Volná sdělení - Arytmogenní kardiomyopatie jako příčina komorové tachykardie

Fakultní nemocnice Olomouc a Lékařská fakulta Univerzity Palackého v Olomouci

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1. Interní klinika - kardiologická FNOL

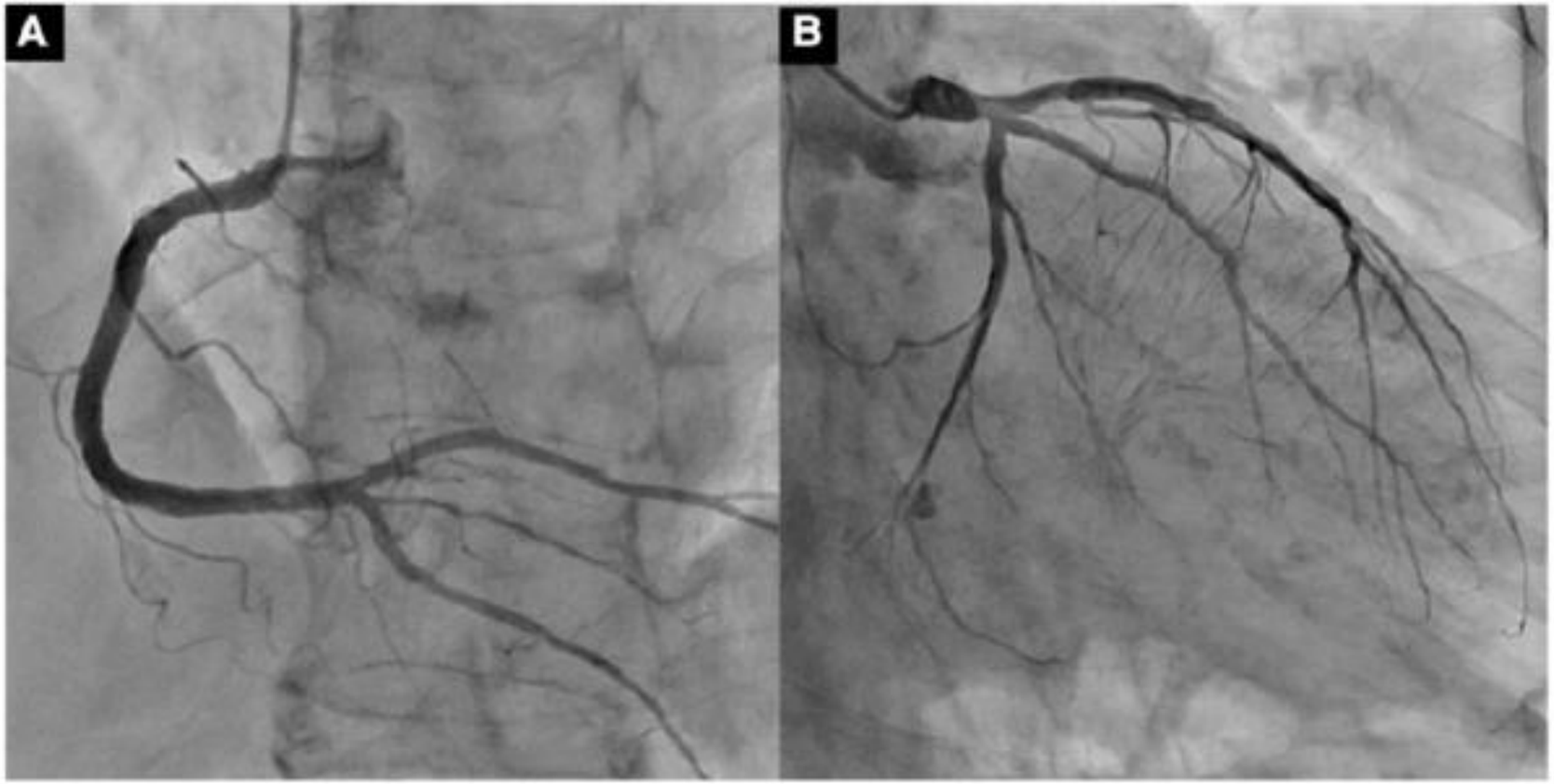
XXXIV. Výroční sjezd české kardiologické společnosti, 9.-12.květen 2026

Vladimír M. – muž (1974)

Anamnéza:

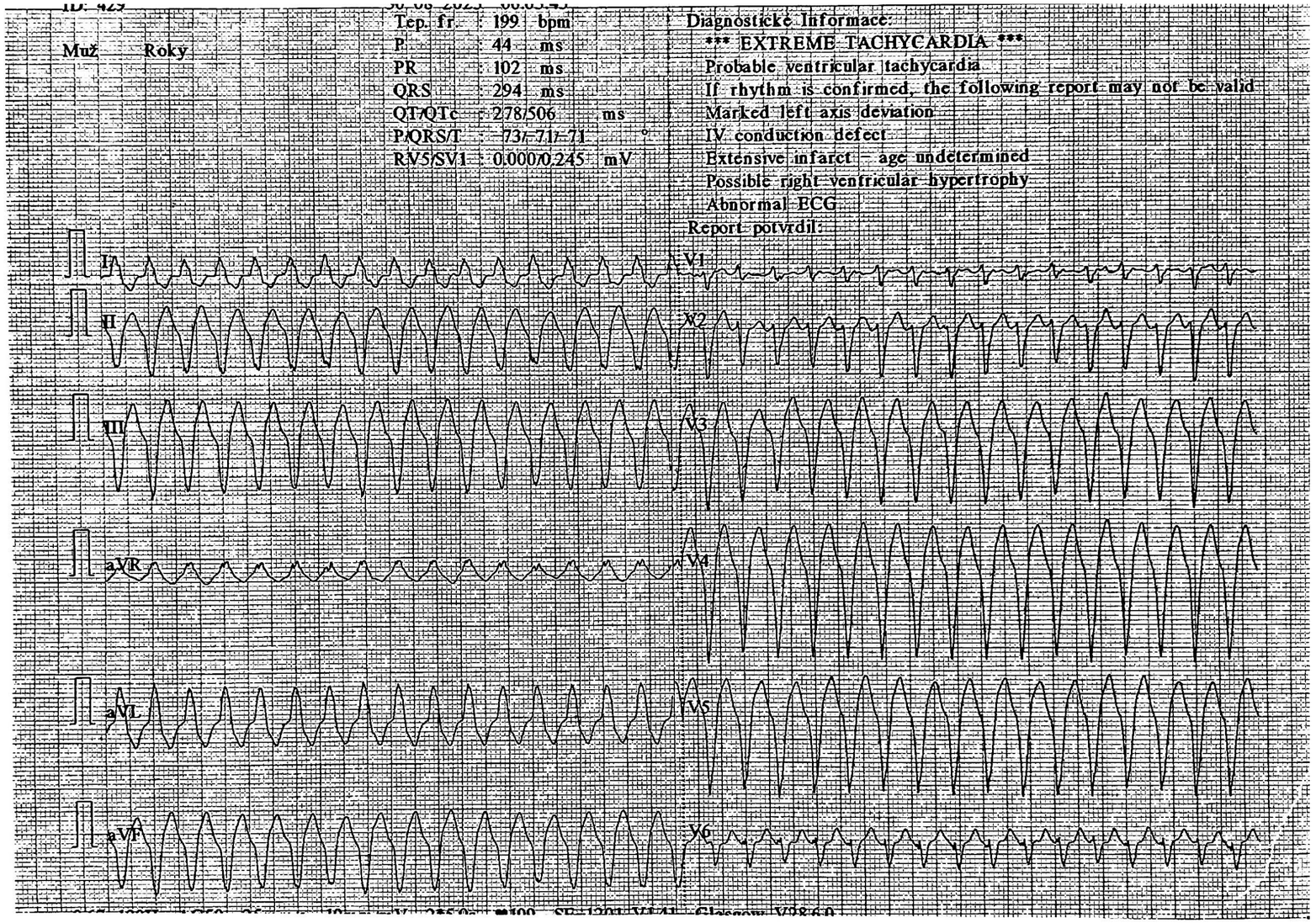
- Anamnesticky v 90. letech vyšetřován pro komorovou ektopii, provedeno echokardiografické vyšetření a EMB s normálním nálezem
- Bez interních onemocnění
- Bez chronické medikace
- V mládí vrcholový sportovec - přestal na doporučení lékaře
- Nekuřák

- Pacient byl přijatý překladem ze spádové nemonice pro hemodynamickou dobře tolerovanou setrvalou monomorní komorovou tachykardii, ve spádu provedena farmakologická kardioverze amiodaronem na SR
- Subjektivně palpitace, vertigo a píchavá bolest v oblasti srdečního hrotu
- Dle laboratoře ze spádu: mírná dynamika v troponinu (500...540ng/l)



https://www.researchgate.net/figure/Coronary-angiography-revealed-no-significant-stenosis-in-the-right-coronary-artery-A_fig2_374473342

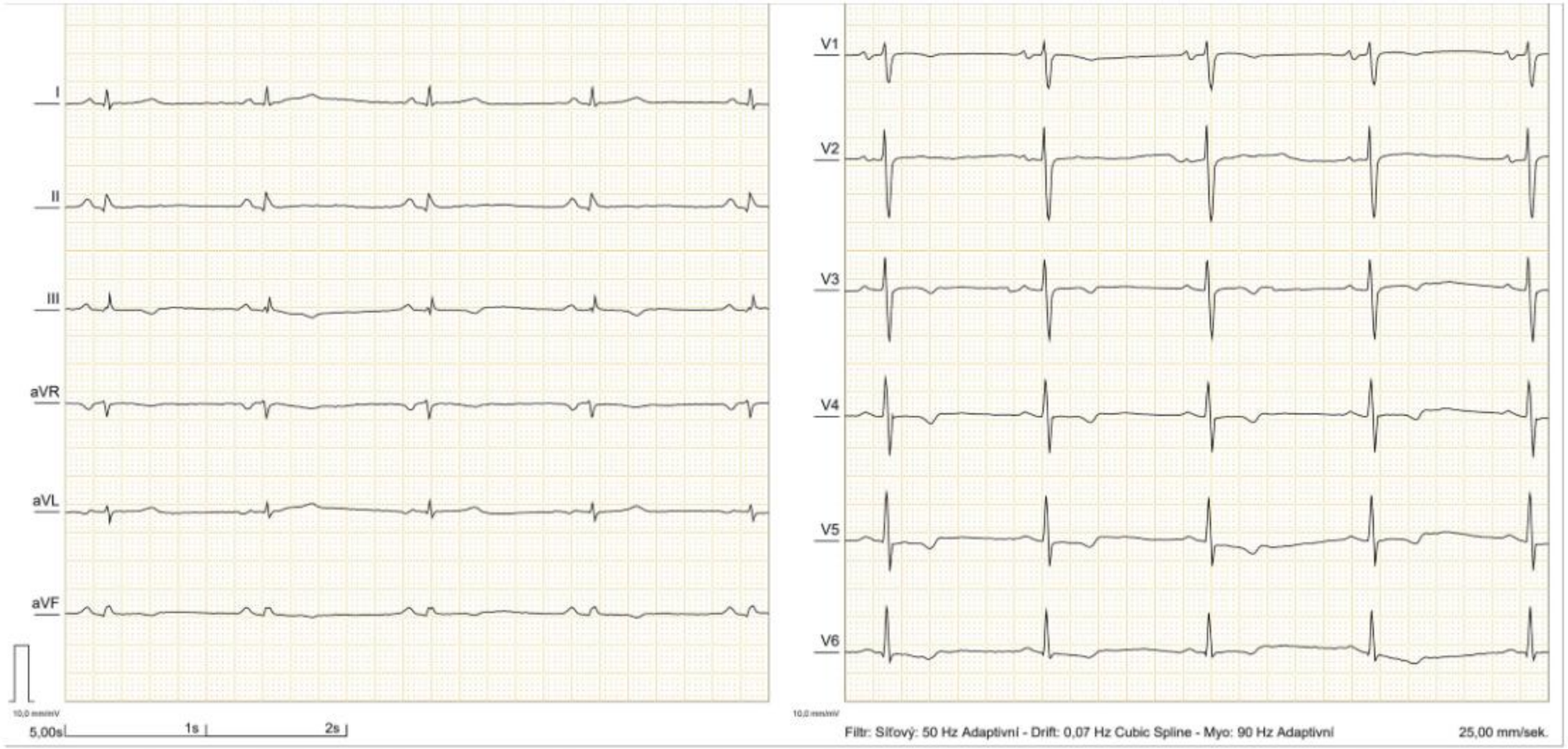
- Provedena selektivní koronarografie bez průkazu významných stenóz



Nemocnice AGEL - Jeseník

Komorová tachykardie

- Setrvalá KT
- Monomorfní
- Tvar LBBB
- Frekvence 180-200/min
- Hemodynamicky relativně dobře tolerovaná
- Farmakologická kardioverze na SR Amiodaronem i.v.
- Extrémní deviace osy srdeční



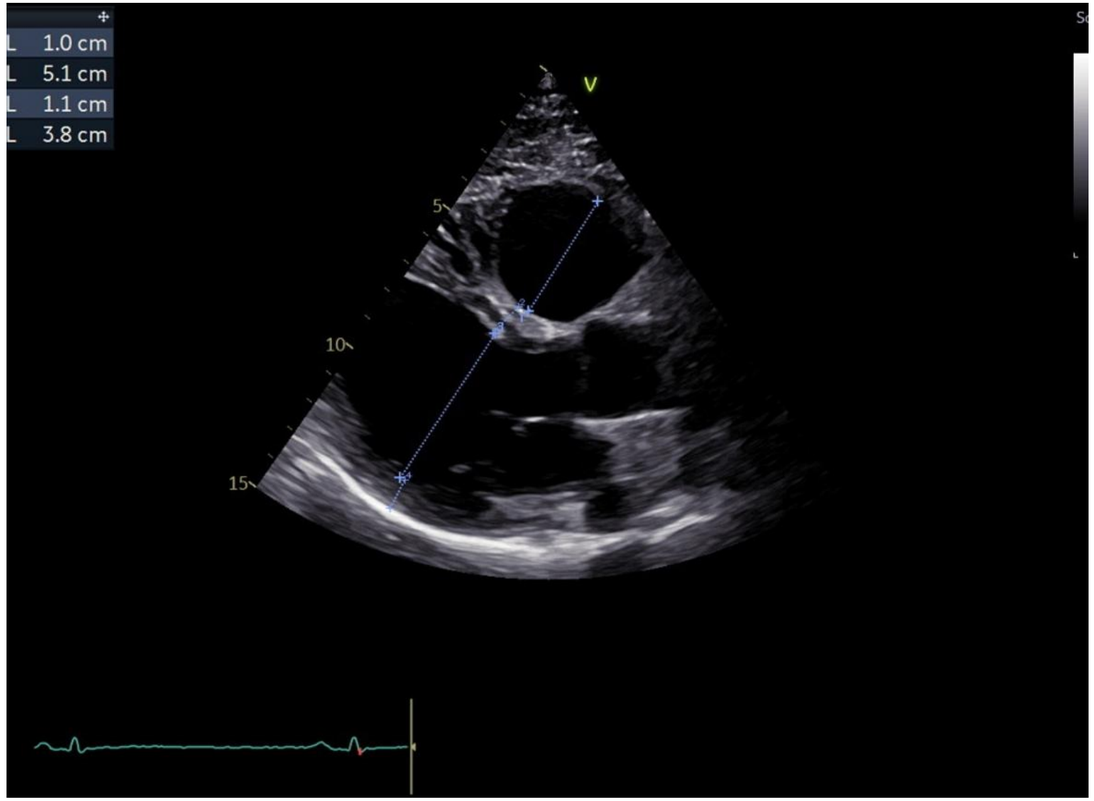
BTL EKG 1.interní klinika kardiologická FNOL

Klidové EKG při příjmu

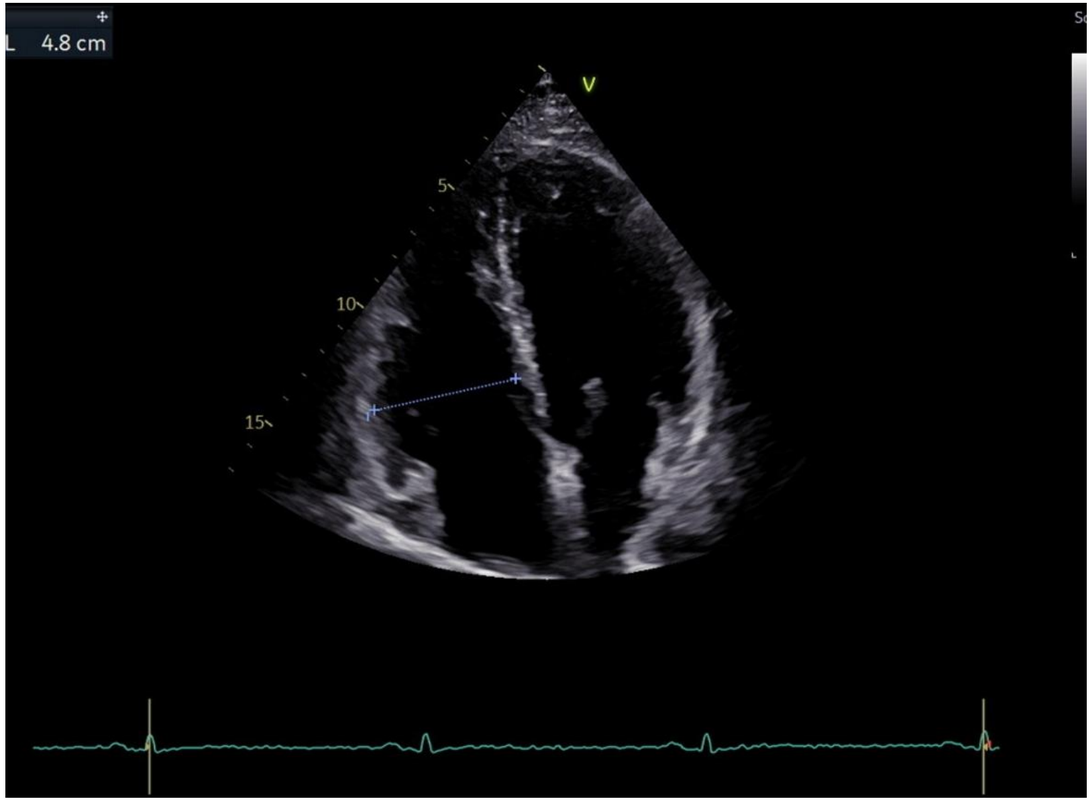
Vstupní laboratorní odběry

- **Biochemie:** Na 135 mmol/l, K 4,3 mmol/l, Mg 0,74 mmol/l, urea 9,3 mmol/l, kreatinin 98 mmol/l, qGFkrea 1,27 ml/s/1,73m2, NTproBNP 1480 ng/l, CRP 26,5 mmol/l, kyselina močová 511 mmol/l, troponin 733 ng/l, Cholesterol 3,91 mmol/l, TAG 0,6 mmol/l, HDL 1,15 mmol/L, LDL 2,49 mmol/l
- **Krevní obraz:** leukocyty 17,35 x 10 na 9/L, hemoglobin 139 g/L, trombocyty 196 x 10 na 9/L
- **Koagulace:** INR: 1,36, aPTT 29,8 s

Echokardiografické vyšetření



Z archívu I . interní kardiologické kliniky FNOL, dr. Koubek

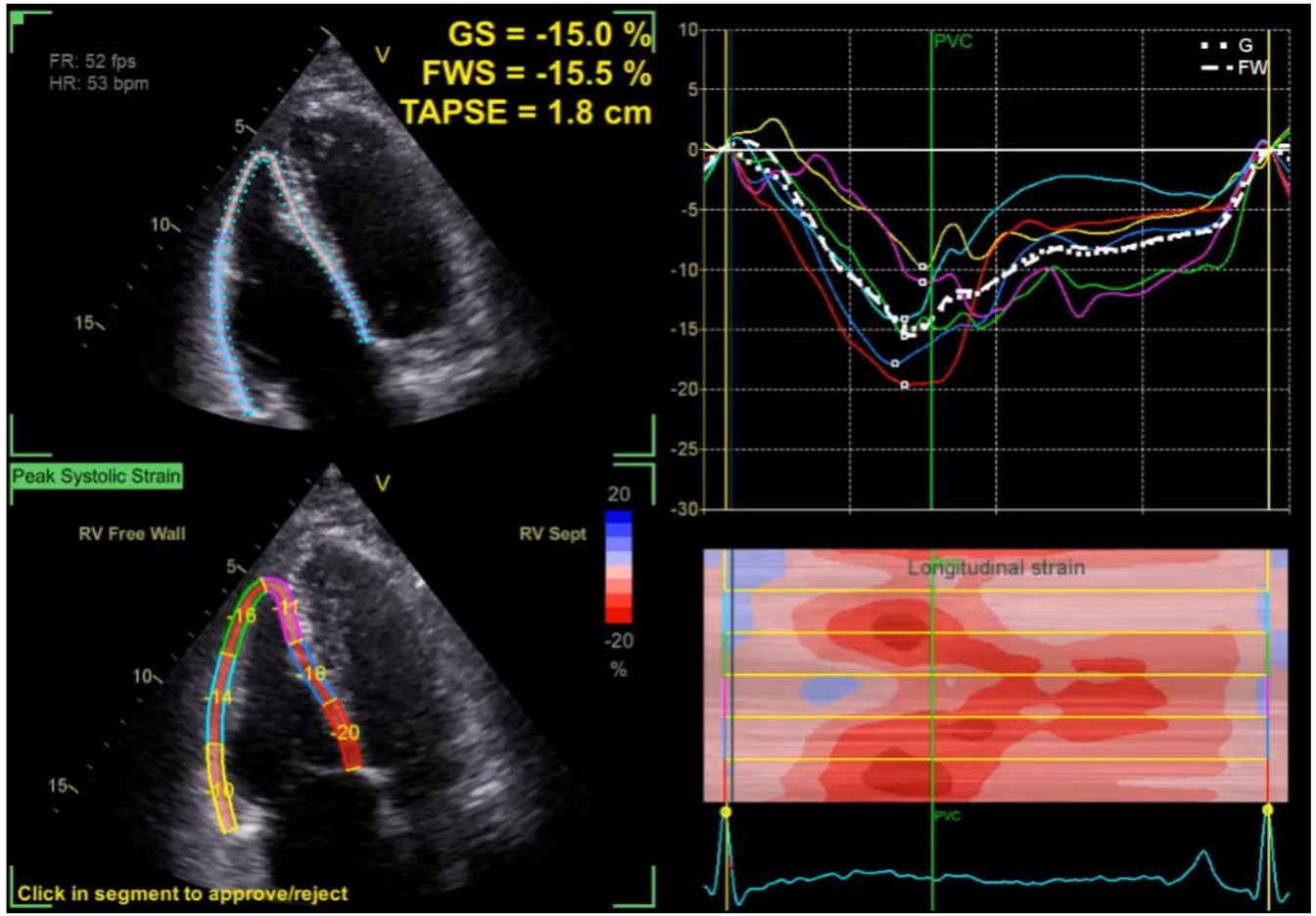


Z archívu I . interní kardiologické kliniky FNOL, dr. Koubek

Dilatace pravé komory včetně RVOT z PLAX a 4C

Echokardiografické vyšetření

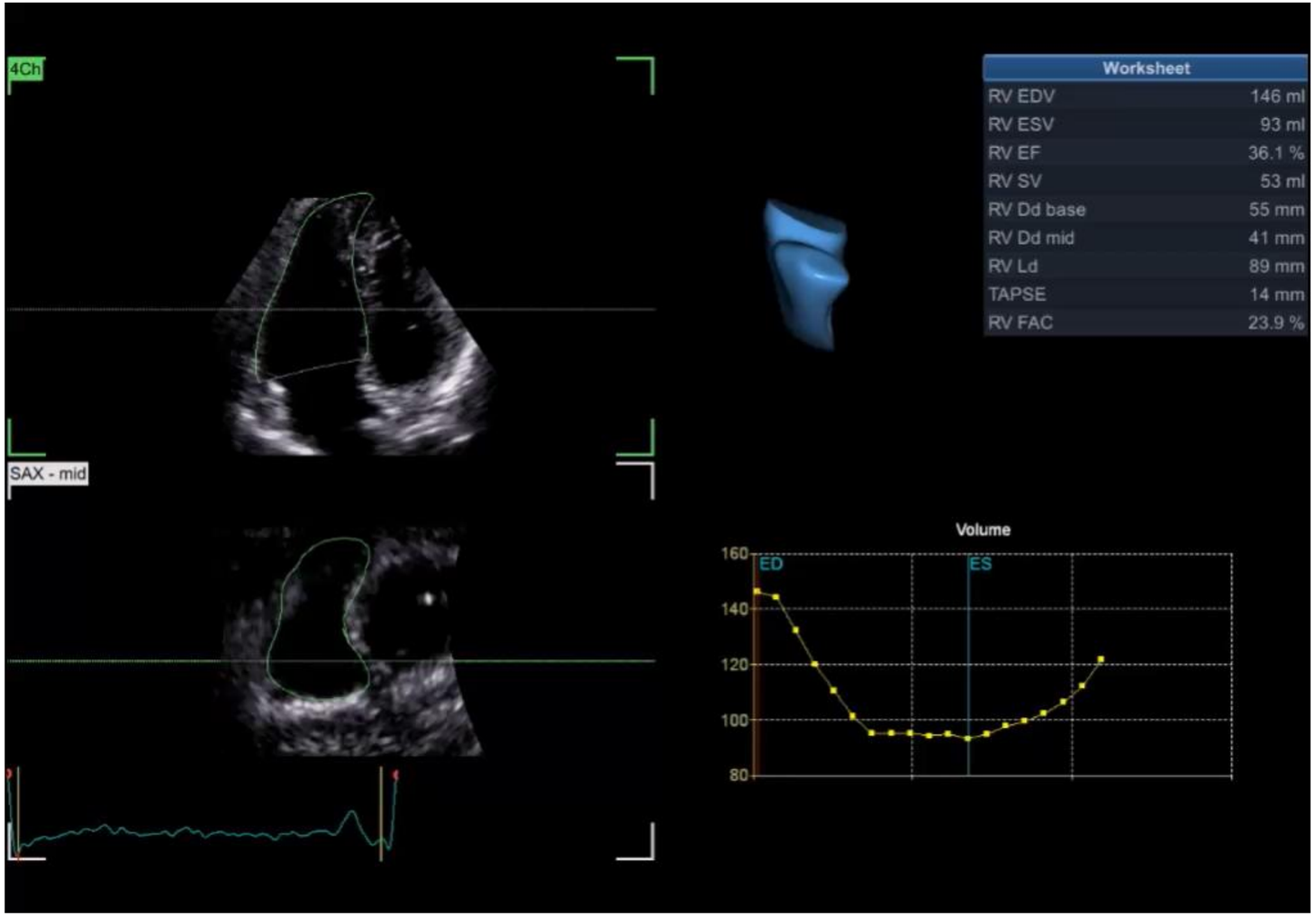
Dysfunkce pravé komory



Z archívu I . interní kardiologické kliniky FNOL, dr. Koubek

TAPSE dle specle tracking 18mm, FWS – 15,5%

Dysfunkce pravé komory



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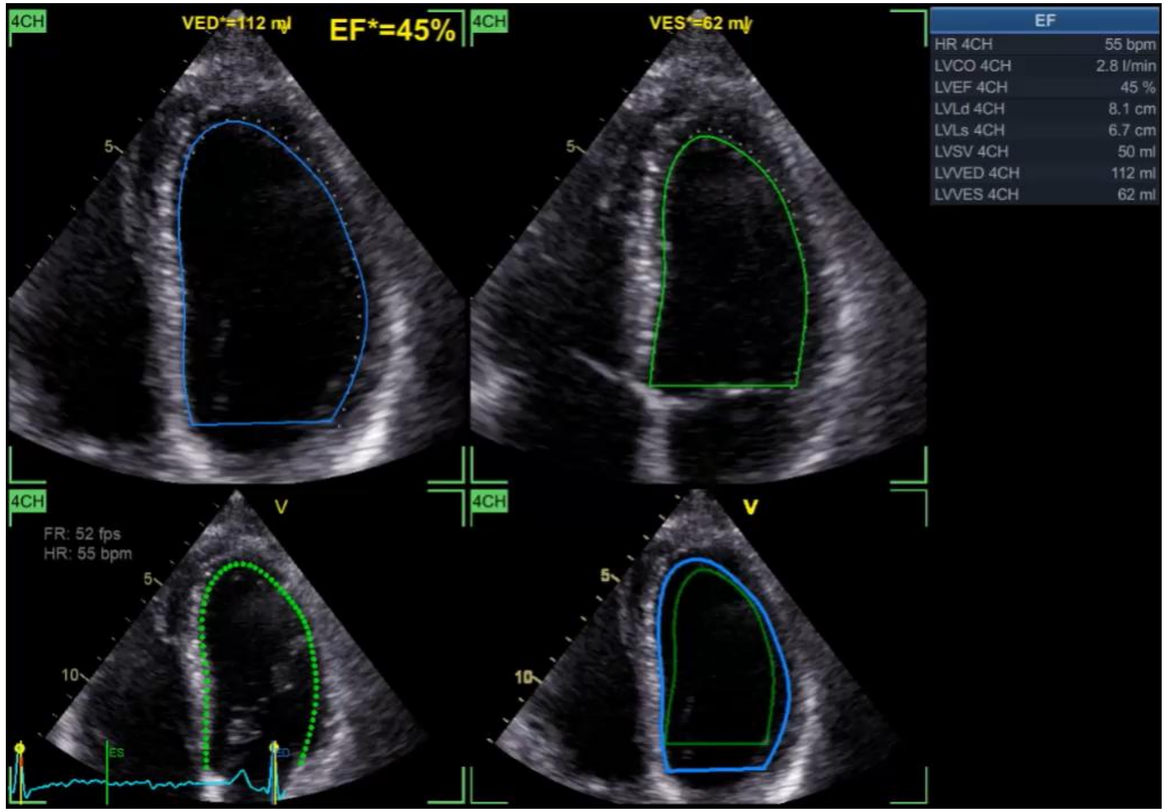
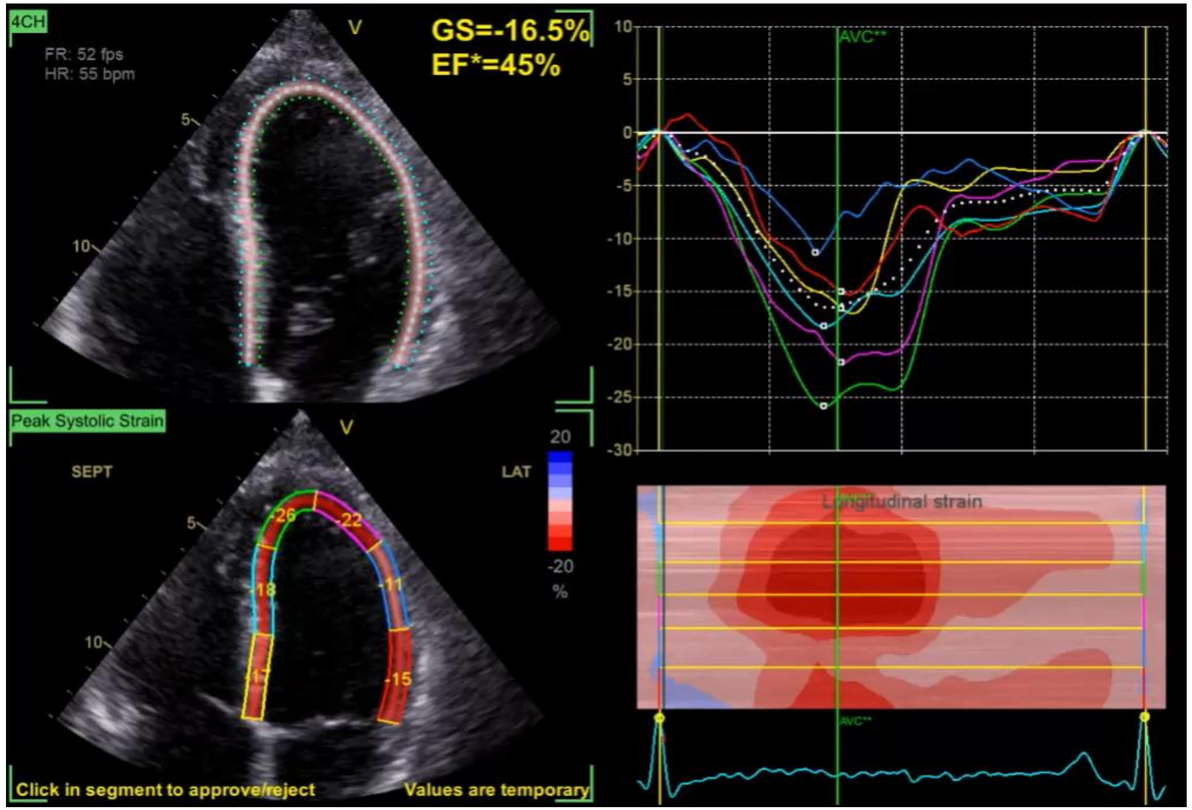
FAC dle volumetrie 23,9%, EF PK dle volumetrie 36%

Echokardiografické vyšetření

Dysfunkce levé komory





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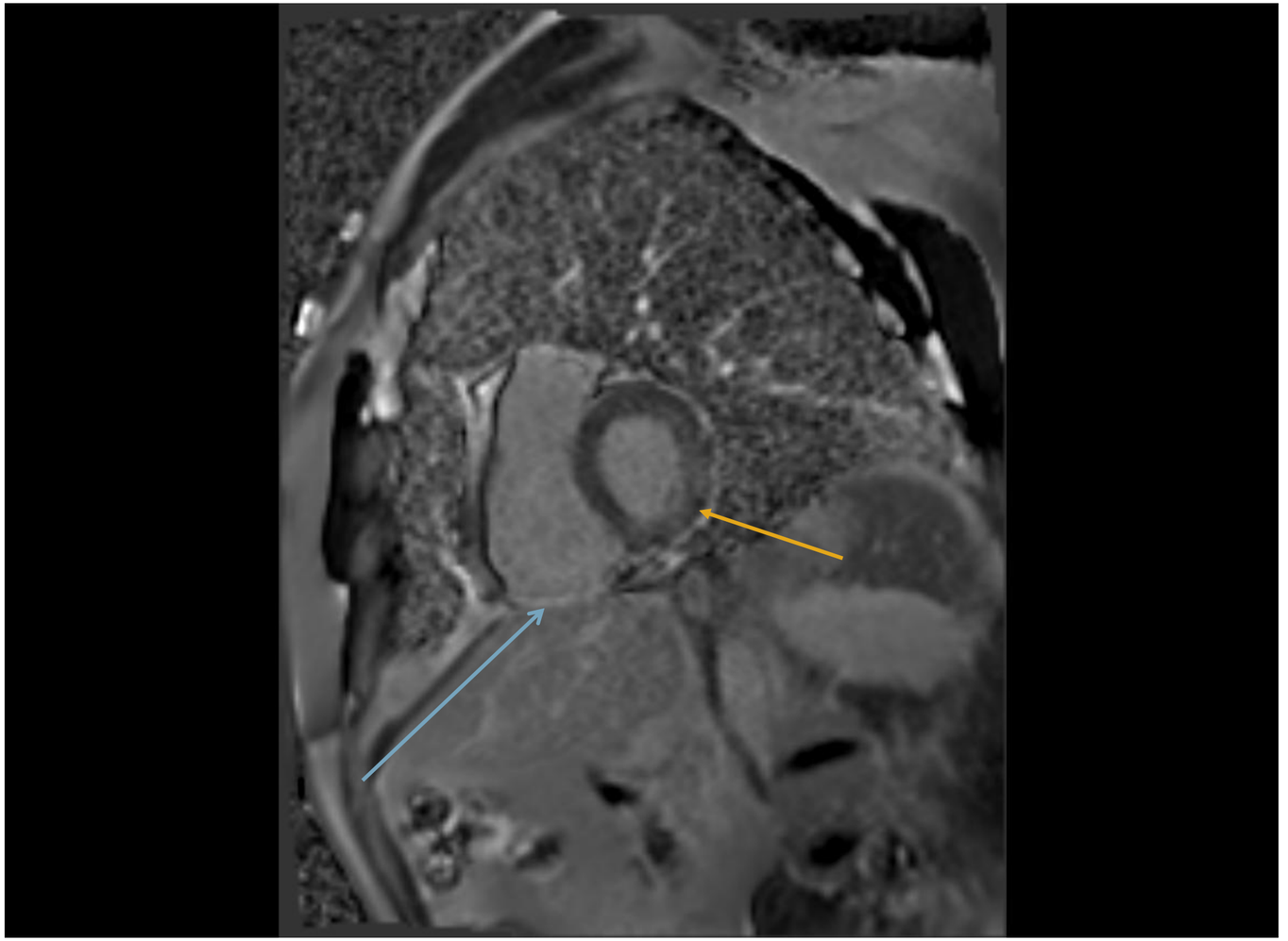
Mírná globální systolická dysfunkce a difůzní hypokinéza levé komory, speckle tracking EF LK 45%, GLS -16,5 %

MRI srdce



Z archívu Radiologické kliniky FNOL

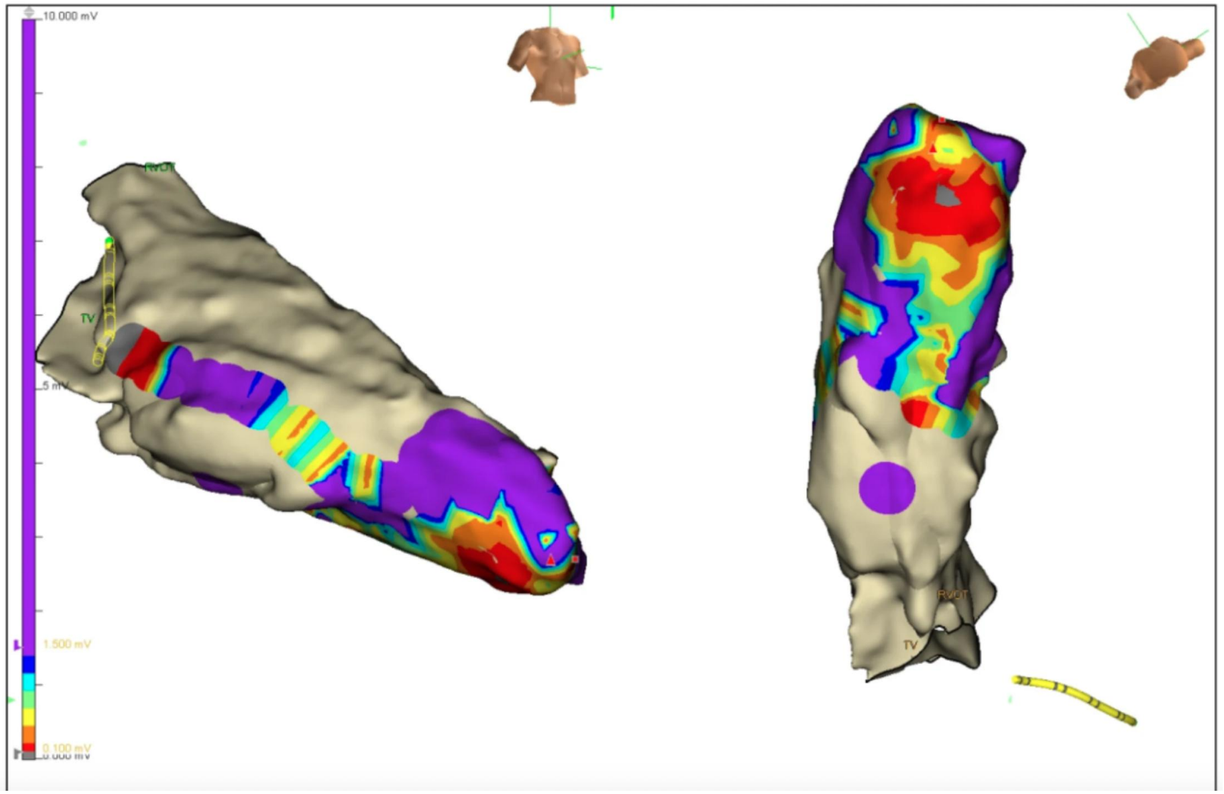
Systolická dysfunkce PK i LK, pozdní sycení inferolaterální stěny LK, pozdní sycení epikardiálně na spodní stěně PK

Další management

- Definitivně potrvzena diagnóza arytmogenní kardiomyopatie (AVC)
- Implantace jednodutinového kardioverter - defibrilátoru ze sekundárně preventivní indikace

Další management

- S odstupem provedena katetrizační ablace
- Během výkonu provedena programovaná stimulace komor s opakovaně vyvolatelnými setrvalými monomorfními tachykardiemi (s poklesem STK k 60mmHg)
- Následně podrobný mapping pravé komory a identifikovány nízkovoltážní zóny v oblasti výtokového traktu pod Pu chlopní, vtokově na spodní stěně, a u hrotu na spodní stěně pravé komory – zde RF aplikace
- Následně již bez vyvolatelných setrvalých komorových tachykardii, maximálně vyvolatelné 3 otočky KT na agresivním protokolu PSK



https://link.springer.com/article/10.1186/s13019-024-03094-y

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	

https://pmc.ncbi.nlm.nih.gov/articles/PMC8746198/

Padauova diagnostická kritéria

Padauova kritéria (2020), rozšířená ITF kritéria (2010), pracující s termínem Arytmogenní kardiomyopatie (ACM) zahrnující nejen klasickou pravostranně dominantní variantu (ARVC), ale i levostrannou (ALVC) či biventrikulární variantu ACM.

Morpho-functional ventricular abnormalities

- ECHO: Dilatace pravé komory včetně RVOT, systolická dysfunkce, porucha kinetiky volné stěny a baze pravé komory – **MAJOR CRITERIA**
- MRI: systolická dysfunkce PK, pozdní sycení epikardiálně na spodní stěně PK

Ventricular arythmias

- Setrvalá komorová tachykardie tvaru LBBB – **MAJOR CRITERIA**

Depolarization/Repolarization abnormalities

- Inverze vlny T ve svodech V1-V3 – **MAJOR CRITERIA**

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 echocardiographic 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
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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)
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https://pmc.ncbi.nlm.nih.gov/articles/PMC8746198/

Biventrikulární fenotyp ACM?

Morpho-functional ventricular abnormalities

ECHO: Mírná globální systolická dysfunkce a difůzní hypokineza, specle traking EF LK 45% – MINOR CRITERIA
MRI: systolická dysfunkce, pozdní sycení inferolaterální stěny LK

- **MINOR CRITERIA**

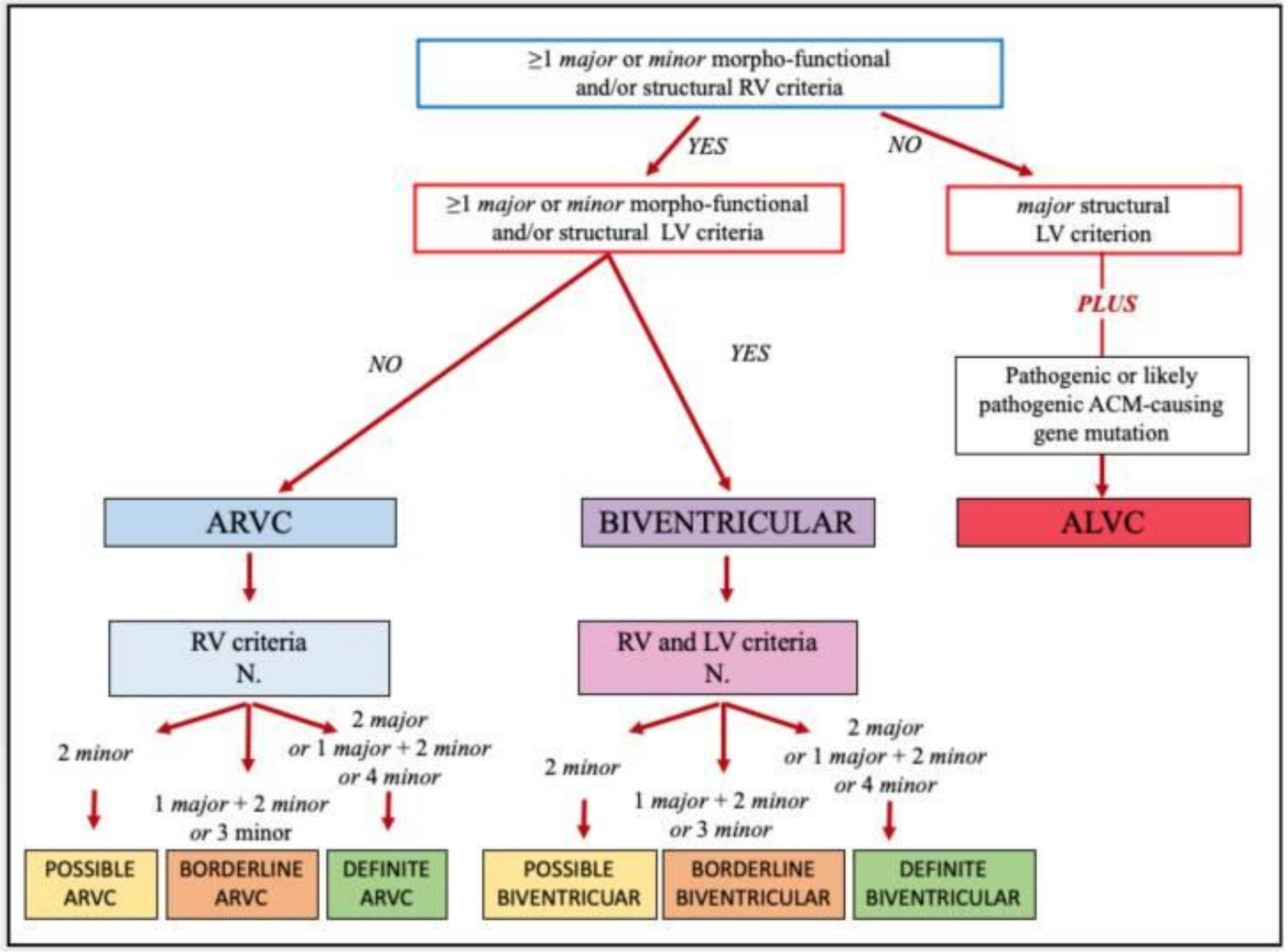
Repolarization abnormalities

Inverze vln T v prekordiálních svodech V4-V6 - **MINOR CRITERIA**

Depolarization abnormalities

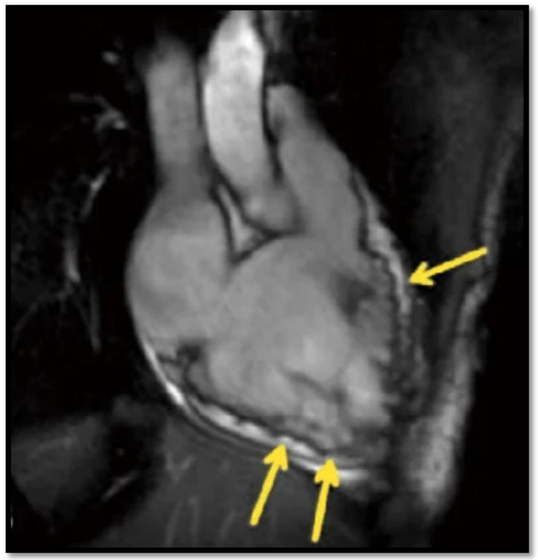
Nízká voltáž QRS (pod 0,5mV peak to peak) v končetinových svodech - **MINOR CRITERIA**

Postup diferenciace fenotypu ACM



https://pmc.ncbi.nlm.nih.gov/articles/PMC8746198/

Fenotypy Arytmogenní kardiomyopatie

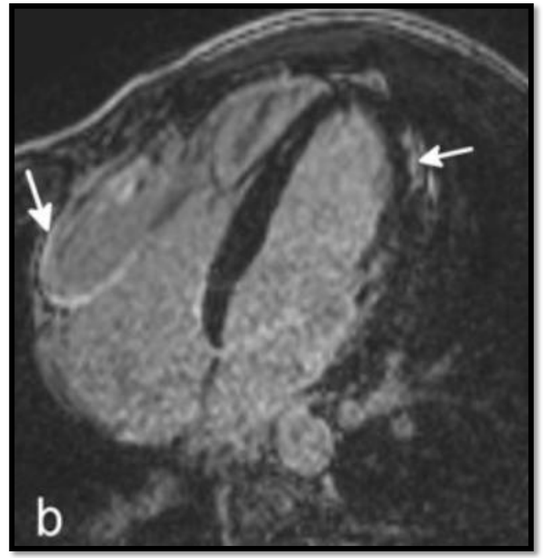


https://www.researchgate.net/figure/Cardiac-MRI-of-a-patient-with-ARVC-Bright-blood-images-in-the-right-ventricular-outflow_fig7_275053412

Dominant right variant – klasická ARVC

https://heart.bmj.com/content/108/9/733

Dominant left variant - ALVC



https://www.researchgate.net/figure/LGE-images-of-an-athlete-with-biventricular-ARVC_fig1_328641643

Biventricular disease variant

ESC guidelines 2023

- ARVC je charakterizovaná atrofií myokardu a náhradou myokardu tukovou a fibrózní tkání vedoucí k dilataci a poruše kinetiky pravé komory
- ESC guidelines používají pojem ARVC, ale uvádějí Padauova kritéria
- Epsilon vlna bývá často naddiagnostikována, vyskytuje se většinou u velkého strukturálního postižení

Recommendation Table 28 — Recommendations for the antiarrhythmic management of patients with arrhythmogenic right ventricular cardiomyopathy

Recommendations	Class[a]	Level[b]
Beta-blocker therapy is recommended in ARVC patients with VE, NSVT, and VT.[920–922]	I	C
Amiodarone should be considered when regular beta-blocker therapy fails to control arrhythmia-related symptoms in patients with ARVC.[921,922]	IIa	C
Flecainide in addition to beta-blockers should be considered when single agent treatment has failed to control arrhythmia-related symptoms in patients with ARVC.[923,924]	IIa	C
Catheter ablation with availability for epicardial approach guided by 3D electroanatomical mapping of VT should be considered in ARVC patients with incessant VT or frequent appropriate ICD interventions for VT despite pharmacological therapy with beta-blockers.[925,929–934]	IIa	C

3D, three-dimensional; ARVC, arrhythmogenic right ventricular cardiomyopathy; ICD, implantable cardioverter defibrillator; NSVT, non-sustained ventricular tachycardia; VE, ventricular ectopic beats; VT, ventricular tachycardia.
[a]Class of recommendation.
[b]Level of evidence.

https://www.escardio.org/guidelines/clinical-practice-guidelines/all-esc-practice-guidelines/cardiomyopathy-guidelines/

ESC guidelines 2023

Doporučení stran prevence náhlé srdeční smrti

Recommendation Table 29 — Recommendations for sudden cardiac death prevention in patients with arrhythmogenic right 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 ARVC who have survived a cardiac arrest or have recovered from a ventricular arrhythmia causing haemodynamic instability.[939,943,944,948,949]	I	A
An ICD should be considered in ARVC patients who have suffered a haemodynamically tolerated VT.[522,939,943–945,948–950]	IIa	B
Primary prevention		
High-risk features[c] should be considered to aid individualized decision-making for ICD implantation in patients with ARVC.[538,939]	IIa	B
The updated 2019 ARVC risk calculator should be considered to aid individualized decision-making for ICD implantation in patients with ARVC.[d,524,526,536–539]	IIa	B

ARVC, arrhythmogenic right ventricular cardiomyopathy; ICD, implantable cardioverter defibrillator; LVEF, left ventricular ejection fraction; NSVT, non-sustained ventricular tachycardia; PES, programmed electrical stimulation; RVEF, right ventricular ejection fraction; SMVT, sustained monomorphic ventricular tachycardia; VT, ventricular tachycardia.

[a]Class of recommendation.

[b]Level of evidence.

[c]High-risk features: arrhythmic syncope, NSVT, RVEF <40%, LVEF <45%, SMVT at PES.

[d]See text for discussion of gene-specific differences in the performance of the updated 2019 ARVC risk calculator.

https://www.escardio.org/guidelines/clinical-practice-guidelines/all-esc-practice-guidelines/cardiomyopathy-guidelines/

Závěr

- Arytmogenní kardiomyopatie (dle Padauových kritérii) s fenotypy ARVC, ALVC a s biventrikulárním postižením
- Kontrola ICD s odstupem 3 měsíců od primoimplantace - bez záchytu recidivy komorových arytmií
- Katetrizační ablace může vést ke snížení rizika recidiv komorových tachykardií
- Doporučeno vyšetření prvostupňových příbuzných, provedeno genetické vyšetření – t.č. bez výsledků

DĚKUJI ZA POZORNOST

FAKULTNÍ NEMOCNICE OLOMOUC