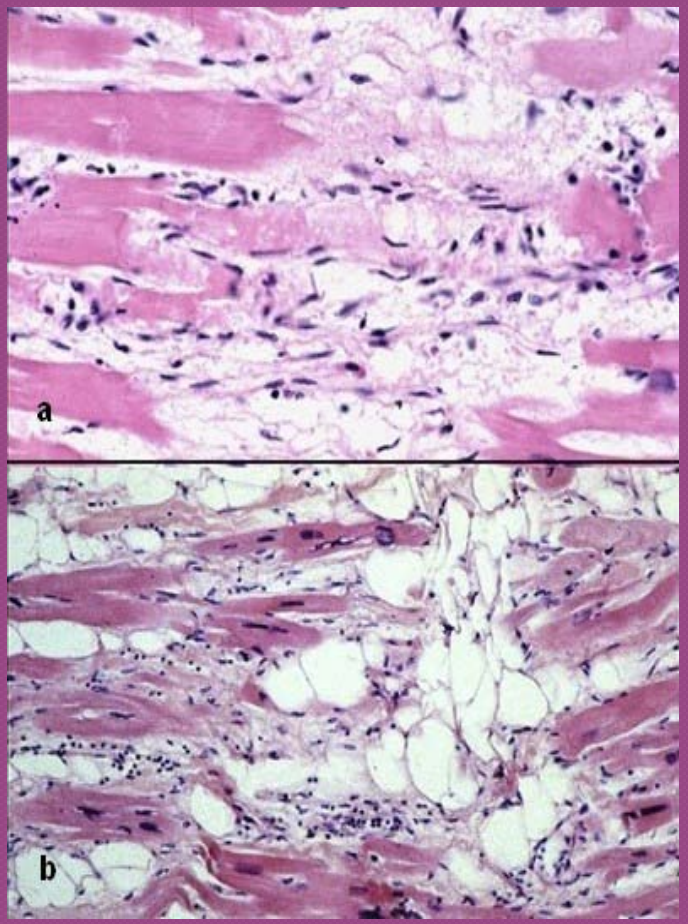
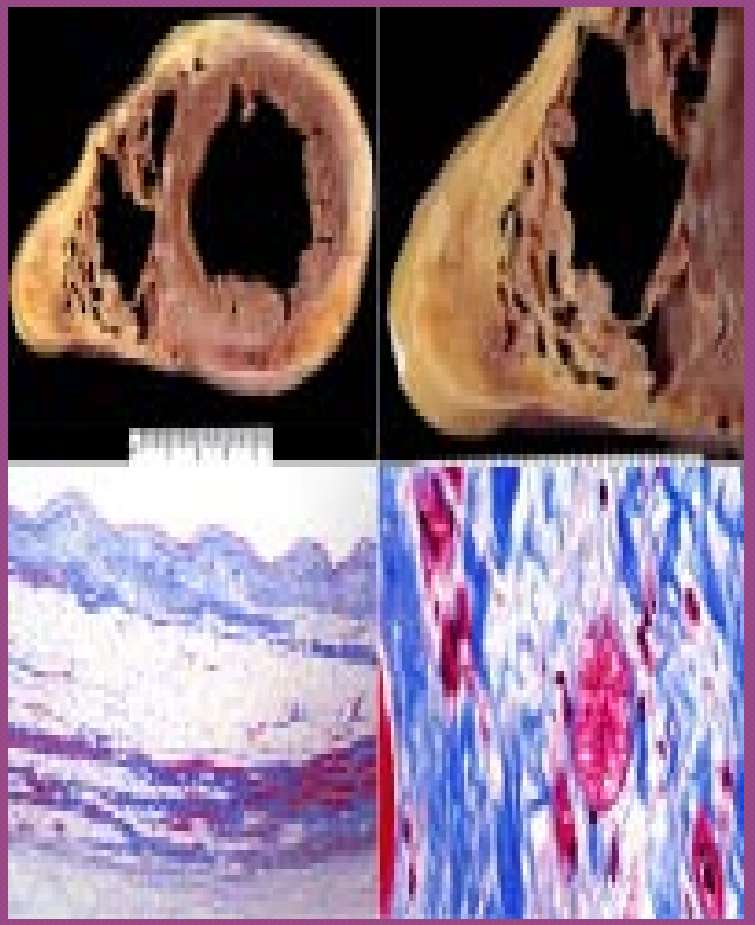
Arrhytmogenic right ventricular dysplasia (ARVD)/Arrhytmogenic right ventricular cardiomyopathy (ARVC)

Retka nasledna bolest na miokardot(1:5000) na desen ventrikul,koja se karakterizira so progresivno gubenje na myocitite so apoptoza I infiltracija so masno ili svrzno tkivo I so segmentalni(anevrizmatski prosiruvanja) na desen ventrikul

Dr.Natasa Taneska


a
b



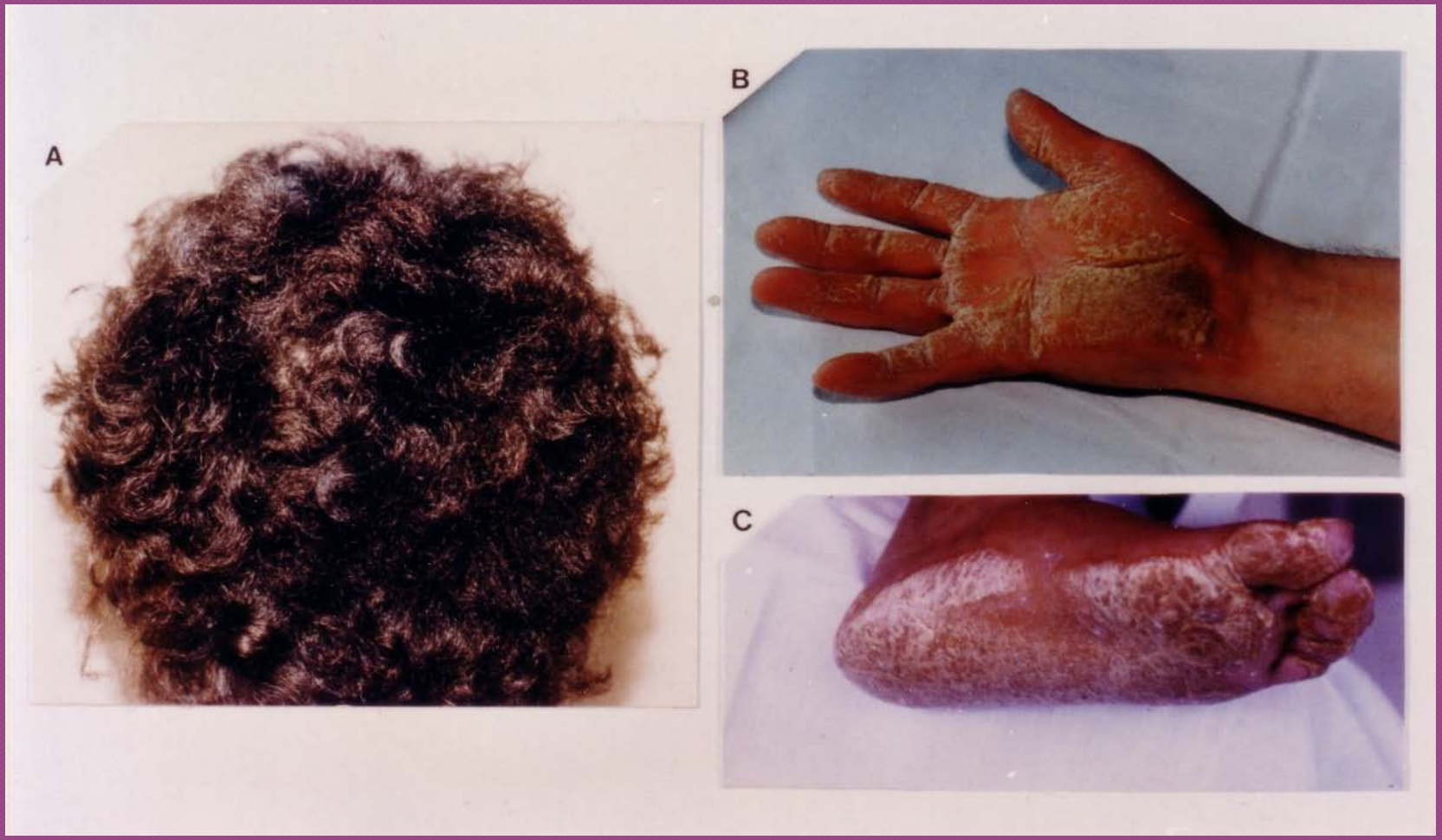
- ARVD e tip na vrodena neishemicna cardiomyopatija,kade sto najcesto e zafaten desniot ventrikul.
- Se karakterizira so hipokineticni arei na desniot ventrikul,so fibrozno-masna degeneracija,koi se asocirani so teski ventrikularni aritmii.

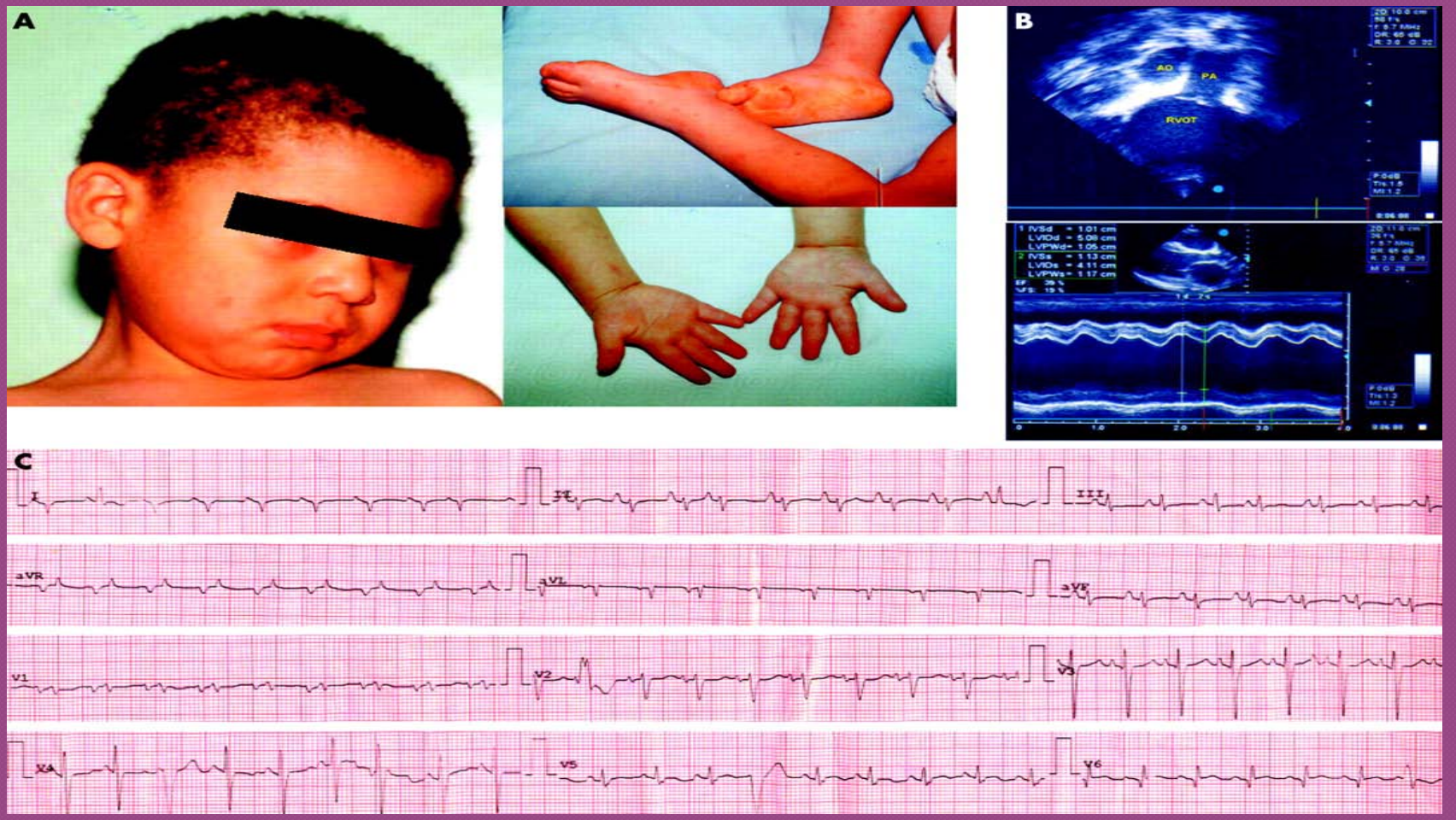
- ARVD e predizvikana od genetski defekti na kletkite na myocardium poznati kako (desmosomes)koi sluzat za povrzuvanje na kletkite megu sebe I koi mozat da pretrpat teski mutacii na myocardot.

Kaj najgolem broj od zabolenite se nasleduva avtosomnodominantno. Otkrieni se poveke mutacii na geni koi go kontroliraat kardijalniot receptor (RyR2),dezmoplakin,plakofilin-2.

Type	OMIM	Gene	Locus
ARVD1lpl	107970	*TGFB3*	14q23-q24
ARVD2	600996	*RYR2*	1q42-q43
ARVD3	602086	?	14q12-q22
ARVD4	602087	?	2q32.1-q32.3
ARVD5	604400	*TMEM43*	3p23
ARVD6	604401	?	10p14-p12
ARVD7	609160	*DES*	10q22.3
ARVD8	607450	*DSP*	6p24
ARVD9	609040	*PKP2*	12p11
ARVD10	610193	*DSG2*	18q12.1-q12
ARVD11	610476	*DSC2*	18q12.1
ARVD12	611528	*JUP*	17q21

- Opisani se I dva rececivni oblici na bolesta,povrzani so difuzna-palmoplantarna keratodermija I wolly hair(Sy Naxos).
- Iako e cesto povrzana so myocarditis(enterovirusi,adenovirusi),se smeta deka nema vospalitelna etiologija.


A
B
C


A
B
AO
PA
RVOT
C
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6

Klinicka slika:

Dyspnea

- Atypical chest pain(27%)
- Sudden cardiac death (SCD)(10-26%)
- Syncope (26-32%)
- Palpitation(27-67%)

- Kaj 80 % od slucaite ARVD se prezentira so sinkopa ili SCD (nenadejna srceva smrt).
- Cesto se prisutni palpitacii ili simptomi povrzani so RVOT (right ventricular outflow tract),tachycardia(monomorfna ventrikularna tachycardia).
- Simptomite se najcesto prisutni kaj sportisti,I se javuvaat kaj adolescenti.

Patogeneza

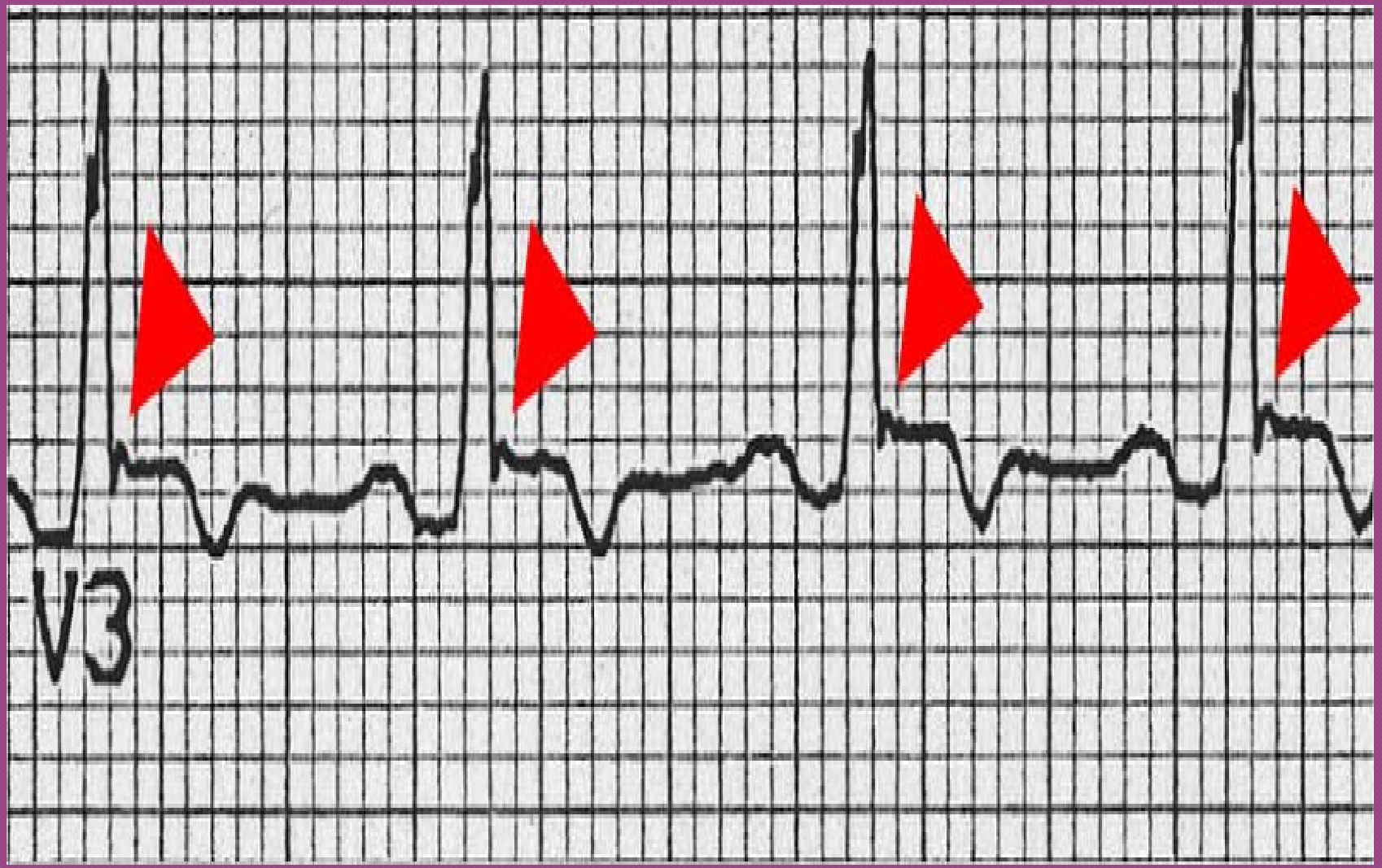
- Patogenezata kaj ARVD e nepoznata.
- Patoloskiot proces najcesto gi zafaka bazalniot,apikalniot I (RVOT)izlezniot del od desniot ventrikul (triagolna displazija),kade sto nastanuva apoptoza na myocitite,a myokardot e potpolno zamenet so masno ili svrzno tkivo I se dobivaat segmentalni(anevrizmatski prosiruvanja) na desen ventrikul.

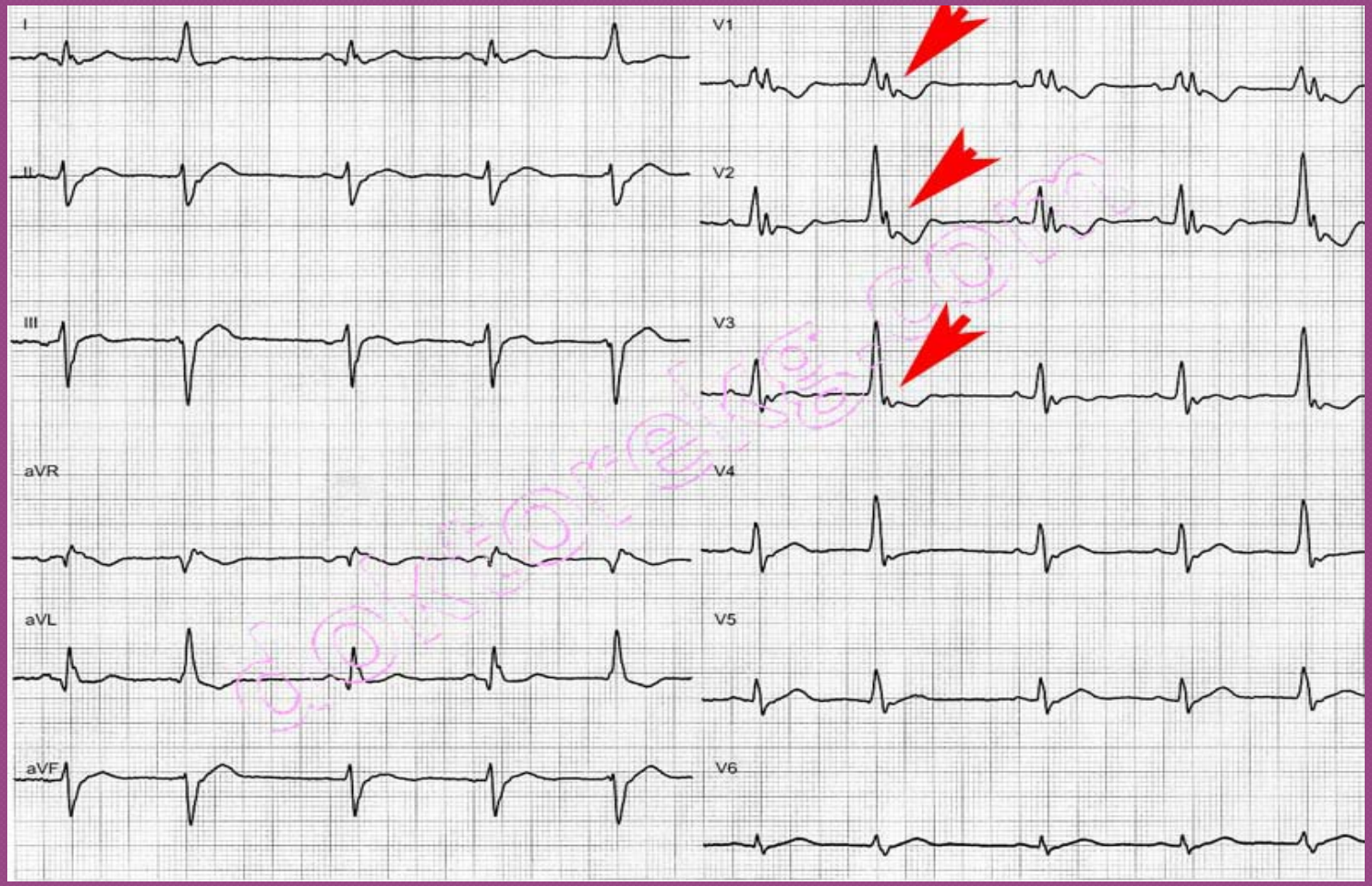
Dijagnoza:

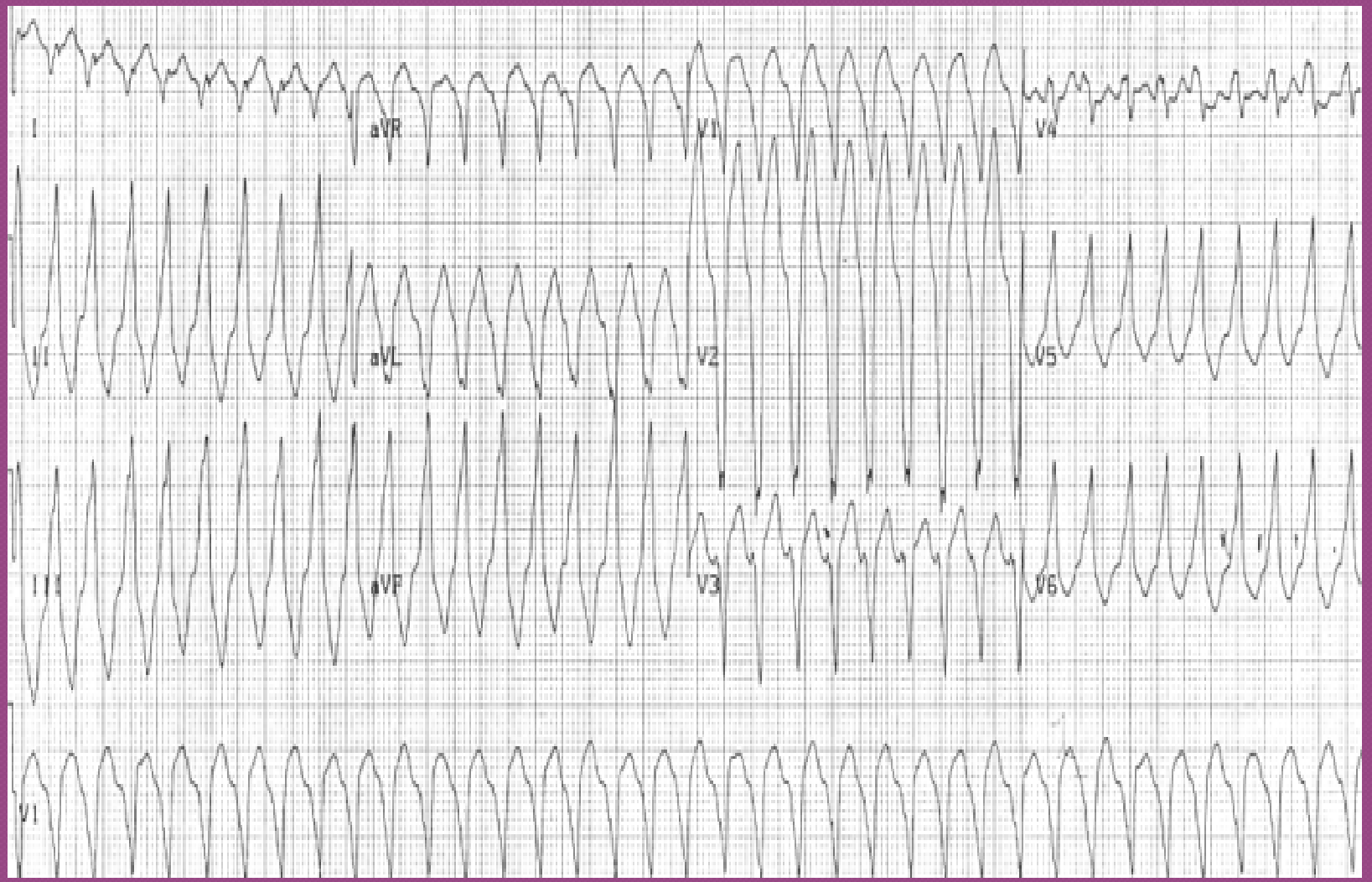
- EKG kriteriumi:

- EKG ima morfologija na RBBB, negativni T branovi
- Od V1-V4 I docen ventrikularen (epsilon)potencijal,I cesti VES.

- Ventrikularnite aritmii kaj ARVD mozat da bidat:
- PVCs(premature ventricular complexes)
- VT (ventricular tachycardia)
- VF (ventricular fibrilation)

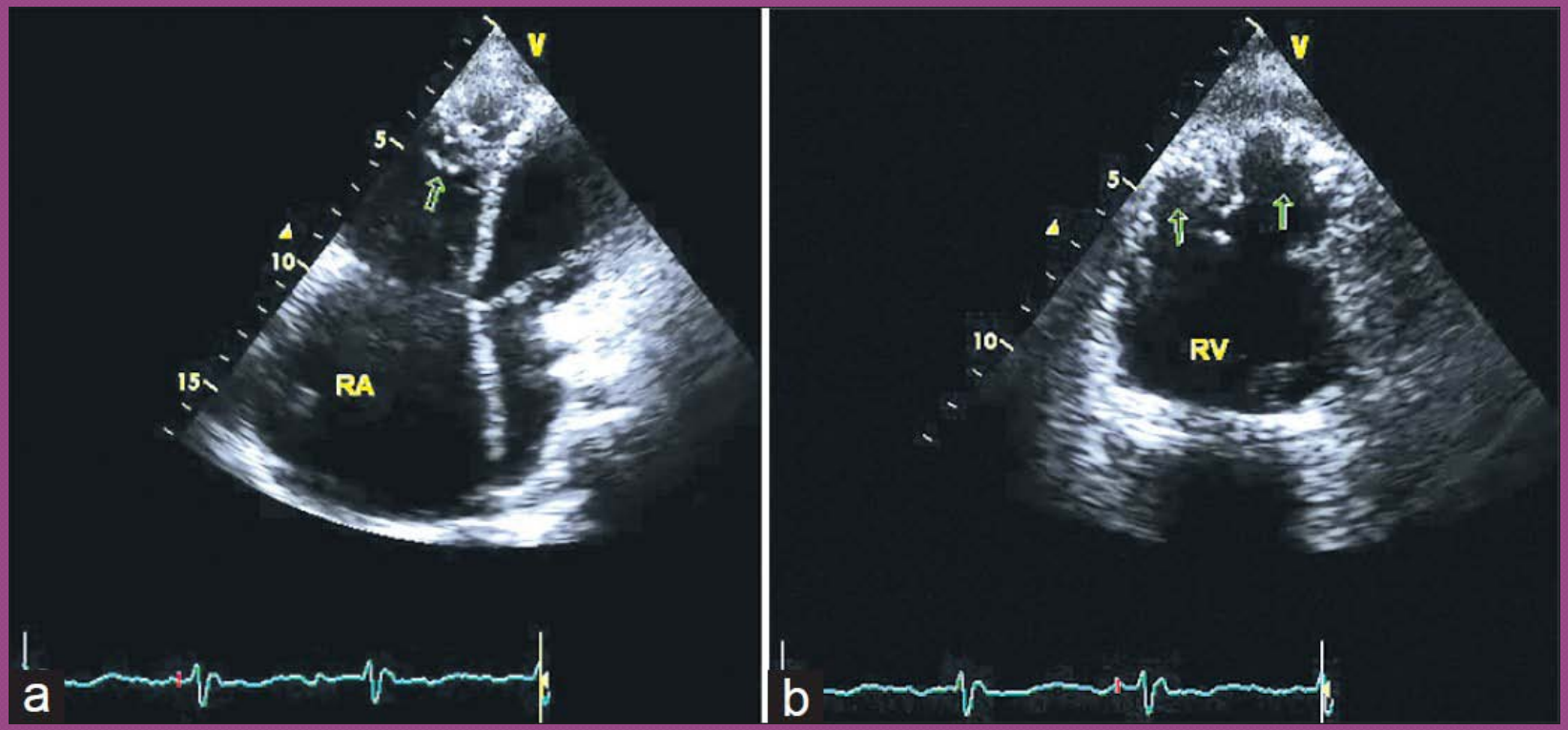

V3

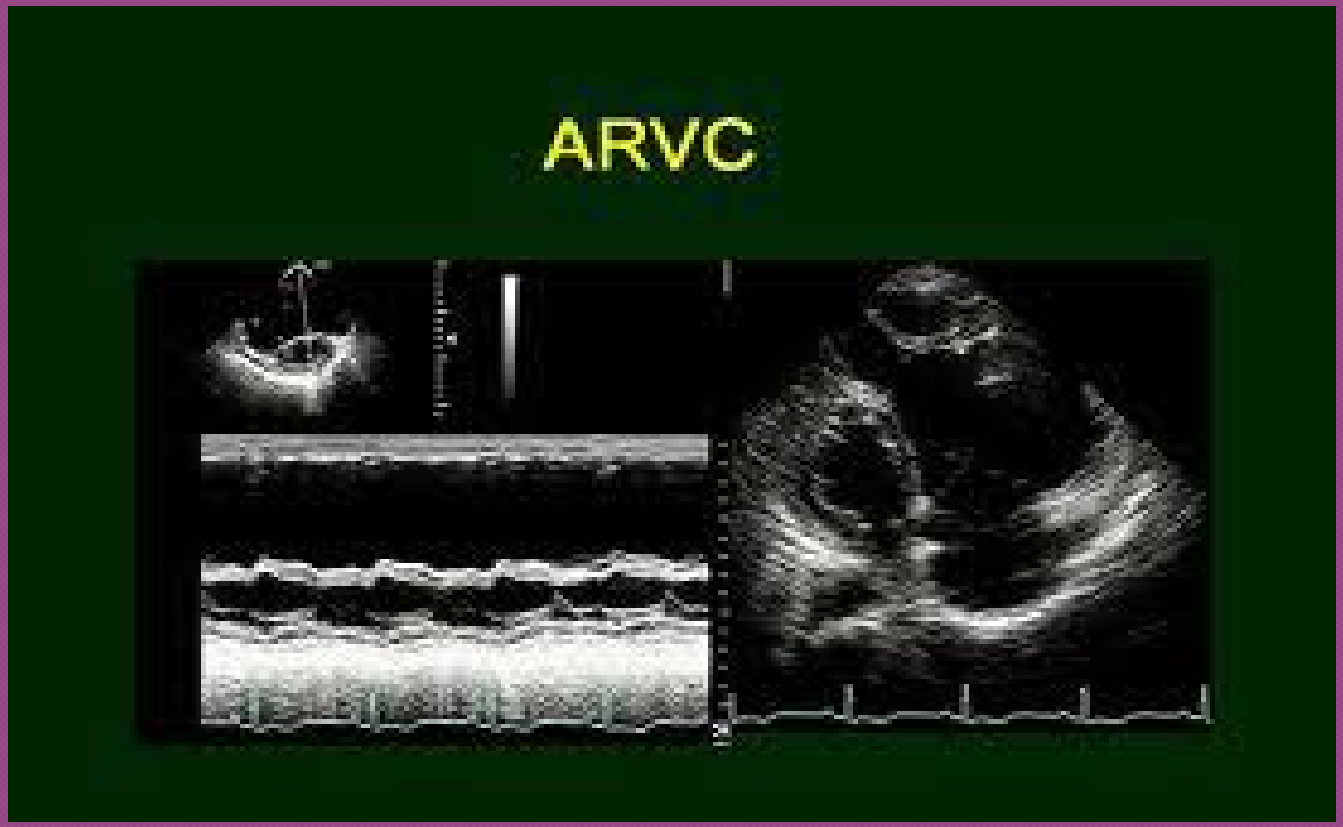

I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6


I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
V1

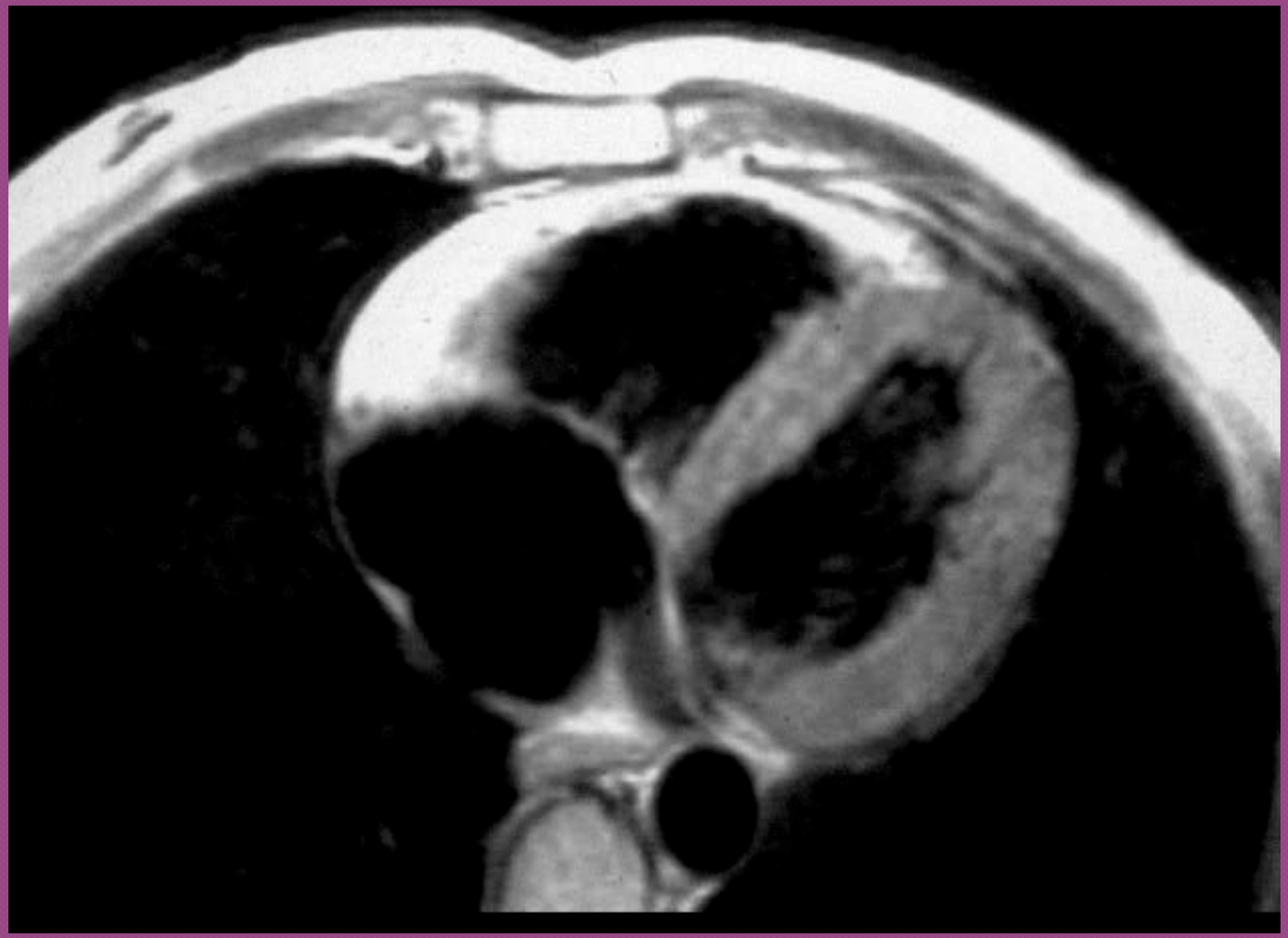
- **Echocardiografski kriteriumi:**

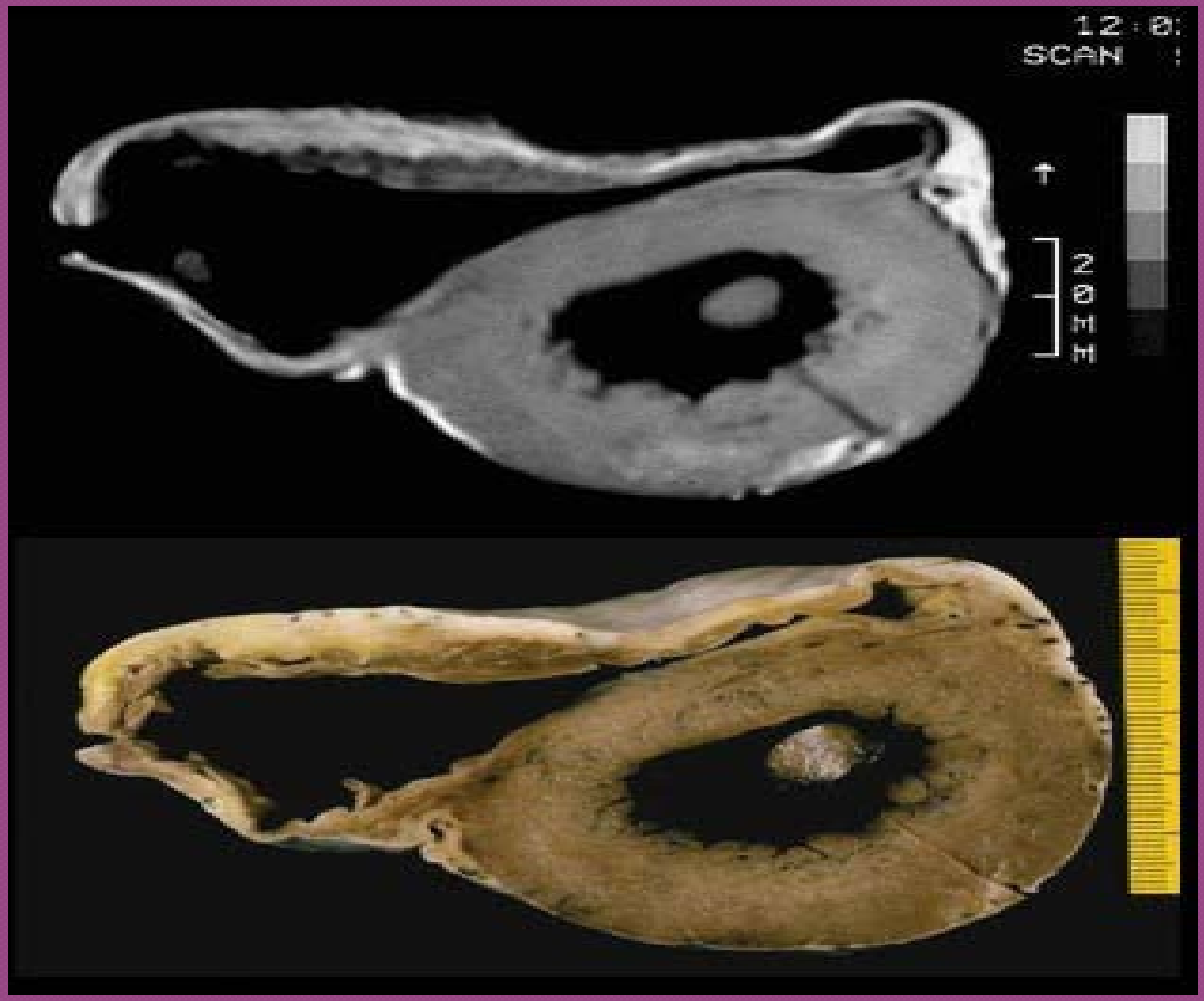
- Normalna dimenzija i funkcija na LV
- Zgolemeni dimenzii na RV
- (RV outflow tract >30 mm vo dijastola)
- Namalena funkcija na RV
- RV (zgolemen,hypokineticen desen ventrikul, so trabekularni i anevrizmatski lokalizirani prosiruvanja), dilatacija na tricuspid valve anulus I tricuspid regurgitation.


V
5
10
15
RA
a
V
5
10
RV
b


ARVC

Cardijalna MRI,
Right ventricular angiography,
Right ventricular biopsy,
Autopsy




12:0:
SCAN :
2 mm

Diff.Dg

- Differential diagnosis for ARVD vklucuva:
- Congenital heart disease
  - Repaired tetralogy of Fallot
  - Ebstein's anomaly
  - Uhl's anomaly
  - Atrial septal defect
  - Partial anomalous venous return
- Acquired heart disease
  - Tricuspid valve disease
  - Pulmonary hypertension
  - Right ventricular infarction
  - Bundle-branch re-entrant tachycardia
- Miscellaneous
  - Pre-excited AV re-entry tachycardia
  - Idiopathic RVOT tachycardia
  - Sarcoidosis

Lekuvanje

- Celta na lekuvanjeto e da se namali incidencata na SCD.
- Lica so visok rizik za SCD:
- Mladi lica
- Sportisti
- Familijarna predispozicija
- Lica so namalena EF na desen ventrikul
- Syncopa
- Cesti epizodi na ventrikularni aritmii

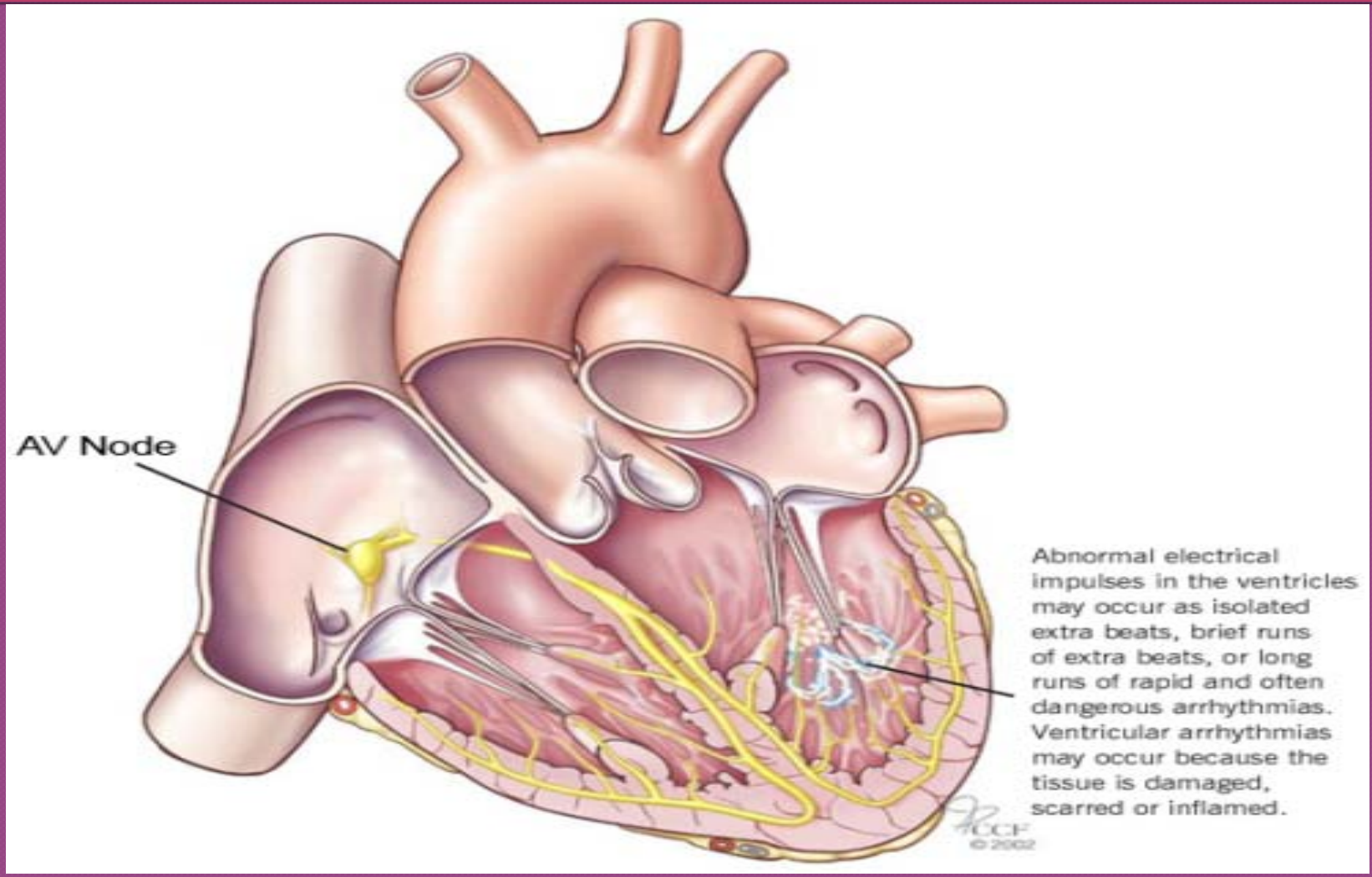
- Lekuvanjeto vklucuva:

  Farmakoloski,
- Hirurski tretman,
- Catheter ablacija,
- Vgraduvanje na implantable cardioverter defibrillator (ICD)

- Farmakoloski tretman:

- Beta blocker, Sotalol (class III antiarrhythmic agent),amiodarone.

- Anticoagulation(warfarin),da se preveniraat trombi I pulmonalna embolija.


AV Node
Abnormal electrical impulses in the ventricles may occur as isolated extra beats, brief runs of extra beats, or long runs of rapid and often dangerous arrhythmias. Ventricular arrhythmias may occur because the tissue is damaged, scarred or inflamed.
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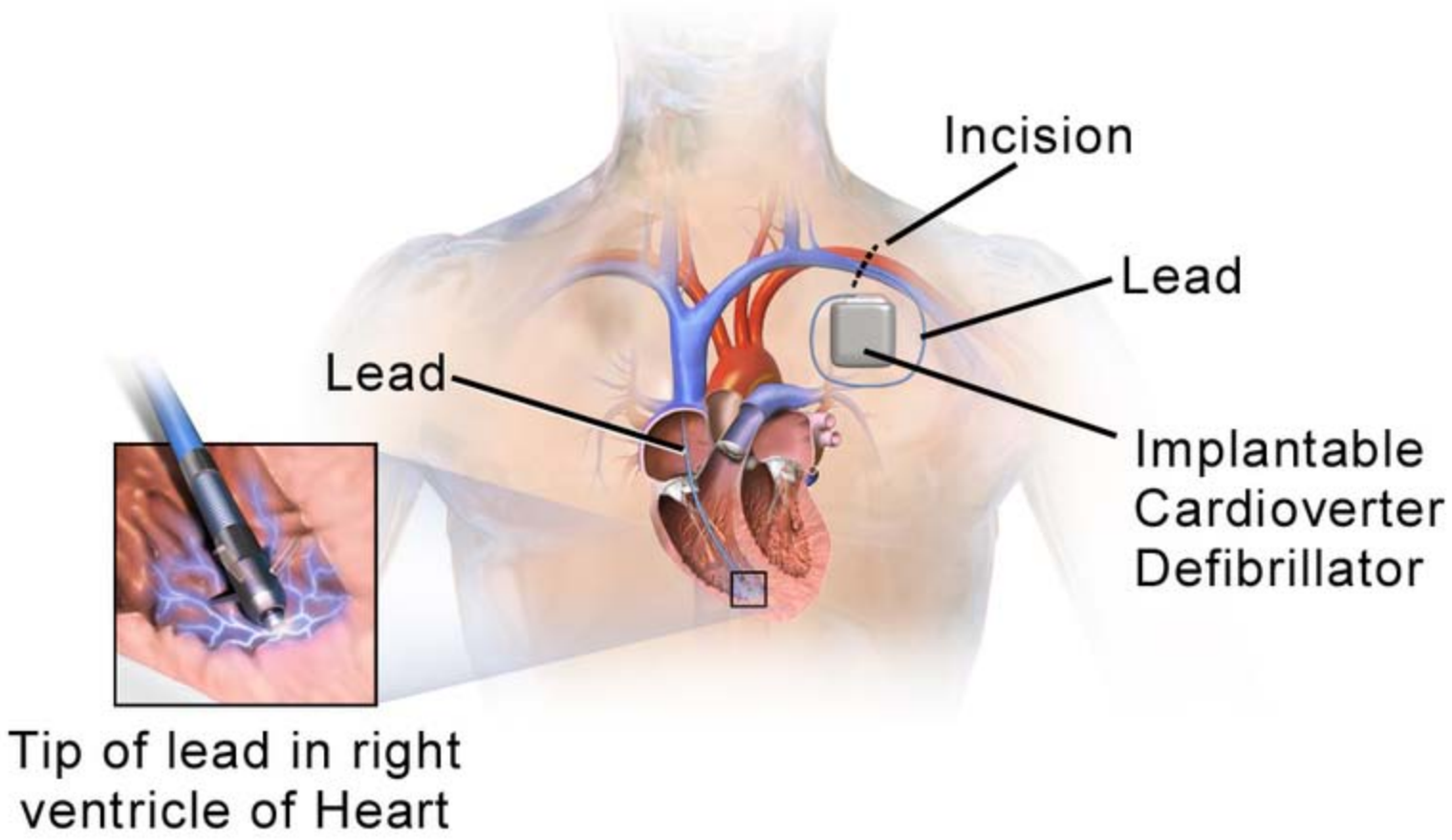
- Catheter ablation:

- se pravi kaj teski ventrikularni tachycardii.
- Indikacija za catheter ablacija vklucuva:
- Drug-refractory VT I
- Frequent recurrence of VT posle ICD
- Placement

Implantable Cardioverter Defibrillator



- Indikacii za implantable cardioverter-defibrillator se:

- Cardiac arrest posle VT iliVF
- Simptomatska VT koja ne reagira na medikamentozna terapija
- Severe RV involvement
- SCD kaj clenovi vo familijata
- ICD se aplicira preku venozen pat do desen ventrikul I najcesto nesakana reakcija e pericardial tamponade.

- Cardiac transplant surgery
- Lica so bi-ventricular heart failure

- Family screening
- (EKG,Echocardiogram,Holter monitoring,exercise stress test,cardiac MRI)