PŘINÁŠÍ SOUČASNÁ DOPORUČENÍ NOVÉ INDIKACE K IMPLANTACI ICD?

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Klinika Kardiologie IKEM

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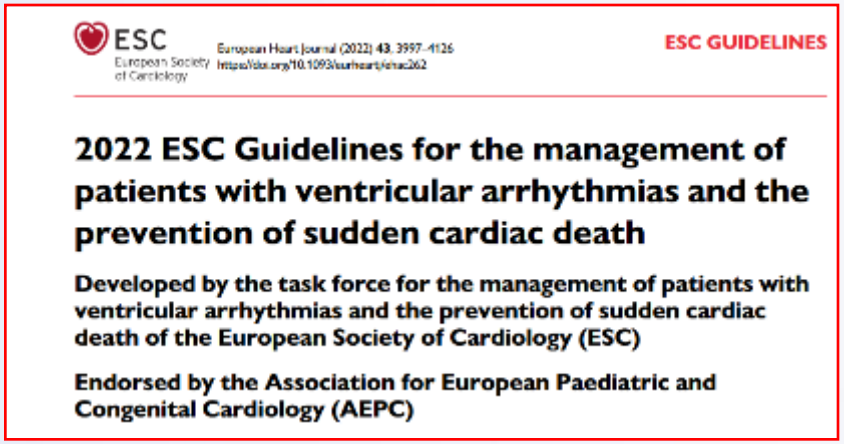

ESC European Society of Cardiology

European Heart Journal (2022) 43, 3997–4126 https://doi.org/10.1093/eurheartj/ehac262

ESC GUIDELINES

2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

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

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

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ČESKÁ KARDIOLOGICKÁ SPOLEČNOST THE CZECH SOCIETY OF CARDIOLOGY

ESC GUIDELINES

Doporučení pro... I Guidelines

Doporučení ESC pro diagnostiku a léčbu pacientů s komorovými arytmiemi a prevenci náhlé srdeční smrti 2022.

Zeppenfeld K, Tfelt-Hansen J, de Riva M, Winkel BG, Behr ER, Blom NA, Charron P, Corrado D, Dagres N, de Chillou C, Eckardt L, Friede T, Haugaa KH, Hocini M, Lambiase PD, Marijon E, Merino JL, Peichl P, Priori SG, Reichlin T, Schulz-Menger J, Sticherling C, Tzeis S, Verstrael A, Volterrani M; ESC Scientific Document Group.

Nahradila původní Guidelines z roku 2015

ESC European Society of Cardiology

European Heart Journal (2023) 44, 3503–3626 https://doi.org/10.1093/eurheartj/ehad194

ESC GUIDELINES

2023 ESC Guidelines for the management of cardiomyopathies

Developed by the task force on the management of cardiomyopathies of the European Society of Cardiology (ESC)

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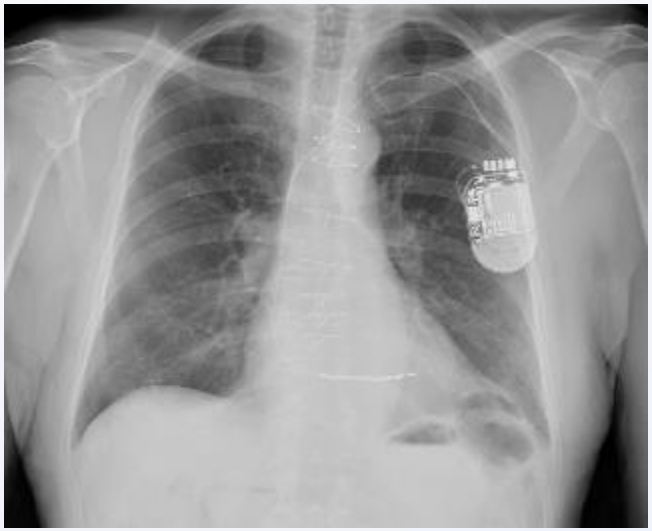
Nová Guidelines, update jen u HKMP z roku 2014

Implantabilní kardioverter defibrilátory

- Indikovat u pts s předpokládanou dobou přežití alespoň 1 rok
- CAVE: těžká CHRI, HD, DM

  zvažovat NSS vs. úmrtí z jiných příčin
- kombinován s bradykardickou nebo s resynchronizační léčbou



Prevence → **primární**

sekundární

Studie	AVID
	SIDS
	CASH

Sekundární prevence u ICHS

Sekundární prevence NSS a léčba KA		
Implantace ICD je doporučena u pacientů bez stávající ischemie, s dokumentovanou FK nebo hemodynamicky netolerovanou KT, která se vyskytla více než 48 hodin po IM.	I	A
U pacientů s ICHS a hemodynamicky tolerovanou SMKT a EF LK > 40 % lze zvážit místo implantace ICD ablaci v expertním centru za předpokladu, že byly dosaženy uznávané cílové ukazatele.[c]	IIa	C
U pacientů s hemodynamicky tolerovanou SMKT a EF LK > 40 % by měla být zvážena implantace ICD v případech, kdy katetrizační ablace selže nebo ji nelze provést, případně není vhodná.	IIa	C

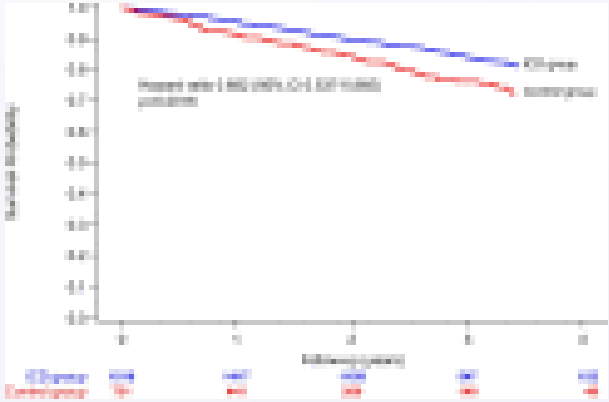
2022 ESC/2023 ČKS Guidelines: Ventricular arrhythmias and the prevention of sudden cardiac death

Primární prevence

- Po IM, hlavním rizikovým faktorem výrazně snížená EF LK pod 35%
- Riziko maligních arytmií stoupá s časem po IM
- Největší přínos pro pacienty s ND NYHA II-III

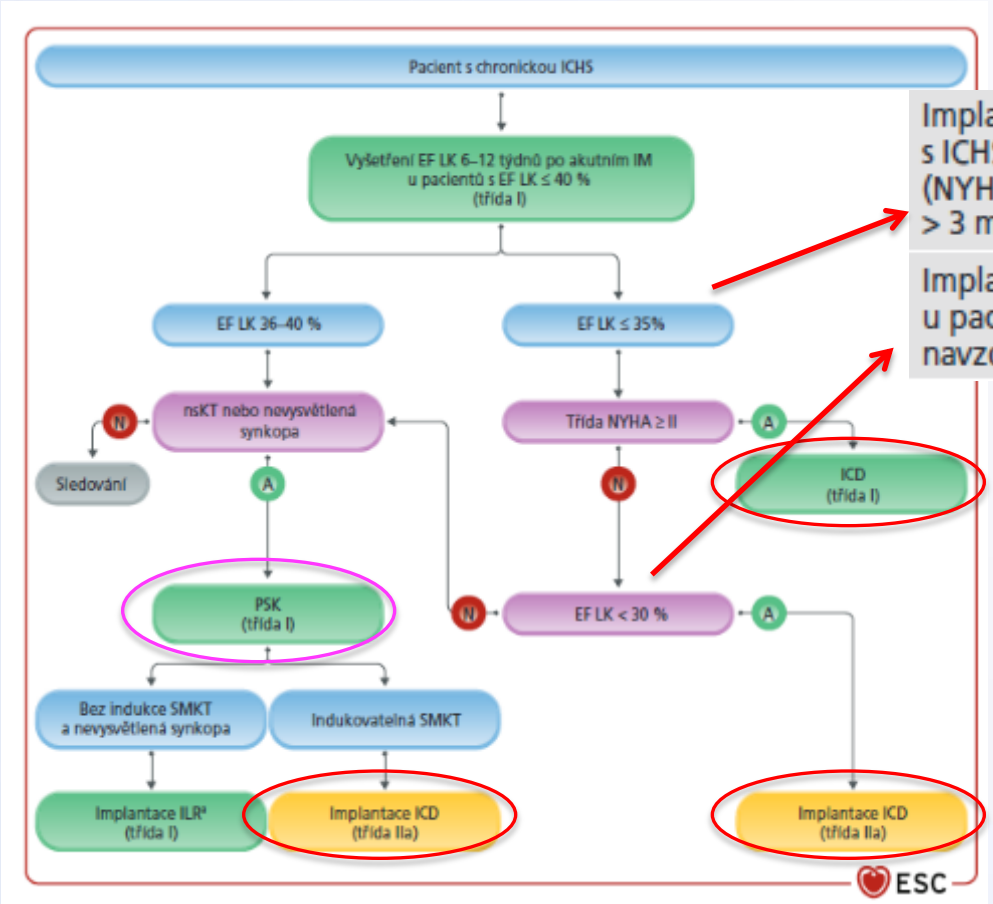
Studie	MADIT
	MADIT II
	Dinamit
	SCD-Heft
	Companion

EU-CERT-ICD (2327 pts 2014-2018)
ICD v ppNSS - velký prospektivní registr – 27% redukce mortality
Eur Heart J 2020 Sep 21;41(36):3437-3447



Probíhá Studie PROFID – u pacientů po IM s dysfunkcí LK a s nastavenou medikací
CHSS – změna v pp NSS ?

Riziková stratifikace a primární prevence NSS u chronické ICHS



Implantace ICD je doporučena u pacientů s ICHS, symptomatickým srdečním selháním (NYHA II–III) a EF LK < 35 % navzdory OMT > 3 měsíce.	I	A
Implantace ICD by měla být zvážena u pacientů s ICHS, NYHA I a EF LK < 30 %, navzdory OMT > 3 měsíce.	IIa	B

EF LK< 35%
NYHA II-III

EF LK < 30%
NYHA I

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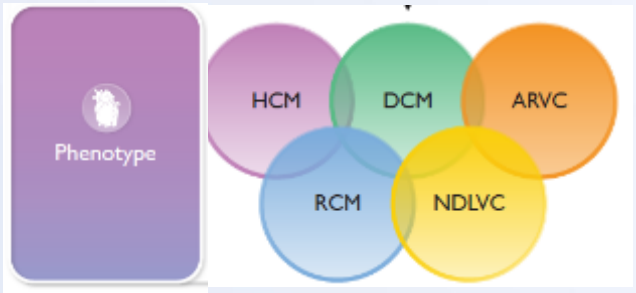
Nová klasifikace KMP podle fenotypu

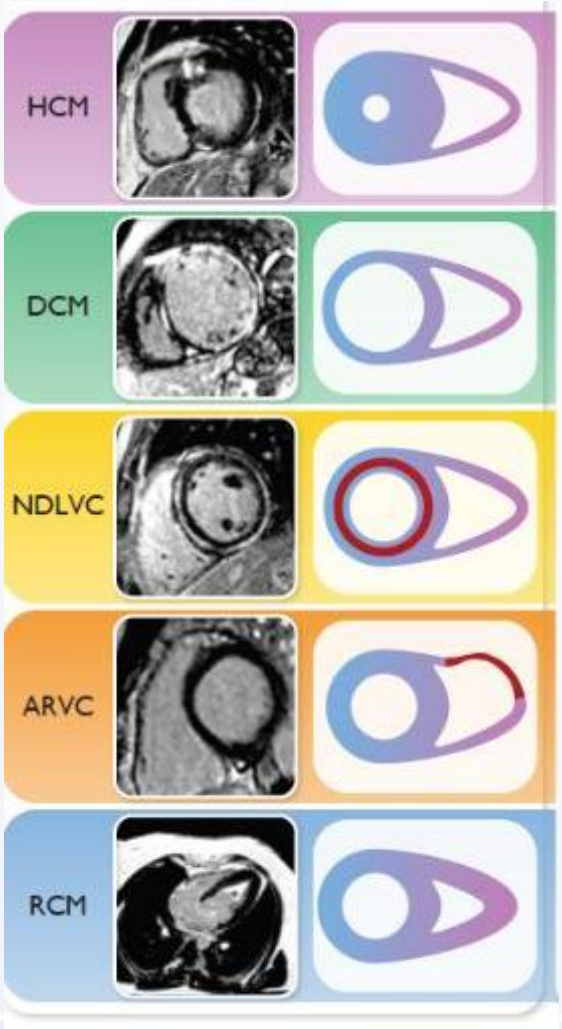


CMR

+

-

genetické testování



HKMP

DKMP

NDLVC

ARVC

RKMP

2023 ESC Guidelines for the management of cardiomyopathies

Nová klasifikace KMP podle fenotypu

- **NDLVC – nedilatovaná KMP LK** (dříve HNDKMP – hypokinetická nedilatovaná kardiomyopatie)

- Fenotyp je definován jako přítomnost **neischemických jizev** nebo **tukové tkáně při absenci dilatace LK** bez ohledu na poruchu kinetiky
  - **Arytmogenní KMP LK- ALVC (ACM)**

  nebo **izolvaná globální hypokineze** LK bez jizev (LGE na CMR)
  - **Hypokinetická nedilatovaná KMP**

- **Non-kompaktní KMP (LVNC) – zařazena mezi DKMP - není vlastní fenotypovou jednotkou**
- **Tako-tsubo – neřadí se mezi KMP**

2023 ESC Guidelines for the management of cardiomyopathies

KMP - riziková stratifikace

Kalkulátory pro výpočet rizika NSS

- **HKMP –** doporučeno
- **DKMP, NDLVC, ARVC** - mělo by být zváženo
- **indikován PM** - zvážit rizikovou stratifikaci k ICD

Primary prevention		
Comprehensive SCD risk stratification is recommended in all cardiomyopathy patients who have not suffered a previous cardiac arrest/sustained ventricular arrhythmia at initial evaluation and at 1–2 year intervals, or whenever there is a change in clinical status.	I	C
The use of validated SCD algorithms/scores as aids to the shared decision-making when offering ICD implantation, where available:[e]		
• is recommended in patients with HCM.[81,525,535]	I	B
• should be considered in patients with DCM, NDLVC, and ARVC.[185,186,524,526,536–542]	IIa	B
If a patient with cardiomyopathy requires pacemaker implantation, comprehensive SCD risk stratification to evaluate the need for ICD implantation should be considered.	IIa	C

2023 ESC Guidelines for the management of cardiomyopathies

Sekundární prevence u KMP

Secondary prevention		
Implantation of an ICD is recommended:[d]		
• in patients with HCM, DCM, and ARVC who have survived a cardiac arrest due to VT or VF, or who have spontaneous sustained ventricular arrhythmia causing syncope or haemodynamic compromise in the absence of reversible causes.[528–534]	I	B
• in patients with NDLVC and RCM who have survived a cardiac arrest due to VT or VF, or who have spontaneous sustained ventricular arrhythmia causing syncope or haemodynamic compromise in the absence of reversible causes.	I	C
ICD implantation should be considered in patients with cardiomyopathy presenting with haemodynamically tolerated VT, in the absence of reversible causes.	IIa	C

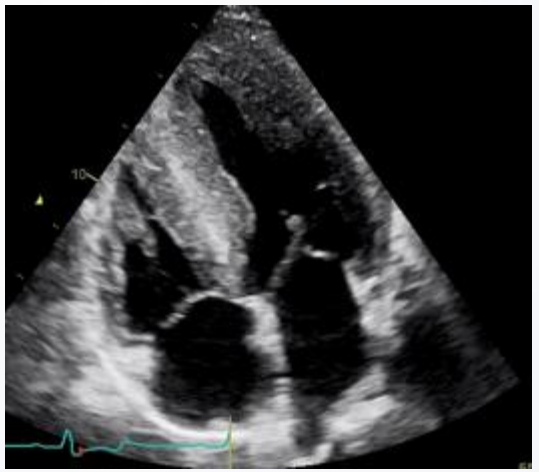
2023 ESC Guidelines for the management of cardiomyopathies

HKMP - primární prevence kalkulátor rizika NSS

HCM Risk-SCD Calculator

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

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



https://doc2do.com/hcm/webHCM.html

In patients with LV apical aneurysms, decisions about primary prevention ICD based on an assessment of risk using the HCM Risk-SCD or a validated paediatric risk-prediction (e.g. HCM Risk-Kids) tool and not solely on the presence of the aneurysm should be considered.[580,728,737,791,792]	**IIa**	**B**

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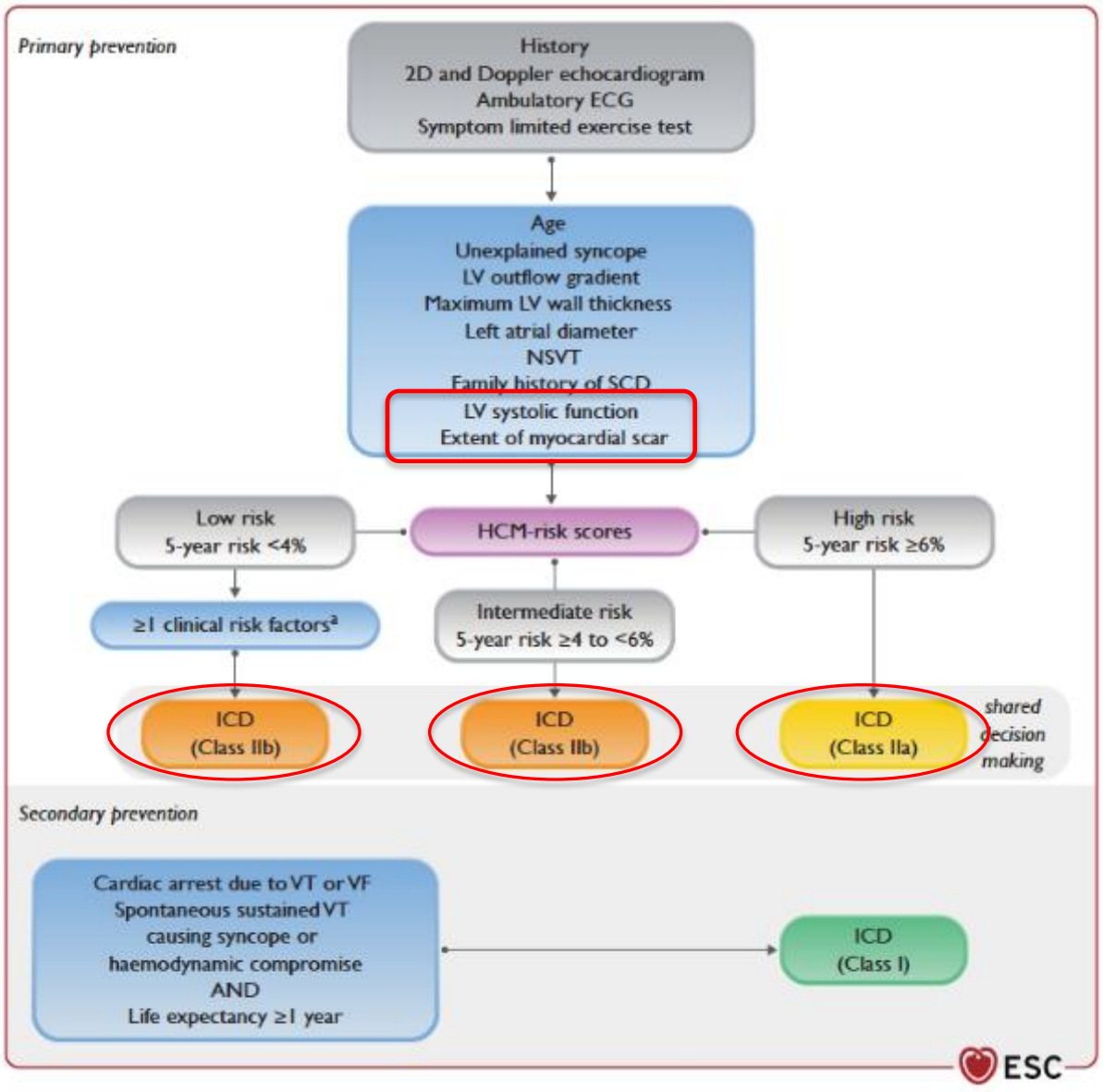
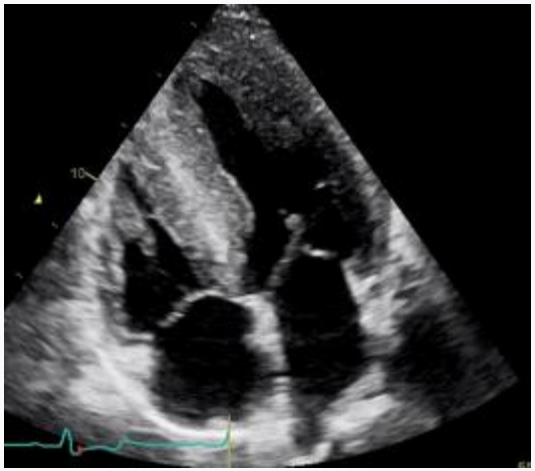
Doporučení u HKMP





Přídatné RF:

- **přítomnost LGE nad 15%**
- **EF LK pod 50%**

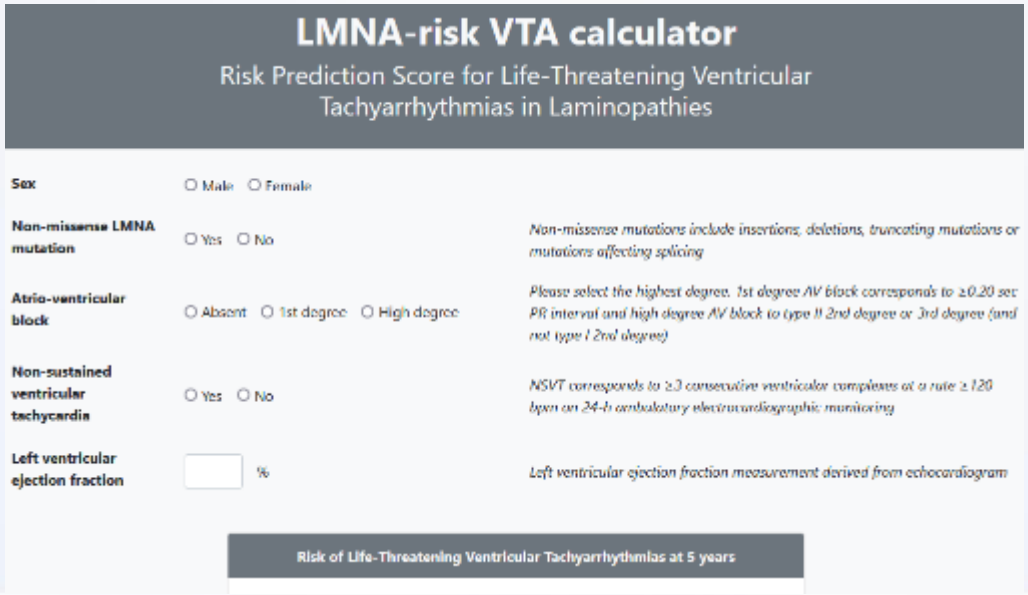
Neakceptované RF:

- Apikální aneurysma
- Patologická tlaková reakce na zátěž
- patologické sarkomerické mutace

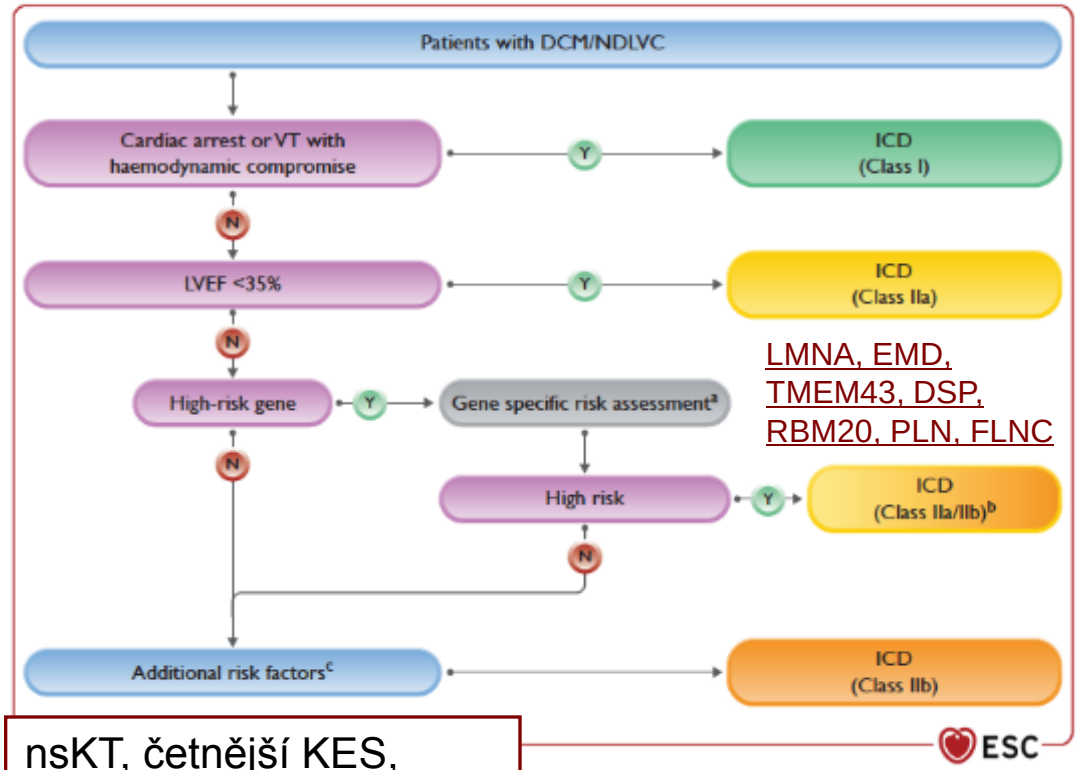
Kardiomyopatie - DKMP

- Při OMT 5-letá mortalita 21-28%, NSS ve 12%
- Patogenní mutace genu pro TTN, LMNA, sarkomerické a desmosomální mutace (AD)
- Vyšší riziko KT a NSS mutace pro LMNA, PLN, RBM20 a FLNC
- CMR

- ***https://lmna-risk-vta.fr/***



Kardiomyopatie – DKMP/NDLVC



LMNA, EMD, TMEM43, DSP, RBM20, PLN, FLNC

nsKT, četnější KES, muži, LGE na CMR

Table 21 High-risk genotypes and associated predictors of sudden cardiac death

Gene	Annual SCD rate	Predictors of SCD
LMNA[185,186,438,541,865,878,879]	5–10%	Estimated 5-year risk of life-threatening arrhythmia using *LMNA* risk score (https://lmna-risk-vta.fr)
FLNC-truncating variants[866,867,880]	5–10%	LGE on CMR LVEF < 45%
TMEM43[868,881]	5–10%	Male Female and any of the following: LVEF <45%, NSVT, LGE on CMR, >200 VE on 24h Holter ECG
PLN[542,882,883]	3–5%	Estimated 5-year risk of life-threatening arrhythmia using *PLN* risk score (https://plnriskcalculator.shinyapps.io/final_shiny) LVEF < 45% LGE on CMR NSVT
DSP[185,186]	3–5%	LGE on CMR LVEF < 45%
RBM20[869]	3–5%	LGE on CMR LVEF < 45%

CMR, cardiac magnetic resonance; DSP, desmoplakin; ECG, electrocardiogram; FLNC, filamin C; LGE, late gadolinium enhancement; LMNA, lamin A/C; LVEF, left ventricular ejection fraction; NSVT, non-sustained ventricular tachycardia; PLN, phospholamban; RMB, RNA binding motif protein; SCD, sudden cardiac death; VE, ventricular ectopic beats.

Kardiomyopatie – DKMP/NDLVC

Primary prevention		
An ICD should be considered to reduce the risk of sudden death and all-cause mortality in patients with DCM, symptomatic heart failure, and LVEF ≤35% despite >3 months of OMT.[861,885]	IIa	A
The patient's genotype should be considered in the estimation of SCD risk in DCM.[185,186,869,886]	IIa	B
An ICD should be considered in patients with DCM with a genotype associated with high SCD risk and LVEF >35% in the presence of additional risk factors (see *Table 21*).[541,542,867,869,873,878,881,886]	IIa	C
An ICD may be considered in selected patients with DCM with a genotype associated with high SCD risk and LVEF >35% without additional risk factors (see *Table 21*).[869,873,881,886]	IIb	C
An ICD may be considered in patients with DCM without a genotype associated with high SCD risk and LVEF >35% in the presence of additional risk factors.[c,138,873,874]	IIb	C

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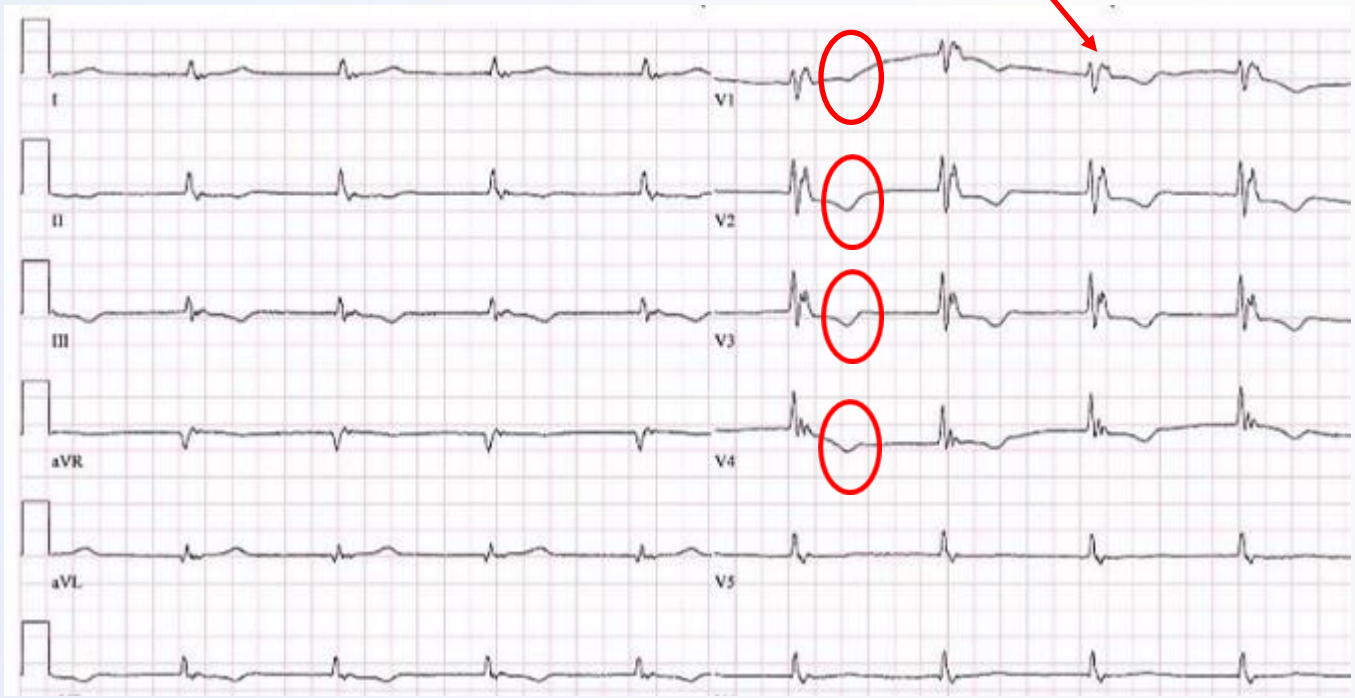
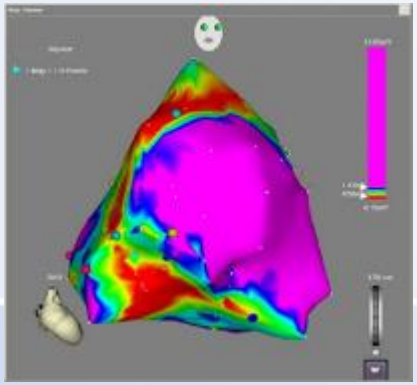
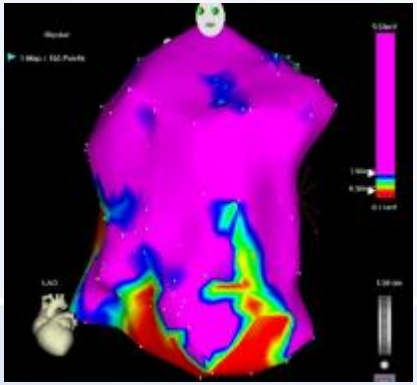
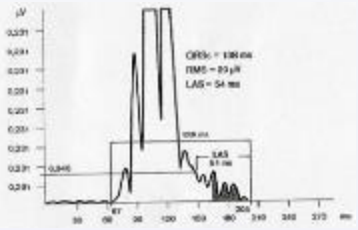
Arytmogenní kardiomyopatie

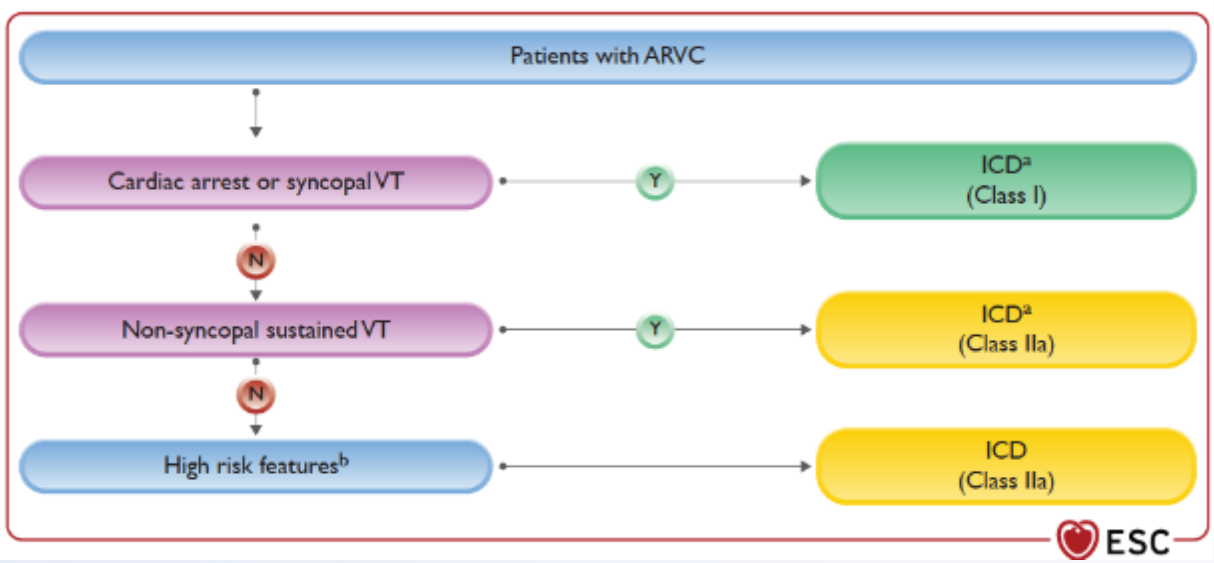


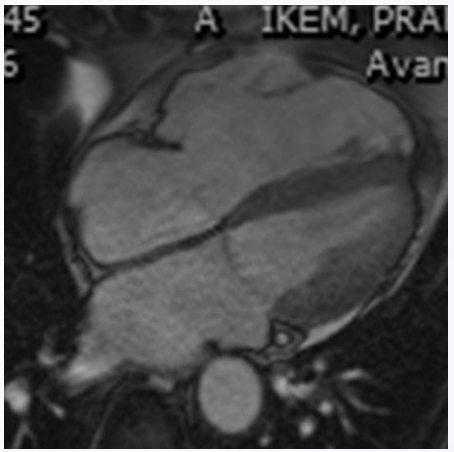






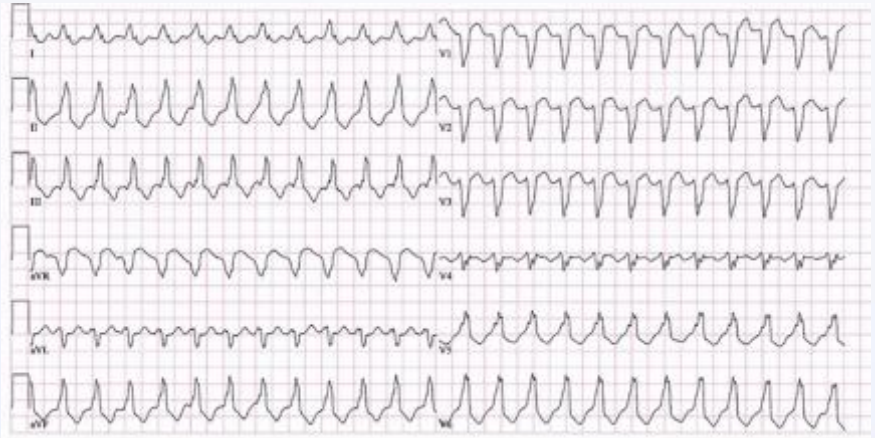
ARVC – prevence NSS





Rizikové faktory:

kardiální synkopa, nsKT, EF PK pod 40%, EF LK pod 45%,
SMKT při PES nebo dle 2019 ARVC kalkulátoru



https://arvcrisk.com

Restriktivní kardiomyopatie

Recommendation Table 30 — Recommendations for the management of patients with restrictive cardiomyopathy

Recommendations	Class[a]	Level[b]
It is recommended that multimodality imaging be used to differentiate RCM from HCM or DCM with restrictive physiology.	I	C
It is recommended that baseline cardiac and non-cardiac investigations are performed to assess involvement of the neuromuscular system or other syndromic disorders.	I	C
Cardiac catheterization is recommended in all children with RCM to measure pulmonary artery pressures and PVR at diagnosis and at 6–12 monthly intervals to assess change in PVR.[953]	I	B
ICD implantation is recommended to reduce the risk of sudden death and all-cause mortality in patients with RCM who have survived a cardiac arrest or have recovered from a ventricular arrhythmia causing haemodynamic instability.	I	C
Endomyocardial biopsy should be considered in patients with RCM to exclude specific diagnoses (including iron overload, storage disorders, mitochondrial cytopathies, amyloidosis, and granulomatous myocardial diseases) and to diagnose restrictive myofibrillar disease caused by desmin variants.	IIa	C
ICD implantation may be considered in *children* with RCM who have evidence of myocardial ischaemia and syncope.[969]	IIb	C

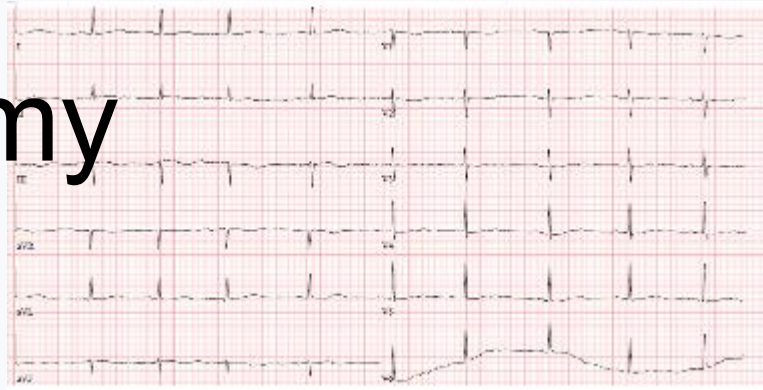
DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; ICD, implantable cardioverter defibrillator; PVR, pulmonary vascular resistance; RCM, restrictive cardiomyopathy.
[a]Class of recommendation.
[b]Level of evidence.

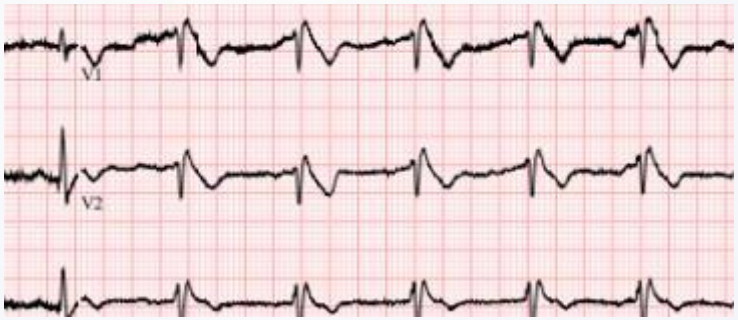
Hereditární syndromy

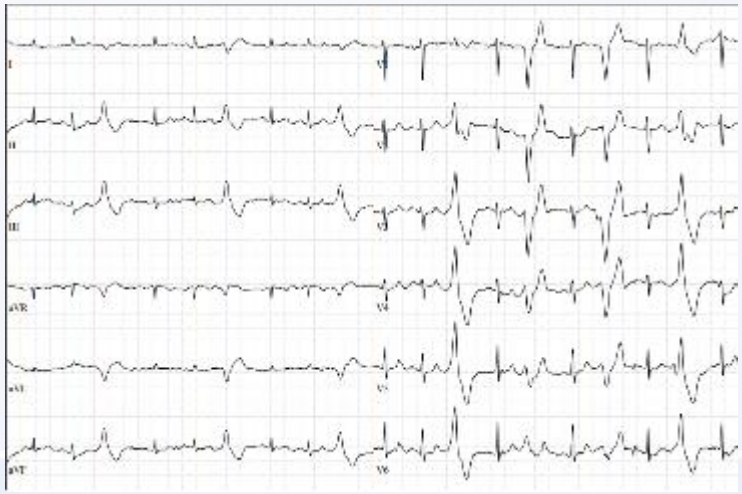
LQTS 1,2,3 (KCNQ1, KCNH2,SCN5A)
neselektivní BB - nadolol

Brugada syndrom (SCN5A) –
ajmalinový test, event. chinidin

Katecholaminergní polymorfní KT
(RYR2, kalsequestrin) - nadolol,
flecainid







- 12 svodové EKG, vyšší svody u BG
- Rodinná anamnéza
- Genetické vyšetření
- Specifická léčba
- Implantace ICD - sekundární prevence
- NSS

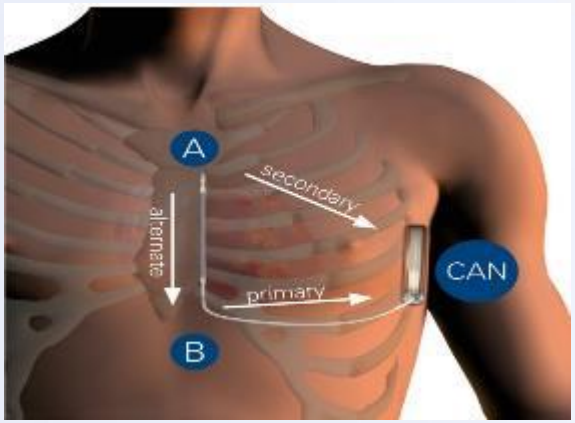
Typ ICD

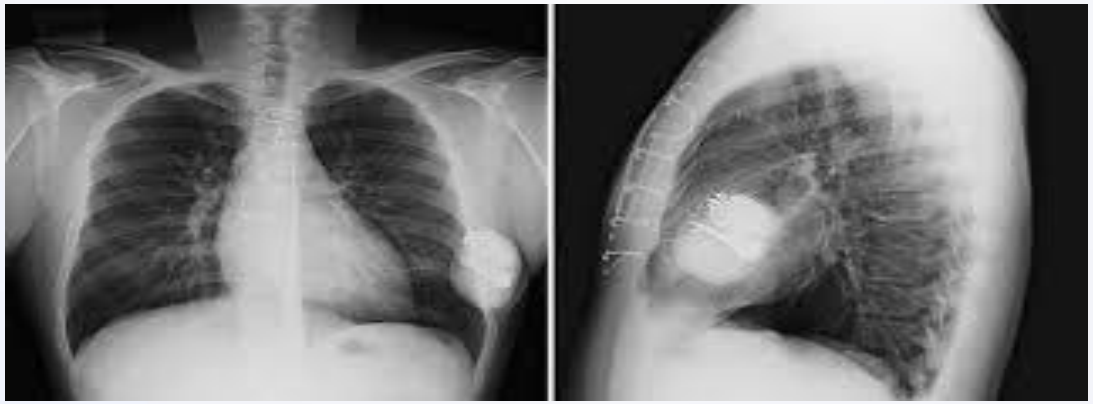
Choice of ICD		
When an ICD is indicated, it is recommended to evaluate whether the patient could benefit from CRT.[533]	I	A
Subcutaneous defibrillators should be considered as an alternative to transvenous defibrillators in patients with an indication for an ICD when pacing therapy for bradycardia, cardiac resynchronization, or antitachycardia pacing is not anticipated.[543]	IIa	B

Indikace k S-ICD

Podkožní defibrilátor by měl být považován za alternativu transvenózního ICD u pacientů s indikací k ICD, kdy není potřeba stimulace pro bradykardii, srdeční resynchronizace nebo ATP.	IIa	B

- **indikovaní:**
  mladí nemocní
  problémy s cévním přístupem
  po explantaci transvenózního ICD pro infekci nebo infrakci elektrod
- pacienti s rizikem komplikací





Závěr

- ICD indikováno v primární prevenci u pacientů s těžkou dysfunkcí LK
- U pacientů se zachovalou nebo středně sníženou EF LK stále snaha o identifikaci pacientů s nejvyšším rizikem NSS
- CMR a genetické vyšetření významnou roli ve stanovování rizika NSS
- Pomoc kalkulátorů rizikového skóre NSS
- Při zvažování benefitu léčby ICD hodnotíme i riziko nekardiálního úmrtí
- Nutno zvážit typ ICD s ohledem na onemocnění, věk pacienta a komorbidity

Děkuji za pozornost