

CARDIOMIOPATIA ARITMOGENA: WHAT'S NEW?

**Impatto diagnostico e prognostico
dell'elettrocardiogramma nella
valutazione del paziente con
cardiomiopatia aritmogena**

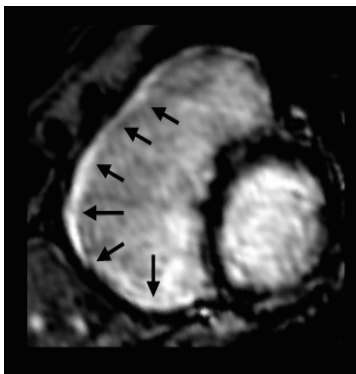
**Dott.ssa Cinzia Crescenzi
Policlinico Casilino (Roma)**



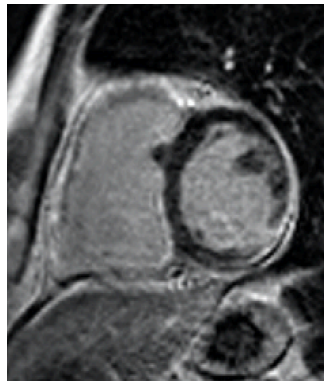
Arrhythmogenic cardiomyopathy (ACM) is a genetically determined heart disease characterized by progressive myocyte death and substitution by fibrofatty or fibrous tissue of the ventricular myocardium.

Electrical instability and **progressive HF** are the typical phenotypes of ACM

Right Dominant



Biventricular

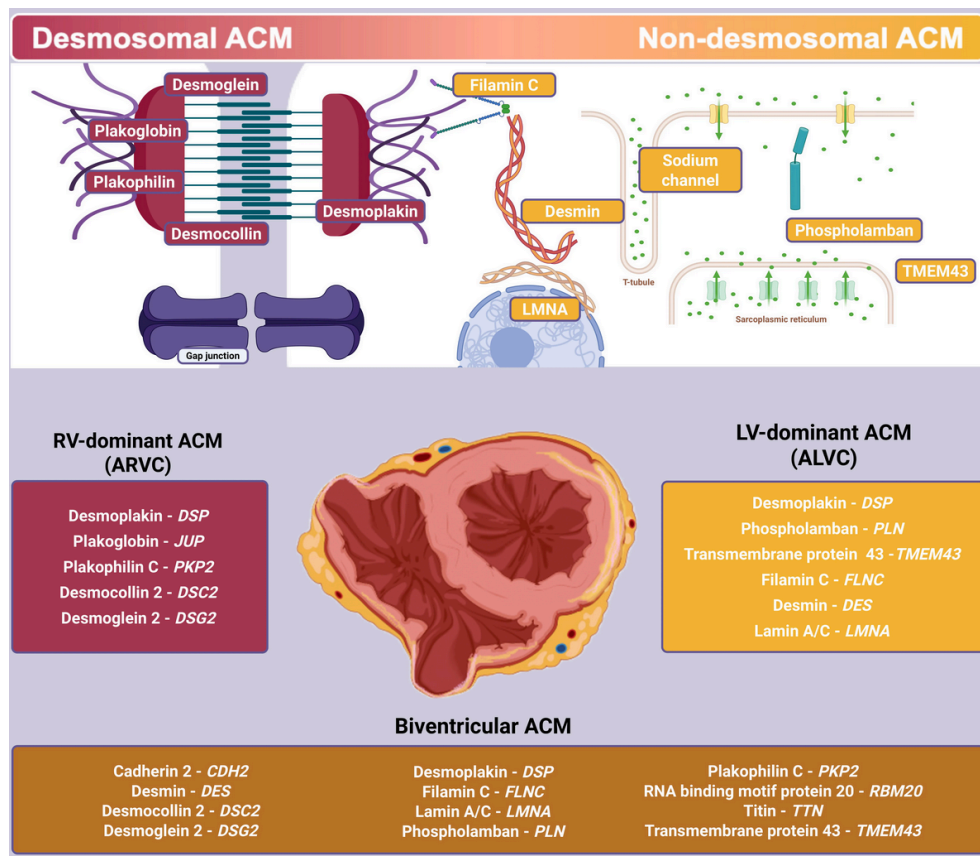


Left Dominant



ACM Phenotypes





2020

Diagnosis of arrhythmogenic cardiomyopathy: The Padua criteria

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Proposed diagnostic criteria for arrhythmogenic cardiomyopathy: European Task Force consensus report[☆]

2024

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2010

Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia

Proposed Modification of the Task Force Criteria

Frank I. Marcus, MD, Chair; William J. McKenna, MD, DSc, Co-Chair; Duane Sherrill, PhD; Cristina Basso, MD, PhD; Barbara Baucé, MD; David A. Bluemke, MD, PhD; Hugh Calkins, MD; Domenico Corrado, MD, PhD; Moniek G.P.J. Cox, MD; James P. Daubert, MD; Guy Fontaine, MD, PhD; Kathleen Gear, RN; Richard Hauer, NW, MD; Andrea Nava, MD; Michael H. Picard, MD; Nikos Protonotarios, MD; Jeffrey E. Saffitz, MD, PhD; Danita M. Yoerger Sanborn, MD, MMSc; Jonathan S. Steinberg, MD; Harikrishna Tandri, MD; Gaetano Thiene, MD; Jeffrey A. Towbin, MD; Adalena Tsatsopoulou, MD; Thomas Wichter, MD; Wojciech Zareba, MD, PhD



ESC

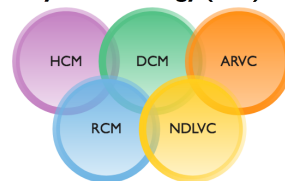
European Society
of Cardiology

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

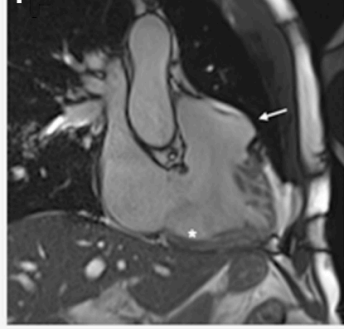
ESC GUIDELINES

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)



LA DIAGNOSI NEL TEMPO ...



CHARACTERIZATION OF PHENOTYPIC VARIANT

Corrado D et al. Int J Cardiol 2024 Jan 15;395:131447

≥1 major or minor morpho-functional or structural (tissue-characterization) **RV criteria**

YES

NO

≥1 major or minor morpho-functional or structural (tissue-characterization) **LV criteria**

major or minor structural (tissue-characterization) **LV criterion**

YES

NO

Consider the total number of criteria from **only** the right ventricle

Consider the total number of criteria from **both** the right and left ventricle

Consider the total number of criteria from **only** the left ventricle

1 major or 2 minor

1 major + 1 minor or 3 minor

2 major or 1 major + 2 minor or 4 minor

POSSIBLE ARVC

BORDERLINE ARVC

DEFINITE ARVC

2 minor

1 major + 1 minor or 3 minor

2 major or 1 major + 2 minor or 4 minor

POSSIBLE BIV-ACM

BORDERLINE BIV-ACM

DEFINITE BIV-ACM

1 major or 2 minor

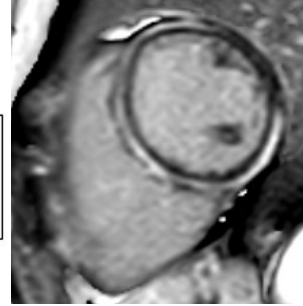
1 major + 1 minor or 3 minor

2 major or 1 major + 2 minor or 4 minor

POSSIBLE ALVC

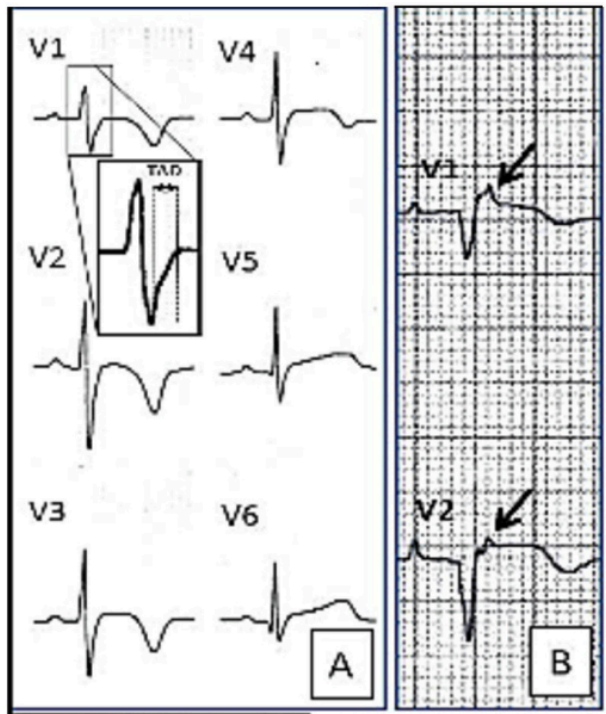
BORDERLINE ALVC

DEFINITE ALVC



European Task Force criteria for diagnosis of Arrhythmogenic Cardiomyopathy

RV PHENOTYPE : ECG CRITERIA



Major

- Negative T waves in right precordial leads (V_1 , V_2 , and V_3) or beyond in individuals ≥ 14 year-old (in the absence of complete RBBB and not preceded by J-point/ST-segment elevation)

Minor

- Negative T waves in leads V_1 and V_2 in males ≥ 14 year-old (in the absence of RBBB and not preceded by J-point/ST-segment elevation)
- Negative T waves beyond V_3 in the presence of complete RBBB
- Negative T waves beyond V_3 in individuals < 14 year-old

Minor

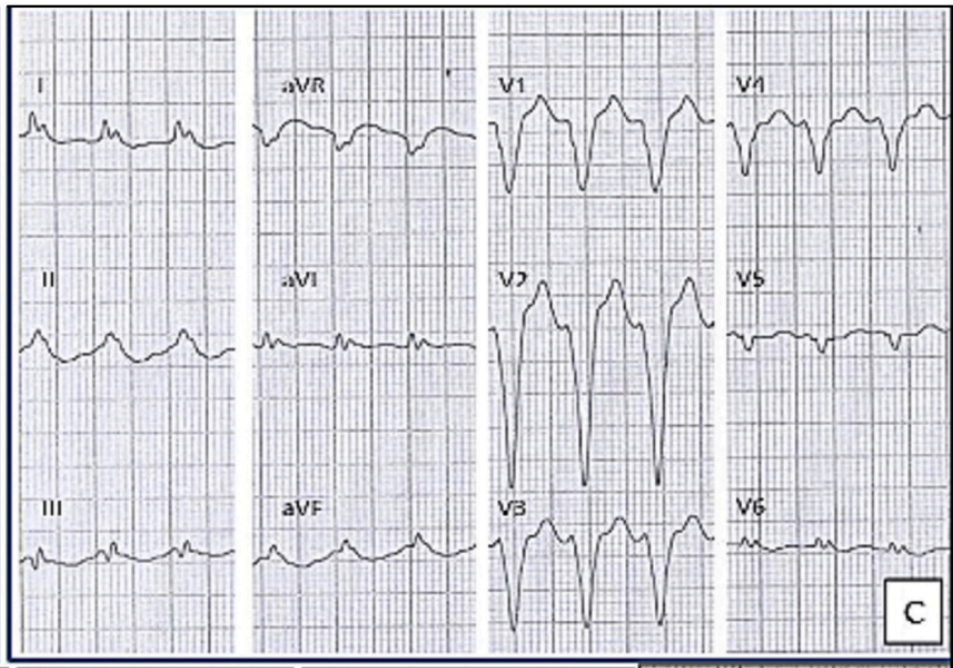
- Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads (V_1 to V_3)
- Terminal activation duration of QRS ≥ 55 ms measured from the nadir of the S wave to the end of the QRS, including R' , in V_1 , V_2 , or V_3 (in the absence of complete RBBB)

Corrado D et al. Int J Cardiol 2024 Jan 15:395:131447



European Task Force criteria for diagnosis of Arrhythmogenic Cardiomyopathy

RV PHENOTYPE : ARRHYTHMIAS



Major

- Frequent ventricular extrasystoles (>500 per 24 h), non-sustained or sustained ventricular tachycardia of LBBB morphology with non-inferior axis

Minor

- Frequent ventricular extrasystoles (>500 per 24 h), non-sustained or sustained ventricular tachycardia of LBBB morphology with inferior axis ("RVOT pattern")
- History of cardiac arrest due to ventricular fibrillation or sustained ventricular tachycardia of unknown morphology

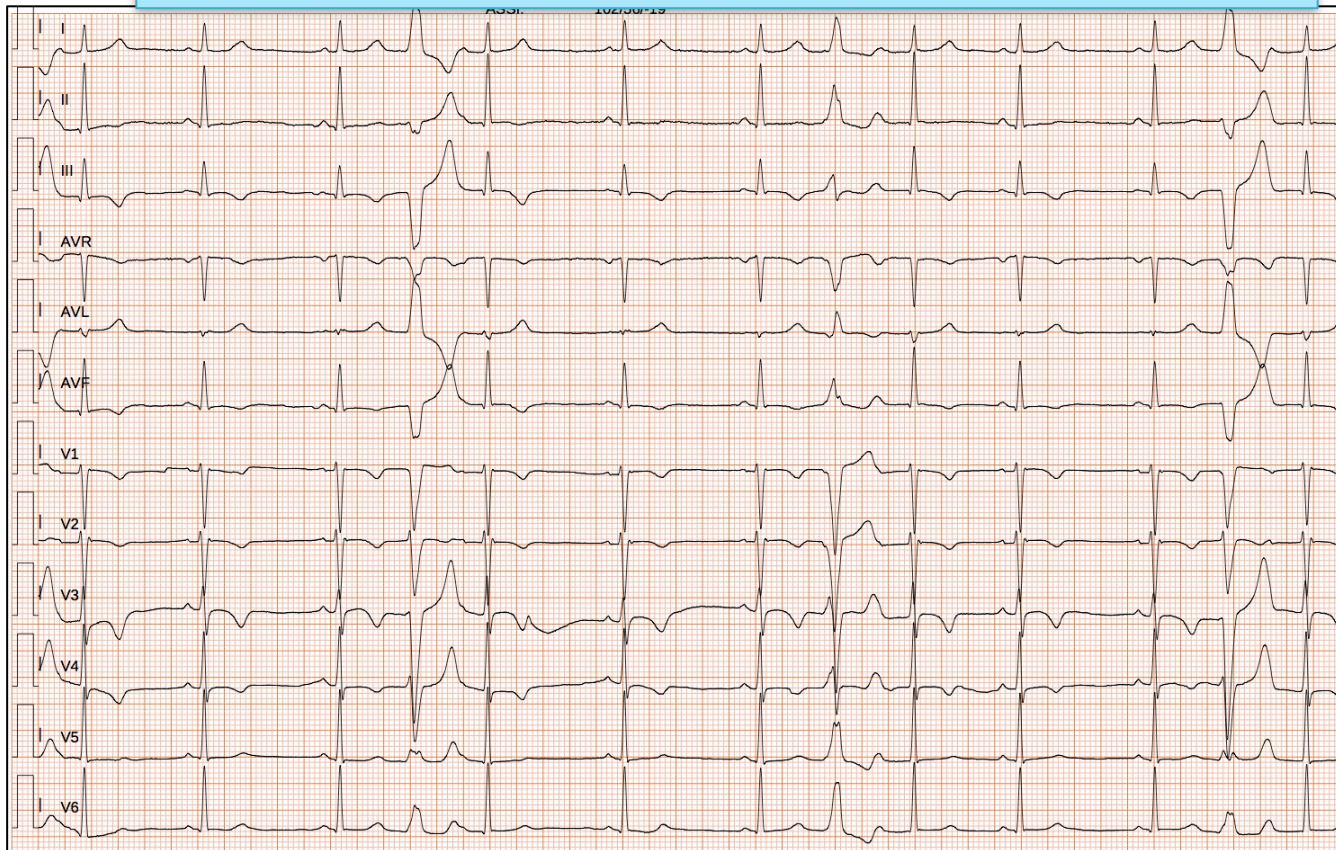


Alice, 19 anni.

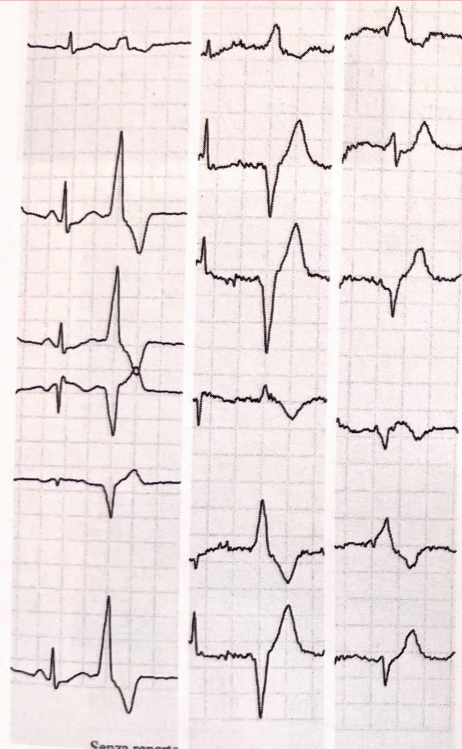
Ginnastica ritmica a livello agonistico

17 anni stop idoneità. Riscontro di anomalie ECG e Aritmie ventricolari.

*Mutazione
Gene DSP*



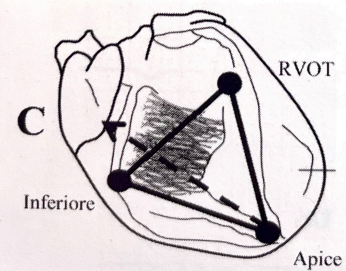
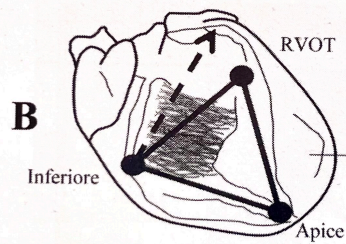
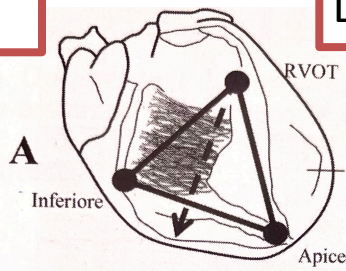
DERIVAZIONI PERIFERICHE



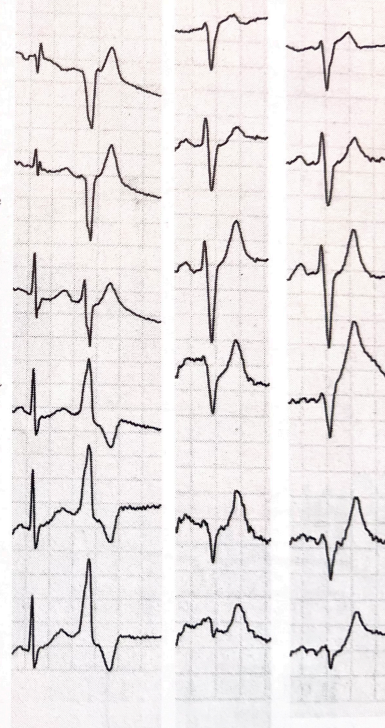
A

B

C



DERIVAZIONI PRECORDIALI



A

B

C



European Task Force criteria for diagnosis of Arrhythmogenic Cardiomyopathy

LV PHENOTYPE : ECG CRITERIA

Major

- Low QRS voltages (<0.5 mV peak to peak) in all limb leads in the absence of other causes (e.g. cardiac amyloidosis, obesity, emphysema, or pericardial effusion)

D

Minor

- Negative T waves in left precordial leads (V_4 - V_6) (in the absence of complete LBBB)

E

Corrado D et al. Int J Cardiol 2024 Jan 15;395:131447

LV PHENOTYPE : ARRHYTHMIAS

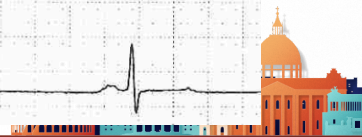
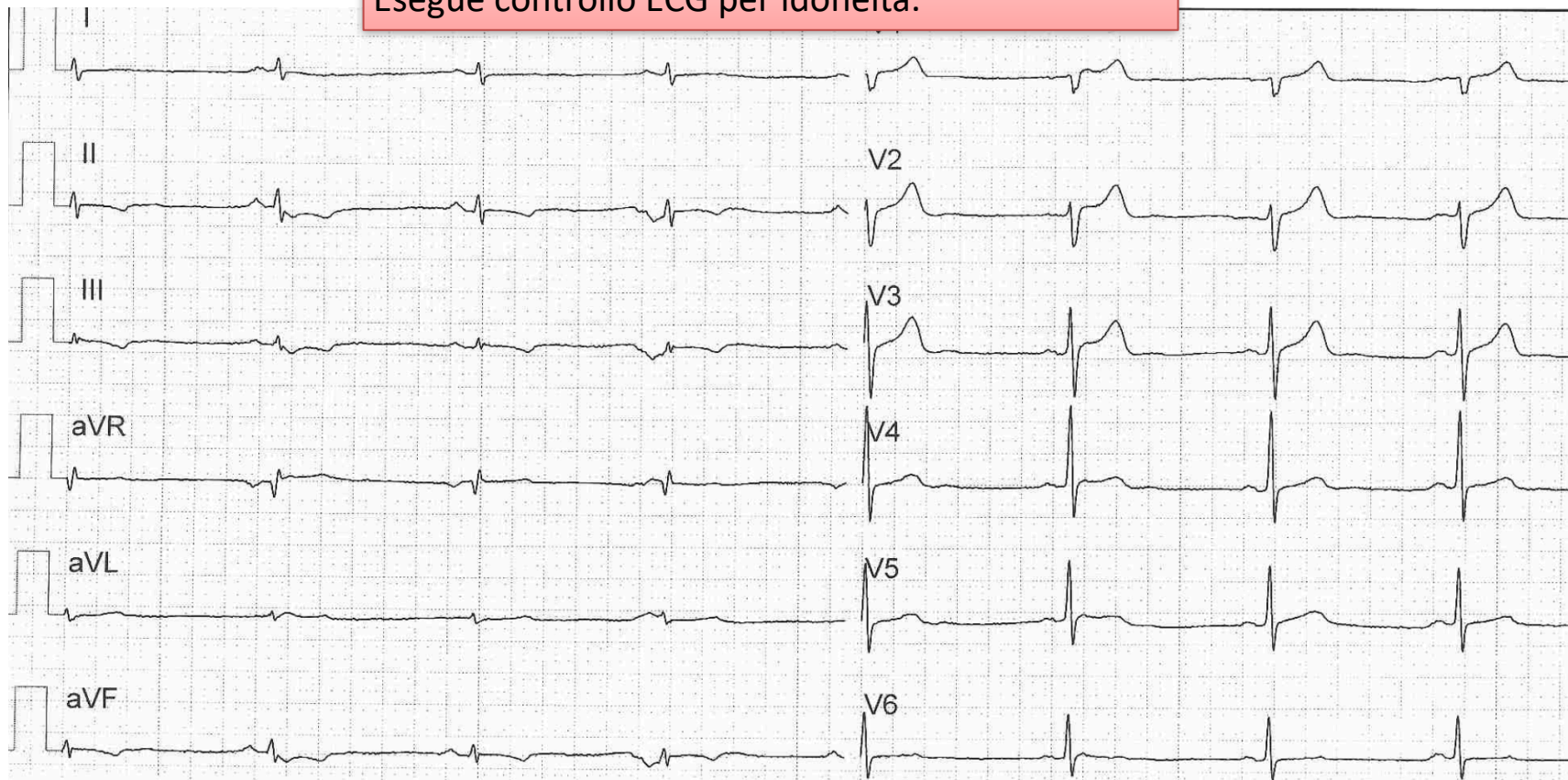
Minor

- Frequent (>500 per 24 h) or exercise-induced ventricular extrasystoles with a RBBB morphology or multiple RBBB morphologies (excluding the “fascicular pattern”)
- Non-sustained or sustained ventricular tachycardia with a RBBB morphology (excluding the “fascicular pattern”)
- History of cardiac arrest due to ventricular fibrillation or sustained ventricular tachycardia of unknown morphology

Corrado D et al. Int J Cardiol 2024 Jan 15:395:131447



Alessandro, 32 anni. Familiarità per CMD (Madre).
Calciatore non agonista.
Esegue controllo ECG per idoneità.



Riscontro di **extrasistolia ventricolare
polimorfa e complessa da sforzo**
ASINTOMATICO

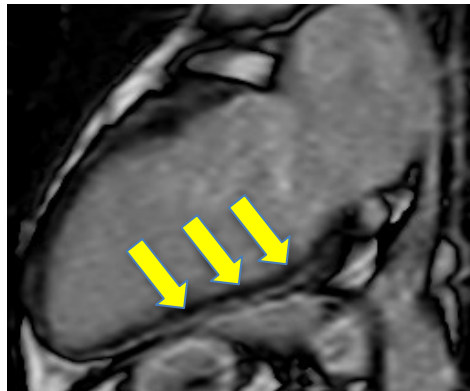
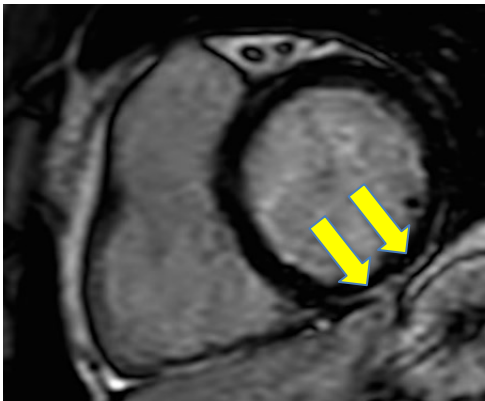
Alessandro, 32 anni



EcocardiogrammaColorDoppler nei limiti di norma



Test genetico: variante P gene DSP



Arrhythmogenic cardiomyopathy diagnosis and management: a systematic review of clinical practice guidelines and recommendations with insights for future research



Galanti K et al *Cardiogenetics* 2025, 15(3), 22;



Diagnostic delay in arrhythmogenic cardiomyopathy

Giacomo Tini ^{1*†}, Maddalena Graziosi^{2†}, Beatrice Musumeci¹, Mattia Targetti³, Domitilla Russo¹, Vanda Parisi², Alessia Argirò ³, Raffaello Ditaranto², Ornella Leone ⁴, Camillo Autore⁵, Iacopo Olivotto ^{3‡}, and Elena Biagini ^{2‡}

- Data of 174 ACM patients from three Italian Cardiomyopathy Referral Centers
- **Almost one-third** of patients with ACM experience a **diagnostic delay >2 years**. These patients are mostly affected by an **ACM phenotype with LV involvement** and present **worse mortality** compared with those without diagnostic delay.



ACM was the cause of SCD in **29%** of athletes

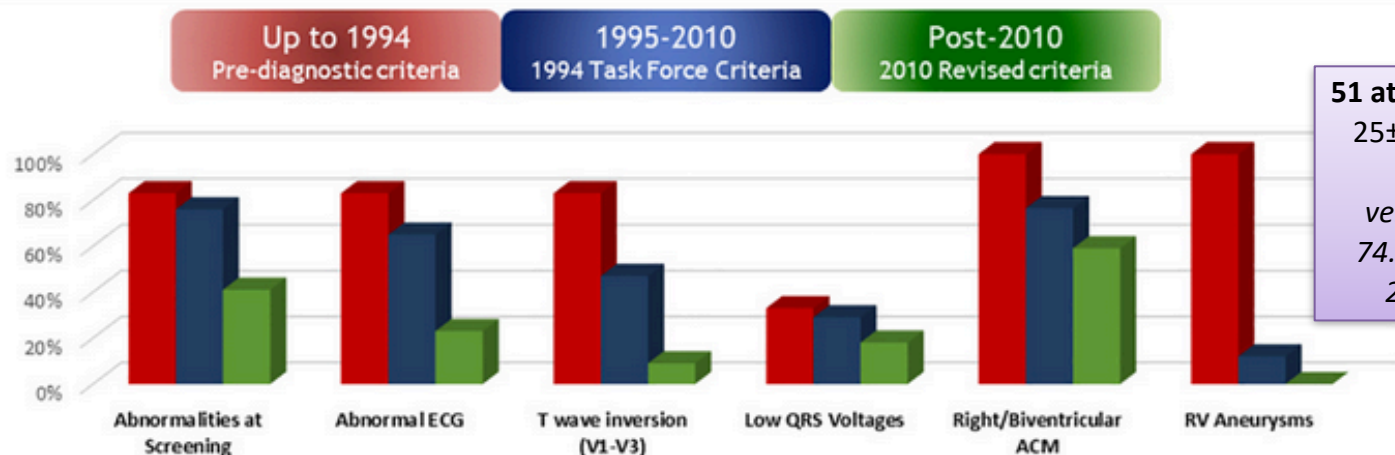
The incidence rate of SCD in athletes was 0.43

versus **0.14 (0.05–0.33) per 100 000/y** before and after 2010, respectively.

Sudden Death in Young Competitive Athletes Due to Arrhythmogenic Cardiomyopathy: A 4-Decade National Referral Center Experience

De Gaspari et al Circ Arrhythm Electrophysiol. 2026;19:e014877

Evolution of Clinical Profile and Phenotype of Athletes who Died Suddenly with Arrhythmogenic Cardiomyopathy Across Diagnostic Eras



51 athletes with ACM (50 male, 25±6.4 years) were enrolled.
The pattern was *right ventricular/biventricular* in 74.5% and *left ventricular* in 25.5% (**41% after 2010**)

A phenotypic shift toward the LEFT ventricular variant



Sudden Death in Young Competitive Athletes Due to Arrhythmogenic Cardiomyopathy: A 4-Decade National Referral Center Experience

Table 3. Analysis of PPS Data by ACM Variant Detected at Autopsy

	RV/BIV, ACM Nr. 38	LV ACM Nr. 13	P value
Abnormalities at PPS, n (%)	28 (73.6)	4 (30.7)	0.0087
Positive family history of SCD or ACM	7 (18.4)	3 (23.1)	0.7015
Previous symptoms	9 (23.7)	3 (23.1)	>0.9999
Abnormal ECG, n (%)	23 (60.5)	3 (23.1)	0.0266
Negative T waves, n (%)	19 (50)	1 (7.7)	0.0083
Low QRS voltage, n (%)	11 (28.9)	2 (15.4)	0.4714
LBBB or polymorphic VA on basal ECG or limited exercise test, n (%)	17 (44.7)	1 (7.7)	0.0193
Further tests prescribed for PPS abnormalities, n (%)	19 (50)	4 (30.7)	0.3355
24-hour Holter monitoring (positive/performed), n (%)	5/10 (50)	0/1 (0)	>0.9999
Maximal stress test (positive/performed), n (%)	11/16 (68.7)	0/4 (0)	0.0260
2D echocardiography (positive/performed), n (%)	1/14 (7.1)	0/3 (0)	>0.9999
CE-CMR (positive/performed), n (%)	1/1 (100)	...	...

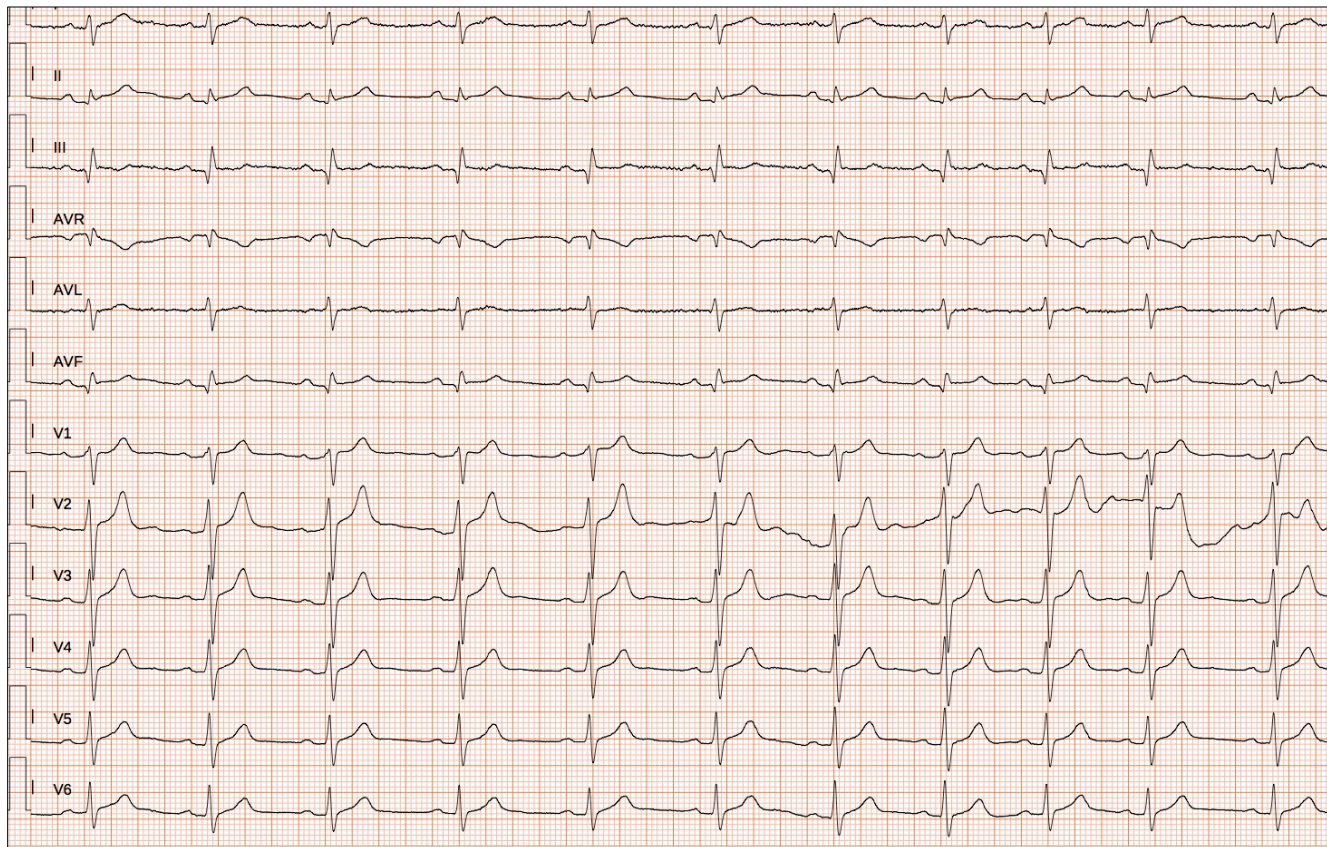
P values were calculated using Fisher exact test for comparison across groups. 2D indicates 2-dimensional; ACM, arrhythmogenic cardiomyopathy; BIV, biventricular; CE-CMR, contrast-enhanced cardiac magnetic resonance; LBBB, left bundle-branch block; LV, left ventricular; PPS, preparticipation screening; RV, right ventricular; SCD, sudden cardiac death; and VA, ventricular arrhythmias.

De Gaspari et al *Circ Arrhythm Electrophysiol.* 2026;19:e014877

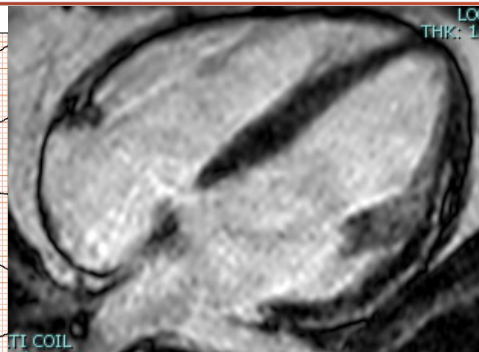
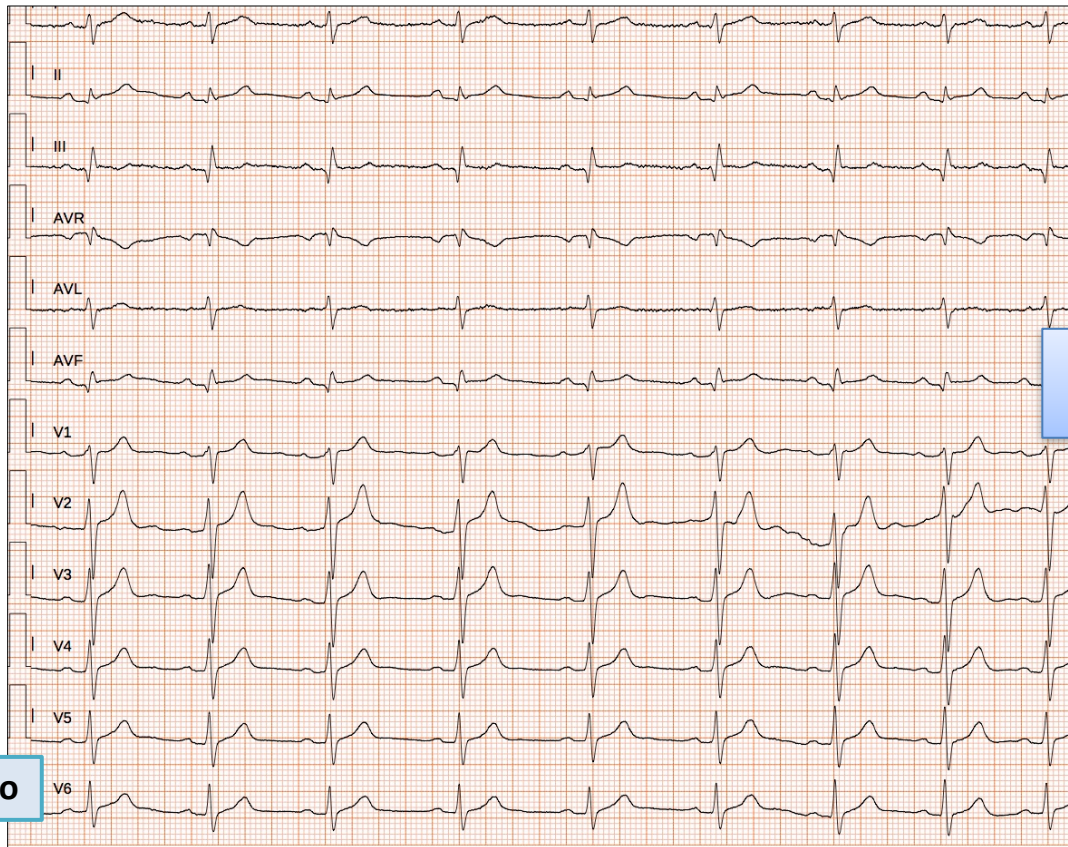


- Alessandro, 38 anni
- Sportivo Agonista (ex ciclista)
- Non familiarità per MCI/cardiomiopatie/canalopatie
- Riscontro di extrasistolia ventricolare isolata durante accertamenti medico-sportivi
- ECG Holter delle 24h : 38 BEV isolati

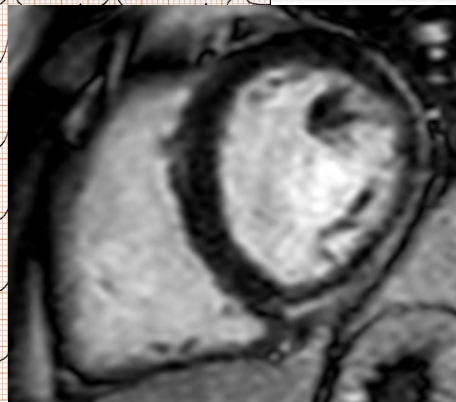




ECG e Imaging



Conservati Volumi e funzione
biventricolare

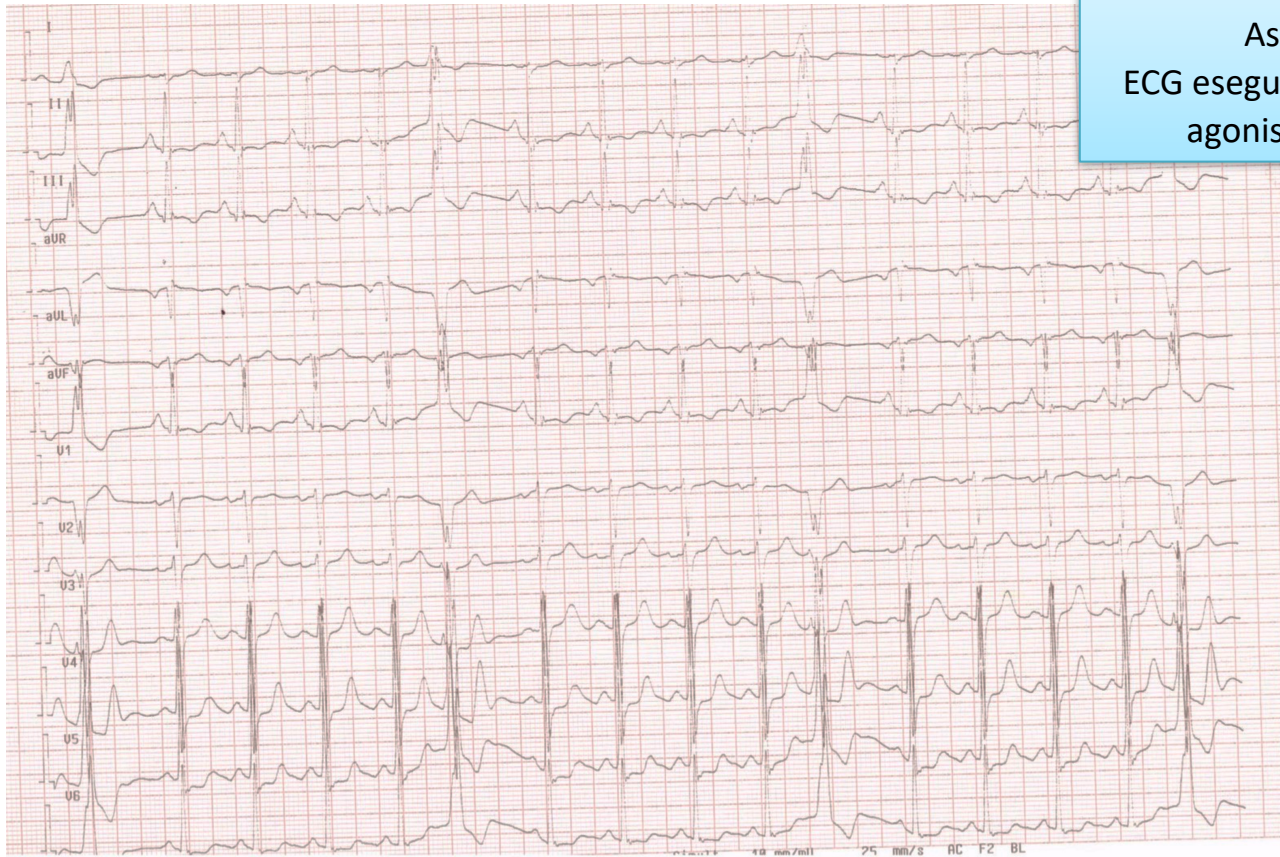


Genetica in corso



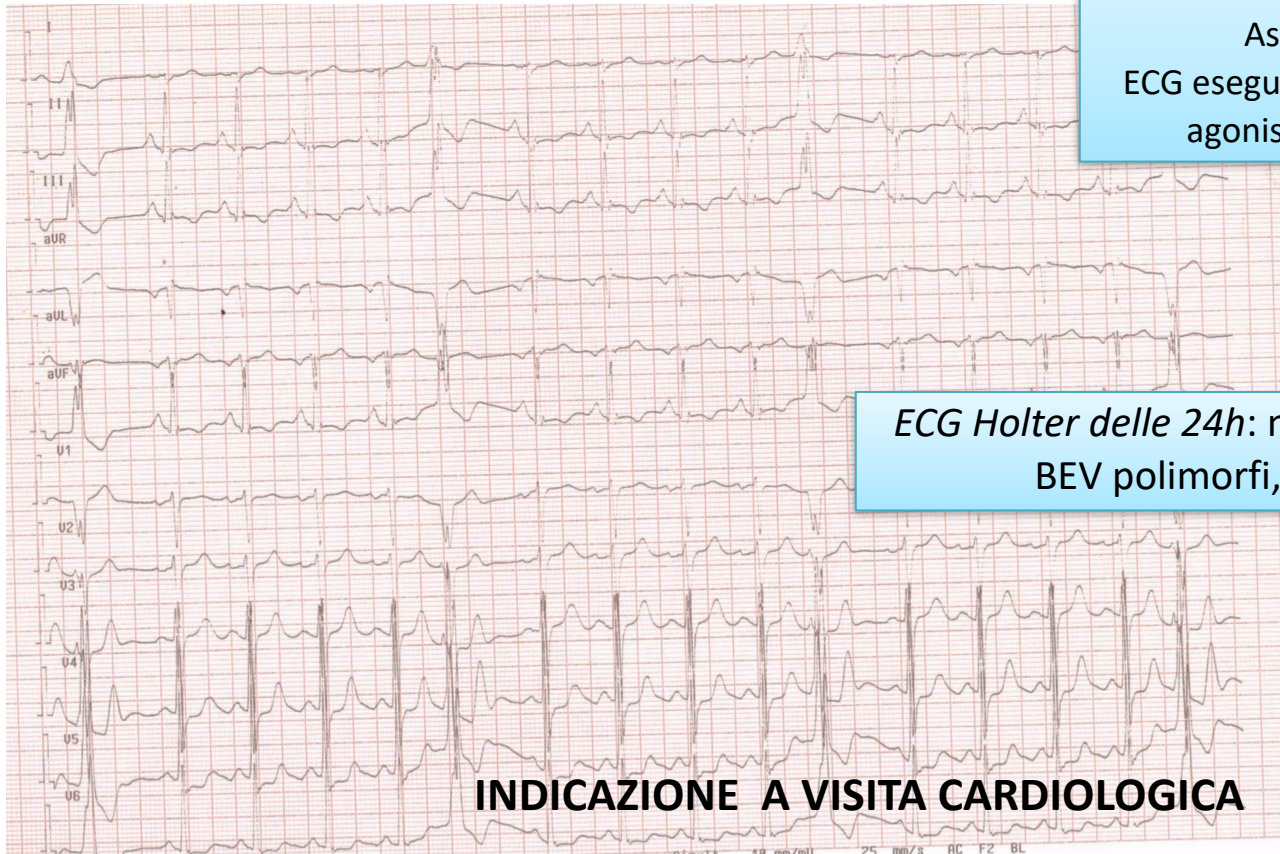
ECG dicembre 2025

Alessandra, 49 anni
Familiarità per aritmie (padre)
Asintomatica
ECG eseguito per attività non
agonistica (palestra)



ECG dicembre 2025

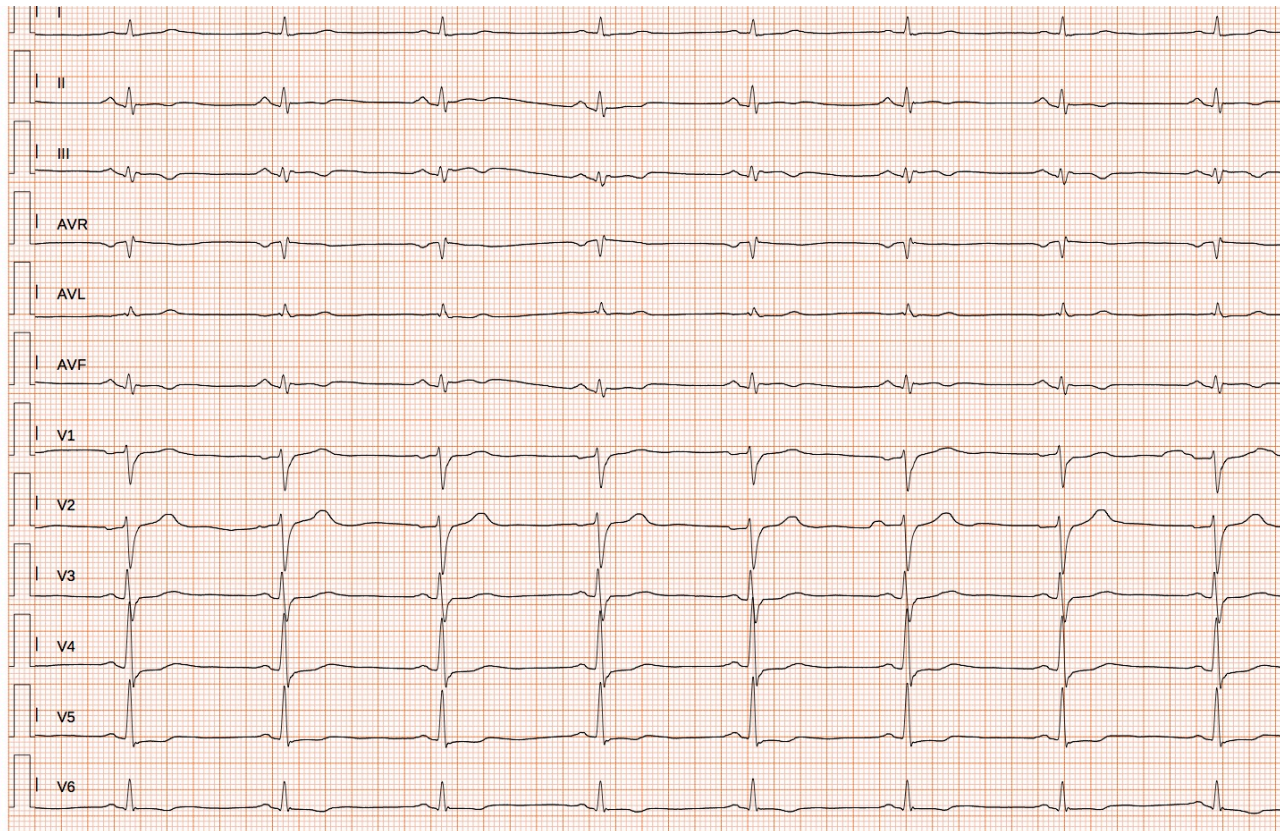
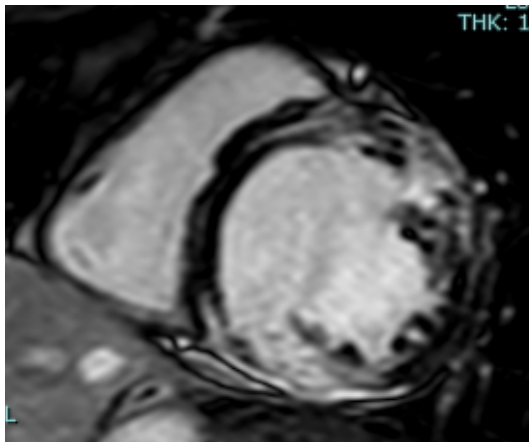
Alessandra, 49 anni
Familiarità per aritmie (padre)
Asintomatica
ECG eseguito per attività non
agonistica (palestra)

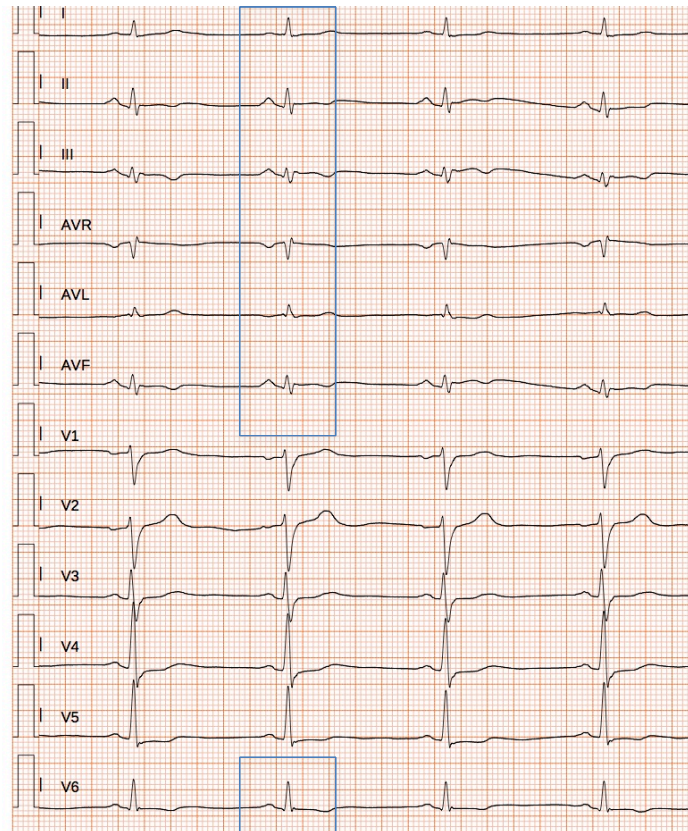
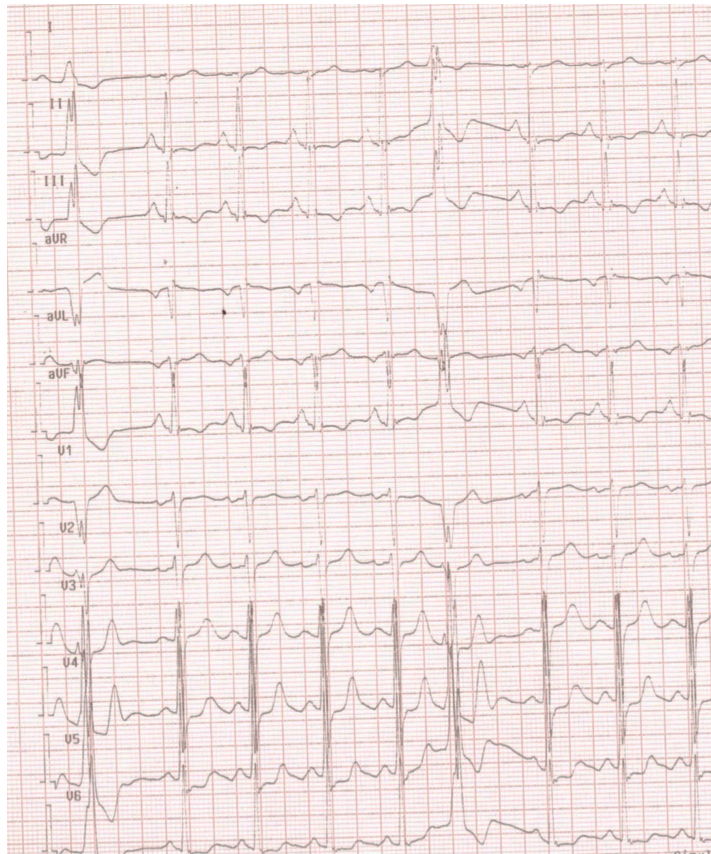


*ECG Holter delle 24h: numerosi 33000
BEV polimorfi, coppie*



Ecocardiogramma C/D e RMC:
Dilatazione ventricolare sinistra
con severa disfunzione
ventricolare (FE 30%)

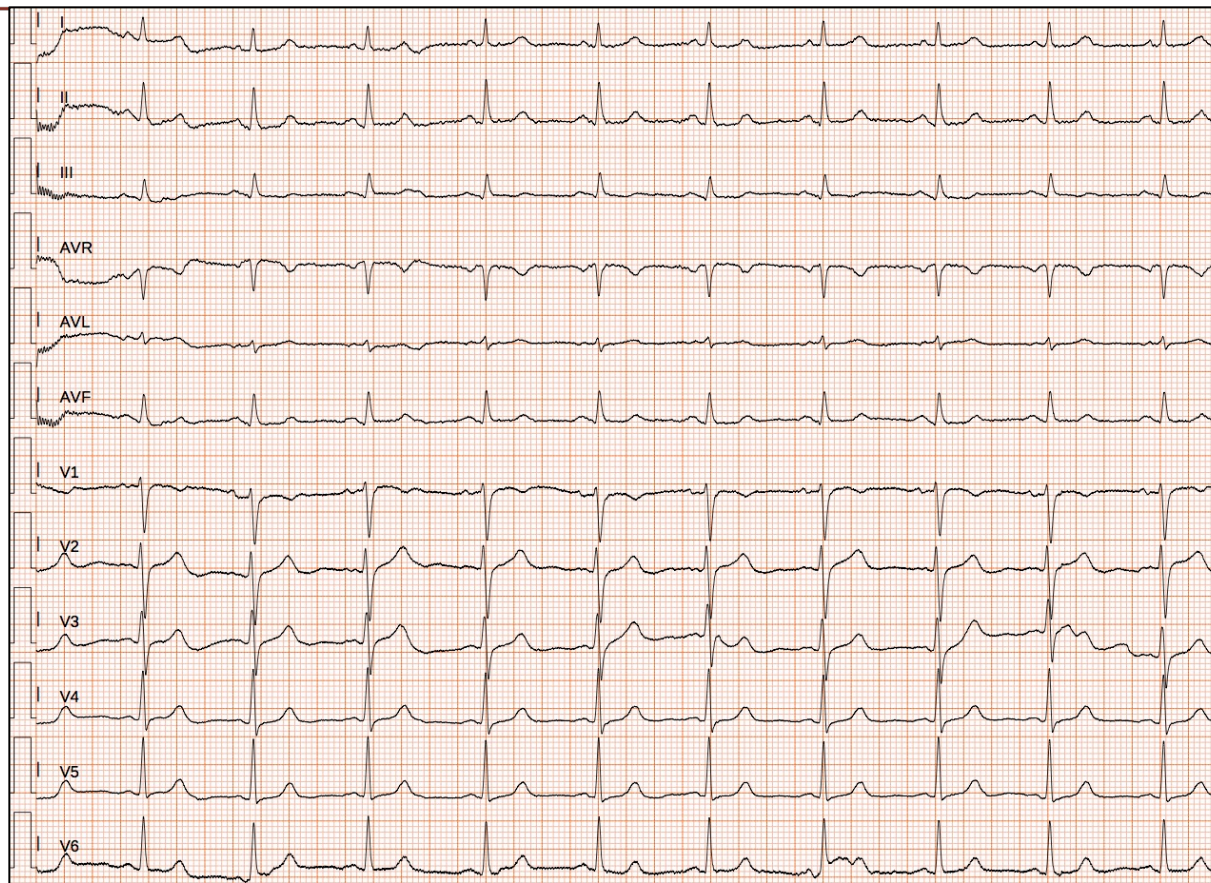




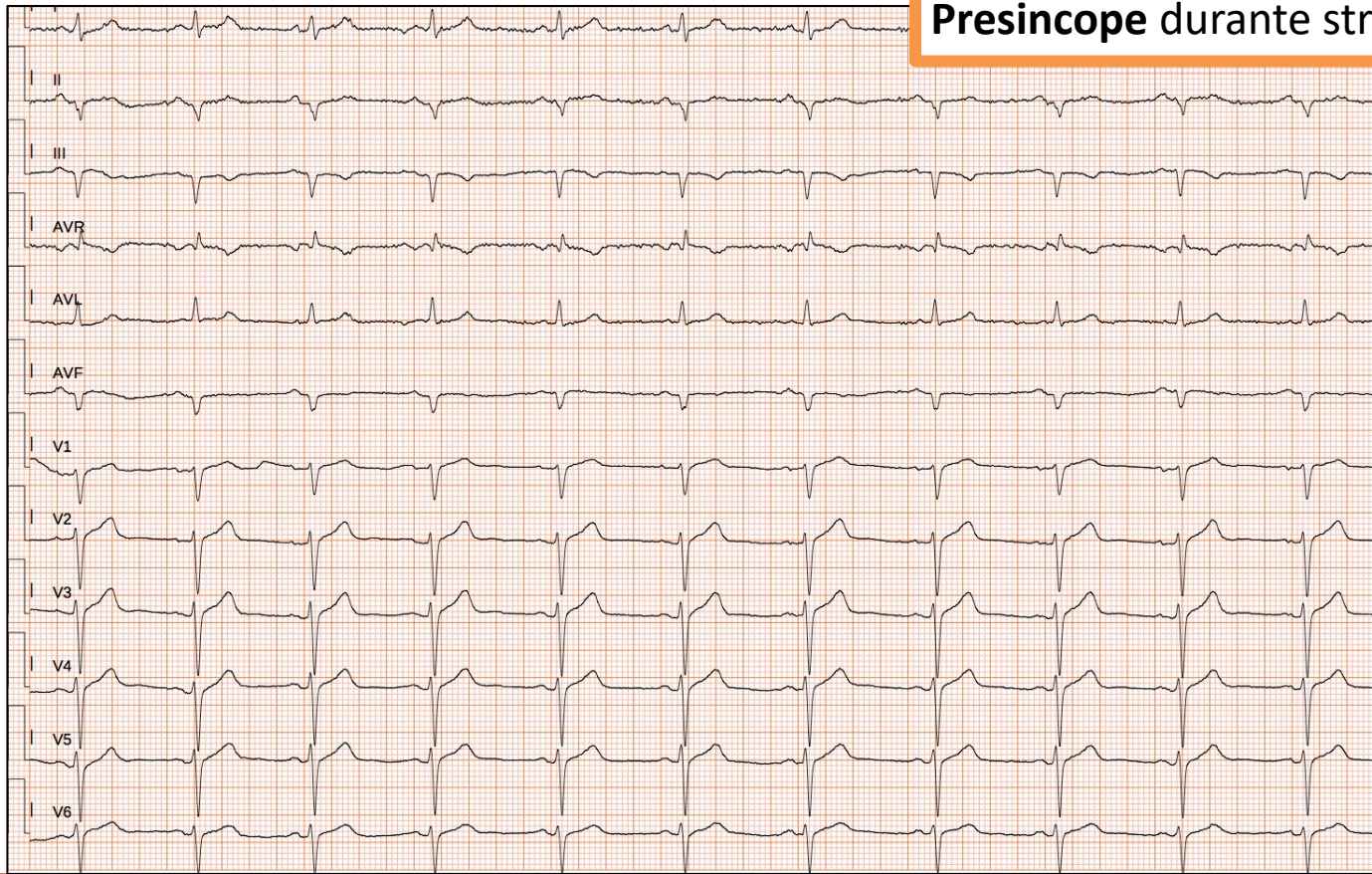
- Mario, 39 anni
- Storia familiare di CMD a variante aritmogena (fratello portatore di ICD, variante patogena **gene DSP**)
- Asintomatico
- *EcocardiogrammaColordoppler* normale
- *ECG Holter delle 24h*: 300 BEV monomorfi (non descritta morfologia)
- Test genetico e Risonanza Magnetica Cardiaca rifiutati



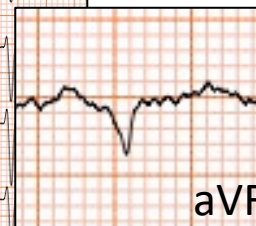
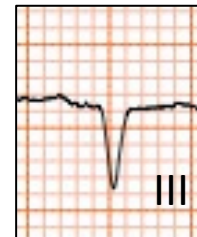
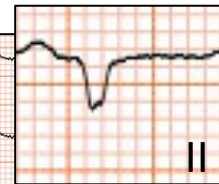
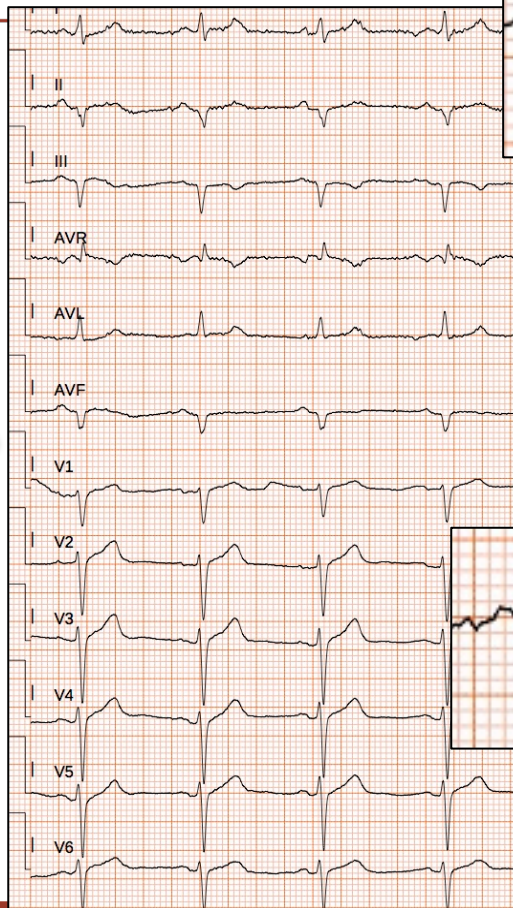
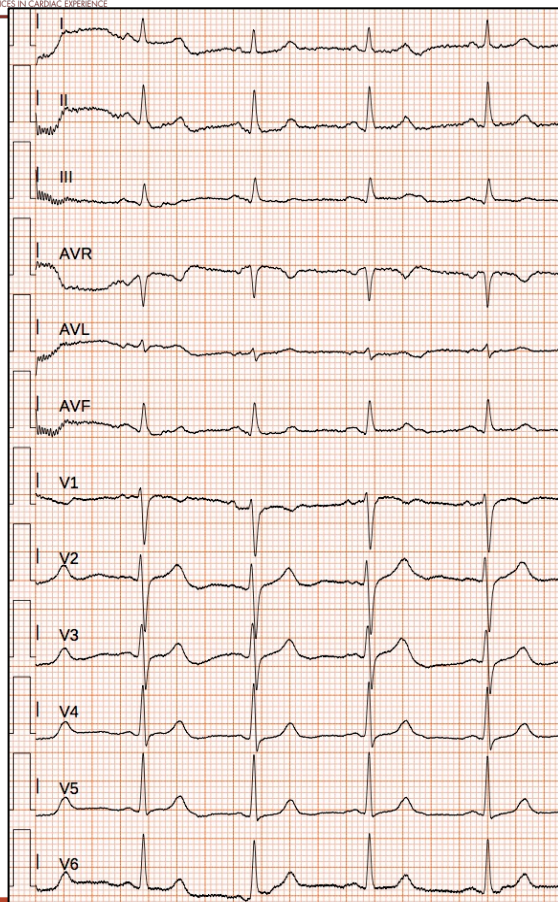
ECG

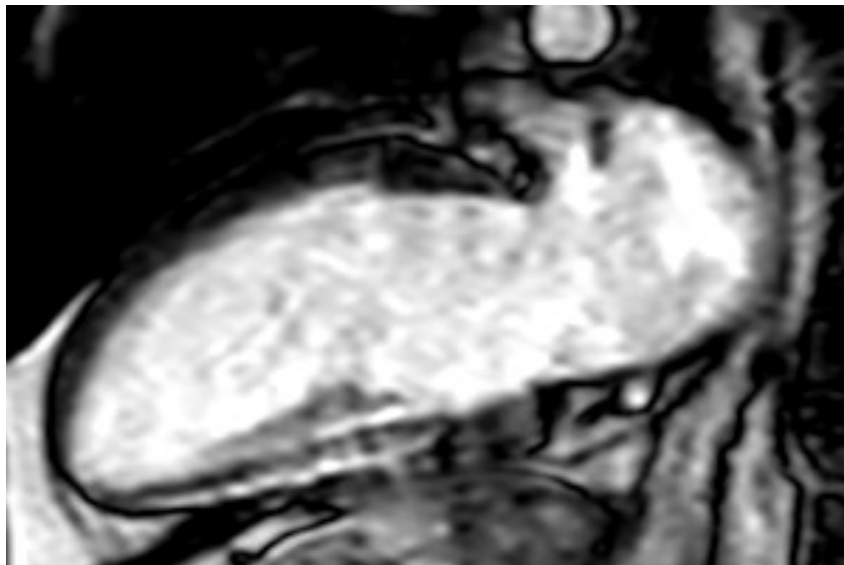


Presincope durante stress emotivo



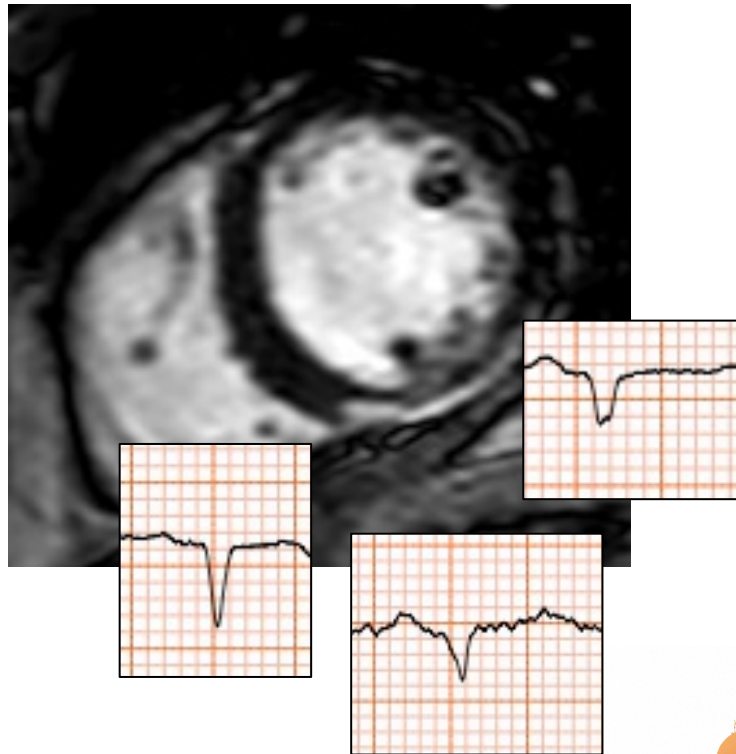
ECG nel tempo





Normali dimensioni, volumi e funzione biventricolare
LGE inferiore ed inferolaterale medio-apicale

Segregazione genica positiva per DSP



D. , 21 anni, donna

Familiarità per aritmie da parte paterna

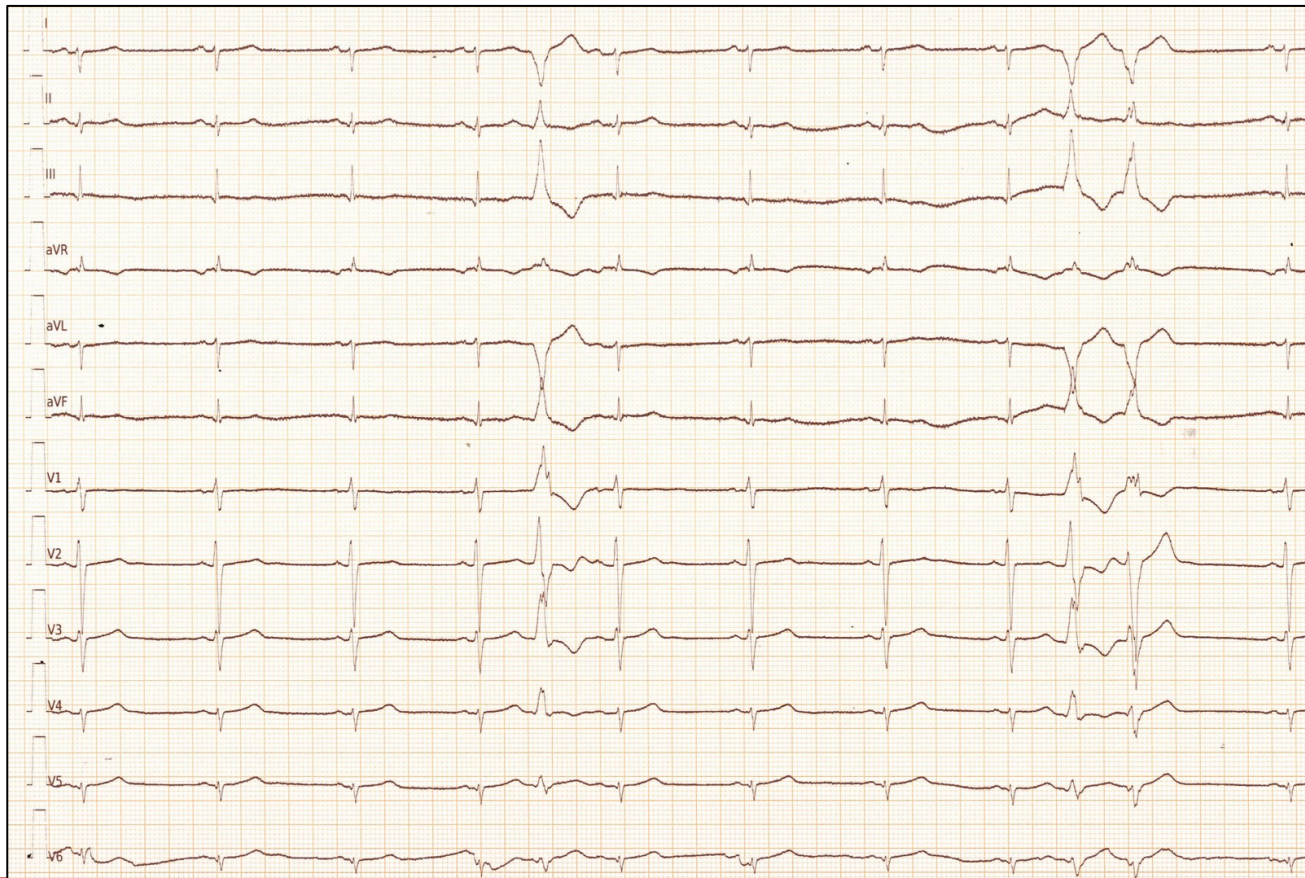
Non precedenti cardiologici

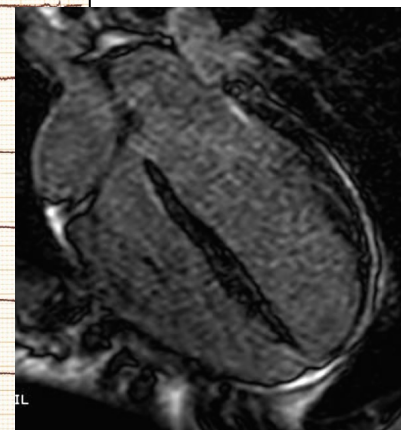
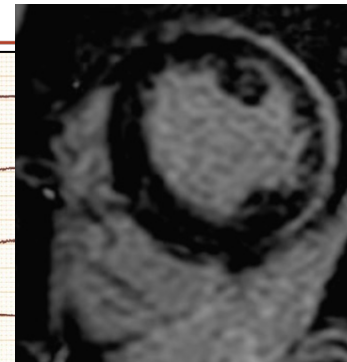
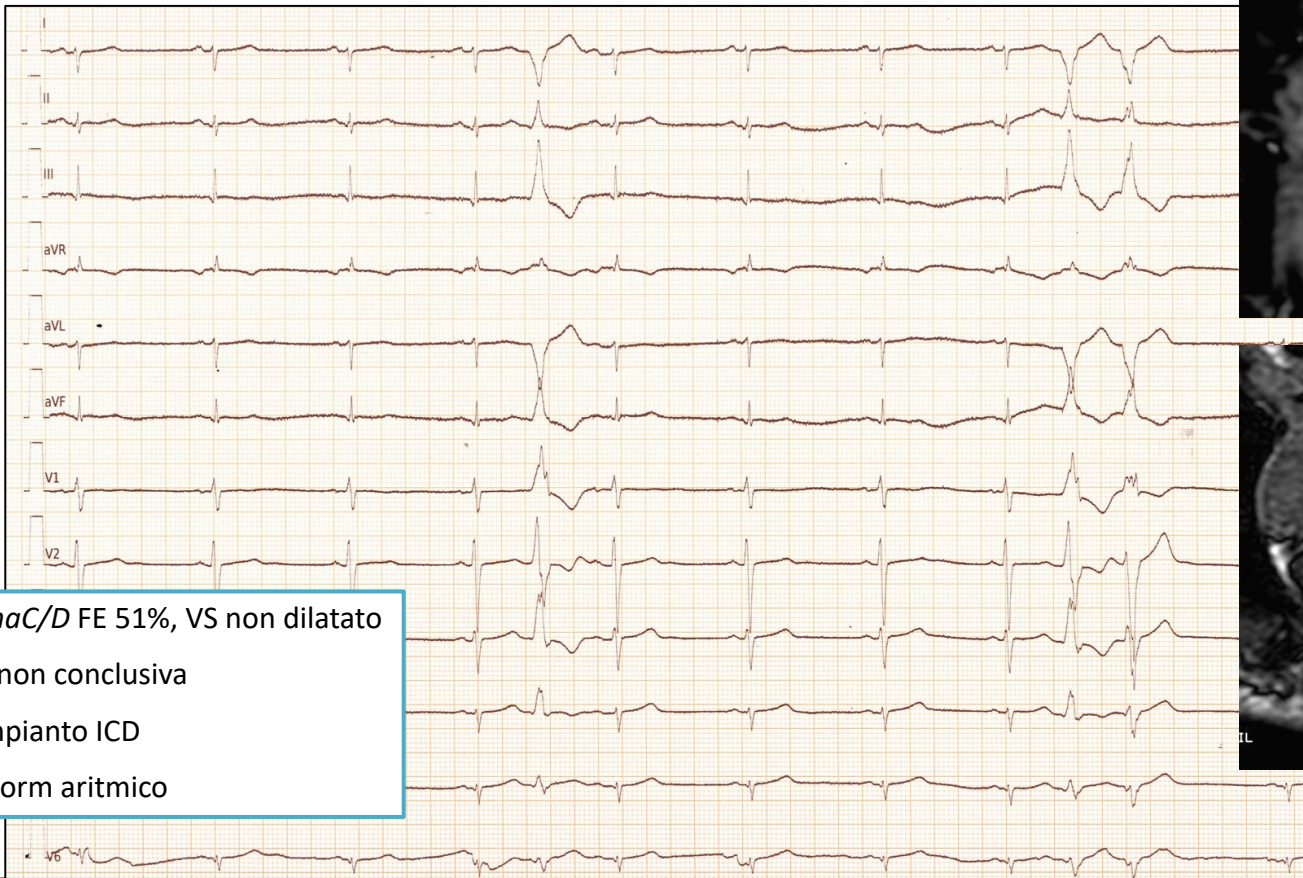
Arresto cardiaco testimoniato mentre teneva lezione in piscina

Ritmo defibrillabile → Dc shock

In PS FV/TdP multipli Dc shock







Ecocardiogramma C/D FE 51%, VS non dilatato

Analisi genetica non conclusiva

Sottoposta ad impianto ICD

Un anno dopo storm aritmico



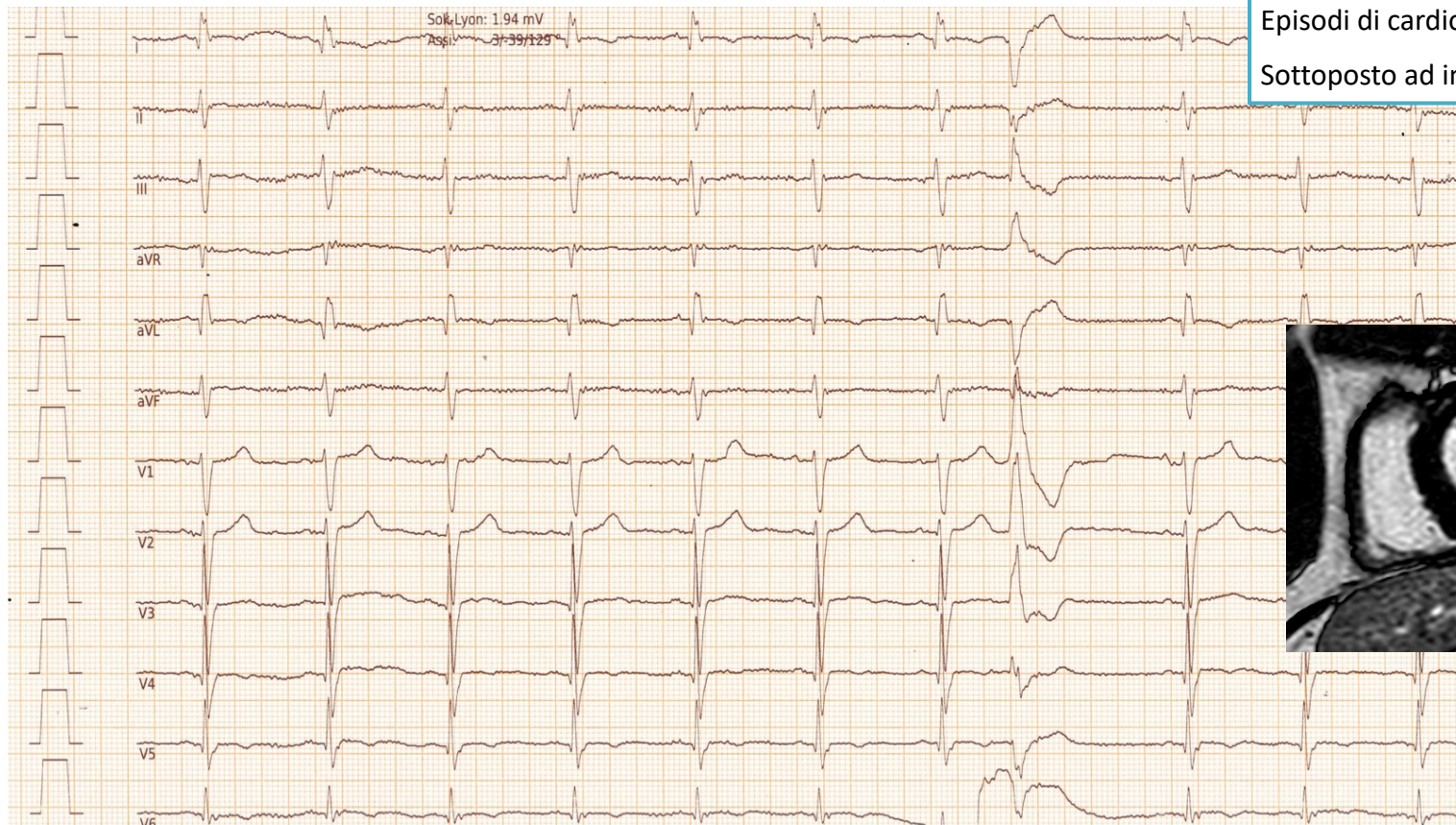
ECG nel papà

Ecocardiogramma C/D FE 40%,

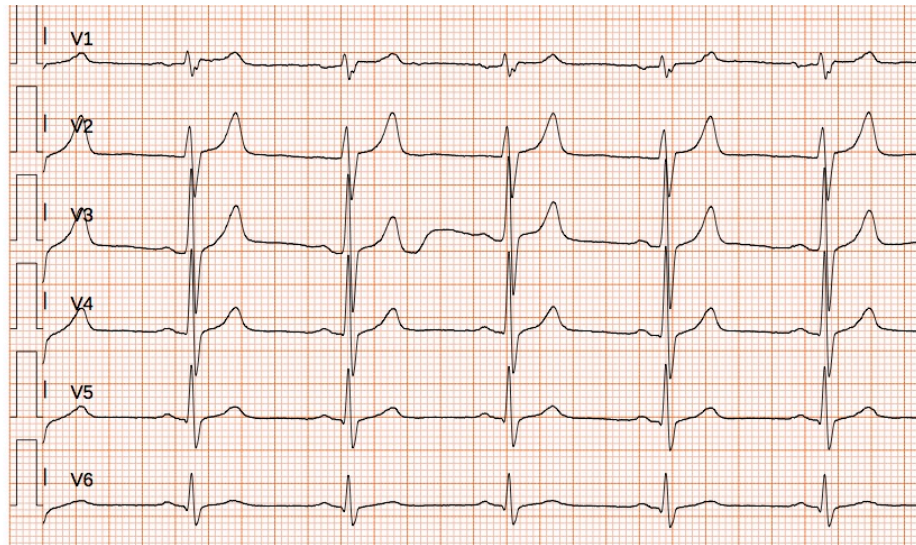
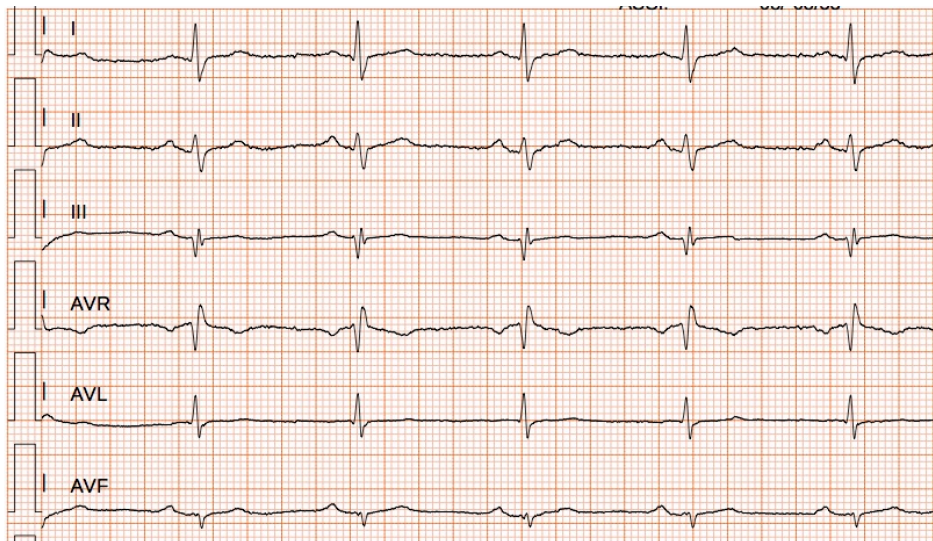
VS non dilatato

Episodi di cardiopalmo

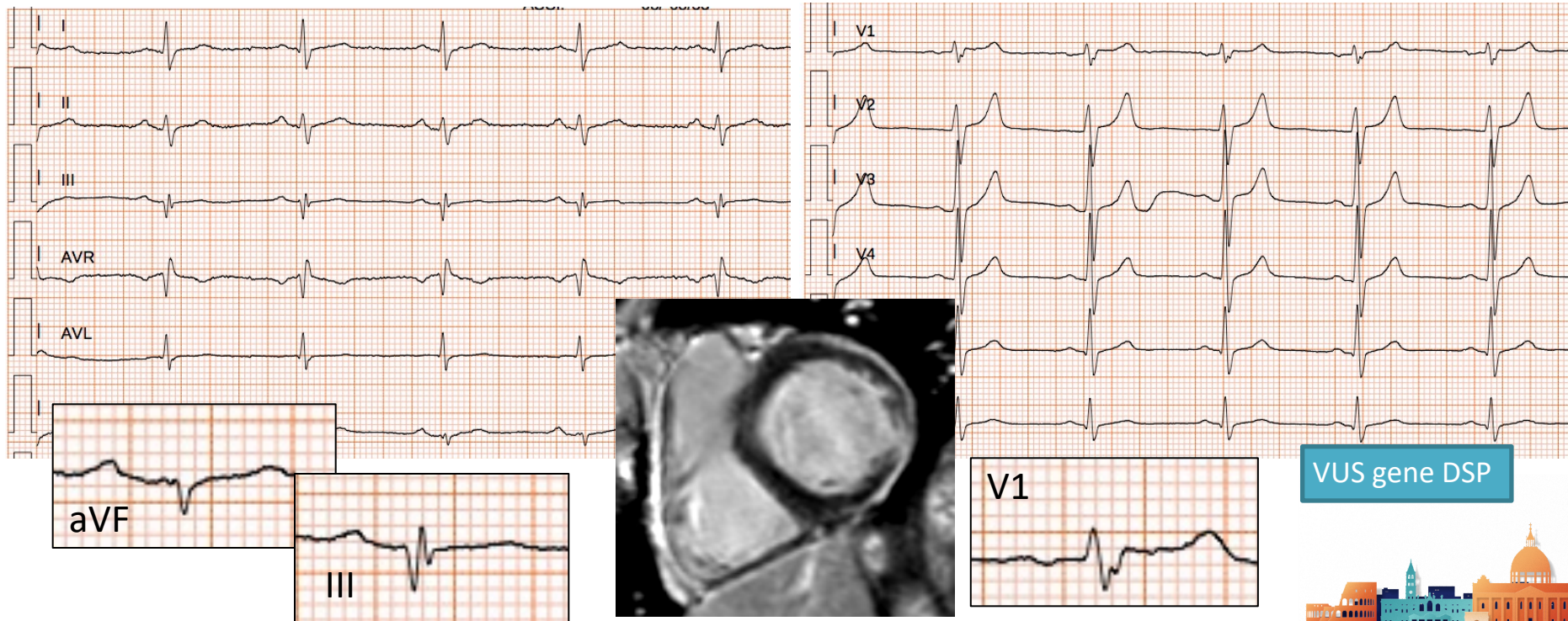
Sottoposto ad impianto ICD



Andrea, 35 anni. Non precedenti cardiologici
Esordio clinico arresto cardiaco extraospedaliero



Andrea, 35 anni. Non precedenti cardiologici
Esordio clinico arresto cardiaco extraospedaliero



ORIGINAL RESEARCH

CLINICAL ELECTROPHYSIOLOGY - ELECTROCARDIOGRAPHY

The Diagnostic Value of the 12-Lead ECG in Arrhythmogenic Left Ventricular Cardiomyopathy



Novel ECG Signs

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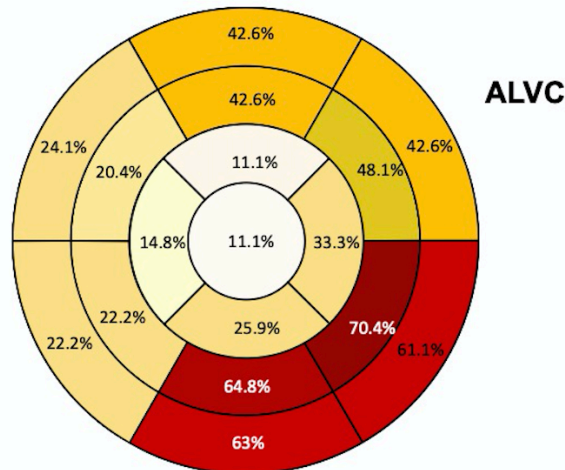


TABLE 1 Baseline Clinical, Structural, and Genetic Characteristics of the Study Population (ALVC, N = 54)

Age at diagnosis, y	39 ± 15
Male	32 (59.3)
Probands	40 (74.1)
Family history of ACM/DCM	23 (42.6)
Family history of SCD	18 (33.3)
NYHA functional class I to II	52 (96.3)
NYHA functional class III	2 (3.7)
Atrial fibrillation	4 (7.4)
Unexplained syncope	8 (14.8)
NSVT	26 (48.1)
SVT	9 (16.7)
VF	1 (1.9)
Cardiac magnetic resonance	
LVEDVi, mL/m ²	97.6 ± 24.5
LVEF, %	49.5 ± 10.0
LV WMA, %	35 (64.8)
RVEDVi, mL/m ²	85.8 ± 18.8
RVEF, %	54.3 ± 9.3
RV WMA, %	13 (24.1)
Intramyocardial fat signal	22 (40.7)
Segments with LGE	6 ± 3, 6 (4-8)
LGE pattern	
Ringlike	28 (51.9)
LGE distribution	
Subepicardial	35 (64.8)
Midmural	10 (18.5)
Transmural	9 (16.7)

54 pz con ALVC (32 uomini, età 39 ±15 anni) vs 84 soggetti sani (normale RMN cardiaca)

Mutazioni P/LP in geni codificanti **proteine desmosomiali (DSP 72.9%; DSG2 14.6%).**



Ringlike pattern was prevalent in DSP alteration carriers (54.3% vs 23.1%; P=0.05)

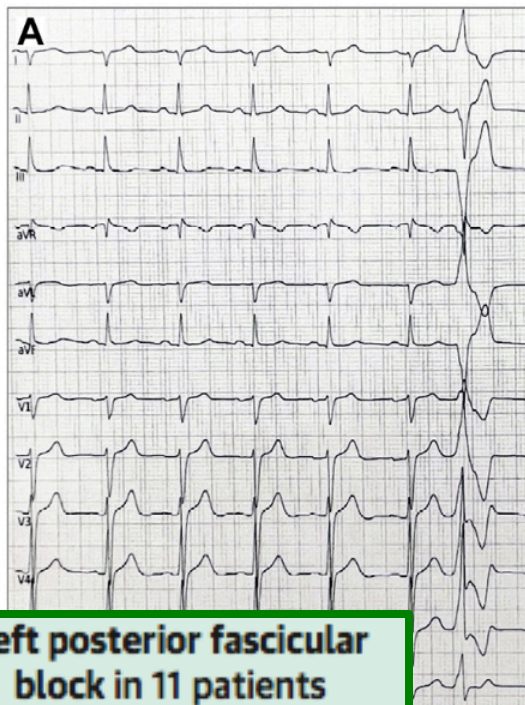
Otto pz (14.8%) ECG apparentemente normale

Bassi voltaggi periferici in 12 paz (22.2%).

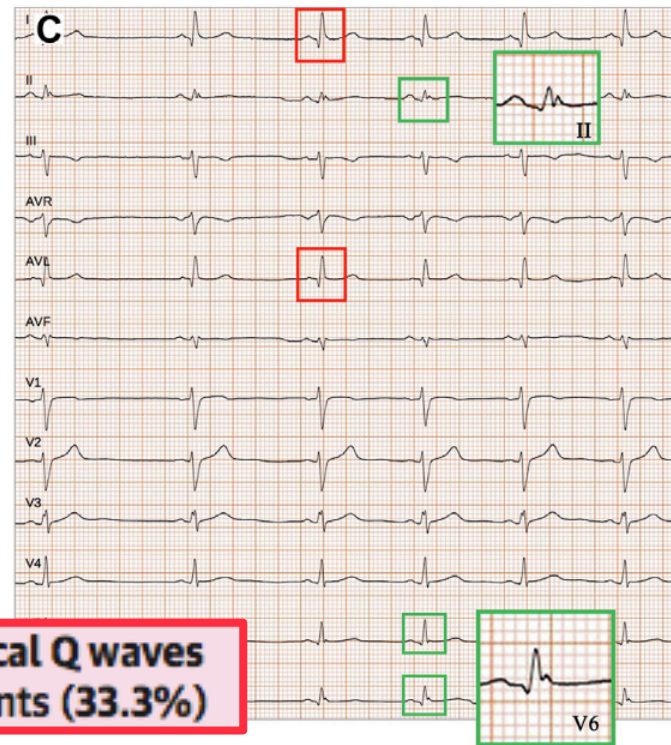
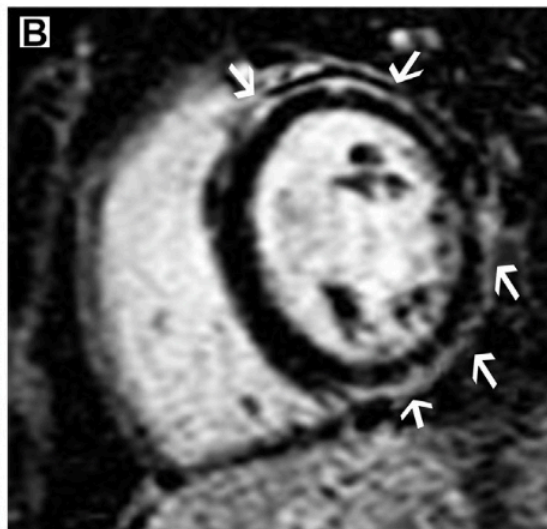
Onde T negative in 31 paz (57.4%),



FIGURE 1 Typical Findings of Arrhythmogenic Left Ventricular Cardiomyopathy Patients

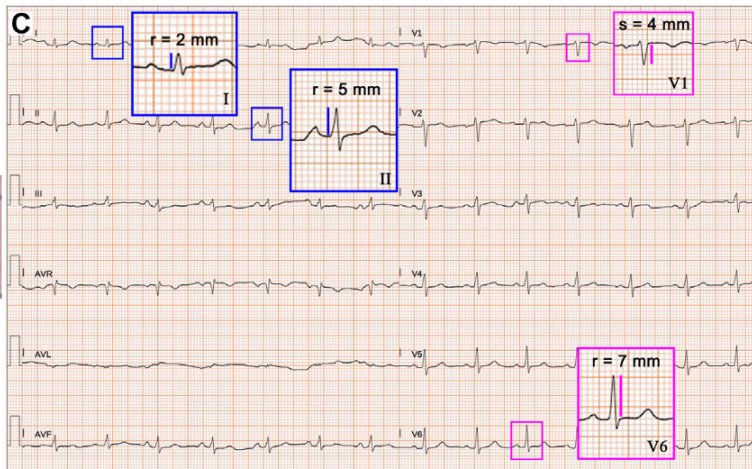
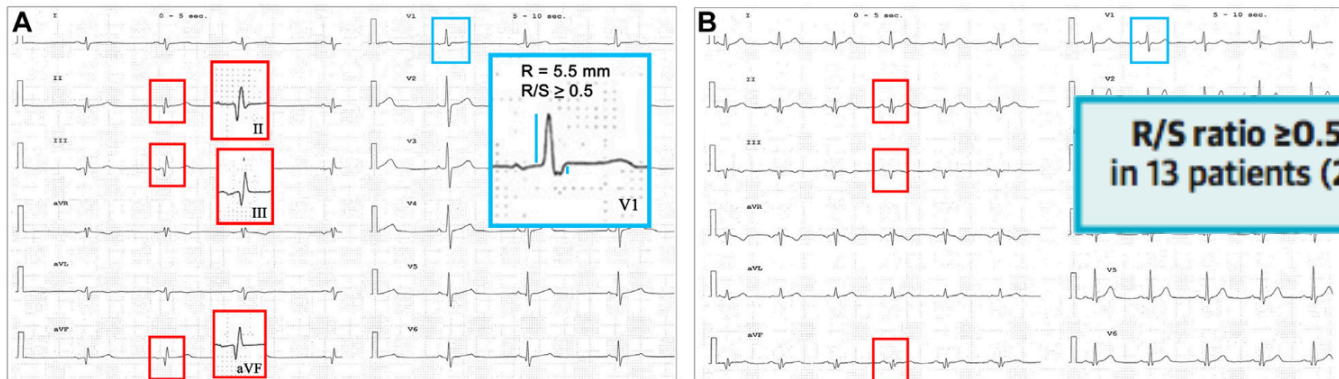


**Left posterior fascicular
block in 11 patients
(20.4%)**

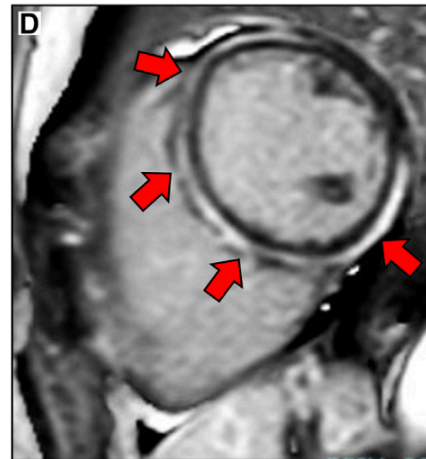


**Pathological Q waves
in 18 patients (33.3%)**

FIGURE 2 Typical Findings of ALVC Patients



Sum of the R-wave in I-II ≤ 8 mm
AND S-wave in V₁ plus R-wave
in V₆ ≤ 12 mm in 24 (44.4%)



Case ID	References	Figure	12 lead ECG findings	Genetic analysis	CMR data	Endomyocardial biopsy data/ Histologic data
Case 1	Protonotarios A et al ¹ <i>J Electrocardiol.</i> 2013	Fig. 1	LPFB , LQRSV in limb and left precordial leads, TW1 V5-V6, rQRS in lead V1.	Not performed	Not performed	Fibrotic subepicardial and midwall bands on anterolateral and postero-apical LV walls and on the interventricular septum. Myocyte loss with fibro-fatty replacement and myocyte abnormalities
Case 2	Zorzi A et al ² <i>Circ Arrhythm Electrophysiol.</i> 2016	Fig. 4	LPFB Inferolateral TW1 Pathological Q in II-III-aVF Prominent R wave V1 LQRSV in left precordial leads	Not performed	Sub/midmyocardial LGE with a striated pattern involving the infero-lateral LV wall	Extensive fibrosis in the sub- and midmyocardial layers (inferolateral LV), focal and patchy fatty infiltration. Cardiomyocytes hypertrophic with dysmorphic and dysmorphic nuclei, with cytoplasmic vacuolization.
Case 3	Oloriz T et al ³ <i>Europace.</i> 2016	Fig. 1 -right panel	LPFB - R/S ratio V1 ≥ 0.5	Not performed	Infero-lateral scar	Not performed
Case 4	Miles C et al ⁴ <i>Circulation.</i> 2019	Fig. 4	LPFB - rQRS infero-septal leads First-degree AV block, Inferolateral TW1 - LQRSV in limb leads, prolonged terminal activation duration in V1	Not performed	Extensive LV LGE, including near-transmural LGE of lateral wall and midwall of anterior wall.	Myocyte degeneration and fibrocytic infiltration within the posterolateral wall of the LV (extending transmurally).
Case 5	Saguner AM et al ⁵ <i>Circulation.</i> 2015	Fig. 2	LPFB Early repolarization in the inferior leads and QRS fragmentation in aVL	Heterozygous pathogenic variant in the plakophilin-2 (c.2392A>G, p.T798A) and desmoglein-2 (c.877A>G, p.I293V) genes.	Fibrofatty infiltration involving epi- and midmyocardial layers of the inferolateral, antero-lateral, and septal LV wall.	Unremarkable
Case 6	d'Amati G et al ⁶ <i>Int J Cardiol.</i> 2016	Fig. 1	LPFB	Not pathogenic mutation	Not performed	Fibro-adipose replacement (LV postero-lateral wall). Myocytes enlarged, dysmorphic nuclei
Case 7	Blom LJ et al ⁷	Fig. 1	LPFB - Intraventricular conduction delay.	Unclassified variant in the DSG2 gene and	Not performed	Normal

	<i>Heart Rhythm Case Rep.</i> 2018		1-point elevation in inferior leads	a p.Leu729del mutation in the gene SCN5A.		
Case 8	Blom LJ et al ⁷ <i>Heart Rhythm Case Rep.</i> 2018	Fig.2	LPFB TW1 V5-V6, I, II, aVF R/S ratio V1 ≥ 1	Unclassified variant plakophilin-2 gene and a pathogenic c.40-42delAGA mutation in the phospholamban (PLN) gene.	Not performed	Normal
Case 9	Singh SM et al ⁸ <i>JACC Case Rep.</i> 2021	Fig.3 (C)	LPFB - R/S ratio V1 ≥ 1	Not performed	Biventricular apical and anterolateral LGE in the epi- to mid-myocardium	Not performed
Case 10	Pirioni N et al ⁹ <i>ESC Heart Fail.</i> 2020	Fig.2 (B)	LPFB - TW1 V4-V6 LQRSV in limb leads	Pathogenic variant in desmoplakin c.3924del	High T2 intensity and subepicardial circumferential LGE.	Not performed
Case 11	Reichl K et al ¹⁰ <i>Circ Genom Precis Med.</i> 2018	Fig.1 (A)	LPFB	Heterozygous variant was identified in exon 23 of the DSP gene—c.3415_3417del TATinsG.	Epi- and midmyocardial LGE and fatty replacement in anterior, lateral and inferior LV and basal inferior RV segments.	Not performed
Case 12	Chmielewski P et al ¹¹ <i>Diagnostics</i> 2020	Fig.2 (A)	LPFB TW1 V1-V3 LQRSV, prolonged terminal activation duration in V1, rQRS II-III-aVF.	DSP NM_004415.4:c.3737dupA (p.Asn1246LysfsTer7) PKP2 NM_004572.3:c.2636T>C (p.Leu879Pro) NLRP3 NM_004895.4:c.1469G>A (p.Arg490Lys)	Moderate subepicardial and midwall areas of LGE with a ringlike pattern	
Case 13	Poller W et al ¹² <i>J Am Heart Assoc.</i> 2020	Fig.3 (C)	LPFB TW1 inferolateral leads	Dystrophin c.3970C>T, p.Arg1324Cys, desmoplakin c.4372C>T, p.Arg1458Ter, nexilin F-actin-binding protein c.154G>C,	Multifocal subepicardial posteroseptal and lateral LGE.	Low-level immune cell infiltration in the absence of intramyocardial virus genomes



The Diagnostic and Prognostic Value of the 12-Lead ECG in Arrhythmogenic Left Ventricular Cardiomyopathy



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The Diagnostic and Prognostic Value of the 12-Lead ECG in Arrhythmogenic Left Ventricular Cardiomyopathy

125 consecutive ALVC patients: 64 men, mean age 37 ± 15 years. 89 **probands (71%)**.
81.5% DSP and 18.5% non-DSP P/LP variants (PKP2 n=10; DSG2 n=8; JUP n=3; DSC2 n=1)

OBJECTIVES:



ECG findings in ALVC



The ECG correlation with CMR and genetic data

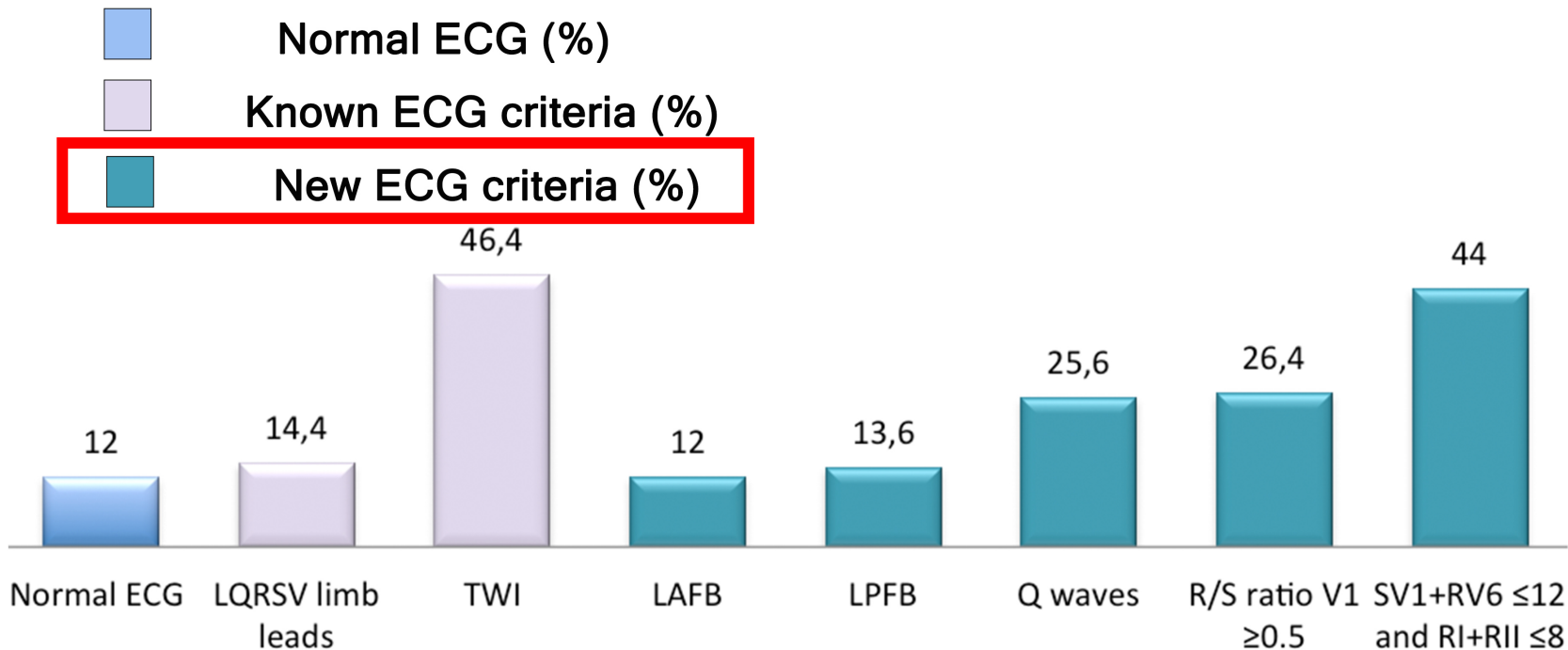
The prognostic value of ECG in ALVC

ALVC defined by the presence of the following criteria:

1. Presence of LGE in the LV, manifesting as a stria (or band) pattern affecting >1 segment.
2. Pathogenic/ likely pathogenic variants in genes responsible for desmosomal proteins associated with ALVC.
3. Confirmation of ALVC diagnosis through endomyocardial or at autopsy, for those cases in which genetic screening did not identify any pathogenic mutation



ECG analysis



In comparison with probands, **relatives had more often a normal ECG**



Follow-up

	Major Arrhythmic Events (n = 35)	No Major Arrhythmic Events (n = 90)	P Value
ECG			
Normal ECG	1 (2.9)	14 (15.6)	0.051
QRS (msec)	99 ± 17	95 ± 14	0.004
First degree AV block	4 (11.4)	6 (6.7)	0.32
NSICD	0	2 (2.2)	0.65
RBBB	2 (5.7)	2 (2.2)	0.15
LAFB	6 (17.1)	9 (10.0)	0.15
LPFB	10 (28.6)	7 (7.8)	<0.001
LBBB	1 (2.9)	0	0.11
Pathological Q waves	9 (25.7)	23 (25.6)	0.99
Lateral distribution	4 (11.4)	8 (8.9)	0.64
Inferior distribution	4 (11.4)	11 (12.2)	0.89
Precordial distribution	0	2 (2.2)	0.65
More 2 localizations	1 (2.9)	2 (2.2)	0.98
Fragmented QRS	12 (34.3)	34 (37.8)	0.56
Lateral distribution	2 (5.7)	4 (4.4)	0.52
Inferior distribution	6 (17.1)	22 (24.4)	0.22
Precordial distribution	1 (2.9)	1 (1.1)	0.31
More 2 localizations	3 (8.6)	7 (7.8)	0.80
Global LQRSV	6 (17.1)	6 (6.7)	0.091
LQRSV in limb leads	2 (5.7)	16 (17.8)	0.089
Local LQRSV			
Lateral distribution	8 (22.9)	21 (23.3)	0.89
Inferior distribution	5 (14.3)	14 (15.6)	0.84
Inferolateral distribution	2 (5.7)	3 (3.3)	0.31
Precordial and local distribution	2 (5.7)	10 (11.1)	0.20

	Major Arrhythmic Events (n = 35)	No Major Arrhythmic Events (n = 90)	P Value
QTc (msec)	411 ± 27	409 ± 24	0.92
QTc ≥ 440 ms	5 (14.3)	5 (5.6)	0.11
Tzou criteria ^b	7 (20.0)	12 (13.3)	0.18
R > 3 mm V ₁	4 (11.4)	6 (6.7)	0.32
R/S ratio ≥ 0.5 in V ₁	15 (42.9)	18 (20.0)	0.009
R/S ratio ≥ 1 in V ₁	8 (22.9)	7 (7.8)	0.021
Bayés de Luna criteria ^c	4 (11.4)	3 (3.3)	0.071
TWI	21 (60.0)	37 (41.1)	0.052
Inferolateral TWI	4 (11.4)	5 (5.6)	0.34
Anterior TWI	6 (17.1)	5 (5.6)	0.055
Inferior TWI	1 (2.9)	4 (4.4)	0.70
Lateral TWI	2 (5.7)	9 (10.0)	0.30
Anterolateral TWI	6 (17.1)	9 (10.0)	0.15
Inferior-anterior-lateral TWI	2 (5.7)	5 (5.6)	0.98
New ECG criteria			
SV ₁ + RV ₆ ≤ 12 and RI + RII ≤ 8 (mm)	22 (62.9)	33 (36.7)	0.008



Follow-up

FOLLOW-UP: Mean 57 Months, MAE in 35 (28%)

Independent Predictors of MAE

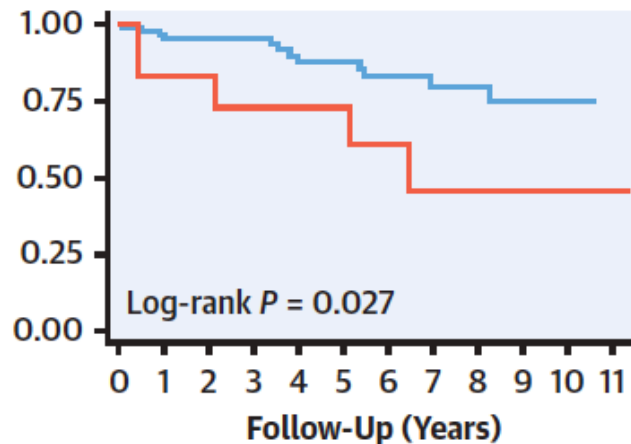
LPFB [OR: 4.7, 95% CI: 1.2-18.3]

Syncope [OR: 84.95, 95% CI: 14-496]

Transmural LGE [OR: 9.95, 95% CI: 2.3-36]

RVEF [OR: 0.92, 95% CI: 0.87-0.97]

Survival Free From MAE



Number at risk

— 96 81 70 57 43 37 28 21 18 15 10 7

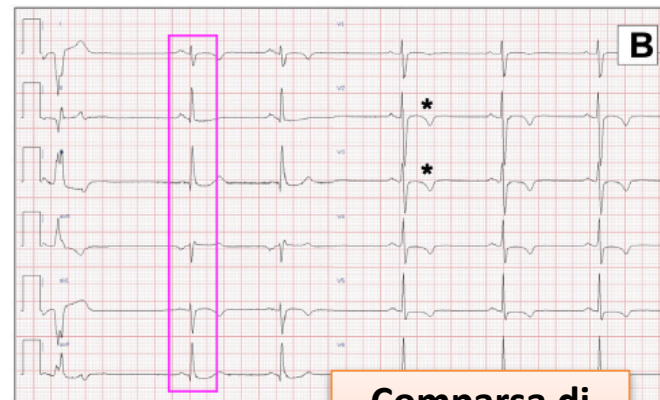
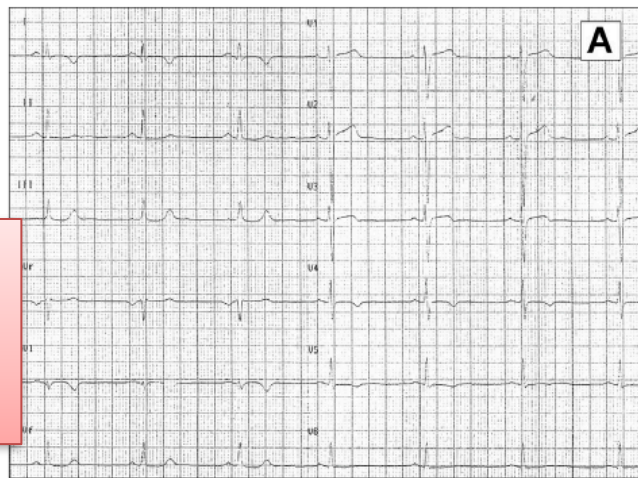
— 12 10 8 7 7 6 4 3 3 3 3 3

— No LPFB — LPFB

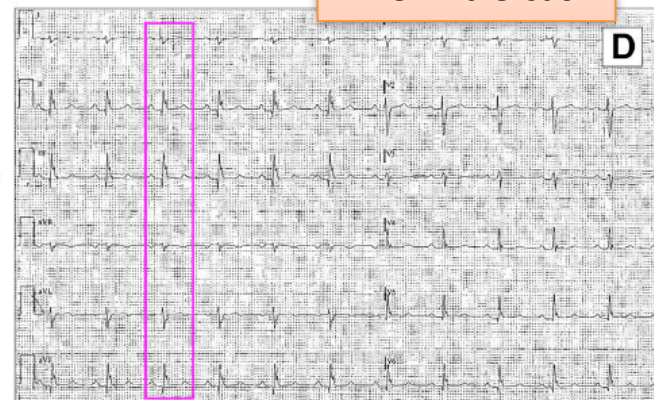
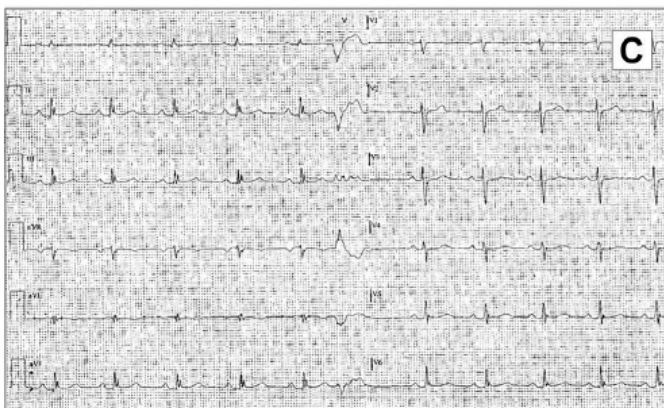


FIGURE 2 Electrocardiographic Changes During Follow-Up

FU medio 45 mesi,
cambiamenti ECG
nel 54,2% dei
pazienti



**Comparsa di
EPS in tre casi**



Conclusion

- Patients with ALVC exhibit distinct ECG characteristics. Some of these ECG signs are **new or little known** such as **pathological Q-waves, LPFB, a R/S ratio in V1 ≥ 0.5 , a sum of the R-wave in I-II ≤ 8 mm and S-wave in V1 plus R-wave in V6 ≤ 12 mm.**
- **Periodic ECG assessments** are necessary in order to evaluate significant ECG changes and to increase diagnostic suspicion of ALVC.
- A **normal ECG doesn't exclude ALVC diagnosis**. It could be more observed in relatives than in probands.
- Further studies are required to evaluate potential prognostic role of ECG in patients with ALVC

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