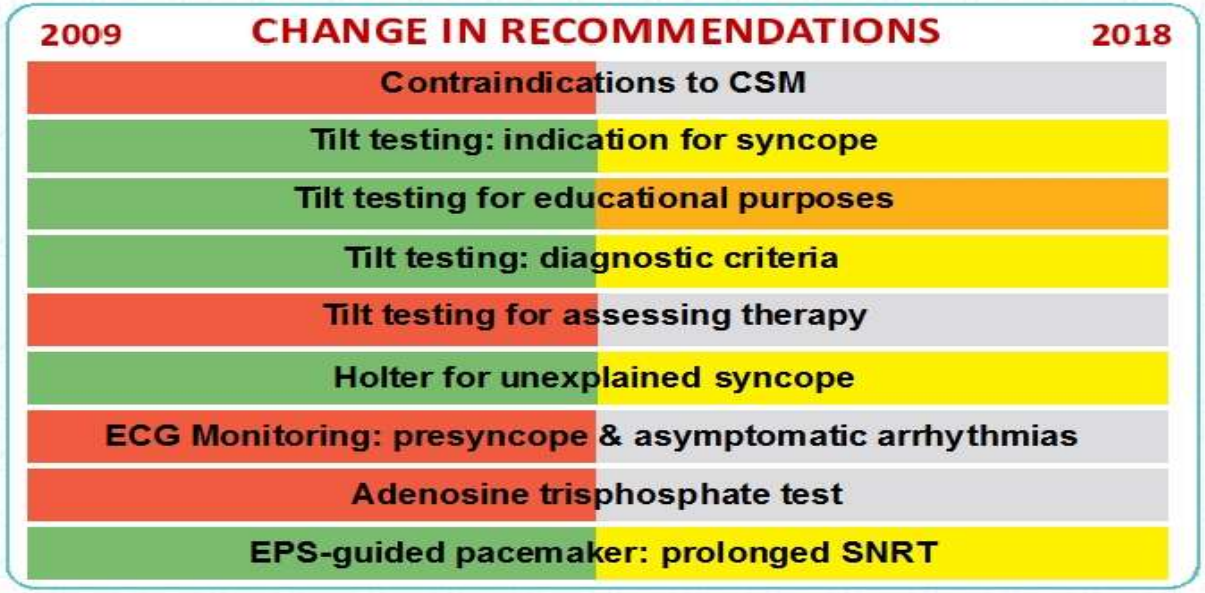

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What is new in 2018 syncope guidelines ? (2)

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CHANGE IN RECOMMENDATIONS

2009	2018
EPS-guided pacemaker: HV >70 ms	
Empiric pacing in bifascicular block	
Therapy of reflex syncope: PCM	
Therapy of OH: PCM	
Therapy of OH: abdominal binders	
Therapy of OH: head-up tilt sleeping	
Syncope & SVT/VT: AA drugs Expert opinion	

CHANGE IN RECOMMENDATIONS

2009	2018
Syncope & AF: catheter ablation Expert opinion	
ICD: LVEF >35% and syncope	
Syncope & high risk HCM: ICD	
Syncope & ARVC: ICD	
Psychiatric consultation for PPS Expert opinion	

I | IIa | IIb | III | Taken out

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What is new in 2018 syncope guidelines ? (3)

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2018 NEW RECOMMENDATIONS (only major included)

Management of syncope in ED (section 4.1.2)

- *Low-risk*: discharge from ED
- *High-risk*: early intensive evaluation in ED, SU versus admission
- *Neither high or low*: observation in ED or in SU instead of being hospitalized

Video recording (section 4.2.5):

- Video recordings of spontaneous events

ILR indications (section 4.2.4.7):

- In patients with suspected unproven epilepsy
- In patients with unexplained falls

ILR indications (section 5.6):

- In patients with primary cardiomyopathy or inheritable arrhythmogenic disorders who are at low risk of sudden cardiac death, as alternative to ICD

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Definition (1)

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- **Syncope** is a TLOC, *due to transient global cerebral hypoperfusion,* characterized by rapid onset, short duration *and* spontaneous complete recovery.

Definition (2)

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- **Transient loss of consciousness** (TLOC) is a state of real or apparent loss of consciousness with loss of awareness, characterized by amnesia for the period of unconsciousness, abnormal motor control, loss of responsiveness, and a short duration.
- **TLOC** is **syncope** when there is:
  a) presence of features specific for reflex, orthostatic hypotension, or cardiac syncope, *and;*
  b) absence of features specific for other forms of TLOC.

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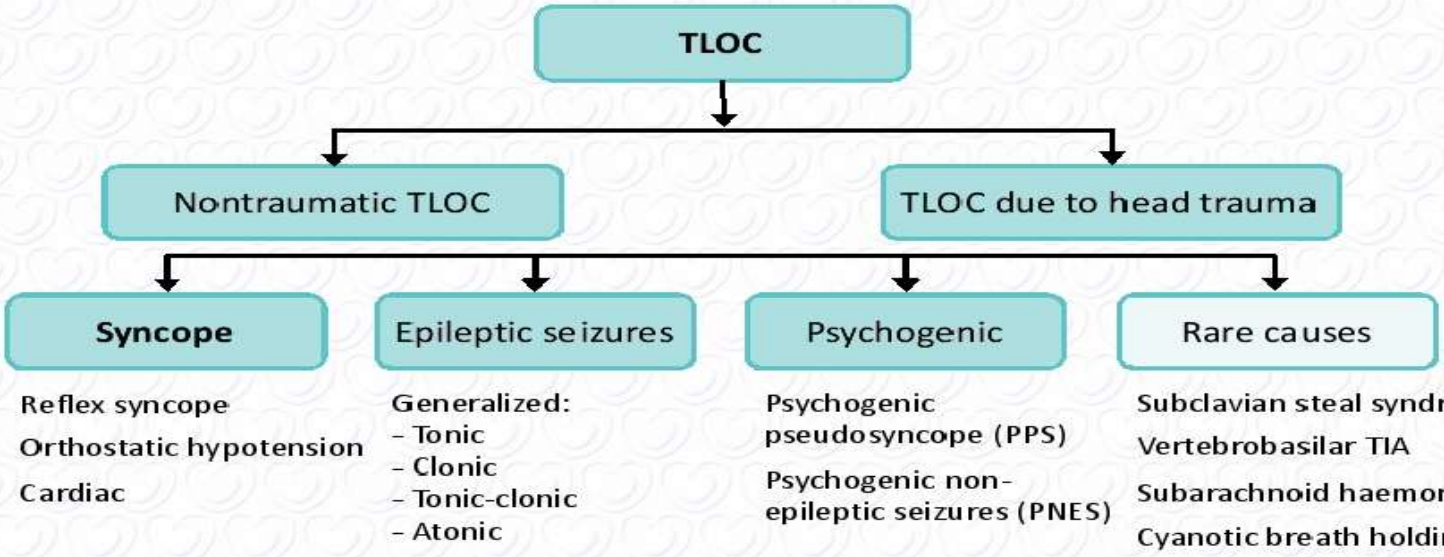
Classification



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Classification
Reflex (neurally-mediated) syncope

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- **Vasovagal:**
  - orthostatic VVS: standing, less common sitting,
  - emotional: fear, pain (somatic or visceral), instrumentation, blood phobia.
- **Situational:**
  - micturition,
  - gastrointestinal stimulation (swallow, defaecation),
  - cough, sneeze,
  - post-exercise,
  - others (e.g. laughing, brass instrument playing).
- **Carotid sinus syndrome.**
- **Non-classical forms** (without prodromes and/or without apparent triggers and/or atypical presentation.

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Classification
Syncope due to orthostatic hypotension

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- **Drug-induced OH (most common cause of OH):**
  - e.g. vasodilators, diuretics, phenothiazine, antidepressants.
- **Volume depletion:**
  - haemorrhage, diarrhoea, vomiting, etc.
- **Primary autonomic failure (neurogenic OH):**
  - pure autonomic failure, multiple system atrophy, Parkinson's disease, dementia with Lewy bodies.
- **Secondary autonomic failure (neurogenic OH):**
  - diabetes, amyloidosis, spinal cord injuries, auto-immune autonomic neuropathy, paraneoplastic autonomic neuropathy, kidney failure.

Note. Hypotension may be exacerbated by venous pooling during exercise (exercise-induced), after meals (postprandial hypotension), and after prolonged bed rest (deconditioning).

Classification
Cardiac syncope

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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 valves dysfunction.
- **Cardiopulmonary and great vessels:** pulmonary embolus, acute aortic dissection, pulmonary hypertension.

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Classification

Conditions (of real or apparent LOC) which may be incorrectly diagnosed as syncope

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- Generalized seizures, complex partial seizures, absence epilepsy.
- Psychogenic pseudosyncope.
- Falls without TLOC.
- Intracerebral or subarachnoid haemorrhage.
- Vertebrobasilar TIA.
- Carotid TIA.
- Subclavian steal syndrome.
- Cataplexy.
- Metabolic disorders including hypoglycaemia, hypoxia, hyper-ventilation with hypocapnia.
- Intoxication.
- Coma.
- Cardiac arrest.

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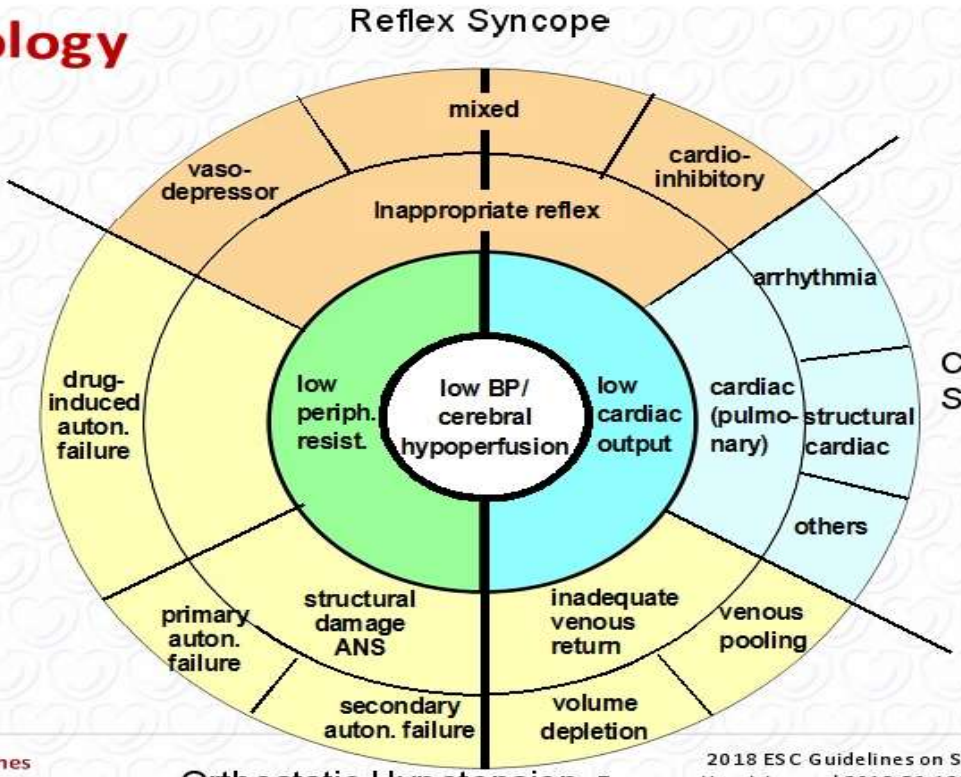
Pathophysiology



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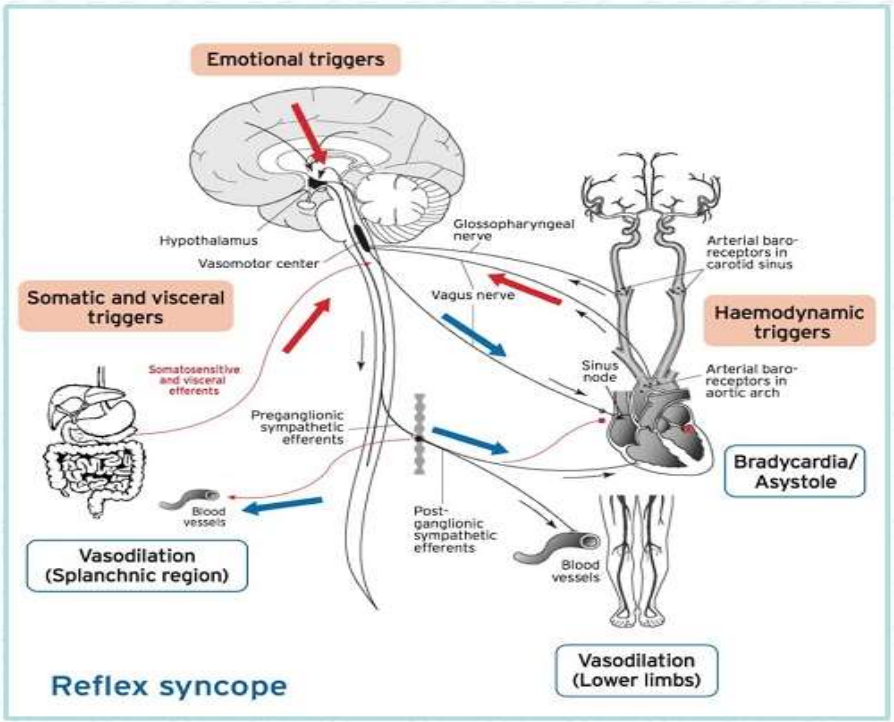
Pathophysiology



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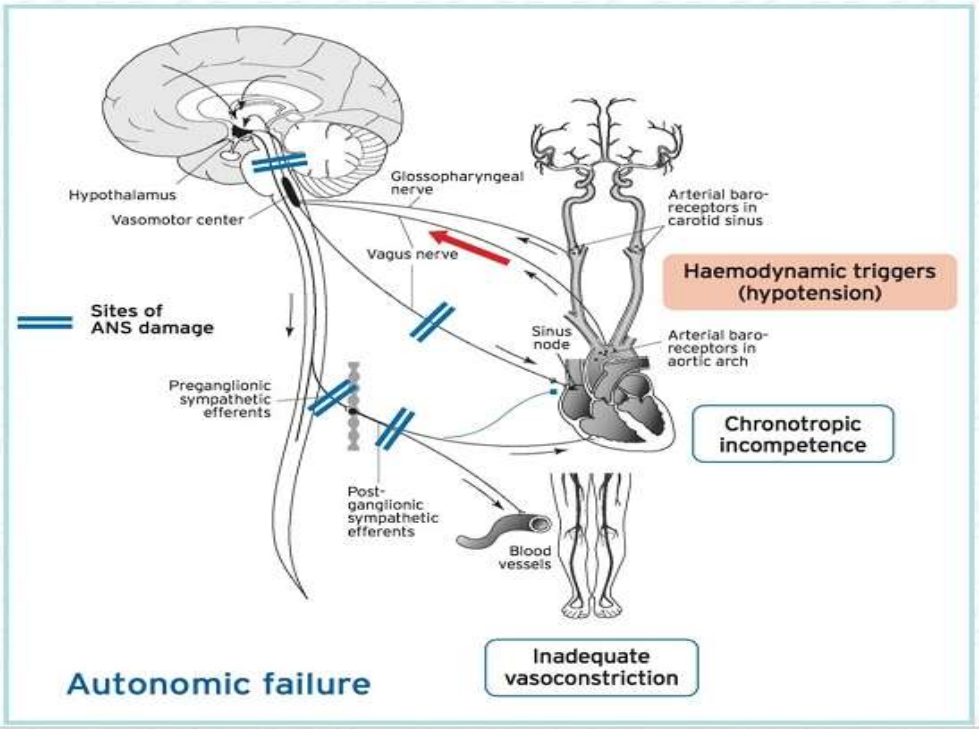
Pathophysiology



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Epidemiology

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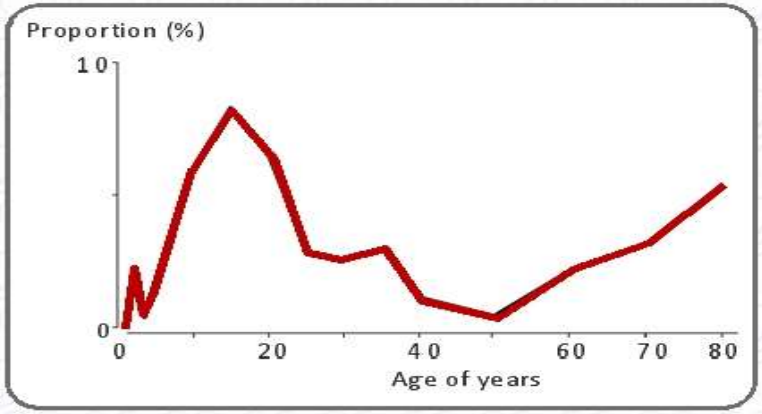
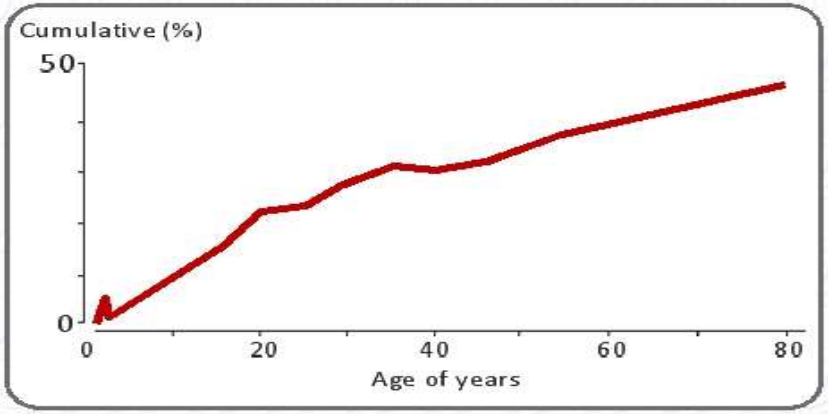
Age of first faint





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Epidemiology - Frequency of the causes of syncope according to the settings (1)

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Setting	Source	Reflex (%)	Orthostatic hypotension (%)	Cardiac (%)	Non syncopal T-LOCs (%)	Un-explained (%)
General population	*Framingham studies*	**21**	**9.4**	**9.5**	**9**	**37**
Emergency department	*Ammirati*	35	6	21	20	17
	Sarasin	38	24	11	8	19
	Blanc	48	4	10	13	24
	Disertori	45	6	11	17	19
	Olde Nordkamp	39	5	5	17	33
	Range	***35-48***	***4-24***	***5-21***	***8-20***	***17-33***

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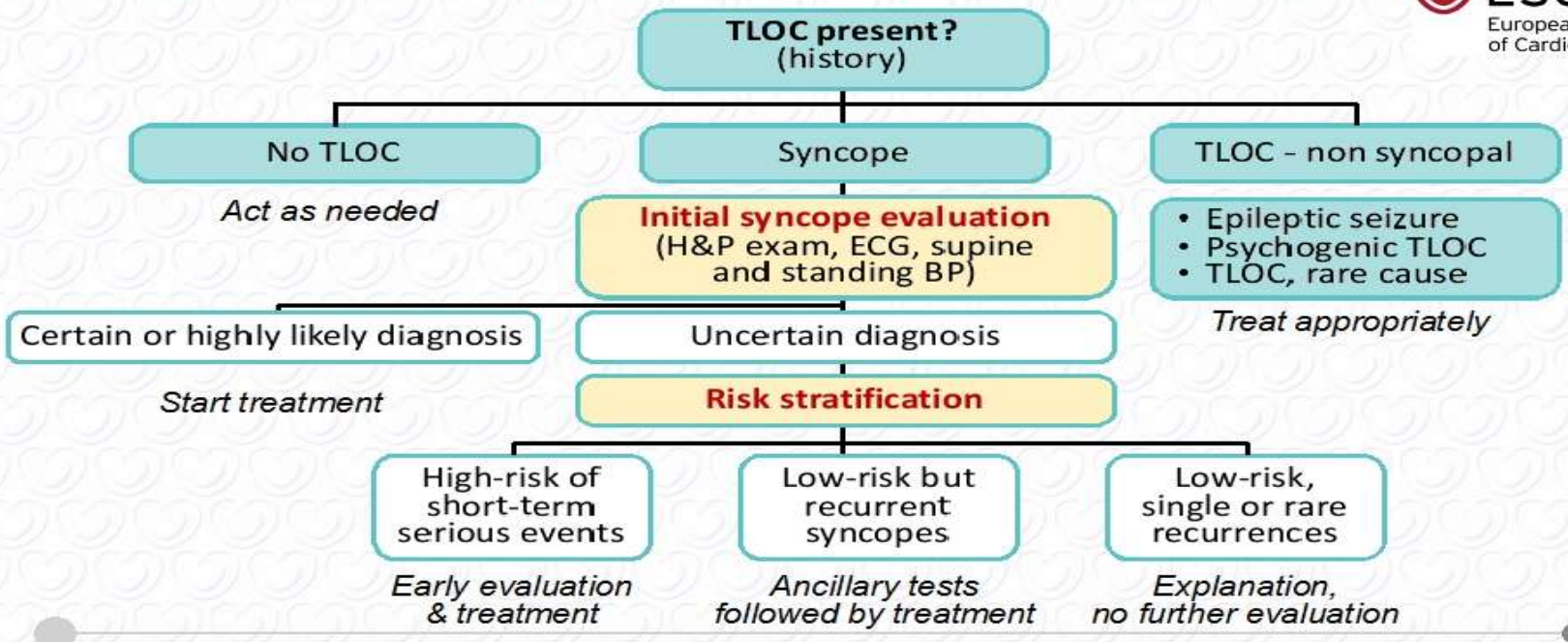
Presentation of patient with probable TLOC



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Risk stratification at the initial evaluation (I)

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Low-risk	High-risk (red flag)
Syncopal event	
1. Associated with prodrome typical of reflex syncope (e.g. light-headedness, feeling of warmth, sweating, nausea, vomiting) 2. After sudden unexpected unpleasant sight, sound, smell, or pain 3. After prolonged standing or crowded, hot places 4. During a meal or postprandial 5. Triggered by cough, defaecation, or micturition 6. With head rotation or pressure on carotid sinus (e.g. tumour, shaving, tight collars) 7. Standing from supine/sitting position	**Major** 1. New onset of chest discomfort, breathlessness, abdominal pain, or headache 2. Syncope during exertion or when supine. 3. Sudden onset palpitation immediately followed by syncope **Minor** (high risk only if associated with structural heart disease or abnormal ECG): 1. No warning symptoms or short (<10 s) prodrome 2. Family history of SCD at young age 3. Syncope in the sitting position

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Risk stratification at the initial evaluation (2)

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Low-risk	High-risk (red flag)
Past medical history	
1. Long history (years) of recurrent syncope with low-risk features with the same characteristics of the current episode 2. Absence of structural heart disease	**Major** 1. Severe structural or coronary artery disease (heart failure, low LVEF or previous myocardial infarction)
Physical examination	
1. Normal examination	**Major** 1. Unexplained systolic BP in the ED <90 mmHg 2. Suggestion of gastrointestinal bleed on rectal examination 3. Persistent bradycardia (<40 b.p.m.) in awake state and in absence of physical training 4. Undiagnosed systolic murmur

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Risk stratification at the initial evaluation (3)

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Low-risk	High-risk (red flag)
ECG	
1. Normal ECG	**Major** 1. ECG changes consistent with acute ischaemia 2. Mobitz II second- and third-degree AV block 3. Slow AF (<40 b.p.m.) 4. Persistent sinus bradycardia (<40 b.p.m.) 5. Bundle branch block or IVCD 6. Q waves consistent with CAD or cardiomyopathy 7. Sustained and non-sustained VT 8. Dysfunction of a pacemaker or ICD 9. Type 1 Brugada pattern 10. Long QT

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Risk stratification at the initial evaluation (4)

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Low-risk	High-risk (red flag)
ECG	
1. Normal ECG	**Minor** (only if history suggests arrhythmic syncope): 1. Mobitz I second-degree AV block and 1° degree AV block with markedly prolonged PR interval 2. Asymptomatic inappropriate mild sinus bradycardia (40–50 b.p.m.), or slow AF (40–50 b.p.m.) 3. Paroxysmal SVT or atrial fibrillation 4. Pre-excited QRS complex 5. Short QTc interval (≤340 ms) 6. Atypical Brugada patterns 7. Negative T waves suggestive of ARVC

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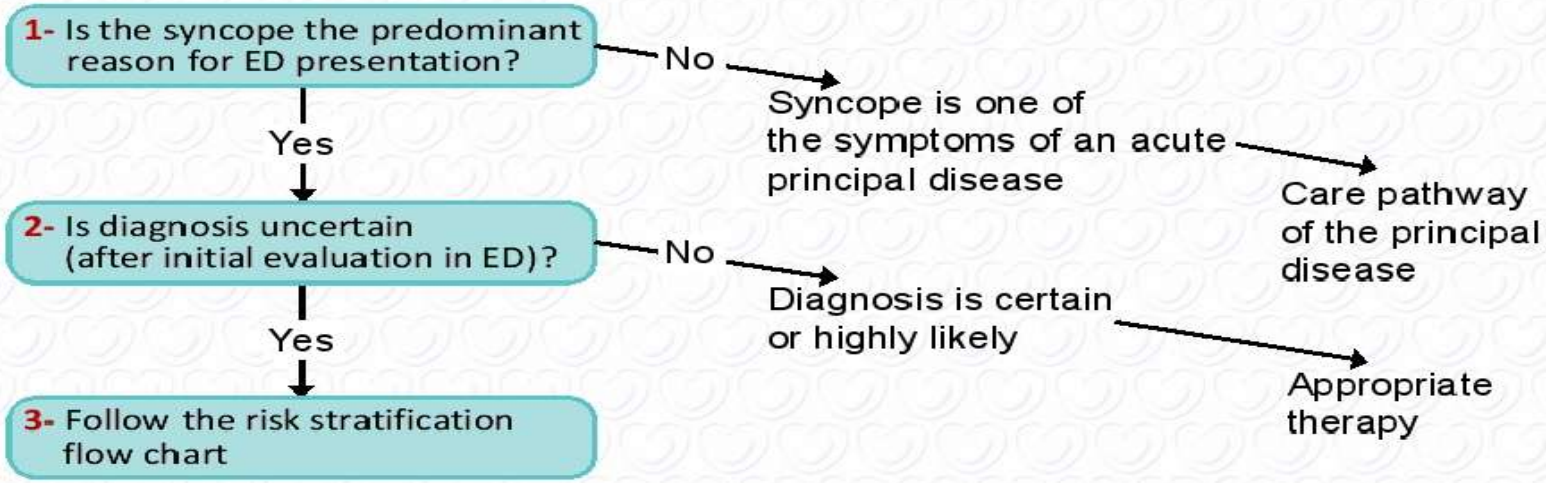
Management of syncope in the ED



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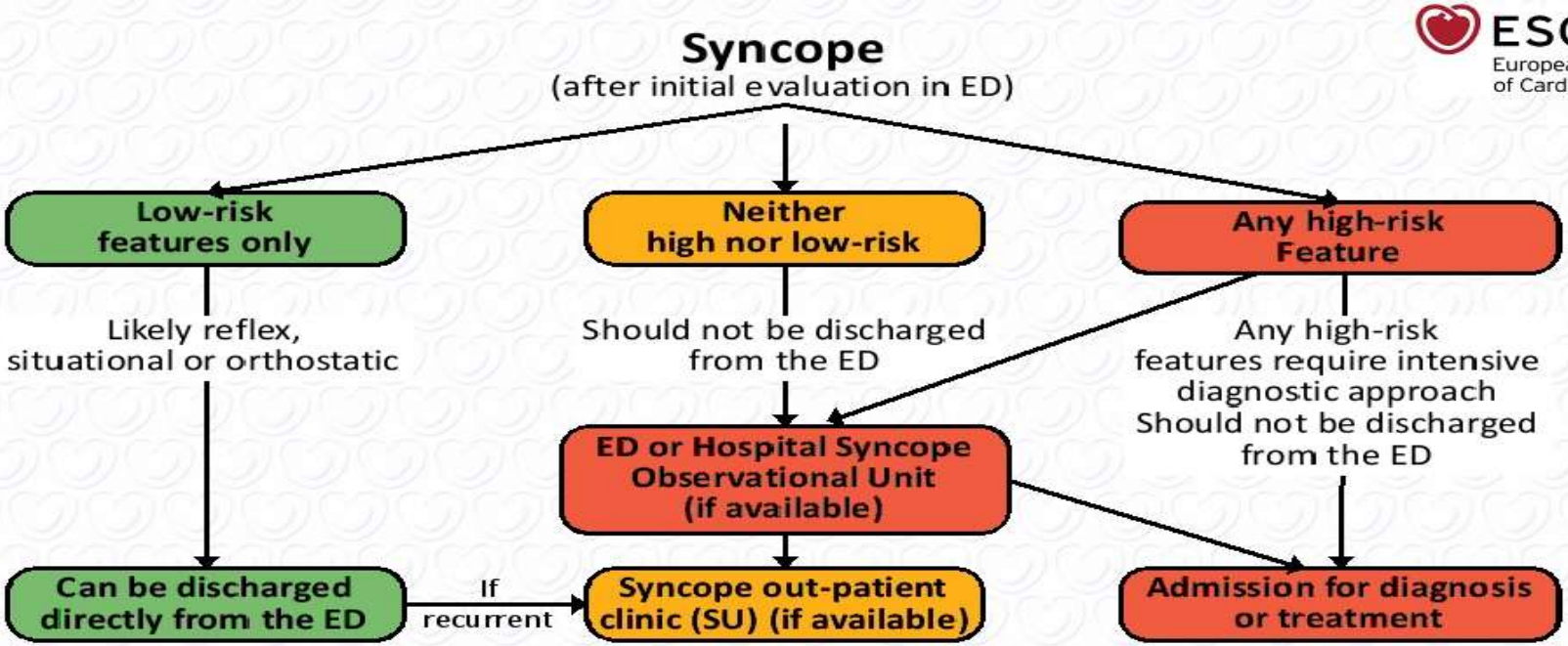

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Management of syncope in the ED

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Recommendations	Class	Level
1. It is recommended that patients with low-risk features, likely to have reflex or situational syncope or syncope due to OH, are discharged from ED.	I	B
2. It is recommended that patients with high-risk features receive an early intensive prompt evaluation in a syncope unit or in an ED observation unit (if available), or are hospitalized.	I	B
3. It is recommended that patients who have neither high- nor low-risk features are observed in the ED or in a syncope unit instead of being hospitalized.	I	B
4. Risk stratification scores may be considered for risk stratification in the ED.	IIb	B

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Management of syncope in the ED

Should the patient be admitted to hospital?

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Favour initial management in ED observation unit and/or fast-track to syncope unit	Favour admission to hospital
High-risk features AND: • Stable, known structural heart disease. • Severe chronic disease. • Syncope during exertion. • Syncope while supine or sitting. • Syncope without prodrome. • Palpitations at the time of syncope. • Inadequate sinus bradycardia or sinoatrial block. • Suspected device malfunction or inappropriate intervention. • Pre-excited QRS complex. • SVT or paroxysmal atrial fibrillation. • ECG suggesting an inheritable arrhythmogenic disorders. • ECG suggesting ARVC.	**High-risk features AND:** • Any potentially severe coexisting disease that requires admission. • Injury caused by syncope. • Need of further urgent evaluation and treatment if it cannot be achieved in another way (i.e. observation unit), e.g. ECG monitoring, echocardiography, stress test, electrophysiological study, angiography, device malfunction, etc. • Need for treatment of syncope.

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Diagnostic criteria with initial evaluation (I)

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Recommendations	Class	Level
Reflex syncope and OH		
1. VVS is highly probable if syncope is precipitated by pain or fear or standing, and is associated with typical progressive prodrome (pallor, sweating, nausea).	I	C
2. Situational reflex syncope is highly probable if syncope occurs during or immediately after specific triggers.	I	C
3. Syncope due to OH is confirmed when syncope occurs while standing and there is concomitant significant OH.	I	C
4. In the absence of the above criteria, reflex syncope and OH should be considered likely when the features that suggest reflex syncope or OH are present and the features that suggest cardiac syncope are absent.	IIa	C

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Diagnostic criteria with initial evaluation (II)

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Recommendations	Class	Level
Cardiac syncope		
1. Arrhythmic syncope is highly probable when the ECG shows: • Persistent sinus bradycardia <40 b.p.m. or sinus pauses >3 seconds in awake state and in absence of physical training, • Mobitz II second- and third-degree AV block, • Alternating left and right BBB, • VT or rapid paroxysmal SVT, • Non-sustained episodes of polymorphic VT and long or short QT interval, • Pacemaker or ICD malfunction with cardiac pauses.	I	C

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Diagnostic criteria with initial evaluation (III)

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Recommendations	Class	Level
Cardiac syncope		
2. Cardiac-ischaemia–related syncope is confirmed when syncope presents with evidence of acute myocardial ischaemia with or without myocardial infarction.	I	C
3. Syncope due to structural cardiopulmonary disorders is highly probable when syncope presents in patients with prolapsing atrial myxoma, left atrial ball thrombus, severe aortic stenosis, pulmonary embolus, or acute aortic dissection.	I	C

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The initial evaluation

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Diagnostic criteria by history

Vasovagal syncope is highly probable if syncope is precipitated by pain or fear or standing, and is associated with typical progressive prodrome (pallor, sweating, nausea).

Situational syncope is reflex syncope is highly probable if syncope occurs during or immediately after specific triggers (e.g., during or immediately after urination, defaecation, cough or swallowing).

Syncope due to ***Orthostatic Hypotension*** is confirmed when syncope occurs while standing and there is concomitant significant orthostatic hypotension.

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The initial evaluation

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ECG diagnostic criteria

Syncope due to cardiac arrhythmia is highly probable in case of:

- Persistent sinus bradycardia <40 beats/min or sinus pauses >3 s in awake state and in absence of physical training,
- Mobitz II 2nd or 3rd degree atrioventricular block,
- Alternating left and right bundle branch block,
- Rapid paroxysmal supraventricular tachycardia or ventricular tachycardia,
- Non-sustained episodes of polymorphic VT and long or short QT interval,
- Pacemaker or ICD malfunction with cardiac pauses.

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The initial evaluation

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ECG diagnostic criteria

Cardiac-ischaemia–related syncope is confirmed when syncope presents with evidence of acute myocardial ischaemia with or without myocardial infarction (*)

** The mechanism can be cardiac (low output or arrhythmia) or reflex (Bezold-Jarish reflex), but management is primarily that of ischemia.*

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The initial evaluation

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ECHO diagnostic criteria

Syncope due to structural cardiopulmonary disorders (*) *is highly probable in patients with:*

- prolapsing atrial myxoma,
- left atrial ball thrombus,
- severe aortic stenosis,
- pulmonary embolus,
- acute aortic dissection.

** The mechanism can be multifactorial, but management is primarily that of the underlying structural disease*

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The initial evaluation

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Indications for blood tests

- Haematocrit or haemoglobin when haemorrhage is suspected,
- Oxygen saturation and blood gas analysis when hypoxia is suspected,
- Troponin when cardiac-ischaemia related syncope is suspected,
- D-dimer when pulmonary embolism is suspected.

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Clinical & ECG features that suggest a cardiac syncope

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- During exertion or when supine.
- Presence of structural heart disease or coronary artery disease.
- Family history of unexplained sudden death at young age.
- Sudden onset palpitations immediately followed by syncope.
- ECG findings suggesting arrhythmic syncope:
  - Bifascicular block?
  - Other intraventricular conduction abnormalities (QRS duration ≥0.12 s),
  - Mobitz I second-degree AV block,
  - 1° degree AV block with markedly prolonged PR interval,
  - Asymptomatic mild inappropriate sinus bradycardia (40–50 b.p.m.) or slow atrial fibrillation (40–50 b.p.m.),
  - Non-sustained VT,
  - Pre-excited QRS complexes,
  - Long or short QT intervals,
  - Early repolarization,
  - Type 1 Brugada pattern,
  - Negative T waves in right precordial leads, epsilon waves suggestive of ARVC,
  - Left ventricular hypertrophy suggesting hypertrophic cardiomyopathy.

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Clinical and ECG features that suggest a reflex (neurally-mediated) syncope

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- Long history of recurrent syncope, in particular occurring before the age of 40 years.
- After unpleasant sight, sound, smell, or pain.
- Prolonged standing.
- During meal.
- Being in crowded and/or hot places.
- Autonomic activation before syncope: pallor, sweating, and/or nausea/vomiting.
- With head rotation or pressure on carotid sinus (as in tumours, shaving, tight collars).
- Absence of heart disease.

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Advice for driving in patients with syncope (I)

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Disorder causing syncope	Group 1 (private drivers)	Group 2 (professional drivers)
Cardiac arrhythmias		
Untreated arrhythmias	Unfit to drive	Unfit to drive
Cardiac arrhythmia, not life-threatening, medical treatment	After successful treatment is established	After successful treatment is established
Cardiac arrhythmia, life-threatening (e.g. inheritable disorders), medical treatment	After successful treatment is established	Permanent restriction
Pacemaker implant	After 1 week	After appropriate function is established (first post-implant visit)

Disclaimer: Country-specific regulations may differ

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Advice for driving in patients with syncope (2)

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Disorder causing syncope	Group 1 (private drivers)	Group 2 (professional drivers)
Catheter ablation.	After successful treatment is established	After successful treatment is established.
Implantable cardioverter defibrillator implant.	After 1 month. The risk may increase in the few months following an implantable cardioverter defibrillator shock (3 months).	Permanent restriction.
Structural cardiac/cardiopulmonary		
	After appropriate function is established.	After appropriate function is established.
Orthostatic hypotension (neurogenic)		
Syncope while sitting.	After successful treatment is established.	After successful treatment is established.

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Advice for driving in patients with syncope (3)

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Disorder causing syncope	Group 1 (private drivers)	Group 2 (professional drivers)
Reflex syncope		
Single/mild	No restrictions unless it occurred during driving.	No restriction unless it occurred during driving or without prodromes.
Recurrent and severe	After successful treatment is established.	After successful treatment is established. Particular caution if it occurred during driving or without prodromes.
Unexplained syncope		
	No restrictions unless absence of prodrome, occurrence during driving, or presence of severe structural heart disease. If yes, after diagnosis and appropriate therapy is established.	After diagnosis and appropriate therapy is established.

Disclaimer: Country-specific regulations may differ

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Clinical and ECG features that suggest a syncope due to orthostatic hypotension

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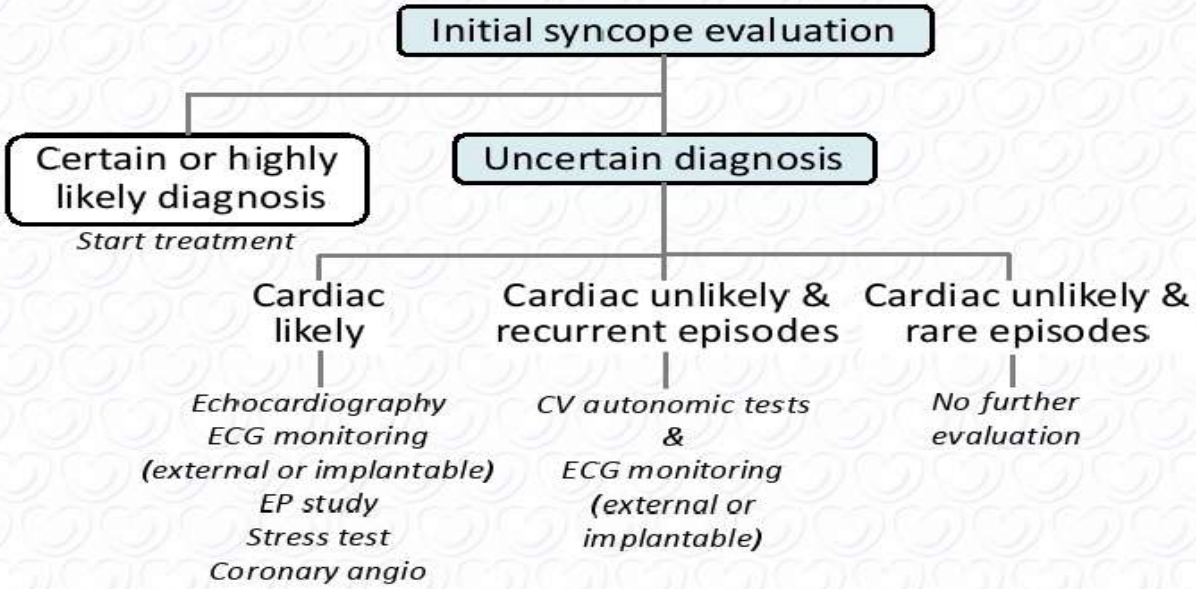
- While or after standing.
- Prolonged standing.
- Standing after exertion.
- Post-prandial hypotension.
- Temporal relationship with start or changes of dosage of vasodepressive drugs or diuretics leading to hypotension.
- Presence of autonomic neuropathy or Parkinsonism.

The initial evaluation: diagnostic strategy



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Basic cardiovascular autonomic function tests

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- Active standing.
- Valsalva manoeuvre & deep breathing.
- Carotid sinus massage.
- Tilt testing.
- Ambulatory BP monitoring.

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Basic cardiovascular autonomic function tests

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Active Standing Test

		History of syncope and orthostatic complaints	
		Highly suggestive of OH: • *syncope and presyncope during standing, not during lying;* • *complaints may get worse immediately after exercise, after meals or in high temperatures;* • *no 'autonomic activation'*	Possibly due to OH: *not all of the features highly suggestive of OH are present*
Supine and standing BP measurement for 3 minutes	Symptomatic abnormal BP fall	**Syncope is due to OH** (class I)	**Syncope is likely due to OH** (class IIa)
	Asymptomatic abnormal BP fall	**Syncope is likely due to OH** (class IIa)	**Syncope may be due to OH** (class IIb)
	No abnormal BP drop	**Unproven**	**Unproven**

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Active standing test (1)

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Recommendations	Class	Level
Indication		
1. Intermittent determination by sphygmomanometer of BP and HR while supine and during active standing for 3 minutes are indicated at initial syncope evaluation.	I	C
2. Continuous beat-to-beat non-invasive BP and HR measurement may be preferred when short-lived BP variations are suspected such as in initial OH.	IIb	C
Diagnostic criteria		
3. Syncope due to OH is confirmed when there is a fall in systolic BP from baseline value ≥20 mmHg or diastolic BP ≥10 mmHg or a decrease in systolic BP to <90 mmHg that reproduces spontaneous symptoms.	I	C
4. Syncope due to OH should be considered likely when there is an asymptomatic fall in systolic BP from baseline value ≥20 mmHg or diastolic BP ≥10 mmHg or a decrease in systolic BP to <90 mmHg and symptoms (from history) are consistent with OH.	IIa	C

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Active standing test (2)

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Recommendations	Class	Level
Diagnostic criteria		
5. Syncope due to OH should be considered likely when there is a symptomatic fall in systolic BP from baseline value ≥20 mmHg or diastolic BP ≥10 mmHg or a decrease in systolic BP to <90 mmHg and not all of the features (from history) are suggestive of OH.	IIa	C
6. POTS should be considered likely when there is an orthostatic HR increase (>30 b.p.m. or to >120 b.p.m. within 10 minutes of active standing) in the absence of OH that reproduces spontaneous symptoms.	IIa	C
7. Syncope due to OH may be considered possible when there is an asymptomatic fall in systolic BP from baseline value ≥20 mmHg or diastolic BP ≥10 mmHg or a decrease in systolic BP to <90 mmHg and symptoms (from history) are less consistent with OH.	IIb	C

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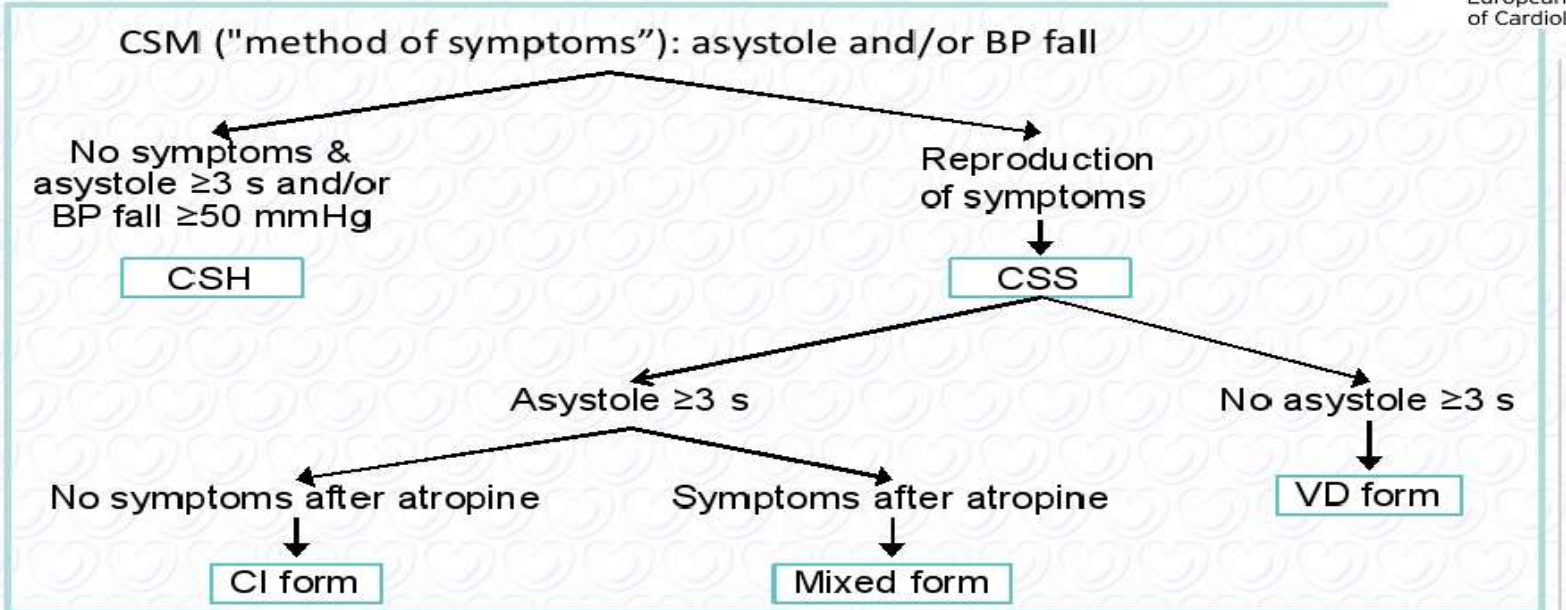
Carotid sinus massage: "Method of symptoms"



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Carotid sinus massage: Cardioinhibitory form

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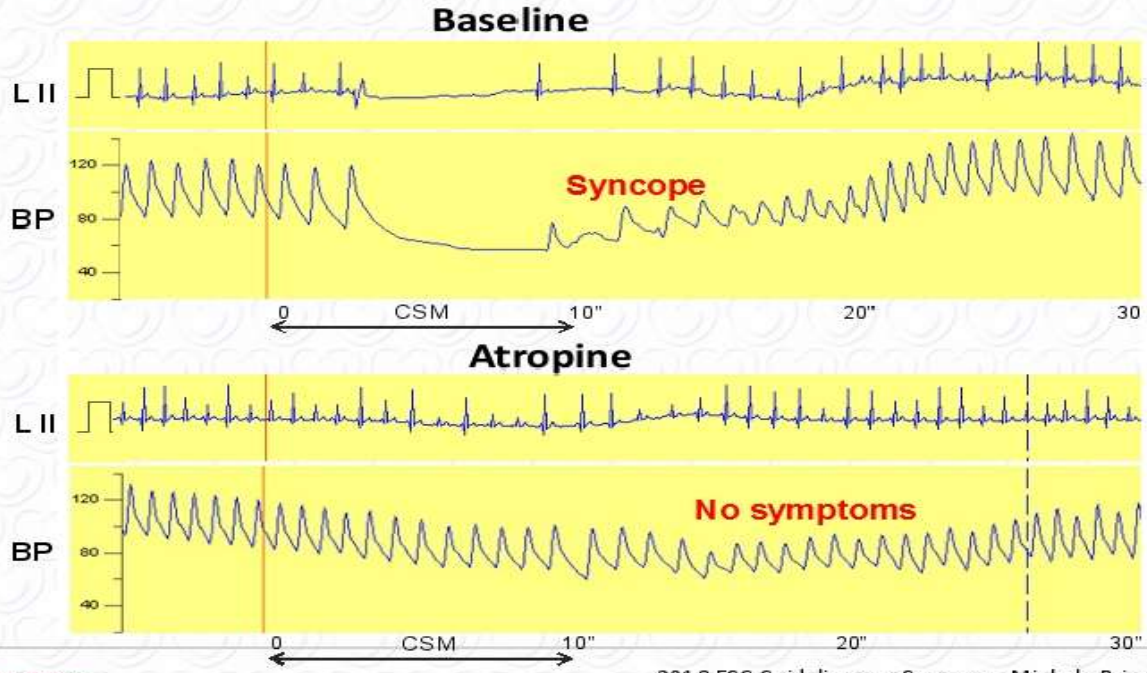

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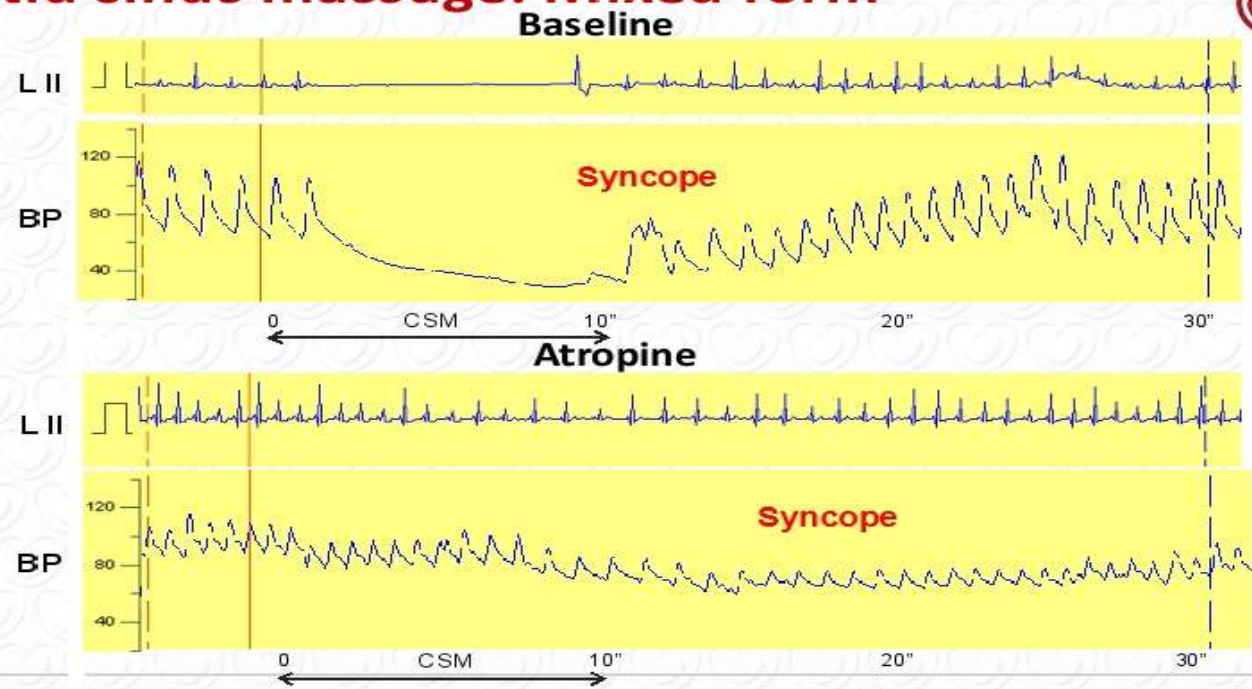
Carotid sinus massage: Mixed form



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Carotid sinus massage

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Recommendations	Class	Level
Indication		
1. CSM is indicated in patients >40 years of age with syncope of unknown origin compatible with a reflex mechanism.	I	B
Diagnostic criteria		
2. CSS is confirmed if CSM causes bradycardia (asystole) and/or hypotension that reproduce spontaneous symptoms and patients have clinical features compatible with a reflex mechanism of syncope.	I	B

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Electrocardiographic monitoring

Type	ECG classification	Suggested pathophysiology
Type 1. **Asystole**	*Type 1A.* Sinus arrest	Probably reflex
	Type 1B. Sinus bradycardia plus AV block	Probably reflex
	Type 1C. Sudden onset AV block	Probably intrinsic or idiopathic ("low adenosine")
Type 2. **Bradycardia**	Decrease in HR >30% or <40 b.p.m. for >10 seconds	Probably reflex
Type 3. **No or slight rhythm variations**	Variations in HR <30% and HR >40 b.p.m	Uncertain
Type 4. **Tachycardia**	*Type 4A.* Progressive sinus tachycardia	Uncertain
	Type 4B. Atrial fibrillation	Cardiac arrhythmia
	Type 4C. SVT (except sinus)	Cardiac arrhythmia
	Type 4D. Ventricular tachycardia	Cardiac arrhythmia

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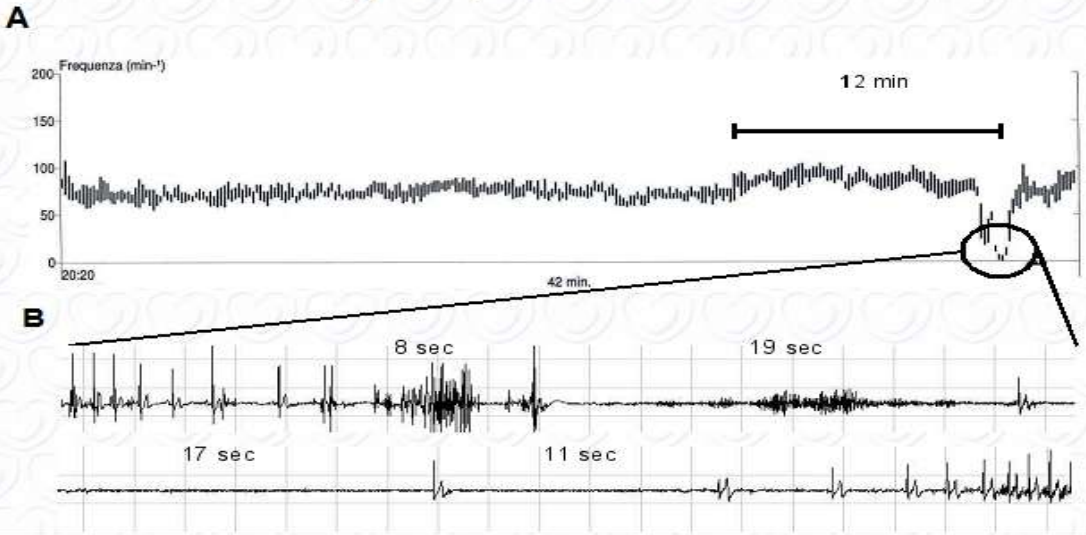
***Type 1A*, sinus arrest**



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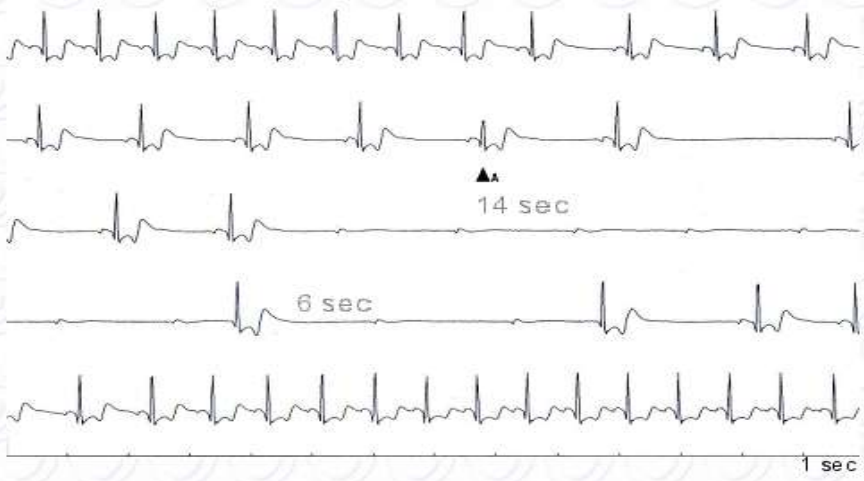
***Type 1B*, sinus bradycardia plus atrioventricular block**



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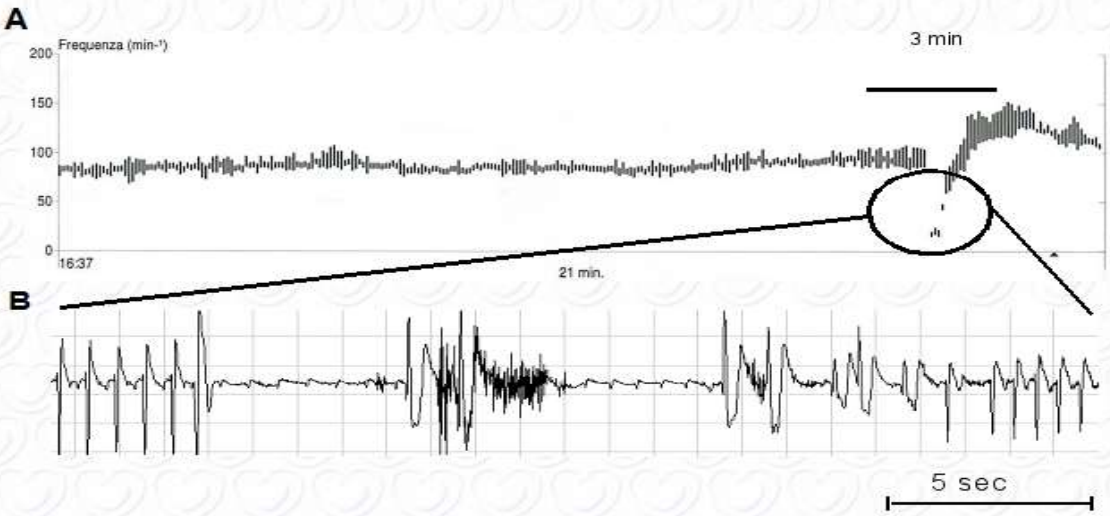
***Type 1C*, intrinsic atrioventricular block**



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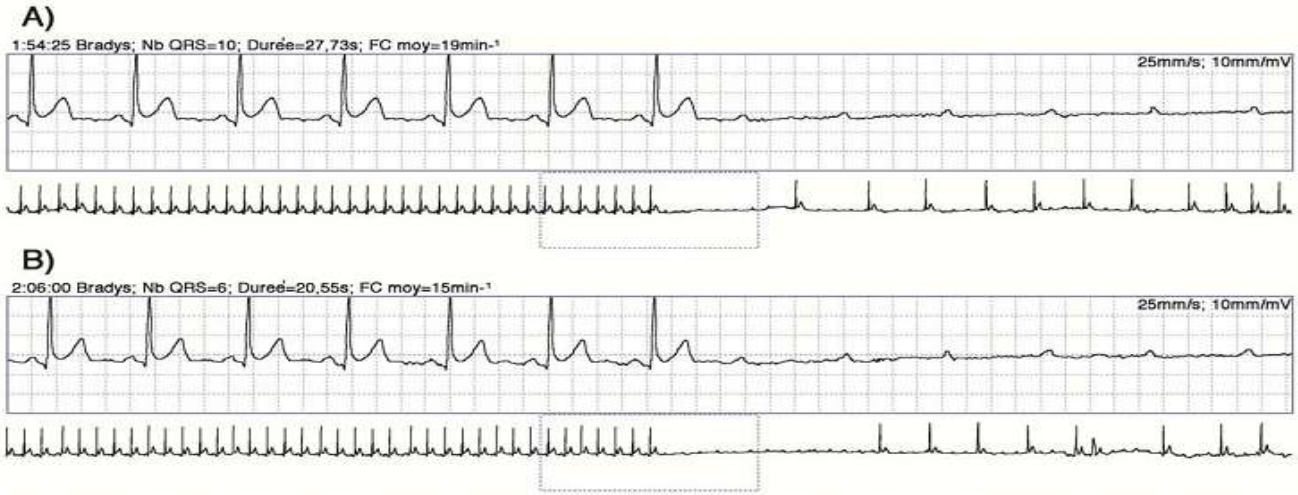
***Type 1C*, idiopathic AV block ("low adenosine")**



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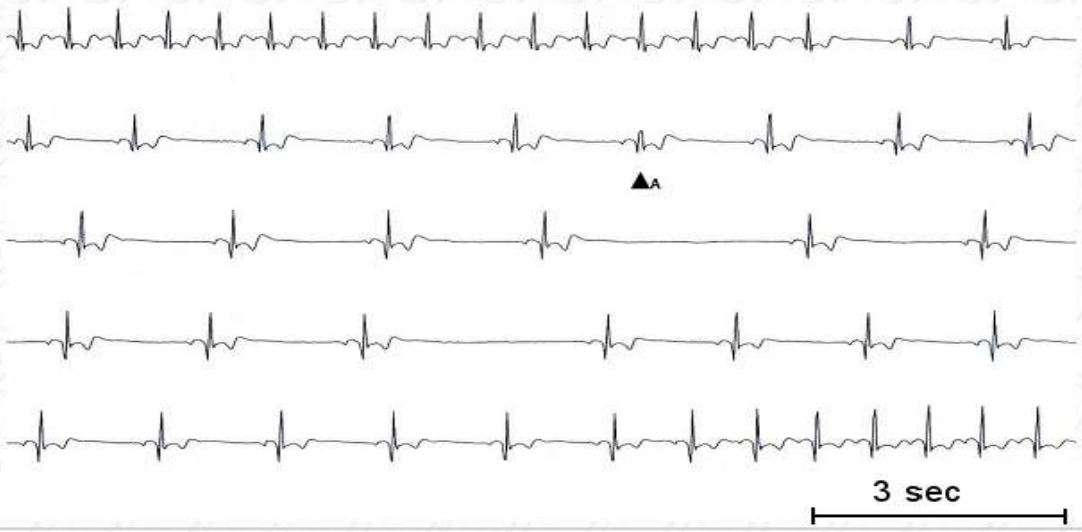
***Type 2*, bradycardia**



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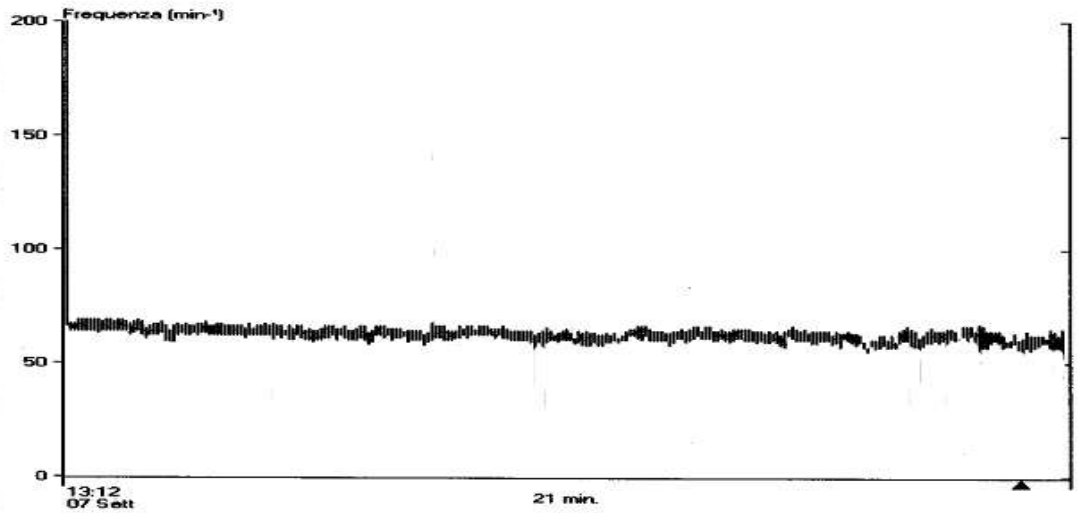
***Type 3*, no or slight rhythm variations**



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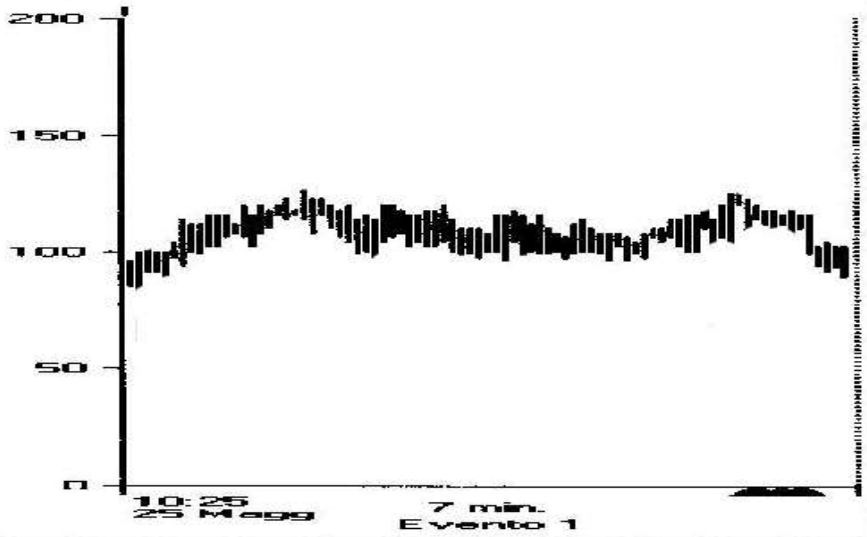
***Type 3*, no or slight rhythm variations**



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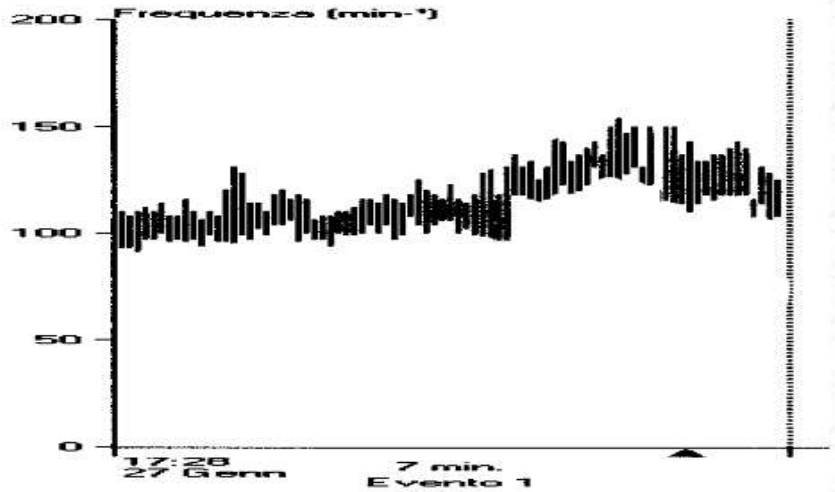
***Type 4*, tachycardia**



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ECG monitoring: indications

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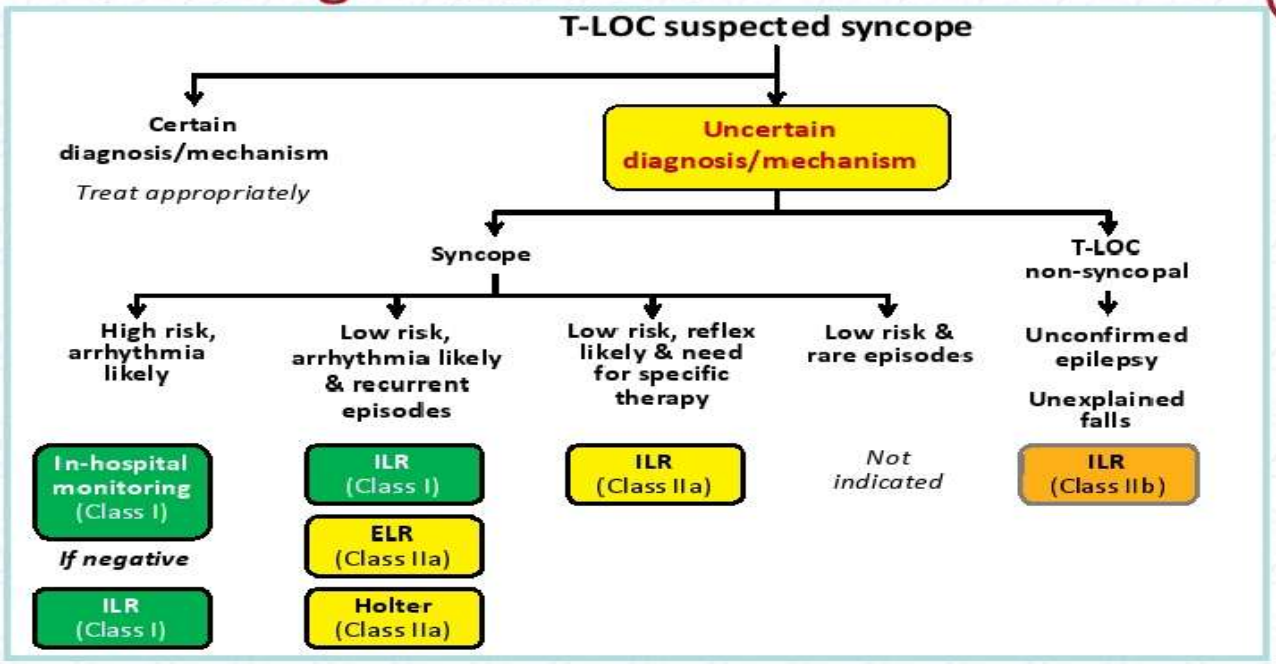
Metanalysis of RCT of ILR vs conventional strategy

Study	ILR n/N	Control n/N	Relative probability	95% CI	p
RAST *Circ 2001*	14/27 (52%)	6/30 (20%)	2.6	1.2-5.8	0.01
EaSyAS *Eur Heart J 2006*	43/101 (41%)	7/97 (7%)	5.9	2.8-12	0.001
Da Costa *Arch Card Dis 2013*	15/41 (37%)	4/37 (11%)	3.4	1.2-9.3	0.01
FRESH *Arch Card Dis 2014*	18/39 (46%)	2/39 (5%)	9	2.2-36	0.001
EaSyAS II *Europace 2016*	62/125 (50%)	21/121 (17%)	2.9	1.9-4.4	0.001
Total	**152/336 (46%)**	**40/324 (12%)**	**3.6**	**2.4-5.3**	**0.001**

Test for heterogeneity: p=0.26

ECG monitoring: indications



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ECG monitoring: Indications (I)

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Recommendations	Class	Level
In-hospital monitoring		
1. *Immediate in-hospital monitoring* (in bed or by telemetry) is indicated in high-risk patients.	I	C
Holter monitoring		
2. *Holter monitoring* should be considered in patients who have frequent syncope or presyncope (≥1 episode per week).	IIa	B
External loop recorder		
3. External loop recorders should be considered, early after the index event, in patients who have an inter-symptom interval ≤4 weeks	IIa	B

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ECG monitoring: Indications (II)

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Recommendations	Class	Level
Implantable loop recorder		
4. ILR is indicated in an early phase of evaluation in patients with recurrent syncope of uncertain origin, absence of high-risk criteria (listed in *Table 6*), and a high likelihood of recurrence within the battery life of the device.	I	A
5. ILR should be considered in patients with suspected or certain reflex syncope presenting with frequent or severe syncopal episodes.	IIa	B
6. ILR may be considered in patients in whom epilepsy was suspected but the treatment has proven ineffective.	IIa	B
7. ILR may be considered in patients with unexplained falls.	IIb	B

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ECG monitoring: Diagnostic criteria

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Recommendations	Class	Level
1. Arrhythmic syncope is confirmed when a correlation between syncope and an arrhythmia (bradyarrhythmia or tachyarrhythmia) is detected.	I	B
2. In the absence of syncope, arrhythmic syncope should be considered likely when periods of Mobitz II second- or third-degree AV block or a ventricular pause >3 seconds (with possible exception of young trained persons, during sleep or rate-controlled atrial fibrillation), or rapid prolonged paroxysmal SVT or VT are detected.	IIa	C

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Electrophysiological study: **Indications**

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Recommendations	Class	Level
1. In patients with syncope and previous myocardial infarction or other scar-related conditions, EPS is indicated when syncope remains unexplained after non-invasive evaluation.	I	B
2. In patients with syncope and bifascicular BBB, EPS should be considered when syncope remains unexplained after non- invasive evaluation.	IIa	B
3. 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.	IIb	B
4. In patients with syncope preceded by sudden and brief palpitations, EPS may be considered when syncope remains unexplained after non-invasive evaluation.	IIb	C

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EPS-guided therapy

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Recommendations	Class	Level
1. 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, or second- or third-degree His-Purkinje block during incremental atrial pacing, or with pharmacological challenge.	I	B
2. In patients with unexplained syncope and previous myocardial infarction or other scar-related conditions, it is recommended to manage induction of sustained monomorphic VT according to the current ESC guidelines for VA.	I	B
3. In patients without structural heart disease with syncope preceded by sudden and brief palpitations, it is recommended to manage the induction of rapid SVT or VT, which reproduces hypotensive or spontaneous symptoms, with appropriate therapy according to the current ESC Guidelines.	I	C
4. In patients with syncope and asymptomatic sinus bradycardia, a pacemaker should be considered if a prolonged corrected SNRT is present.	IIa	B

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Echocardiography

Recommendations	Class	Level
Indications		
1. Echocardiography is indicated for diagnosis and risk stratification in patients with suspected structural heart disease	I	B
2. Two-dimensional and Doppler echocardiography during exercise in the standing, sitting, or semi-supine position to detect provocable left ventricular outflow tract obstruction is indicated in patients with HCM, a history of syncope, and a resting or provoked peak instantaneous left ventricular outflow tract gradient <50 mmHg	I	B
Diagnostic criteria		
3. Aortic stenosis, obstructive cardiac tumours or thrombi, pericardial tamponade, and aortic dissection are the most probable causes of syncope when the echocardiography shows the typical features of these conditions	I	C

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Exercise testing

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Recommendations	Class	Level
Indications		
1. Exercise testing is indicated in patients who experience syncope during or shortly after exertion.	I	C
Diagnostic criteria		
2. Syncope due to second- or third-degree AV block is confirmed when the AV block develops during exercise, even without syncope.	I	C
3. Reflex syncope is confirmed when syncope is reproduced immediately after exercise in the presence of severe hypotension.	I	C

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Coronary angiography

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Recommendations	Class	Level
1. In patients with syncope, the same indications for coronary angiography should be considered as in patients without syncope.	IIa	C

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Treatment of syncope: General principles

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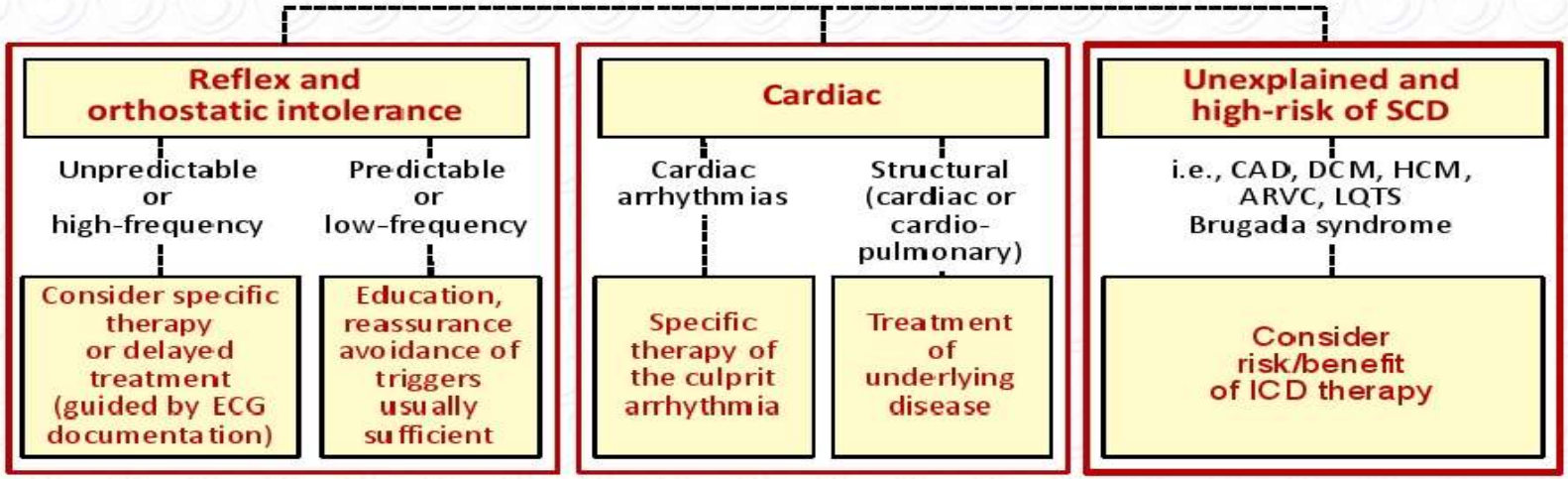
Diagnostic evaluation



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Treatment syncope: **Reflex syncope**

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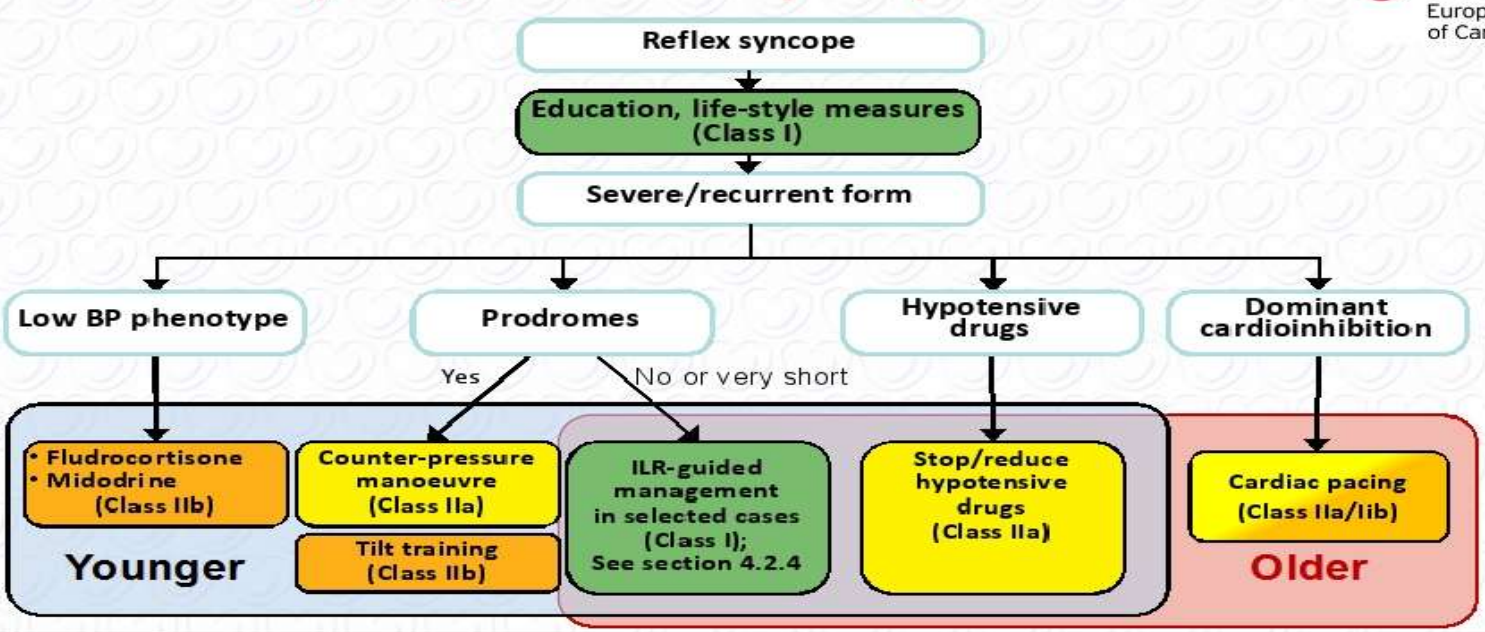

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Treatment of **Reflex syncope (I)**

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Recommendations	Class	Level
Education and life-style modification		
1. Explanation of the diagnosis, provision of reassurance, explanation of risk of recurrence, avoidance of triggers and situations are indicated in all patients.	I	B
Discontinuation/reduction of hypotensive therapy		
2. Modification or discontinuation of hypotensive drug regimen should be considered in patients with vasodepressor syncope, if possible.	IIa	B
Physical manoeuvres		
3. Isometric PCM should be considered in patients with prodromes who are less than 60 years of age.	IIa	B
4. Tilt training may be considered for the education of young patients.	IIb	B

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Treatment of Reflex syncope (II)

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Recommendations	Class	Level
Pharmacological therapy		
5. Fludrocortisone may be considered in young patients with the orthostatic form of VVS, low-normal values of arterial BP, and absence of contraindication to the drug.	IIb	B
6. Midodrine may be considered in patients with the orthostatic form of VVS.	IIb	B
7. Beta-adrenergic blocking drugs are not indicated.	III	B

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Treatment of syncope: **Orthostatic hypotension**

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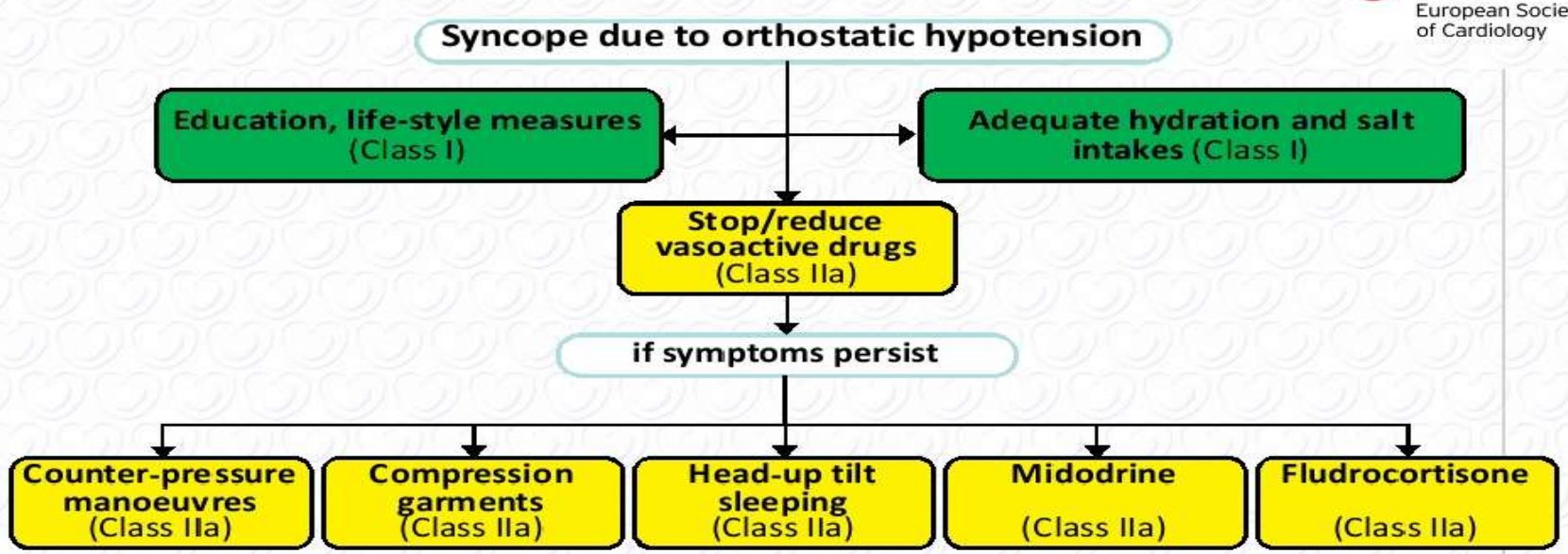

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Treatment of syncope: **Orthostatic Hypotension**

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Recommendations	Class	Level
1. Explanation of the diagnosis, provision of reassurance, explanation of risk of recurrence, and avoidance of triggers and situations are indicated in all patients.	I	C
2. Adequate hydration and salt intake are indicated.	I	C
3. Modification or discontinuation of hypotensive drugs regimen should be considered.	IIa	B
4. Isometric PCM should be considered.	IIa	B
5. Abdominal binders and/or support stockings to reduce venous pooling should be considered.	IIa	B
7. Head-up tilt sleeping (>10 degrees) to increase fluid volume should be considered.	IIa	B
8. Midodrine should be considered if symptoms persist.	IIa	B
9. Fludrocortisone should be considered if symptoms persist.	IIa	C

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Treatment of syncope: **Cardiac arrhythmias**

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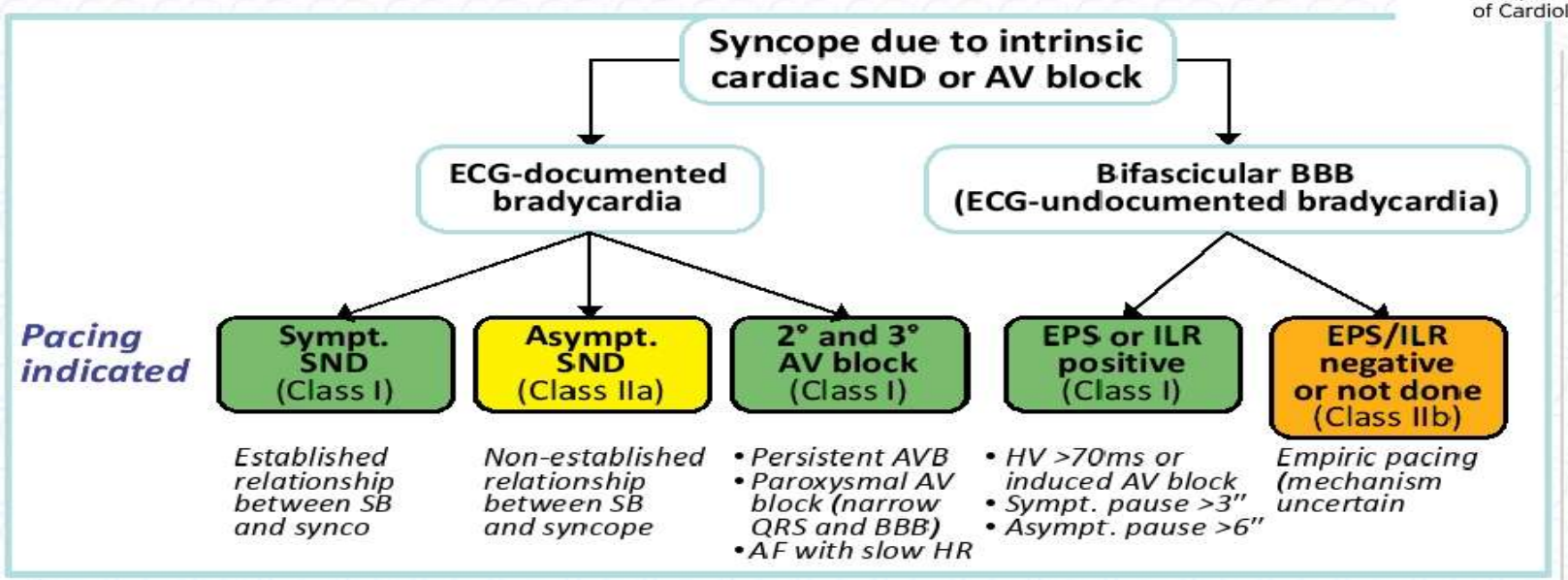

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Treatment of syncope: **Bundle Branch Block**

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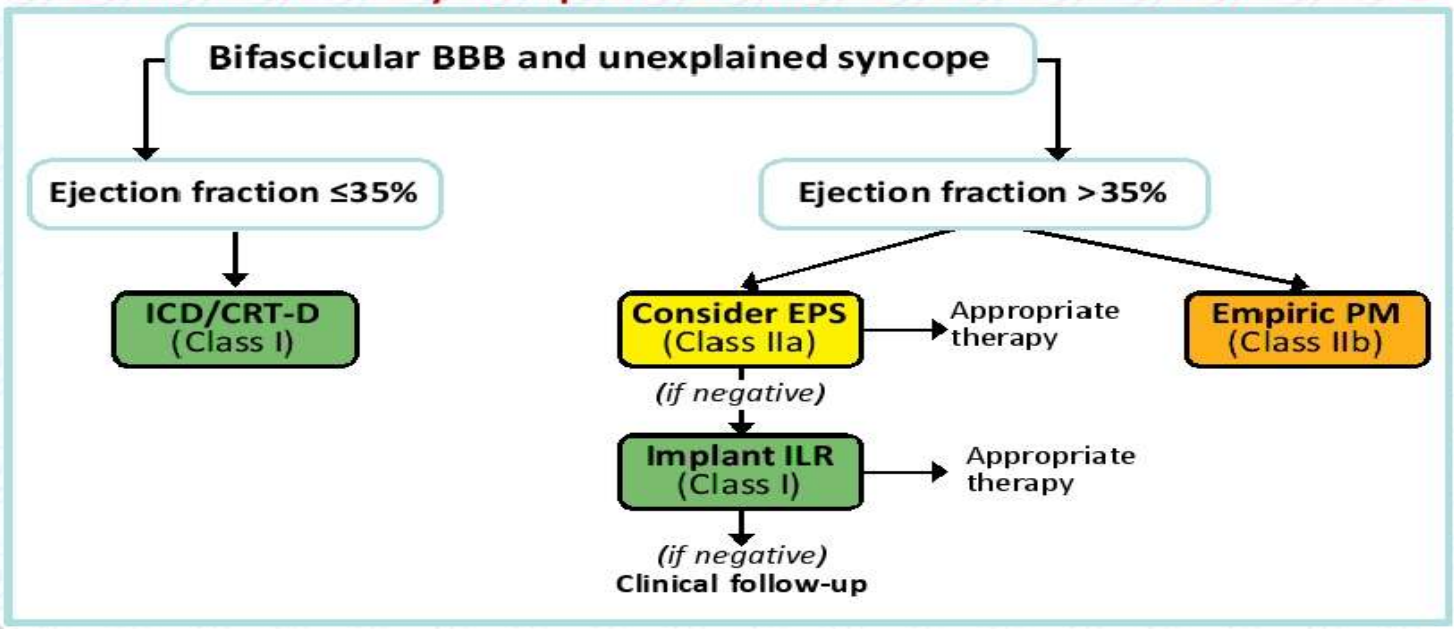

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Treatment of syncope: **Cardiac arrhythmias (I)**

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Recommendations	Class	Level
Bradycardia (intrinsic)		
1. Cardiac pacing is indicated when there is an established relationship between syncope and symptomatic bradycardia due to sick sinus syndrome or intrinsic AV block.	I	B
2. Cardiac pacing is indicated in patients with intermittent/ paroxysmal intrinsic third- or second-degree AV block (including AF with slow ventricular conduction) although there is no documentation of correlation between symptoms and ECG.	I	C
3. Cardiac pacing should be considered when the relationship between syncope and asymptomatic sinus node dysfunction is less established.	IIa	C
4. Cardiac pacing is not indicated in patients when there are reversible causes for bradycardia.	III	C

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Treatment of syncope: **Cardiac arrhythmias (II)**

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Recommendations	Class	Level
Bifascicular BBB		
5. Cardiac pacing is indicated in patients with syncope, BBB, and a positive EPS or ILR-documented AV block.	I	B
6. Cardiac pacing may be considered in patients with unexplained syncope and bifascicular BBB.	IIb	B

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Treatment of syncope:
Cardiac tachyarrhythmias

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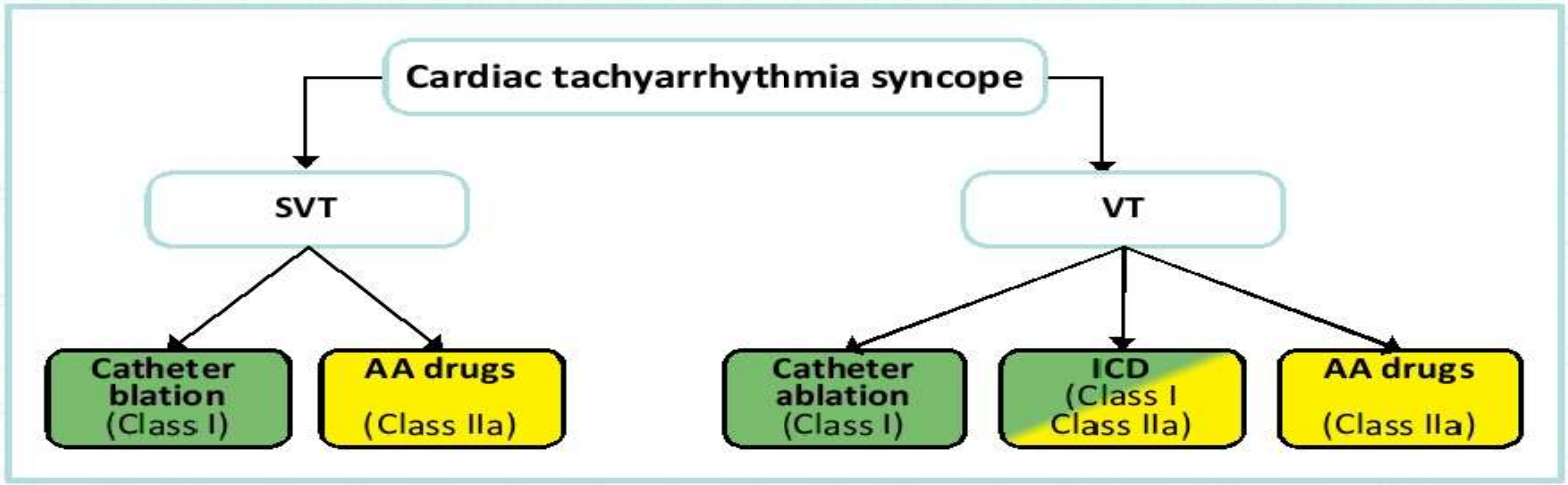

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Treatment of syncope: **Cardiac arrhythmias (III)**

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Recommendations	Class	Level
Tachycardia		
1. Catheter ablation is indicated in patients with syncope due to SVT or VT in order to prevent syncope recurrence.	I	B
2. An ICD is indicated in patients with syncope due to VT and ejection fraction ≤35%.	I	A
3. An ICD is indicated in patients with syncope and previous myocardial infarction who have VT induced during EPS.	I	C
4. An ICD should be considered in patients with ejection fraction >35% with recurrent syncope due to VT when catheter ablation and pharmacological therapy have failed or could not be performed.	IIa	C
5. Antiarrhythmic drug therapy, including rate-control drugs, should be considered in patients with syncope due to SVT or VT.	IIa	C

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Treatment of syncope: **Unexplained syncope in patients at high risk of SCD (I)**

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Recommendations	Class	Level
Left ventricular systolic dysfunction		
1. ICD therapy is recommended to reduce SCD in patients with symptomatic heart failure (NYHA class II–III) and LVEF ≤35% after ≥3 months of optimal medical therapy who are expected to survive for at least 1 year with good functional status	I	A
2. An ICD should be considered in patients with unexplained syncope with systolic impairment but without a current indication for ICD to reduce the risk of sudden death	IIa	C
3. Instead of an ICD, an ILR may be considered in patients with recurrent episodes of unexplained syncope with systolic impairment but without a current indication for ICD	IIb	C
Unexplained syncope is defined as syncope that does not meet a Class I diagnostic criterion defined in the tables of recommendations. In the presence of clinical features described in this section, unexplained syncope is considered a risk factor for ventricular tachyarrhythmias		

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Treatment of syncope: **Unexplained syncope in patients at high risk of SCD (II)**

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Recommendations	Class	Level
Hypertrophic cardiomyopathy		
1. It is recommended that the decisions for ICD implantation in patients with unexplained syncope are made according to the ESC HCM Risk-SCD score http://www.doc2do.com/hcm/webHCM.html	I	B
2. Instead of an ICD, an ILR may be considered in patients with recurrent episodes of unexplained syncope with systolic impairment but without a current indication for ICD.	IIa	C
Arrhythmogenic right ventricular cardiomyopathy		
3. ICD implantation may be considered in patients with ARVC and a history of unexplained syncope.	IIb	C
4. Instead of an ICD, an ILR should be considered in patients with recurrent episodes of unexplained syncope with systolic impairment but without a current indication for ICD.	IIa	C
Unexplained syncope is defined as syncope that does not meet a Class I diagnostic criterion defined in the tables of recommendations. In the presence of clinical features described in this section, unexplained syncope is considered a risk factor for ventricular tachyarrhythmias.		

Treatment of syncope: **Unexplained syncope in patients at high risk of SCD (III)**

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Recommendations	Class	Level
Long QT syndrome		
1. ICD implantation in addition to beta-blockers should be considered in LQTS patients who experience unexplained syncopea while receiving an adequate dose of beta-blockers.	IIa	B
2. Left cardiac sympathetic denervation should be considered in patients with symptomatic LQTS when: (a) beta-blockers are not effective, not tolerated, or are contraindicated; (b) ICD therapy is contraindicated or refused; or (c) when patients on beta-blockers with an ICD experience multiple shocks.	IIa	C
3. Instead of an ICD, an ILR may be considered in patients with recurrent episodes of unexplained syncope with systolic impairment but without a current indication for ICD.	IIa	C
Unexplained syncope is defined as syncope that does not meet a class I diagnostic criterion defined in the tables of recommendations. In the presence of clinical features described in this section, unexplained syncope is considered a risk factor for ventricular tachyarrhythmias.		

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Treatment of syncope: **Unexplained syncope in patients at high risk of SCD (IV)**

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Recommendations	Class	Level
Brugada syndrome		
1. ICD implantation should be considered in patients with a spontaneous diagnostic type I ECG pattern and a history of unexplained syncope.	**IIa**	C
4. Instead of an ICD, an ILR may be considered in patients with recurrent episodes of unexplained syncope with systolic impairment but without a current indication for ICD.	**IIa**	C
Unexplained syncope is defined as syncope that does not meet a Class I diagnostic criterion defined in the tables of recommendations. In the presence of clinical features described in this section, unexplained syncope is considered a risk factor for ventricular tachyarrhythmias.		

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Syncope in patients with comorbidity and frailty

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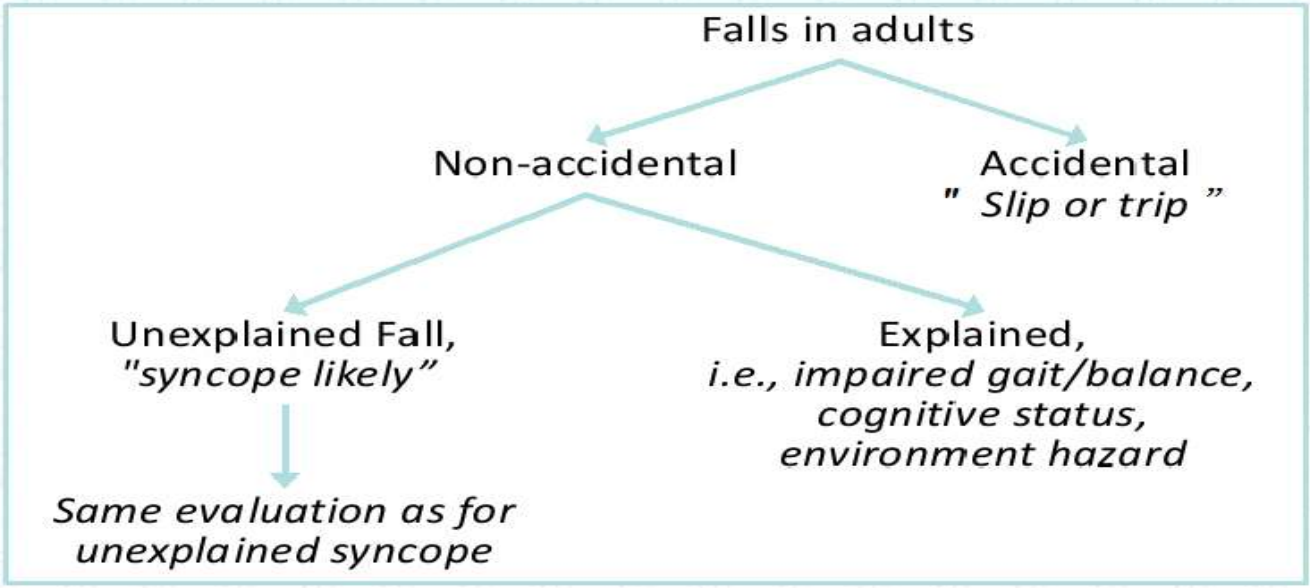

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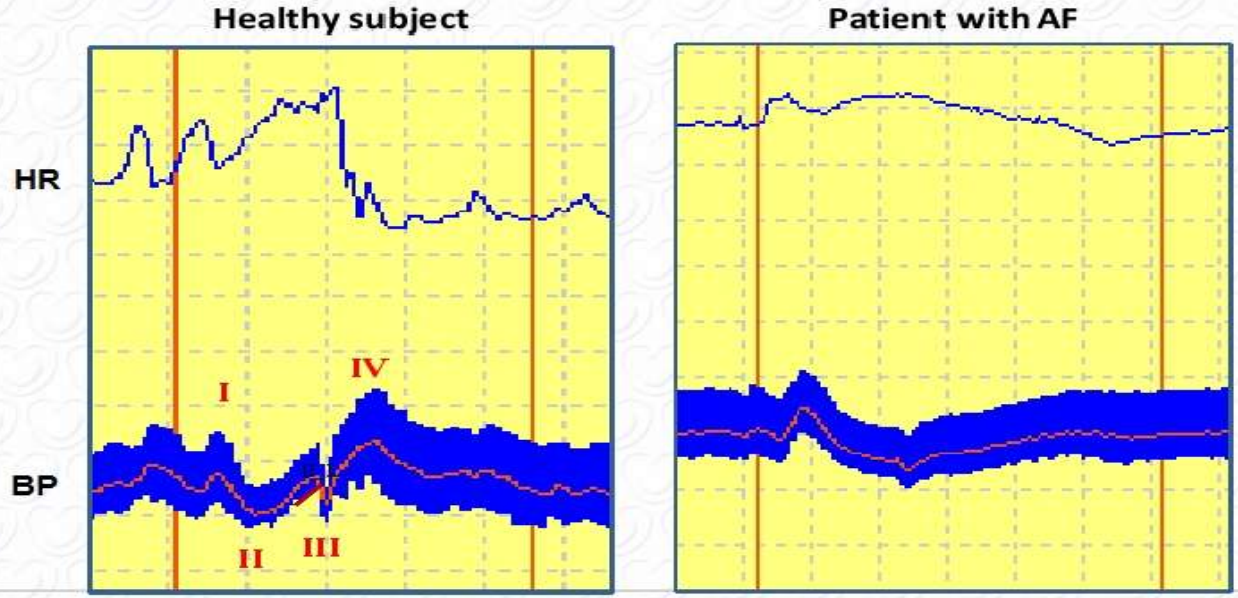
Supplemental Slides

Basic cardiovascular autonomic function tests

Valsalva manoeuvre



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Basic cardiovascular autonomic function tests

Valsalva manoeuvre

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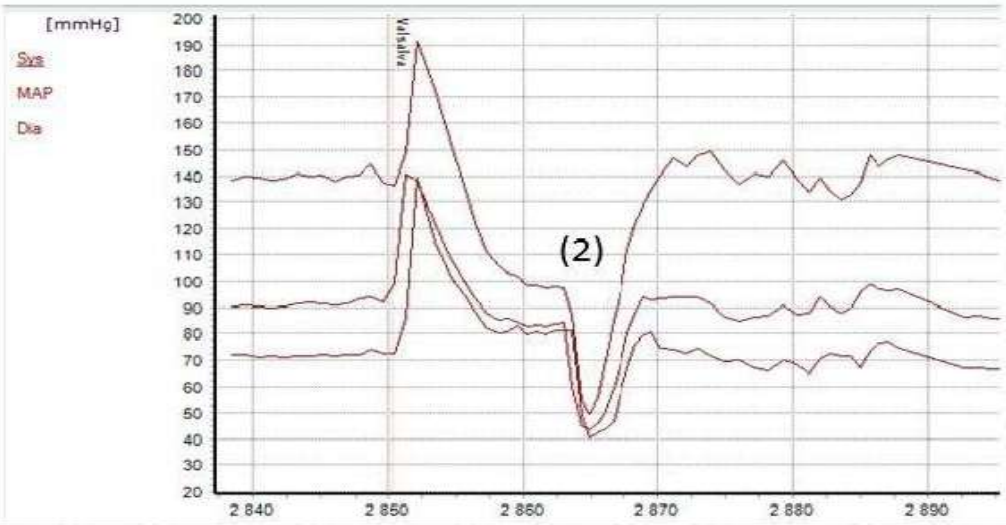
Patient with Situational syncope (e.g., cough)



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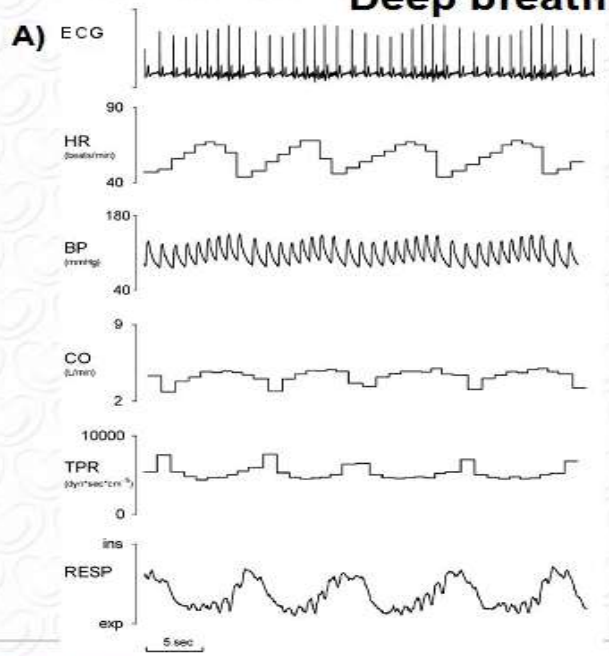
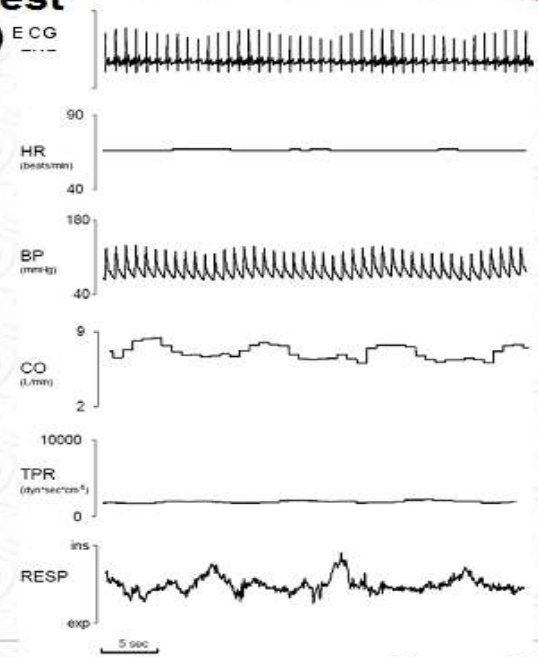
Basic cardiovascular autonomic function tests

Deep breathing test





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Basic cardiovascular autonomic function tests

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Recommendations	Class	Level
Valsalva manoeuvre		
1. Valsalva manoeuvre should be considered for assessment of autonomic function in patients with suspected neurogenic OH.	IIa	B
2. Valsalva manoeuvre may be considered for confirming the hypotensive tendency induced by some forms of situational syncope, e.g. cough, brass instrument playing, singing and weight lifting.	IIb	C
Deep breathing test		
3. Deep breathing test should be considered for assessment of autonomic function in patients with suspected neurogenic OH.	IIa	B
Other autonomic function tests		
4. Other autonomic function tests (30:15 ratio, cold pressure test, sustained hand grip test, and mental arithmetic test) may be considered for assessment of autonomic function in patients with suspected neurogenic OH.	IIb	C

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Tilt testing: **positivity rate**

92%	Typical VVS, emotional trigger (Clom)
78%	Typical VVS, situational trigger (TNG)
73%-65%	Typical VVS, miscellaneous (Clom) (TNG)
56%-51%	Likely reflex, atypical (TNG)
47%	Cardiac syncope (TNG)
45%	Likely tachyarrhythmic syncope (Passive)
36%-30%	Unexplained syncope (TNG) (Clom)
13%-8%	Subjects without syncope (Passive) (Clom) (TNG)

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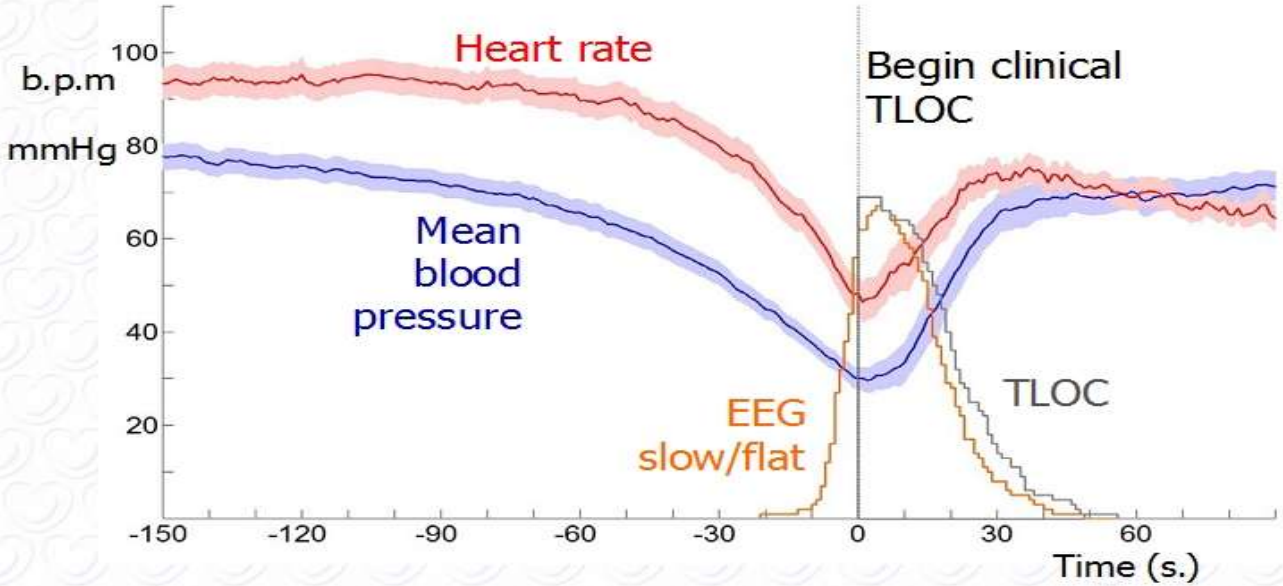
Tilt testing: **Reflex syncope**



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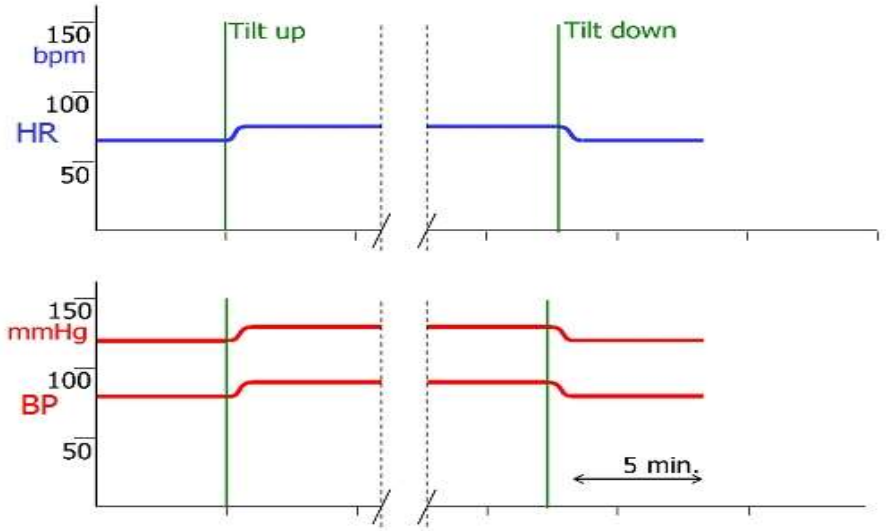
Tilt testing: **Normal result**



Normal tilt table test result

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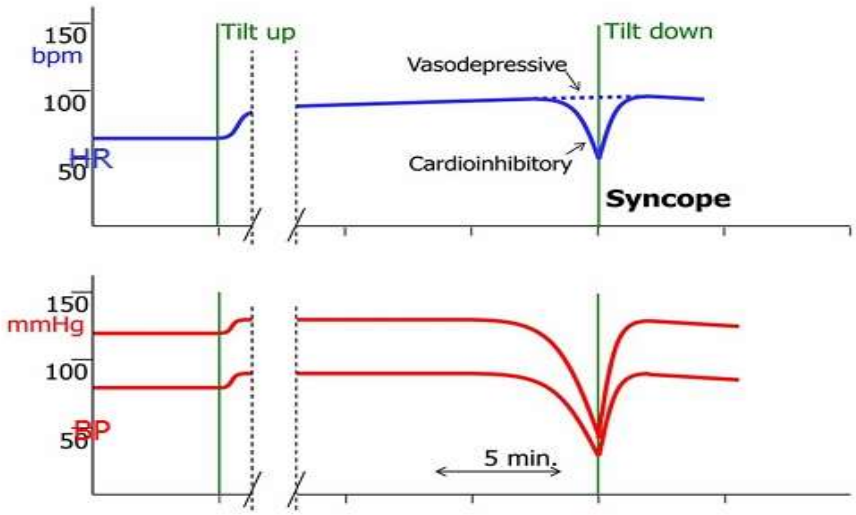
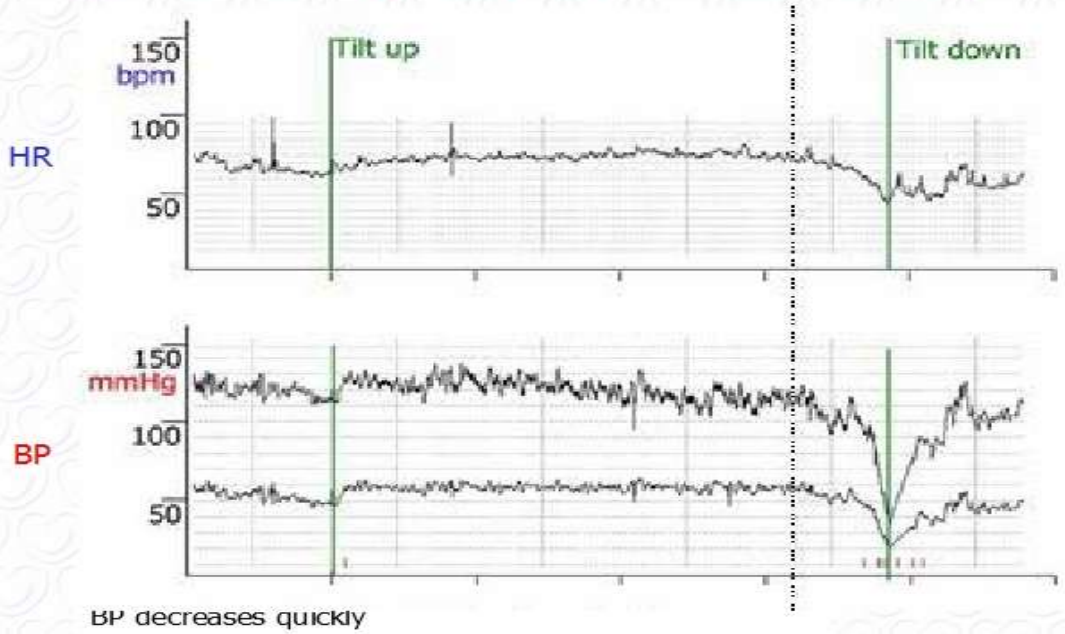
Tilt testing: **Reflex syncope**



Tilt-induced reflex syncope

Tilt testing: **Reflex syncope (mixed form)**



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Tilt testing: **Reflex syncope (asystolic form)**

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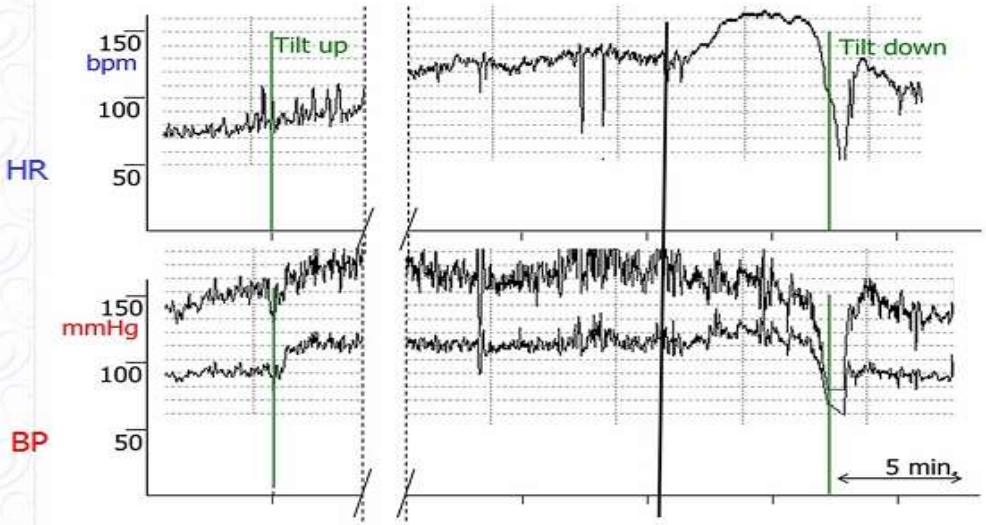

HR increases briefly when BP starts to decrease, but this makes way for a very steep decrease in HR, ending in asystole

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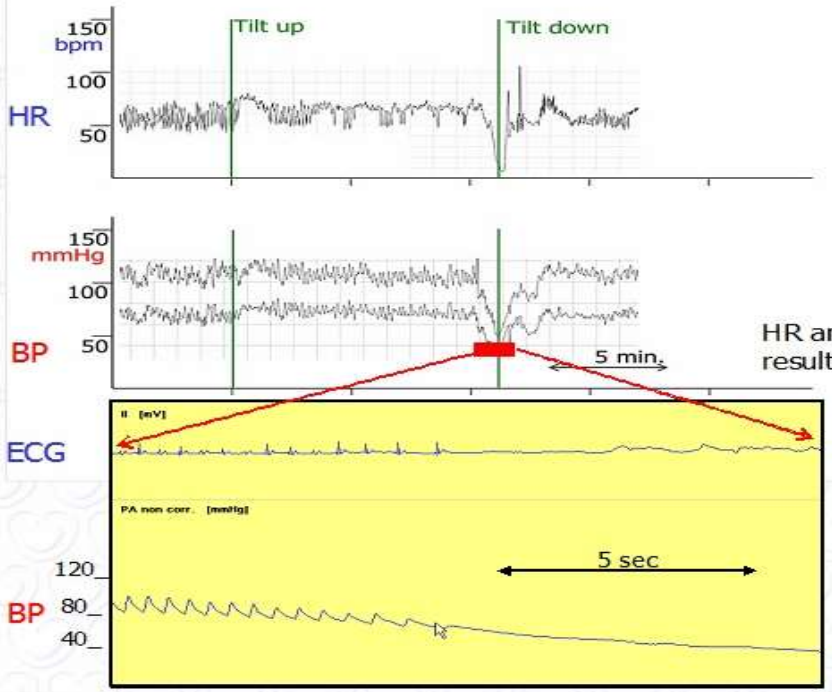
Tilt testing: **Reflex syncope (asystolic form)**



HR and BP fall quickly resulting in asystole

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Tilt testing: **Classical OH**

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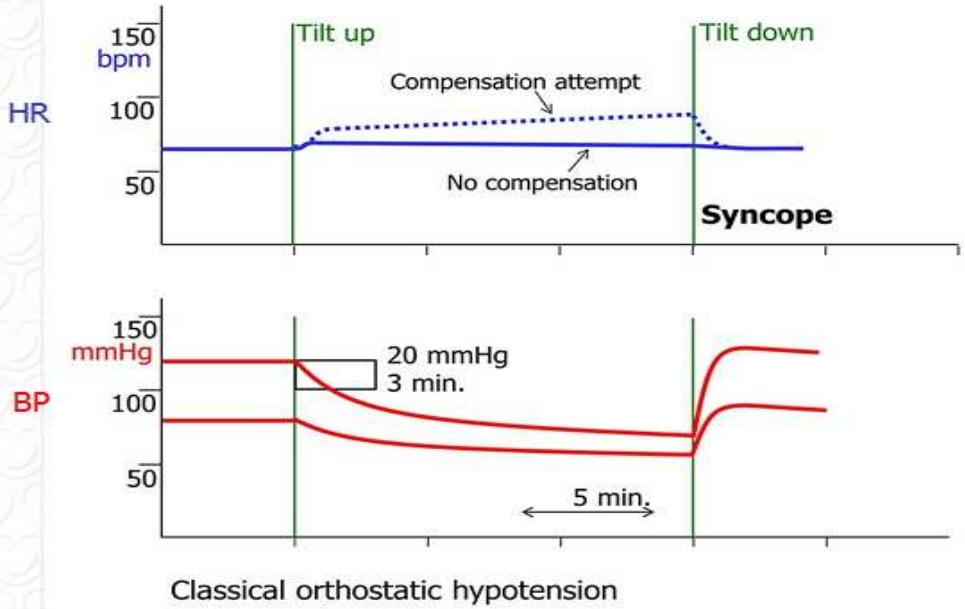

Classical orthostatic hypotension

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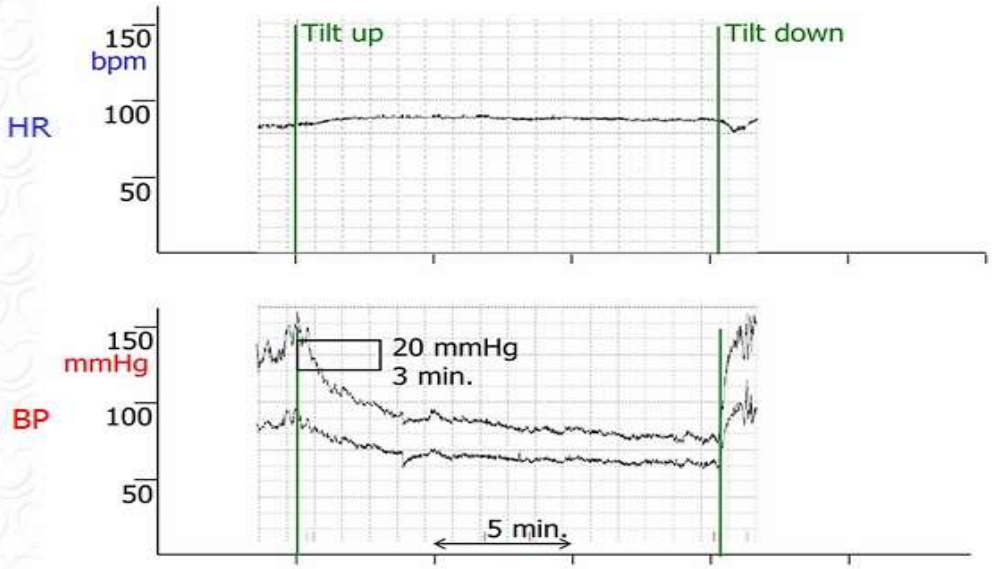
Tilt testing: **Classical OH**



Example #1 of classical orthostatic hypotension

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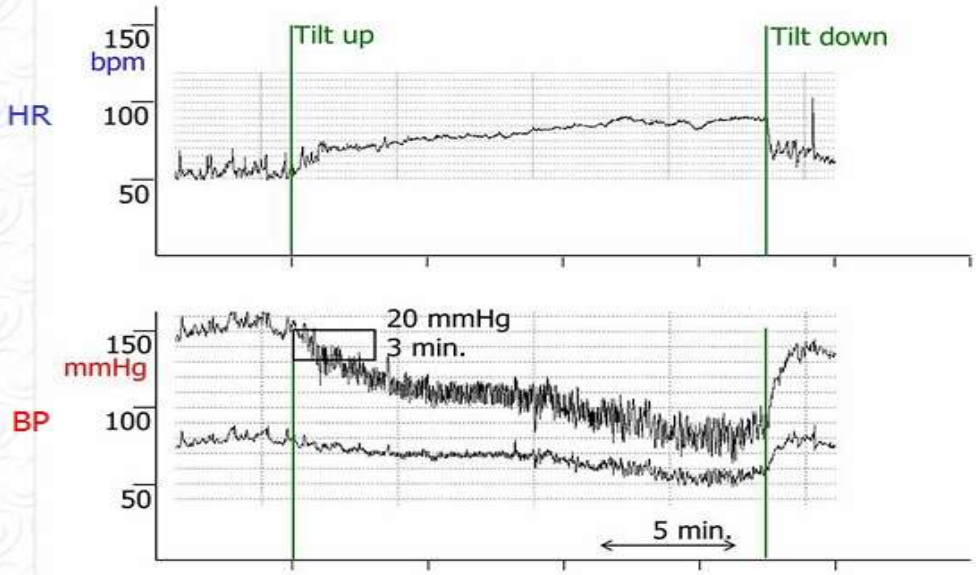
Tilt testing: **Classical OH**



Example #2 of classical orthostatic hypotension

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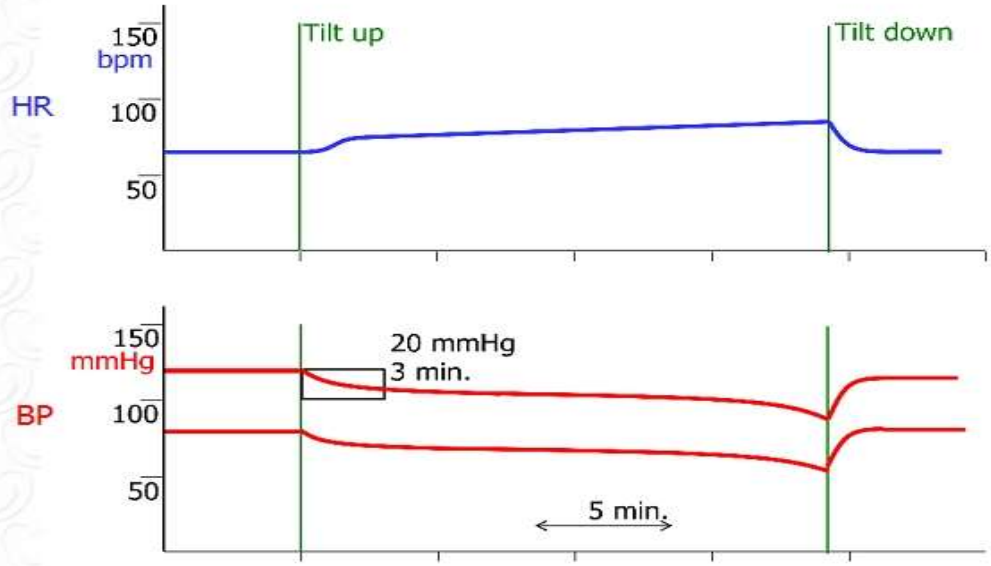
Tilt testing: **Delayed OH**



Delayed orthostatic hypotension

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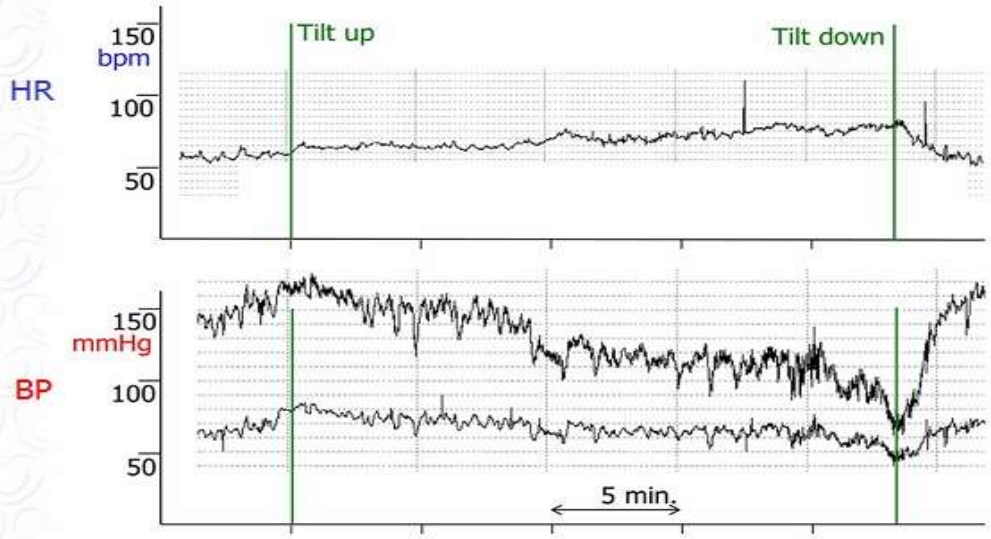
Tilt testing: **Delayed OH**



Example of delayed orthostatic hypotension

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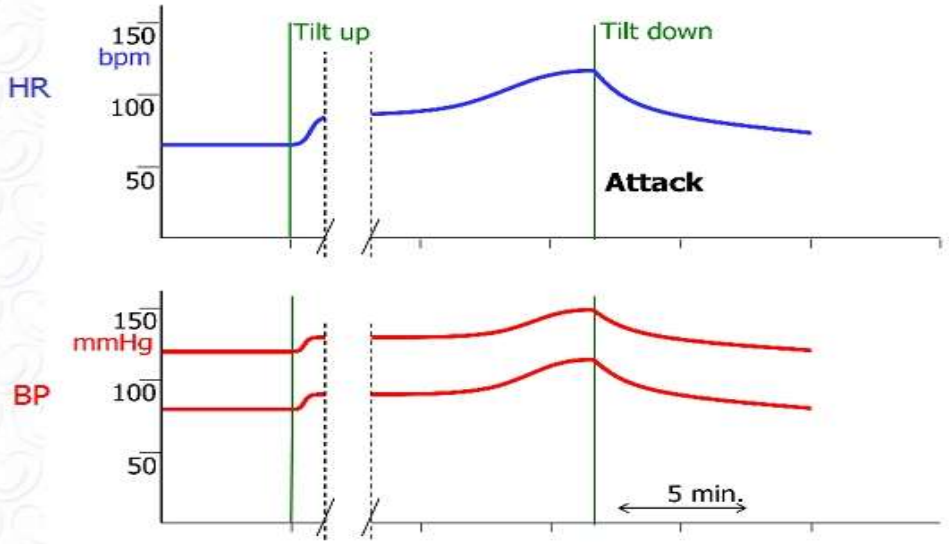
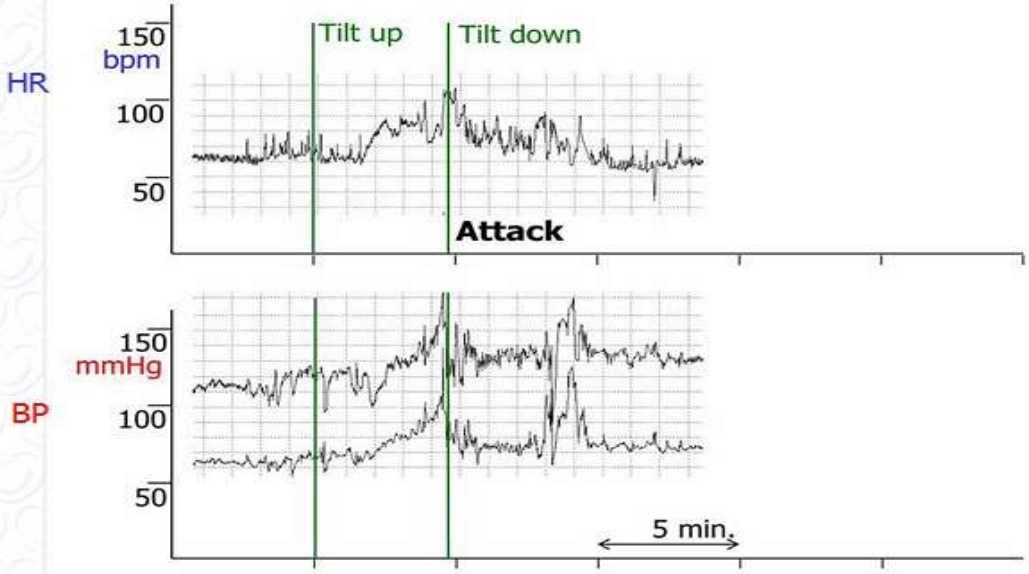
Tilt testing: **Psychogenic pseudosyncope**



Tilt testing: **Psychogenic pseudosyncope**



Example of psychogenic pseudosyncope

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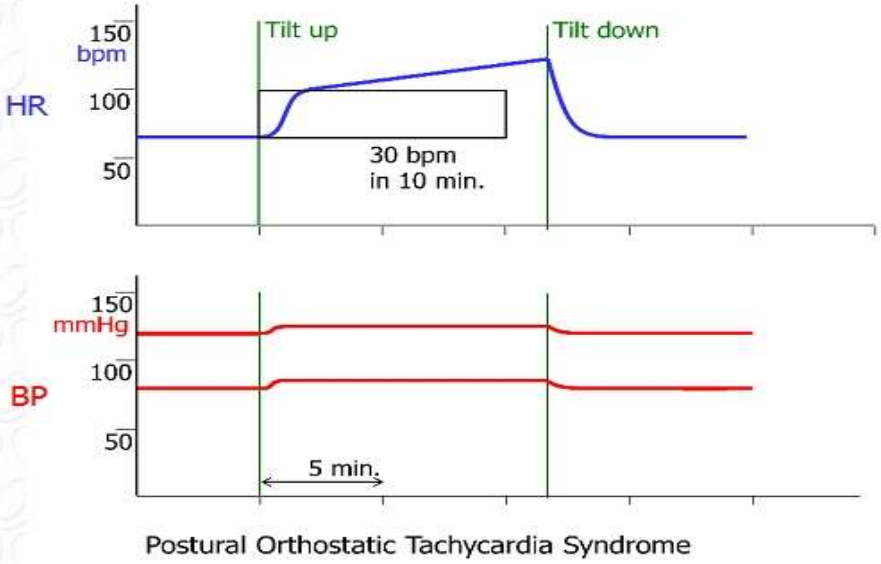
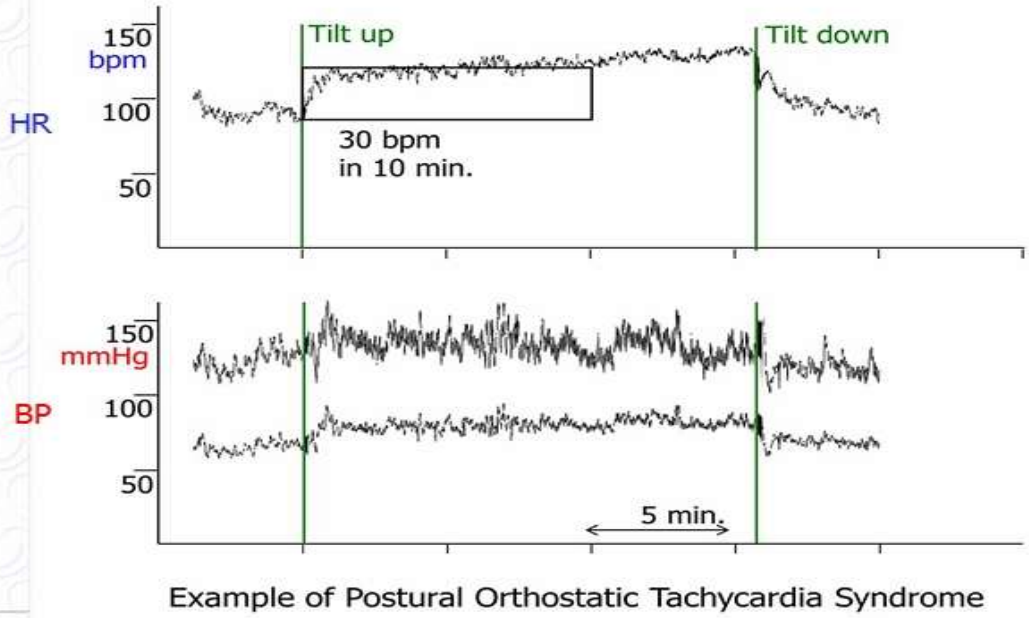
Tilt testing: **POTS**



Postural Orthostatic Tachycardia Syndrome

Tilt testing: **POTS**



Example of Postural Orthostatic Tachycardia Syndrome

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Tilt testing

Recommendations	Class	Level
Indications		
1. Tilt testing should be considered in patients with suspected reflex syncope, OH, POTS, or PPS.	IIa	B
2. Tilt testing may be considered to educate patients to recognize symptoms and learn physical manoeuvres.	IIb	B
Diagnostic criteria		
3. Reflex syncope, OH, POTS, or PPS should be considered likely if tilt testing reproduces symptoms along with the characteristic circulatory pattern of these conditions.	IIa	B

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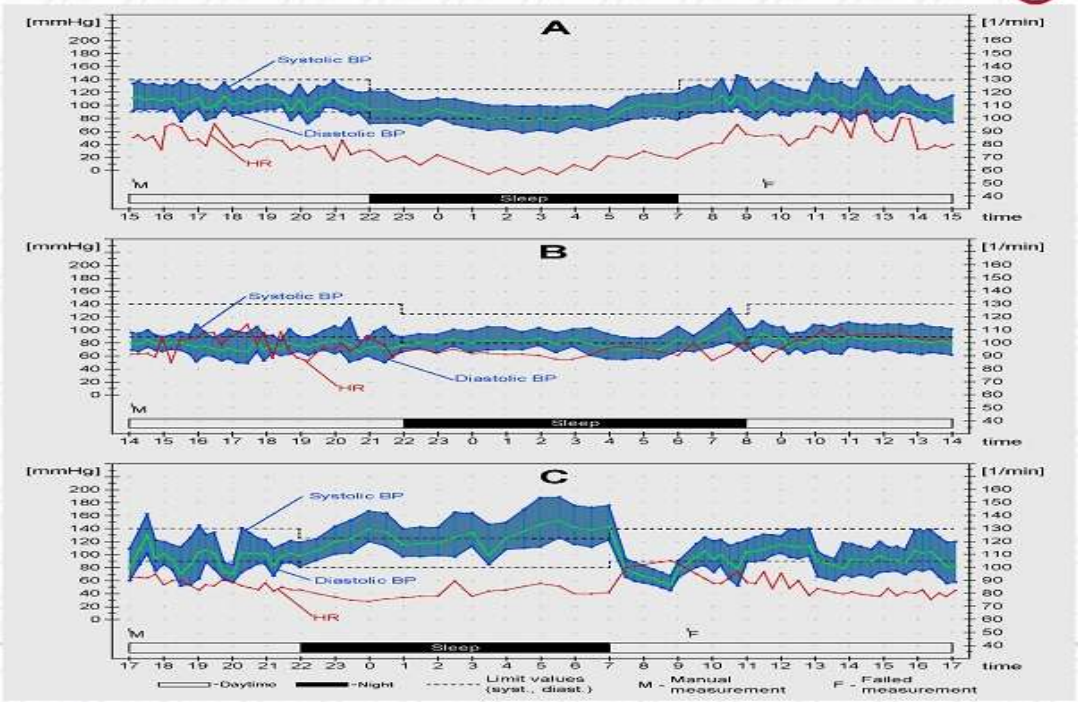
Basic cardiovascular autonomic function tests

ABPM

Nocturnal dipping

Non-dipping

Reverse dipping



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24-hour ambulatory blood pressure monitoring (ABPM)

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Recommendations	Class	Level
Indication		
1. ABPM is recommended to detect nocturnal hypertension in patients with autonomic failure.	I	B
2. ABPM should be considered to detect and monitor degree of OH and supine hypertension in daily life in patients with autonomic failure.	IIa	C
3. ABPM and home BP monitoring may be considered to detect whether BP is abnormally low during episodes suggestive of orthostatic intolerance.	IIb	C

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Video recording

Recommendations	Class	Level
1. Home video recordings of spontaneous events should be considered. Physicians should encourage patients and their relatives to obtain home video recordings of spontaneous events.	IIa	C
2. Adding video recording to tilt testing may be considered in order to increase reliability of clinical observation of induced events.	IIb	C

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Treatment of syncope: **General principles**

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Recurrence of syncope in untreated patients in RCT

Reference	Aetiology	Syncopes before evaluation	Syncopes after evaluation (%)
VPS I	VVS -Tilt +	6 (3–40) last 1 year	70% at 1 year
PC-Trial	VVS	3 (2–5) last 2 years	51% at 14 months
VASIS-Etilefrine	VVS -Tilt +	4 (3–17) last 2 years	24% at 1 year
POST	VVS - Tilt +	3 (1–6) last 1 year	35% at 1 year
Madrid *et al*	VVS - Tilt +	Median 3 per year	46% at 1 year
VPS II	VVS - Tilt +	4 (3–12) last 1 year	40% at 6 months
SYNPACE	VVS - Tilt +	4 (3–6) last 6 months	44% at 1 year
VASIS	Reflex – CI tilt +	3 (3–4.5) last 2 years	50% at 2 years
SPAIN	Reflex – CI tilt +	>5 during life	46% at 2 years
ISSUE 3	Reflex	5 (3–6) last 2 years	57% at 2 years
ATP Study	Unexplained – ATP +	Na	69% at 2 years
PRESS	Cardiac – BBB	1 last 6 months	14% at 2 years
THEOPACE	Sick sinus syndrome	3.2 ± 4.3	30% at 4 years

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ESC information sheet for patients affected by reflex syncope (1)

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Actions to take to avoid an impending attack of reflex syncope

- When you feel symptoms of syncope coming on, the best response is to lie down. If this is not possible, then sit down and do counter manoeuvres. The final warning symptom is when everything goes dark and you lose vision: then you *only have seconds in which to prevent syncope*.
- Your doctor will have shown you how to do the counter manoeuvres. They all concern tensing large muscles in the body. One way is to press the buttocks together and straighten the knees forcefully; another is to cross your legs and press them together over their entire length. Others make fists and tense the arm muscles.
- Drink around 2 litres of fluid a day and do not use salt sparingly (unless there are medical reasons not to!). A simple way to tell your fluid intake is high enough is to *check the colour of your urine*: if it is dark yellow there is little fluid in your body, so try to keep it very lightly coloured.
- Inform those in your immediate surroundings what to do during a spell: in typical spells there is *no need to call a doctor or an ambulance*. Of course, if you hurt yourself in the fall, this may change.

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Treatment syncope: **Counterpressure manoeuvres**

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Treatment syncope: **Counterpressure manoeuvres**

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Cardiac pacing in different settings (1)

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Setting/ condition	Diagnostic tool	Bradycardic mechanism of syncope	Recurrence of syncope with pacing	*Reference*
Documented paroxysmal AVB	ECG (standard or prolonged monitoring)	Established	**0%** at 3.5 yrs **0%** at 4 yrs **1%** at 5 yrs **7%** at 5 yrs	*Sud Brignole* *Aste* *Langenfeld H*
BBB-positive EPS	Positive EPS	Likely	**≈7%** at 2 yrs	*B4*
BBB-empirical pacing	Clinical evaluation	Suspected	**13.5%** at 2 yrs **14%** at 5 yrs	*PRESS* *Aste*
Sick sinus syndrome	Clinical evaluation	Suspected	**15%** at 5 yrs **22%** at 5 yrs **28%** at 5 yrs	*Sgarbossa* *DANPACE* *Langenfeld*

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Cardiac pacing in different settings (2)

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Setting/ condition	Diagnostic tool	Bradycardic mechanism of syncope	Recurrence of syncope with pacing	*Reference*
Carotid sinus syndrome (cardio-inhibitory form)	Carotid sinus massage	Likely	**10%** at 1 yr **11%** at 5 yrs **16%** at 3 yrs **16%** at 4 yrs **20%** at 5 yrs	*Claesson Lopes* *SUP 2* *Brignole* *Gaggioli*
Tilt-induced syncope (asystolic form)	Tilt test	Likely	**6%** at 5 yrs **7%** at 3 yrs **9%** at 2 yrs **23%** at 3 yrs	*VASIS-PM SYDIT* *SPAIN SUP 2*
Asystolic pause, no structural heart disease	ECG (standard or prolonged monitoring)	Established	**12%** at 2 yrs **24%** at 3 yrs **25%** at 2 yrs	*ISSUE 2* *SUP 2* *ISSUE 3*

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Cardiac pacing in different settings (3)

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Setting/ condition	Diagnostic tool	Bradycardic mechanism of syncope	Recurrence of syncope with pacing	*Reference*
Unexplained syncope	ATP test	Suspected	**23%** at 3 yrs	*ATP*
Tilt-induced Syncope (non-asystolic form)	Tilt test	Possible	**22%** at 1 yr **33%** at 6 months **44%** at 1 yr	*VPS I* *VPS II* *SYNPACE*

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Treatment of syncope: **General principles**

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Cardiac pacing in different clinical settings

Expected 2-year syncope recurrence rate	Clinical setting	
High efficacy (≤5% recurrence rate)	Established bradycardia	no hypotensive mechanism
Moderate efficacy (5% to 25% recurrence rate)	Established bradycardia	*and* hypotensive mechanism
Low efficacy (>25% recurrence rate)	Suspected bradycardia	*and* hypotensive mechanism

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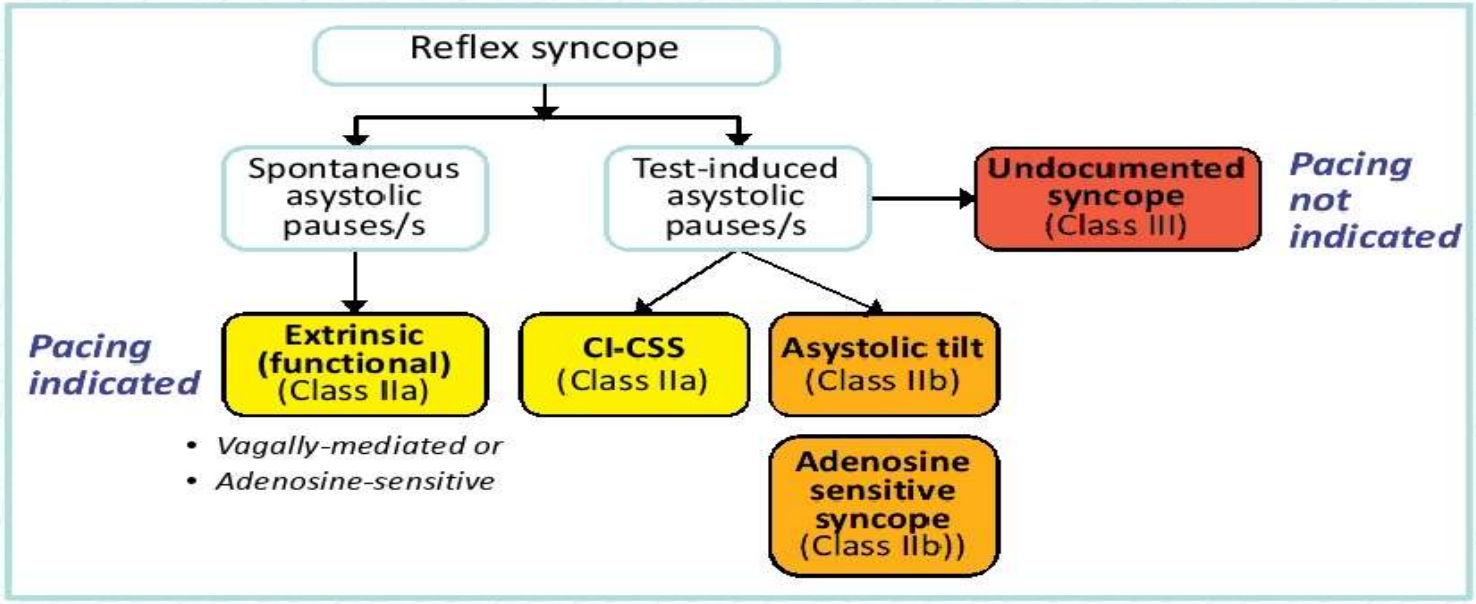
Pacing for reflex syncope



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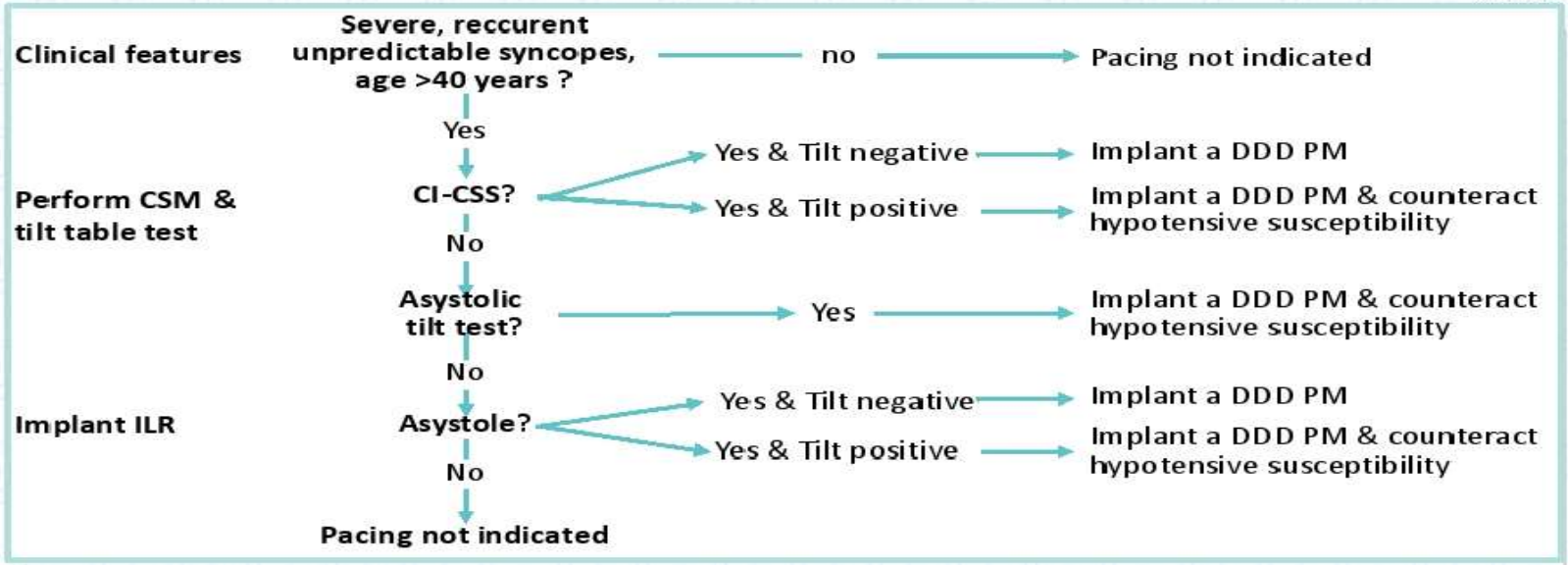
Pacing for reflex syncope: decision pathway



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Treatment of **Reflex syncope (III)**

Recommendations	Class	Level
Cardiac pacing		
1. Cardiac pacing should be considered to reduce syncopal recurrences in patients aged >40 years, with spontaneous documented symptomatic asystolic pause/s >3 seconds or asymptomatic pause/s >6 seconds due to sinus arrest or AV block or the combination of the two.	IIa	B
2. Cardiac pacing should be considered to reduce syncope recurrence in patients with cardioinhibitory carotid sinus syndrome who are >40 years with recurrent frequent unpredictable syncope.	IIa	B
3. Cardiac pacing may be considered to reduce syncope recurrences in patients with tilt-induced asystolic response who are >40 years with recurrent frequent unpredictable syncope.	IIb	B
4. Cardiac pacing may be considered to reduce syncope recurrences in patients with the clinical features of adenosine-sensitive syncope.	IIb	B
5. Cardiac pacing is not indicated in the absence of a documented cardioinhibitory reflex.	III	B

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Syncope in patients with comorbidity and frailty

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Recommendations	Class	Level
1. A multifactorial evaluation and intervention is recommended in older patients because more than one possible cause for syncope and unexplained fall may be present.	I	B
2. Cognitive assessment and physical performance tests are indicated in older patients with syncope or unexplained fall.	I	C
3. Modification or discontinuation of possible culprit medications, particularly hypotensive drugs and psychotropic drugs, should be considered in older patients with syncope or unexplained fall.	IIa	B
4. In patients with unexplained fall, the same assessment as for unexplained syncope should be considered.	IIa	C

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Psychogenic pseudosyncope (PPS)

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Recommendations	Class	Level
Diagnosis		
1. Recording of spontaneous attacks with a video by eyewitness should be considered for diagnosis of PPS.	IIa	C
2. Tilt testing, preferably with concurrent EEG recording and video monitoring may be considered for diagnosis of PPS.	IIb	C
Management		
3. Doctors who diagnose PPS should present the diagnosis of PPS to the patients.	IIa	C
4. Cognitive behavioural therapy may be considered in the treatment of PPS if attacks persist after explanation.	IIb	C

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Psychogenic pseudosyncope (PPS)

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How to present diagnosis to the patient and relatives

- Relatives or colleagues should know what a typical attack looks like (usually patients look as if they are asleep but cannot be woken).
- Relatives or colleagues should know beforehand what to do during a typicalattack.
- The attacks are not a medical emergency, so it is not necessary to call an ambulance.
- The attacks will pass by themselves, but some patience is required.
- Patients may be moved during an attack, if necessary.
- While waiting for the attack to end, patients may be put in a comfortable position, such as lying on their side with a pillow under the head.
- People close to the patient may stay next to the patient and comfort them when they recover, as they are then often emotionally distressed.

Humility and empathy is needed with these patients !

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Neurological causes and mimics of syncope

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Differentiating syncope from epileptic seizures

Clinical feature	Syncope	Epileptic seizures
Useful features		
Presence of trigger	Very often.	Rare.
Nature of trigger	Emotions for VVS; specific trigger for situational syncope; standing for OH.	Flashing lights is best known; also range of rare triggers.
Prodromes	Atonomic activation in reflex syncope, light-headedness in OH, palpitations in cardiac syncope).	Epileptic aura: repetitive (includes *déjà vu*) Epigastric aura and/or an unusual unpleasant smell.
Myoclonus	• <10, irregular in amplitude, asynchronous, asymmetrical; • Starts after the onset of LOC.	• 20–100, synchronous, symmetrical, hemilateral. • The onset mostly coincides with LOC. • Clear long-lasting automatisms as chewing or lip smacking at the mouth.

Neurological causes and mimics of syncope

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Differentiating syncope from epileptic seizures

Clinical feature	Syncope	Epileptic seizures
Useful features *(contd)*		
Tongue bite	Rare, tip of tongue	Side of tongue (rarely bilateral)
Duration of LOC	10–30 seconds	May be many minutes
Confusion after attack	No understanding of situation for <10 seconds in most syncope,	Memory deficit, i.e. repeated questions without imprinting for many minutes
Features of limited utility		
Incontinence	Not uncommon	Common
Myoclonus	Very often	~60%,
Eyes open	Frequent	Nearly always
Fatigue and sleep afterwards	Common, particularly in children	Very common

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Neurological tests or autonomic failure

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History & First evaluation
Neurological examination

Isolated autonomic failure

- Anti-ganglionic acetylcholine receptor antibodies
- Neoplasm-associated antibodies (anti-Hu)
- ^{123}I-MIBG cardiac SPECT

Autonomic failure + peripheral neuropathy

- Nerve conduction studies
- Laboratory tests: blood cells count, fasting glucose, Hb1AC, anti SS-A and anti SS-B antibodies, neoplasm-associated antibodies (anti-Hu, anti-PCA-2, anti-CRMP-5), serum/ urinary protein electrophoresis, HIV.
- Punch skin biopsy
- Genetic testing: familial amyloid neuropathy, hereditary sensory-autonomic neuropathy (in case of positive family history)

Autonomic failure

+

CNS involvement

(parkinsonism, ataxia, cognitive impairment)

- Neuroimaging (MRI)
- Cognitive tests
- DAT scan

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Neurological evaluation and tests

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Recommendations	Class	Level
1. Neurological evaluation is indicated when syncope is due to autonomic failure to evaluate the underlying disease.	I	C
2. Neurological evaluation is indicated in patients in whom TLOC is suspected to be epilepsy.	I	C
3. Brain magnetic resonance imaging is recommended if neurological examination indicates Parkinsonism, ataxia, or cognitive impairment.	I	C
4. Screening for paraneoplastic antibodies and antiganglionic acetylcholine receptor antibodies is recommended in cases of acute or subacute onset of multidomain autonomic failure.	I	B
5. EEG, ultrasound of neck arteries, and computed tomography or magnetic resonance imaging of the brain are not indicated in patients with syncope.	III	B

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Organizational aspects: **Syncope Unit**

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Key components

- The syncope unit should take the lead in service delivery for syncope, and in education and training of healthcare professionals who encounter syncope.
- The syncope unit should be led by a clinician with specific knowledge of TLOC and additional necessary team members (i.e. clinical nurse specialist) depending on the local model of service delivery.
- The syncope unit should provide minimum core treatments for reflex syncope and OH, and treatments or preferential access for cardiac syncope, falls, psychogenic pseudosyncope, and epilepsy.
- Referrals should be directly from family practitioners, EDs, in-hospital and out-hospital services, or self-referral depending on the risk stratification of referrals. Fast-track access, with a separate waiting list and scheduled follow-up visits, should be recommended.
- Syncope units should employ quality indicators, process indicators, and desirable outcome targets.

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Organizational aspects: **Structure of the SU**

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Staffing of an SU is composed of:

1. One or more physicians of any specialty who are ***syncope specialists***.
2. A team comprised of professionals who will advance the care of syncope patients.

Equipment:

1. Essential Equipment/tests:
   - 12-lead ECG and 3-lead ECG monitoring,
   - non-invasive beat-to-beat blood pressure monitor,
   - tilt-table,
   - Holter monitors,
   - external loop recorders,
   - follow-up of implantable loop recorders (*),
   - 24-hour blood pressure monitoring,
   - Basic autonomic function tests.
2. Established procedures for:
   - Echocardiography
   - Electrophysiological studies
   - Stress test
   - Neuroimaging tests
3. Specialists' consultancies (cardiology, neurology, internal medicine, geriatric medicine, psychology)

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Organizational aspects: Test and assessments in a SU

Initial assessment	
History & physical evaluation 12-lead standard ECG	
Subsequent tests and assessments (only when indicated)	
Blood tests	Electrolytes, Haemoglobin, troponin, BNP, glucose, D-dimer, Hemogasanalysis/O2 saturation.
Provocative tests	Carotid sinus massage, Tilt table test.
Monitoring	External loop recording, Implantable loop recording, Ambulatory 1-7 days ECG monitoring, 24-48 hour BP monitoring.
Autonomic function tests	Standing test, Valsalva manoeuvre, deep breathing test.
Cardiac evaluation	Established procedures for access to echocardiogram, stress test, electrophysiological study, coronary angiography.
Neurological evaluation	Established procedures for access to neurological tests (CT, MRI, EEG, video-EEG).
Geriatric evaluation	Established procedures for access to fall risk assessment (cognitive, gait and balance, visual, environmental).
Psychological or psychiatric evaluation	Established procedures for access to psychological or psychiatric consultancy.

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Organizational aspects: **Role of physician and staff in a SU**

Procedure or test	**SU Physician**	**SU Staff**	**Non-SU personnel**
History taking	x		
Structured history taking (e.g., application of software technologies)		x	
12-lead ECG		x	
Blood tests		x	
Echocardiogram and imaging			x
Carotid sinus massage	x		
Active standing test		x	
Tilt table test	(x)	x	
Basic autonomic function test		x	
ECG monitoring (Holter, ELR): administration and interpretation	x	x	
Implantable loop recorder	x	(x)	
Remote monitoring		x	
Others: stress test, electrophysiological study, angiograms			x
Neurological tests (CT, MRI, EEG, video-EEG)			x
Pacemaker and ICD implantation, catheter ablation			x
Patient's education, biofeedback training. and instructions	x	x	
Final report and clinic note	x		
Communication with patients, referring physicians	x	x	
Follow-up	x	x	

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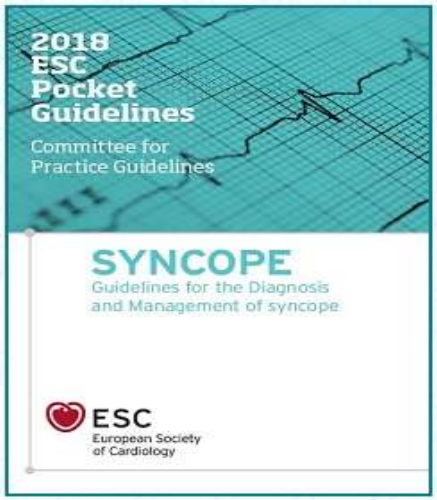

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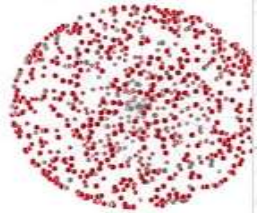

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