Co přinesla nová KMP Guidelines 2023?

Jan Krejčí

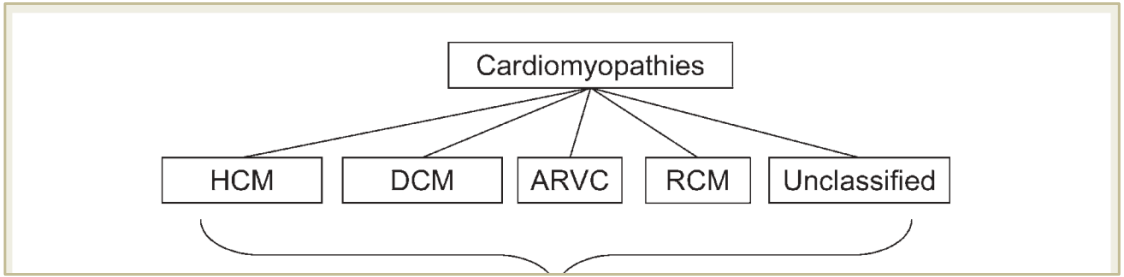
FAKULTNÍ NEMOCNICE U SV. ANNY V BRNĚ

I. INTERNÍ KARDIOANGIOLOGICKÁ KLINIKA FNUSA A LF MU

MUNI MED

Do roku 2023 platilo dělení kardiomyopatií z roku 2008

Classification of the cardiomyopathies: a position statement from the european society of cardiology working group on myocardial and pericardial diseases



European Heart Journal (2008) **29**, 270–276

V roce 2023 byla představena ESC Guidelines pro KMP

Eur Heart J. 2023 Oct 1;44(37):3503-3626.

...co se změnilo a co zůstalo?

Obecná definice kardiomyopatie zůstává…

Kardiomyopatie je definována jako onemocnění srdeční svalu způsobující jeho strukturální či funkční poruchy v nepřítomnosti ischemické choroby srdeční, hypertenze, chlopenní či vrozené vady, které by byly dostatečně významné, aby mohly být podkladem dané patologie.

Eur Heart J. 2023 Oct 1;44(37):3503-3626.

Přístup k rozdělení do značné míry také…

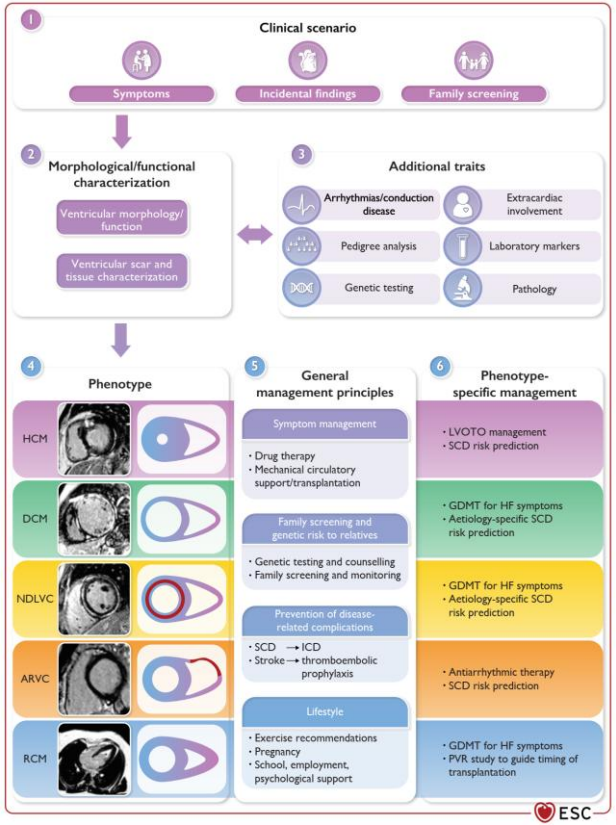
ESC European Society of Cardiology
European Heart Journal (2023) **00**, 1–124
https://doi.org/10.1093/eurheartj/ehad194

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)

Eur Heart J. 2023 Oct 1;44(37):3503-3626.



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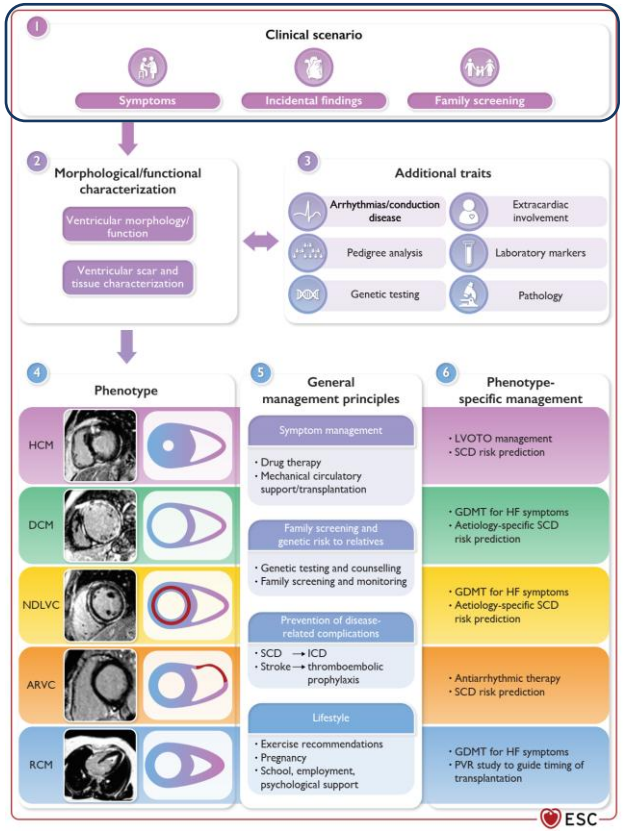
Diagnostická cesta dle ESC Guidelines 2023



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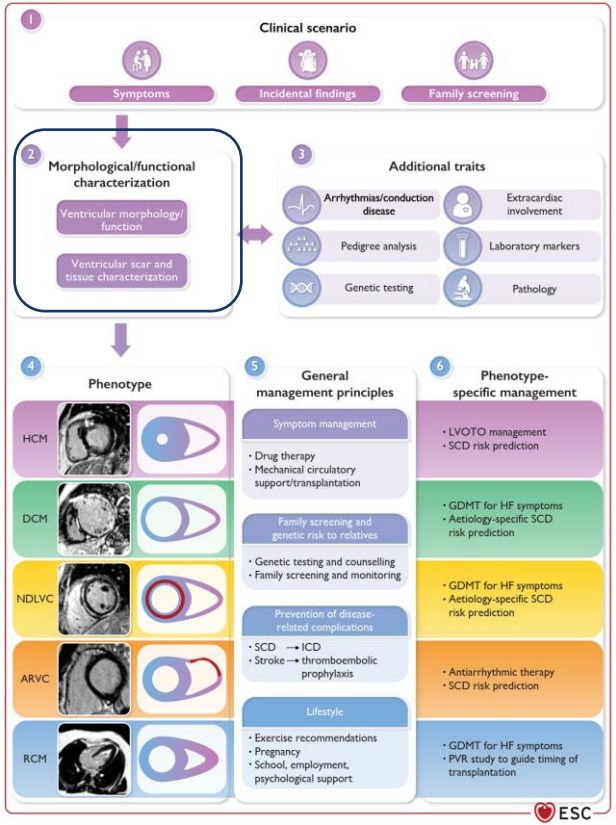
Přístup k rozdělení kardiomyopatií

Morphological/functional characterization

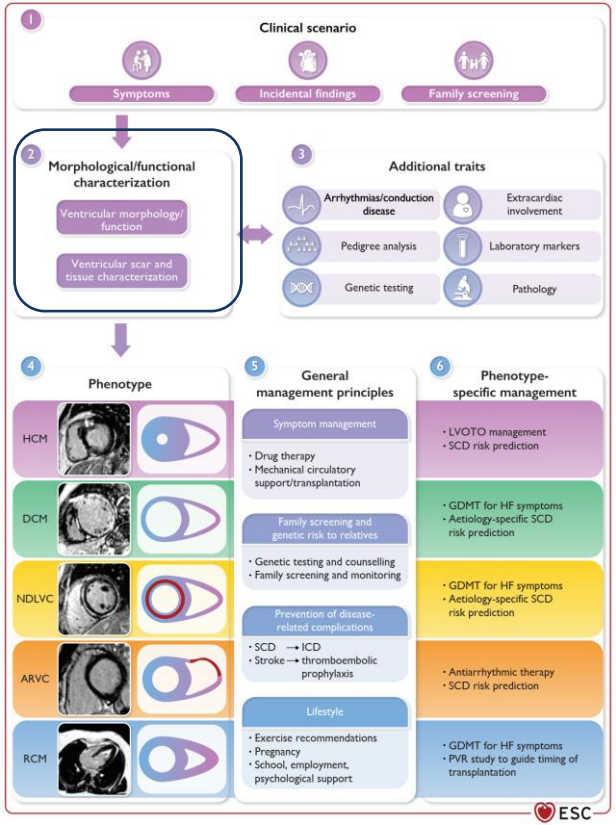
Ventricular morphology/ function



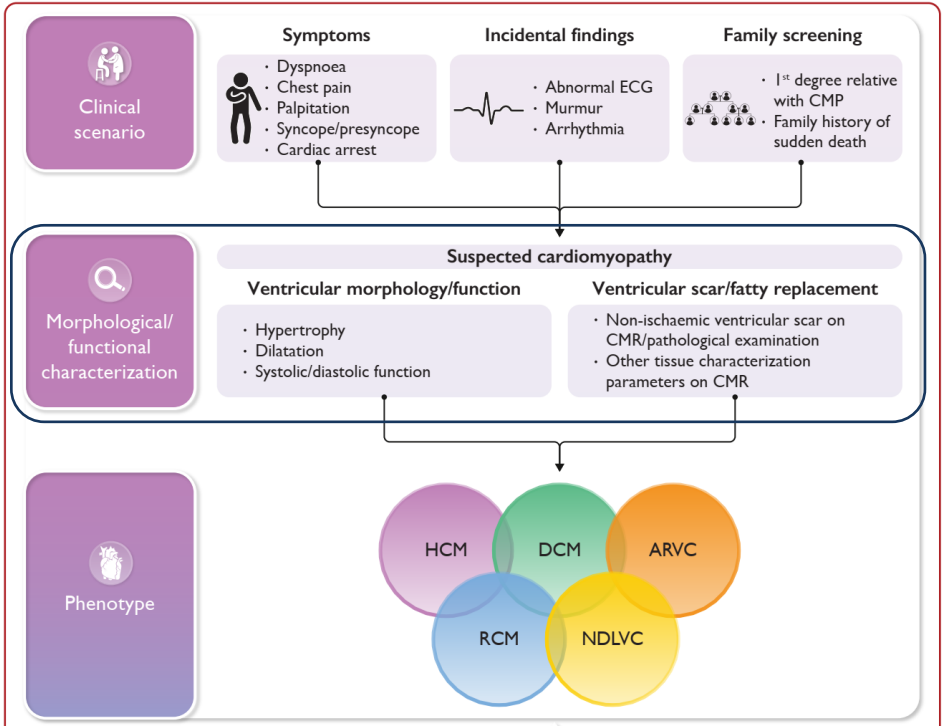
Přístup k rozdělení kardiomyopatií

Morphological/functional characterization

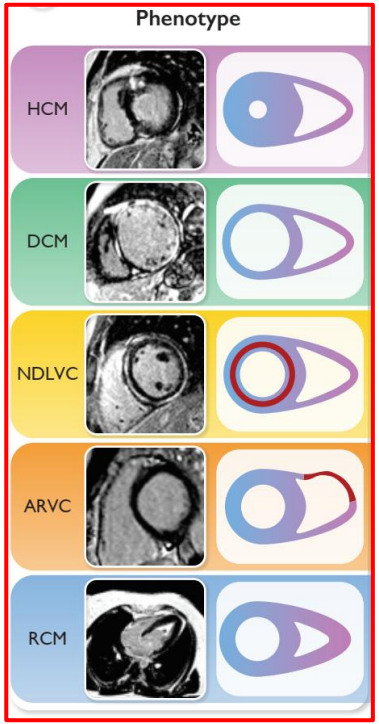
- Ventricular morphology/ function
- Ventricular scar and tissue characterization



Diagnostický algoritmus kardiomyopatií



CMR



Eur Heart J. 2023 Oct 1;44(37):3503-3626.

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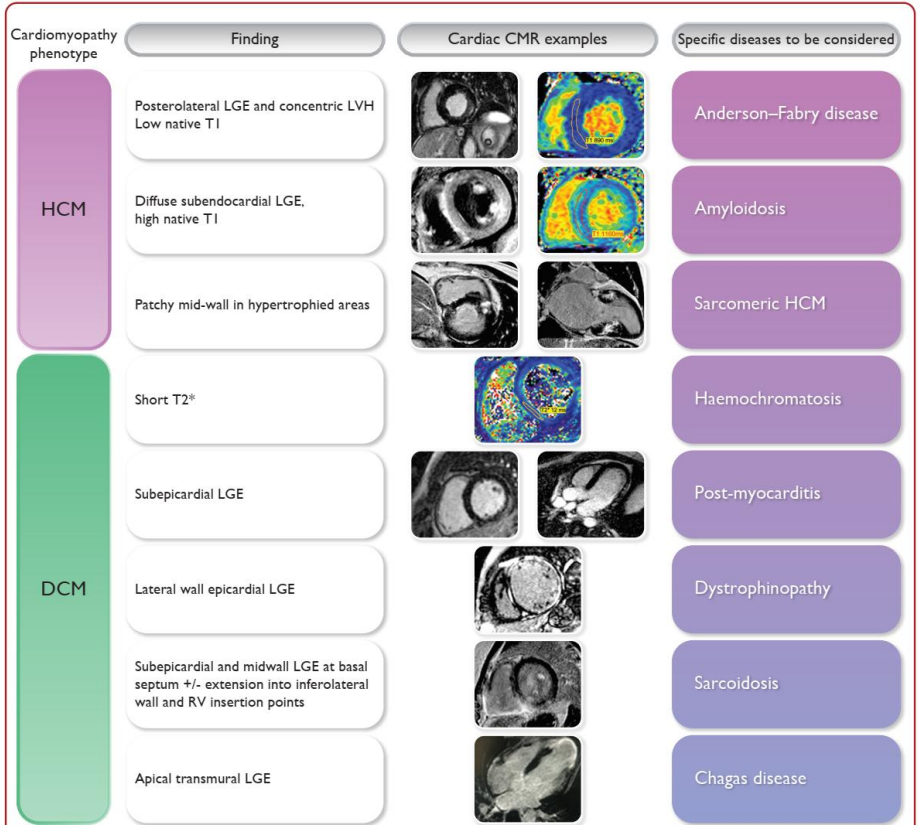
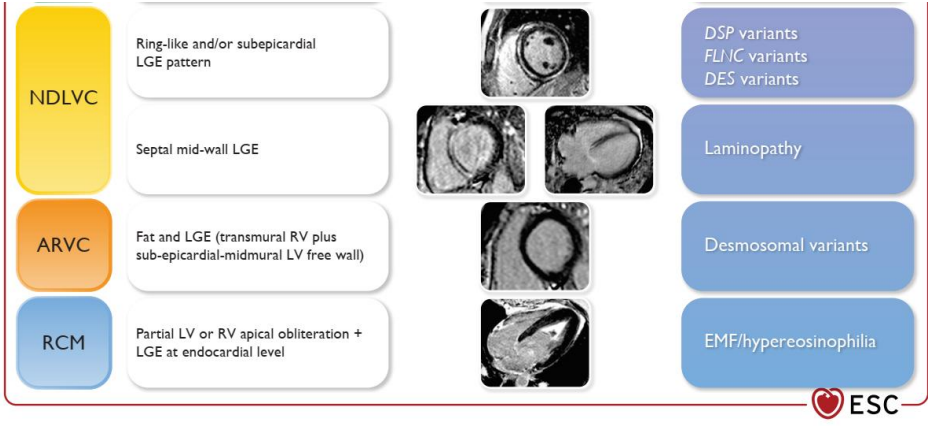
Klíčová role CMR v dg a dif dg všech typů KMP





Eur Heart J. 2023 Oct 1;44(37):3503-3626.

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Klíčová role CMR v dg a dif dg všech typů KMP

URČENÍM FENOTYPU KARDIOMYOPATIE DIAGNOSTIKA NEKONČÍ, NAOPAK JDE O JEJÍ ZAČÁTEK !

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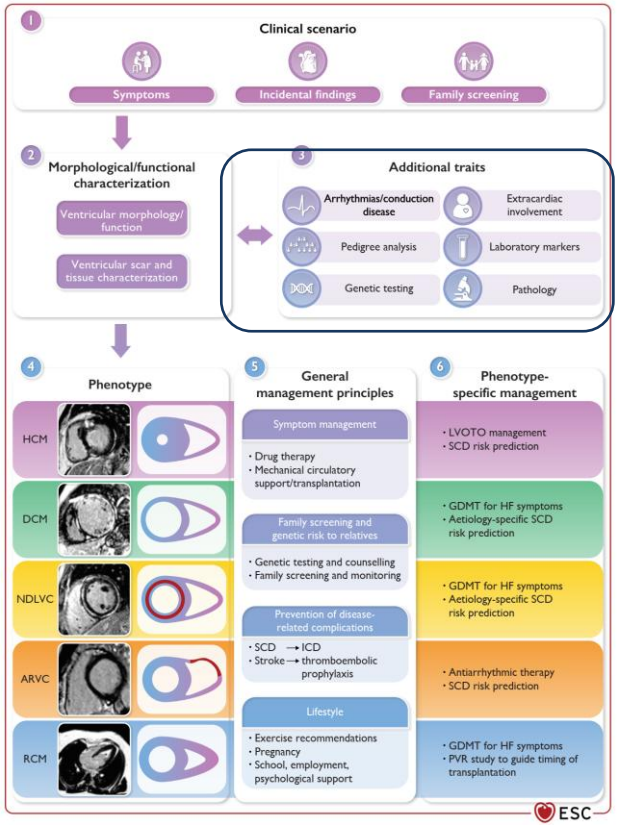
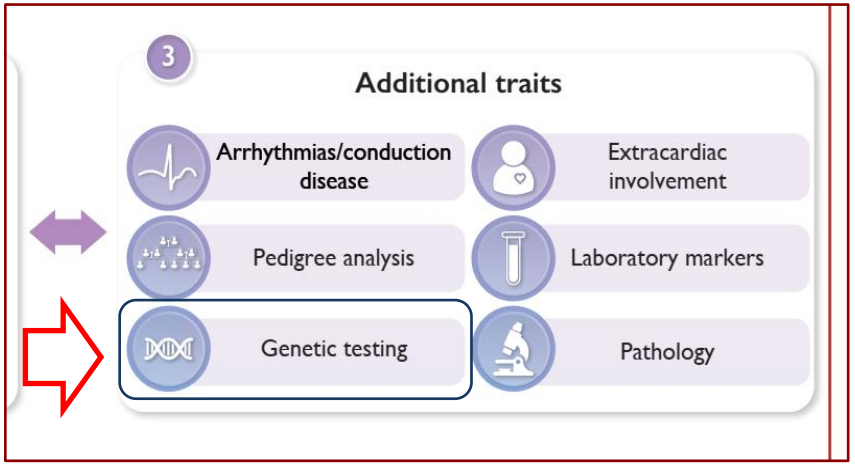
V dg. KMP hraje stále významnější roli genetická diagnostika





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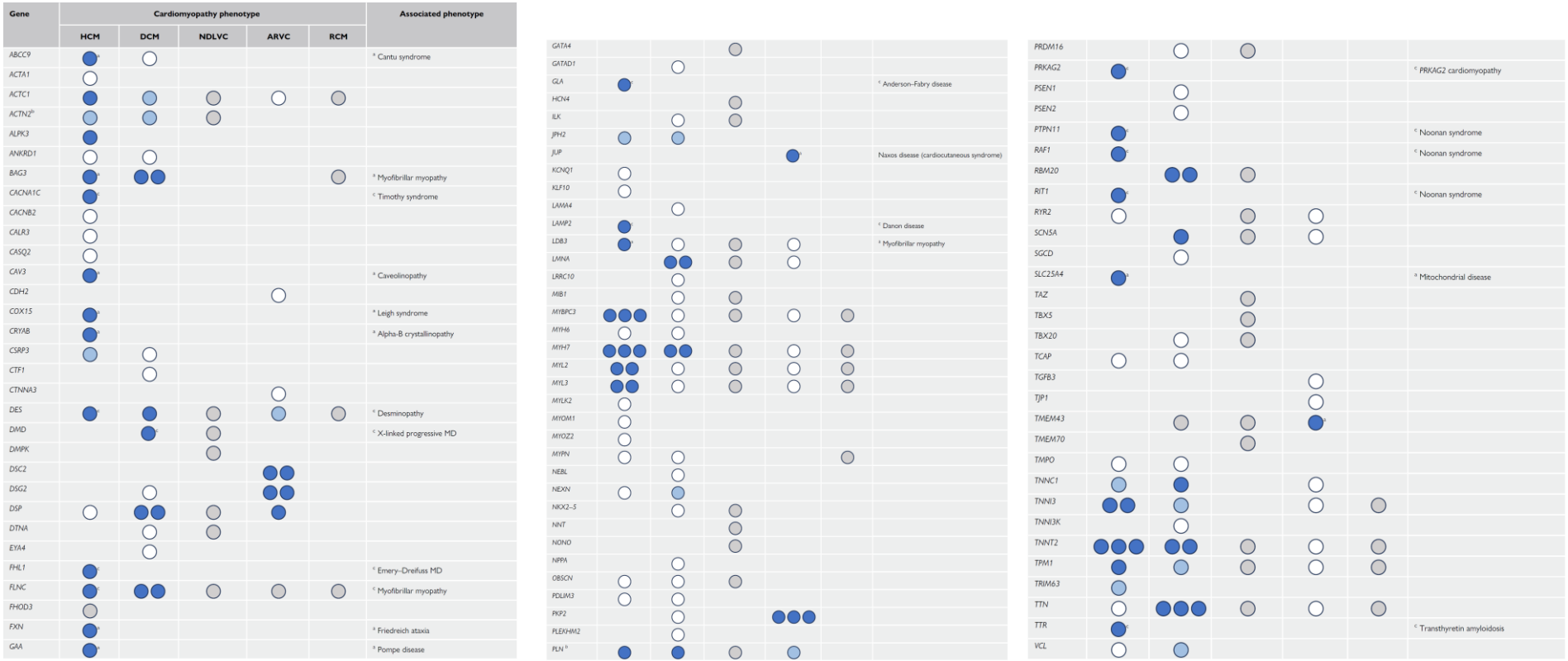
Role genetického testování

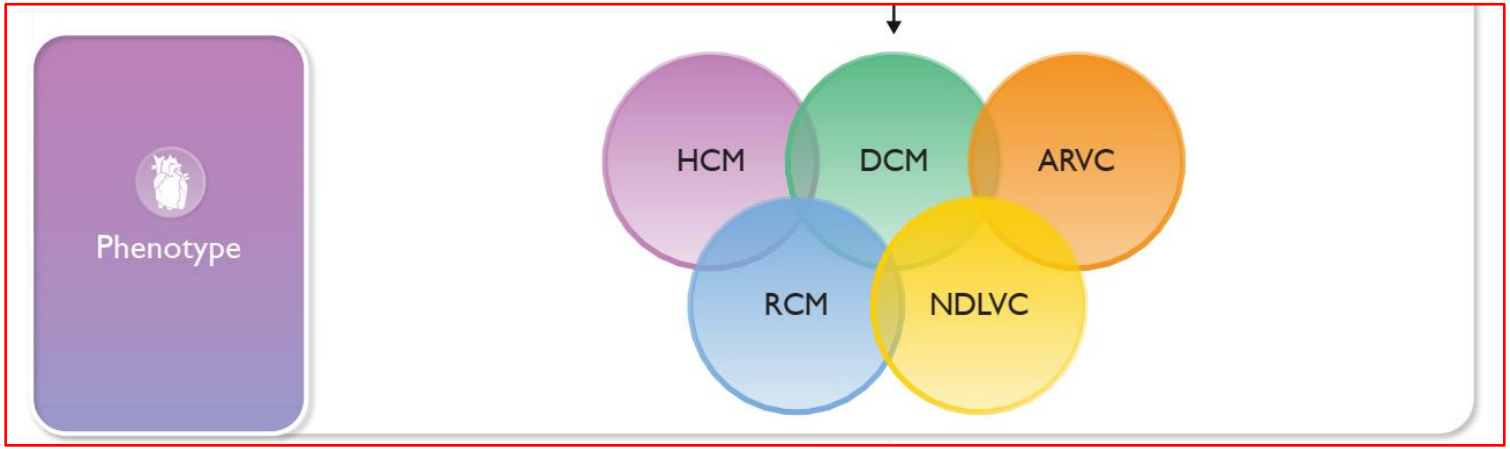


Genetika a identifikace specifické diagnózy

Gene	Cardiomyopathy phenotype					Associated phenotype
	HCM	DCM	NDLVC	ARVC	RCM	
GLA	●[c]					[c] Anderson–Fabry disease
LAMP2	●[c]					[c] Danon disease

Eur Heart J. 2023 Oct 1;44(37):3503-3626.

Fenopyty KMP - model 2023



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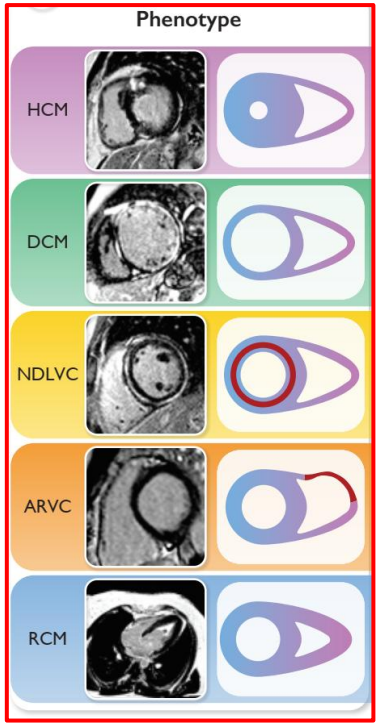
Hypertrofická kardiomyopatie

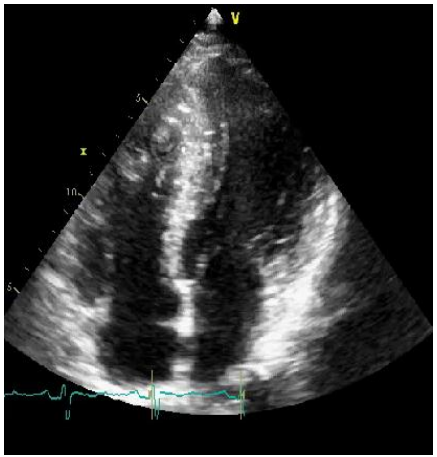


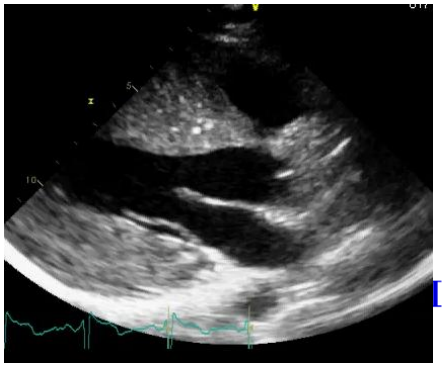
7.1.1. Diagnosis

7.1.1.1. Diagnostic criteria

Adults: in an adult, HCM is defined by an LV wall thickness ≥15 mm in any myocardial segment that is not explained solely by loading conditions. Lesser degrees of wall thickening (13–14 mm) require evaluation of other features including family history, genetic findings, and ECG abnormalities.

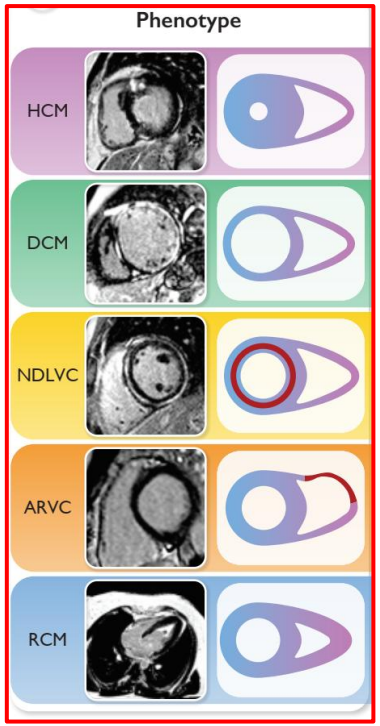
Children: the diagnosis of HCM requires an LV wall thickness more than 2 standard deviations greater than the predicted mean (z-score >2).[578]





Eur Heart J. 2023 Oct 1;44(37):3503-3626.

Dilatační kardiomyopatie

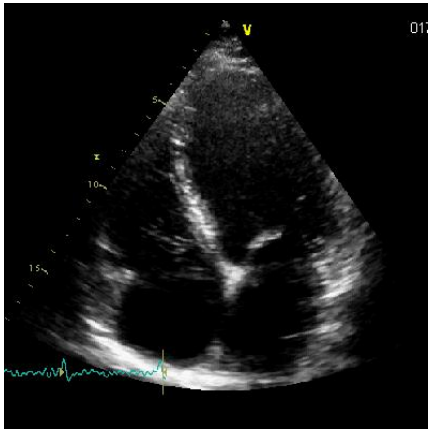


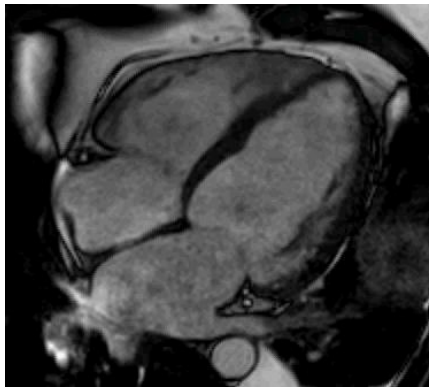
7.2. Dilated cardiomyopathy

7.2.1. Diagnosis

7.2.1.1. Index case

Dilated cardiomyopathy is defined by the presence of LV dilatation and systolic dysfunction unexplained solely by abnormal loading conditions or CAD. Left ventricular dilatation is defined by LV end-diastolic dimensions or volumes >2 z-scores above population mean values corrected for body size, sex, and/or age. For adults this represents an LV end-diastolic diameter >58 mm in males and >52 in females and an LVEDV index of ≥75 mL/m^2 in males and ≥62 mL/m^2 in females by ECHO.[9,845,846] Left ventricular global systolic dysfunction is defined by LVEF <50%.[9]





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Prevence SCD u DCM/NDLVC dle CMP Guid 2023

Recommendation Table 24 — Recommendations for an implantable cardioverter defibrillator in patients with dilated cardiomyopathy

Recommendations	Class[a]	Level[b]
Secondary prevention		
An ICD is recommended to reduce the risk of sudden death and all-cause mortality in patients with DCM who have survived a cardiac arrest or have recovered from a ventricular arrhythmia causing haemodynamic instability.[530,531,884]	I	B
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	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

Recommendation Table 26 — Recommendations for an implantable cardioverter defibrillator in patients with non-dilated left ventricular cardiomyopathy

Recommendations	Class[a]	Level[b]
Secondary prevention		
An ICD is recommended to reduce the risk of sudden death and all-cause mortality in patients with NDLVC who have survived a cardiac arrest or have recovered from a ventricular arrhythmia causing haemodynamic instability.	I	C
Primary prevention		
An ICD should be considered to reduce the risk of sudden death and all-cause mortality in patients with NDLVC, heart failure symptoms, and LVEF ≤35% despite >3 months of OMT.[861,885]	IIa	A
The patient's genotype should be considered in the	IIa	C
An ICD should be considered in patients with NDLVC with a genotype associated with high SCD risk and LVEF >35% in the presence of additional risk factors (see *Table 21*).[185,186,438,541,542,865–869,878–883]	IIa	C
An ICD may be considered in selected patients with NDLVC with a genotype associated with high SCD risk and LVEF >35% without additional risk factors (see *Table 21*).	IIb	C
An ICD may be considered in patients with NDLVC without a genotype associated with high SCD risk and LVEF >35% in the presence of additional risk factors.[c]	IIb	C

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%

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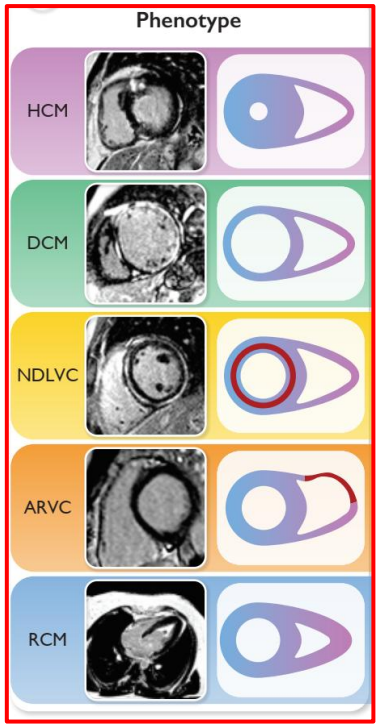
Prevence SCD u DCM dle Arytmo Guid 2022

Recommendation Table 28 — Recommendations for risk stratification, sudden cardiac death prevention, and treatment of ventricular arrhythmias in dilated cardiomyopathy/hypokinetic non-dilated cardiomyopathy

Risk stratification and primary prevention of SCD		
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.[357,359,635,650]	IIa	A
ICD implantation should be considered in DCM/HNDCM patients with a pathogenic mutation in *LMNA* gene, if the estimated 5-year risk of life-threatening VA is ≥10%[c] *and* in the presence of NSVT or LVEF < 50% or AV conduction delay.[80,652,653]	IIa	B
ICD implantation should be considered in DCM/HNDCM patients with a LVEF <50% and ≥2 risk factors (syncope, LGE on CMR, inducible SMVT at PES, pathogenic mutations in *LMNA*,[d] *PLN*, *FLNC*, and *RBM20* genes).	IIa	C

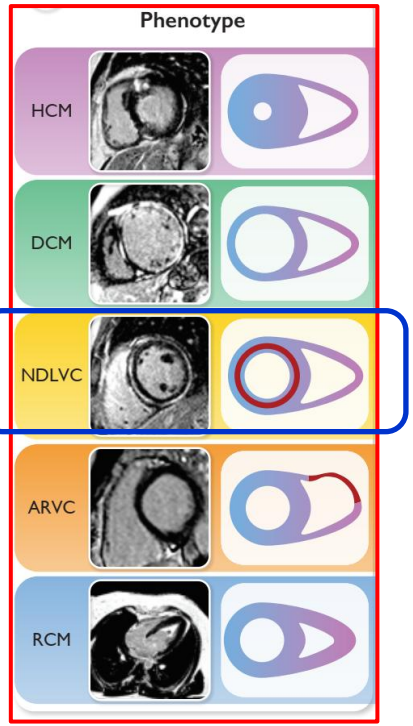
Nedilatovaná kardiomyopatie levé komory – novinka roku 2023



In this guideline, we propose replacement of this term with non-dilated left ventricular cardiomyopathy (NDLVC), which can be further characterized by the presence or absence of systolic dysfunction (regional or global). Isolated LV dysfunction (regional or global) without scarring should also be considered under this diagnostic category.

The NDLVC phenotype is defined as the presence of non-ischaemic LV scarring or fatty replacement regardless of the presence of global or regional wall motion abnormalities (RWMAs), or isolated global LV hypokinesia without scarring.

ESC Guidelines 2023



Morphological/functional characterization

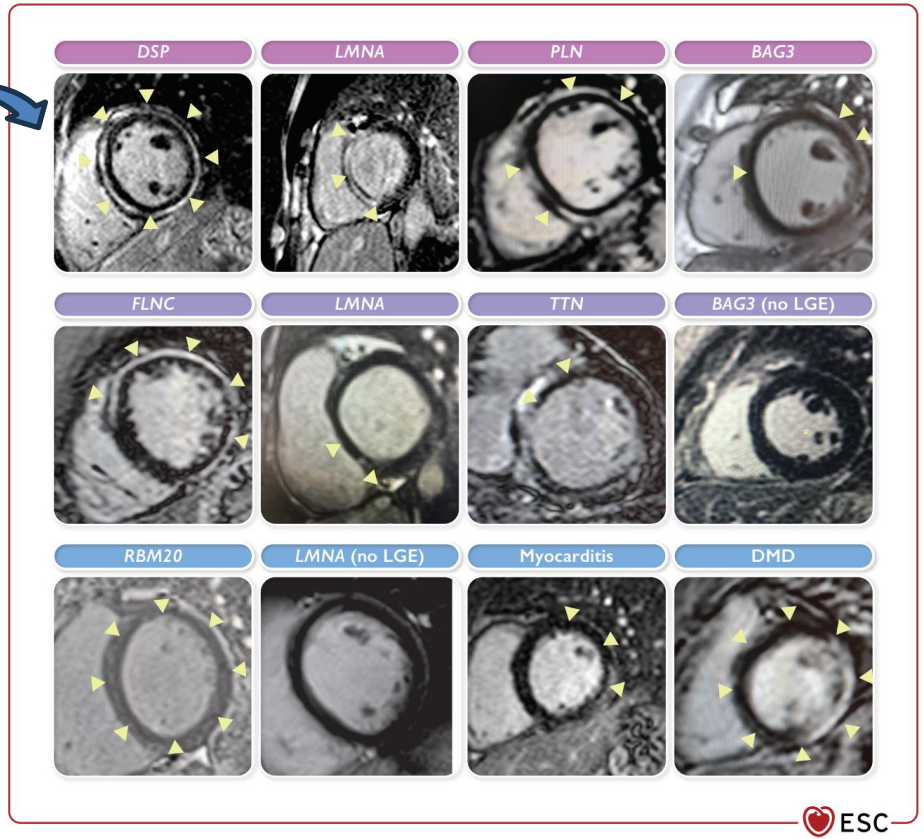
- Ventricular morphology/ function
- Ventricular scar and tissue characterization

Řada různých diagnóz s odlišnou prognózou a léčbou

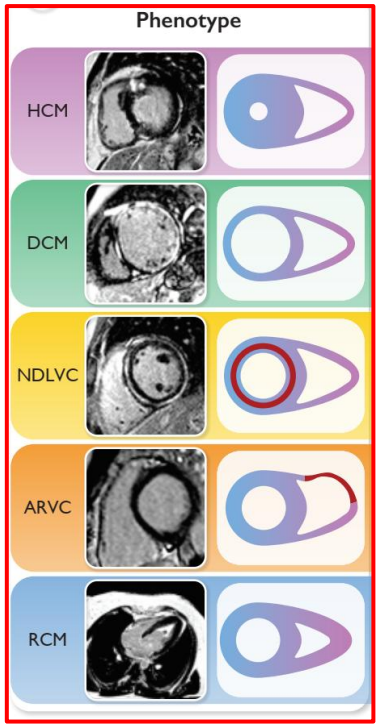
Eur Heart J. 2023 Oct 1;44(37):3503-3626.

Nedilatovaná kardiomyopatie levé komory (NDLVC)

Levokomorová či biventrikulární forma ACM



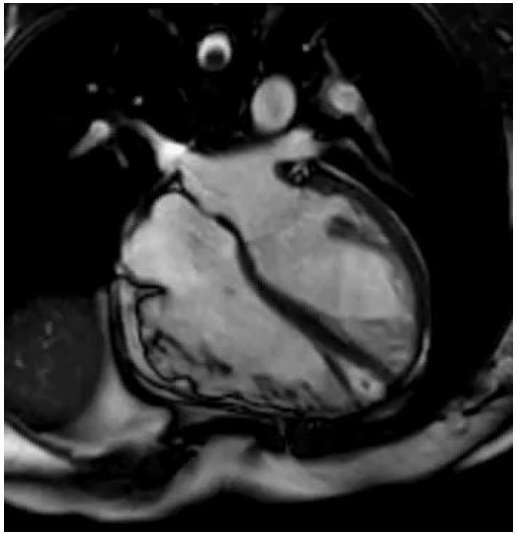
Arytmogenní kardiomyopatie pravé komory



3.2.4. Arrhythmogenic right ventricular cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is defined as the presence of predominantly RV dilatation and/or dysfunction in the presence of histological involvement and/or electrocardiographic abnormalities in accordance with published criteria.[10]

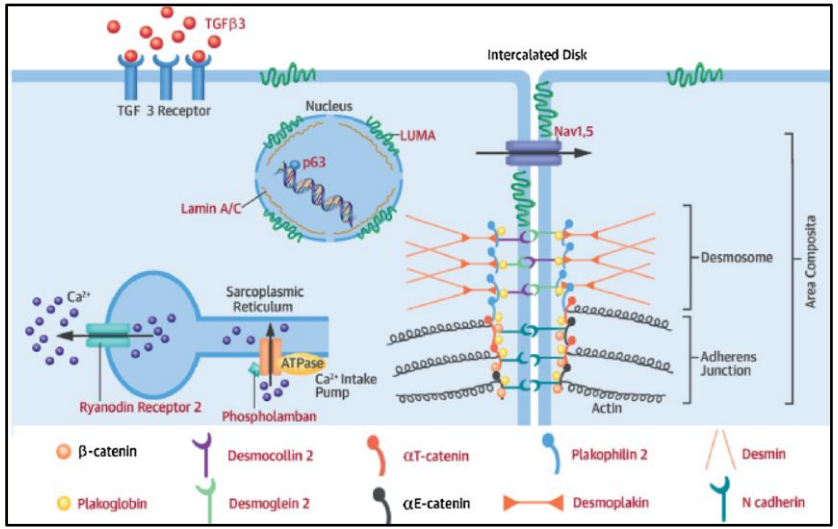
For decades, ARVC has been one of the principal cardiomyopathy subtypes. It has been defined in accordance with published consensus criteria that comprise RV dysfunction (global or regional), histological abnormalities in the form of fibro-fatty replacement of cardiomyocytes, electrocardiographic characteristics, ventricular arrhythmia of RV origin, and the presence of familial disease and/or pathogenic variants in desmosomal protein genes.

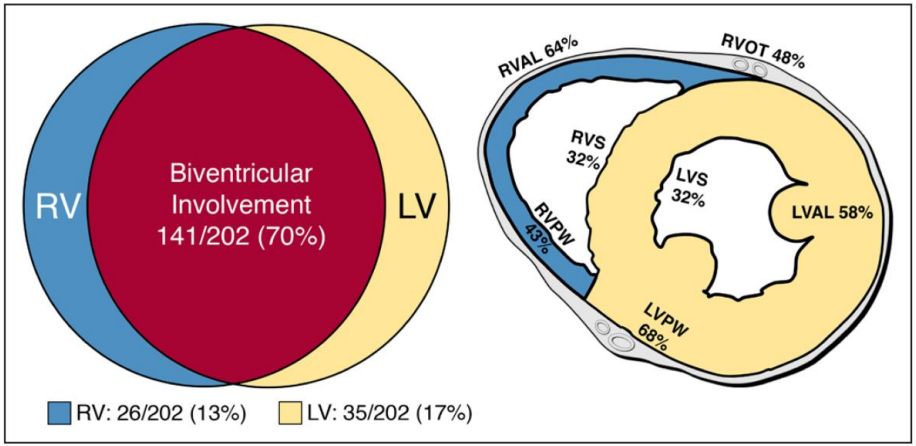


Eur Heart J. 2023 Oct 1;44(37):3503-3626.

Arytmogenní kardiomyopatie nepostihuje jen pravou komoru…

Arrhythmogenic Cardiomyopathy in 2018: ARVC/ALVC or both?



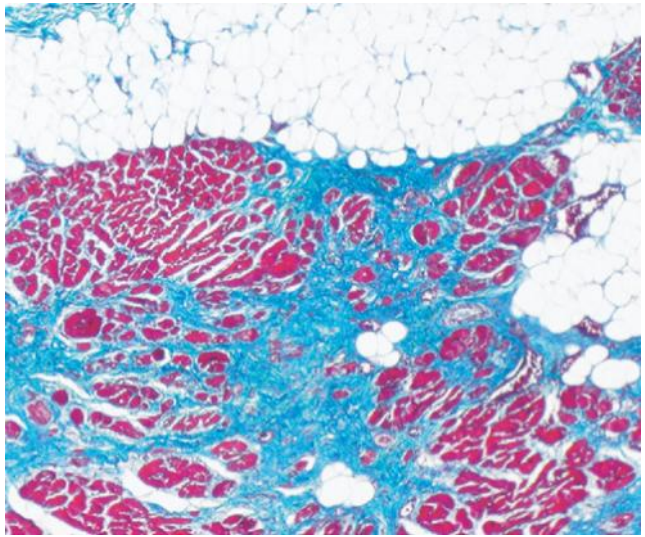


Circulation. 2019;139:1786–1797.

Diagnostika biventrikulárních forem ACM – „Padua criteria“

Diagnosis of arrhythmogenic cardiomyopathy: The Padua criteria

D. Corrado et al. / International Journal of Cardiology 319 (2020) 106–114



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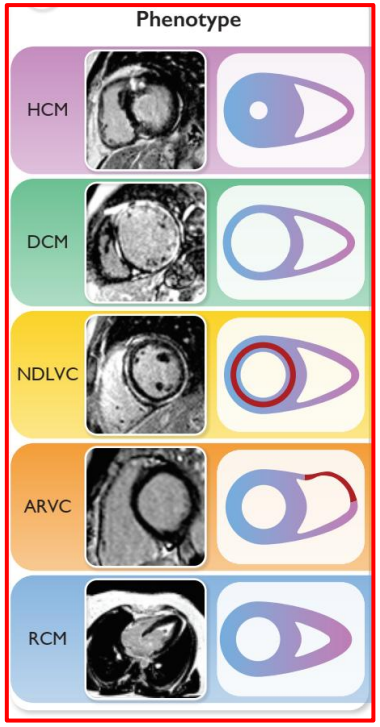
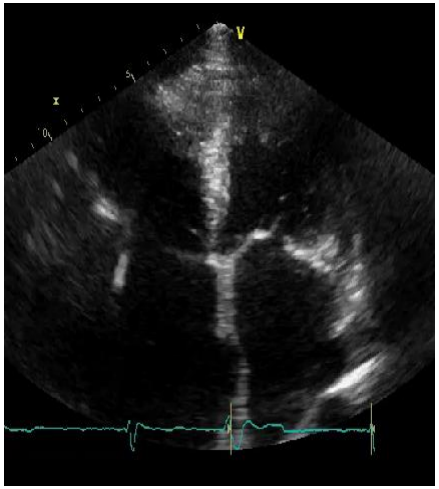
"Padua criteria" for diagnosis of Arrhythmogenic Cardiomyopathy.

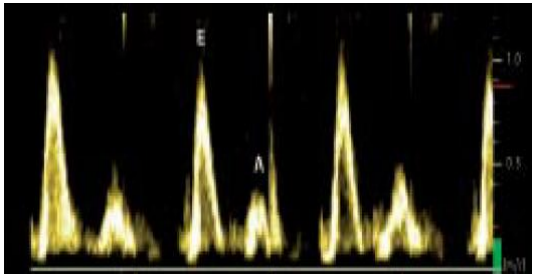
Category	Right ventricle (upgraded 2010 ITF diagnostic criteria)	Left ventricle (new diagnostic criteria)
I. Morpho-functional ventricular abnormalities	*By echocardiography, CMR or angiography:* *Major* • Regional RV akinesia, dyskinesia, or bulging *plus* one of the following: - global RV dilatation (increase of RV EDV according to the imaging test specific nomograms) - global RV systolic dysfunction (reduction of RV EF according to the imaging test specific nomograms) *Minor* • Regional RV akinesia, dyskinesia or aneurysm of RV free wall	*By echocardiography, CMR or angiography:Minor* • Global LV systolic dysfunction (depression of LV EF or reduction of echo-cardiographic global longitudinal strain), with or without LV dilatation (increase of LV EDV according to the imaging test specific nomograms for age, sex, and BSA) *Minor* • Regional LV hypokinesia or akinesia of LV free wall, septum, or both
II. Structural myocardial abnormalities	*By CE-CMR:Major* • Transmural LGE (stria pattern) of ≥1 RV region(s) (inlet, outlet, and apex in 2 orthogonal views) *By EMB (limited indications):Major* • Fibrous replacement of the myocardium in ≥1 sample, with or without fatty tissue	*By CE-CMR:Major* • LV LGE (stria pattern) of ≥1 Bull's Eye segment(s) (in 2 orthogonal views) of the free wall (subepicardial or midmyocardial), septum, or both (excluding septal junctional LGE)
III. Repolarization abnormalities	*Major* • Inverted T waves in right precordial leads (V_1,V_2, and V_3) or beyond in individuals with complete pubertal development (in the absence of complete RBBB) *Minor* • Inverted T waves in leads V1 and V2 in individuals with completed pubertal development (in the absence of complete RBBB) • Inverted T waves in V1,V2,V3 and V4 in individuals with completed pubertal development in the presence of complete RBBB.	*Minor* • Inverted T waves in left precordial leads (V_4-V_6) (in the absence of complete LBBB)
IV. Depolarization abnormalities	*Minor* • Epsilon wave (reproducible low-amplitude signalsbetween end of QRS complex to onset of the T wave) in the right precordial leads (V1 to V3) • Terminal activation duration of QRS ≥55 ms measured from the nadir of the S wave to the end of the QRS, including R', in V1, V2, or V3 (in the absence of complete RBBB)	*Minor* • Low QRS voltages (<0.5 mV peak to peak) in limb leads (in the absence of obesity, emphysema, or pericardial effusion)
V. Ventricular arrhythmias	*Major* • Frequent ventricular extrasystoles (>500 per 24 h), non-sustained or sustained ventricular tachycardia of LBBB morphology *Minor* • Frequent ventricular extrasystoles (>500 per 24 h), non-sustained or sustained ventricular tachycardia of LBBB morphology with inferior axis ("RVOT pattern")	*Minor* • Frequent ventricular extrasystoles (>500 per 24 h), non-sustained or sustained ventricular tachycardia with a RBBB morphology (excluding the "fascicular pattern")
VI. Family history/genetics	*Major* • ACM confirmed in a first-degree relative who meets diagnostic criteria • ACM confirmed pathologically at autopsy or surgery in a first degree relative • Identification of a pathogenic or likely pathogenetic ACM mutation in the patient under evaluation *Minor* • History of ACM in a first-degree relative in whom it is not possible or practical to determine whether the family member meets diagnostic criteria • Premature sudden death (<35 years of age) due to suspected ACM in a first-degree relative • ACM confirmed pathologically or by diagnostic criteria in a second-degree relative	

Restriktivní kardiomyopatie



- je charakterizovaná restriktivní hemodynamikou
- morfologicky jde o onemocnění s normální (či téměř normální) LVEF a normálními/sníženými objemy jedné nebo obou komor
- tloušťka stěn LK by neměla být zvýšená



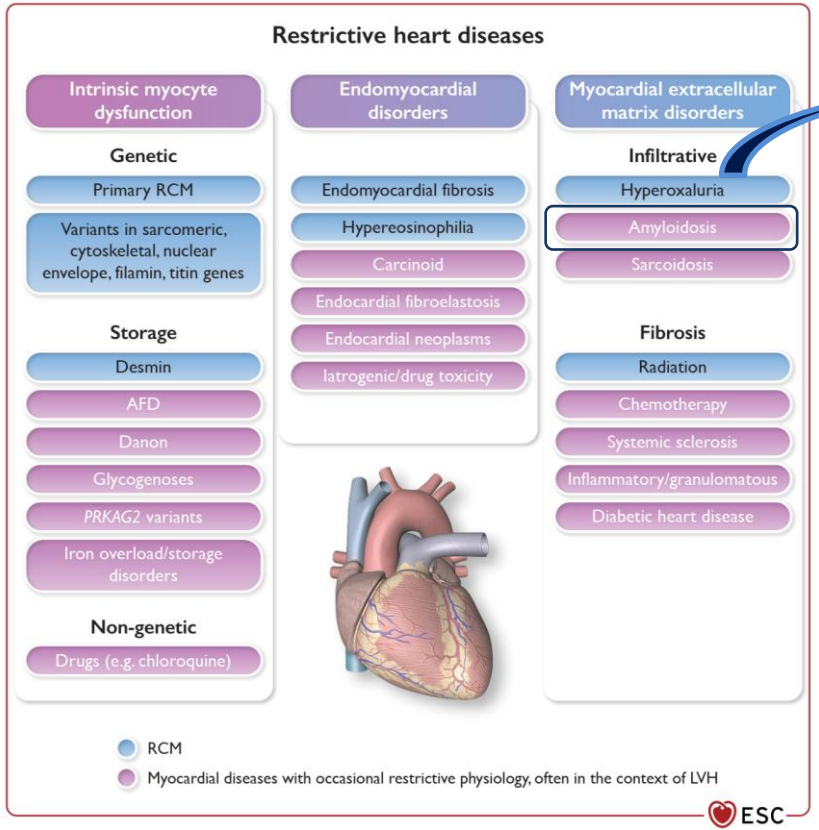


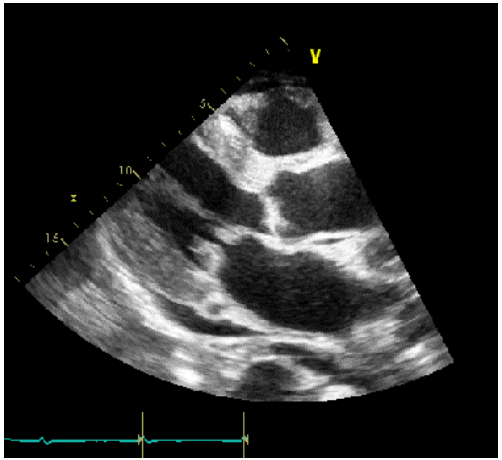
FAKULTNÍ NEMOCNICE U SV. ANNY V BRNĚ

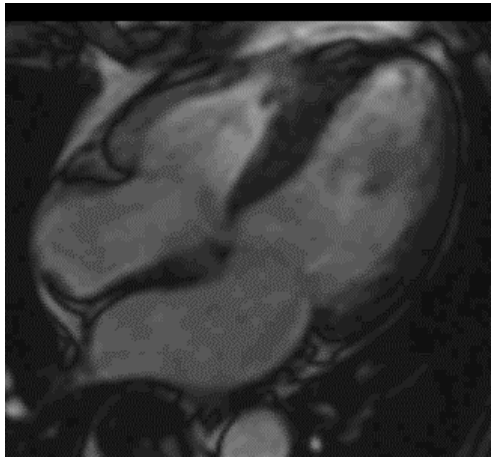
I. INTERNÍ KARDIOANGIOLOGICKÁ KLINIKA FNUSA A LF MU

Eur Heart J. 2023 Oct 1;44(37):3503-3626.

Má srdeční amyloidóza typicky fenotyp RKMP?

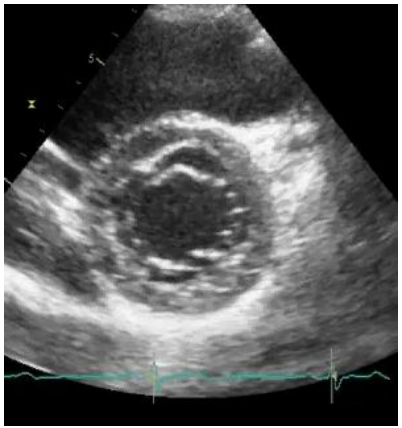


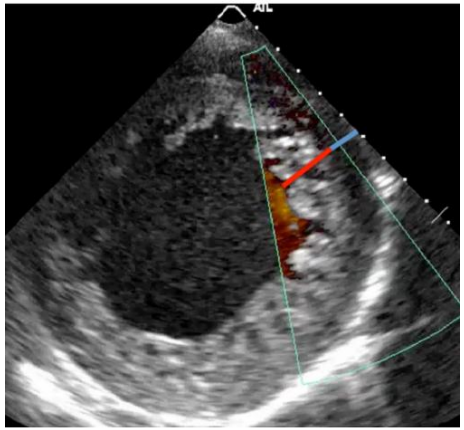




MUNI MED

Co už dnes nepokládáme za kardiomyopatii?





LVNC již nepoužívat, nově „hypertrabekularizace“ levé komory

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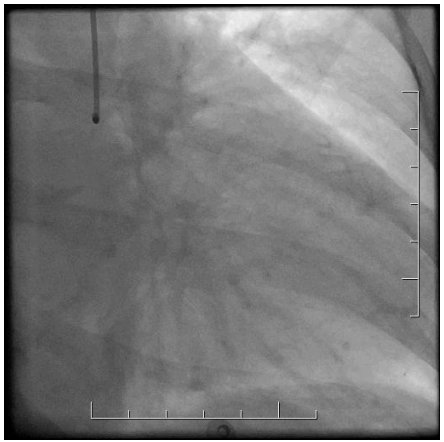
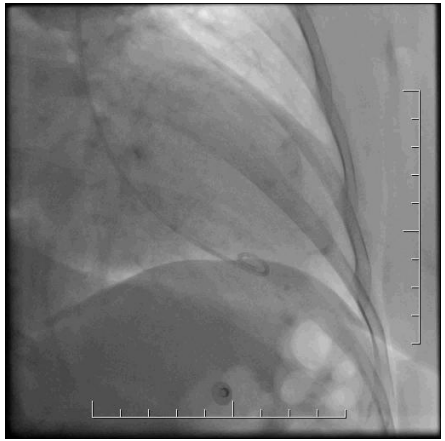
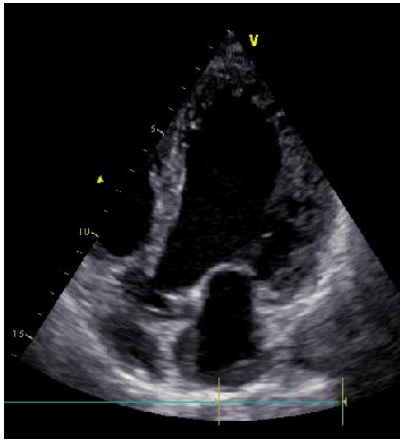
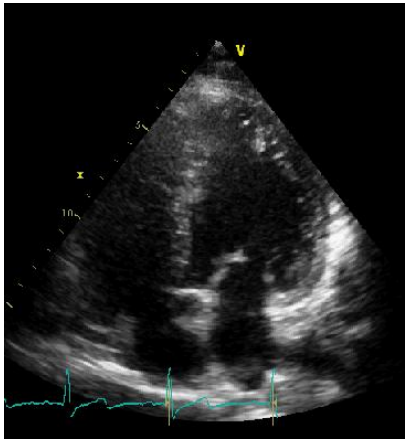
Co už dnes nepokládáme za kardiomyopatii?

Tako-tsubo sy není pokládán za KMP

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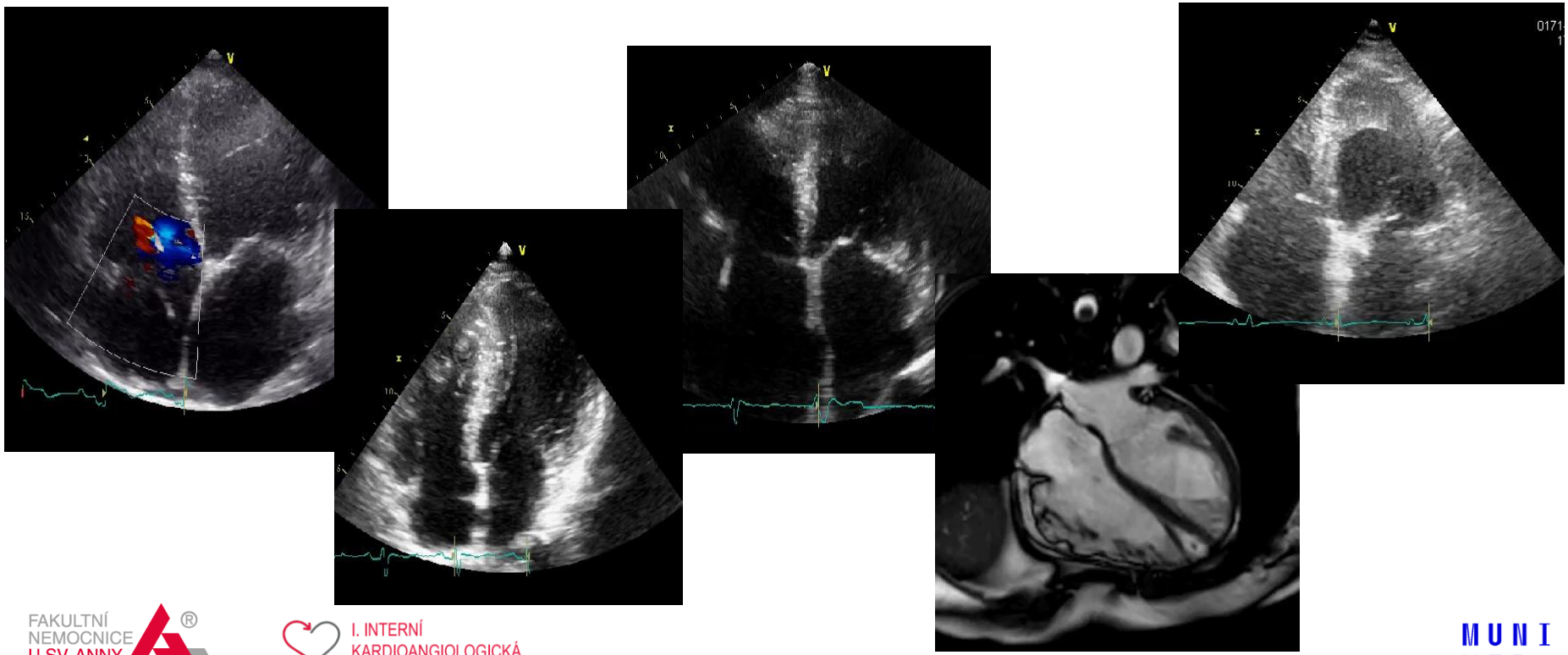
Kardiomyopatie zůstávají velmi různorodou skupinou onemocnění…



Kardiomyopatie jsou velmi různorodá skupina onemocnění…

…jejichž diagnostika, diferenciální diagnostika a léčba jsou náročný a komplexní proces.

Děkuji za pozornost!