

ECG e cardiomiopatie

L. Calò, Roma

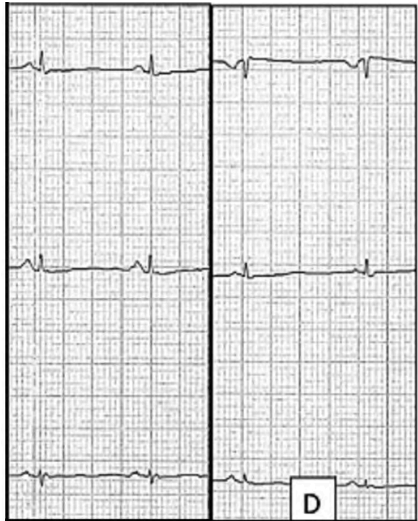
Università degli Studi di Roma «Foro Italico»

Cardiologia - Policlinico Casilino Roma



European Task Force criteria for diagnosis of Arrhythmogenic Cardiomyopathy

LV PHENOTYPE : ECG CRITERIA (3. e 4.)



Major

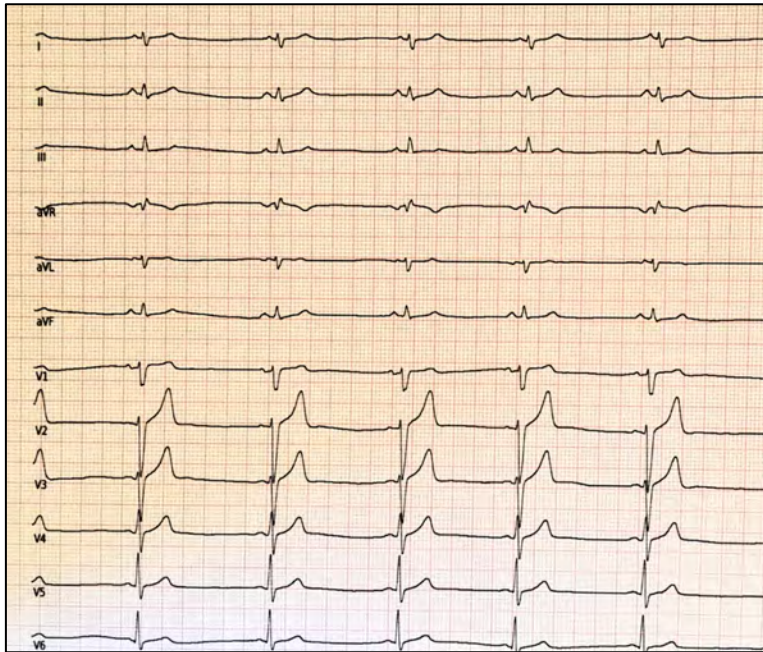
- Low QRS voltages (<0.5 mV peak to peak) in all limb leads in the absence of other causes (e.g. cardiac amyloidosis, obesity, emphysema, or pericardial effusion)



Minor

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

Low QRS voltages



Defined as QRS complexes with a **peak-to-peak amplitude <0.5 mV in all peripheral leads.**

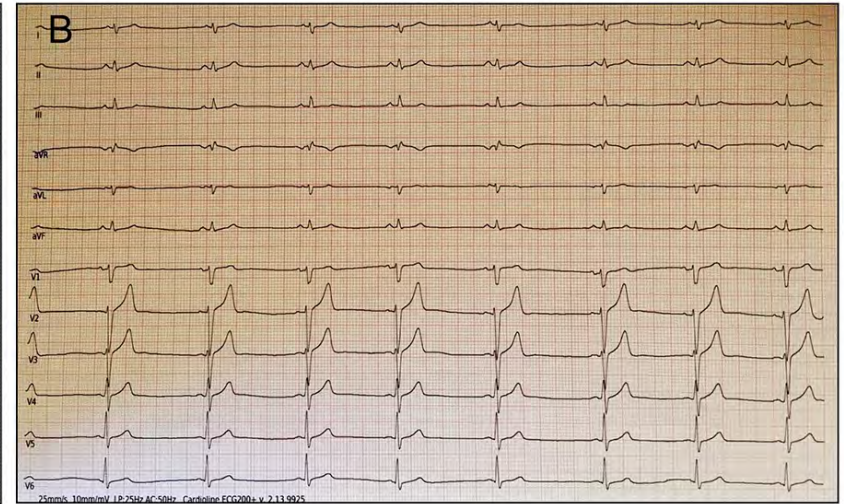
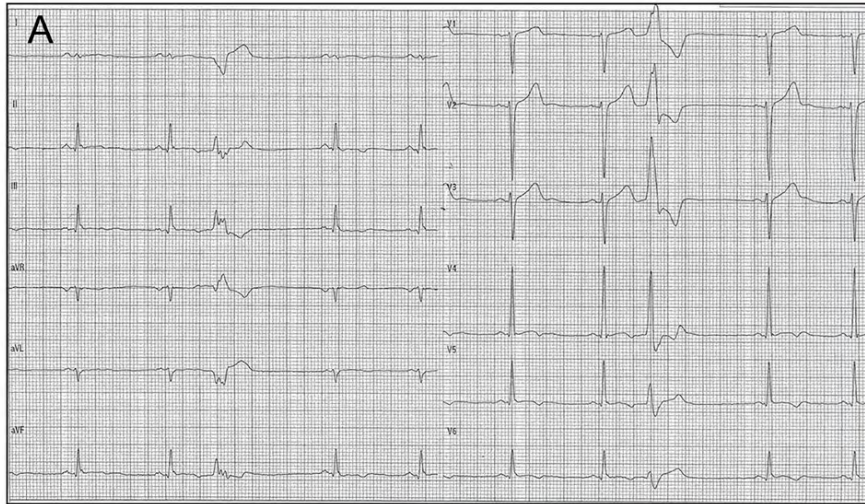
LQSV may be observed in pts with conditions that lead to a diseases characterized by loss of myocytes with replacement fibrosis (ACM, cardiac sarcoidosis, healed myocarditis, or idiopathic NILVS).

Low QRS voltages are characteristic of **FLNC, PLN, ?**  **DSP** and may precede any echocardiographic changes

Two brothers in comparison

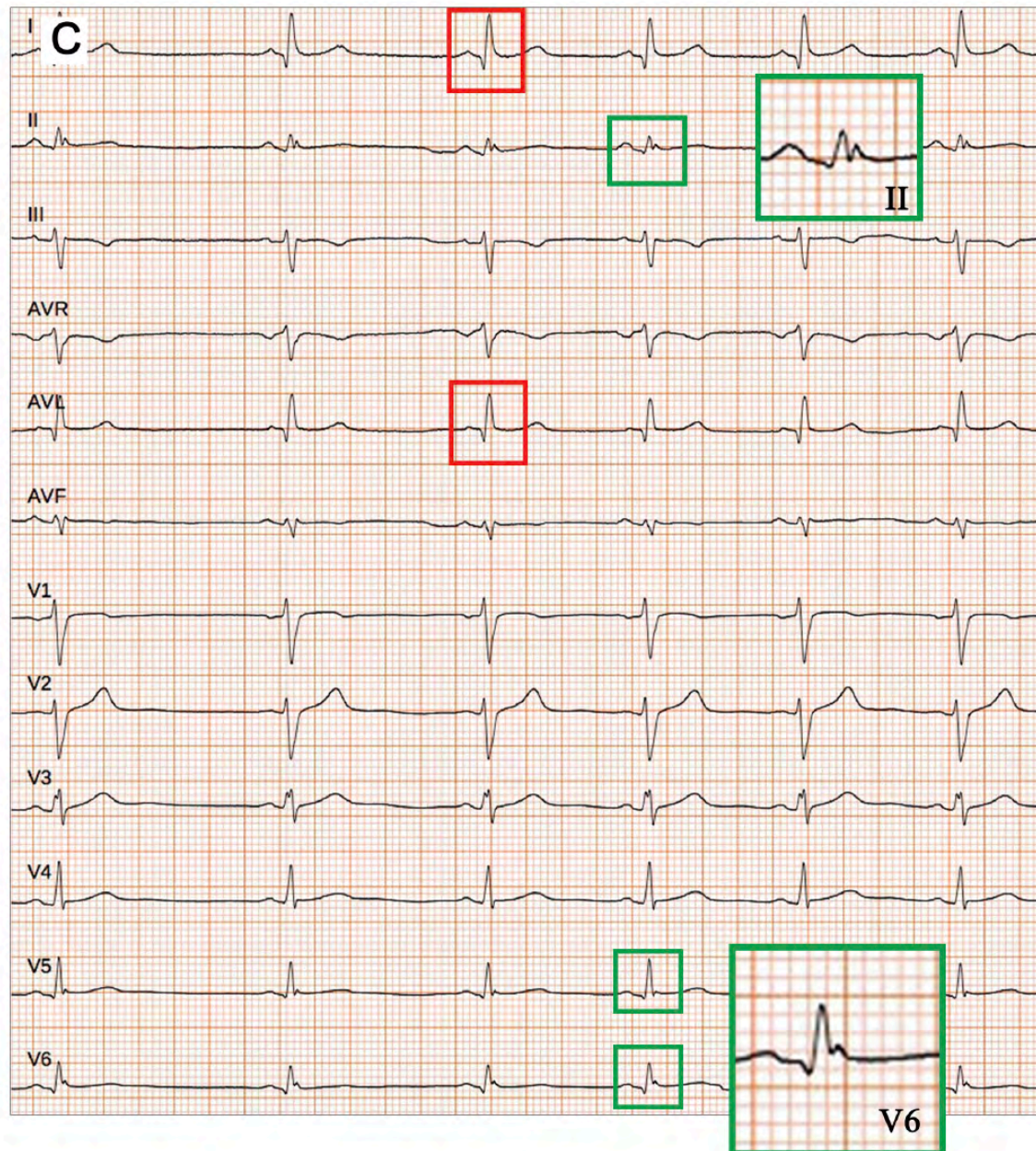
BROTHER

PATIENT 9



Pt 9 CMR reveals **subepicardial/intramycardial circumferential LGE**.

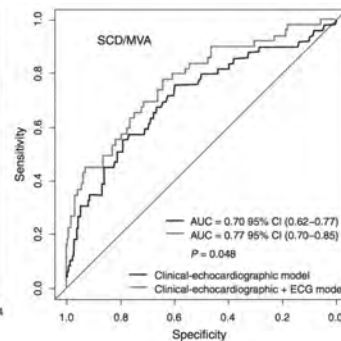
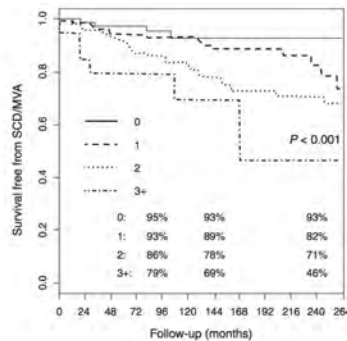
Autopsy of his brother shows the same **LV circumferential scar**, mostly located in the subepicardium, with gross features of fibroadipose myocardial replacement.



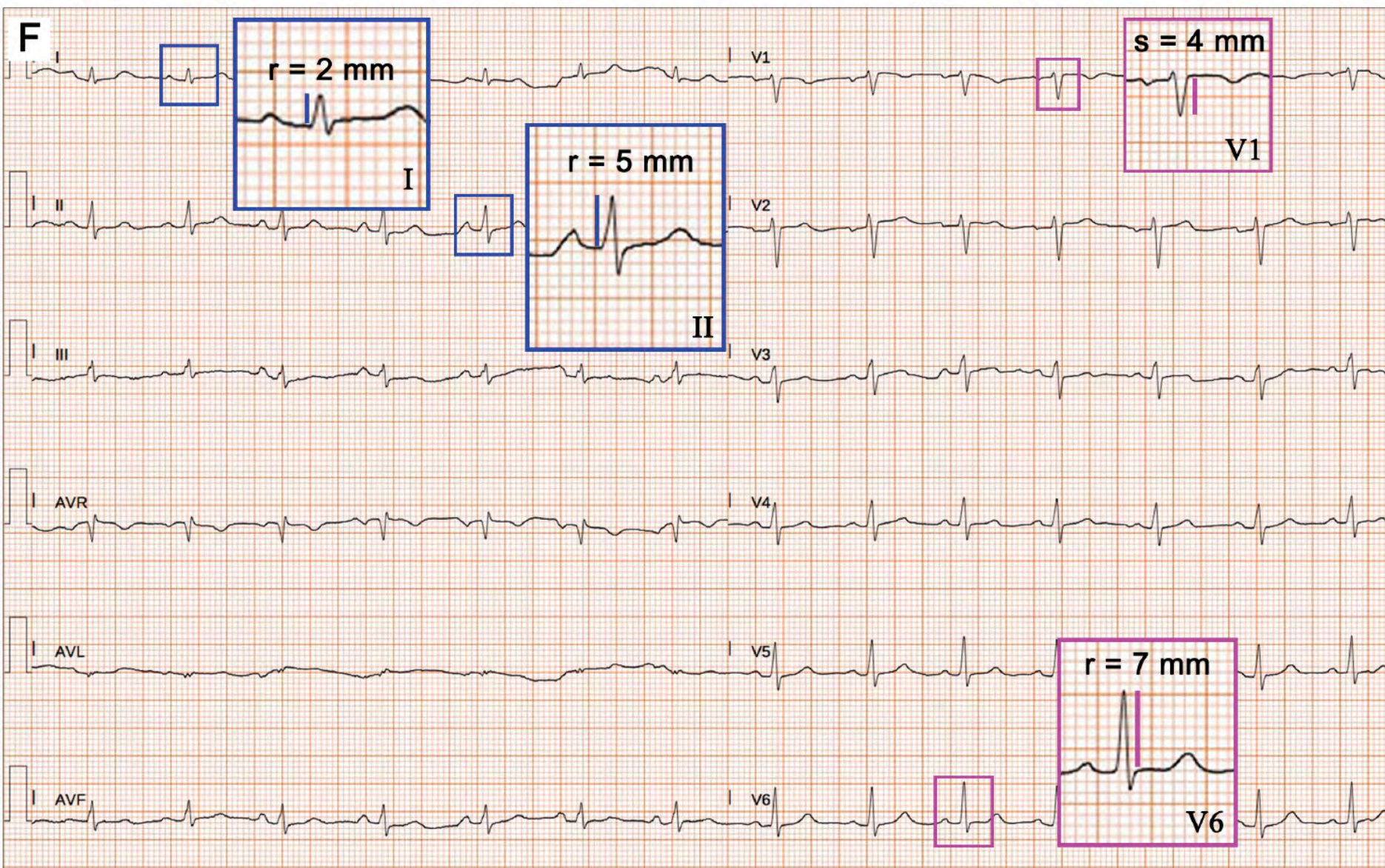
Patient #8, 20-year-old man, pathogenic variant in desmoplakin (c.5851 C>T, p.Arg1951Ter)

ECG in dilated cardiomyopathy: specific findings and long-term prognostic significance

Marco Merlo^a, Denise Zaffalon^a, Davide Stolfo^a, Alessandro Altinier^a, Giulia Barbatì^b, Massimo Zecchin^a, Stefano Bardari^a and Gianfranco Sinagra^a



- **14%** of pts **SD/MVA**. Predictors at multivariate analysis:
 - **inverted T-waves in anterolateral leads** (HR 2.141; 95% CI 1.148–3.991; $P = 0.017$)
 - **higher amplitude of S wave in V2** (HR 0.961; 95% CI 0.933 – 0.990; $P = 0.008$) and **of R wave on II** (HR 0.901; 95% CI 0.834–0.972; $P = 0.007$) **protective** factors.



Patient #20, 23-year-old woman with a pathogenic variant in desmoplakin (c.5210del, p.Gly1737AspfsTer16)

Sinus rhythm QRS amplitude and fractionation in patients with nonischemic cardiomyopathy to identify ventricular tachycardia substrate and location



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BACKGROUND Ventricular tachycardia (VT) substrate in left ventricular (LV) nonischemic cardiomyopathy (NICM) consists of fibrosis with surviving myocardium.

OBJECTIVE The purpose of this study was to determine whether, in patients with LV NICM and sustained VT, reduced QRS amplitude and QRSf during sinus rhythm can identify the presence and location of abnormal septal (S-NICM) and/or free-wall (FW-NICM) VT substrate.

METHODS We compared patients with NICM and VT (group 1) with electroanatomic mapping septal (S-NICM; $n = 21$) or free-wall (FW-NICM; $n = 20$) VT substrate to a 38-patient reference cohort (group 2) with cardiac magnetic resonance imaging (cMRI) and NICM but no VT referred for primary prevention implantable cardioverter-defibrillator (26 [68.4%] with late gadolinium enhancement).

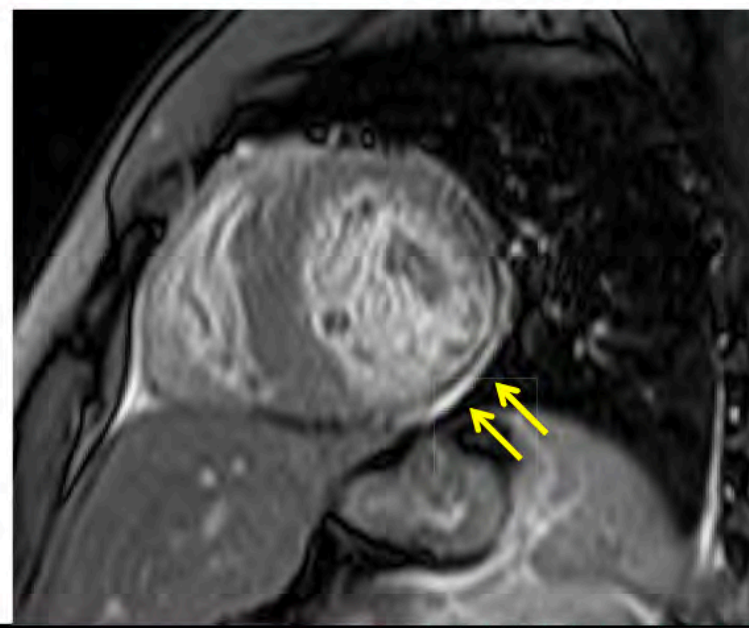
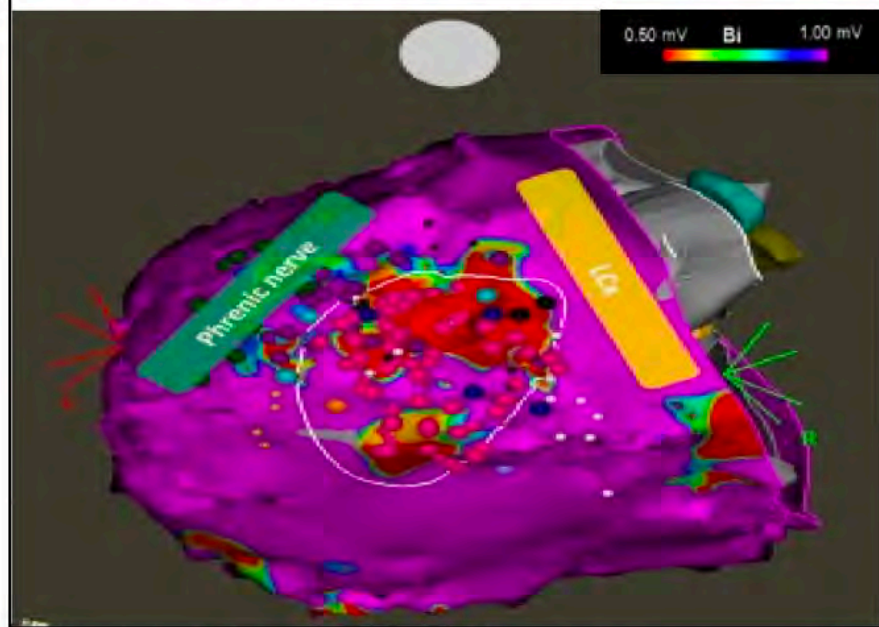
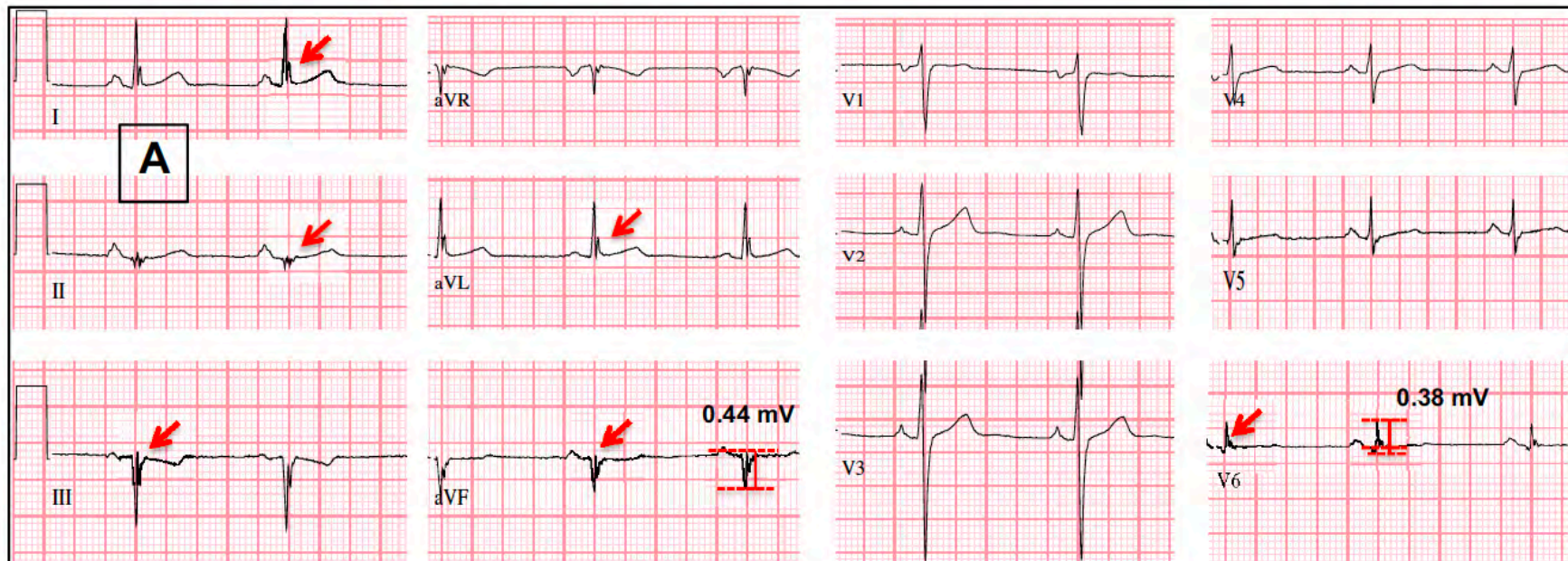
RESULTS Group 1 had lower QRS amplitude in leads II (0.60 ± 0.22 vs 0.86 ± 0.35 , $P < .001$), aVR (0.60 ± 0.24 vs 0.75 ± 0.31 , $P = .002$), aVF (0.48 ± 0.20 vs 0.70 ± 0.28 , $P < .001$), and V_2 (1.09 ± 0.52 vs 1.38 ± 0.55 , $P = .001$) than group 2. QRS

<0.55 mV in lead aVF identified VT and accompanying substrate with sensitivity 70% and specificity 71%. Most group 1 and group 2 patients had 12-lead ECG QRS fractionation (QRSf) in ≥ 2 contiguous leads (78% vs 63.2%, $P = .14$). Sensitivity and specificity for ≥ 2 QRSf leads identifying respective regional electroanatomic or cMRI abnormalities were 76% and 50% for inferior, 44% and 87% for lateral, and 21% and 89% for anterior leads.

CONCLUSION In LV NICM, low frontal plane QRS (<0.55 mV in aVF) is associated with VT substrate. Although multilead QRS fractionation is associated with the presence and location of VT substrate, it is frequently identified in patients without VT with cMRI abnormalities.

KEYWORDS Nonischemic cardiomyopathy; QRS amplitude; QRS fractionation; Sinus rhythm; Ventricular tachycardia

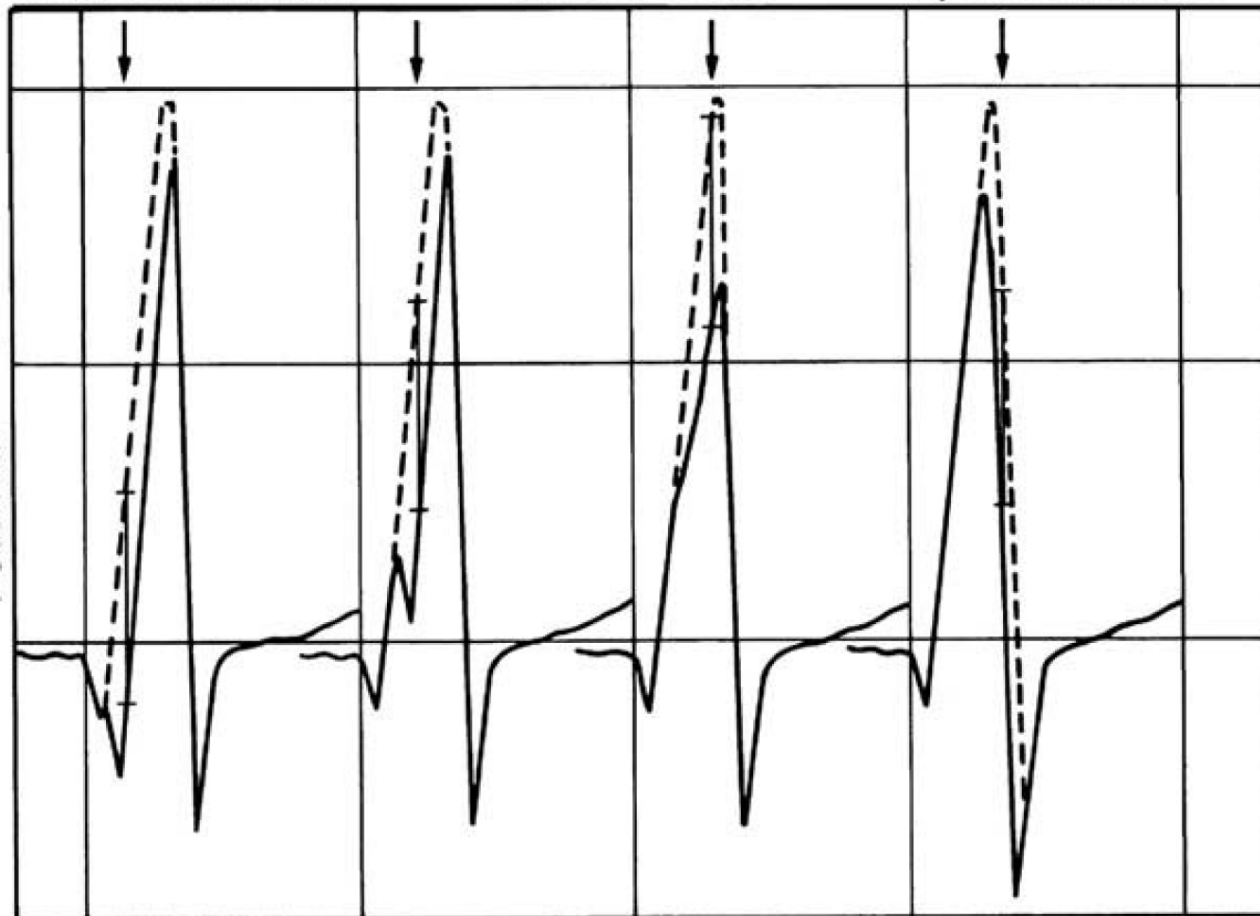
(Heart Rhythm 2022;19:187–194) © 2021 Heart Rhythm Society. All rights reserved.

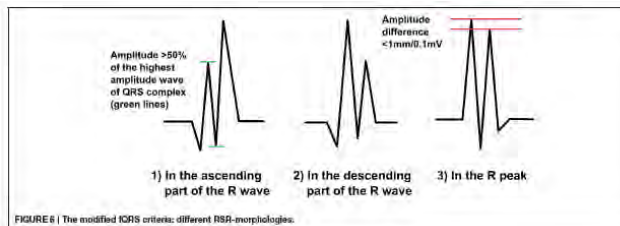
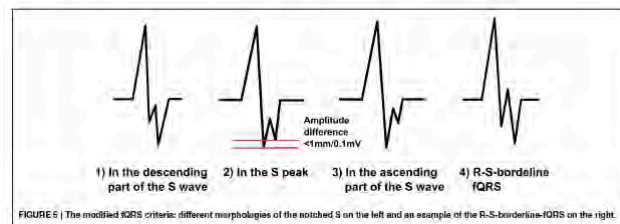
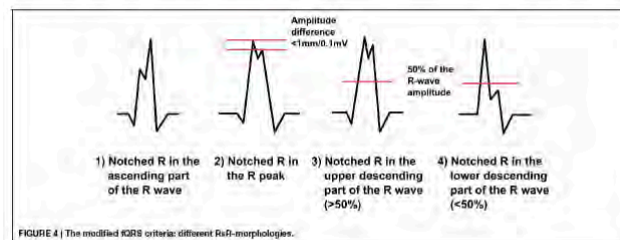
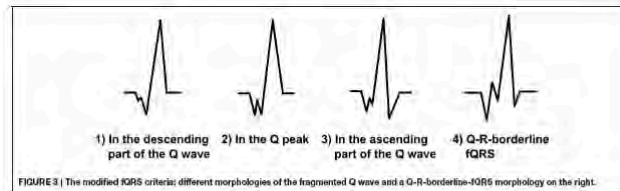


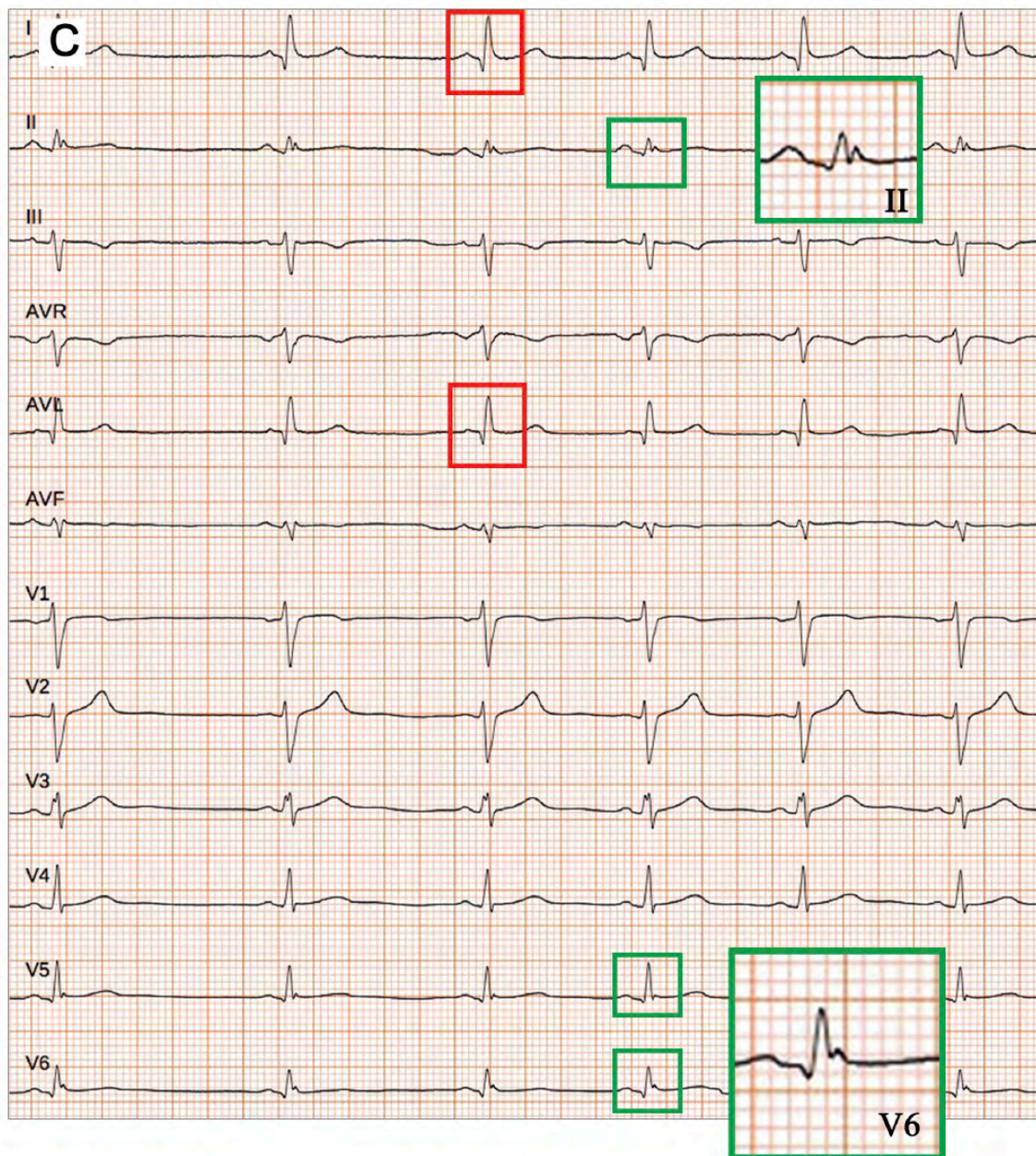
Endocardial Infarct

Midwall Infarct

Epicardial Infarct







The J wave and fragmented QRS complexes in inferior leads associated with sudden cardiac death in patients with chronic heart failure

Juanhui Pei¹, Ning Li¹, Yonghong Gao², Zengwu Wang¹, Xian Li¹, Yinhui Zhang¹, Jingzhou Chen¹, Ping Zhang^{3†}, Kejiang Cao^{4†}, and Jieli Pu^{1*}

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Aims

To investigate the relationship between electrocardiogram (ECG) parameters [J wave, fragmented QRS (fQRS), QTc, the peak-to-end interval of T wave (Tp–Te)], and sudden cardiac death (SCD) in chronic heart failure (CHF).

Methods and results

The ECGs of 1570 CHF patients, 572 cases with dilated cardiomyopathy (DCM) and 998 cases with ischaemic cardiomyopathy (ICM) were analysed with the endpoint being an SCD or non-SCD (NSCD). During a median follow-up period of 36 months (0.40–65 months), 438 (27.89%) patients died, of which 158 (35.84%) were SCD. Overall, the occurrence of J wave, fQRS, and long Tp–Te were greater in SCD patients than that of NSCD patients (all $P < 0.01$). For DCM cases, more SCD patients had J waves observed in the inferior leads than that in the NSCD group (26.78 vs. 13.07%, $P < 0.001$). However, ICM cases with SCD did have more fQRS in the inferior leads than that with NSCD (42.16 vs. 26.67%, $P = 0.01$). After adjusting for other risk factors, Cox regression analysis revealed that presence of J wave or fQRS in the inferior leads predicted a higher risk for SCD in DCM [hazard ratio (HR), 4.095; 95% confidence interval (CI), 2.132–7.863] and ICM (HR, 2.714; 95% CI, 1.809–4.072) patients. A left ventricular ejection fraction $\leq 30\%$ also predicted SCD and NSCD in DCM and ICM patients. In contrast, the predictive value of QTc and Tp–Te for SCD was not significant.

Conclusions

Presence of J wave or fQRS in the inferior leads predicted higher risk of SCD in DCM and ICM patients and might serve as independent predictors for SCD in patients with CHF.

Keywords

J wave • fQRS • Sudden cardiac death • Chronic heart failure • Tp–Te

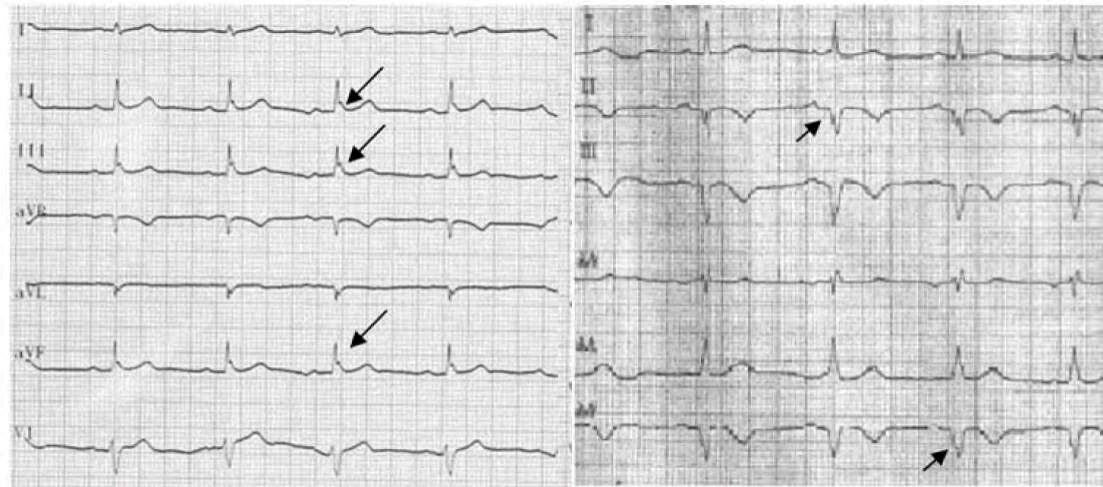
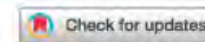


Figure 1 Patient with prominent J waves in inferior leads (left arrow) and fragmented QRS complex in inferior leads (right arrow).

CONTEMPORARY REVIEW

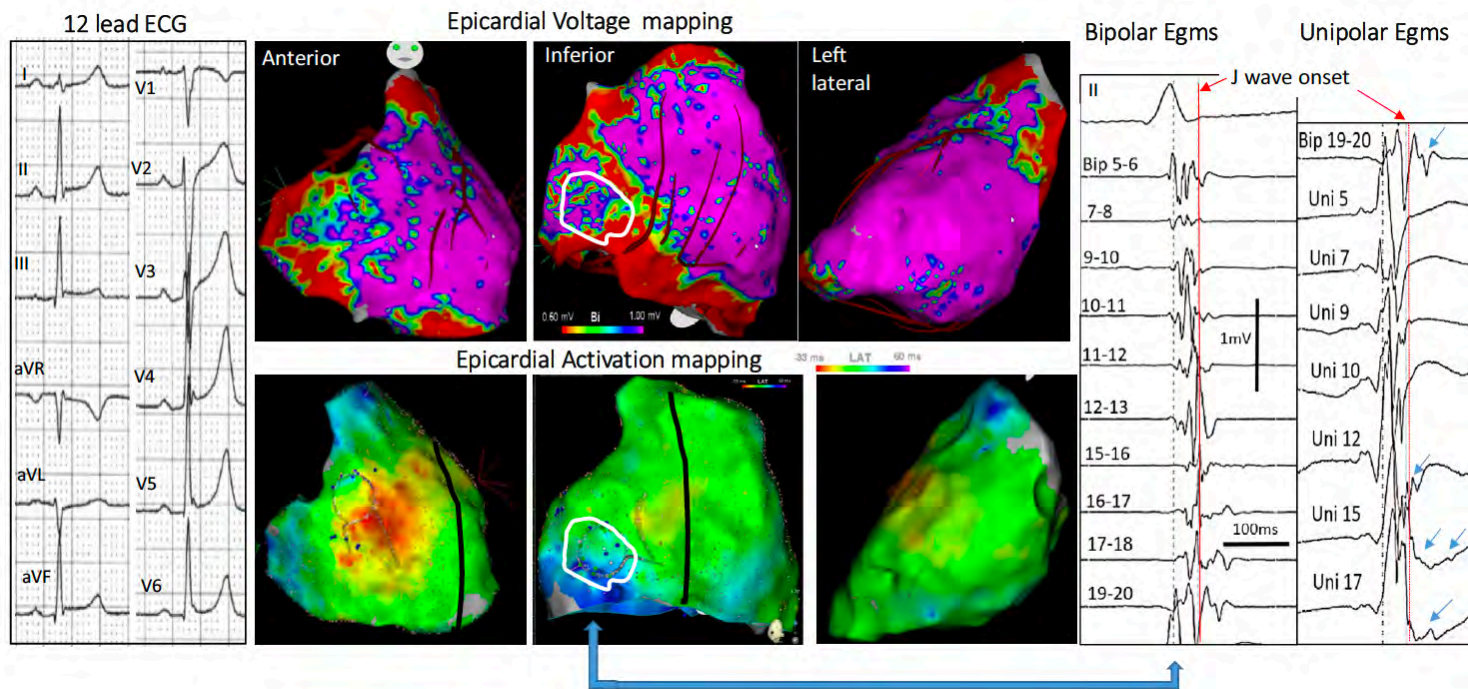
Depolarization versus repolarization abnormality underlying inferolateral J-wave syndromes: New concepts in sudden cardiac death with apparently normal hearts



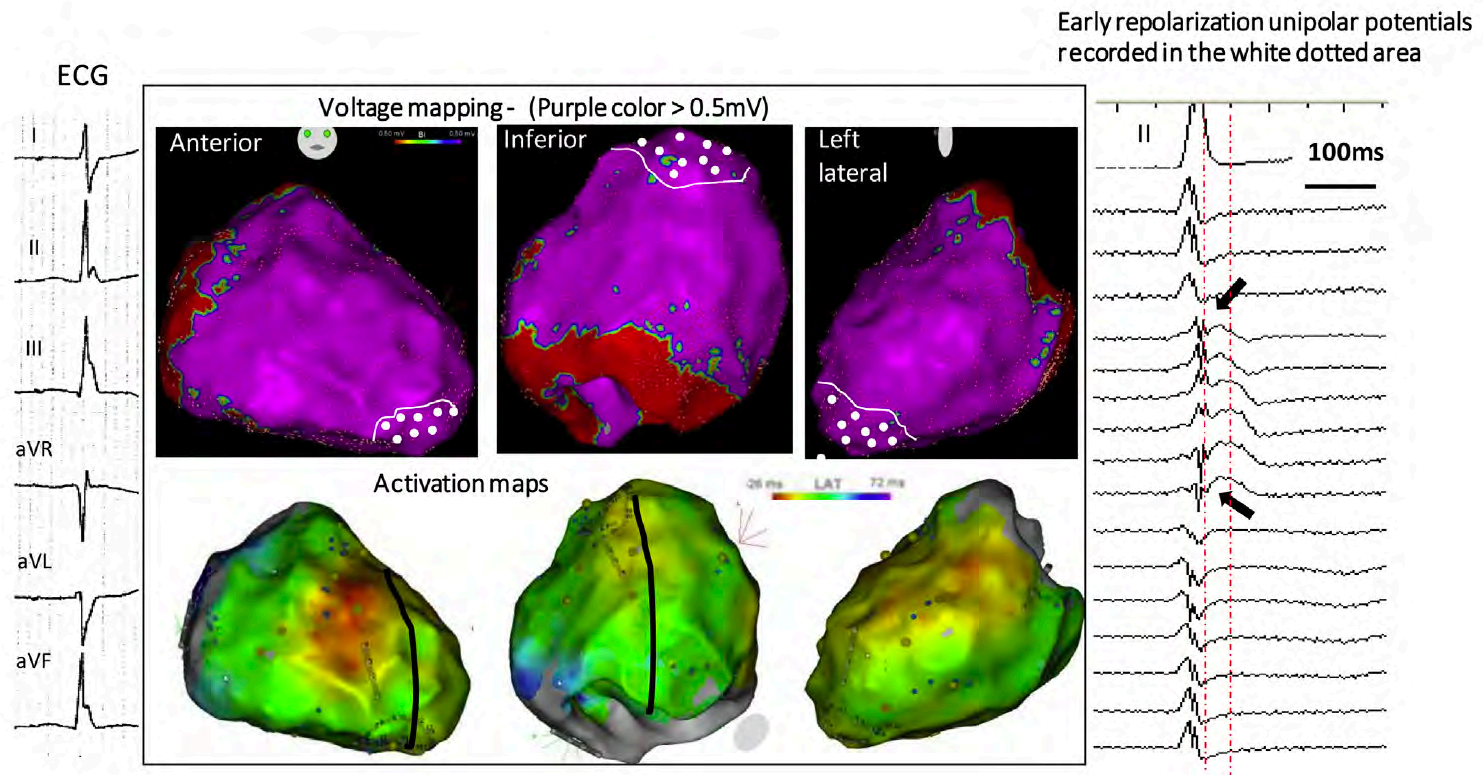
Michel Haïssaguerre, MD,^{*†‡1} Koonlawee Nademanee, MD,^{§1} Méléze Hocini, MD,^{*†‡} Ghassen Cheniti, MD,^{*} Josselin Duchateau, MD,^{*†‡} Antonio Frontera, MD,^{*} Frédéric Sacher, MD,^{*†‡} Nicolas Derval, MD,^{*†‡} Arnaud Denis, MD,^{*†‡} Thomas Pambrun, MD,^{*†‡} Rémi Dubois, PhD,^{†‡} Pierre Jaïs, MD,^{*†‡} David Benoist, PhD,^{†‡} Richard D. Walton, PhD,^{†‡} Akihiko Nogami, MD,^{||} Ruben Coronel, MD, PhD,[†] Mark Potse, PhD,[†] Olivier Bernus, PhD^{†‡}

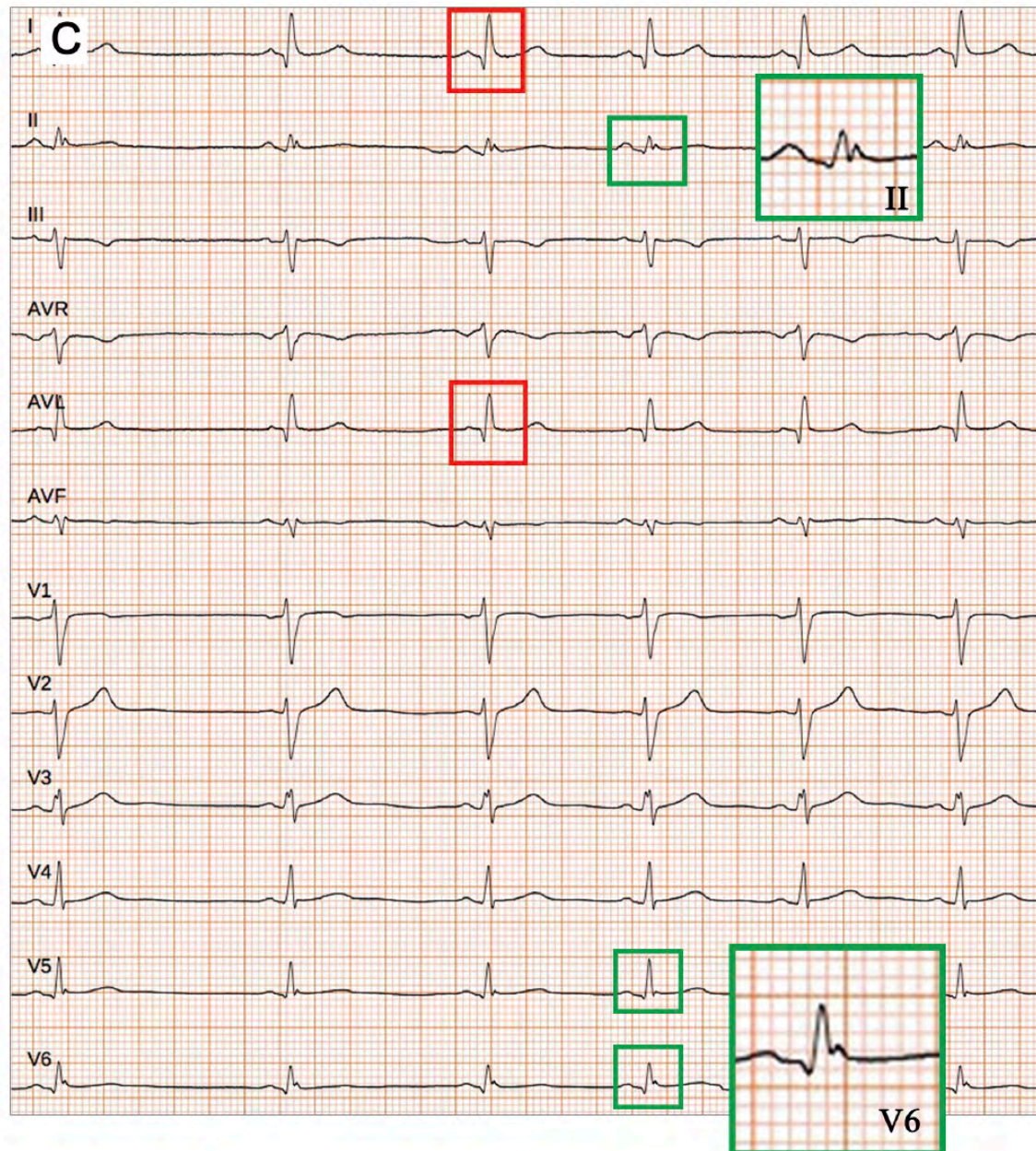
From the ^{}Bordeaux University Hospital, Bordeaux, France, [†]IHU LIRYC, Electrophysiology and Heart Modeling Institute, Bordeaux, France, [‡]University of Bordeaux, U1045, Bordeaux, France, [§]Bumrungrad Hospital, Bangkok, Thailand, and ^{||}University of Tsukuba, Ibaraki, Japan.*

Inferolateral J-wave syndrome due to abnormal depolarization

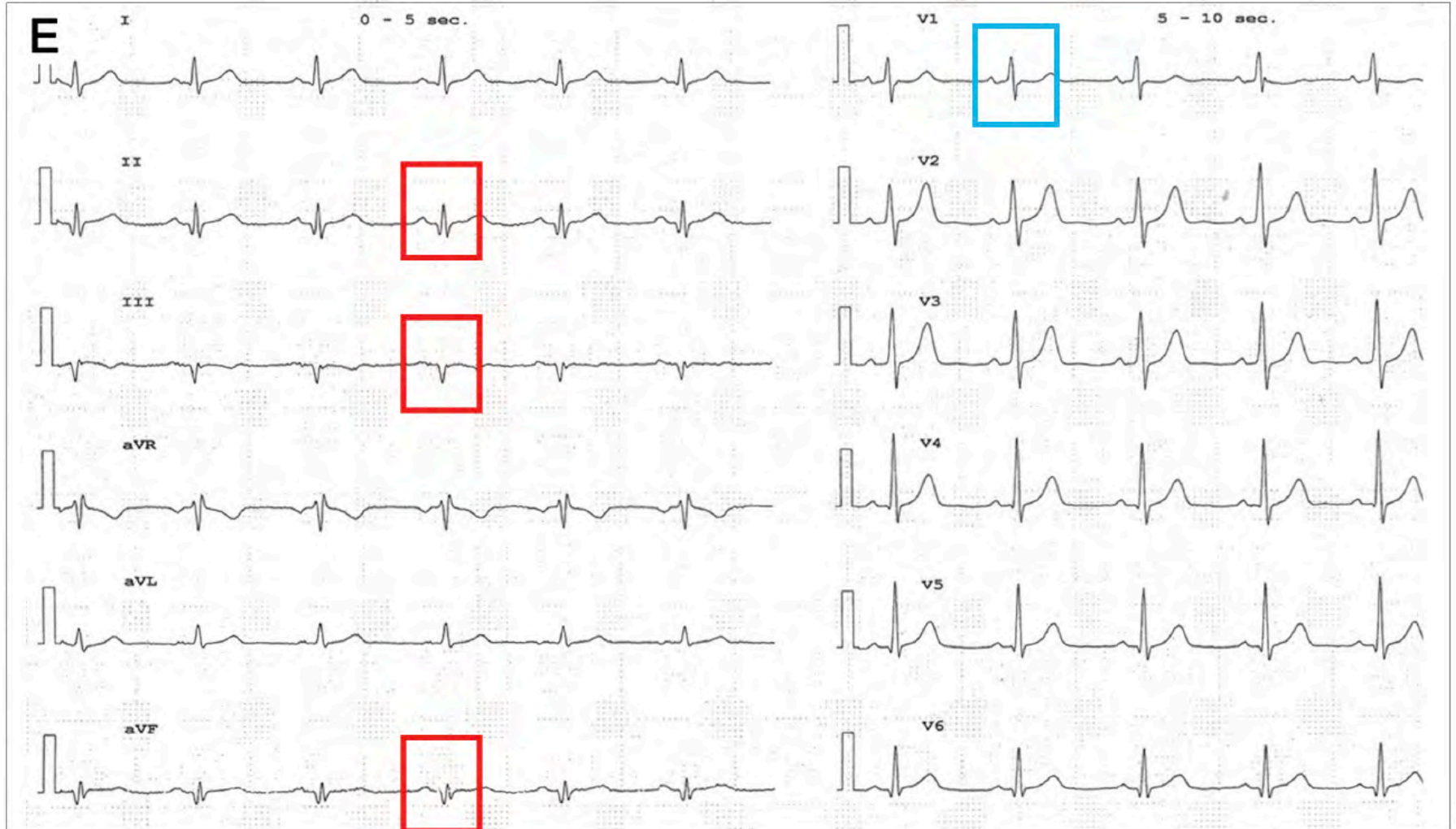


Inferolateral J-wave syndrome due to early repolarization





Patient #8, 20-year-old man, pathogenic variant in desmoplakin (c.5851 C>T, p.Arg1951Ter)



The family member (Patient #36, 37-year-old man) has the same pathogenic variant in desmoglein-2 (c.1912G>A, p.Gly638Arg)

The Distinctive Electrocardiogram of Duchenne's Progressive Muscular Dystrophy*

An Electrocardiographic-Pathologic Correlative Study

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The clinical, electrocardiographic, hemodynamic and cardiac pathologic features are described in two patients with the rapidly progressive, pseudohypertrophic, sex-linked form of Duchenne's muscular dystrophy. Previous studies have shown that the electrocardiogram is distinctive enough to aid in identifying this particular type of heredofamilial myopathic disorder even when the clinical features of the systemic myopathy are inconclusive. Tall right precordial R waves and deep limb lead and left precordial Q waves characterize these electrocardiograms. Information thus far has not provided explanations for either the distinctive morphology of the Duchenne electrocardiogram or for the variety of rhythm disturbances that have been observed. Accordingly, an electrocardiographic-pathologic correlative study was undertaken in order to determine whether a relationship existed between the distribution of myocardial dystrophy, the presence of an unusual type of small vessel coronary arteriopathy, and the electrocardiographic abnormalities of both QRS configuration and rhythm.

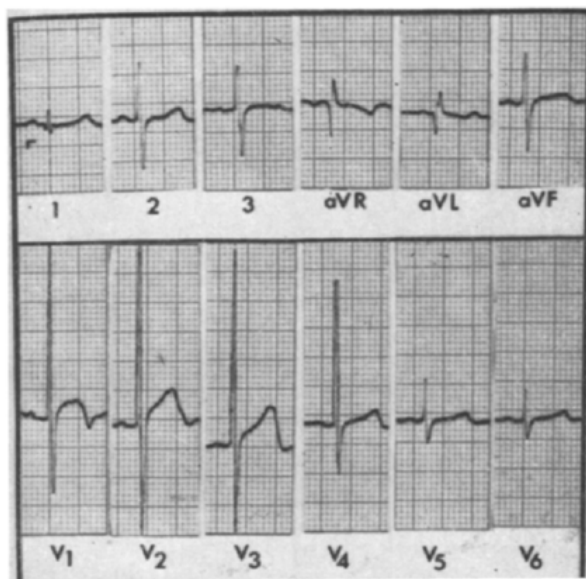


FIG. 1A. Case 1. Electrocardiogram obtained at twelve years of age shows a marked increase in the amplitude of the R wave in lead V₁, but a significant Q wave only in lead aVL.

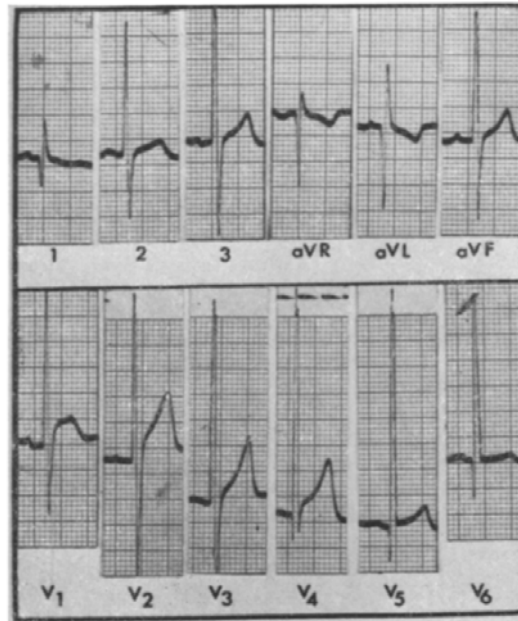


FIG. 1B. Case 1. Electrocardiogram obtained at seventeen years of age (similar to subsequent tracings except for preterminal atrial flutter). The tall R wave in lead V₁ persists. Deep Q waves have now appeared in leads I, aVL, and V₅ to V₆. The amplitude of the R waves in leads V₄ to V₆ has substantially increased.

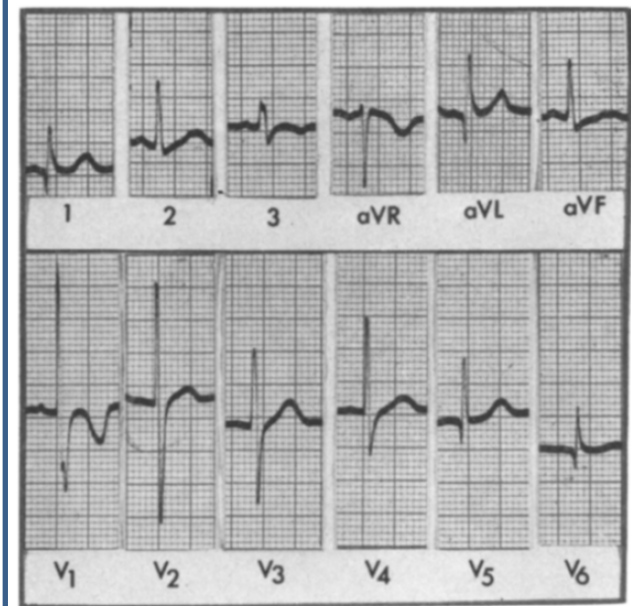
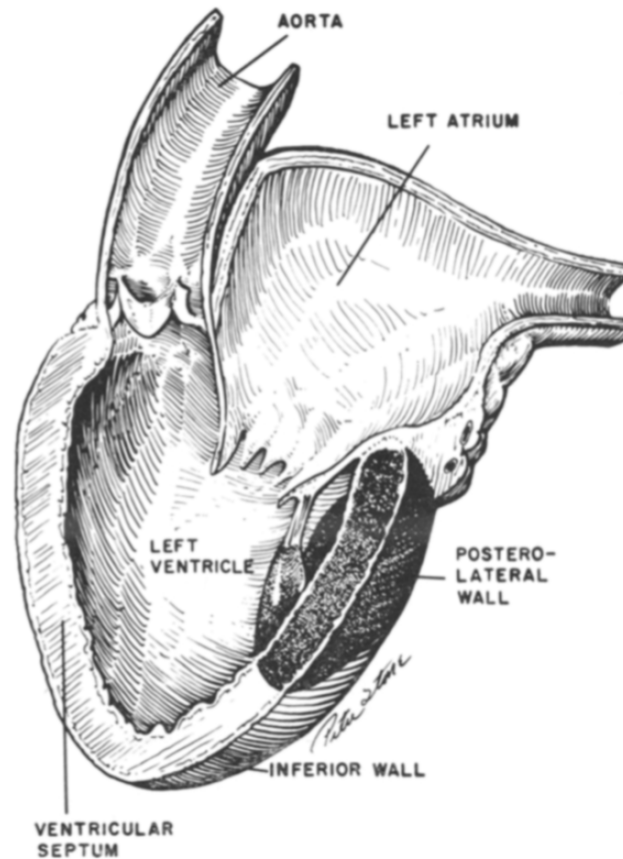
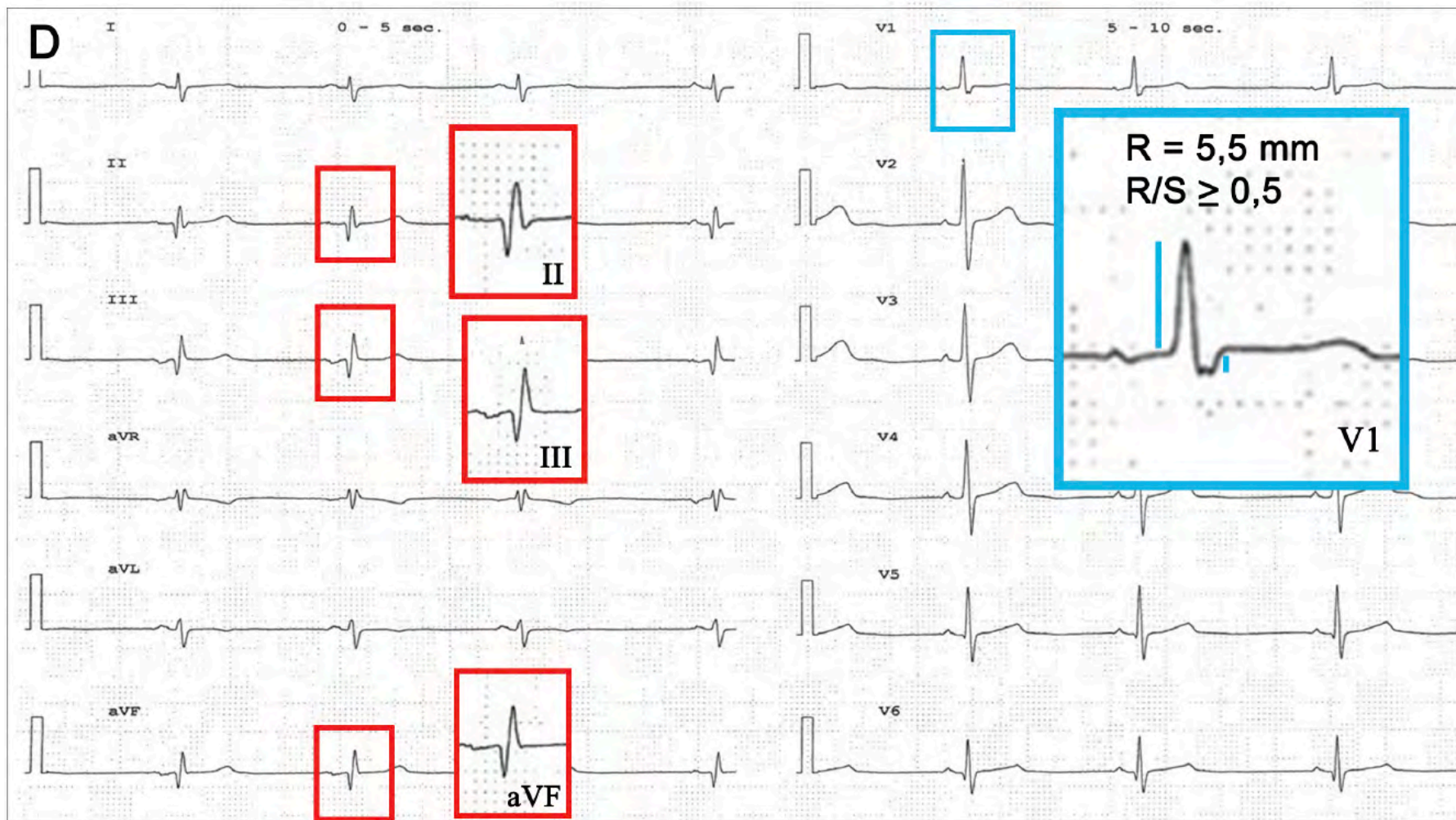


FIG. 2. Case 2. Electrocardiogram obtained at twelve years of age shows a tall R wave in lead V₁ in addition to deep Q waves in leads I and aVL. Leads V₅ to V₆ display moderately deep Q waves that were not present at nine years of age.

FIG. 3. Diagram of the left side of the heart illustrating the distribution of the fibrous scarring in the two patients. The scarring was focal and limited to the posterolateral free wall of the left ventricle. No scarring was observed in the diaphragmatic or inferior free wall or in the ventricular septum. A papillary muscle was involved in one patient (Case 1) who had clinical mitral incompetence.





Patient #35, 42-year-old man), pathogenic variant in desmoglein-2 (c.1912G>A, p.Gly638Arg)

The end of an electrocardiographic dogma: a prominent R wave in V₁ is caused by a lateral not posterior myocardial infarction—new evidence based on contrast-enhanced cardiac magnetic resonance—electrocardiogram correlations

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 Francesc Carreras¹, Diego Goldwasser¹, and Raymond J. Kim⁴**

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Received 6 February 2014; revised 16 October 2014; accepted 6 January 2015



Figure 1 Original drawing of true posterior infarction with the QRS morphology according to Perloff.¹

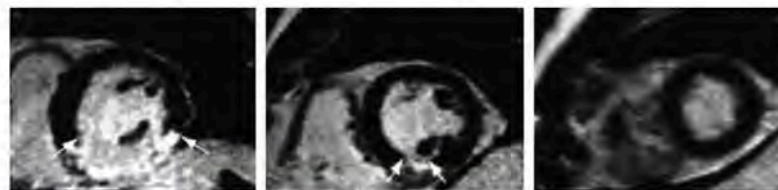
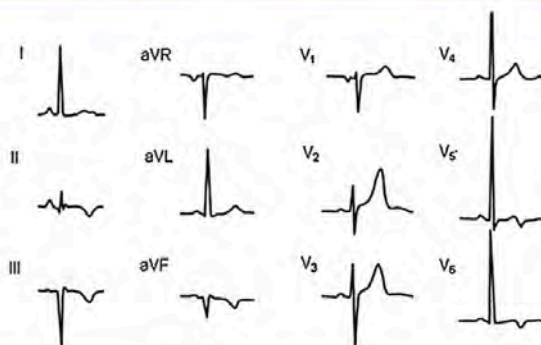


Figure 2 Electrocardiographic and cardiac magnetic resonance images of an inferior infarct. Despite the clear infero-basal location of infarction at contrast-enhanced cardiac magnetic resonance (left-hand panel, between the white arrows), lead V₁ does not show a prominent R wave but an rS morphology.

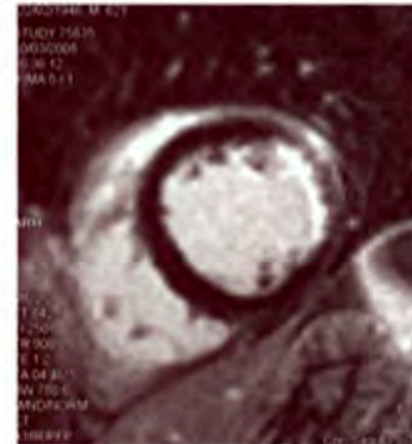
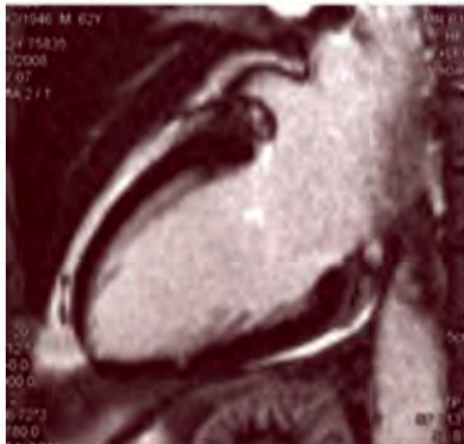
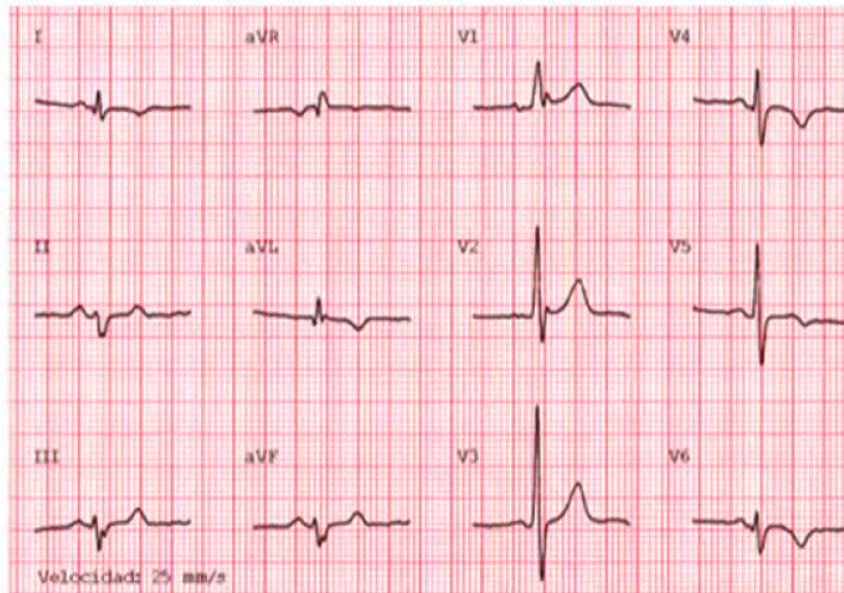


Figure 3 Electrocardiographic and cardiac magnetic resonance images of a lateral infarct. A tall R wave in V_1 corresponds to a lateral infarct at contrast-enhanced cardiac magnetic resonance (lower central and right-hand panels). Of note, the infero-basal segment (segment 4) does not present any sign of necrosis (lower left and right panels).

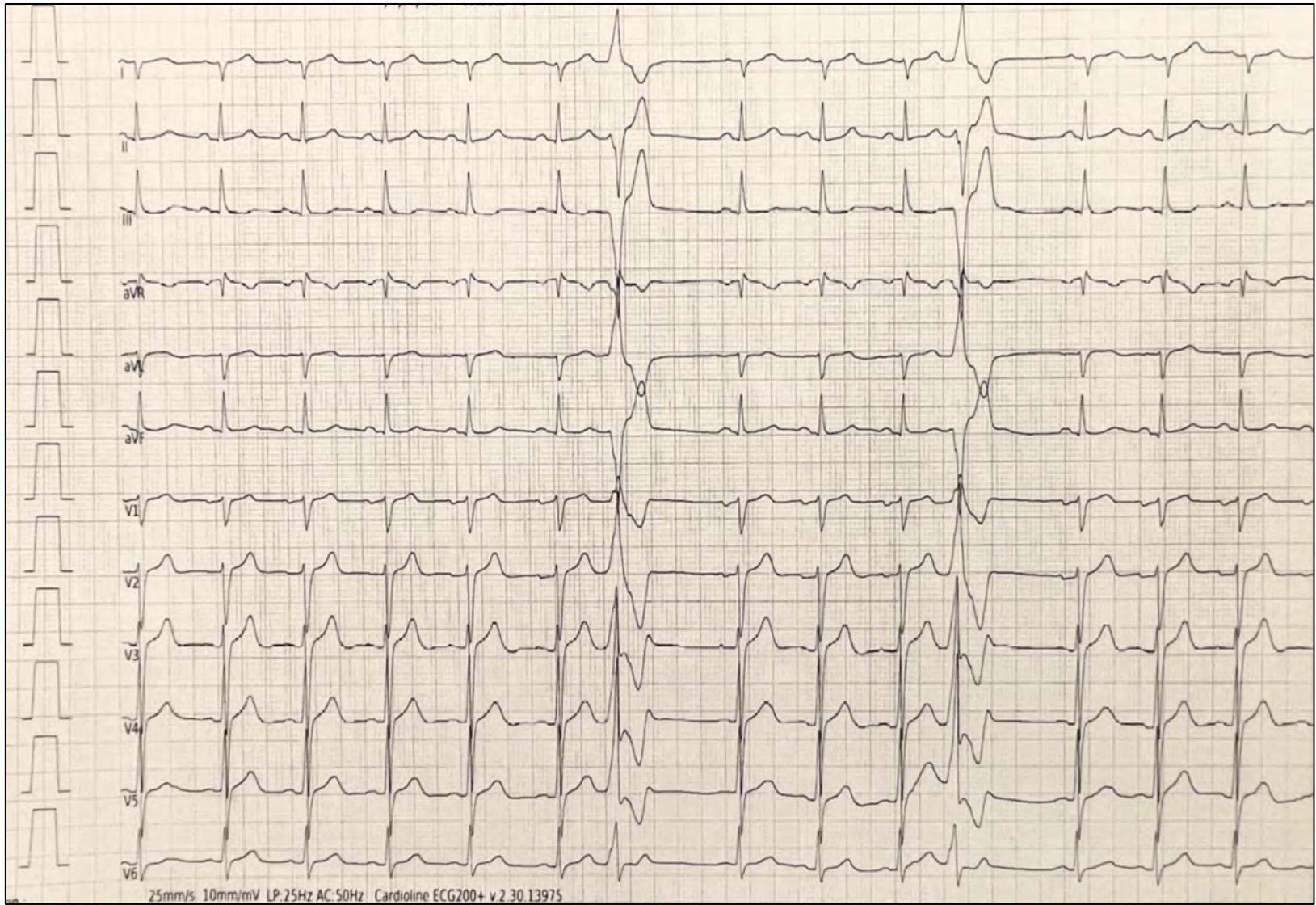
Unrecognized cases of prominent R-wave in V1 detected in the iconography of published papers

Case ID	References	Figure	12 lead ECG findings	Genetic analysis	CMR data	Endomyocardial biopsy data/ Histologic data
Case 1	Rubino M et al ¹ <i>Genes</i> 2021	Fig.1	Prominent R V1 (>3mm) Inferolateral TWI Pathological Q in I-aVL	DSP (c.5428C>T, p.Gln1810Ter)	Subepicardial circumferential LGE involving the entire LV	Not available
Case 2	Zorzi A et al ² <i>Circ Arrhythm Electrophysiol.</i> 2016	Fig. 4	LPFB Inferolateral TWI Pathological Q II-III-aVF Prominent R V1 (>3 mm) LQRSV in left precordial leads	Not performed	Sub/midmyocardial LGE with a stria 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 dysmetric 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	Sakamoto N et al ⁴ <i>Circ Cardiovasc Imaging.</i> 2019	Fig.1	R/S ratio V1 ≥1 Inferolateral TWI LQRSV in limb leads	DSP (c.4650delTG, p.V1551E fs74X) and MYBPC3 (c.2459G>A, p.R820Q)	LGE in the mid-myocardial septum and subepicardial anterolateral LV myocardium.	Fibrofatty replacement, mild hypertrophy, and disarrangement of the myocytes. Electron microscopy of the intercalated discs showed disarrangement of the filaments and widening of the fascia adherens gap
Case 5	Tsuruta Y et al ⁵ <i>Heart Fail.</i> 2020	Fig.1	R/S ratio V1 ≥1 Inferolateral TWI LQRSV in limb leads	Nonsense mutation in DSP (c.5212C>T, p.R1738*)	Fat signals LGE in the mid-wall to subepicardial layers in the LV myocardium	Moderate fibrofatty replacement and mild hypertrophy
Case 6	Groeneweg JA et al ⁶ <i>Heart Rhythm.</i> 2013	Fig.4	R/S ratio V1 ≥1 TWI in II and anterolateral leads	PKP2 variant c.419C4T and the PLN mutation c.40_42delAGA	LGE in the lateral wall of the LV	Normal myocardium, with locally some (<10%) subendocardial fibrosis.

Case 7	Blom LJ et al ⁷ <i>Heart Rhythm Case Rep.</i> 2018	Fig.2	LPFB TWI V5-V6, I, II, aVF R/S ratio V1 ≥1	Unclassified variant PKP2 gene and a pathogenic c.4042delAGA mutation in the PLN gene.	Not performed	Normal
Case 8	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 9	Norman M et al ⁹ <i>Circulation</i> 2005	Fig.3	R/S ratio V1 ≥0.5 Inferolateral TWI	DSP gene identified insertion of a single adenine base (2034insA)	Not performed	Not performed
Case 10	Chen P et al ¹⁰ <i>Int J Cardiol.</i> 2020	Fig. 2 (A)	LPFB. TWI V1-V3. R/S ratio V1 ≥1	DSG2 p.Leu237Ter mutation	Dilation of both ventricles	Not performed
Case 11	Pilichou K et al ¹¹ <i>Circulation</i> 2014	Fig.3	R/S ratio V1 ≥1	DSP c.448C>T mutation	Focal bulging on anterolateral RV apex. LGE (midepicardial stria) in the inferior LV wall	Not performed

Caso 4. Marco

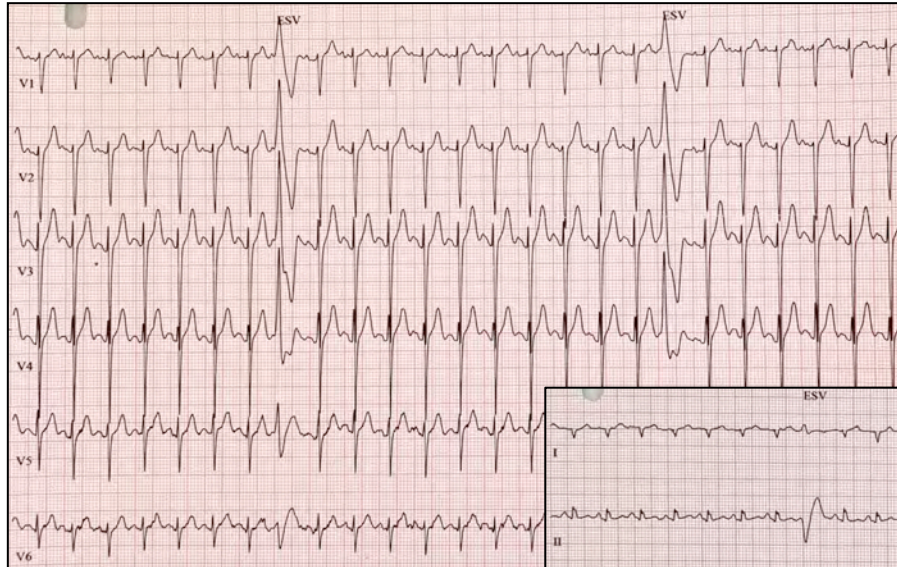
23 anni - Sportivo agonista (Calcio)- Storia negativa familiare per MCI/Cardiomiopatie
Asintomatico - Accertamenti per idoneità medico-sportiva



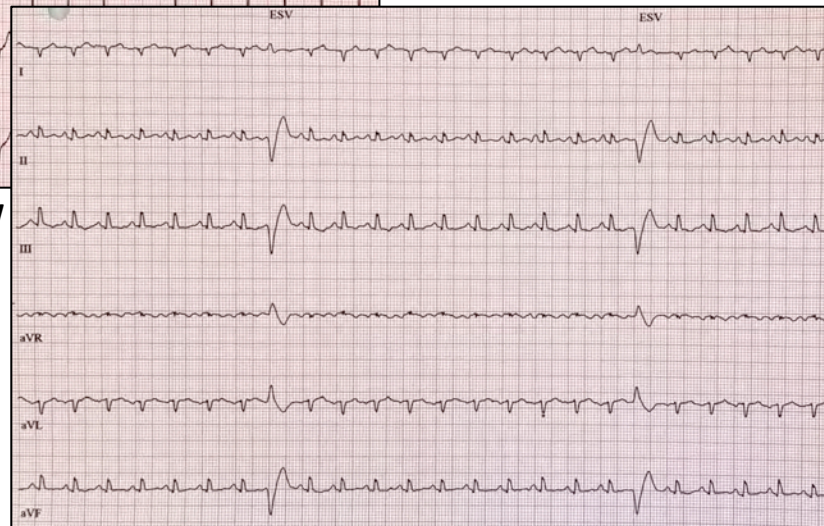
Ecocardiogramma CD: VSx normali dimensioni, volumi, spessori e funzione. Non deficit della cinesi. VDx normale.
*ECG Holter delle 24h: RS costante, **46 BEV isolati monomorfi e tardivi***

Caso 4. Marco

Test Ergometrico



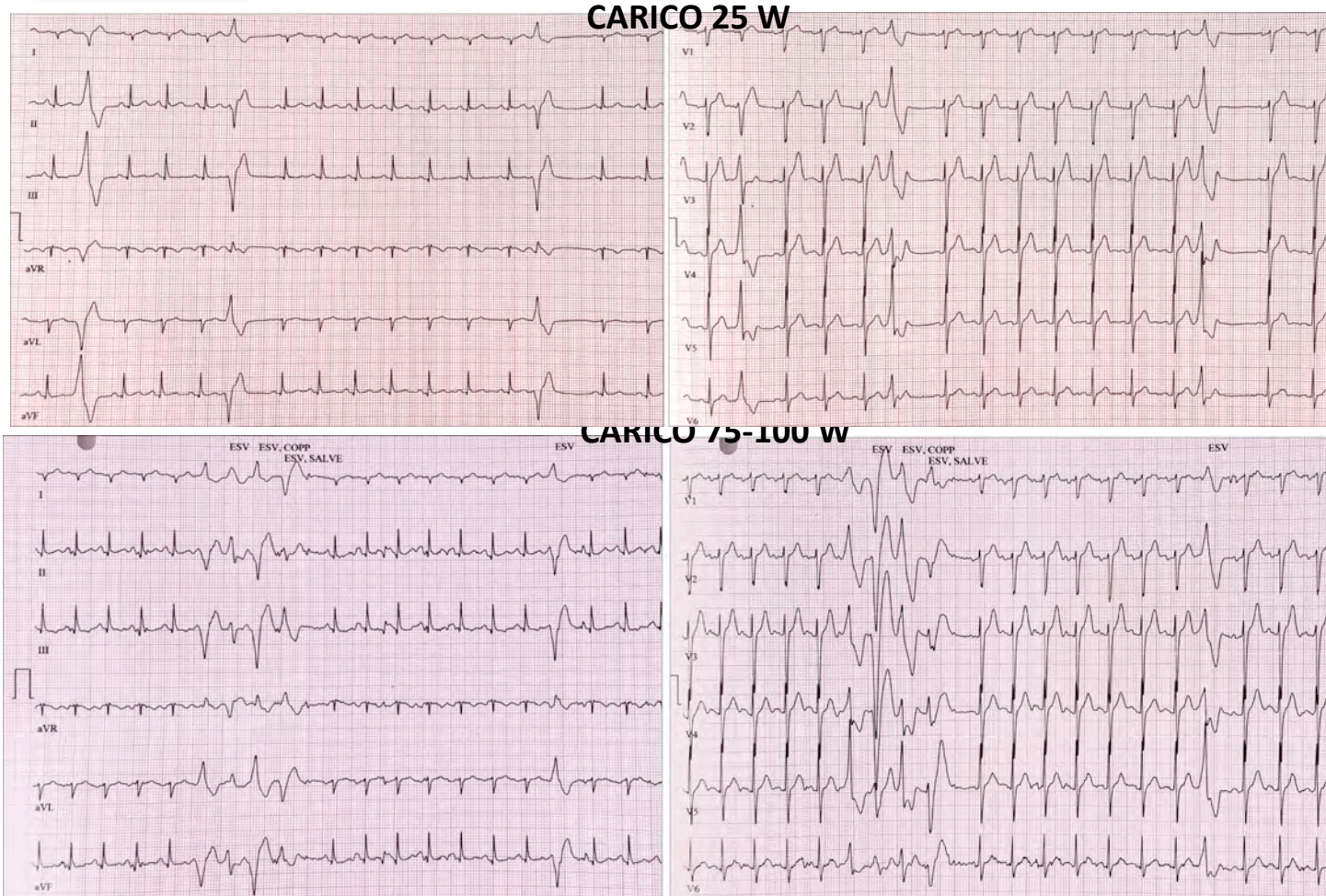
75-100 W



Rari BEV isolati
indotti da sforzo
morfologia BBDX
asse superiore

Caso 4. Marco

Quattro anni dopo Test ergometrico



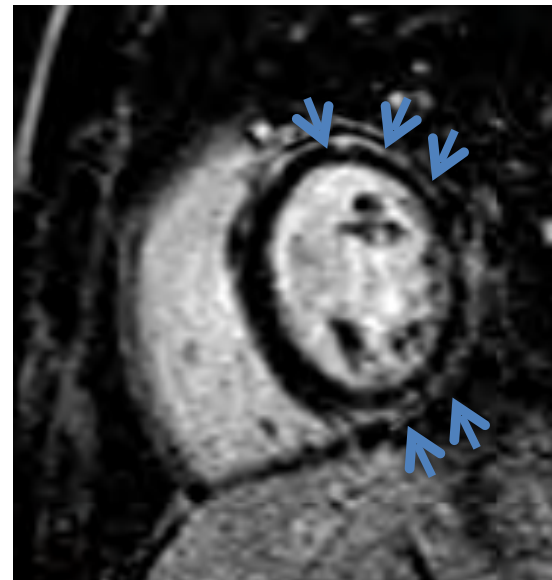
Caso 4. Marco

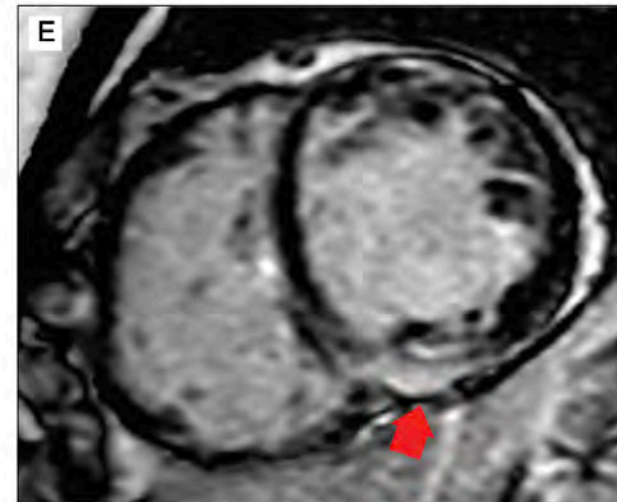
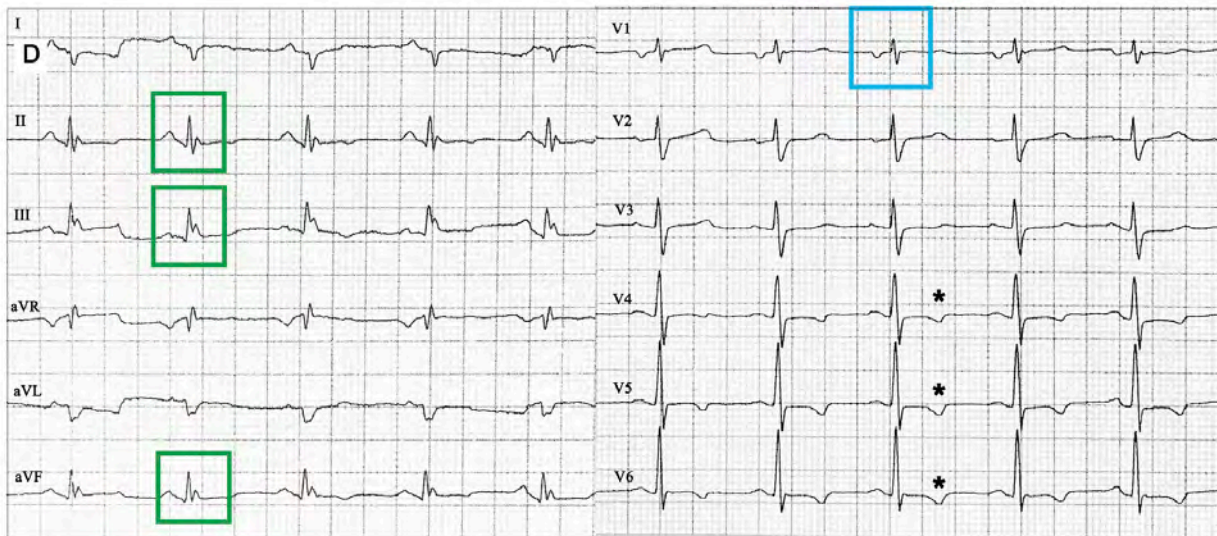
Quattro anni dopo

- *EcocardiogrammaColorDoppler*: invariato
- *ECG Holter delle 24h*: RS costante, frequenti BEV polimorfi, anche in coppie

Variante Patogenetica a carico gene DSP

**Nessun criterio ECG
identificava
la presenza di ALVC !**





Patient #9 is a 33-year-old woman with a pathogenic variant in desmoplakin (c.5851 C>T, p.Arg1951Ter).

Left Posterior Fascicular Block and Increased Risk of Sudden Cardiac Death in Young People



We retrospectively compared the clinical data for 109 consecutive individuals age ≤ 40 years who had ACA or SCD (86 men; 32.3 ± 5.9 years [range: 17 to 40 years]) and who had at least 1 ECG in the 3 years preceding the ACA or SCD to data for 8,892 healthy individuals age ≤ 40 years (6,265 men; 30.5 ± 8.6 years [range: 17 to 40 years]) consecutively referred to our institution for screening. LPFB was defined by the presence of all of the following: frontal axis 100° to 180° ; rS pattern in leads I and aVL; qR pattern in III and aVF; QRS duration < 110 ms; and no QS pattern in I and aVL. The association of LPFB with ACA/SCD was analyzed by nominal logistic regression and was estimated with unadjusted odds ratios (ORs) and 95% confidence intervals (CIs). The study (CARITMO) was approved by our Institutional Review Board.

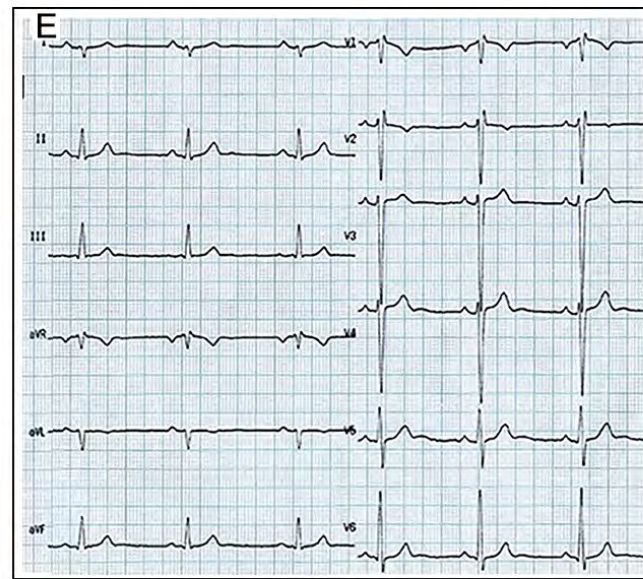
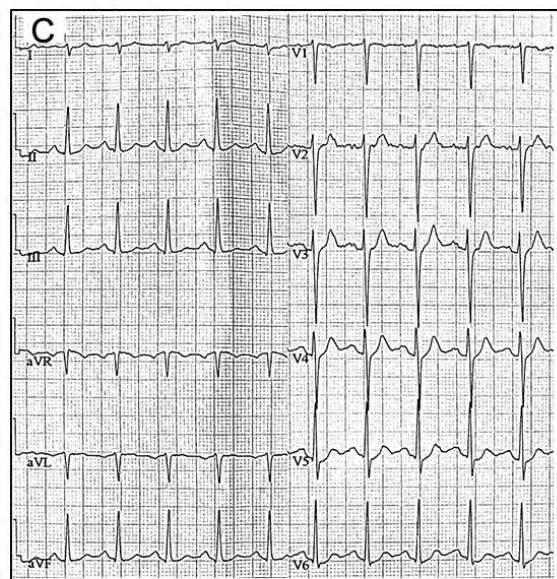
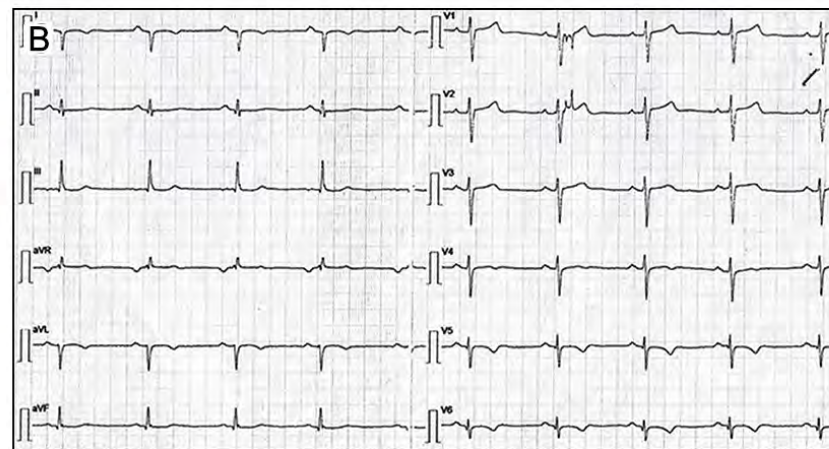
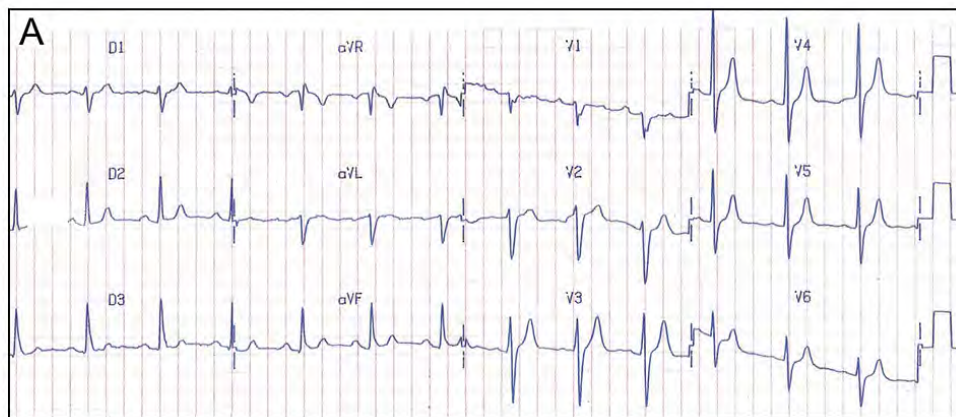
*Leonardo Calò, MD
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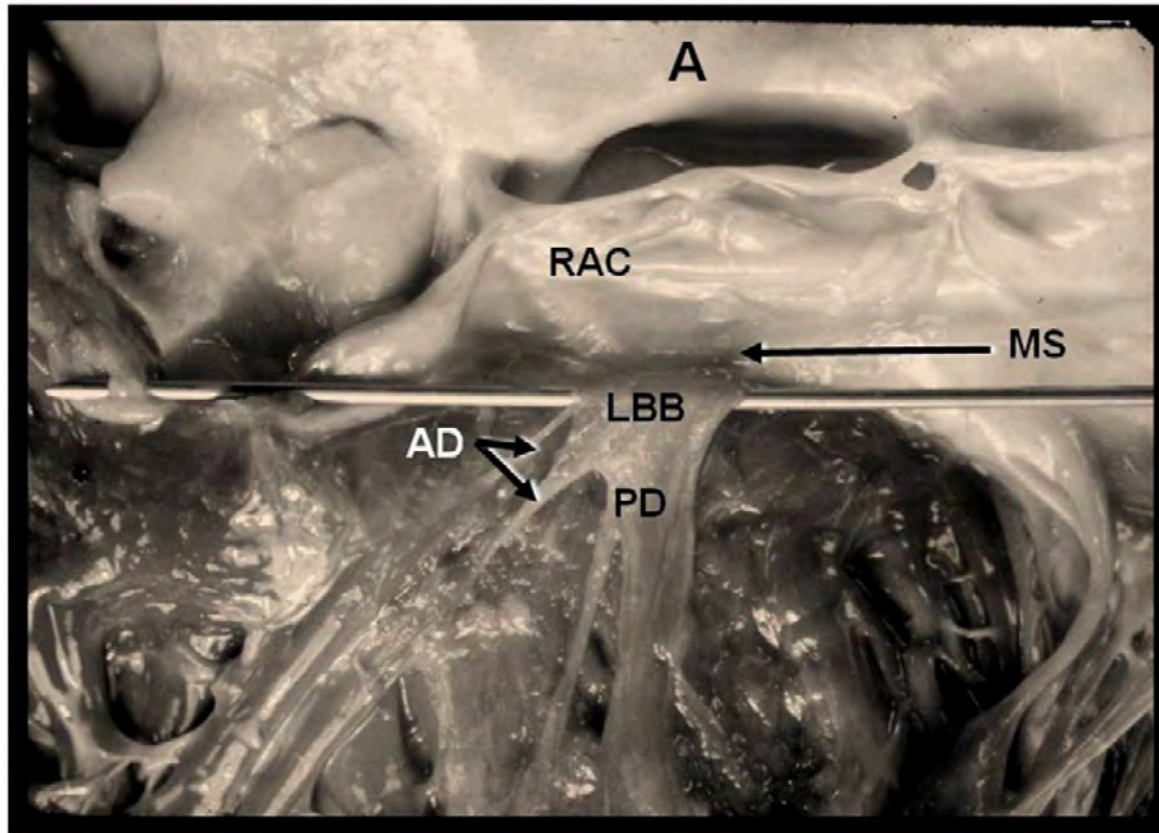
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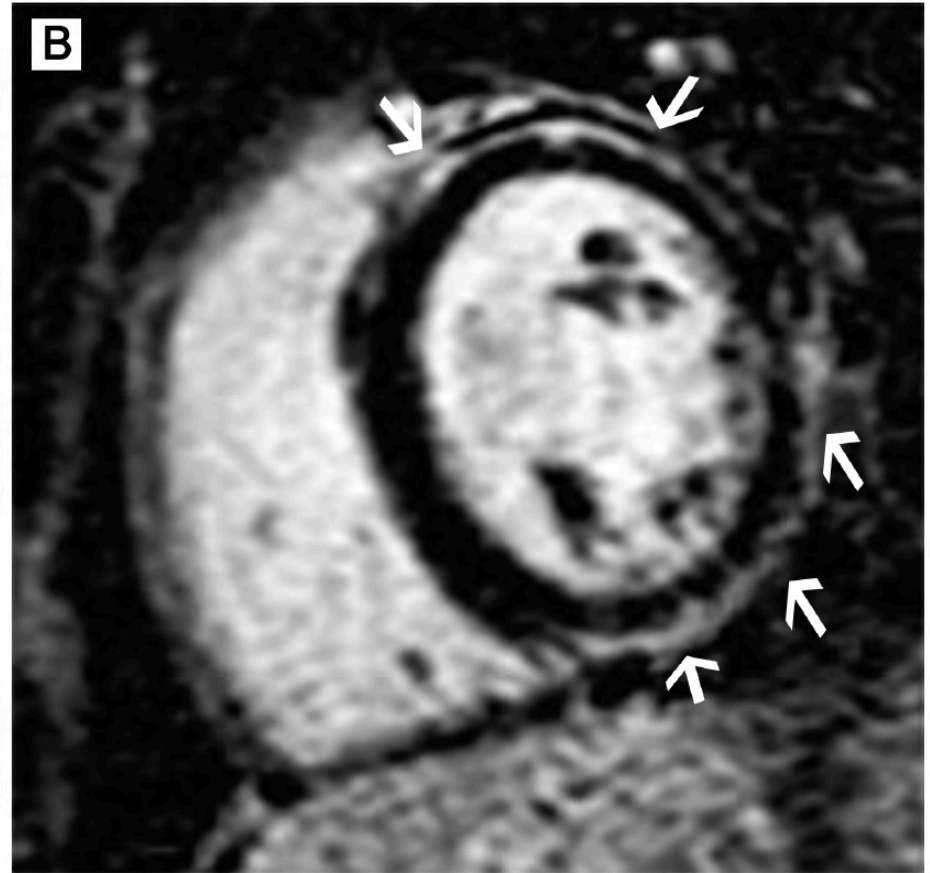
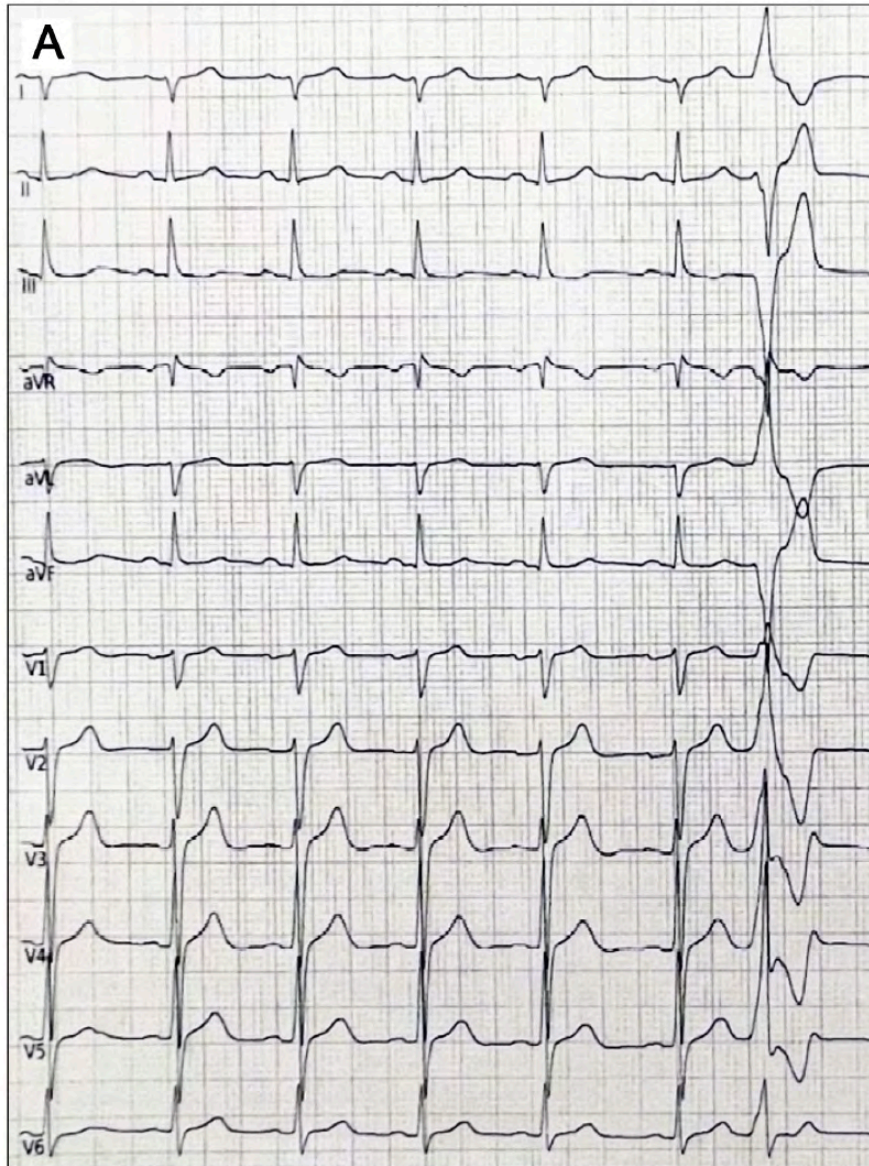
<https://doi.org/10.1016/j.jacc.2020.12.033>

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Human heart. The LBB (LBB) emerges in the subaortic region. The membranous septum (MS) is almost absent and the aortic valve lies directly over the LBB, which gives off the anterior division (AD) and posterior division (PD) from its very beginning. The membranous septum is strikingly small or practically absent in this case. The distance between the branching portion of the bundle of His from the aortic valve depends on the size of the MS. The larger the MS, the lesser the possibility that the aortic valve pathology involves this crucial part of the conducting system. A: aorta; RAC right aortic cusp.



Patient #2, 19-year-old man, pathogenic variant in desmoplakin (c.1707-1708insAC,p.Met571Glnfs*8)

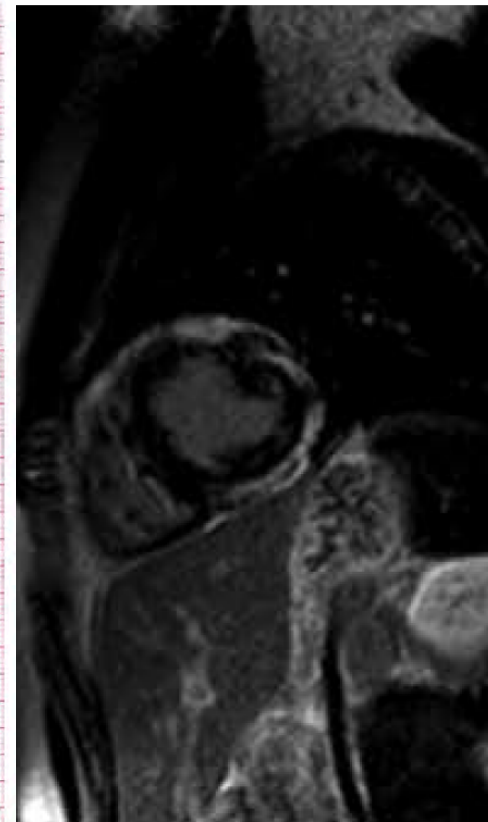
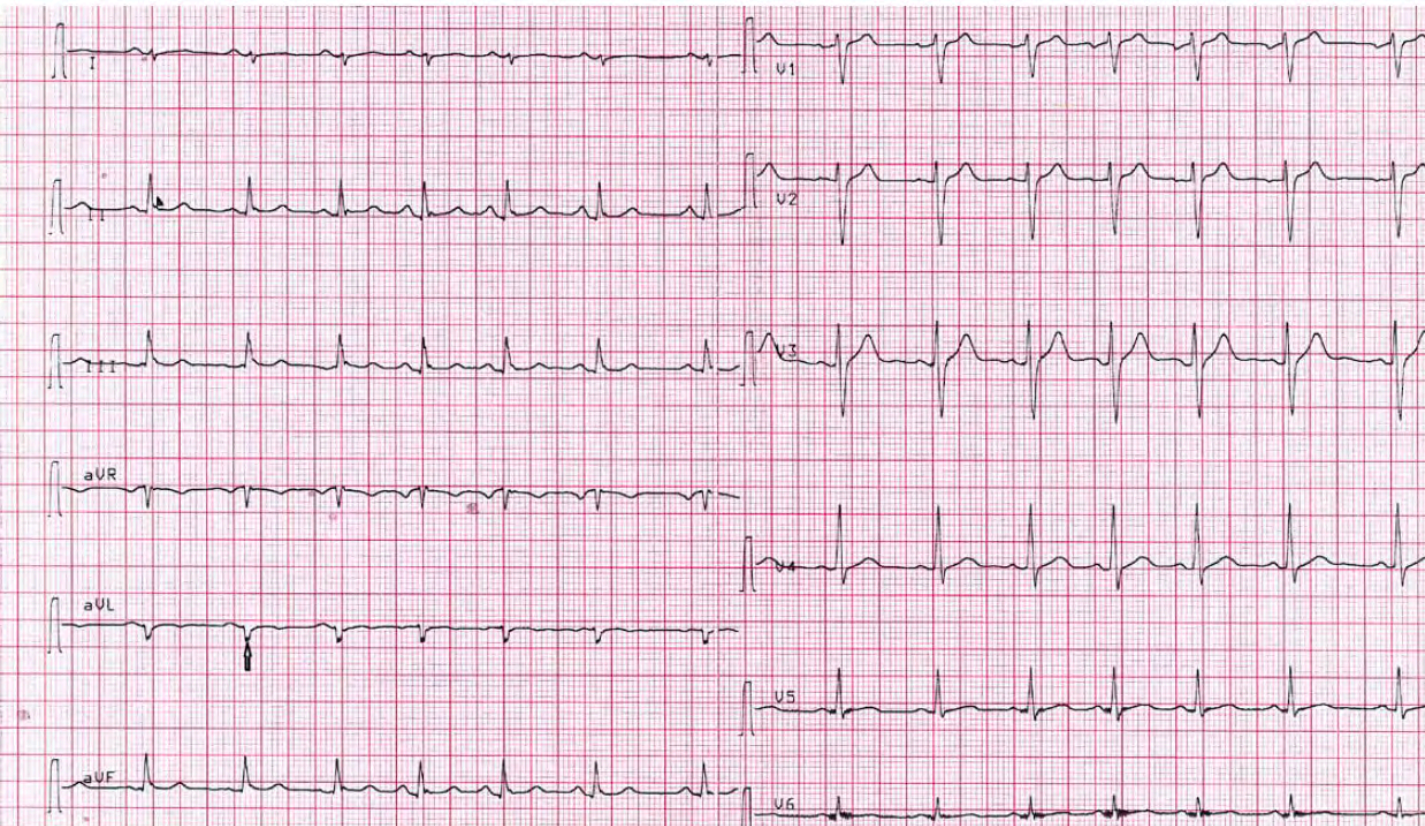
Unrecognized LPFB cases noted in the iconography of published papers

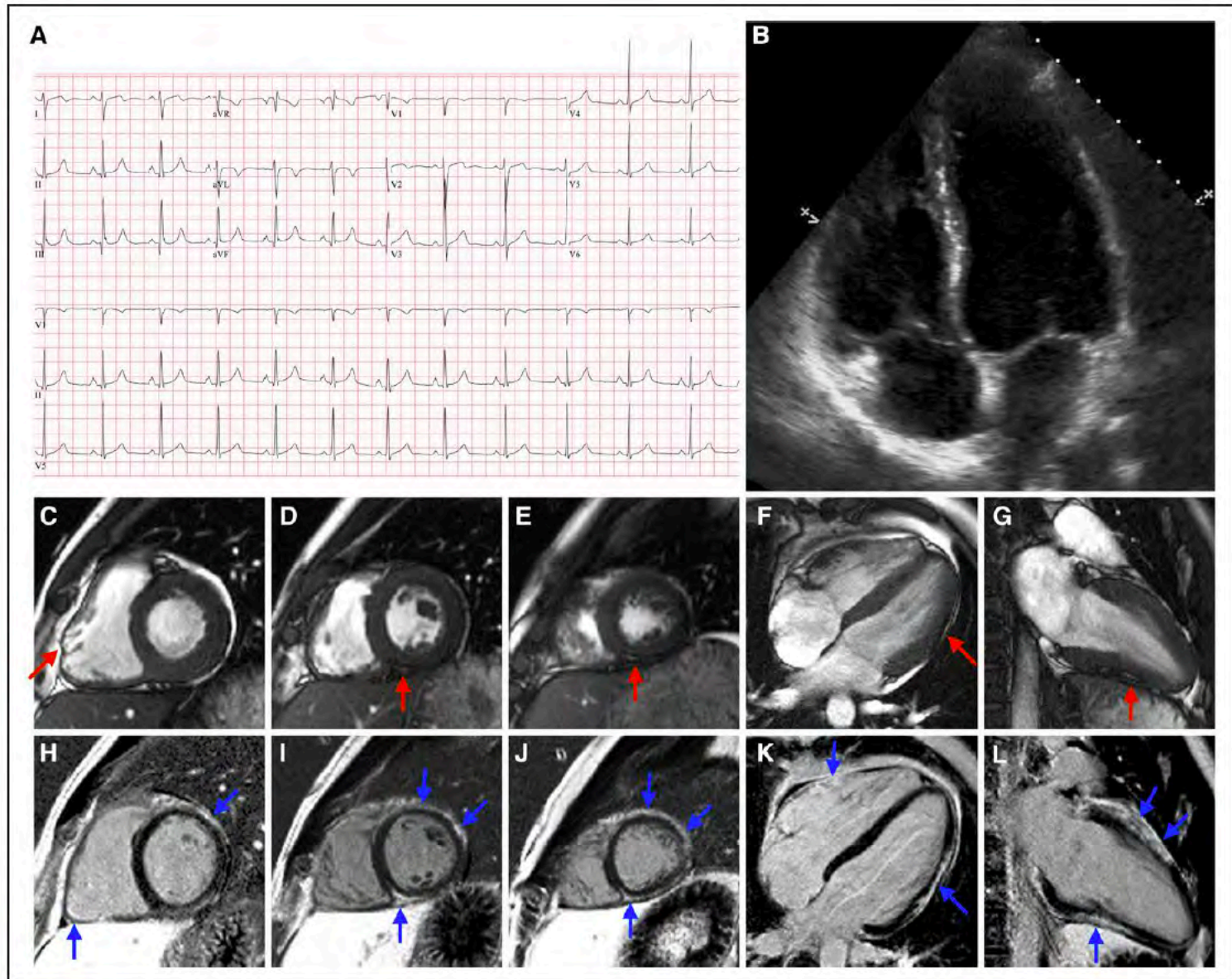
Case ID	References	Figure	12 lead ECG findings	Genetic analysis	CMR data	Endomyocardial biopsy data/ Histologic data
Case 1	Protonotarios A et al ¹ <i>J Electrocardiol.</i> 2013	Fig. 1	LPFB, LQ/RSV in limb and left precordial leads, TWI V5-V6, fQRS in lead V1.	Not performed	Not performed	Fibrotic subepicardial and midwall bands on anterolateral and posterolateral 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 TWI Pathological Q in II-III-aVF Prominent R wave V1 LQ/RSV in left precordial leads	Not performed	Sub/midmyocardial LGE with a stria 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 dysmetric and dysmorphic nuclei, with cytoplasmic vacuolization.
Case 3	Oloriz T et al ⁴ <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 – fQRS inf-septal leads First-degree AV block, Inferolateral TWI – LQ/RSV 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 fibrofatty infiltration within the posterolateral wall of the LV (extending transmurally).
Case 5	Saguner AM et al ³ <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, anterolateral, 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

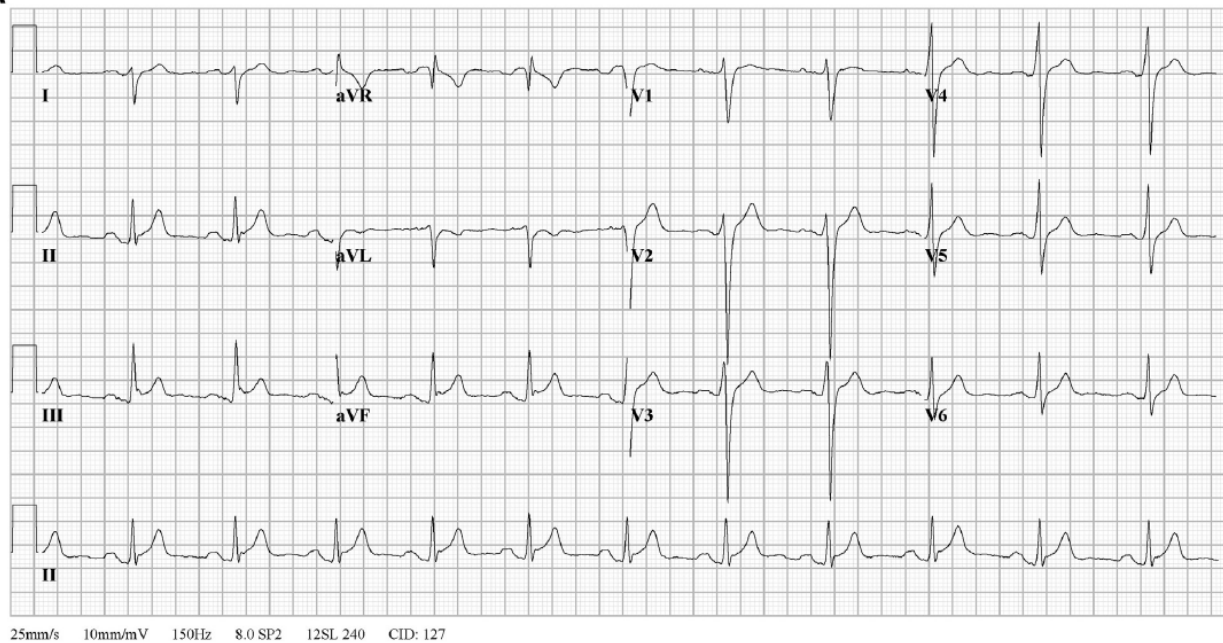
Case 7	Blom LJ et al ⁷ <i>Heart Rhythm Case Rep.</i> 2018	Fig. 1	LPFB - Intra-ventricular conduction delay, J-point elevation in inferior leads	Unclassified variant in the DSG2 gene and a p.Leu729del mutation in the gene SCN5A.	Not performed	Normal
Case 8	Blom LJ et al ⁷ <i>Heart Rhythm Case Rep.</i> 2018	Fig.2	LPFB TWI 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	Pirou N et al ⁹ <i>ESC Heart Fail.</i> 2020	Fig.2 (B)	LPFB - TWI V4-V6 LQ/RSV 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 TWI V1-V3 LQ/RSV, prolonged terminal activation duration in V1, fQRS 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 TWI inferolateral leads	Dystrophin c.3970C>T, p.Arg1324Cys, desmoplakin c.4372C>T, p.Arg1458Ter,	Multifocal subepicardial posteroseptal and lateral LGE.	Low-level immune cell infiltration in the absence of intramyocardial virus genomes
				nexilin F-actin-binding protein c.154G>C, p.Asp52His		
Case 14	Vahidinezhad H et al ¹³ <i>Sci Rep.</i> 2020	Fig.2 (A)	LPFB, TWI V1-V3 Prolonged V3 terminal QRS duration- LQ/RSV limb leads	JUP mutation	Normal	Not performed
Case 15	Chen P et al ¹⁴ <i>Int J Cardiol.</i> 2020	Fig. 2 (A)	LPFB, TWI V1-V3 R/S ratio V1 ≥ 1	DSG2 p.Leu237Ter mutation	Dilation of both ventricles	Not performed
Case 16	Protonotarios N et al ¹⁵ <i>Br Heart J.</i> 1986	Fig. 3 (A)	LPFB, QRS prolongation, LQ/RSV, TWI precordial leads.	Not performed (JUP mutation?)	Not performed	Not performed
Case 17	Chen V et al ¹⁶ <i>Eur Heart J Case Rep</i> 2022	Fig 2	LPFB	Pathogenic heterozygous DSP gene truncation variant (p.R1951X) and the pathogenic HFE variant (p.H63D).	Subepicardial basal-anterior, basal anterolateral, mid-inferior and mid-anteroseptal areas of LGE.	Mild lymphocytic myocarditis, interstitial fibrosis, and myocyte hypertrophy

Arrhythmogenic Left Ventricular Cardiomyopathy Suspected by Cardiac Magnetic Resonance Imaging, Confirmed by Identification of a Novel Plakophilin-2 Variant

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Alexander Gotschy, MD; Argelia Medeiros-Domingo, MD, PhD; Corinna Brunckhorst, MD;
Firat Duru, MD; Kurt A. Mayer, MD





A**B**



Fascicular heart blocks and risk of adverse cardiovascular outcomes: results from a large primary care population.

Benjamin Chris Nyholm, MD, Jonas Ghouse, MD, PhD, Christina Ji-Young Lee, MD, PhD, Peter Vibe Rasmussen, MD, Adrian Pietersen, MD, Steen Møller Hansen, MD, PhD, Christian Torp-Pedersen, MD, DMSci, Lars Køber, MD, DMSci, Stig Haunsø, MD, DMSci, Morten Salling Olesen, MSc, PhD, Jesper Hastrup Svendsen, MD, DMSci, Claus Graff, MSc, PhD, Anders Gaarsdal Holst, MD, PhD, Jonas Bille Nielsen, MD, PhD, Morten Wagner Skov, MD, PhD

PII: S1547-5271(21)02216-5

DOI: <https://doi.org/10.1016/j.hrthm.2021.09.041>

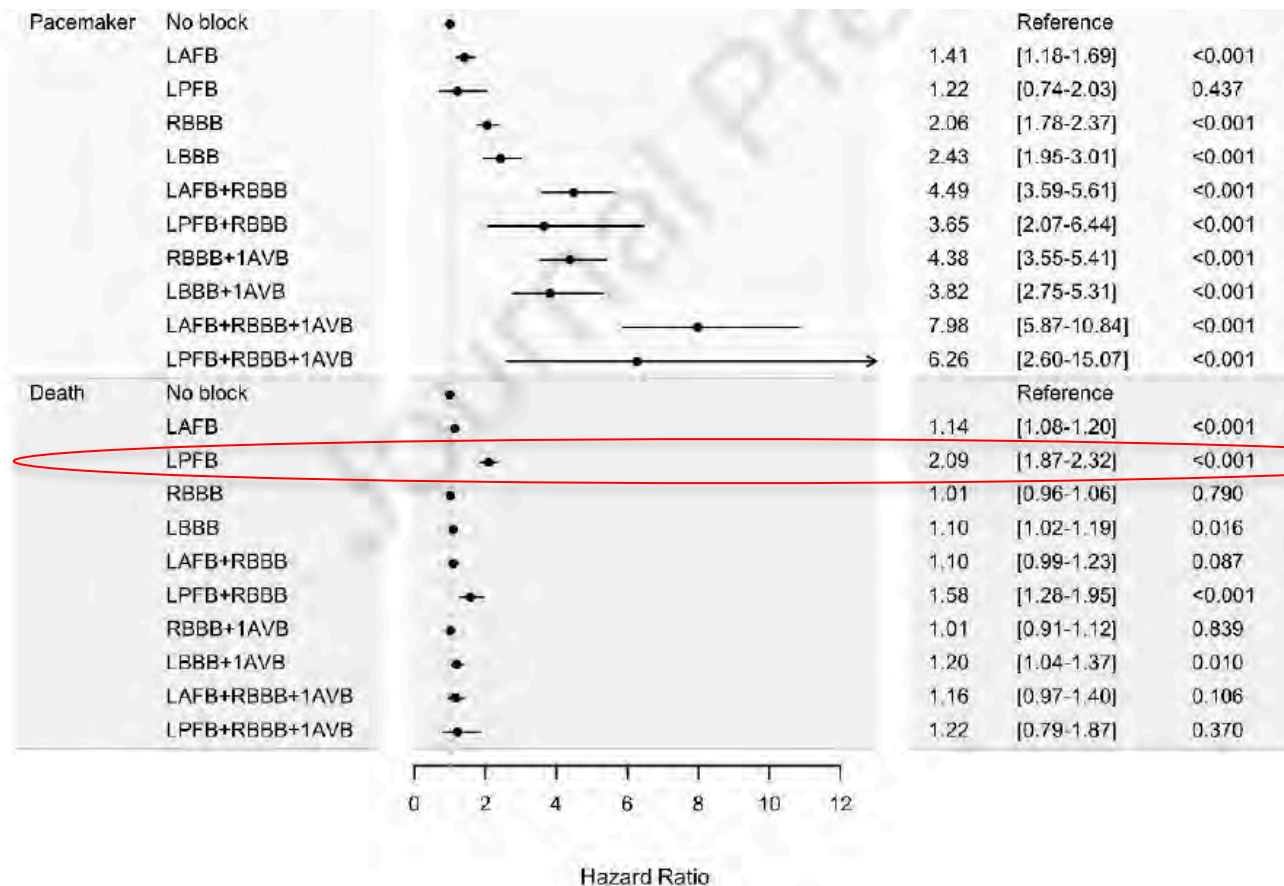


Table 1. Baseline characteristics

Fascicular block subtype n (%)	No conduction defects		LAFB		LPFB		RBBB		LBBB		RBBB + LAFB		RBBB + LPFB		RBBB + 1AVB		LBBB + 1AVB		RBBB + LAFB + 1AVB		RBBB + LPFB + 1AVB	
	345,315	(96.2)	3,526	(0.98)	2,889	(0.80)	3,910	(1.09)	1,390	(0.39)	570	(0.16)	196	(0.05)	643	(0.18)	304	(0.08)	176	(0.05)	32	(0.01)
Age, years, median (IQR)	54	(41-66)	71	(60-80)	35	(26-51)	69	(58-79)	75	(64-82)	76	(68-83)	66	(53-78)	77	(70-84)	80	(72-86)	79	(70-86)	78	(71-84)
Women, n (%)	190,796	55	1,463	41	1,441	50	1,349	35	949	68	169	30	79	40	158	25	136	45	32	18	6	19
Medical history, n (%)																						
Hypertension	66,316	(19)	1,139	(32)	198	(7)	1,349	(35)	625	(45)	241	(42)	48	(25)	333	(52)	174	(57)	101	(57)	15	(47)
Syncope	9,355	(2.7)	135	(3.8)	83	(2.9)	148	(3.8)	60	(4.3)	26	(4.6)	6	(3.1)	37	(5.8)	13	(4.3)	16	(9.1)	<4	(<12.5)*
Atrial fibrillation	5,041	(1.5)	134	(3.8)	38	(1.3)	121	(3.1)	66	(4.8)	20	(3.5)	9	(4.6)	35	(5.4)	27	(8.9)	12	(6.8)	5	(15.6)
Valvular heart disease	1,442	(0.4)	46	(1.3)	10	(0.4)	50	(1.3)	25	(1.8)	12	(2.1)	<4	(<2)*	15	(2.3)	12	(4.0)	<4	(<3)	<4	(<12.5)*
Beta blocker therapy	52,809	(15.3)	736	(20.9)	242	(8.4)	845	(21.6)	408	(29.4)	132	(23.2)	31	(15.8)	193	(30.2)	111	(36.5)	58	(33.0)	11	(34.4)
Charlson Comorbidity Index																						
0 points	265,361	(77)	2,078	(59)	2,413	(84)	2,429	(62)	802	(58)	296	(52)	124	(63)	319	(50)	144	(47)	71	(40)	14	(44)
1 point	41,598	(12)	648	(18)	271	(9)	632	(16)	237	(17)	109	(19)	31	(16)	121	(19)	66	(22)	34	(19)	8	(25)
≥ 2 points	38,356	(11)	800	(23)	205	(7)	849	(22)	351	(25)	165	(29)	41	(21)	203	(31)	94	(31)	71	(40)	10	(31)
ECG variables																						
QRS duration, median (IQR)	92	(84-100)	102	(96-110)	96	(88-104)	136	(128-146)	146	(136-156)	144	(136-154)	140	(130-148)	144	(134-152)	154	(144-164)	151	(142-160)	150	(134-159)
PR interval, median (IQR)	156	(144-172)	168	(152-186)	154	(140-170)	162	(148-178)	168	(154-180)	170	(156-184)	162	(146-178)	220	(208-242)	218	(208-238)	228	(211-251)	221	(207-242)
Heart rate (IQR)	69	(62-79)	72	(63-82)	71	(62-82)	70	(62-80)	73	(65-83)	71	(64-80)	74	(66-84)	68	(60-78)	70	(62-80)	69	(60-77)	76	(68-86)

TABLE 1: LAFB = left anterior fascicular block; LPFB = left posterior fascicular block; LBBB = left bundle branch block; RBBB = right bundle branch block; 1AVB = first degree atrioventricular block;

*Due to the Act on Processing of Personal Data, we are not allowed to report any number less than four observations.

NUOVI SEGNI ECG NELLA DIAGNOSI DI ALVC

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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Obiettivi:

- 1) analizzare **ECG in pazienti affetti da ALVC** confrontandole con quelle di soggetti sani di controllo
- 2) correlare i risultati ECG con i risultati della risonanza magnetica cardiaca e i dati genetici

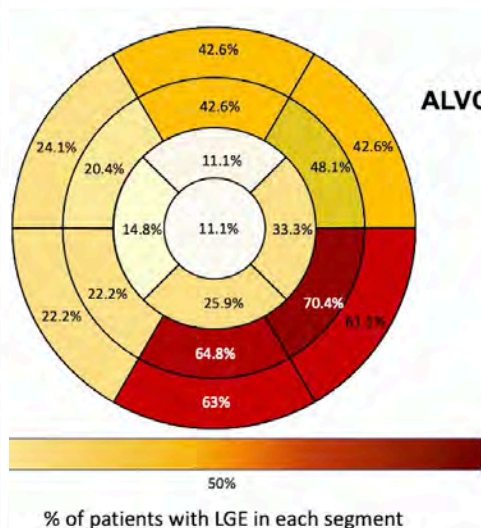
TABLE 1 Baseline Clinical, Structural, and Genetic Characteristics of the Study Population (ALVC, N = 54)

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)
Genetic testing	
Pathogenic/likely pathogenic variant	48/54 (88.9)
DSP	35/48 (72.9)
Non-DSP	13/48 (27.1)

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)



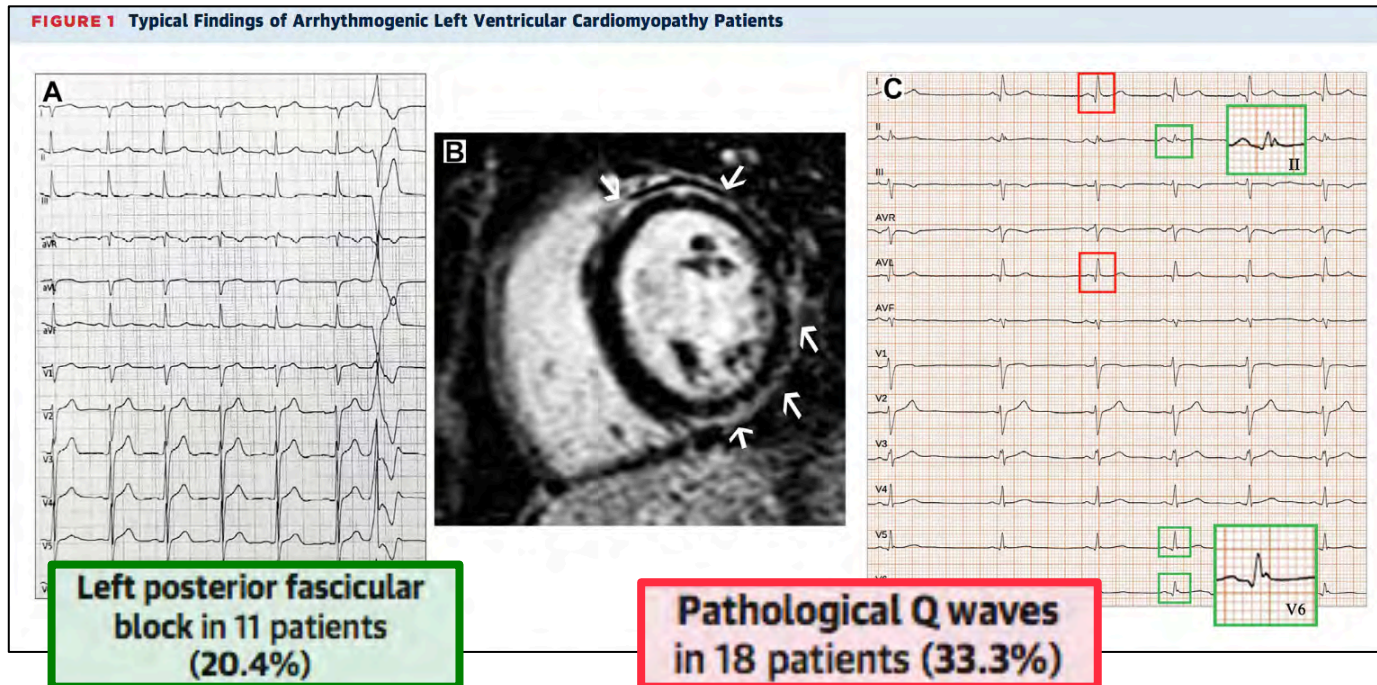
ALVC

Otto (**14.8%**) pazienti ECG apparentemente normale

Bassi voltaggi periferici nelle derivazioni periferiche in 12 paz (**22.2%**).

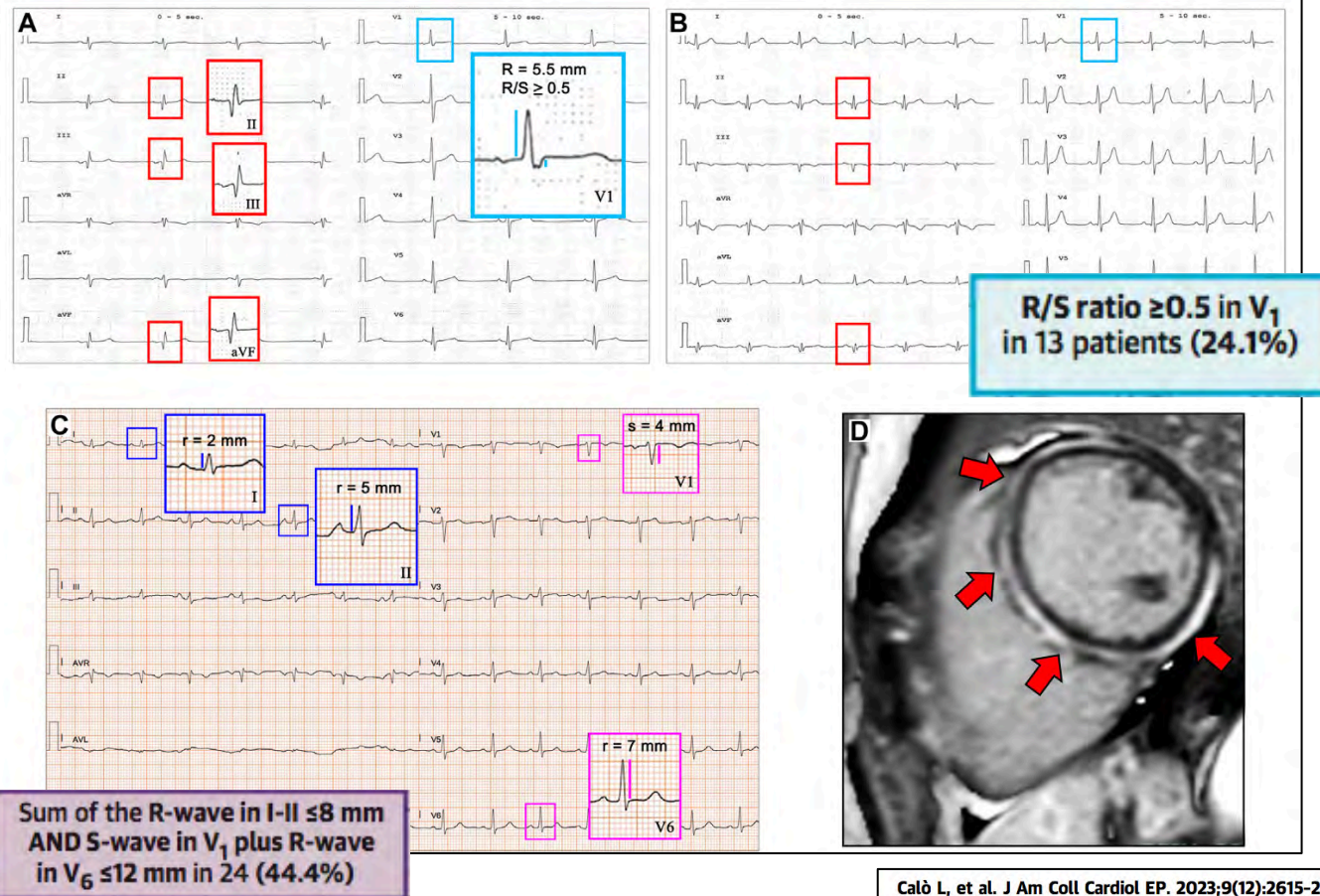
Onde T negative in 31 paz (**57.4%**), particolarmente evidenti nelle derivazioni inferiori e laterali

NUOVI SEGNI ECG NELLA DIAGNOSI DI ALVC

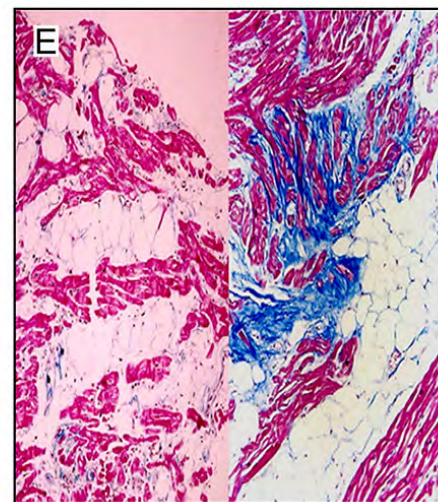
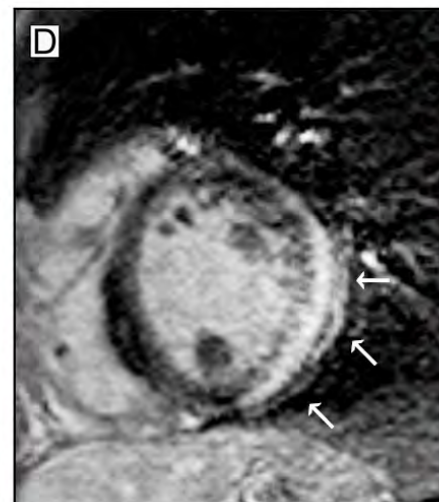
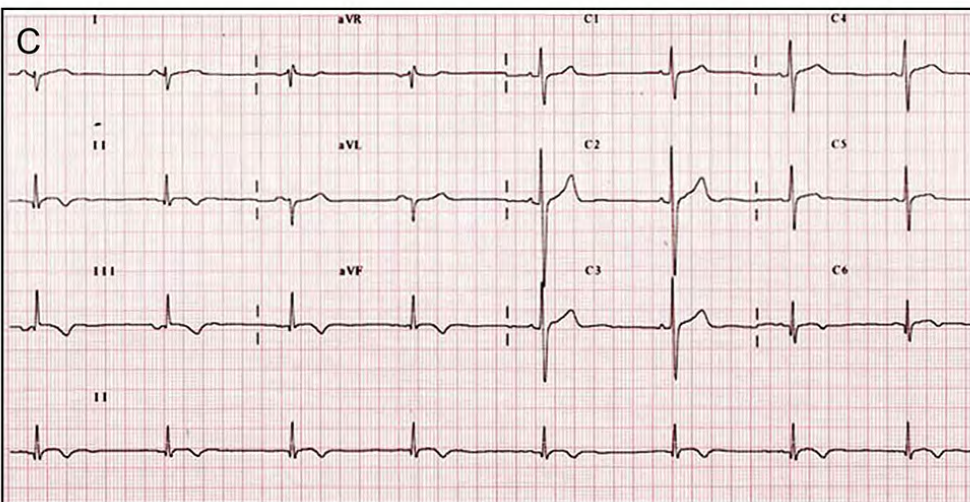
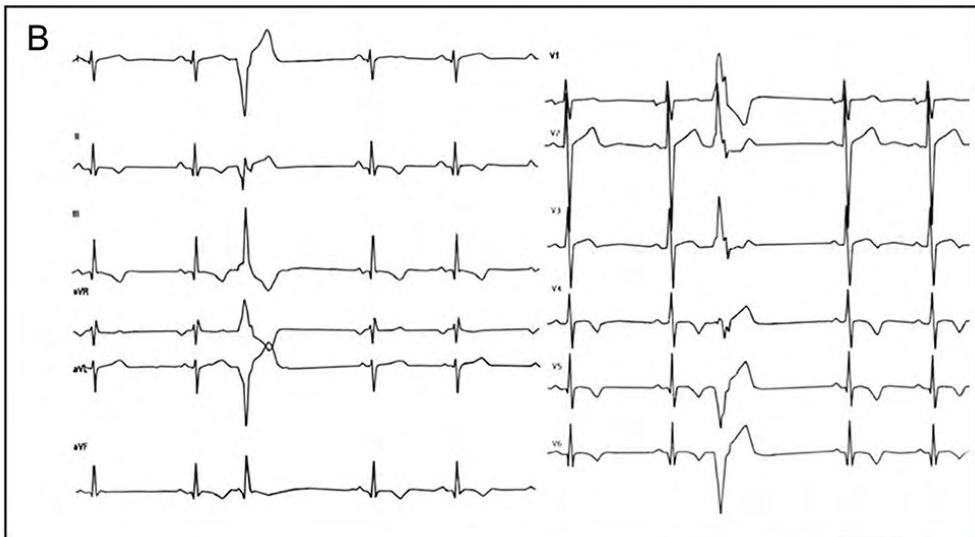


NUOVI SEGNI ECG NELLA DIAGNOSI DI ALVC

FIGURE 2 Typical Findings of ALVC Patients



Analisi temporale dell'ECG nella cardiomiopatia aritmogena



CASE REPORT

ADVANCED

CLINICAL CASE SERIES

Acute Myocardial Infarction-Like Events in Related Patients With a Desmoplakin-Associated Arrhythmogenic Cardiomyopathy

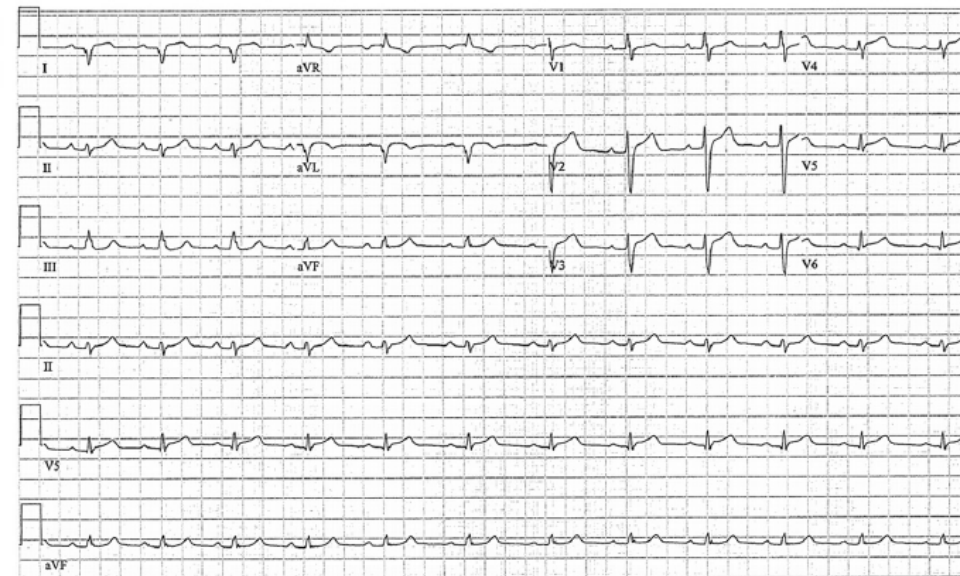
Sajya M. Singh, BS,^a Scott W. Sharkey, MD,^a Susan A. Casey, RN,^a Kevin M. Harris, MD,^a Christina M. Thaler, MD,^a Mina Chung, MD,^b Allison Berg, MS, CGC,^c Mosi K. Bennett, MD,^a Emily R. Ducanson, MD,^a Shannon Mackey-Bojack, MD,^d Jay D. Sengupta, MD^a



B



C

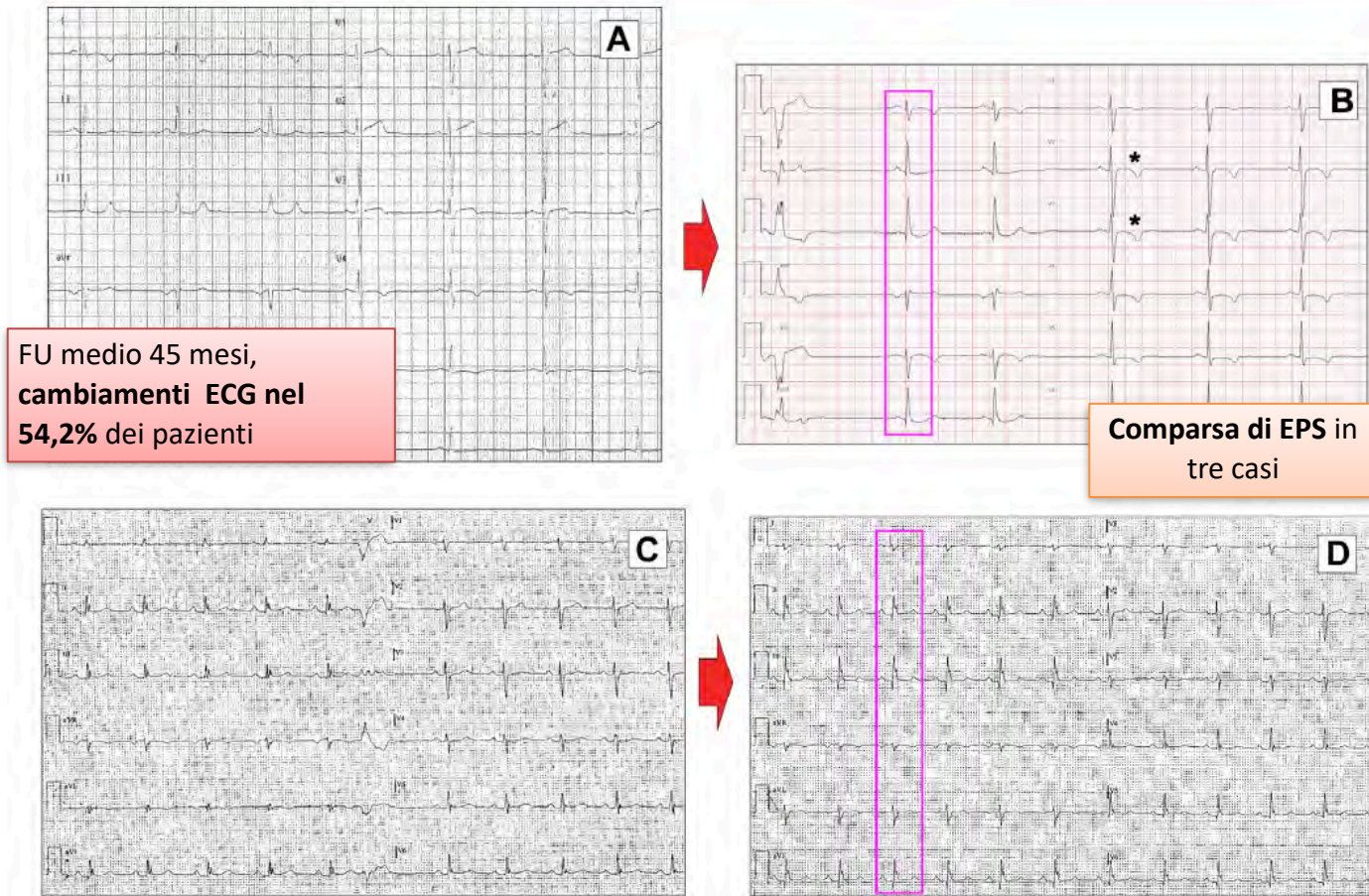


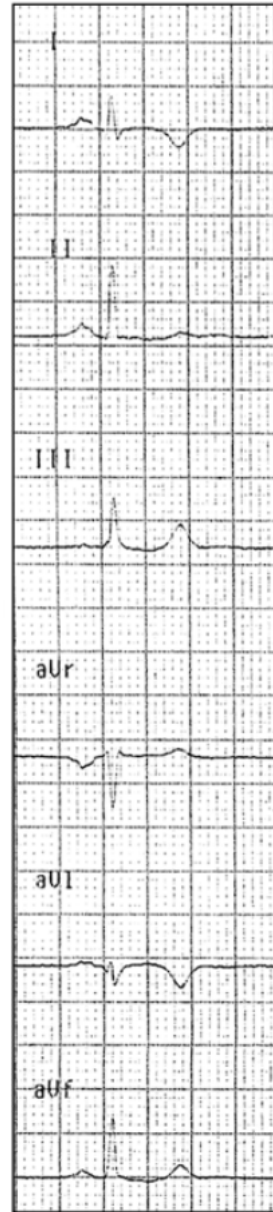
A

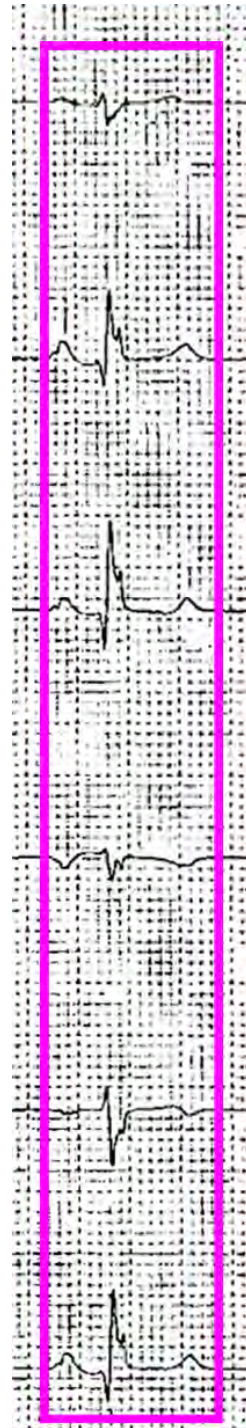
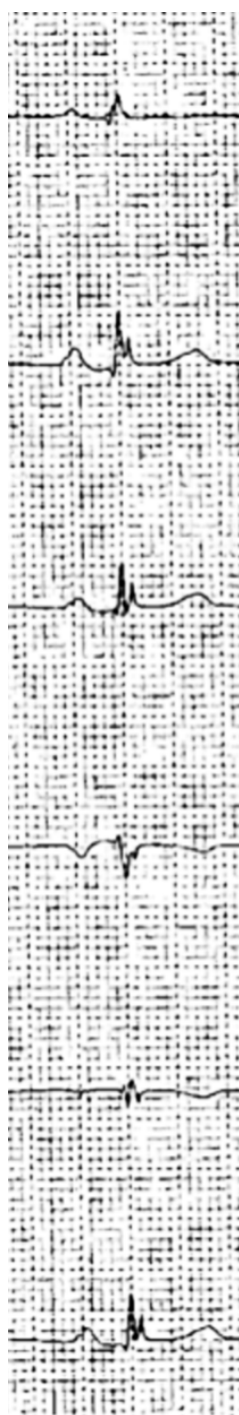


MODIFICHE DELL'ECG NEL TEMPO

FIGURE 2 Electrocardiographic Changes During Follow-Up







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

JACC: ADVANCES
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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:



The ECG correlation with CMR and genetic data



ECG findings in ALVC



The prognostic value of ECG in ALVC

ALVC defined by the presence of the following criteria:

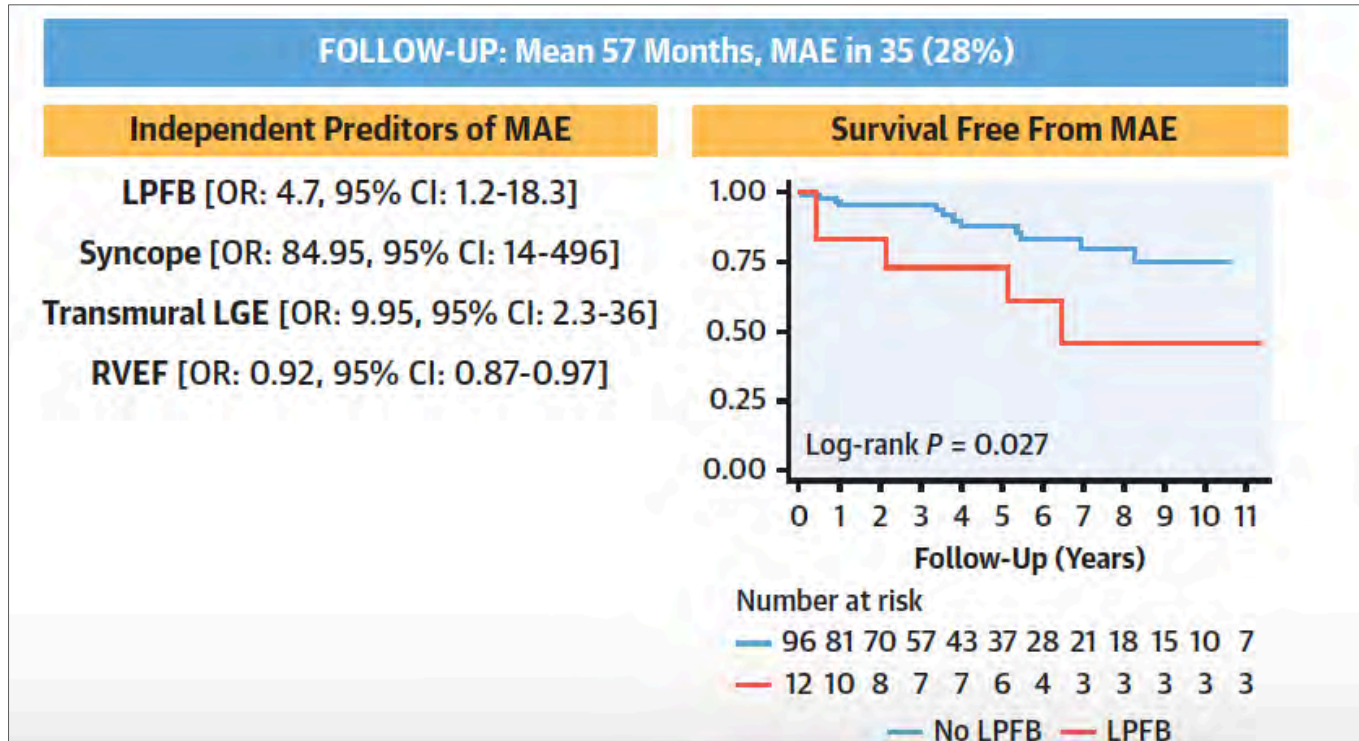
1. Presence of LGE in the LV, manifesting as a stria (or band) pattern affecting >1 segment.
2. Pathogenic (class V)/ likely pathogenic (class IV) variants in genes responsible for desmosomal proteins associated with ALVC.
3. Confirmation of ALVC diagnosis through endomyocardial or at autopsy, for those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

The Role of ECG in Arrhythmogenic Left Ventricular Cardiomyopathy

125 patients with ALVC from 14 Centers; Median Age 37±15years; 51% male; 71% probands.



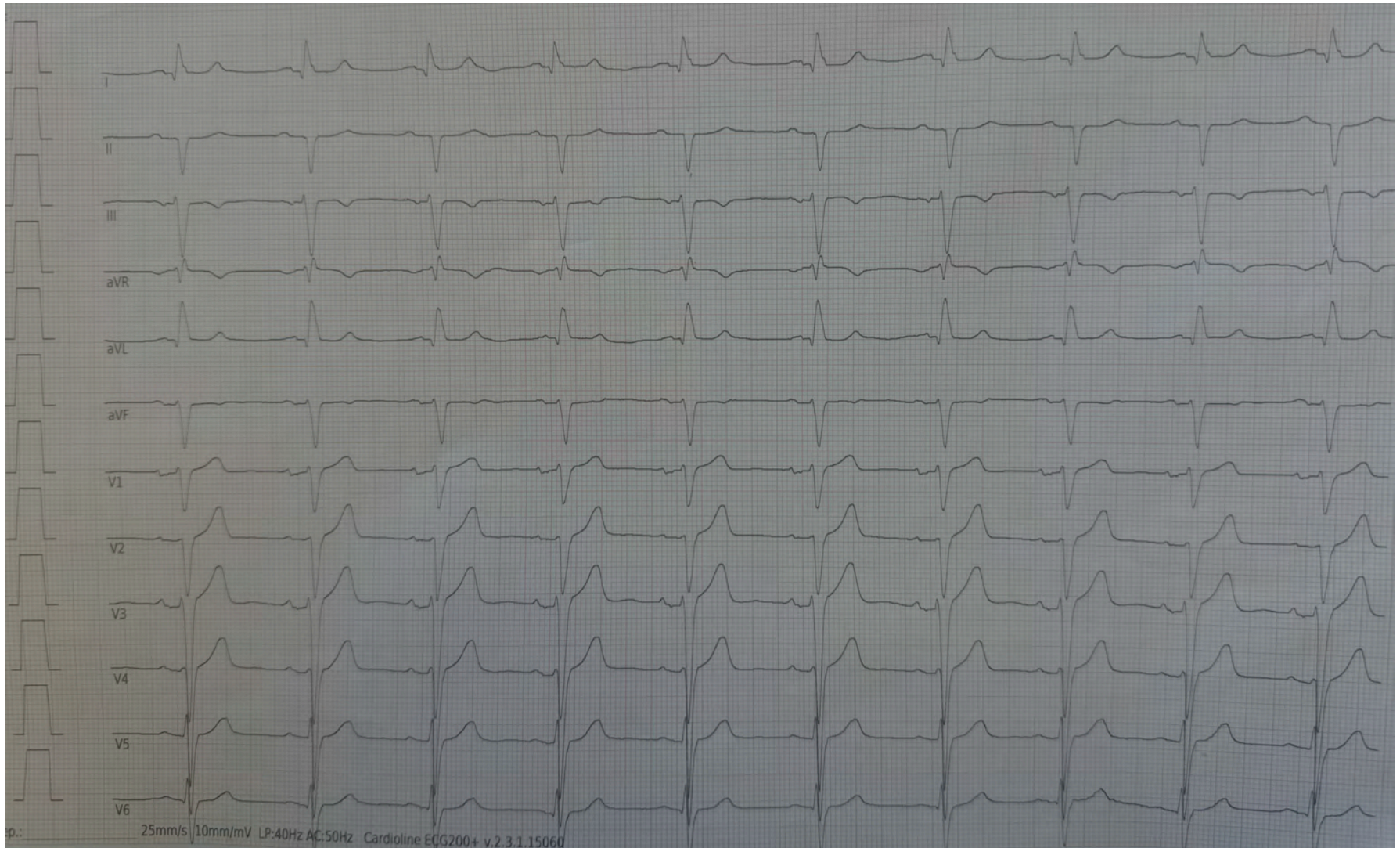
Follow-up

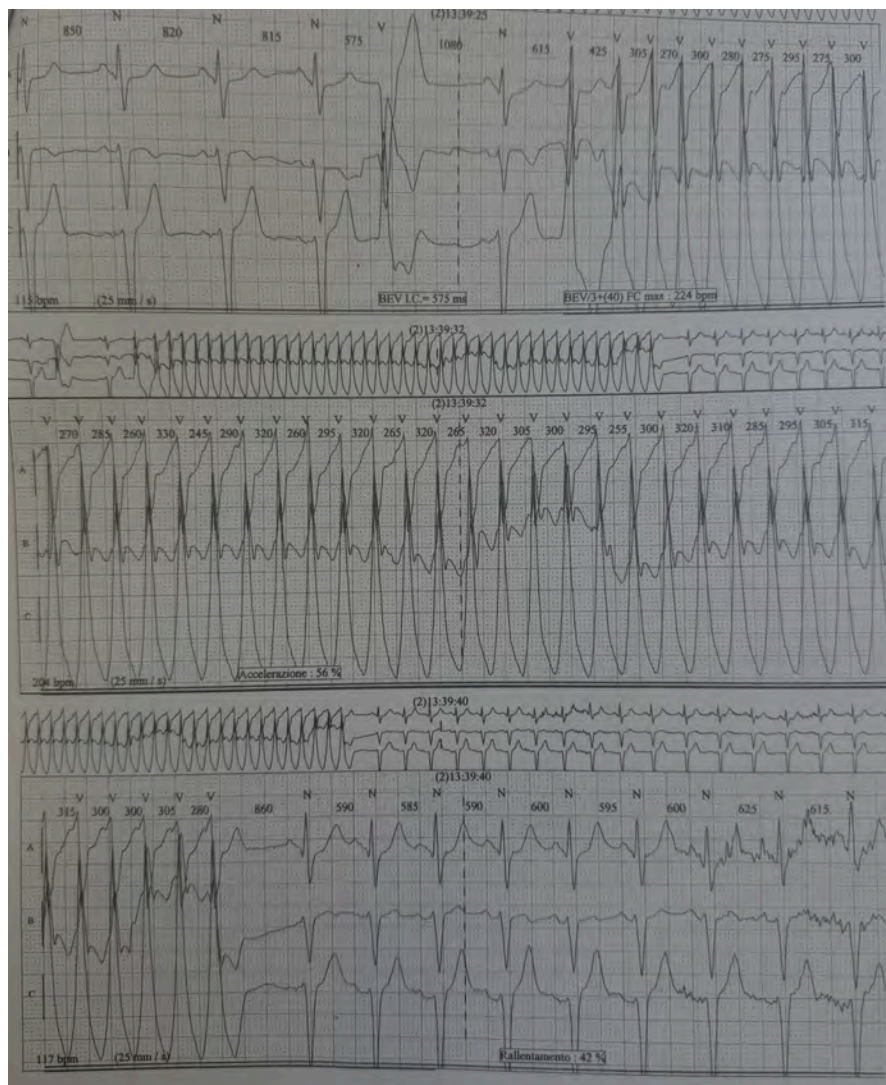


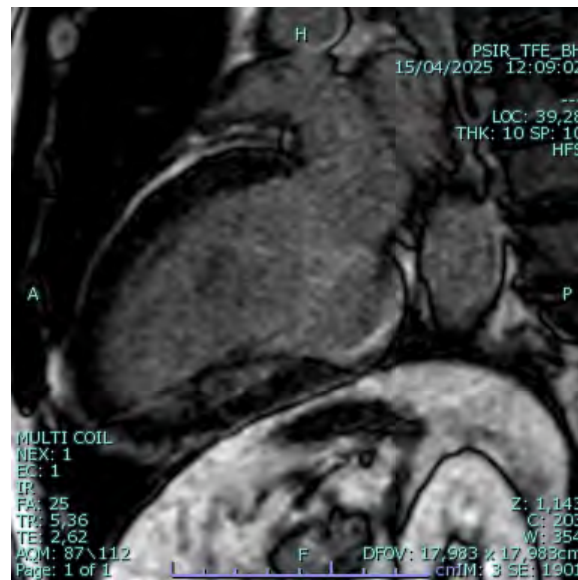
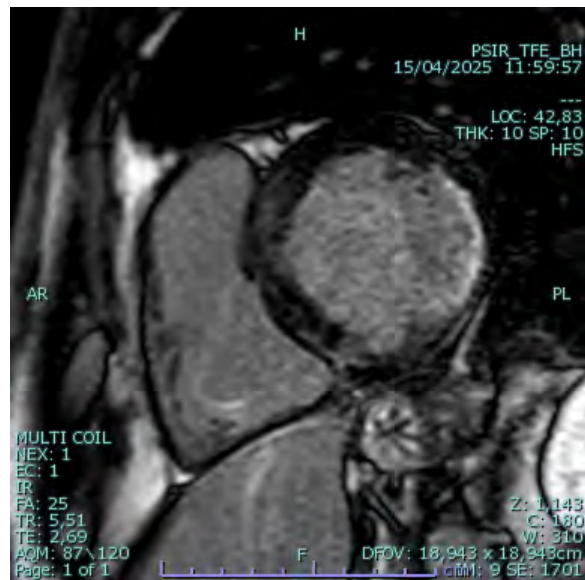
ECG progression during follow-up

During a median follow-up of **45 months** (Q1-Q3: 27-70), ECG changes were observed in 39/72 patients (**54.2%**). In 3 patients, we observed the new appearance of a LPFB.

51-year-old man- Symptomatic for syncope and palpitation- Normal Echocardiogram
-Normal coronary arteries







Subendocardial LGE in the basal infero-lateral wall of the left ventricle with an EF of 45%

Left anterior fascicular block

The Role of ECG in Arrhythmogenic Left Ventricular Cardiomyopathy

125 patients with ALVC from 14 Centers; Median Age 37±15years; 51% male; 71% probands.



Arrhythmogenic Right Ventricular Cardiomyopathy: Characterization of Left Ventricular Phenotype and Differential Diagnosis With Dilated Cardiomyopathy

Alberto Cipriani, MD; Barbara Baucé, MD, PhD; Manuel De Lazzari, MD, PhD; Ilaria Rigato, MD, PhD; Riccardo Bariani, MD; Samuele Meneghin, MD; Kalliopi Pilichou, MD, PhD; Raffaella Motta, MD, PhD; Camillo Aliberti, MD; Gaetano Thiene, MD; William J. McKenna, MD, DSc; Alessandro Zorzi, MD, PhD; Sabino Iliceto, MD; Cristina Basso, MD, PhD; Martina Perazzolo Marra, MD, PhD; *Domenico Corrado , MD, PhD*

Background—This study assessed the prevalence of left ventricular (LV) involvement and characterized the clinical, electrocardiographic, and imaging features of LV phenotype in patients with arrhythmogenic right ventricular cardiomyopathy (ARVC). Differential diagnosis between ARVC-LV phenotype and dilated cardiomyopathy (DCM) was evaluated.

Methods and Results—The study population included 87 ARVC patients (median age 34 years) and 153 DCM patients (median age 51 years). All underwent cardiac magnetic resonance with quantitative tissue characterization. Fifty-eight ARVC patients (67%) had LV involvement, with both LV systolic dysfunction and LV late gadolinium enhancement (LGE) in 41/58 (71%) and LV-LGE in isolation in 17 (29%). Compared with DCM, the ARVC-LV phenotype was statistically significantly more often characterized by low QRS voltages in limb leads, T-wave inversion in the inferolateral leads and major ventricular arrhythmias. LV-LGE was found in all ARVC patients with LV systolic dysfunction and in 69/153 (45%) of DCM patients. Patients with ARVC and LV systolic dysfunction had a greater amount of LV-LGE (25% versus 13% of LV mass; $P<0.01$), mostly localized in the subepicardial LV wall layers. An LV-LGE $\geq 20\%$ had a 100% specificity for diagnosis of ARVC-LV phenotype. An inverse correlation between LV ejection fraction and LV-LGE extent was found in the ARVC-LV phenotype ($r=-0.63$; $P<0.01$), but not in DCM ($r=-0.01$; $P=0.94$).

Conclusions—LV involvement in ARVC is common and characterized by clinical and cardiac magnetic resonance features which differ from those seen in DCM. The most distinctive feature of ARVC-LV phenotype is the large amount of LV-LGE/fibrosis, which impacts directly and negatively on the LV systolic function. (*J Am Heart Assoc.* 2020;9:e014628. DOI: 10.1161/JAHA.119.014628.)

Key Words: arrhythmogenic right ventricular cardiomyopathy • cardiac magnetic resonance • dilated cardiomyopathy • late gadolinium enhancement • left ventricular involvement • left ventricular phenotype

	ARVC Patients (n=87)	LV Involvement No (n=29)	LV Involvement Yes (n=58)	P Value
Age at diagnosis, y	34 (22–47)	36 (25–43)	34 (19–50)	0.689
Male sex	56 (64)	20 (69)	36 (62)	0.527
Symptoms				
Chest pain	10 (11)	3 (10)	7 (12)	1.00
Syncope	29 (33)	13 (45)	16 (28)	0.108
Palpitations	13 (15)	1 (3)	12 (21)	0.06
Exertional dyspnea	20 (23)	4 (14)	16 (28)	0.149
Family history				
Clinical ARVC diagnosis in relatives	57 (66)	20 (69)	37 (64)	0.632
Family history of SCD	22 (25)	10 (35)	12 (21)	0.163
Gene mutation carriers	59 (68)	23 (79)	36 (62)	0.105
Arrhythmic history				
Atrial fibrillation	6 (7)	1 (3)	5 (9)	0.659
Major ventricular arrhythmias	33 (38)	11 (38)	22 (38)	1.00
Electrocardiographic features				
First degree atrioventricular block	6 (7)	0	6 (10)	0.07
Complete left bundle branch block	0	0	0	1.00
LV hypertrophy (Sokolow-Lyon Index)	1 (1)	0	1 (2)	0.333
Left axis deviation	8 (9)	1 (3)	7 (12)	0.260
Left anterior fascicular block	5 (6)	0	5 (9)	0.103
Left atrial enlargement	10 (12)	2 (7)	8 (14)	0.485
Strain pattern	1 (1)	0	1 (2)	1.00
Low QRS voltages in limb leads	29 (33)	5 (17)	24 (41)	0.024
TWI in anterolateral leads (V1–V6)	16 (18)	3 (10)	13 (22)	0.243
TWI in lateral leads (V5–V6±V4, I, aVL)	24 (28)	3 (10)	21 (36)	0.011
TWI in inferolateral leads (II, III, aVF+V5–V6±V4 or I, aVL)	17 (20)	2 (7)	15 (26)	0.032

Among 1658 consecutive pts treated in a cardiological service, LAH found in 76 cases (4.58%); 53 cases showed RBBB (3.19%), and 17 cases showed LBBB (1.02%).¹

In a retrospective study 247 cases of LAH were detected among a presumable healthy population of 8915 individuals engaged in civilian flying activities in Argentina (prevalence, 2.77%).²

1. Rosenbaum MB, Elizari MV, Lázari JO. The Hemiblocks. Oldsmar, Florida: Tampa Tracings; 1970.

2. Canaveris G, Halpern MS, Elizari MV. Intraventricular conduction disturbances in civilian flying personnel: left anterior hemiblock. Aviat Space Environ Med. 1992;63:292–298.

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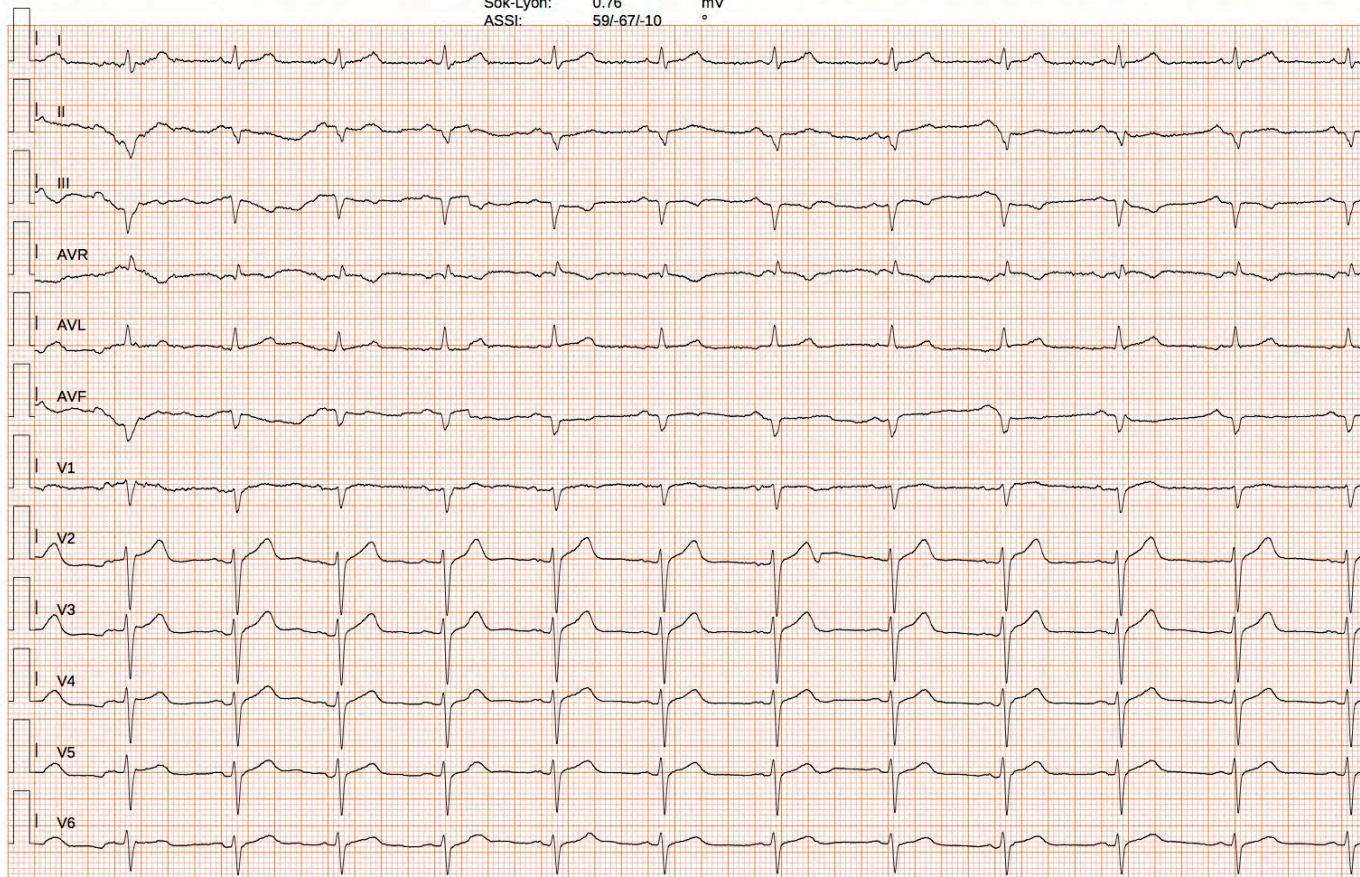
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TECNICO:
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NOTE: -

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FC:	72	bpm
PR:	162	ms
QRSd:	80	ms
QT/QTc:	370/391	ms
QTcB:	405	ms
QTcF:	393	ms
Rv5-6/Sv1	0.34/0.42	mV
Sok-Lyon:	0.76	mV
ASSI:	59/67/10	°

Rs a 72 bpm conduzione AV nei limiti, emblocco anteriore sinistro. Lievi anomalie aspecifiche della fase di RV in sede inferiore. QTc nei limiti

Confermato: CICOGNAFRANCESCO 13/06/2023 11:12

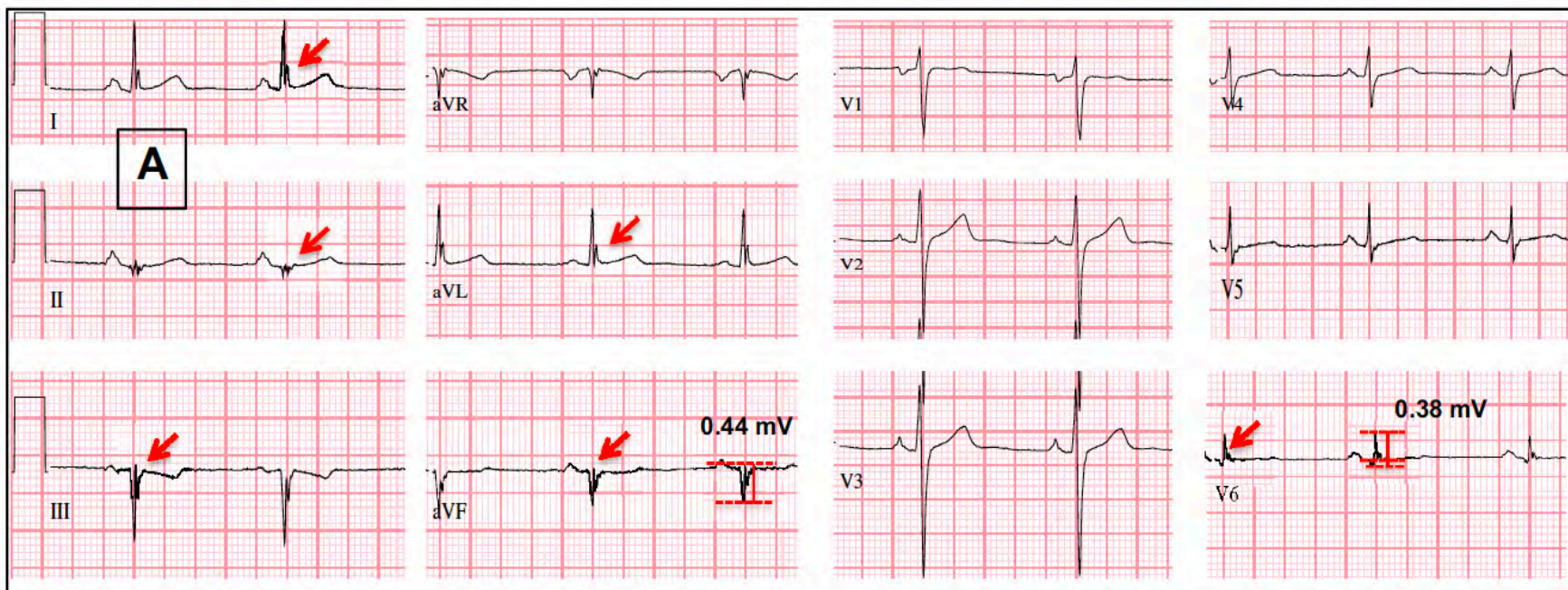


Sinus rhythm QRS amplitude and fractionation in patients with nonischemic cardiomyopathy to identify ventricular tachycardia substrate and location

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Heart Rhythm, Vol 19, No 2, February 2022



Sinus Rhythm ECG Criteria Associated with Basal-Lateral Ventricular Tachycardia Substrate in Patients with Nonischemic Cardiomyopathy

WENDY S. TZOU, M.D., ERICA S. ZADO, P.A.-C., DAVID LIN, M.D., DAVID J. CALLANS, M.D., SANJAY DIXIT, M.D., JOSHUA M. COOPER, M.D., RUPA BALA, M.D., FERMIN GARCIA, M.D.,

MATHEW D. F.
EDWARD

NICM and VT (A and B)

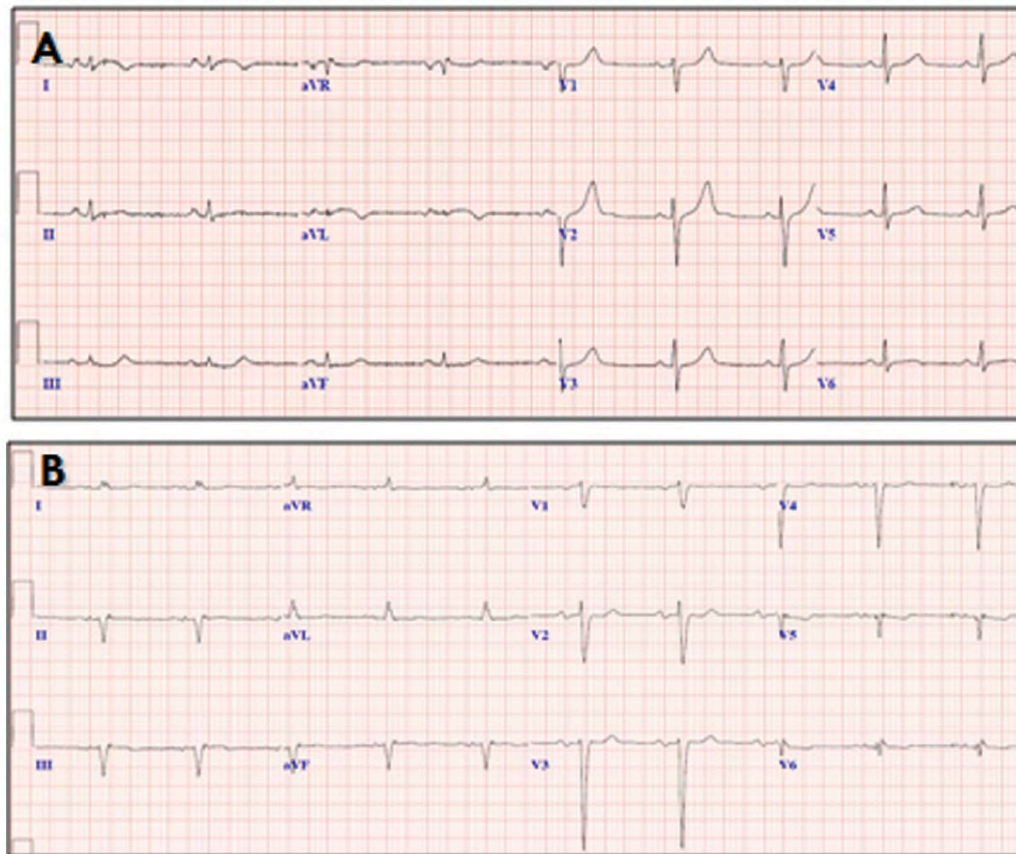
AT DEO, M.D.,
KI, M.D.

From the Section of

ECG Criteria
thy (NICM) and
sought to determ
basal-lateral scar

Methods and R
lateral LV low vo
with NICM, and
atrial rhythm EC
prospectively, bli
and endocardial/
R and R:S ratio,
VT. Controlling f
V6 S ≥ 0.15 mV (
In Phase II, the f
sensitivity and sp

Conclusions: A
V6 S ≥ 0.15 mV
information may have
patients. (*J Cardiovasc Electrophysiol*, Vol. 22, pp. 1351-1358, December 2011)

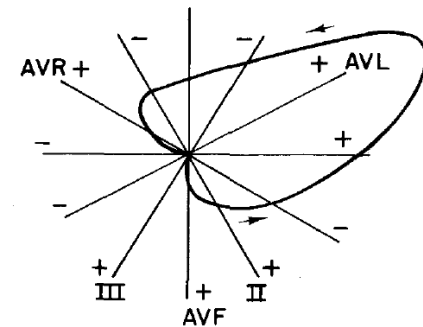
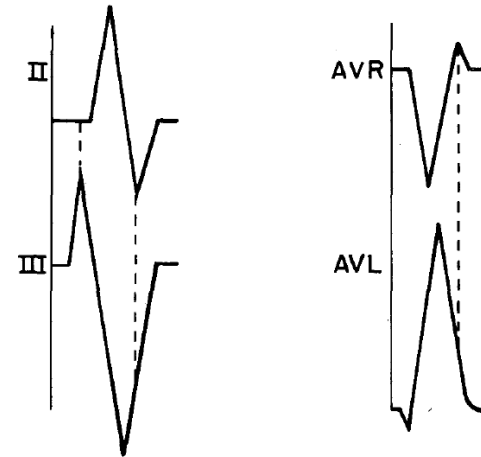
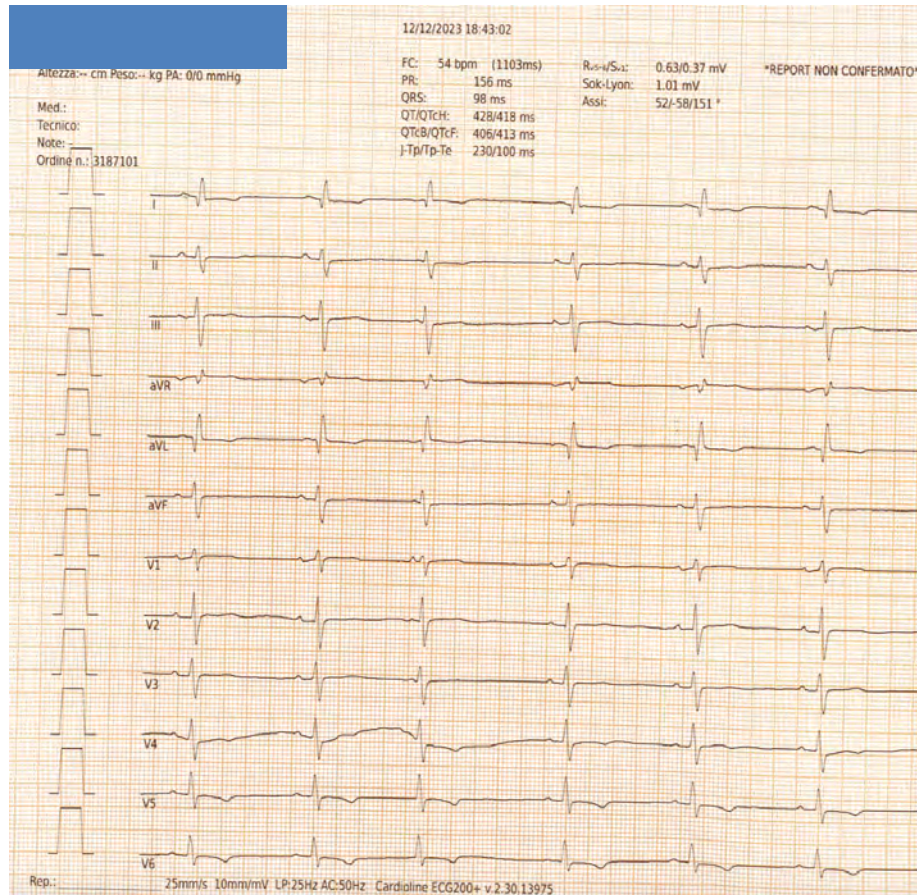


ital of the University

nic cardiomyopa-
ventricle (LV). We
CM patients with

/epicardial basal-
controls (n = 18)
ner sinus or paced
ria were evaluated
patients had VT
ristics studied, V1
al-scar associated
V (P = 0.001) and
basal-lateral scar.
-lateral scar, with

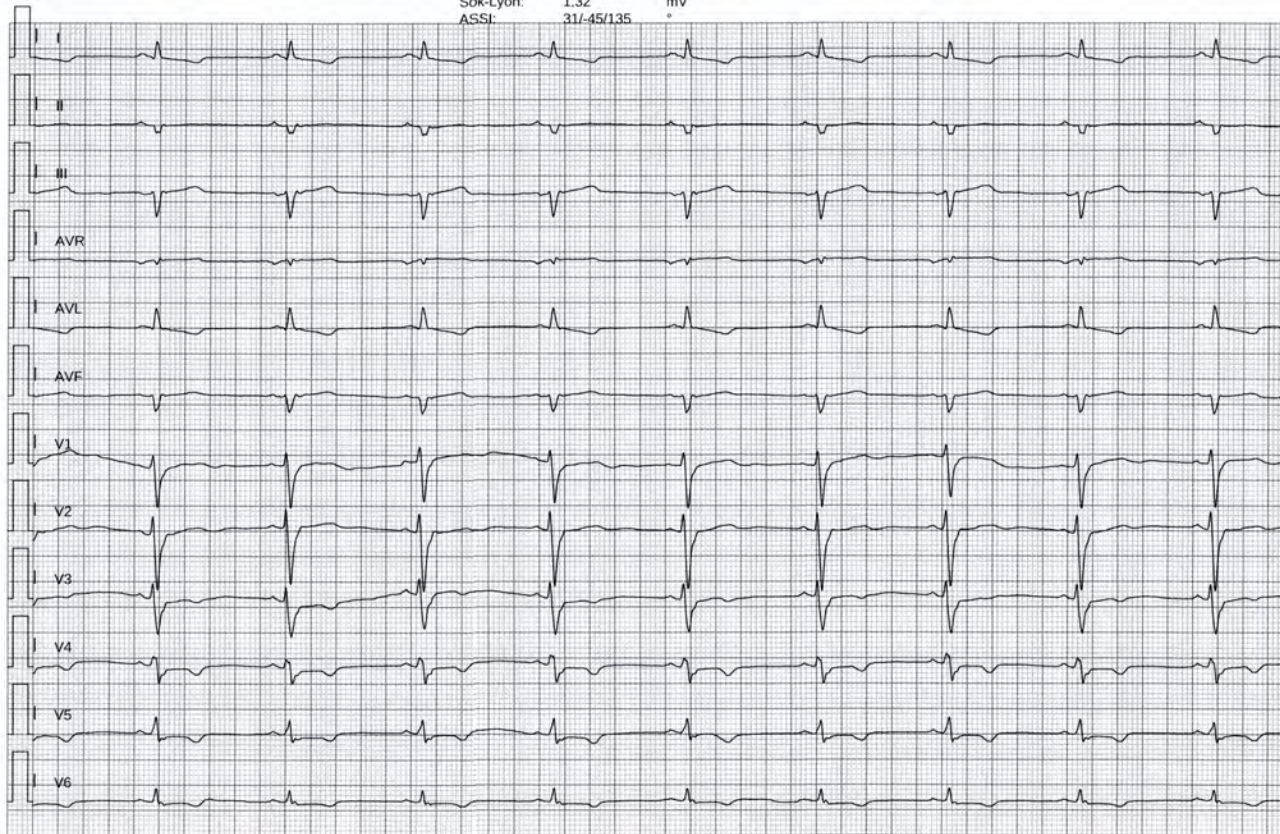
≥ 0.15 mV and
This ECG infor-
for VT in NICM



In a typical LAFB, an R wave always observed in leads II, III and aVF due to early activation of the left ventricle in the inferior wall, supplied by the left posterior fascicle.

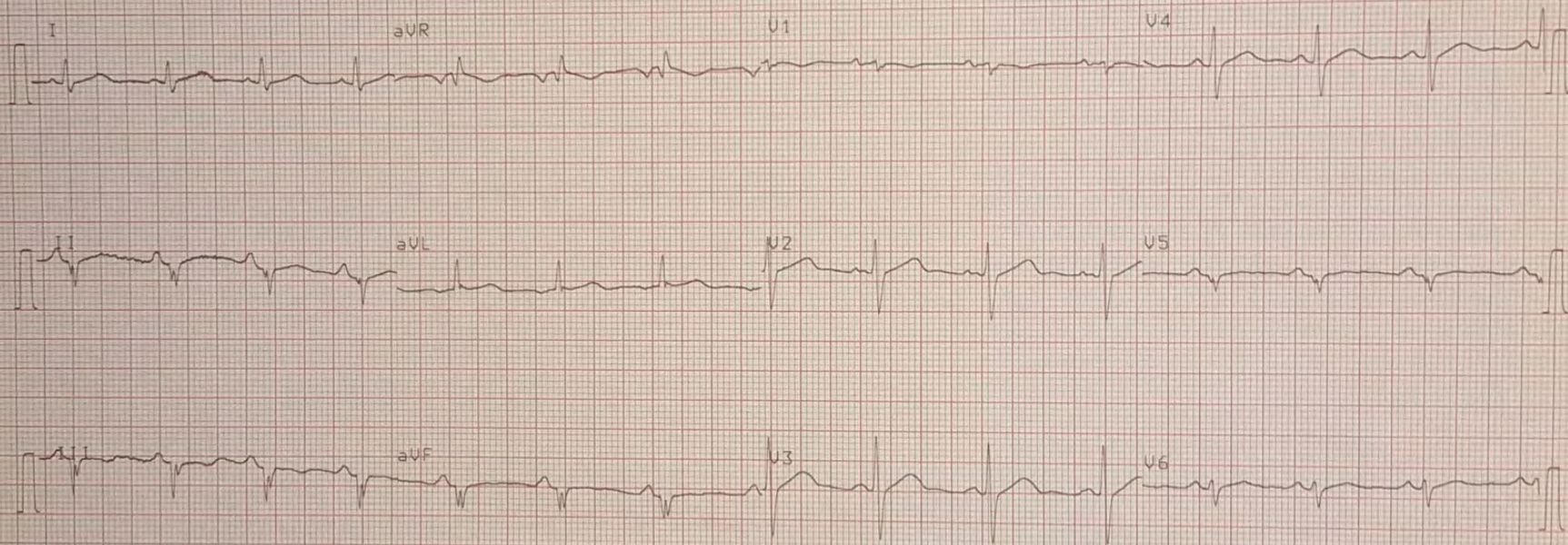
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PR:	130	ms	Non Confermato
QRSd:	132	ms	
QT/QTc:	458/453	ms	
QTcB:	446	ms	
QTcF:	450	ms	
Rv5-6/Sv1	0.41/0.91	mV	
Sokol-Lyon:	1.32	mV	
ASSI:	31/-45/135	°	



10 mm/mV

10 mm/mV



**Left anterior fascicular block
and Q-wave in inferior leads**

True LAFB ?

DIAGNOSIS OF LEFT ANTERIOR
HEMIBLOCK AND LEFT POSTERIOR
HEMIBLOCK IN THE PRESENCE OF
INFERIOR WALL MYOCARDIAL
INFARCTION*

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University of Miami School of Medicine
Miami, Fla.

Bull. N. Y. Acad. Med. Vol. 47, No. 8, August 1971

Inferior infarction without left anterior fascicular block

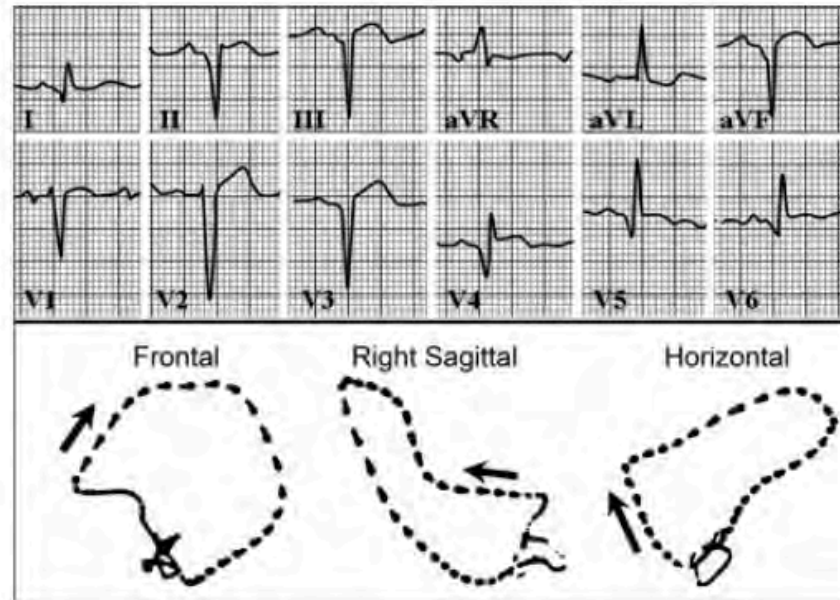
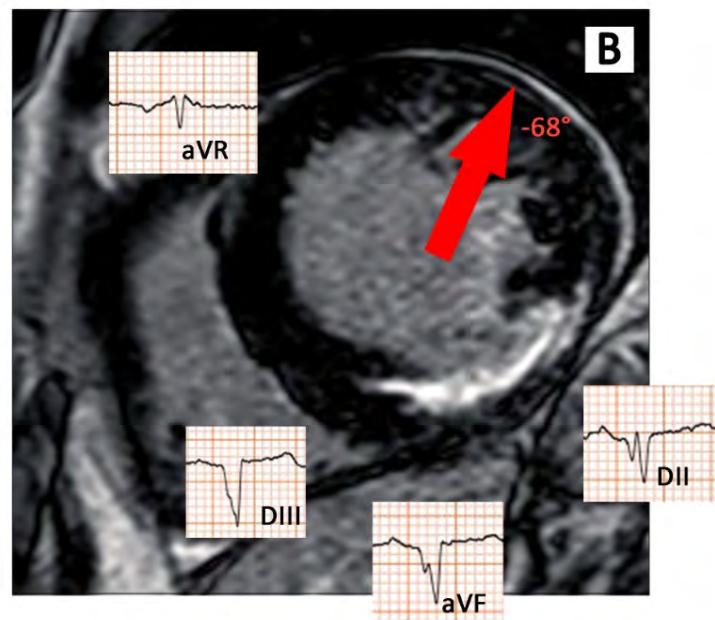
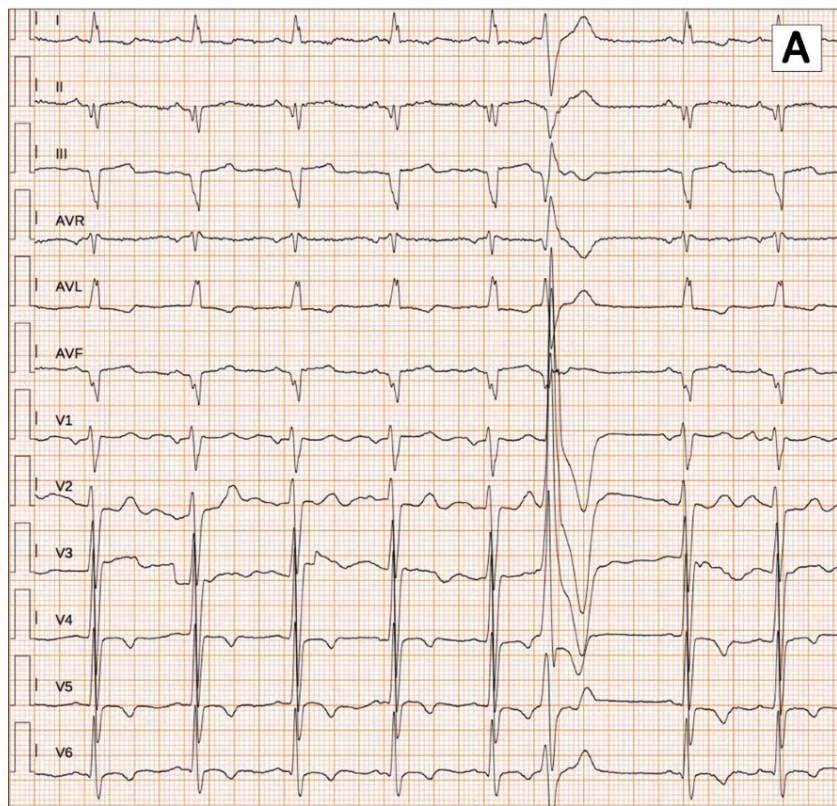


Figure 7. Anterolateral and inferior wall myocardial infarction that simulates LAH. Top, The ECG shows a QRS interval of 110 milliseconds. Bottom, Clockwise rotation of the QRS loop in the frontal plane rules out the presence of LAH.

L'onda R scomparirà totalmente o parzialmente ogni volta che una zona fibrotica coinvolge le aree di attivazione precoce nella parete inferiore ventricolare sinistra. Al contrario, se la fibrosi risparmia queste aree, si verificherà un'onda R iniziale nelle derivazioni inferiori.



**Inferior infarction with
left anterior fascicular block**

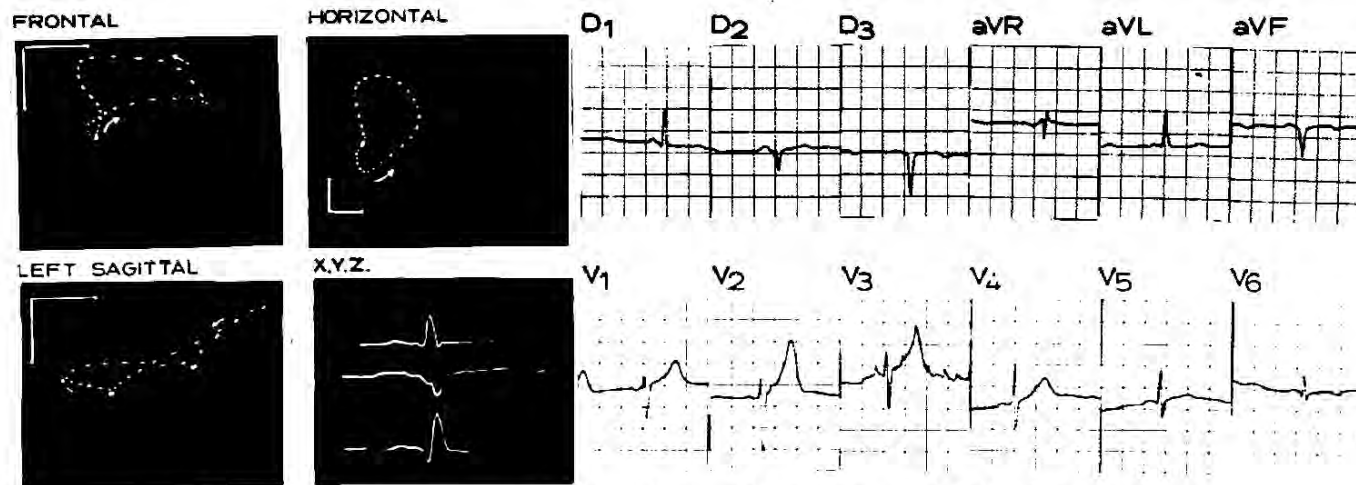
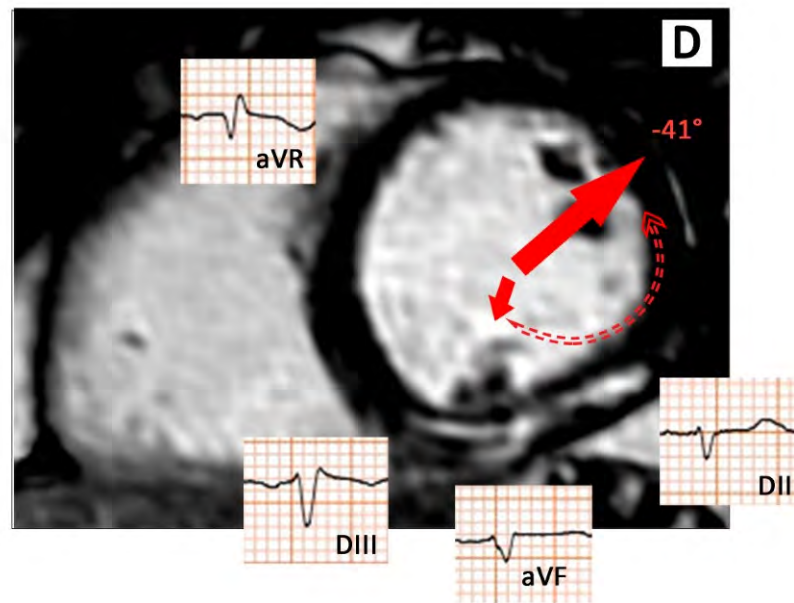
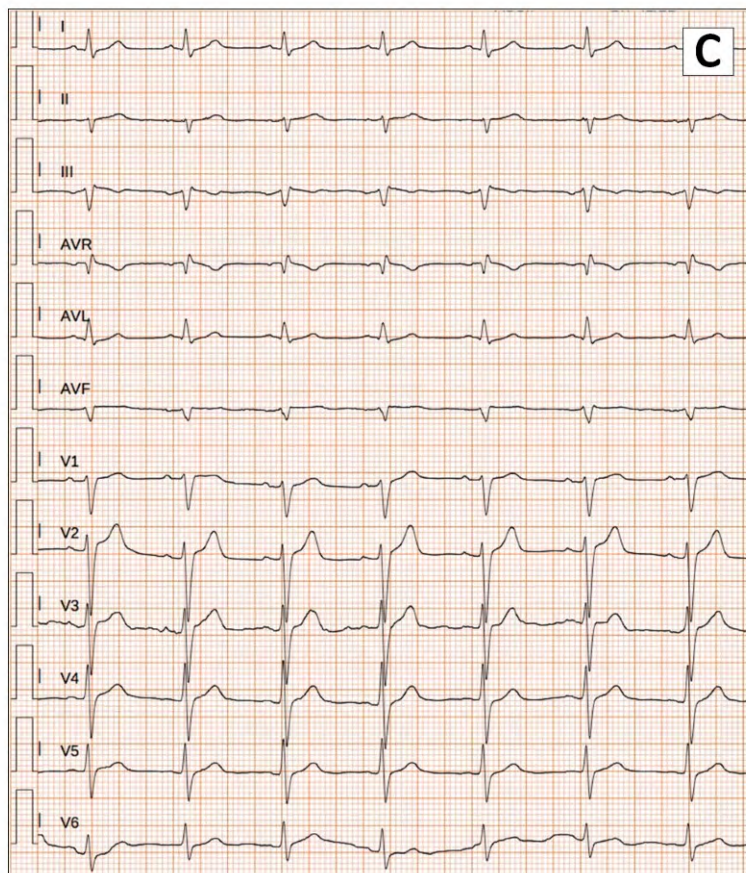
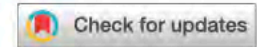


FIGURE 2. Electrocardiogram and vectorcardiogram of a patient aged 53 with inferior infarction and left axis deviation. In the frontal plane, the initial forces are superiorly displaced and clockwise rotated. At about 40 msec, there is an abrupt shift of the loop. The second half of QRS is located superior to the efferent limb and turns in a counterclockwise fashion. The loop is interrupted every 1/400 sec. Calibration 500 μ V.

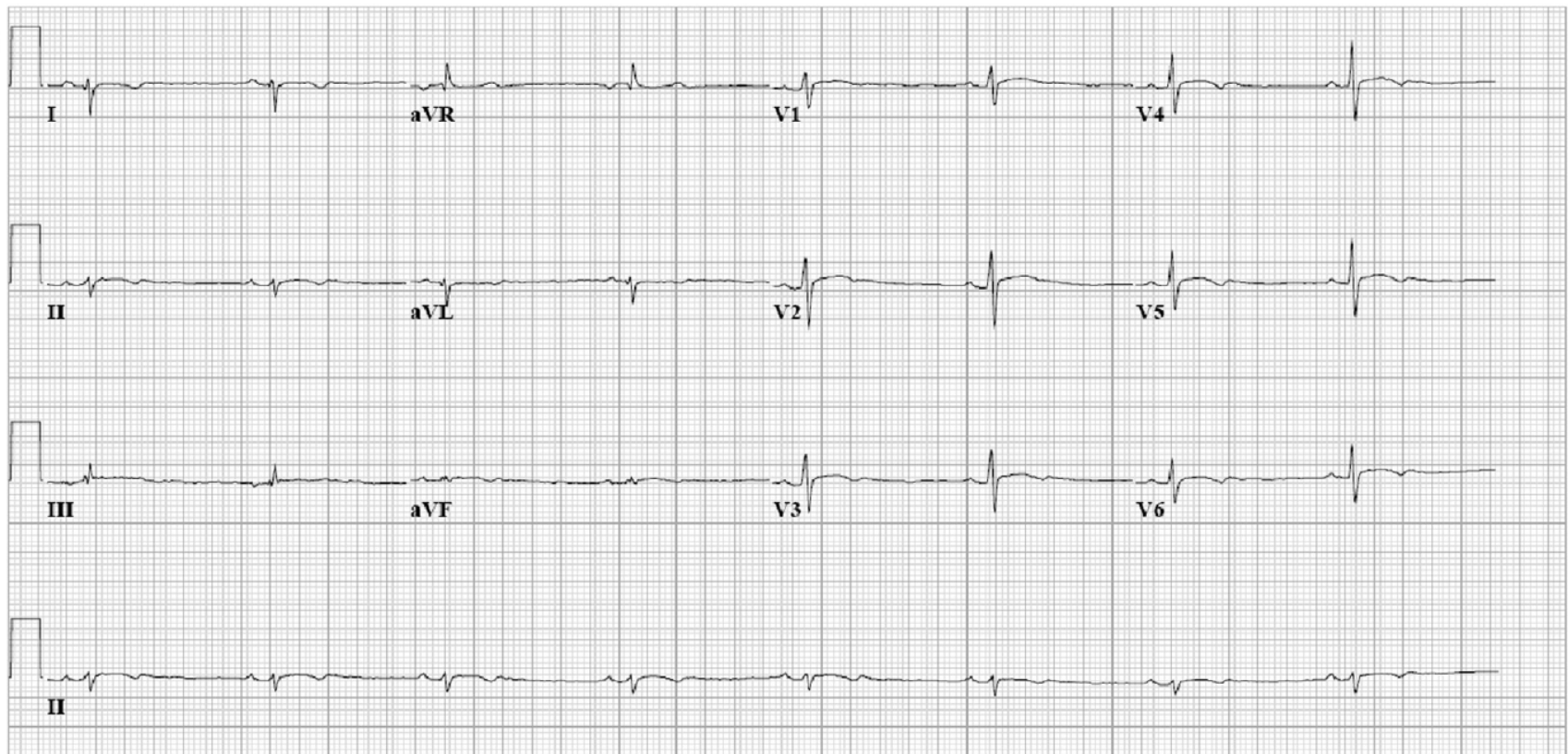


Late evolution of arrhythmogenic cardiomyopathy in patients with initial presentation as idiopathic ventricular fibrillation

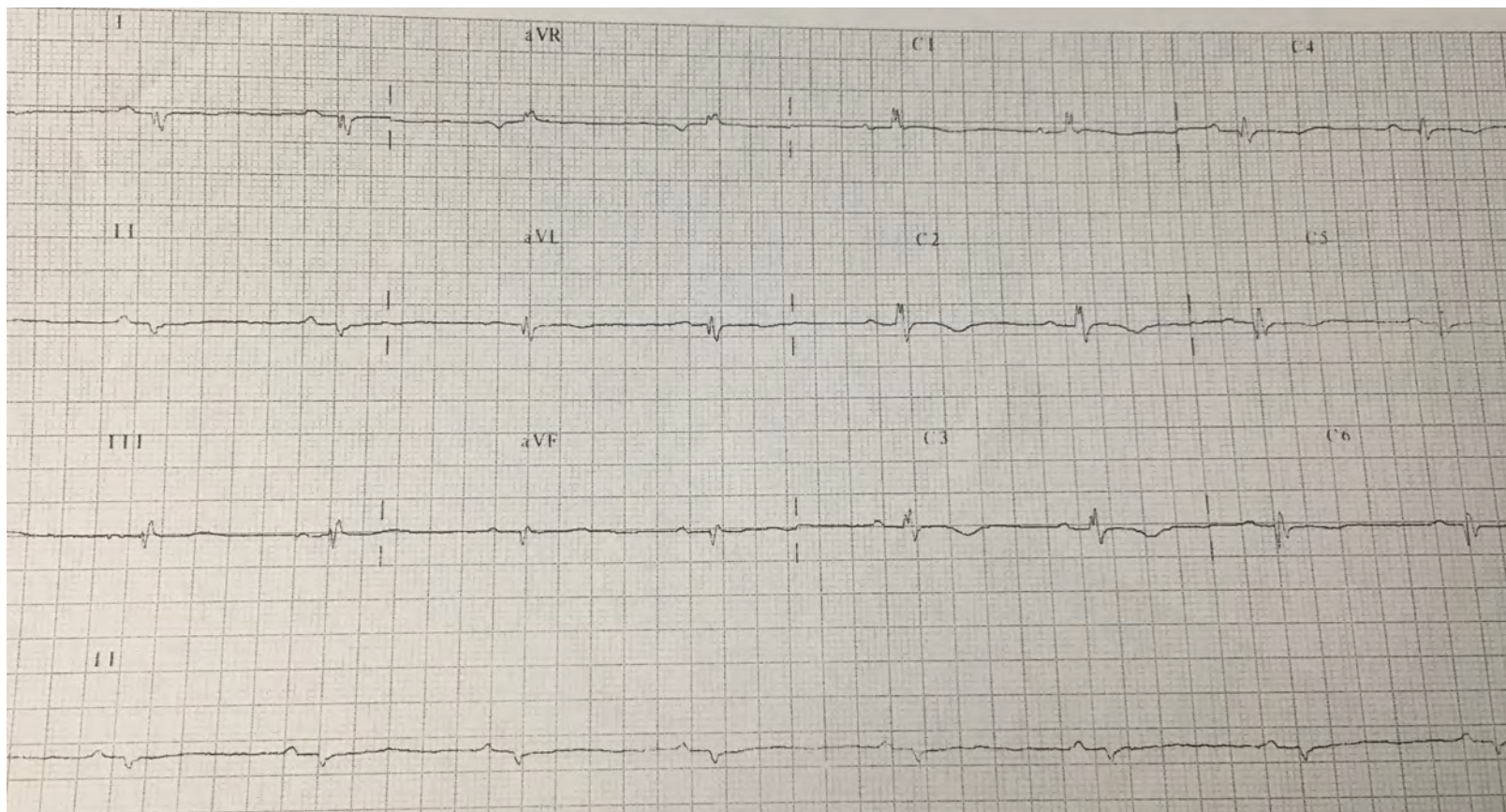


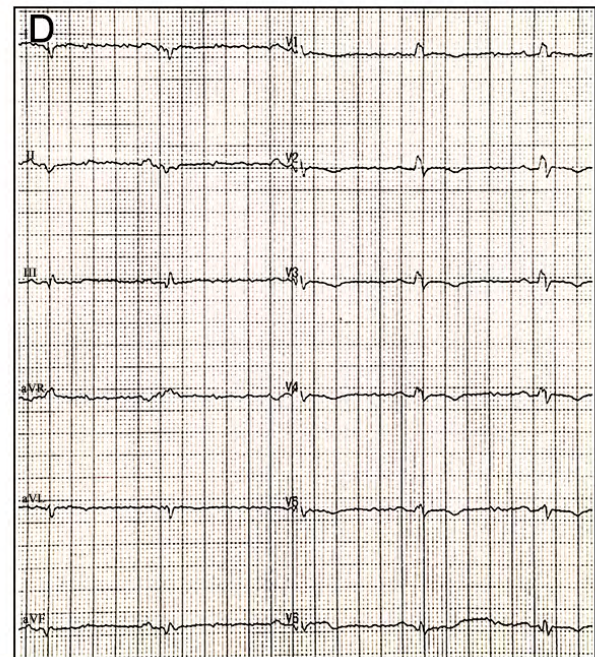
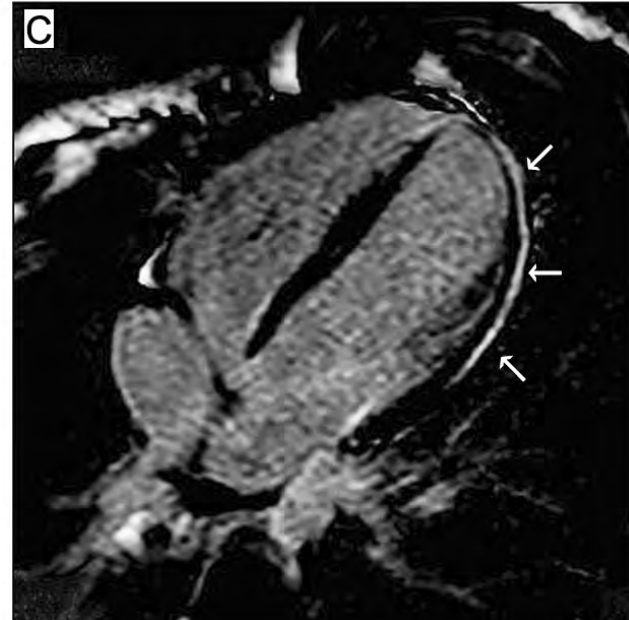
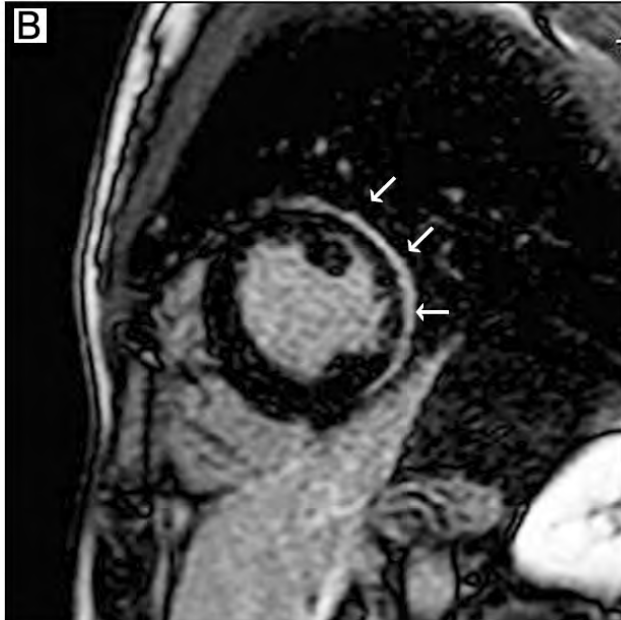
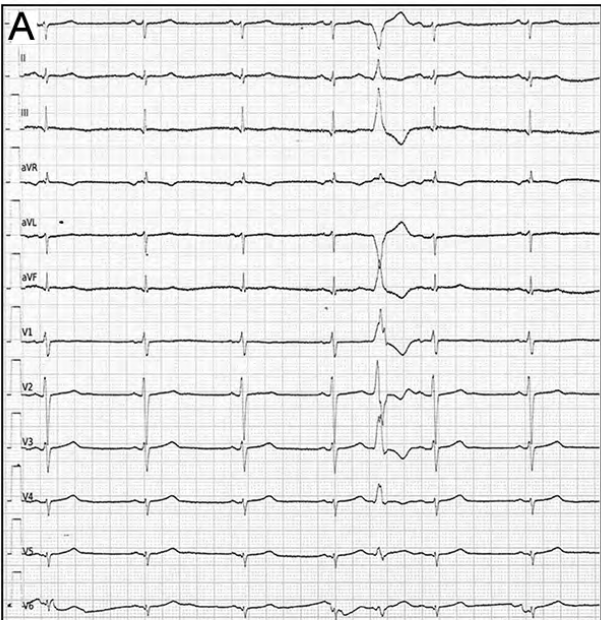
Lennart J. Blom, MD,^{*} Anneline S.J.M. Te Riele, MD, PhD,^{*} Aryan Vink, MD, PhD,[†]
Richard N.W. Hauer, MD, PhD,^{*} Rutger J. Hassink, MD, PhD^{*}

A



25mm/s 10mm/mV 150Hz 8.0 SP2 12SL 240 CID: 127





The tetrafascicular nature of the intraventricular conduction system

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²Coronary Center of the Hospital de Messejana Dr. Carlos Alberto Studart Gomes, Fortaleza, Brazil

³Heart Center, Tampere University Hospital and Faculty of Medicine and Life Sciences, University of Tampere, Tampere, Finland

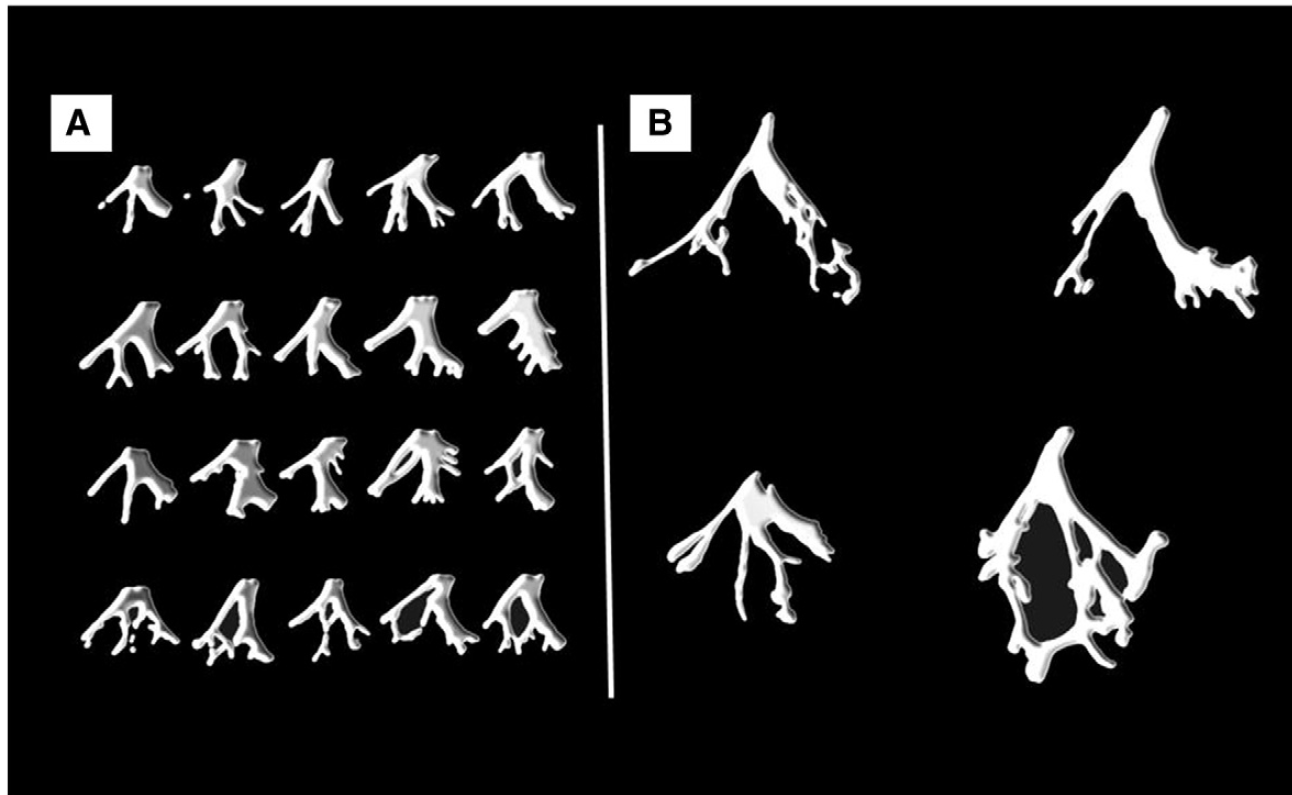
Correspondence

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Email: riera@uol.com.br

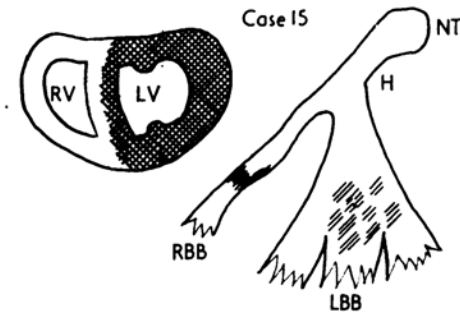
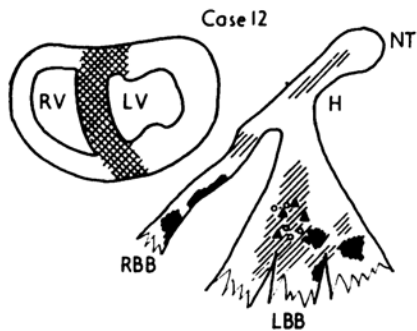
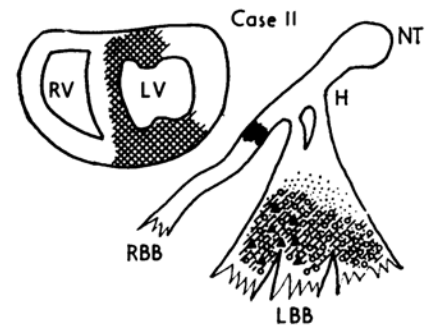
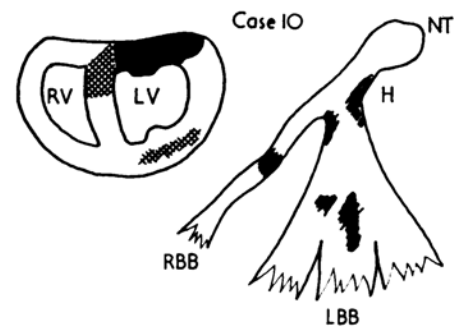
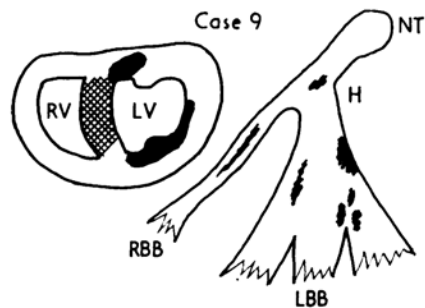
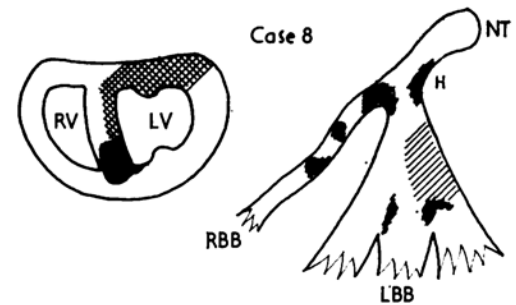
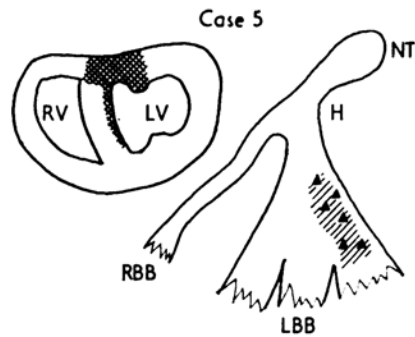
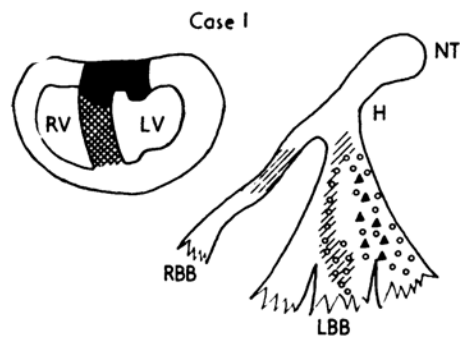
The existence of a tetrafascicular intraventricular conduction system remains debatable. A consensus statement ended up with some discrepancies and, despite agreeing on the possible existence of an anatomical left septal fascicle, the electrocardiographic and vectorcardiographic characteristics of left septal fascicular block (LSFB) were not universally accepted. The most important criteria requested to confirm the existence of LSFB is its intermittent nature. So far, our group has published cases of transient ischemia-induced LSFB and phase 4 or bradycardia-dependent LSFB. Finally, anatomical, anatomopathological, histological, histopathological, electrocardiographic, vectorcardiographic, body surface potential mapping, and electrophysiology studies support the fact that the left bundle branch divides into three fascicles or a “fan-like interconnected network.”

KEYWORDS

intraventricular conduction system, left septal fascicle, left septal fascicular block



A. Fig. 2 from Demoulin JC, Kulbertus HE [24]. Diagrammatic sketches of the LBB conduction system in twenty human normal hearts. These sketches depict the anatomy from serial histologic sections of left septal myocardium. The LBB and its subdivisions were identified by their subendocardial location and histological features typical of the conducting fibers. B: Figure 28, Chapter 2 from Rosenbaum MB et al. [13]. Four human LBB systems dissected and separated from the heart considered the main prototypes observed in our material. In every case, the main LBB is short and its divisions longer depicting a wider posterior division as compared with the anterior one. The bottom left LBB shows what can be considered a medial left septal fascicle arising at the bifurcation of the LBB, although it actually emerges from a wide posterior division, a pattern that can also be observed in the examples of Fig. 14 A in most cases.



Conduction system lesions

Cloudy swelling, vacuolization

Leucocytic infiltration

Haemorrhage

Acute necrosis

Chronic (sclerosis, atrophy)

Myocardial infarction

Acute & subacute stage

Chronic stage

RV=Right ventricle; LV=Left ventricle

NT: Node of Tawara

H: His bundle

RBB: Right bundle-branch

LBB: Left

Electrophysiological abnormalities precede overt structural changes in arrhythmogenic right ventricular cardiomyopathy due to mutations in desmoplakin-A combined murine and human study

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¹Department of Medicine, UCL, London, UK; ²Institute of Cardiovascular Sciences, UCL, London, UK; ³Columbia University, NY, USA; ⁴Beth Israel Deaconess Medical Centre, Boston, MA, USA; ⁵William Harvey Heart Centre, London, UK; and ⁶King's College, London, UK

Received 28 July 2011; revised 2 November 2011; accepted 1 December 2011; online publish-ahead-of-print 11 January 2012

Aims

Anecdotal observations suggest that sub-clinical electrophysiological manifestations of arrhythmogenic right ventricular cardiomyopathy (ARVC) develop before detectable structural changes ensue on cardiac imaging. To test this hypothesis, we investigated a murine model with conditional cardiac genetic deletion of one desmoplakin allele (DSP[±]) and compared the findings to patients with non-diagnostic features of ARVC who carried mutations in desmoplakin.

Methods and results

Murine: the DSP[±] mice underwent electrophysiological, echocardiographic, and immunohistochemical studies. They had normal echocardiograms but delayed conduction and inducible ventricular tachycardia associated with mislocalization and reduced intercalated disc expression of Cx43. Sodium current density and myocardial histology were normal at 2 months of age. **Human:** ten patients with heterozygous mutations in DSP without overt structural heart disease (DSP+) and 12 controls with supraventricular tachycardia were studied by high-density electrophysiological mapping of the right ventricle. Using a standard S₁–S₂ protocol, restitution curves of local conduction and repolarization parameters were constructed. Significantly greater mean increases in delay were identified particularly in the outflow tract vs. controls ($P < 0.01$) coupled with more uniform wavefront progression. The odds of a segment with a maximal activation–repolarization interval restitution slope > 1 was 99% higher (95% CI: 13%; 351%, $P = 0.017$) in DSP+ vs. controls. Immunostaining revealed Cx43 mislocalization and variable Na channel distribution.

Conclusion

Desmoplakin disease causes connexin mislocalization in the mouse and man preceding any overt histological abnormalities resulting in significant alterations in conduction–repolarization kinetics prior to morphological changes detectable on conventional cardiac imaging. Haploinsufficiency of desmoplakin is sufficient to cause significant Cx43 mislocalization. Changes in sodium current density and histological abnormalities may contribute to a worsening phenotype or disease but are not necessary to generate an arrhythmogenic substrate. This has important implications for the earlier diagnosis of ARVC and risk stratification.

Keywords

Arrhythmia • Conduction • ARVC • Repolarization • Desmosome • Desmoplakin

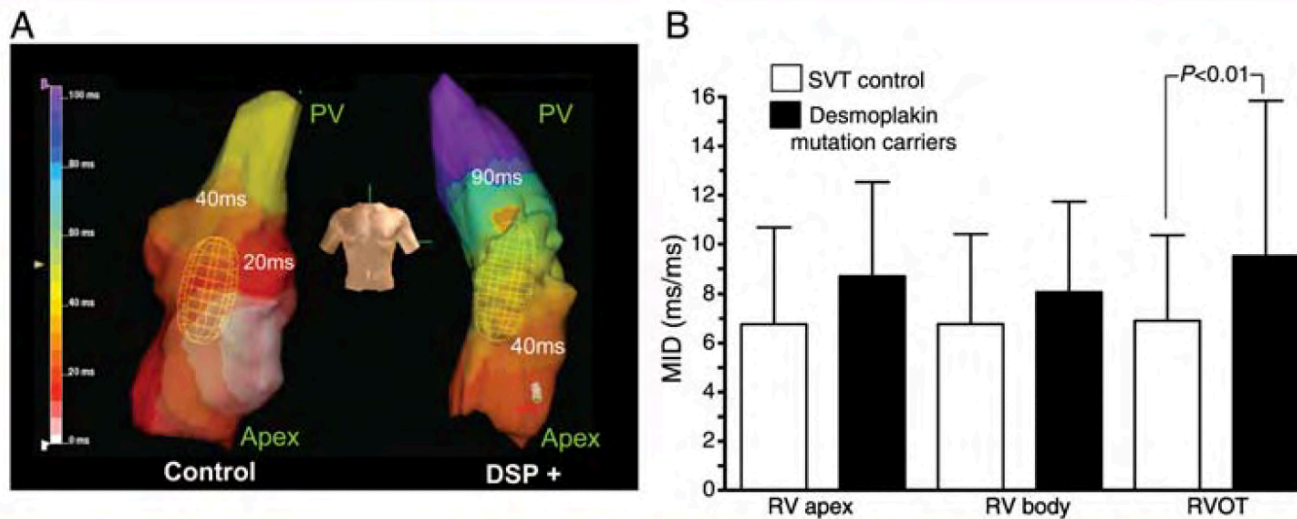
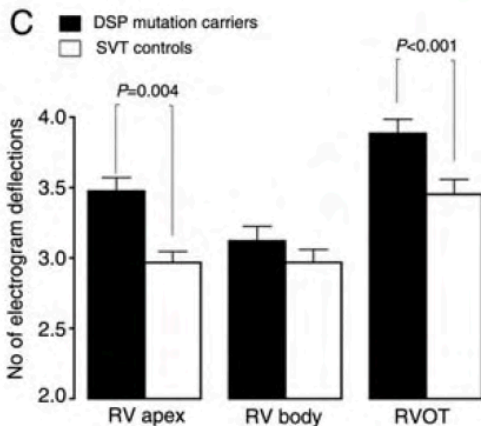
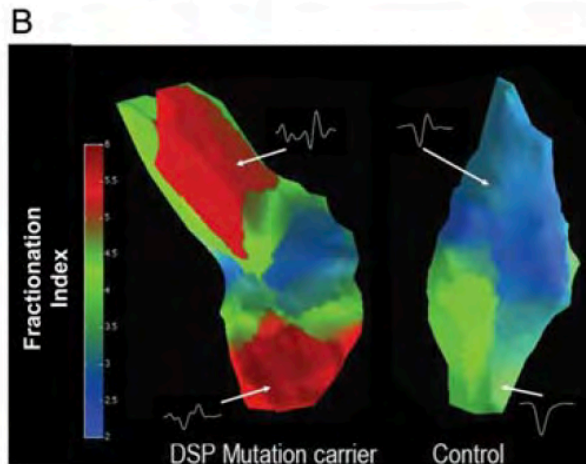
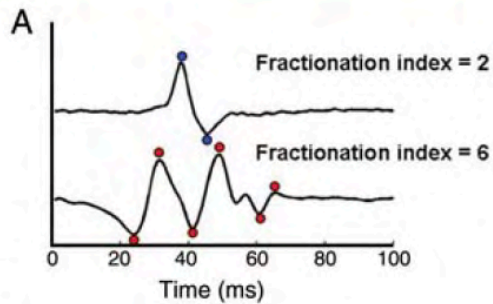


Figure 4 (A) Isochronal map showing RV activation sequence with steady-state pacing from RV apex at 400 ms cycle length (subject 5 vs. control). PV, pulmonary valve. (B) Mean increase delay according to the region in DSP+ and control groups.



In this study of desmoplakin disease, subclinical electrophysiological effects precede any overt structural changes in a murine model and in patients.

These data may explain early phase of the disease when ventricular tachyarrhythmias and sudden death occur at short coupling intervals during stress, exercise, or sports activity (increased adrenergic drive) due to the initial reduction in conduction reserve.

During later phases of ARVC when demonstrable structural changes ensue (fibrosis and myocyte atrophy), ventricular arrhythmias are more common at rest as a monomorphic VT.

Fibrosis, Connexin-43, and Conduction Abnormalities in the Brugada Syndrome



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ABSTRACT

BACKGROUND The right ventricular outflow tract (RVOT) is acknowledged to be responsible for arrhythmogenesis in Brugada syndrome (BrS), but the pathophysiology remains controversial.

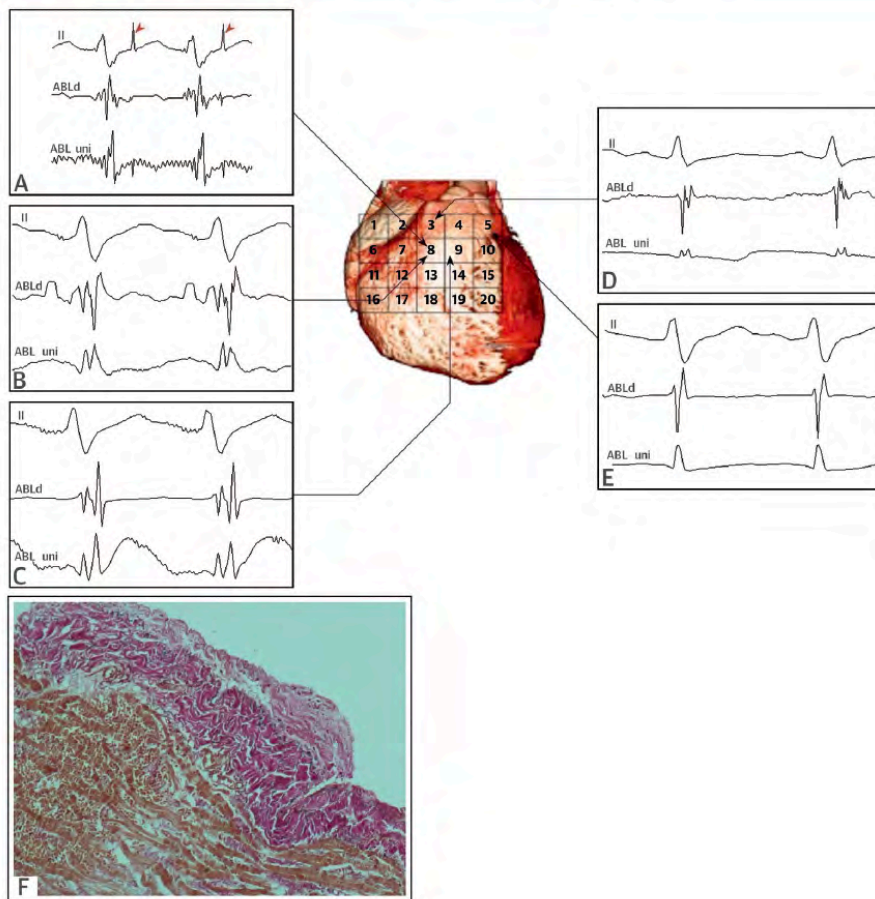
OBJECTIVES This study assessed the substrate underlying BrS at post-mortem and in vivo, and the role for open thoracotomy ablation.

METHODS Six whole hearts from male post-mortem cases of unexplained sudden death (mean age 23.2 years) with negative specialist cardiac autopsy and familial BrS were used and matched to 6 homograft control hearts by sex and age (within 3 years) by random risk set sampling. Cardiac autopsy sections from cases and control hearts were stained with picrosirius red for collagen. The RVOT was evaluated in detail, including immunofluorescent stain for connexin-43 (Cx43). Collagen and Cx43 were quantified digitally and compared. An in vivo study was undertaken on 6 consecutive BrS patients (mean age 39.8 years, all men) during epicardial RVOT ablation for arrhythmia via thoracotomy. Abnormal late and fractionated potentials indicative of slowed conduction were identified, and biopsies were taken before ablation.

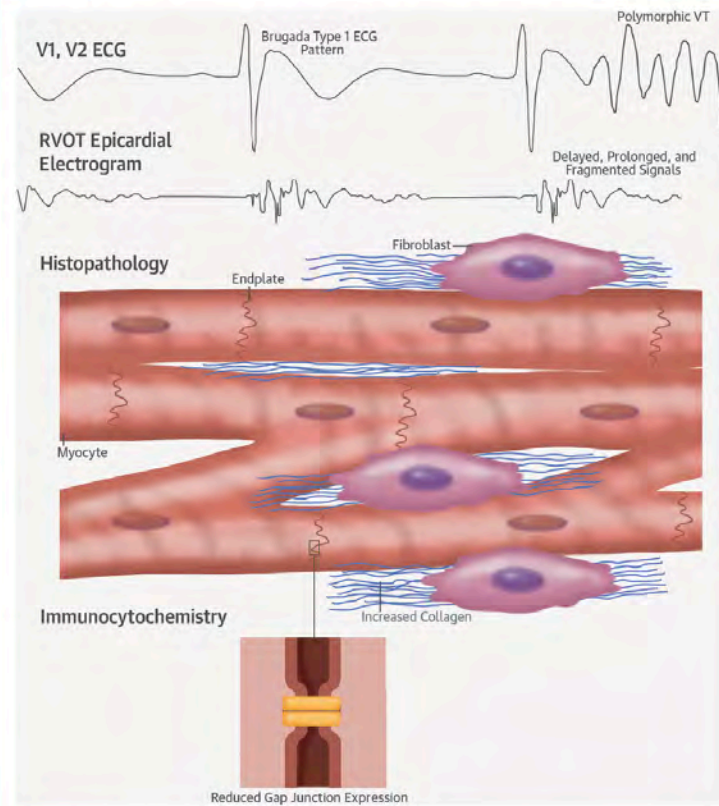
RESULTS Collagen was increased in BrS autopsy cases compared with control hearts (odds ratio [OR]: 1.42; $p = 0.026$). Fibrosis was greatest in the RVOT (OR: 1.98; $p = 0.003$) and the epicardium (OR: 2.00; $p = 0.001$). The Cx43 signal was reduced in BrS RVOT (OR: 0.59; $p = 0.001$). Autopsy and in vivo RVOT samples identified epicardial and interstitial fibrosis. This was collocated with abnormal potentials in vivo that, when ablated, abolished the type 1 Brugada electrocardiogram without ventricular arrhythmia over 24.6 ± 9.7 months.

CONCLUSIONS BrS is associated with epicardial surface and interstitial fibrosis and reduced gap junction expression in the RVOT. This collocates to abnormal potentials, and their ablation abolishes the BrS phenotype and life-threatening arrhythmias. BrS is also associated with increased collagen throughout the heart. Abnormal myocardial structure and conduction are therefore responsible for BrS. (J Am Coll Cardiol 2015;66:1976-86) © 2015 by the American College of Cardiology Foundation.

FIGURE 2 Computed Tomography Scan, Epicardial Electrograms, and Histology of RVOT of In Vivo BrS Patient



CENTRAL ILLUSTRATION Pathophysiology of Brugada Syndrome: Conduction Delay Due to Fibrosis and Connexin-43 Abnormalities



Nademanee, K. et al. J Am Coll Cardiol. 2015; 66(18):1976-86.

Conduction delay in the right ventricular outflow tract (RVOT) is caused by myocyte electrical uncoupling due to a reduction in connexin-43 at endplates and subtle interstitial and replacement fibrosis. As a result, epicardial electrograms are abnormal, slowed, and fragmented. This provides the substrate for the Brugada type 1 electrocardiographic (ECG) pattern, re-entry, and the generation of polymorphic ventricular tachycardia (VT) and ventricular fibrillation.

ORIGINAL RESEARCH

VENTRICULAR TACHYCARDIA - PATHOPHYSIOLOGY

Functional Epicardial Conduction Disturbances Due to a SCN5A Variant Associated With Brugada Syndrome



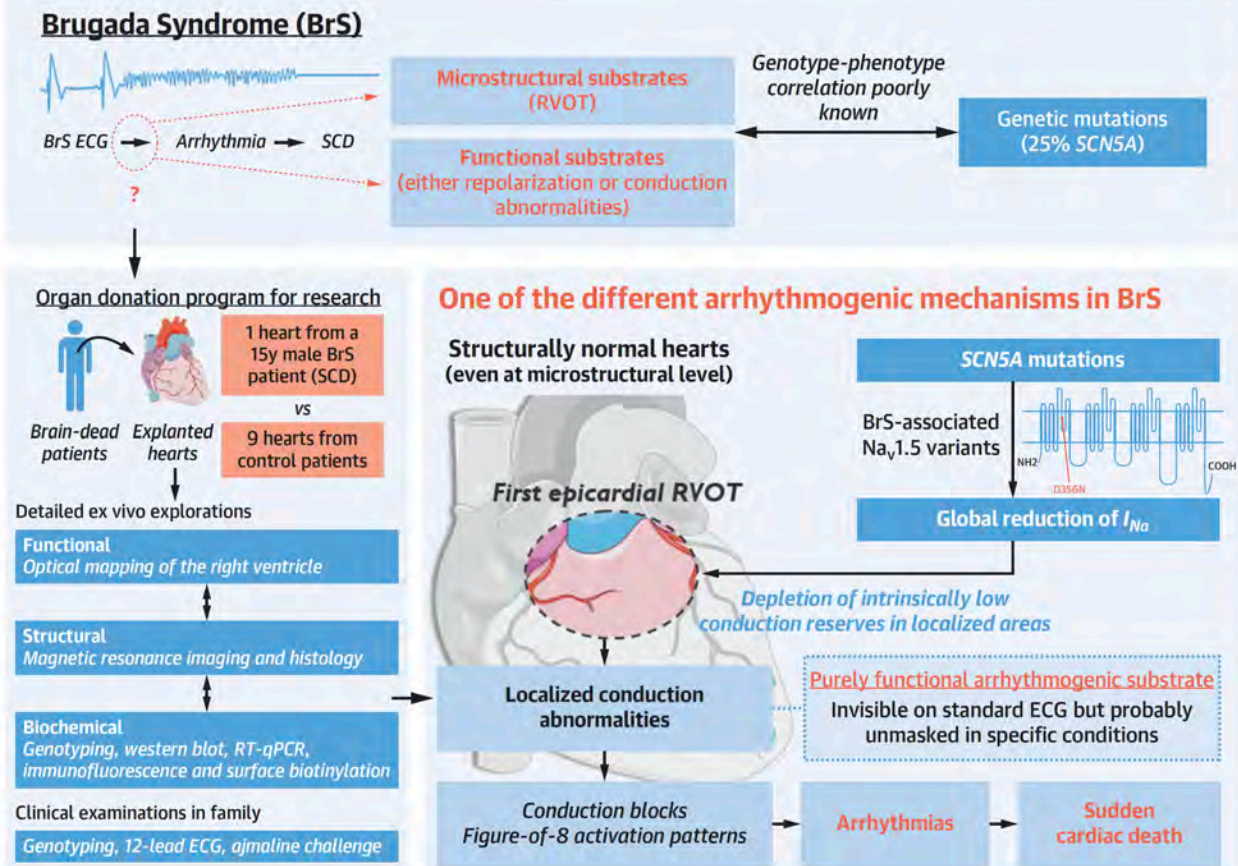
Estelle Renard, PhD,^{a,b} Richard D. Walton, PhD,^{a,b} David Benoist, PhD,^{a,b} Fabien Brette, PhD,^{a,b} Gilles Bru-Mercier, PhD,^{a,b} Sébastien Chaigne, PhD,^{a,b} Sabine Charron, MSc,^{a,b} Marion Constantin, MSc,^{a,b} Matthieu Douard, PhD,^{a,b} Virginie Dubes, PhD,^{a,b} Bastien Guillot, PhD,^{a,b} Thomas Hof, PhD,^{a,b} Julie Magat, PhD,^{a,b} Marine E. Martinez, PhD,^{a,b} Cindy Michel, PhD,^{a,b} Néstor Pallares-Lupon, PhD,^{a,b} Philippe Pasdois, PhD,^{a,b} Alice Récalde, PhD,^{a,b} Fanny Vaillant, PhD,^{a,b} Frédéric Sacher, MD,^{a,b,c} Louis Labrousse, MD,^{a,d} Julien Rogier, MD,^e Florence Kyndt, PHARMAD, PhD,^{f,g} Manon Baudic, MSc,^h Jean-Jacques Schott, PhD,^f Julien Barc, PhD,ⁱ Vincent Probst, MD,^f Marine Sarlandie, MSc,^h Céline Marionneau, PhD,^h Jesse L. Ashton, PhD,^j Méléze Hocini, MD,^{a,b,c} Michel Haïssaguerre, MD,^{a,b,c} Olivier Bemus, PhD^{a,b}

METHODS A heart was obtained from a 15-year-old adolescent boy with normal electrocardiogram who experienced SCD. Postmortem genotyping was performed, and clinical examinations were done on first-degree relatives. The right ventricle was optically mapped, followed by high-field magnetic resonance imaging and histology. Connexin-43 and Na_v1.5 were localized by immunofluorescence, and RNA and protein expression levels were studied. HEK-293 cell surface biotinylation assays were performed to examine Na_v1.5 trafficking.

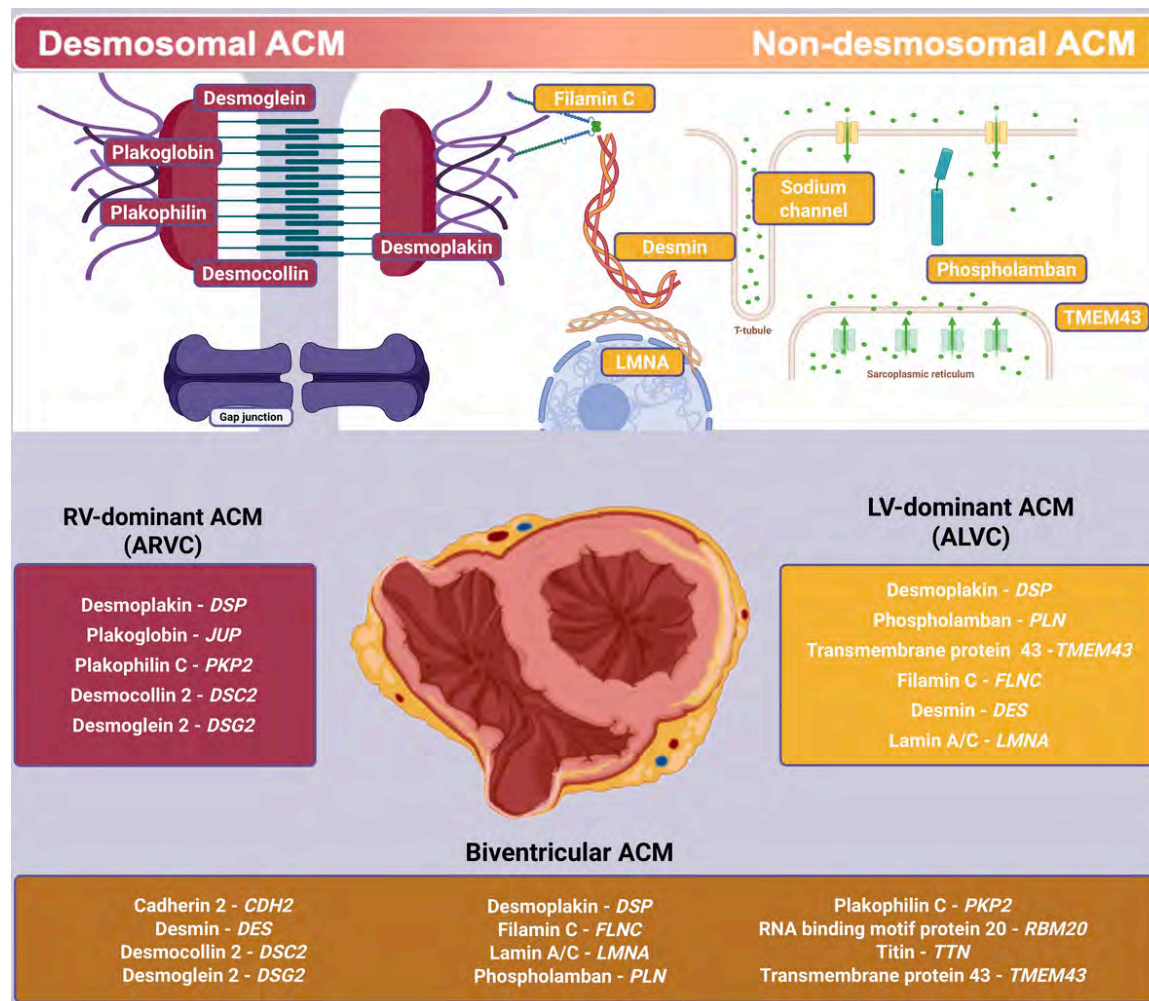
RESULTS A Brugada-related SCD diagnosis was established for the donor because of a *SCN5A* Brugada-related variant (p.D356N) inherited from his mother, together with a concomitant *NKX2.5* variant of unknown significance. Optical mapping demonstrated a localized epicardial region of impaired conduction near the outflow tract, in the absence of repolarization alterations and microstructural defects, leading to conduction blocks and figure-of-8 patterns. Na_v1.5 and connexin-43 localizations were normal in this region, consistent with the finding that the p.D356N variant does not affect the trafficking, nor the expression of Na_v1.5. Trends of decreased Na_v1.5, connexin-43, and desmoglein-2 protein levels were noted; however, the RT-qPCR results suggested that the *NKX2-5* variant was unlikely to be involved.

CONCLUSIONS This study demonstrates for the first time that SCD associated with a Brugada-*SCN5A* variant can be caused by localized functionally, not structurally, impaired conduction. (J Am Coll Cardiol EP 2023;9:1248-1261) © 2023

CENTRAL ILLUSTRATION Localized Functional Conduction Abnormalities as Arrhythmogenic Mechanism in Brugada Syndrome



Renard E, et al. J Am Coll Cardiol EP. 2023;9(8):1248-1261.



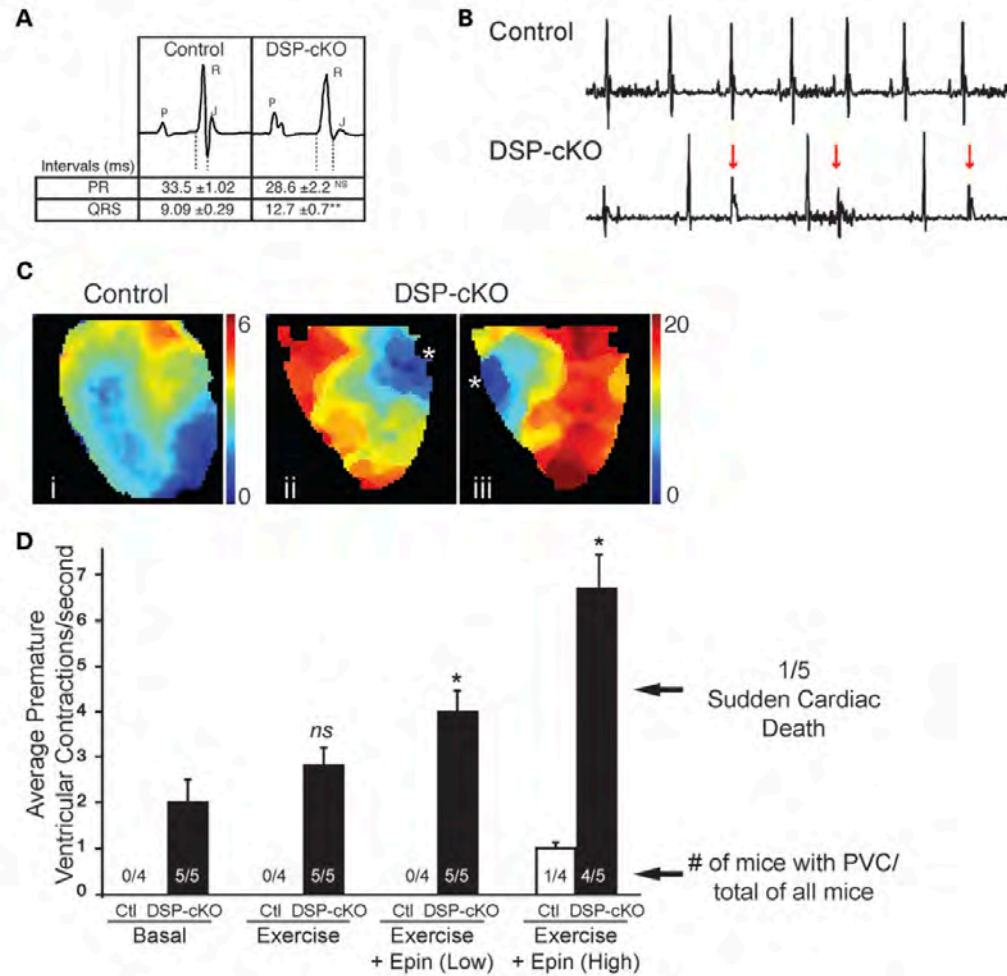
Connexin defects underlie arrhythmogenic right ventricular cardiomyopathy in a novel mouse model

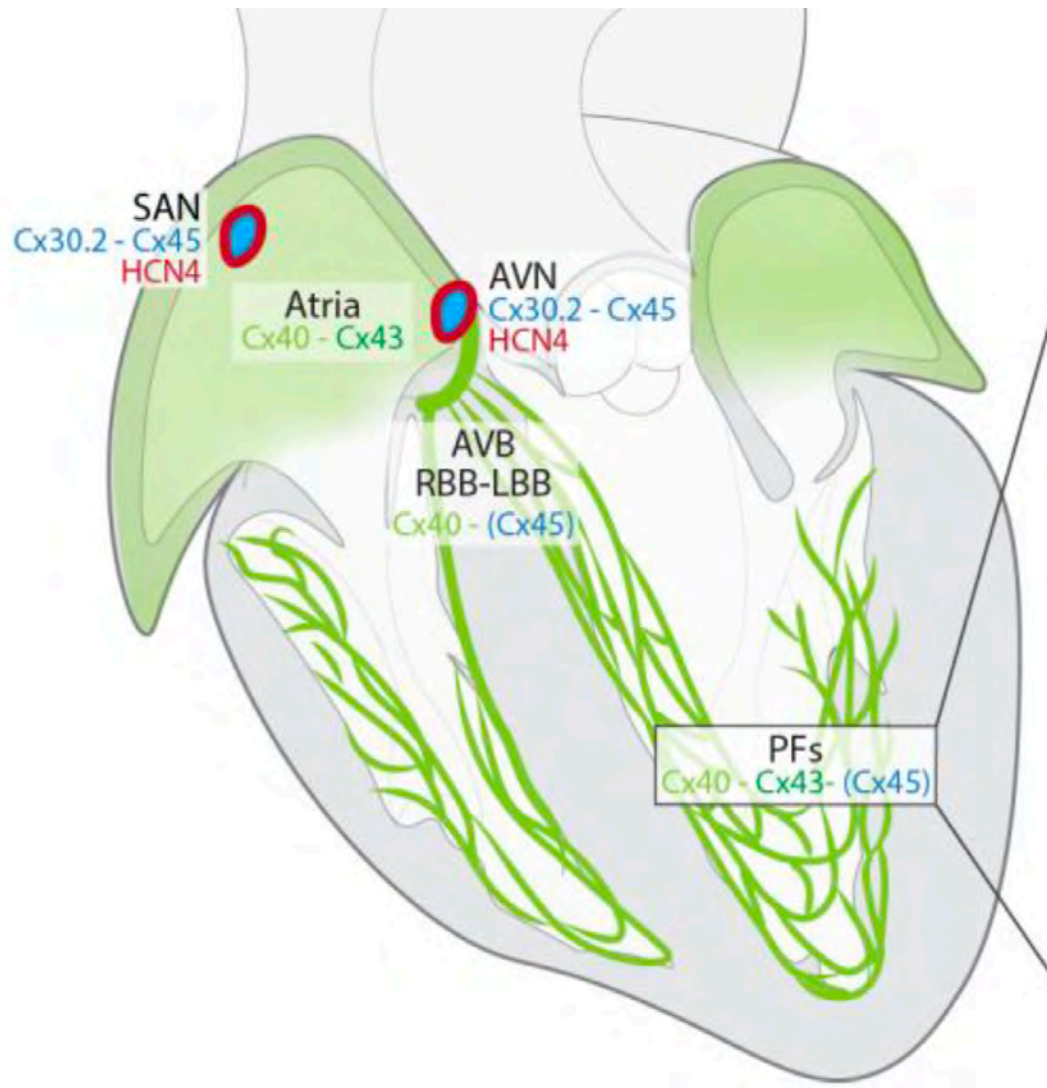
Robert C. Lyon^{1,†}, Valeria Mezzano^{1,†}, Adam T. Wright², Emily Pfeiffer², Joyce Chuang², Katherine Banares¹, Allan Castaneda¹, Kunfu Ouyang¹, Li Cui³, Riccardo Contu¹, Yusu Gu¹, Sylvia M. Evans³, Jeffrey H. Omens^{1,2}, Kirk L. Peterson¹, Andrew D. McCulloch² and Farah Sheikh^{1,*}

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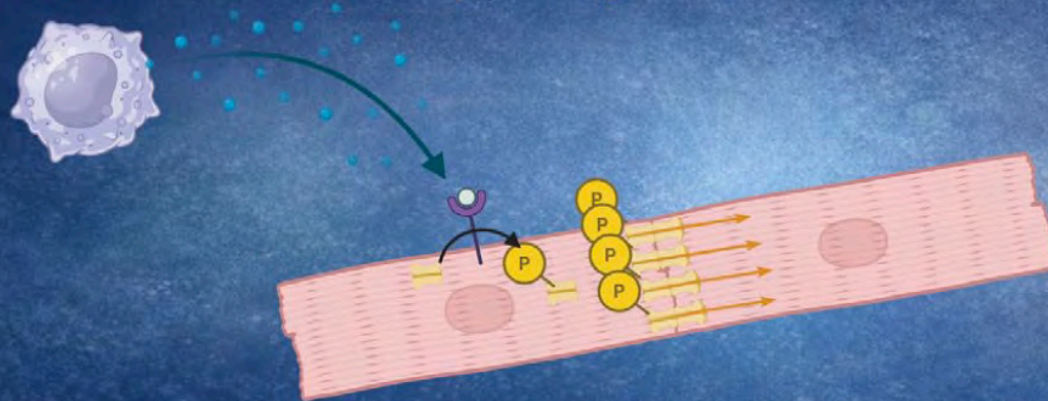
Received July 30, 2013; Revised September 6, 2013; Accepted October 4, 2013

Arrhythmogenic right ventricular cardiomyopathy (ARVC) termed a ‘disease of the desmosome’ is an inherited cardiomyopathy that recently underwent reclassification owing to the identification of left-dominant and biventricular disease forms. Homozygous loss-of-function mutations in the desmosomal component, desmoplakin, are found in patients exhibiting a biventricular form of ARVC; however, no models recapitulate the postnatal hallmarks of the disease as seen in these patients. To gain insights into the homozygous loss-of-function effects of desmoplakin in the heart, we generated cardiomyocyte-specific desmoplakin-deficient mice (DSP-cKO) using ventricular myosin light chain-2-Cre mice. Homozygous DSP-cKO mice are viable but display early ultrastructural defects in desmosomal integrity leading to a cardiomyopathy reminiscent of a biventricular form of ARVC, which includes cell death and fibro-fatty replacement within the ventricle leading to biventricular dysfunction, failure and premature death. DSP-cKO mice also exhibited ventricular arrhythmias that are exacerbated with exercise and catecholamine stimulation. Furthermore, DSP-cKO hearts exhibited right ventricular conduction defects associated with loss of connexin 40 expression and electrical wavefront propagation defects associated with loss of connexin 43 expression. Dose-dependent assessment of the effects of loss of desmoplakin in neonatal ventricular cardiomyocytes revealed primary loss of connexin 43 levels, phosphorylation and function independent of the molecular dissociation of the mechanical junction complex and fibro-fatty manifestation associated with ARVC, suggesting a role for desmoplakin as a primary stabilizer of connexin integrity. In summary, we provide evidence for a novel mouse model, which is reminiscent of the postnatal onset of ARVC while highlighting mechanisms underlying a biventricular form of human ARVC.

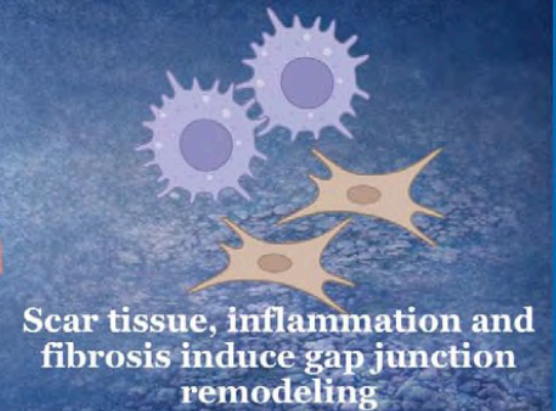




**Macrophage-derived Amphiregulin
Promotes Cx43 phosphorylation
via EGFR pathway**



Conduction Disturbances



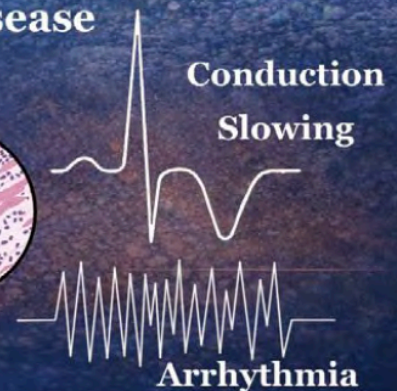
Healthy Heart



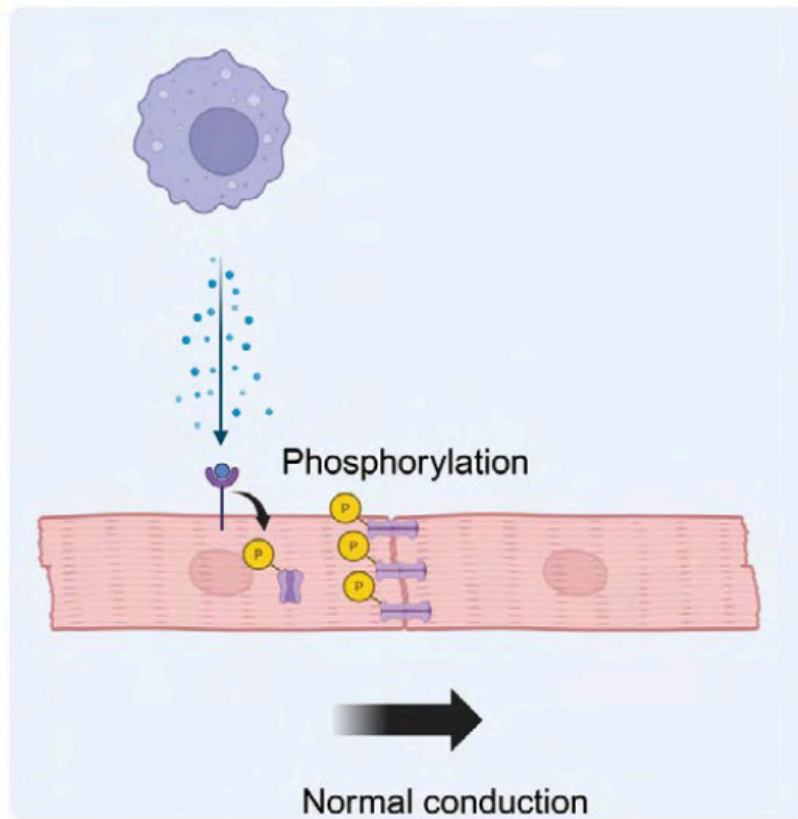
Normal Conduction



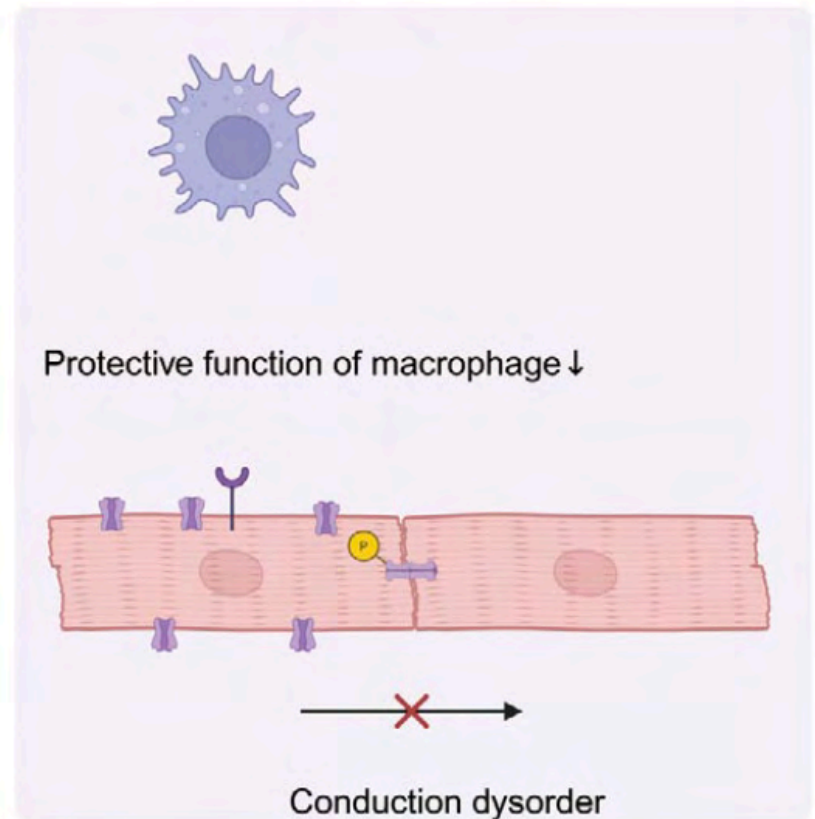
Cardiac Disease



Healthy condition



Under cardiac stress



Conduction disturbances

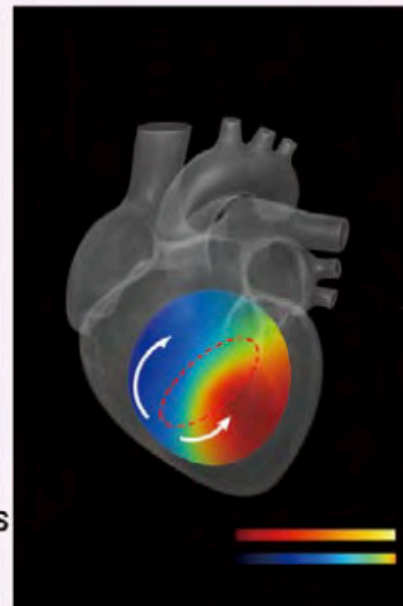
- gap junction dysfunction
- Purkinje network delay
- ventricular arrhythmias



3D Optical mapping
Tissue clearing



AI-based analysis
Mathematical model
Mathematical data analysis



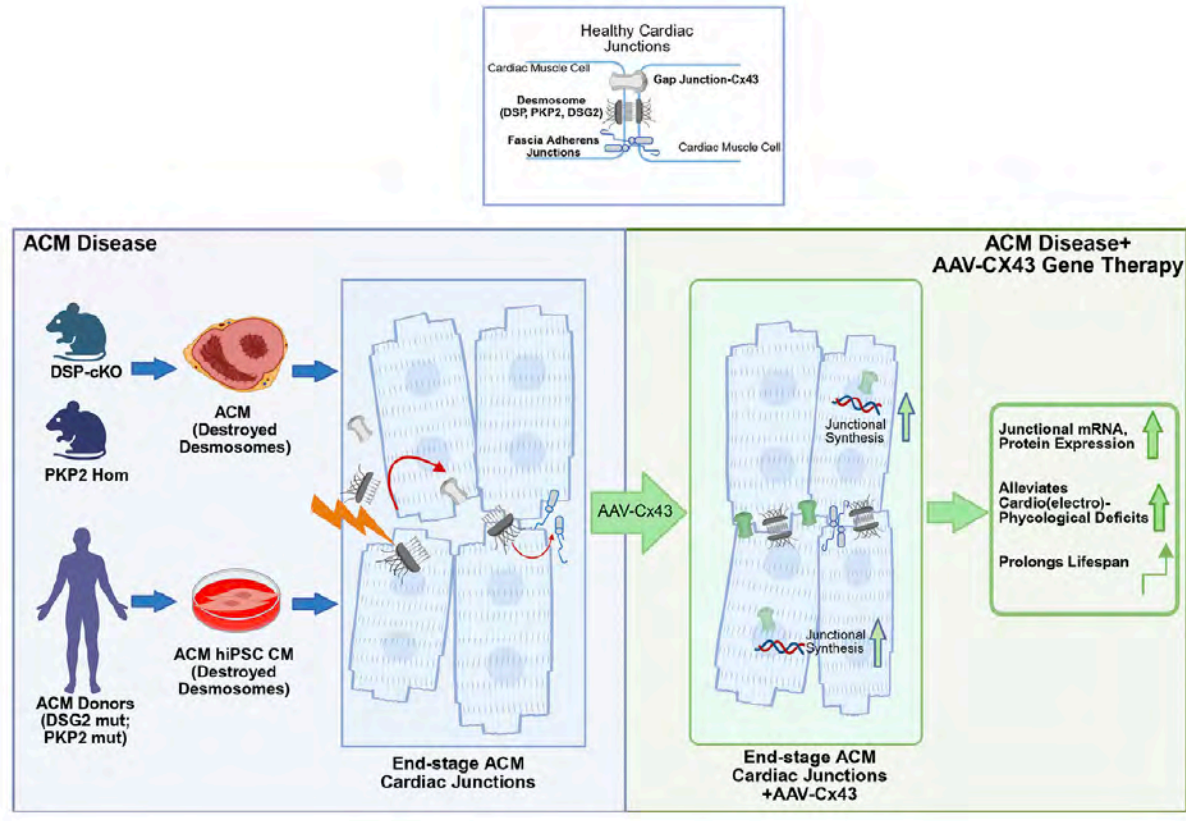
Uncovering
previously
hidden
**dynamic
arrhythmogenic
substrates**

Published in final edited form as:

Circ Heart Fail. 2026 April ; 19(4): e013801. doi:10.1161/CIRCHEARTFAILURE.125.013801.

Connexin-43 Restoration Alleviates Desmosomal Arrhythmogenic Cardiomyopathy

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In advanced stages of ACM disease, these models display severe desmosomal structural deficits that lead to severe loss of Cx43 (thicker red arrow). This Cx43 loss was associated with downregulation of cell-cell junction gene and protein expression leading to structural cardiomyopathy, encompassing ventricular arrhythmias, dilatation and dysfunction.

Adeno-associated viral (AAV)-Cx43 gene therapy targeted to restore Cx43 in cardiomyocytes in early and late ACM disease is sufficient to restore mechanical junctional protein synthesis, potentially through nuclear-mediated mechanisms, and re-localization, reinforcing desmosomal and overall cell-cell junction integrity. This restoration alleviates electrical and physiological deficits to prolong lifespan in desmosomal ACM mouse models while alleviating physiological deficits in desmosomal ACM human models

From Argentina to Denmark—The wine is still good

Reginald T. Ho, MD, FHRS

From the Division of Cardiology, Department of Medicine, Thomas Jefferson University Hospital, Philadelphia, Pennsylvania.

In 1968, Dr Mauricio Rosenbaum published a book dedicated entirely to the intraventricular conduction system.¹ In this classic monograph and its subsequent English version, he coined the term “hemiblock” and introduced the concept of a trifascicular conduction system after analyzing electrocardiograms from a 58-year-old man who had suffered an anterior myocardial infarction and demonstrated right bundle branch block (RBBB) with alternating left anterior fascicular block (LAFB) and left posterior fascicular block (LPFB) (now called Rosenbaum’s syndrome).² He referred to the conduction system as a “detector” of the heart, showing the association between various conduction blocks and heart disease (commonly coronary artery disease and Chagas cardiomyopathy in his home country of Argentina). He described the unequal “anatomic vulnerability” of the bundle branches (right more than left; left anterior more than posterior) and the relative “immunity” of the left posterior fascicle because of its thick structure and dual blood supply (indicating that the presence of LPFB generally signified more severe heart disease). His book was followed by a flurry of studies in the mid-1970s and early 1980s evaluating the value of His-ventricular intervals in predicting impending atrioventricular block (AVB) in patients with bifascicular block—research that today remain the foundation for our current pacemaker guidelines.³ Since then, however, research on the natural history of fascicular block and its progression to AVB has been relatively quiet.

In this issue of *Heart Rhythm Journal*, Nyholm et al⁴ breathe new life into the study of fascicular blocks by providing the largest population-based study on its natural progression to AVB. Among 358,958 primary care patients in a large Danish registry (Copenhagen ECG Study), the authors studied 13,636 patients with fascicular block (3.8%) and compared them with a reference group of patients without block. Not surprisingly, RBBB and isolated LAFB were most common. With the longest follow-up approaching 16 years, they found that syncope, pacemaker implantation, and third AVB increased with increasing complexity of

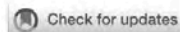
fascicular block. Depending on gender and age, the 10-year absolute risk of developing third-degree AVB increased from 0% to 2% (hazard ratio [HR] 1.60) for isolated LAFB to 23% (HR 10.98) for multicomplex block (first-degree AVB + RBBB + LAFB). While this dose-response relationship between worsening fascicular block and AVB is not unexpected, their data provide clearer granularity about the long-term risk of developing AVB among 10 different block subtypes. True bilateral BBB (eg, alternating BBB and Rosenbaum’s syndrome) was not represented. However, this subtype is already an established high-risk group carrying a class I indication for pacemaker implantation. While a higher burden of comorbidity occurred with increasing block complexity, LAFB was not associated with worse mortality. This has been observed in another study from the same group but not by others.^{5–7} Curiously, isolated LPFB was associated with the youngest age group (median age 35 years) and the highest risk of death (HR 2.09). A recent case-control study of 10 young patients (median age 27.5 years) with LPFB and aborted cardiac arrest/sudden cardiac death found left ventricular fibrosis (particularly along the inferolateral wall) in all patients undergoing cardiac magnetic resonance imaging (n = 6) or histopathological analysis/autopsy (n = 4).⁸ Further investigation is required into this small but worrisome group of young patients.

In the preface to his book, Dr Rosenbaum wrote “Like good wines, some research improves after resting for a while.” By allowing many years for their registry to mature, Nyholm et al have produced an excellent bottle of wine for a slowly aging cellar.

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but not by others.^{2–7} Curiously, isolated LPFB was associated with the youngest age group (median age 35 years) and the highest risk of death (HR 2.09). A recent case-control study of 10 young patients (median age 27.5 years) with LPFB and aborted cardiac arrest/sudden cardiac death found left ventricular fibrosis (particularly along the inferolateral wall) in all patients undergoing cardiac magnetic resonance imaging (n = 6) or histopathological analysis/autopsy (n = 4).⁸ Further investigation is required into this small but worrisome group of young patients.



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The pivotal role of ECG in cardiomyopathies

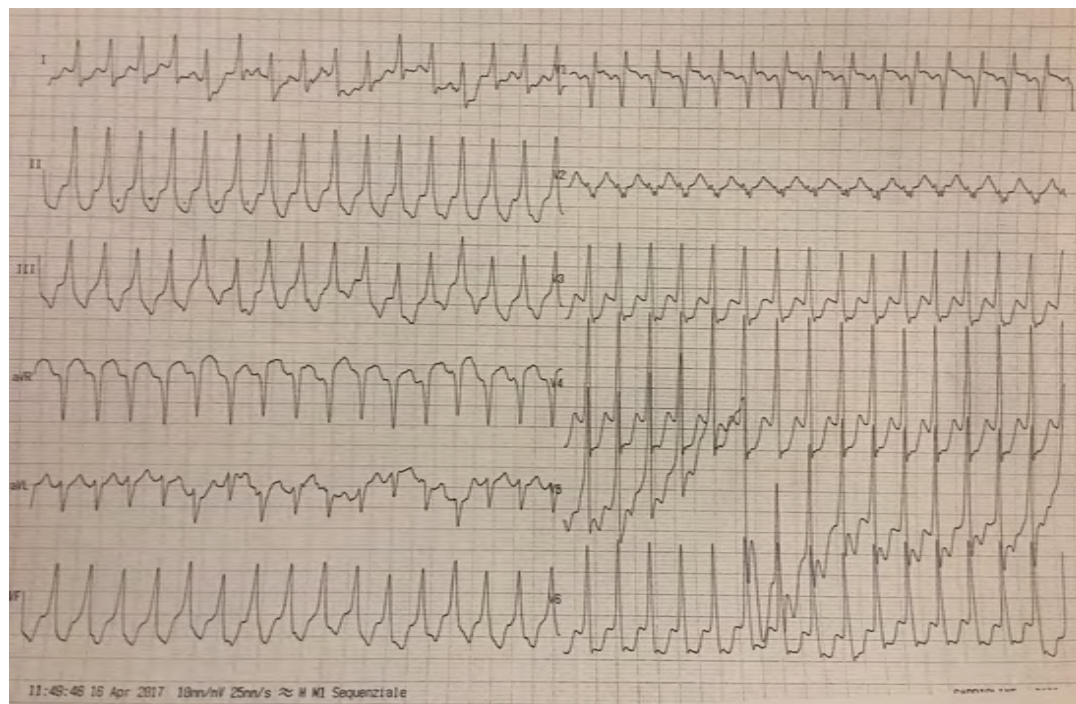
Elisa Silvetti, Oreste Lanza, Fabiana Romeo, Annamaria Martino,
Elisa Fedele, Chiara Lanzillo, Cinzia Crescenzi, Francesca Fanisio
and Leonardo Calò*

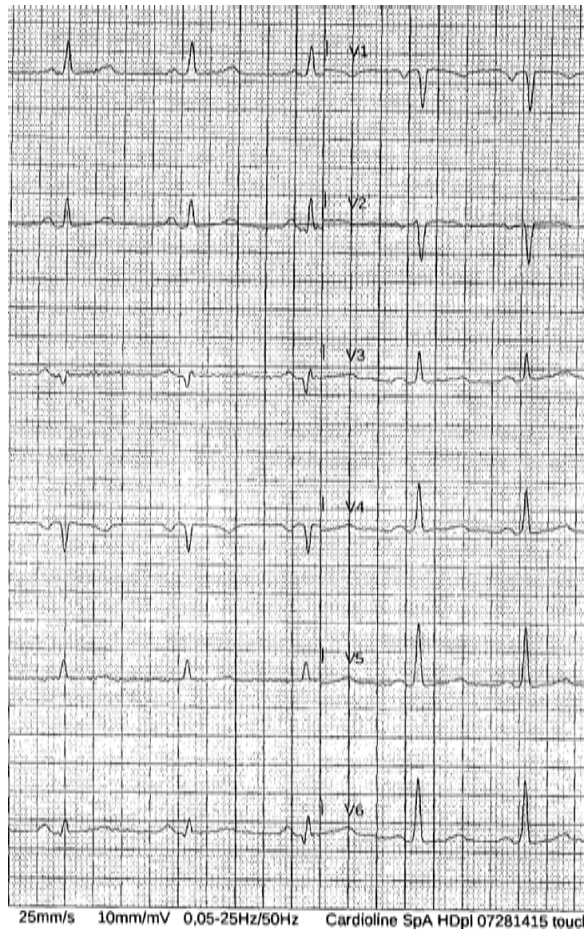
Division of Cardiology, Policlinico Casilino, Rome, Italy

TABLE 1 Main ECG findings in cardiomyopathies.

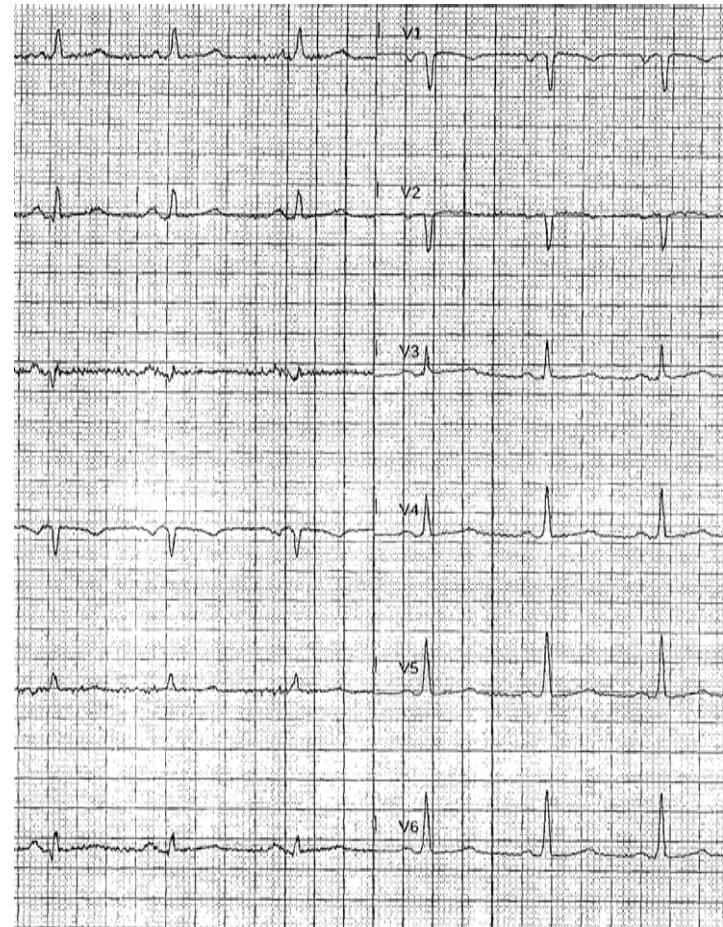
	DCM/HDCM	NMD	ACM	HCM	AFD and storage disease	CA	Sarcoidosis
Depolarization abnormalities	fQRS; pathological Q-wave; occasionally LQRSV (e.g., in FLNC mutation)	LQRSV in the presence of scar	fQRS; epsilon wave; SAECEG; TAD > 55 ms in V1; LQRSV in limb leads	Sokolow-Lyon (V1 S onda + V5 o V6 R dell'onda $\geq$ 35 mm); Cornell signs (SV3 + RaVL with a cutoff for LHV > 2.0 mV in women and > 2.8 mV in men); LQRSV (if extensive LGE); Q waves; fQRS	Q waves; fQRS; extremely high voltages (Danon disease)	LQRSV; QS pattern in precordial leads	fQRS; epsilon wave
AV conduction disorders	First-degree and advanced AVB especially in some defined genetic form (e.g. involving LMNA gene)	Advanced AVB	—	—	Short PR interval (AFD; Danon disease)	First-degree and advanced AVB	Advanced AVB
IV conduction disorders	RBBB (rare); LBBB	LBBB	RBBB-like pattern (ARVC); LPFB (ALVC)	—	—	IV conduction delay	RBBB
Repolarization abnormalities	TWI in inferior, antero-lateral, and infero-lateral leads	TWI in infero-lateral leads	TWI in right precordial (ARVC); TWI in lateral leads	ST strain ($\geq$ 1 mm concave downsloping ST-segment depression); asymmetrical TWI in lateral leads; "pseudo-STEMI" pattern (ST-segment elevations in antero-lateral leads); deep T-wave inversion ($\geq$ 2 mm) in infero-lateral leads; "giant T waves" (> 10 mm), in the apical form of HCM	ST-segment depression and TWI in infero-lateral leads	ST-segment depression and TWI in infero-lateral leads	TWA; higher T-wave amplitude in lead aVR; TWI, and a longer interval of T-peak to T-end; QT dispersion
P-wave morphology alterations	Increased P-Wave duration and dispersion	—	—	Bifid and large P-wave (total duration > 110 ms); P-wave amplitude > 2.5 mm in inferior leads and > 1.5 mm in V1–V2	—	—	—
Arrhythmias	AF; VT	AF; VT; BBR-VT	VT with LBBB pattern (ARVC); VT with RBBB pattern (ALVC)	AF; VT often with RBBB morphology	AF; VT	AF	VT

AF, atrial fibrillation; AFD, Anderson–Fabry disease; ACM, arrhythmogenic cardiomyopathy; ALVC, arrhythmogenic left cardiomyopathy; ARVC, arrhythmogenic right ventricle cardiomyopathy; AVB, atrioventricular block; BBR-VT, bundle branch reentry ventricular tachycardia; CA, cardiac amyloidosis; DCM/HDCM, dilated cardiomyopathy/hypokinetic dilated cardiomyopathy; FLNC, filamin C; fQRS, fragmented QRS; IV, intraventricular; LBBB, left bundle branch block; LMNA, lamin A; LPFB, left posterior fascicular block; LQRSV, low QRS voltage; NMD, neuromuscular dilated; RBBB, right bundle branch block; TAD, terminal activation delay; TWA, T-wave alternans; TWI, T-wave inversion; VT, ventricular tachycardia.

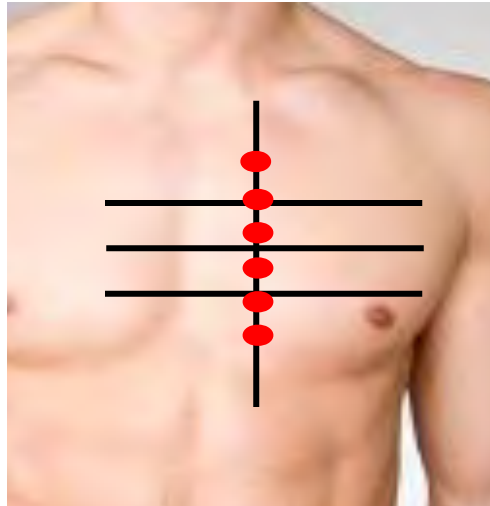


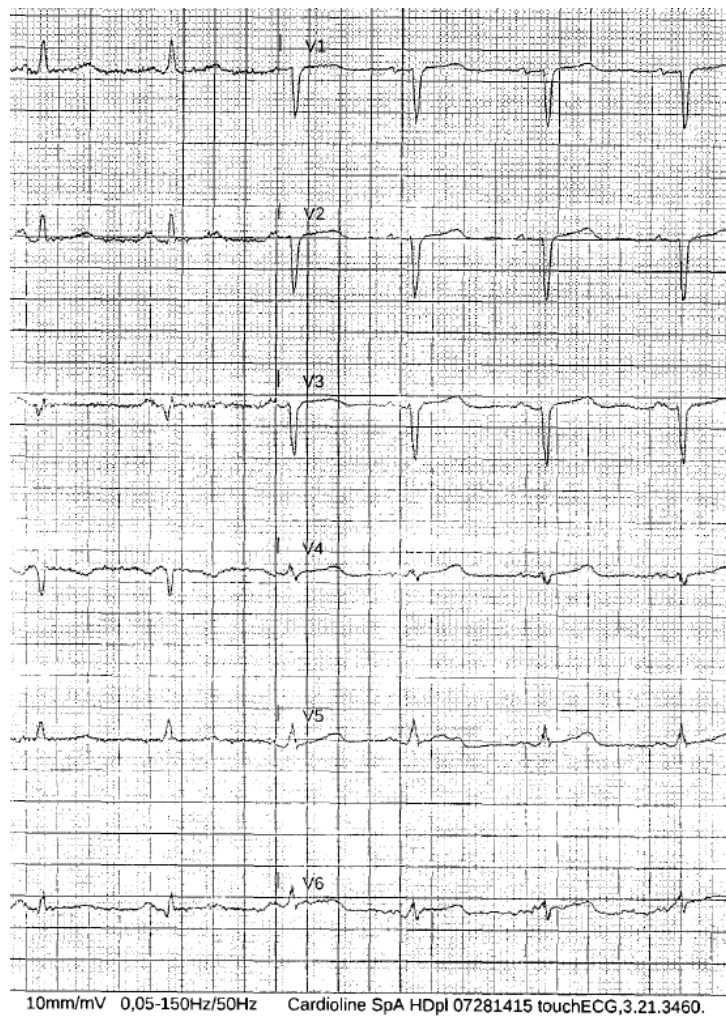


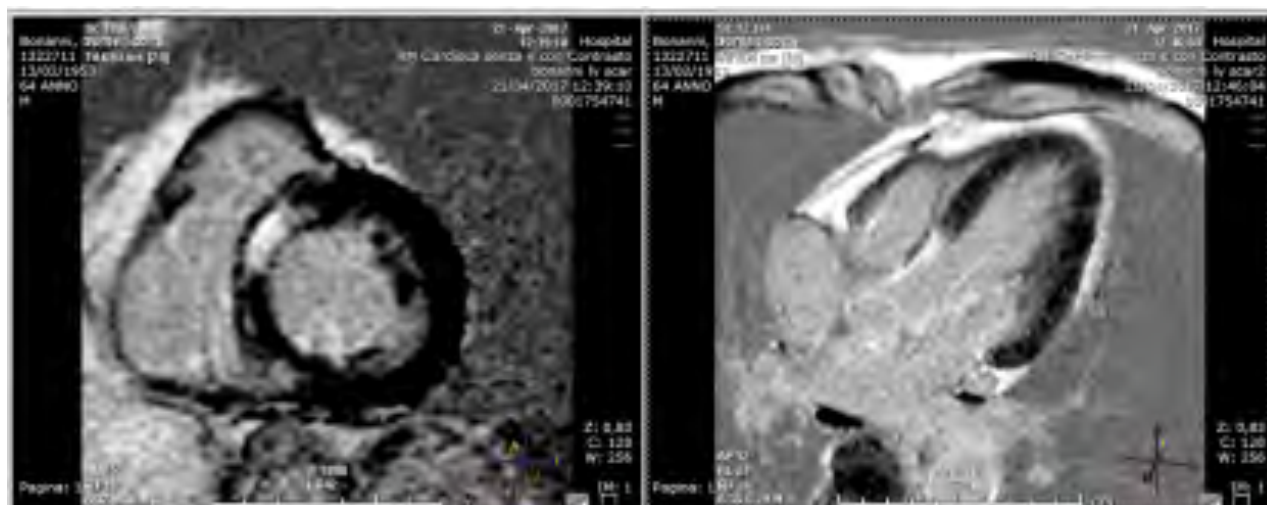
25mm/s 10mm/mV 0,05-25Hz/50Hz Cardioline SpA HDpl 07281415 touch



ino 25mm/s 10mm/mV 0,05-150Hz/50Hz Cardioline SpA HDpl 07281415 touchECG.3.2







Gene: LMNA (Encoding the protein: Prelamin-A/C)
NP_733821.1:p.Arg189Trp/NC_000001.10:g.156104245C>T

Heterozygous carrier: Mutation occurs in only one copy of the gene.

Next Generation Sequencing stats: Depth of coverage: 221. Quality of the variant (0-255): 255.

Mutation nomenclature: Nucleotide code: NM_170707.3:c.565C>T, NC_000001.10:g.156104245C>T. Amino acid code: NP_733821.1:p.Arg189Trp. dbSNP ID: rs267607626. Alternative names at the protein level: NP_733821.1:p.R189W. Located in: exon 3.

Pathogenicity: very likely to be pathogenic or disease-causing (++).

Population frequency: rare variant (found in <1% of controls).

Number of articles/communications that cite it: 10. Number of described families: 7. Number of families with additional

Major phenotypes	Carriers (families)	Non-carriers	Without genetic study	Total
No phenotypic study	5 (3)	0	4	9
Dilated cardiomyopathy	5 (5)	0	0	5
Not affected or healthy	3 (1)	2	4	9
Sudden death	2 (1)	0	1	3
Dilated cardiomyopathy + Left ventricular non-compaction/hypertrabeculation	1 (1)	0	0	1
Dilated cardiomyopathy + Muscular Dystrophy, Limb-Girdle + Peripheral Neuropathy	1 (1)	0	0	1
Left ventricular non-compaction/hypertrabeculation	0 (0)	0	1	1

ARVDC/D

Alterazioni della conduzione intraventricolare

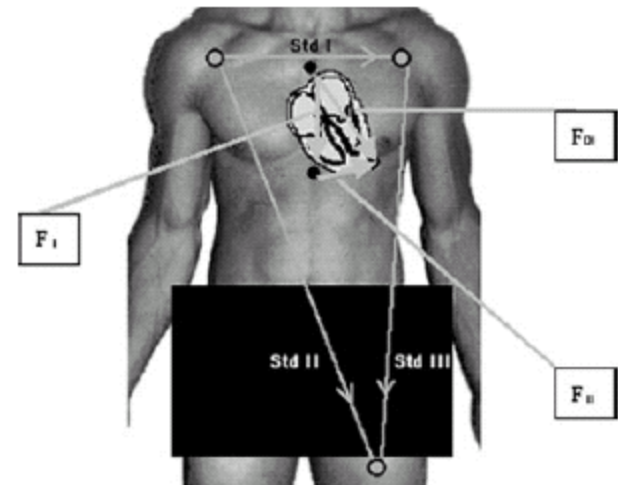
Epsilon Wave in Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy

SHULIN WU, M.D., PEINING WANG, Ph.D., YUESHUANG HOU, M.M., PINGZHEN YANG, M.M., YANPING XIAO, M.S., and XIANZHANG ZHAN, M.M.

From the Department of Cardiology, Guangdong General Hospital, Guangdong Cardiovascular Institute, Guangdong, China

Il primo a registrare l'onda epsilon fu Gui Fontaine nel 1977

Fontaine leads were recorded in order to better recognize the presence of the Epsilon wave. Rhythm strips were obtained at double speed (50 mm/s) and double amplitude (20 mm/mV): the left arm lead was placed over the xyphoid process, the right arm lead on the manubrium sterni, and the left leg lead over a rib at the V4 position



(PACE 2009; 32:59-63)

ARVDC/D

Alterazioni della conduzione intraventricolare

Epsilon Wave in Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy

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