GENERAL UNIVERSITY HOSPITAL IN PRAGUE

FIRST FACULTY OF MEDICINE
Charles University

Doporučení 2022 - Komorové tachykardie

Štěpán Havránek

ESC European Society of Cardiology
European Heart Journal (2022) **00**, 1–130
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2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Developed by the task force for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric and Congenital Cardiology (AEPC)

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Základní změny guidelines

	2015	2022
Coronary artery disease		
In patients with syncope and previous STEMI, PES is indicated when syncope remains unexplained after non-invasive evaluation.	IIa	I
Intravenous amiodarone treatment should be considered for patients with recurrent PVT/VF during the acute phase of ACS.	I	IIa
In patients with CAD eligible for ICD implantation, catheter ablation may be considered just before (or immediately after) ICD implantation to decrease subsequent VT burden and ICD shocks.	IIa	IIb
PVC-induced cardiomyopathy		
In patients with a cardiomyopathy suspected to be caused by frequent and predominately monomorphic PVCs, catheter ablation is recommended.	IIa	I

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

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Základní změny guidelines

Inflammatory diseases		
In patients with haemodynamically not-tolerated SMVT occurring in the chronic phase of myocarditis, ICD implantation is recommended.	IIa	I
ICD implantation is recommended in patients with cardiac sarcoidosis who have an LVEF ≤35%.	IIb	I
ICD implantation is recommended in patients with cardiac sarcoidosis who (1) have documented sustained VT, or (2) aborted CA.	IIb	I
In patients with cardiac sarcoidosis who have an indication for permanent cardiac pacing related to high-degree AV block, ICD implantation should be considered, regardless of LVEF.	IIb	IIa

Primary electrical disease and selected populations		
ICD implantation is recommended in patients with LQTS who are symptomatic[b] while receiving beta-blockers and genotype-specific therapies.	IIa	I
ICD implantation should be considered in patients with CPVT who experience arrhythmic syncope and/or documented bidirectional/PVT while on the highest tolerated beta-blocker dose and on flecainide.	I	IIa
Pre-participation cardiovascular evaluation of competitive athletes should be considered.	I	IIa
Catheter ablation of triggering PVCs and/or RVOT epicardial substrate should be considered in BrS patients with recurrent appropriate ICD shocks refractory to drug therapy.	IIb	IIa
LCSD should be considered in patients with diagnosis of CPVT when the combination of beta-blockers and flecainide at therapeutic dosage are either not effective, not tolerated, or contraindicated.	IIb	IIa

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Diagnostika u nemocných s první manifestací KT bez známé kardiální anamnézy

Scénář 1: Náhodný záchyt nesetrvalé komorové tachykardie

Scénář 2: První manifestace setrvalé monomorfní komorové tachykardie

Scénář 3: Přeživší náhlou srdeční smrt

Scénář 4: Oběť náhlé srdeční smrti

Scénář 5: Příbuzný oběti náhlého úmrtí z arytmické příčiny

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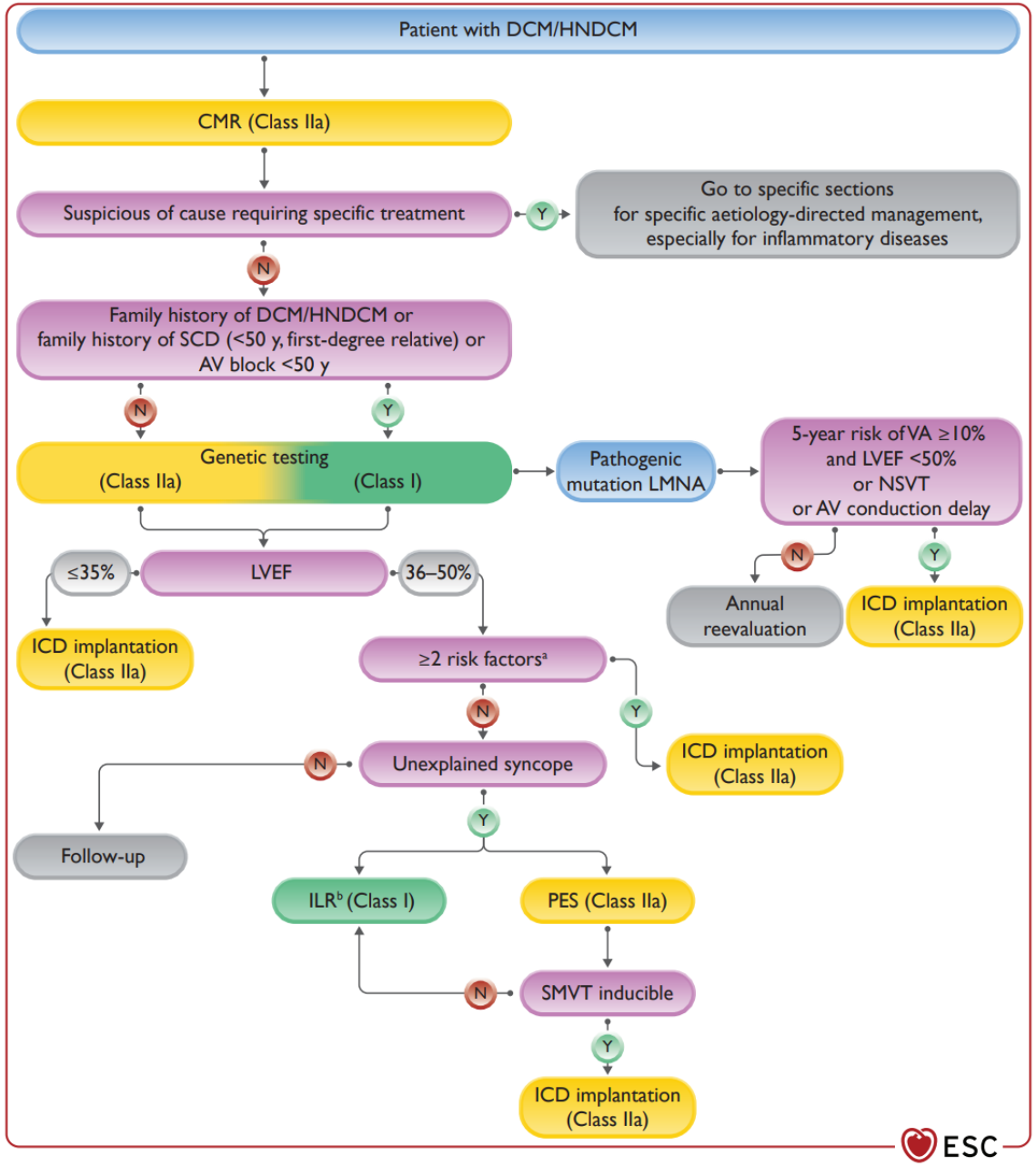
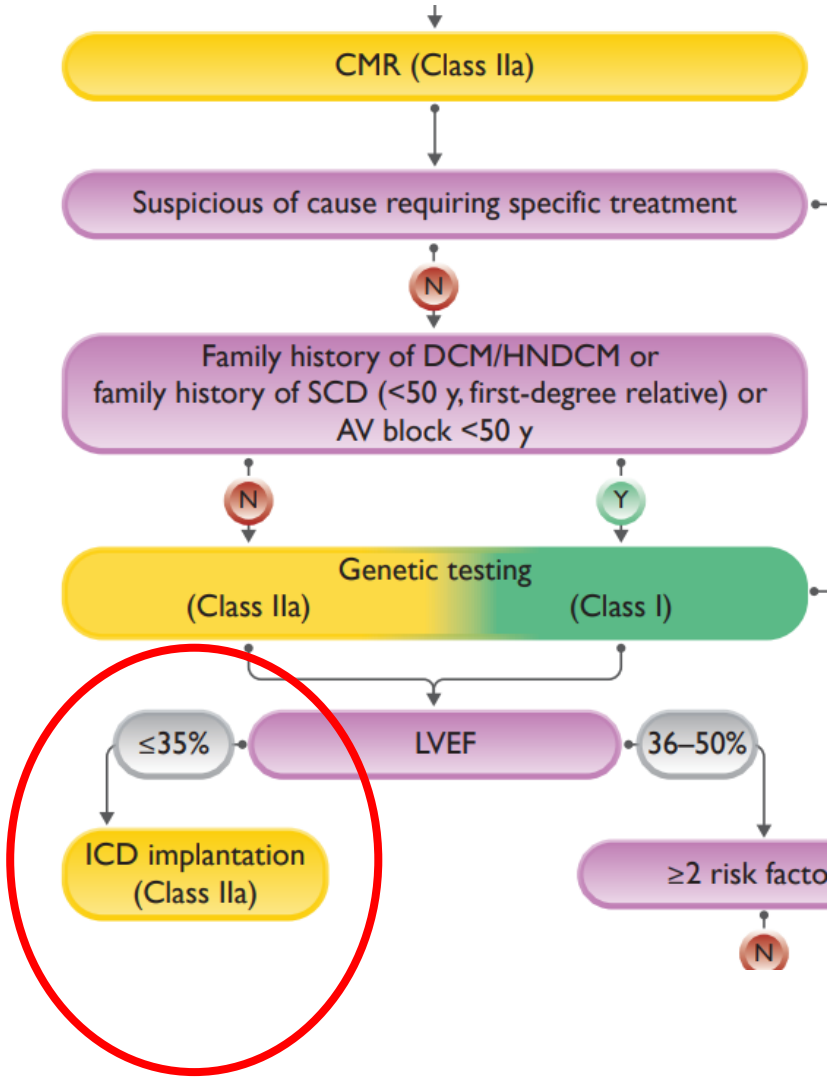
DKMP / HNDCM





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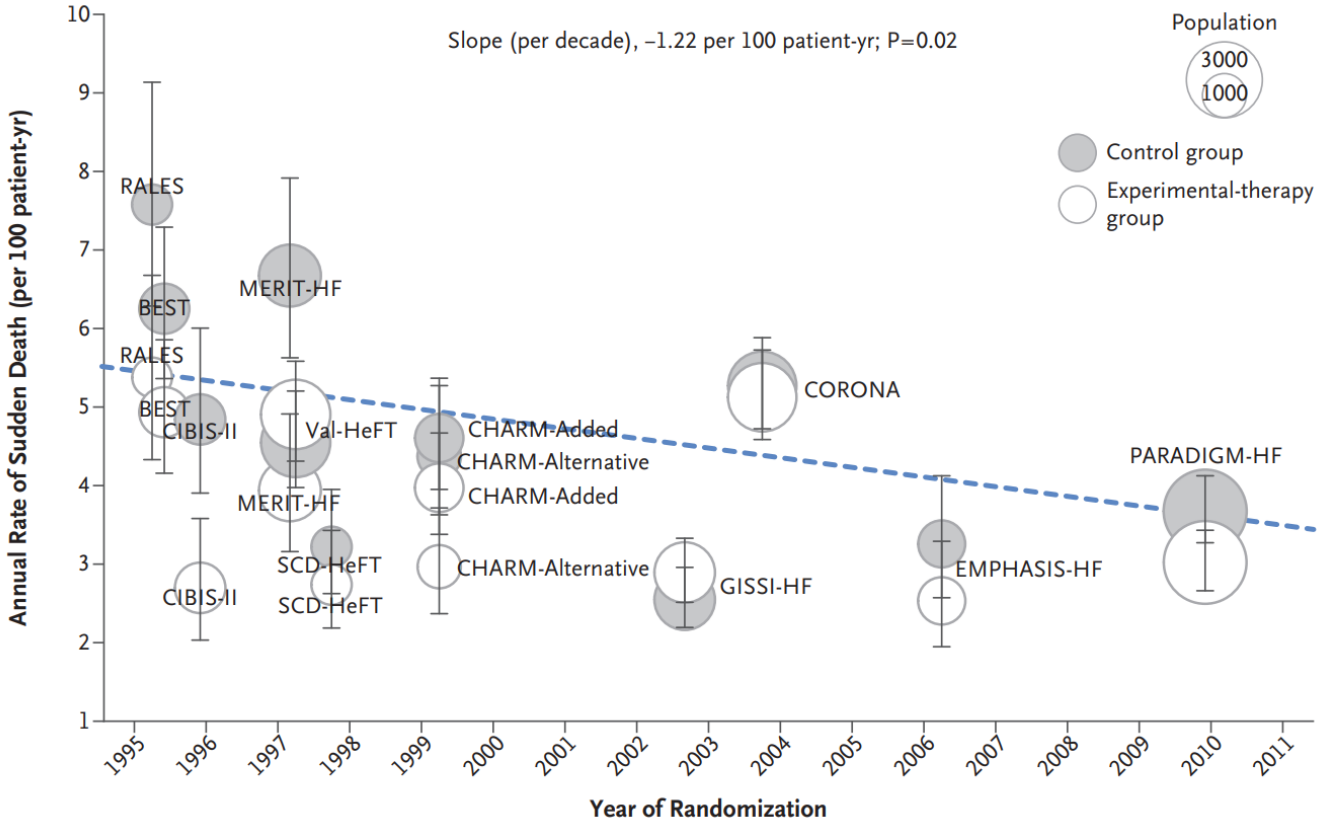
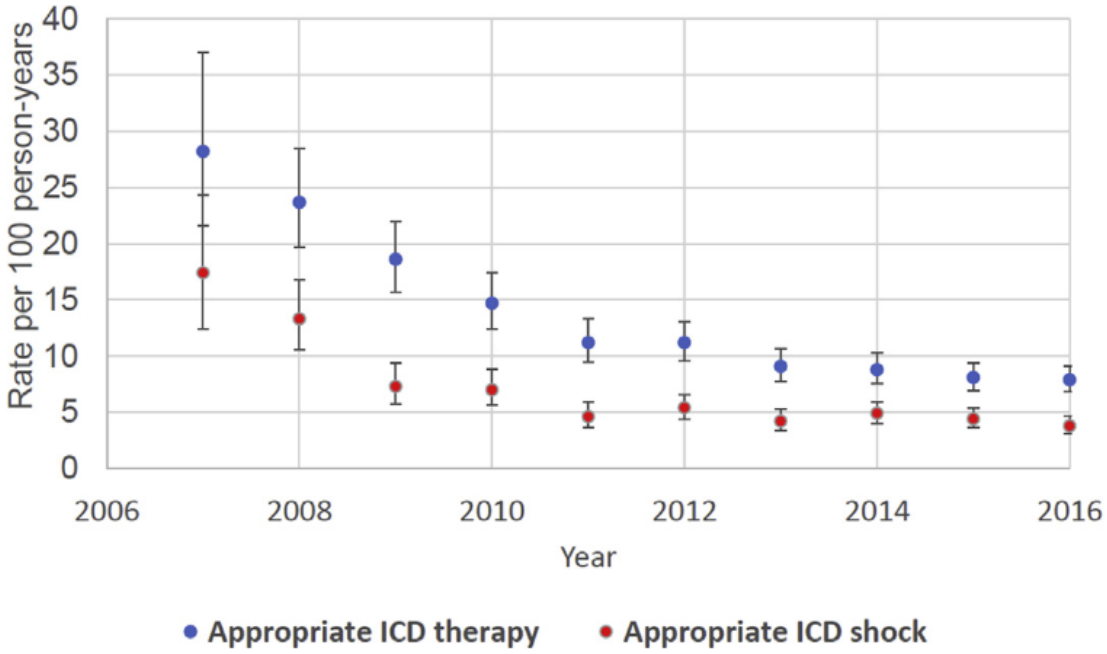
Pokles výskytu náhlé srdeční smrti u pacientů se srdečním selháním





	MADIT II	SCD-HeFT	DANISH
Rok publikace	2002	2005	2016
Number needed to treat	18	14	43

Moss AJ et al. NEJM 2002;346:877-883
Køber L et al. NEJM 2016;375:1221-30
Bardy GH et al. NEJM 2005;352:225-237

Shen L et al. NEJM 2017;377:41-51
Ruwald M et al. J Am Coll Cardiol EP 2021;7:781–92

Nová stratifikační kritéria EF 36 – 50%

Genetické vyšetření

LMNA, PLN, FLNC, RBM20

Magnetická rezonance

LGE

Synkopa

nevysvětlitelná

Programovaná stimulace

pozitivní

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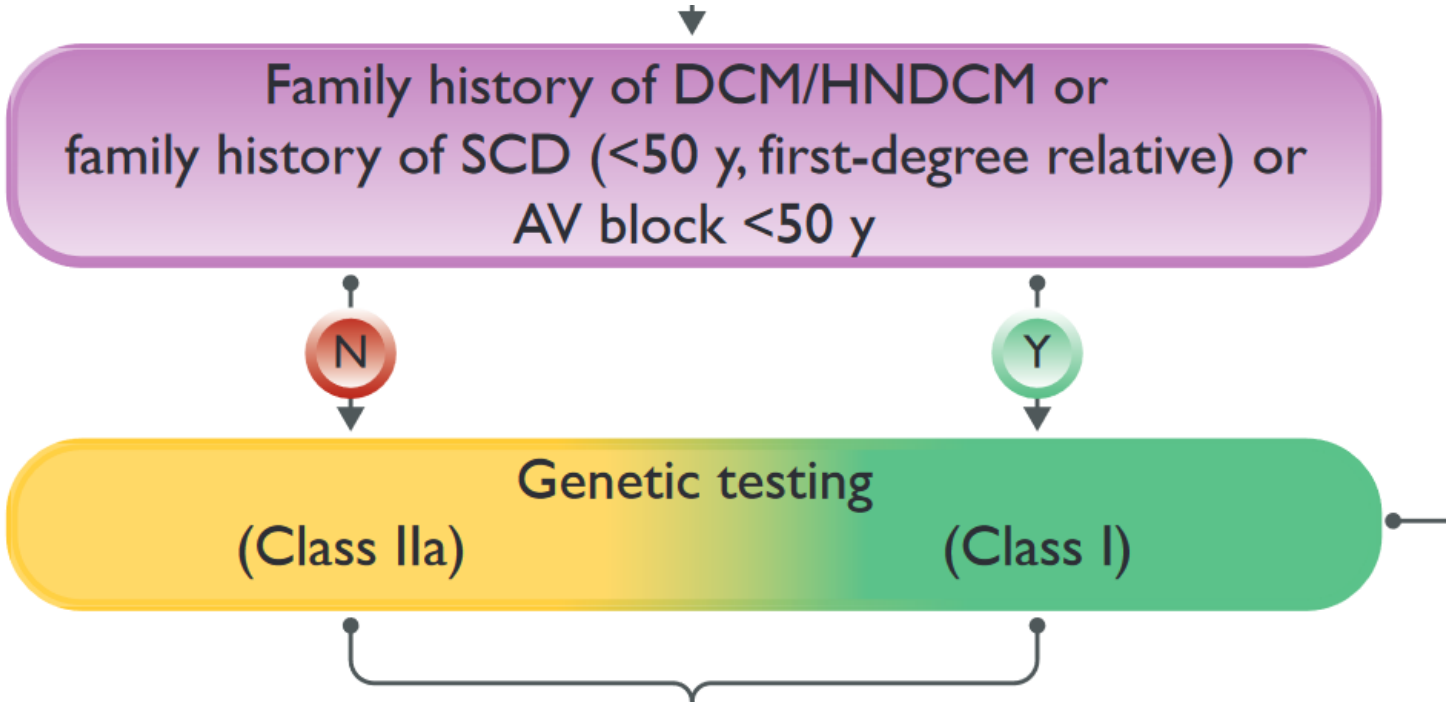
Genetické vyšetření



Lamin (LMNA)

Fosfolamban (PLN)

Titin (RBM20)

Filamin C (FLNC)

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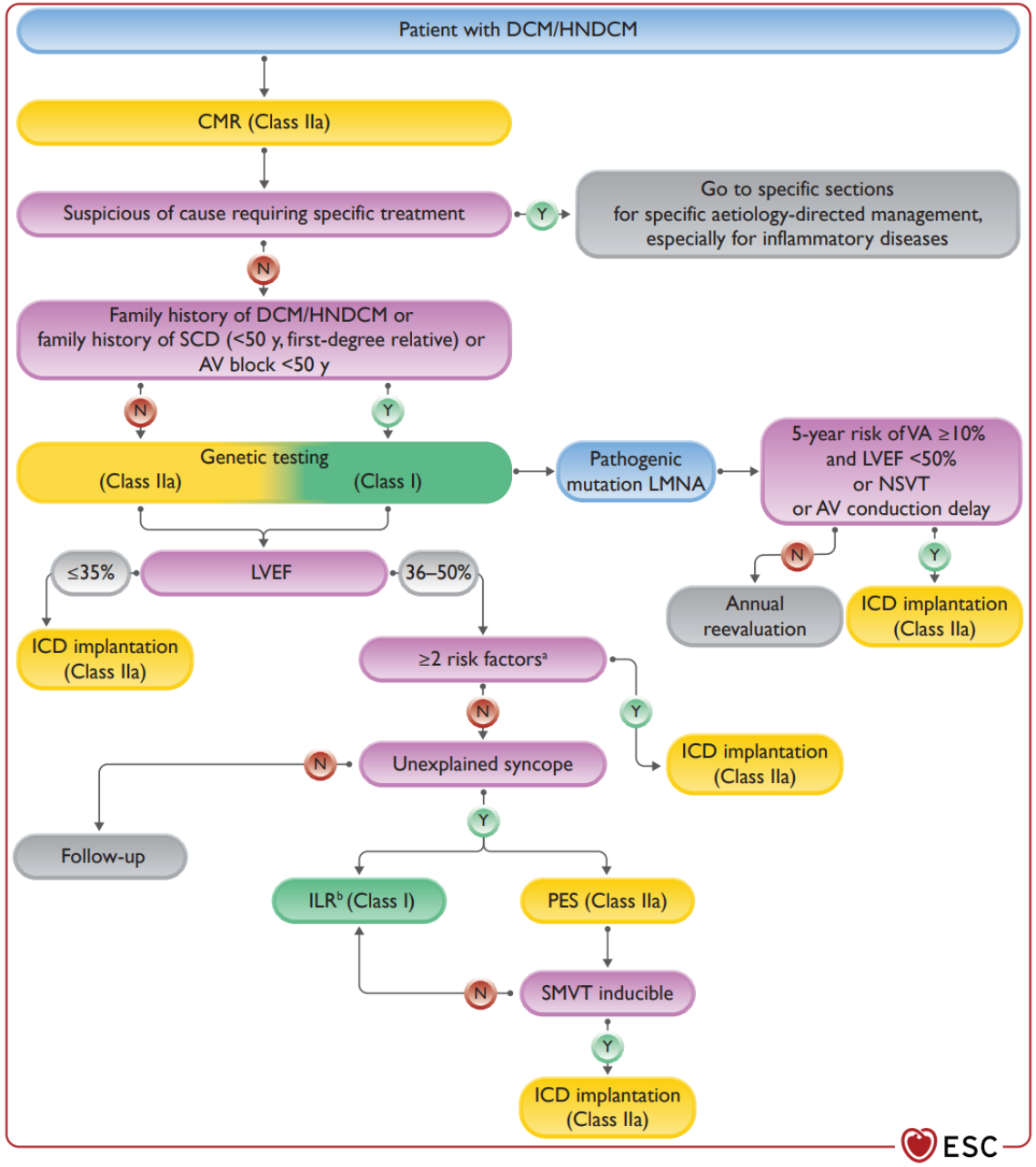
DKMP / HNDCM



LMNA-risk VTA calculator

Risk Prediction Score for Life-Threatening Ventricular Tachyarrhythmias in Laminopathies

Sex	◉ Male ○ Female
Non-missense LMNA mutation	○ Yes ◉ No
Atrio-ventricular block	○ Absent ◉ 1st degree ○ High degree
Non-sustained ventricular tachycardia	○ Yes ◉ No
Left ventricular ejection fraction	60 %

Risk of Life-Threatening Ventricular Tachyarrhythmias at 5 years

13.5 %

lmna-risk-vta.fr

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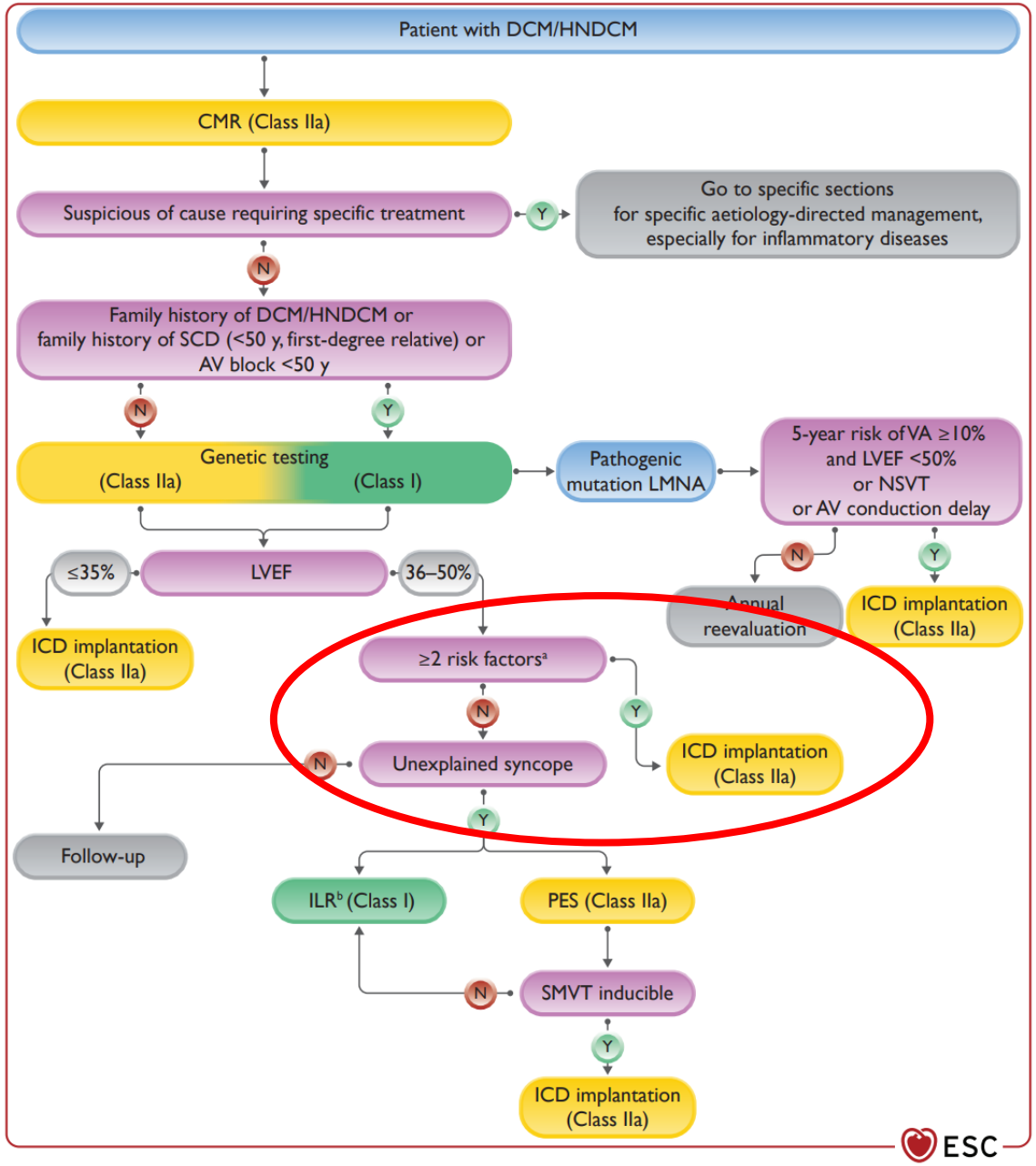

EF LK 36-50%

Rizikové faktory:

- Nevysvětlitelná synkopa
- Patogenní varianta:
  - Lamin (LMNA)
  - Fosfolamban (PLN)
  - Titin (RBM20)
  - Filamin C (FLNC)
- LGE na MRI
- Inducibilní komorová tachykardie

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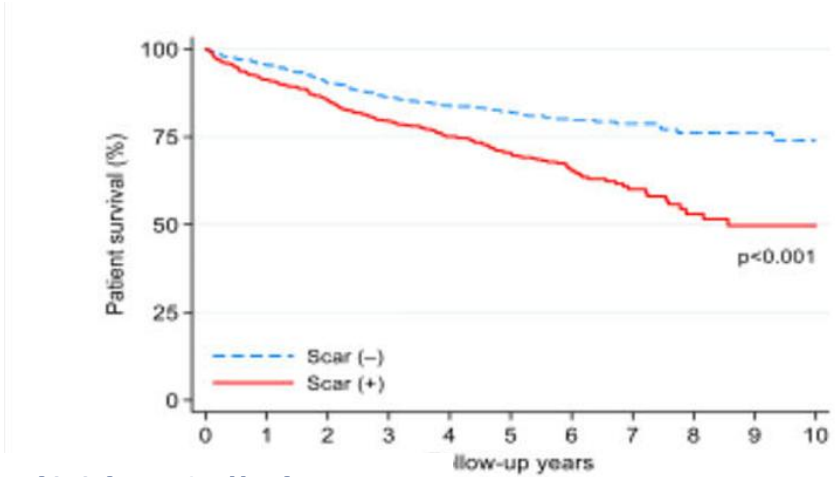
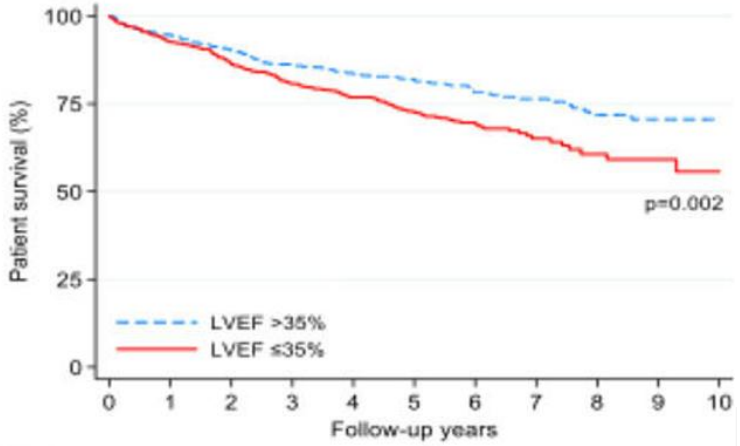
Význam LGE

N = 1020
DKMP

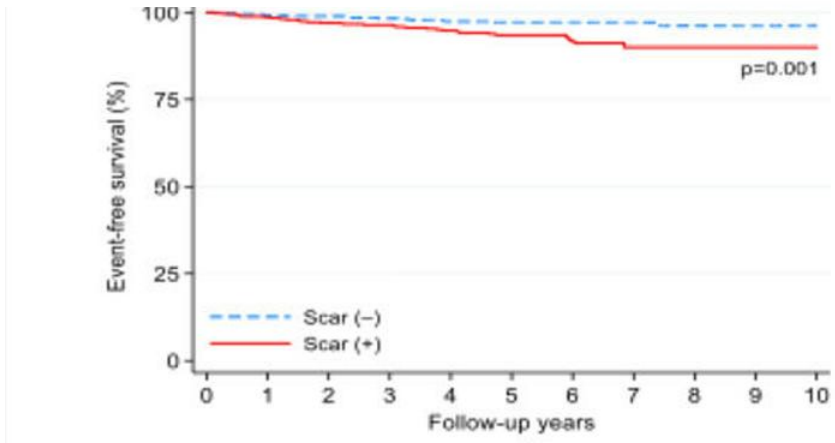
Celková mortalita **Scar (+) vs. Scar (-)**

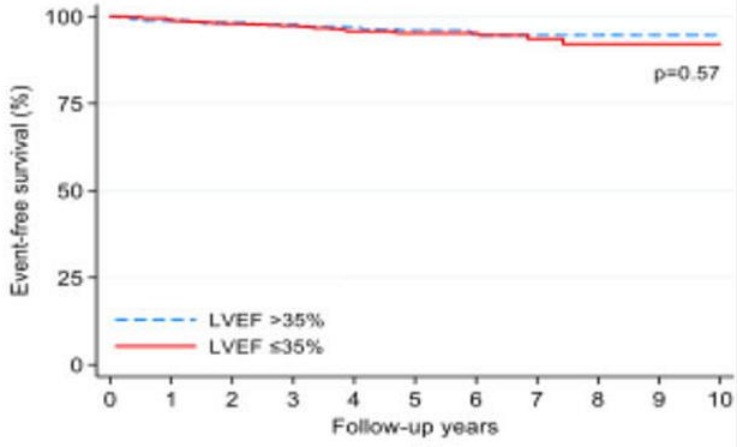


LVEF ≤ 35% vs. LVEF > 35%

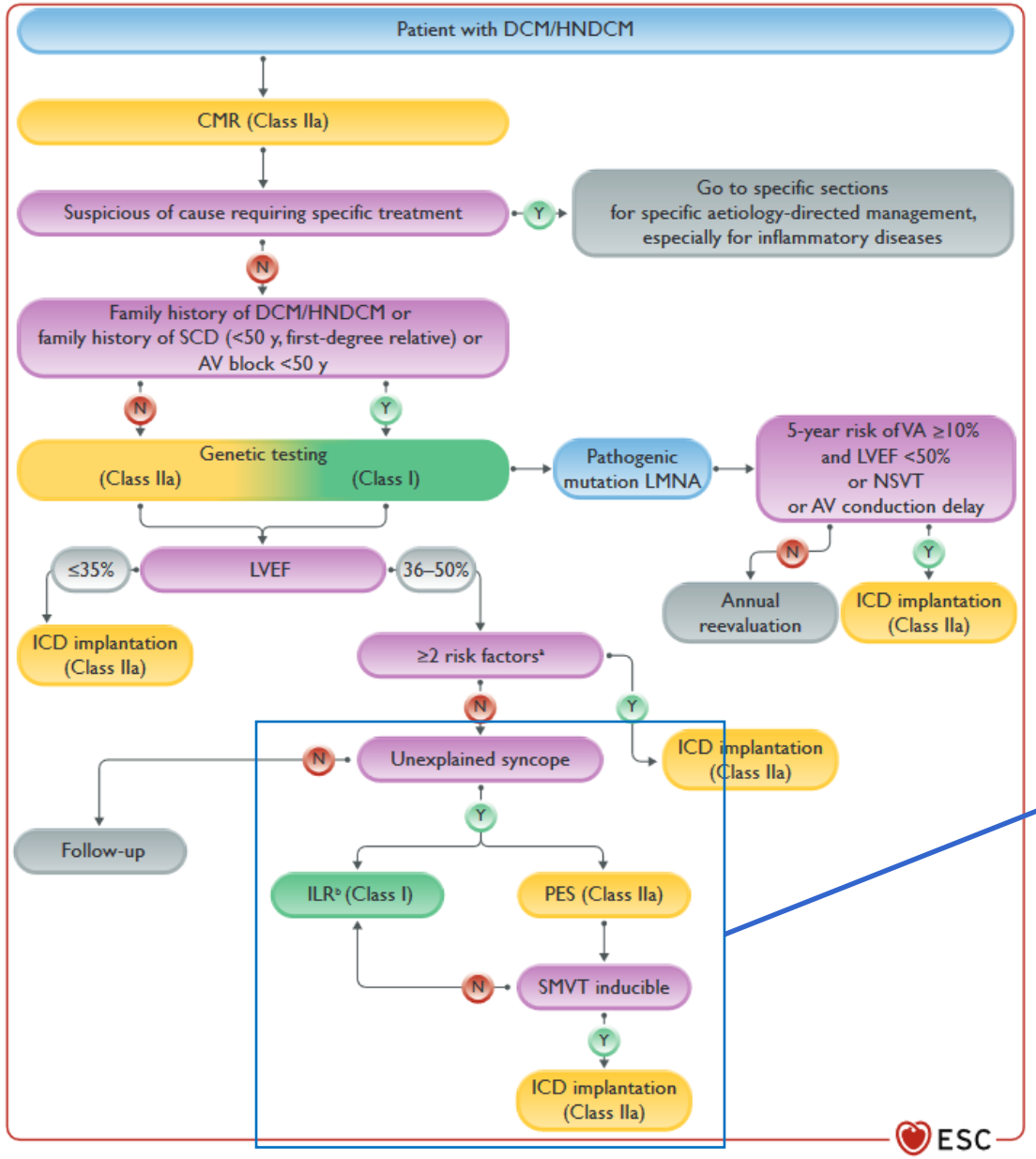


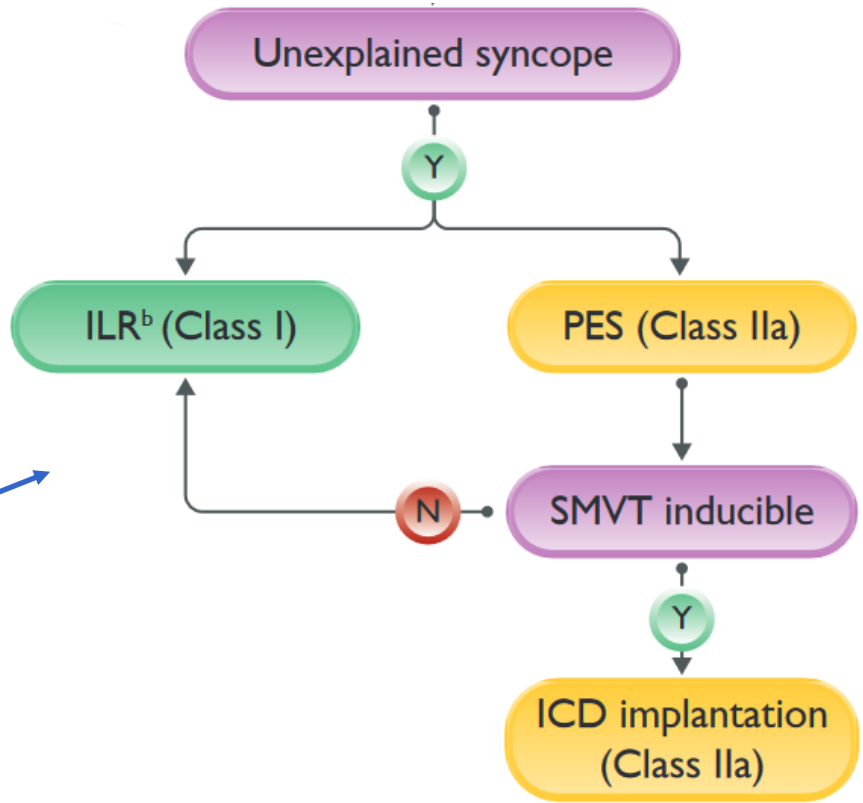
Náhlá srdeční smrt





Klem et al. Circulation 2021;143:1343–1358





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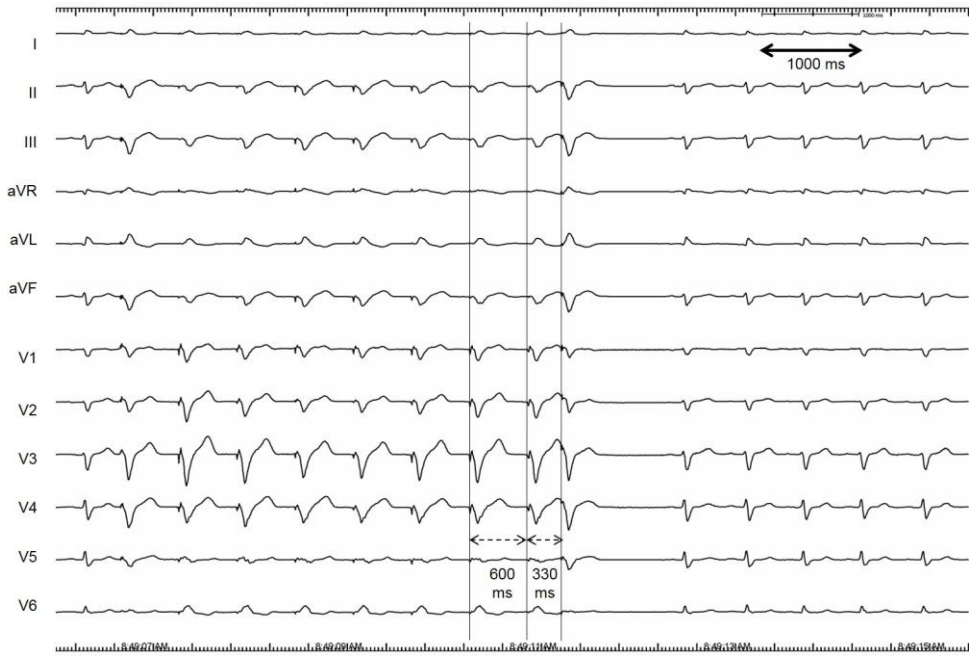
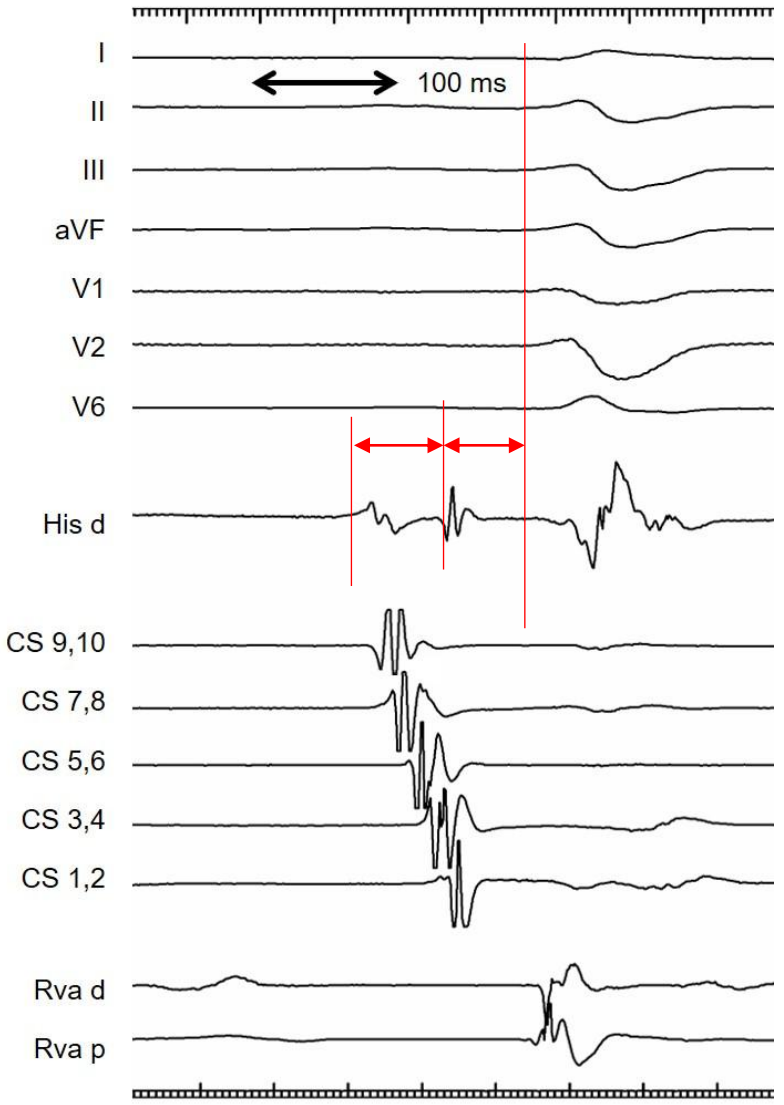
Elektrofyziologie & mapping

Součástí vyšetření

- Převodní intervaly AH, HV
- Programovaná stimulace
- Elektroanatomické mapování
- Podávání isoprenalinu

- Adenosin – rekonekce, provokace KES
- Hadgrip pro provokaci KES





Zdroj: Všeobecná fakultní nemocnice

2020 APHRS/HRS expert consensus statement on the investigation of decedents with sudden unexplained death and patients with sudden cardiac arrest, and of their families.
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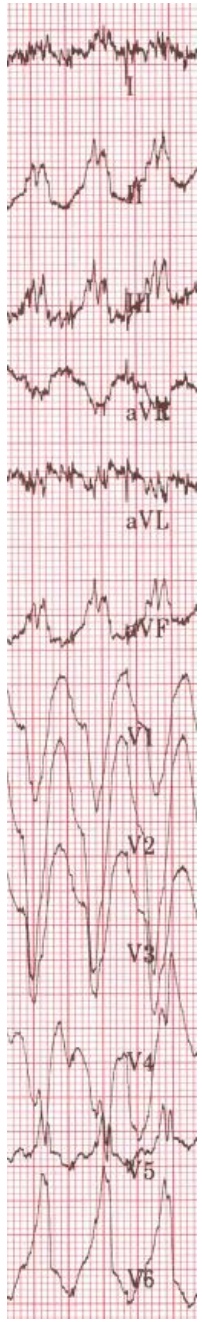
Elektrofyziologie & mapping



Jasný význam

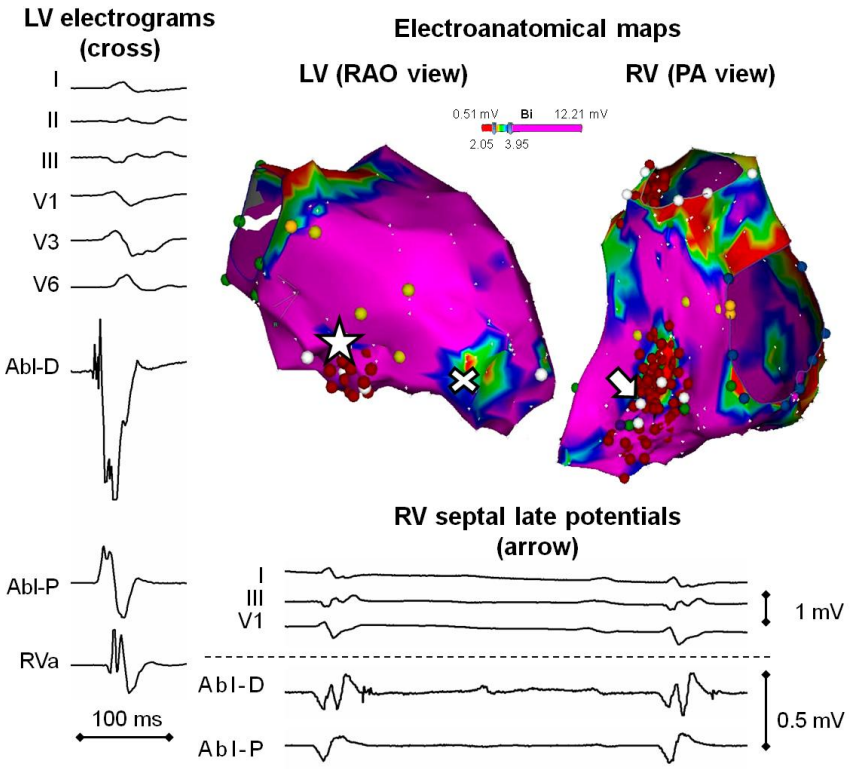
- Preexcitovaná fibrilace síní
- BBR-KT
- Rychlá SVT degenerující do VF
- Součást protokolu katetrizační ablace VT

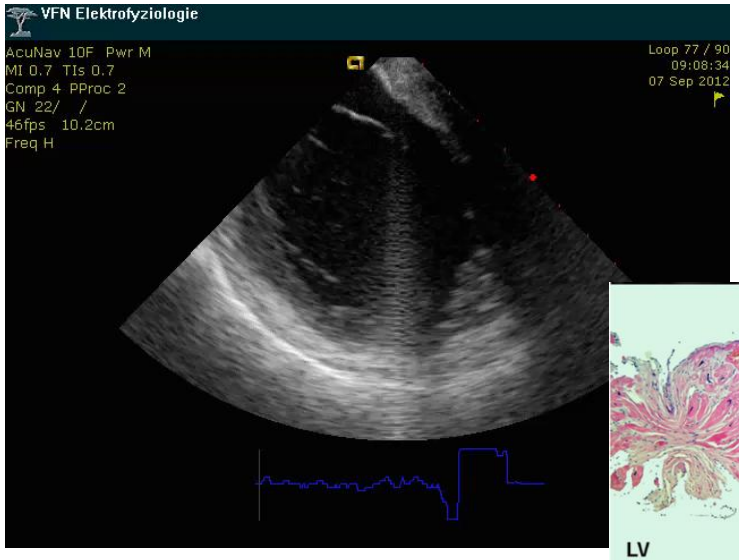
Potenciální význam

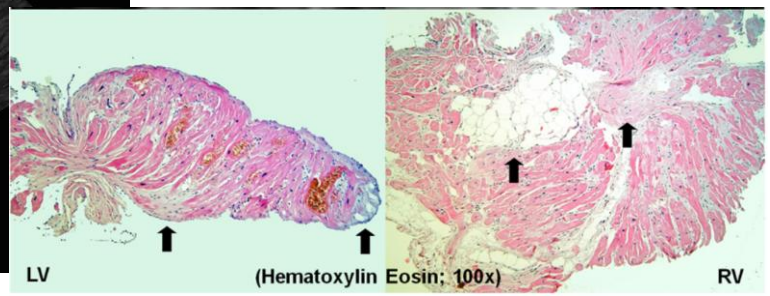
- Nejasná synkopa u pacienta se SHD s mrEF LK
- Brugada syndrom
- Myotonic dystrophy
- Sarkoidosa, ToF

Mapping

- Cíl pro katetrizační ablaci, Purkinje triggers
- Subklinická KMP (ARVC vs. OT VT)
- Cílení biopsie (ARVC, myocarditis, sarcoidosis…)







Havranek S et al. BMC Cardiovasc Disord 2015; 15:18. General University Hospital in Prague

2020 APHRS/HRS expert consensus statement on the investigation of decedents with sudden unexplained death and patients with sudden cardiac arrest, and of their families.

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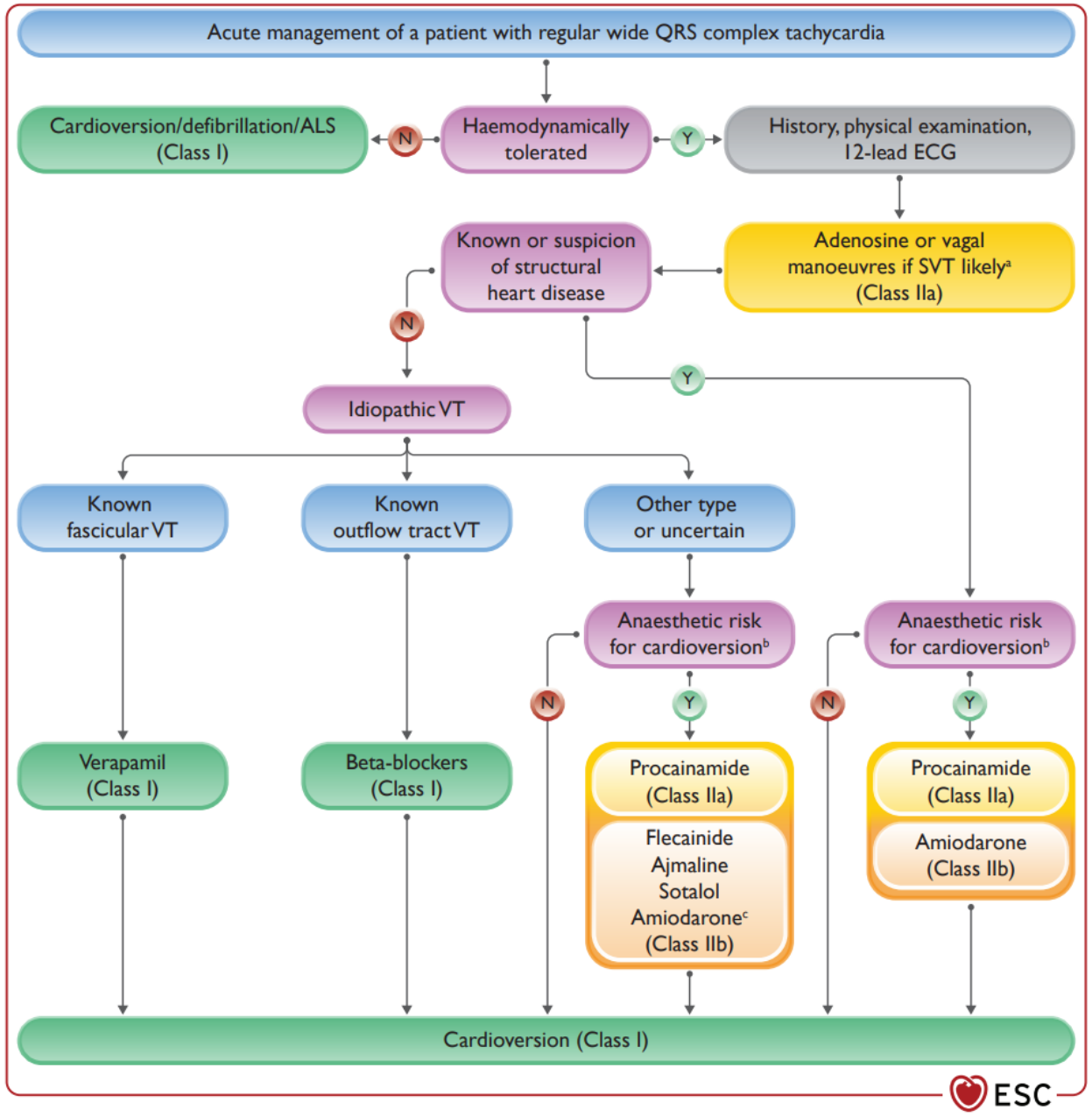
Setrvalá monomorfní komorová tachykardie



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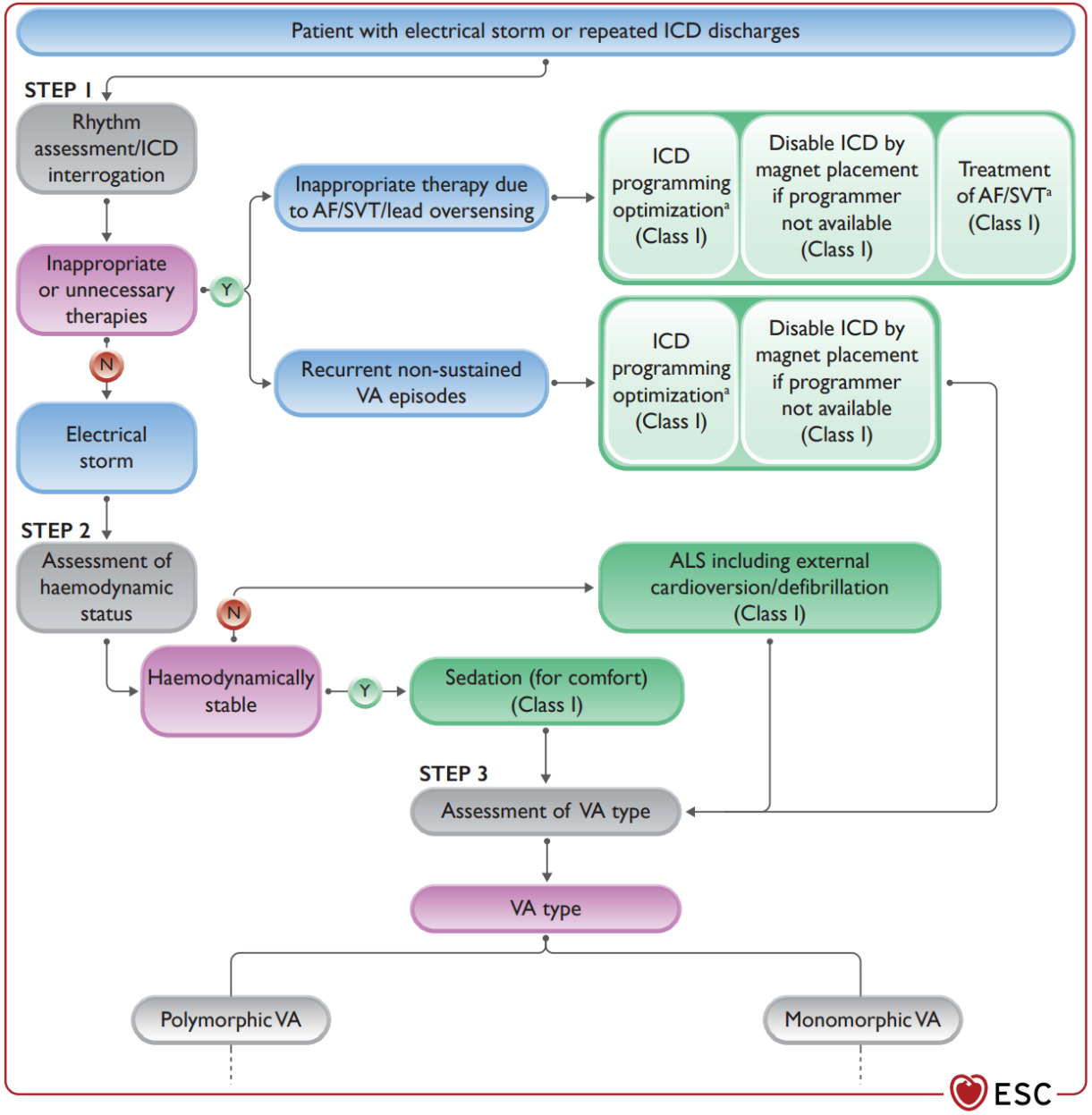
Bouře



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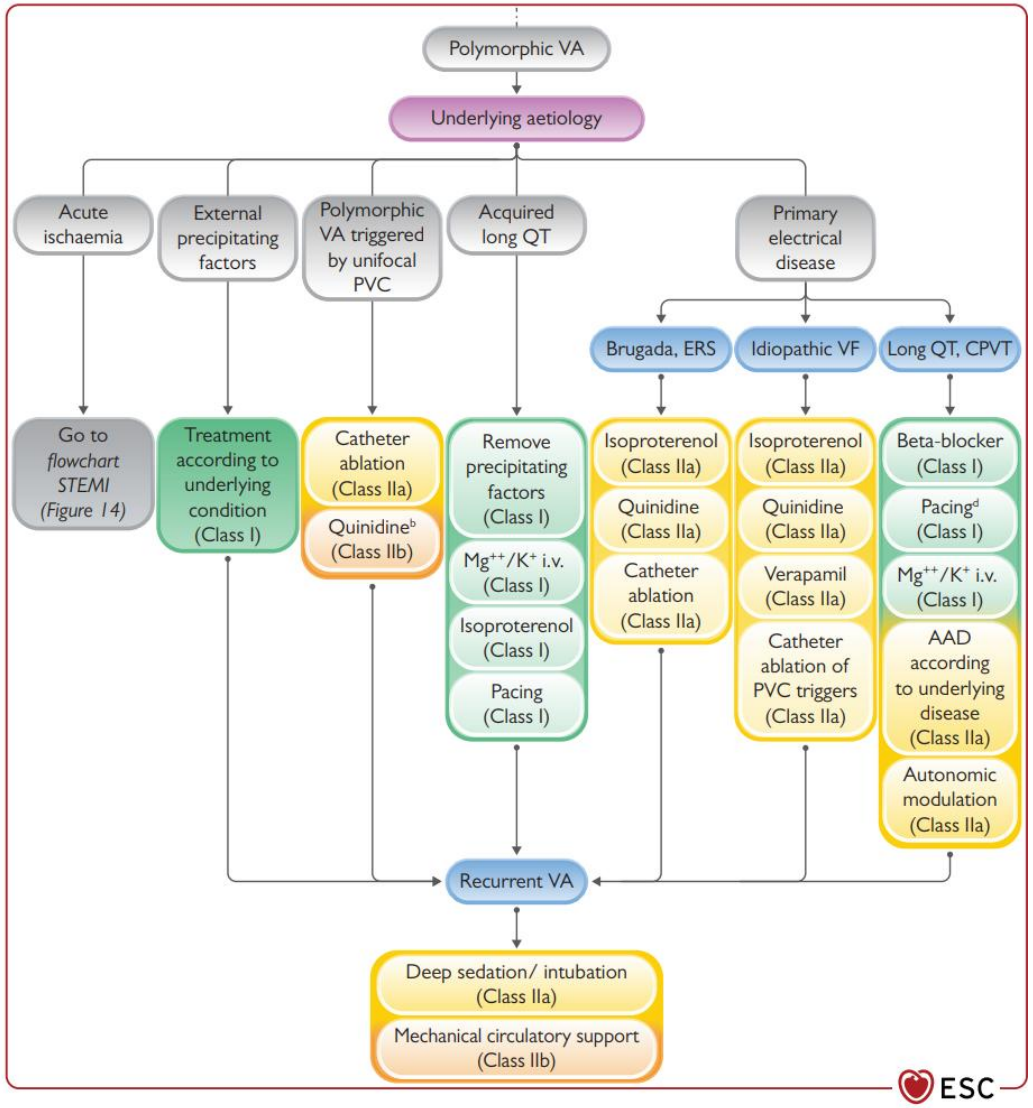

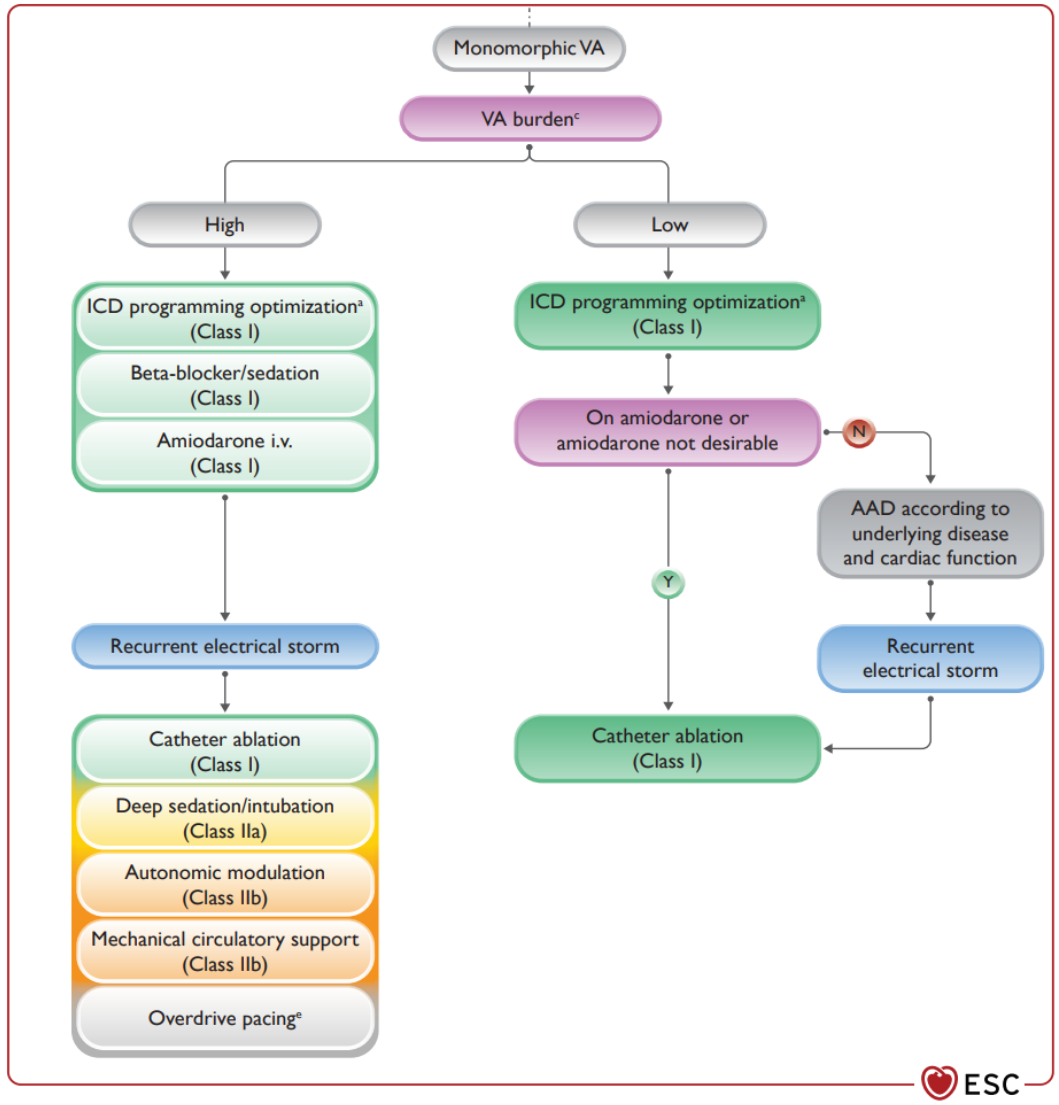

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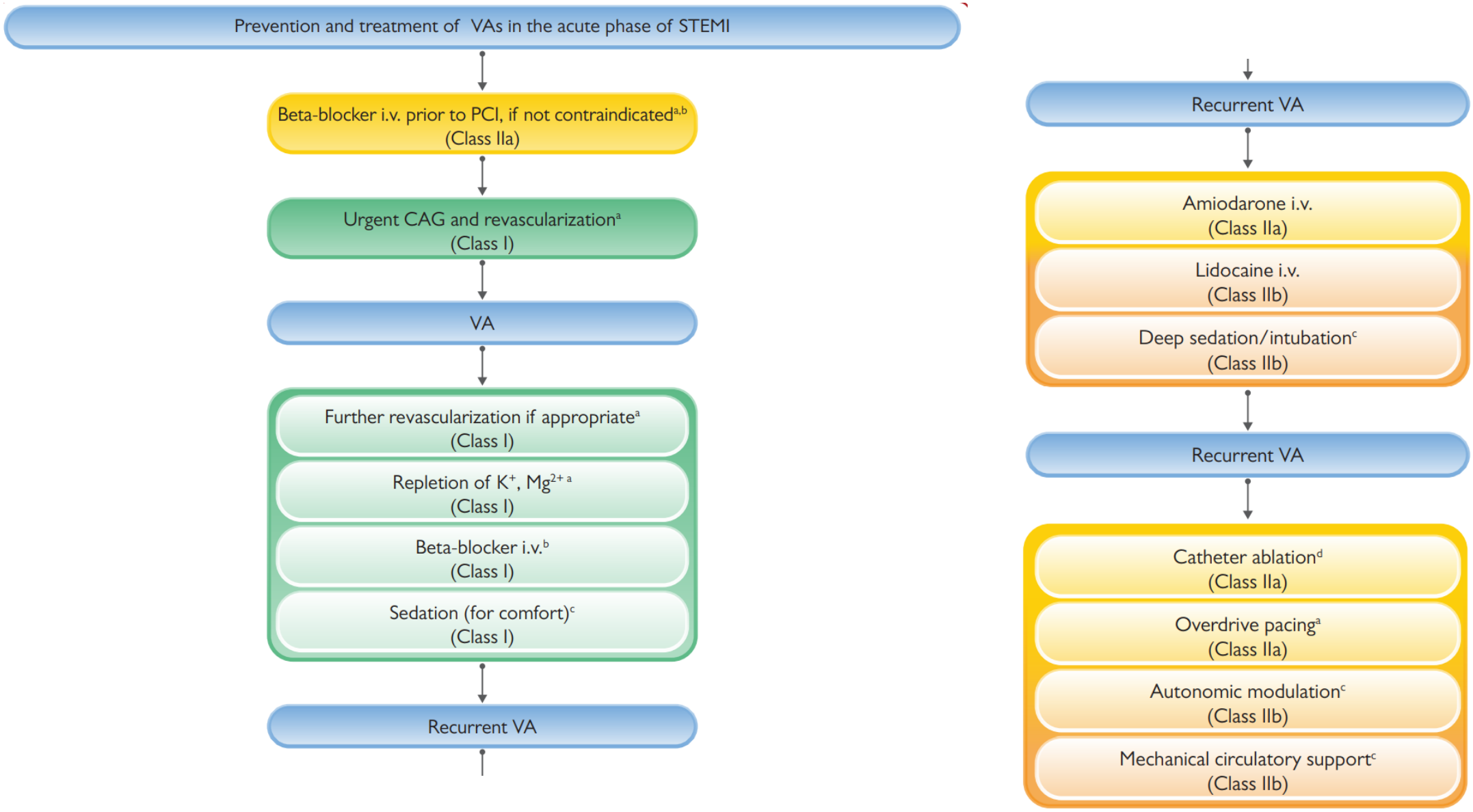
Komorová arytmie v akutní fázi IM



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Závěr

Nová doporučení pro „komorové tachykardie“

Klinické scénáře

Diagnostika

Základy akutní léčby

Chronická léčba dle etiologie

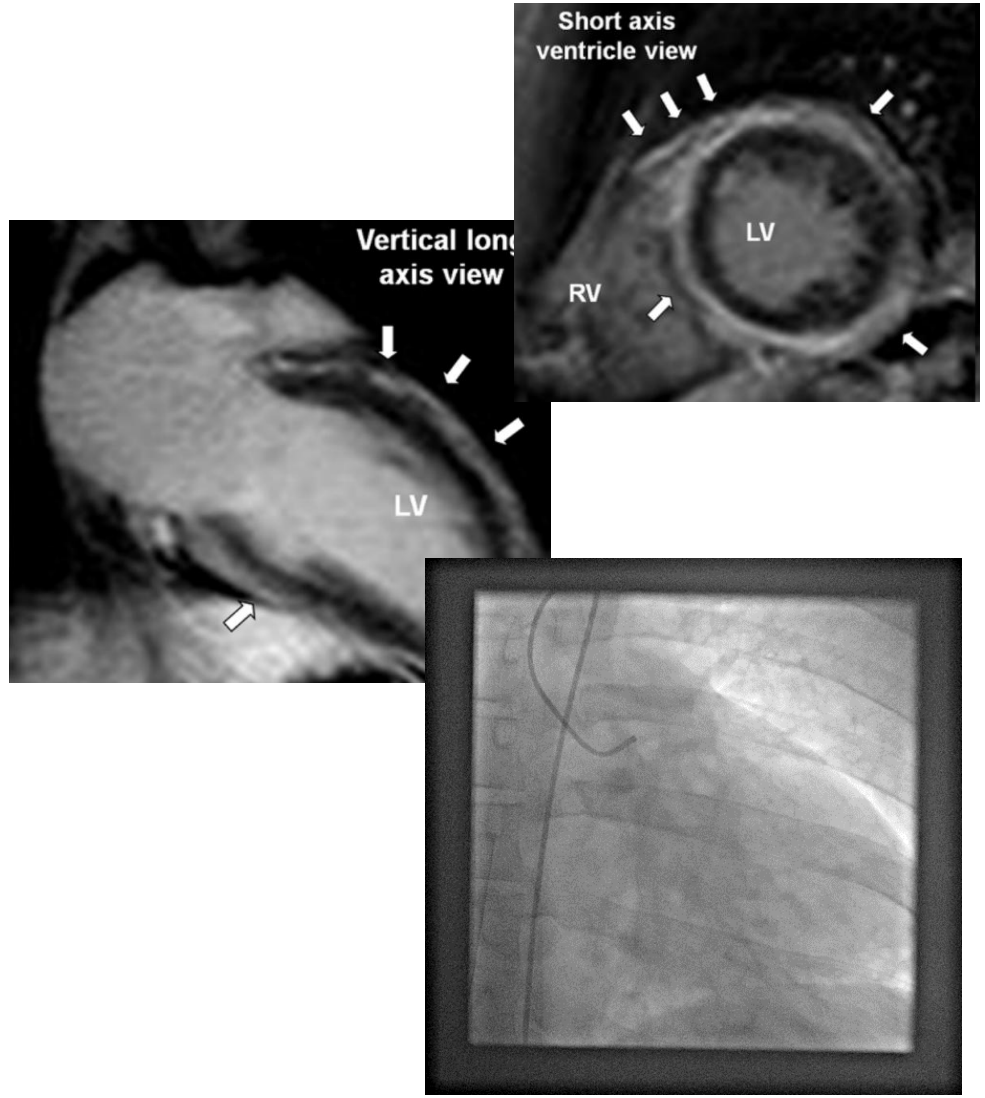
Prevence náhlé srdeční smrti, implantace ICD

Děkuji za pozornost!

Diagnostika u nemocných s první manifestací KT bez známé kardiální anamnézy

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

events.[113] Implantable loop recorders (ILR) can be useful in diagnosing arrhythmias in patients with potentially life-threatening symptoms, such as unexplained syncope.[114]



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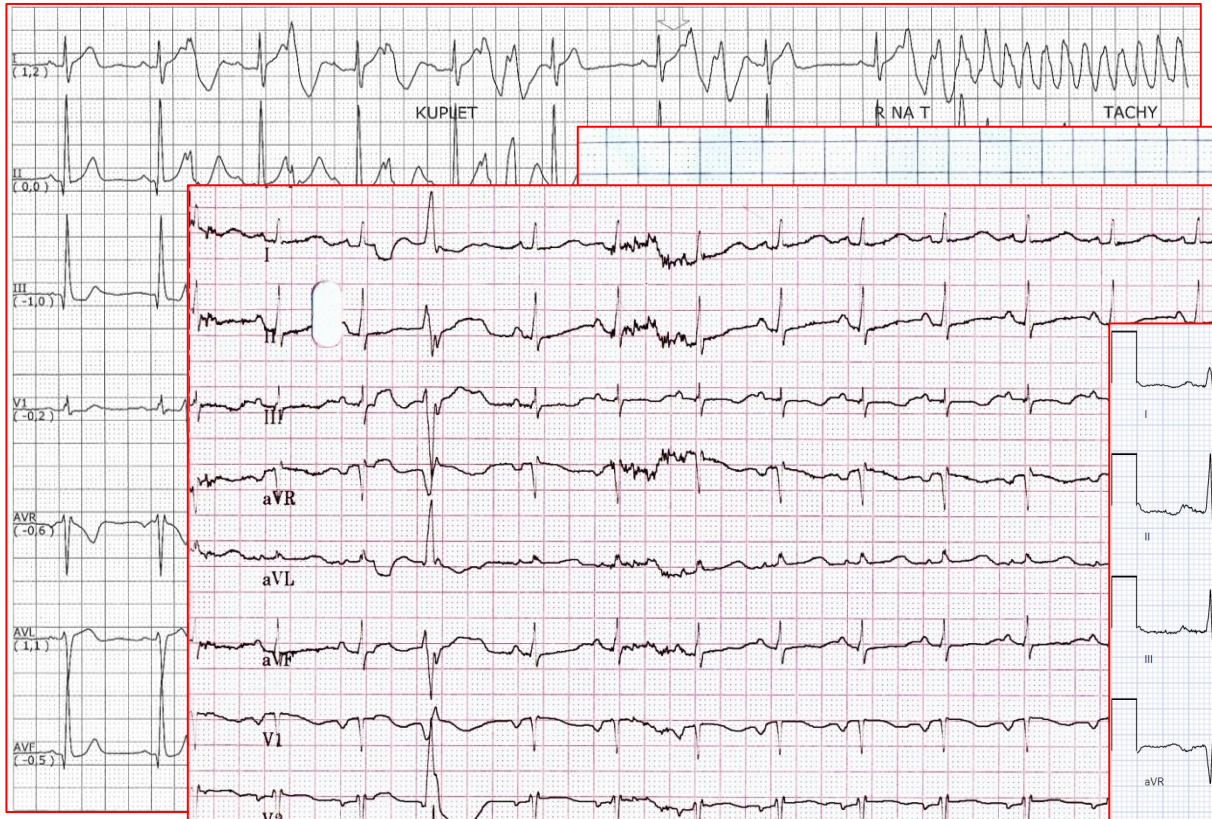
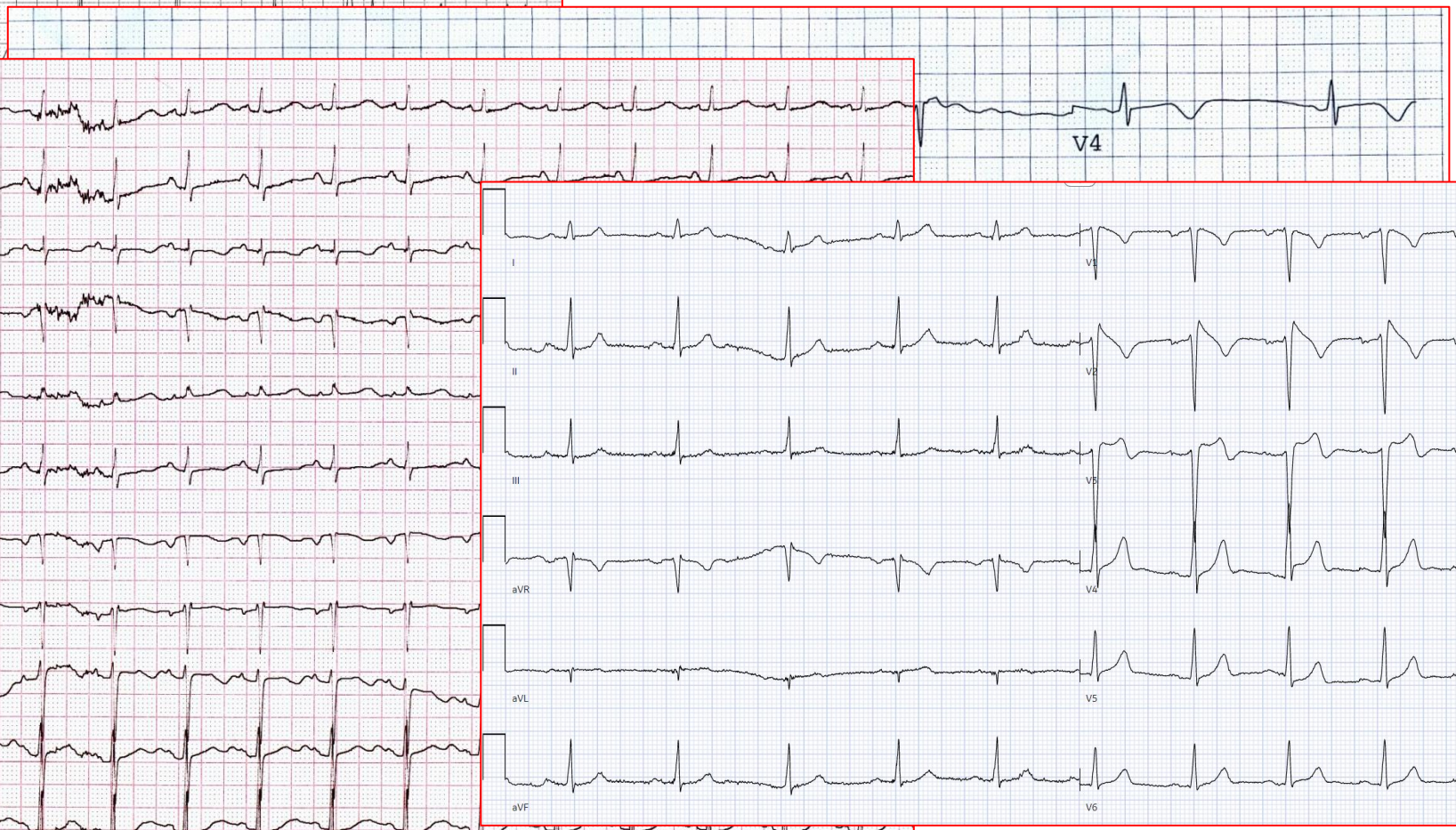
Zdroj: Všeobecná fakultní nemocnice

EKG – opakované, monitorování

Short-couplet TdP

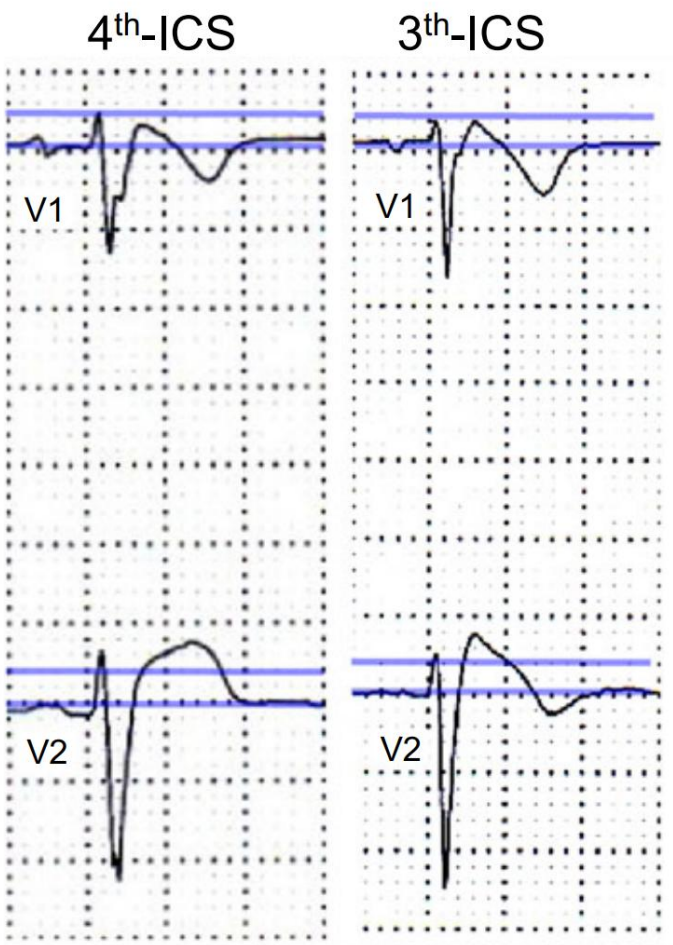


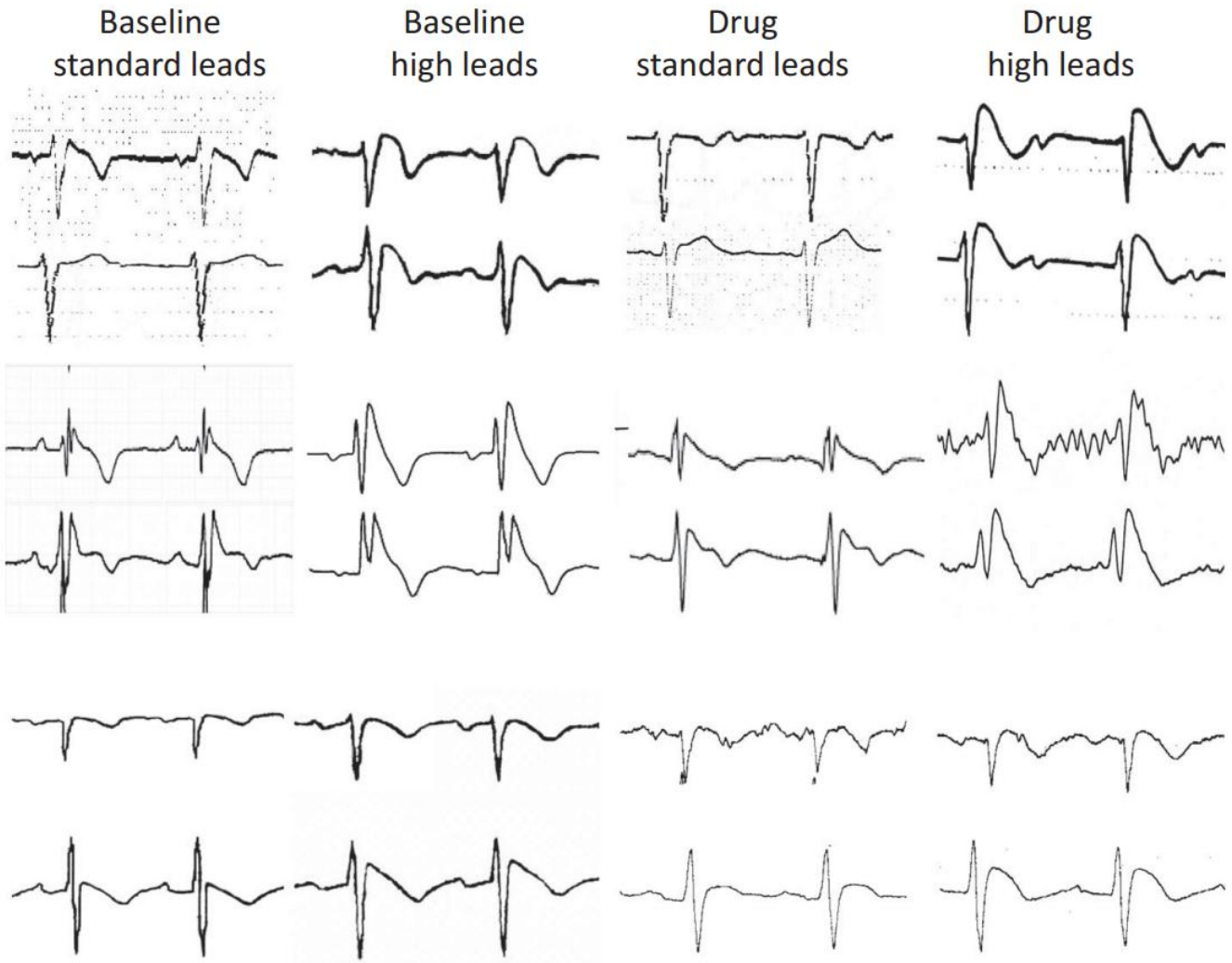
Long QT



EKG ve vyšší etáži

High precordial leads – Brugada syndrome





Curcio A et al. Clinical Presentation and Outcome of Brugada Syndrome Diagnosed with the New 2013 Criteria. JCE 2016; 27:937-43.

Zátěžové testy

Exercise testing in all undiagnosed SCA survivors
(Class 1)

Sodium channel blocker challenge* in undiagnosed SCA survivors with ECG or clinical characteristics suggestive of Brugada syndrome
(Class 1)

Lying to standing ECGs for possible LQTS (note: caution in children)
(Class 2a)

EP study if BBR-VT, pre-excited AF, or SVT is suspected
(Class 2a)

In SCA survivors where no other disorder is identified

- Sodium channel blocker challenge*
(Class 2a)
- Tests for coronary vasospasm†
- Adenosine challenge to unmask pre-excitation
- Electroanatomic right ventricular voltage mapping for detection of subclinical arrhythmogenic cardiomyopathy
- EP study to evaluate potential underlying substrate
- Adrenaline challenge for possible LQTS and CPVT, if unable to exercise
(Class 2b)

Změny navozené adrenergním mechanismem

- Zátěží indukovaná KT (ARVC)
- Bidirecionální KT in CPVT
- Zátěží navozené Epsilon a Typ 1 Brugada
- QTC >480ms ve 4 min recovery - LQTS

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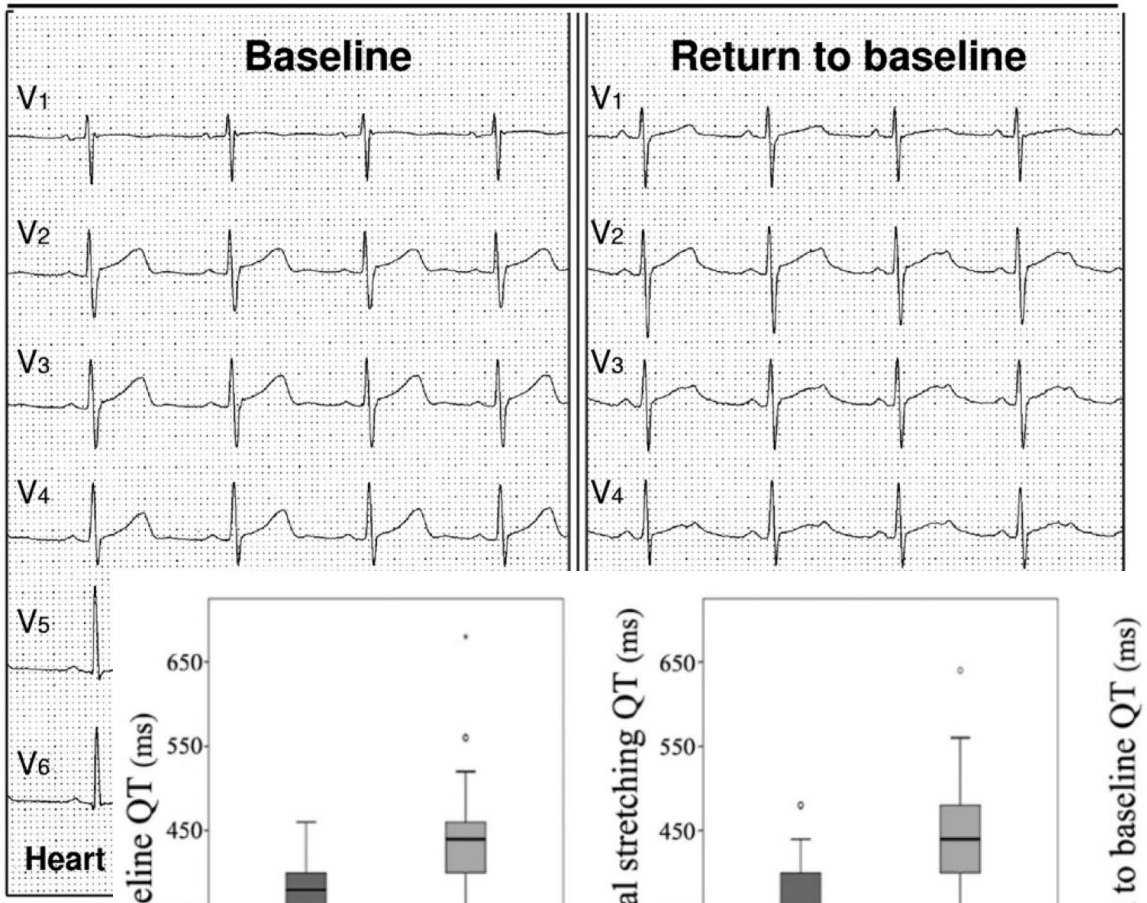
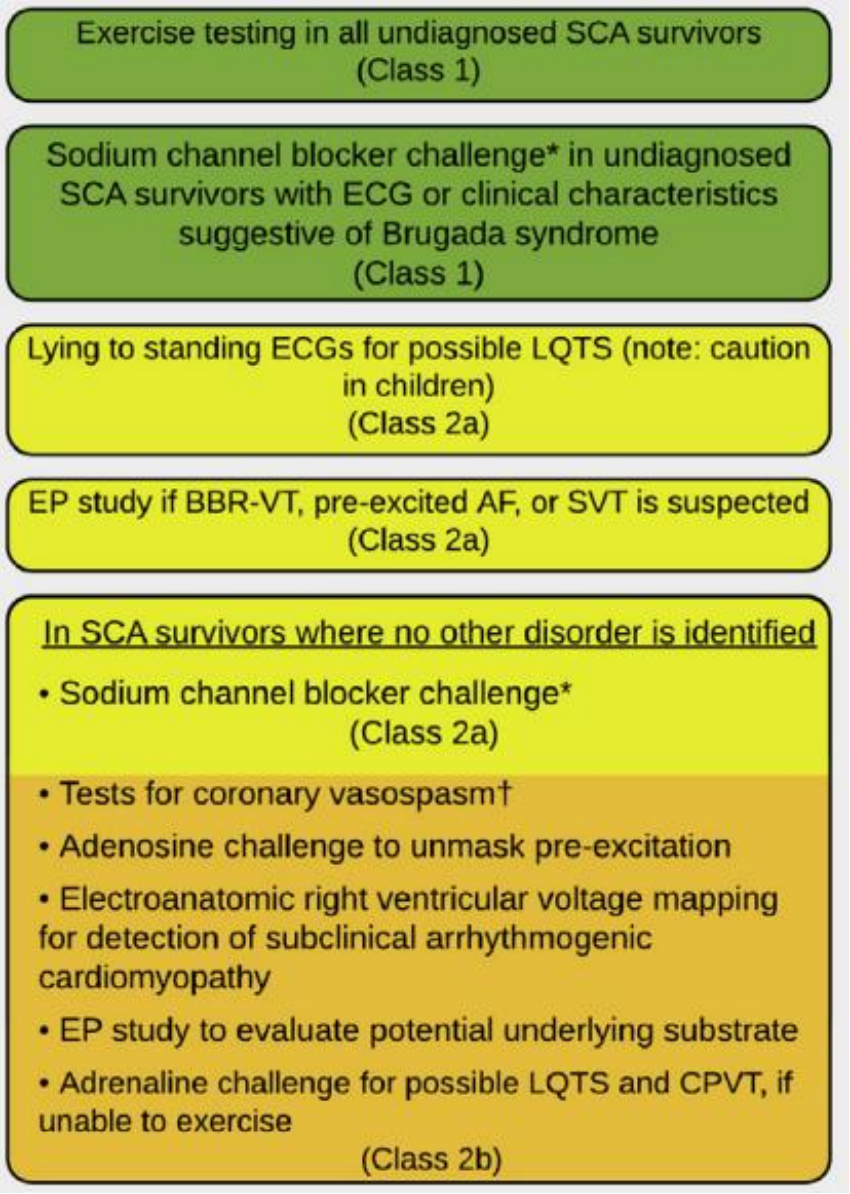
Postavit se, vydržet 5 min; EKG



2020 APHRS/HRS expert consensus statement on the investigation of decedents with sudden unexplained death and patients with sudden cardiac arrest, and of their families.
Adler A et al. The phenomenon of QT stunning: The abnormal prolongation provoked by standing in patients with long QT syndrome. Heart Rhythm 2012; 9:301-8.

Provokační testy



Diagnostic test	Indication	Protocols Dose/infusion rate/duration	Positive test
Ajmaline	Family history of BrS or SADS. Resuscitated CA without SHD.	1 mg/kg over 5–10 min (maximum dose 100 mg) or 1 mg/kg at 10 mg/min. Record in standard and high precordial leads over 30 min.	BrS type 1 ECG.
Flecainide	Same as ajmaline.	2 mg/kg over 10 min (maximum dose 150 mg). Record in standard and high precordial leads over 30 min.	Same as ajmaline.

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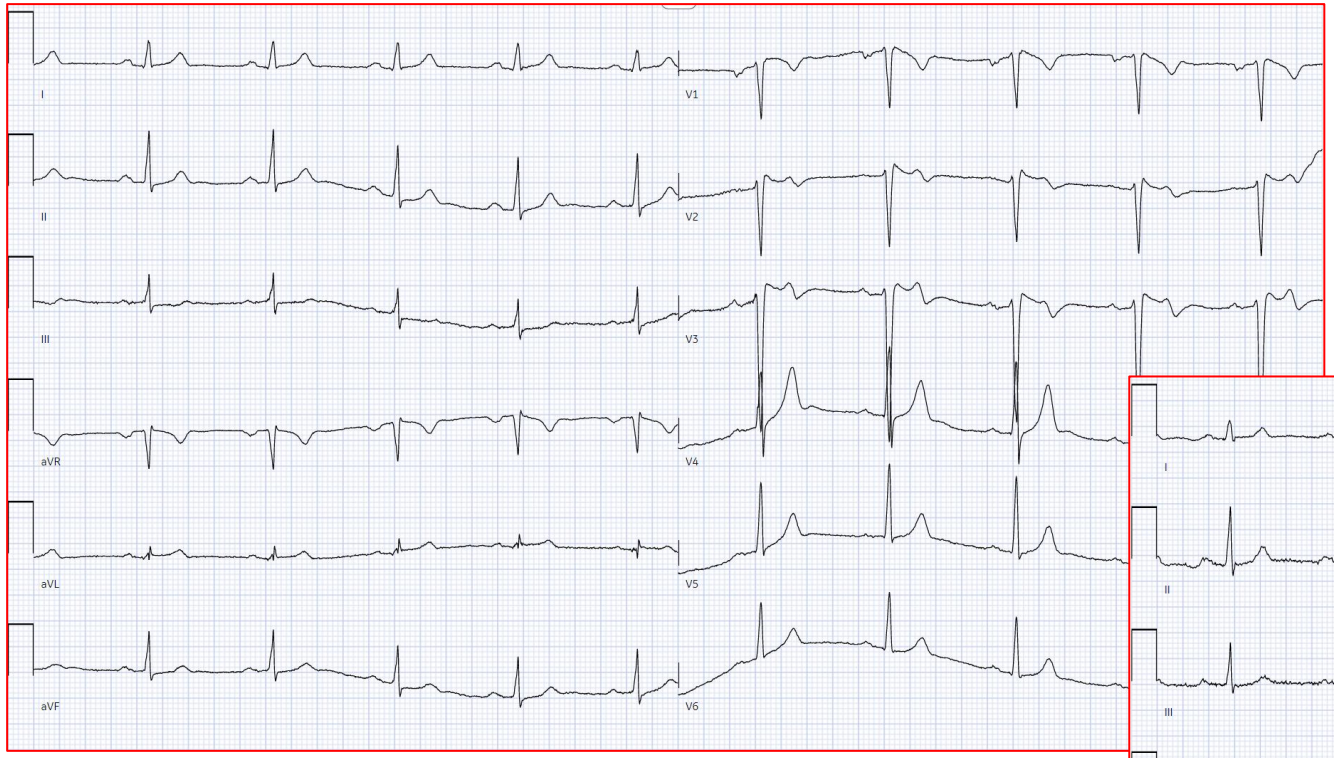
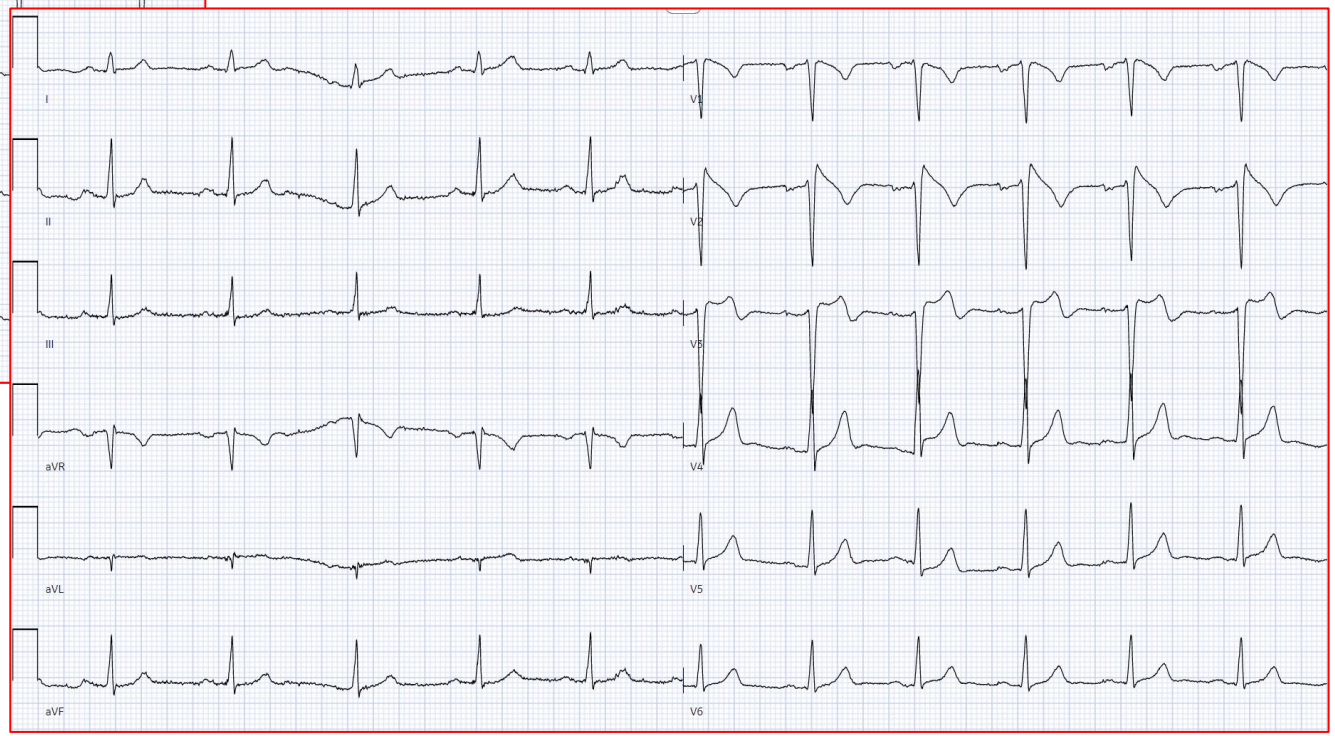
Provokační testy

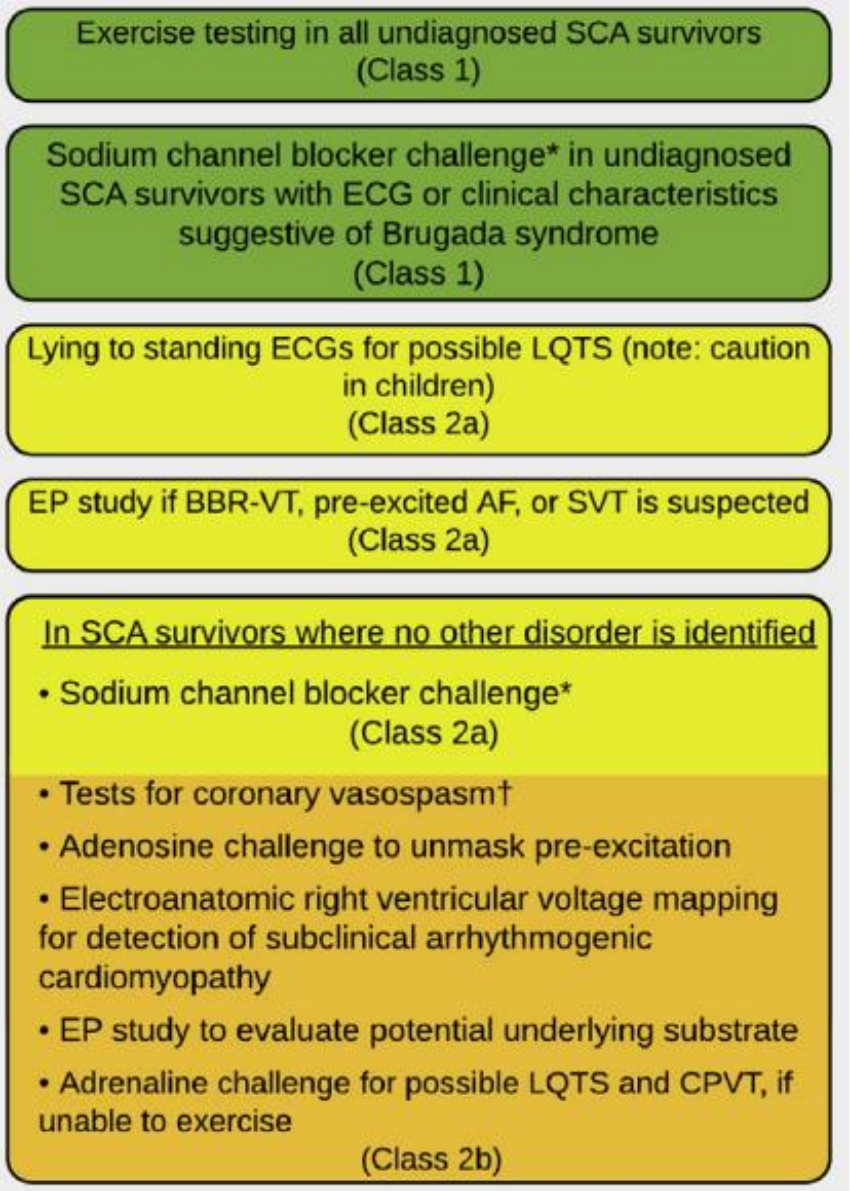




Ajmalin / Flecainid test

Geneticky potvrzený Brugada Syndrom

Provokační testy



Diagnostic test	Indication	Protocols Dose/infusion rate/duration	Positive test
Adenosine	Exclude latent pre-excitation.	6, 12, 18 mg boluses up to maximum dose 24 mg or until AV block or pre-excitation occurs.	Identification of accessory pathway.
Epinephrine	CPVT and resuscitated CA with or without SHD when exercise test not feasible. Family history of SADS.	Rest 10 min. Start at 0.025 µg/ kg/ min for 10 min increase sequentially to 0.05, 0.1 and 0.2 µg/ kg/min in 5 min steps.	≥3 beats of PVT or bidirectional VT.

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Provokační testy

Exercise testing in all undiagnosed SCA survivors
(Class 1)

Sodium channel blocker challenge* in undiagnosed SCA survivors with ECG or clinical characteristics suggestive of Brugada syndrome
(Class 1)

Lying to standing ECGs for possible LQTS (note: caution in children)
(Class 2a)

EP study if BBR-VT, pre-excited AF, or SVT is suspected
(Class 2a)

In SCA survivors where no other disorder is identified

- Sodium channel blocker challenge*
(Class 2a)
- Tests for coronary vasospasm†
- Adenosine challenge to unmask pre-excitation
- Electroanatomic right ventricular voltage mapping for detection of subclinical arrhythmogenic cardiomyopathy
- EP study to evaluate potential underlying substrate
- Adrenaline challenge for possible LQTS and CPVT, if unable to exercise
(Class 2b)

Diagnostic test	Indication	Protocols Dose/infusion rate/duration	Positive test
Acetylcholine	Suspicion of coronary vasospasm.	Intracoronary injection: RCA: 20 and 50 µg. LCA: 20, 50, and 100 µg over 20 s. >3-min intervals between injections. Maximal dose of 50 µg in the RCA and 100 µg in the LCA.	Coronary artery spasm visualized during procedure.
Ergonovine	Same as acetylcholine.	Intracoronary stepwise injection: RCA (20–60 mg) LCA (20–60 mg) over a period of 2–5 min.	Same as acetylcholine

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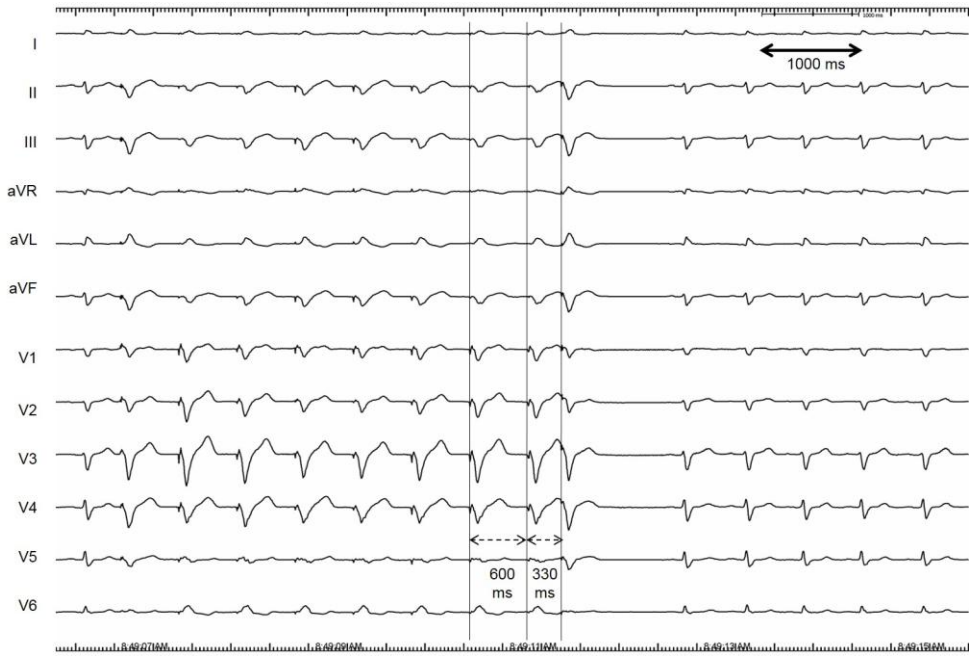
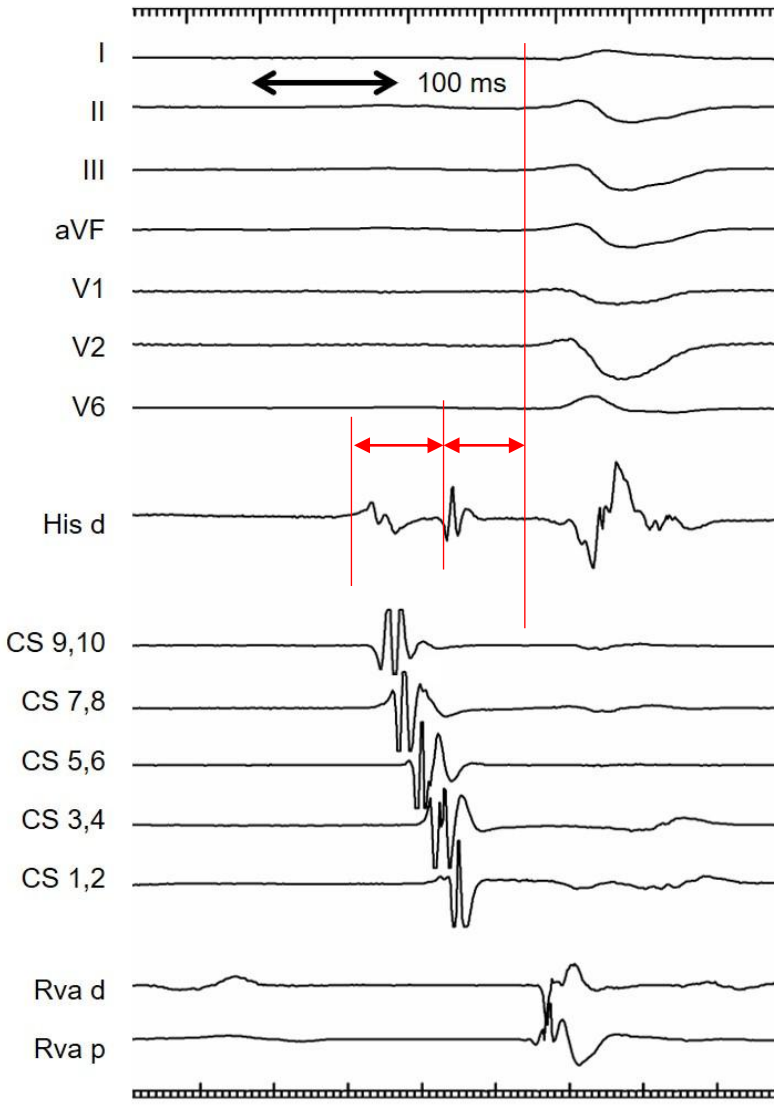
Elektrofyziologie & mapping

Součástí vyšetření

- Převodní intervaly AH, HV
- Programovaná stimulace
- Elektroanatomické mapování
- Podávání isoprenalinu

- Adenosin – rekonekce, provokace KES
- Hadgrip pro provokaci KES





Zdroj: Všeobecná fakultní nemocnice

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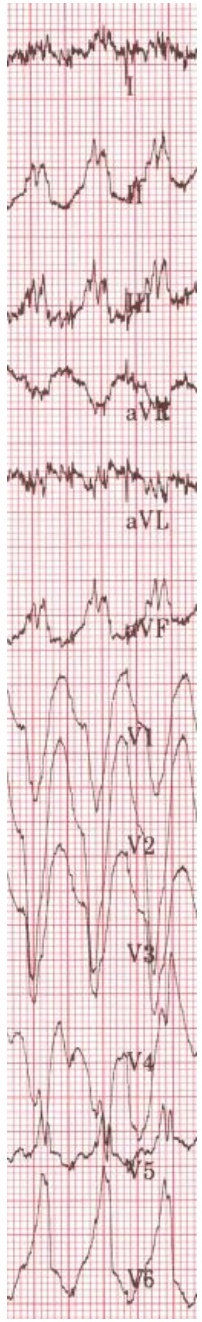
Elektrofyziologie & mapping



Jasný význam

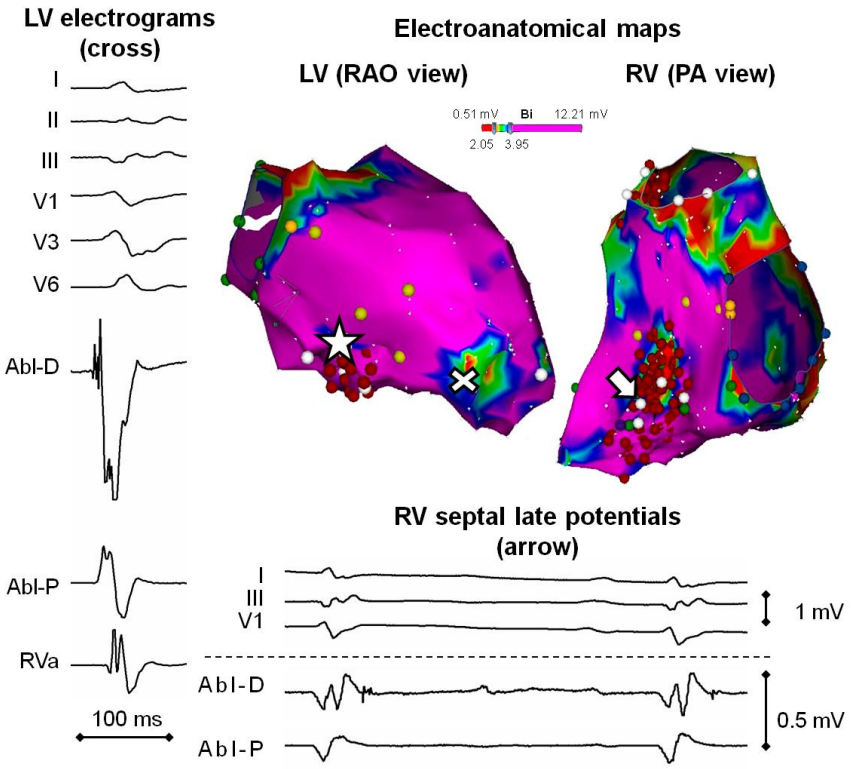
- Preexcitovaná fibrilace síní
- BBR-KT
- Rychlá SVT degenerující do VF
- Součást protokolu katetrizační ablace VT

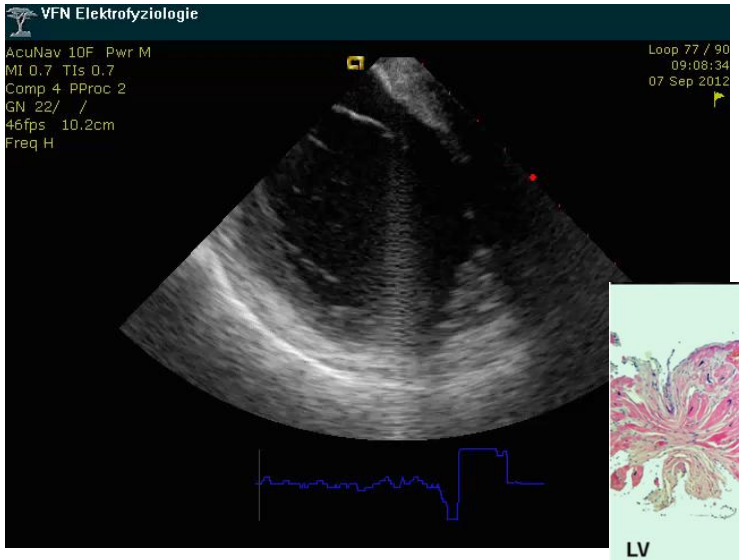
Potenciální význam

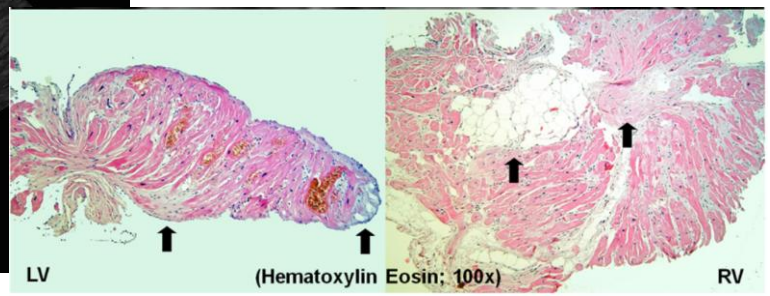
- Nejasná synkopa u pacienta se SHD s mrEF LK
- Brugada syndrom
- Myotonic dystrophy
- Sarkoidosa, ToF

Mapping

- Cíl pro katetrizační ablaci, Purkinje triggers
- Subklinická KMP (ARVC vs. OT VT)
- Cílení biopsie (ARVC, myocarditis, sarcoidosis…)







Havranek S et al. BMC Cardiovasc Disord 2015; 15:18. General University Hospital in Prague

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Genetické testování

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

Optimální je, aby výsledek ovlivnil

- Finální diagnózu
- Další management
- Rodinný screening

Indikované

- Diagnostikované či vysoce suspektní geneticky vázané onemocnění na podkladě fenotypu
- Geny s robustní „gene-disease association“

Není benefit

- Fenotyp s jasnou negenetickou příčinou

Mutation class	Variant	
I	Benign	Nondiagnostic
II	Likely benign	
III	A variant of uncertain origin	Re-evaluation
IV	Likely pathogenic	Diagnostic
V	Pathogenic	

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Genetické testování

			LQTS	BrS	CPVT	Idiopathic VF	ERS
	Genetic test		Class I[a]	Class I	Class I[a]	Class IIb	Class IIb
Proband	**Initial clinical test**	**Cornerstone for diagnosis**	ECG Exercise test	ECG and high precordial lead ECG Sodium channel blockers provocative test[c]	Exercise test	See *Section 5.2.3*, scenario 3	ECG
		Other tests/processes	Exclude acquired LQTS	Exclude phenocopy[b]	Exclude phenocopy[b]/SHD		Holter Echocardiography
	Follow-up		1–3 years dependent on level of risk				
Relatives	**Clinical screening**		ECG Exercise test (when feasible) From birth	ECG and high precordial lead ECGs: start at 10 years Sodium channel blockers provocative test[c]: start >16 years unless clinically indicated[180,181]	ECG Exercise test From birth	ECG and high precordial lead ECGs Exercise test Echocardiogram[182]	ECG Echocardiogram
	Follow-up	Positive phenotype and/or Class IV/V variant	1–3 years dependent on level of risk				
		Negative phenotype and no Class IV/V variant	Discharge				

Genetické testování a management

- Long QT syndrom → β-blokátor
- Long QT3 syndrom → Na+ blokátor
- CPVT → flecainide
- ARVC / ALVC → exercise restrictions

Další zásadní diagnózy

- DKMP (lamin A/C gene, PLN, FLNC, RBM20)
- HKMP (sarkomerická mutace)

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