

10° CONGRESSO NAZIONALE



*Quello che le Linee
Guida Non Dicono*

Napoli
Hotel Excelsior
14-15 aprile 2023

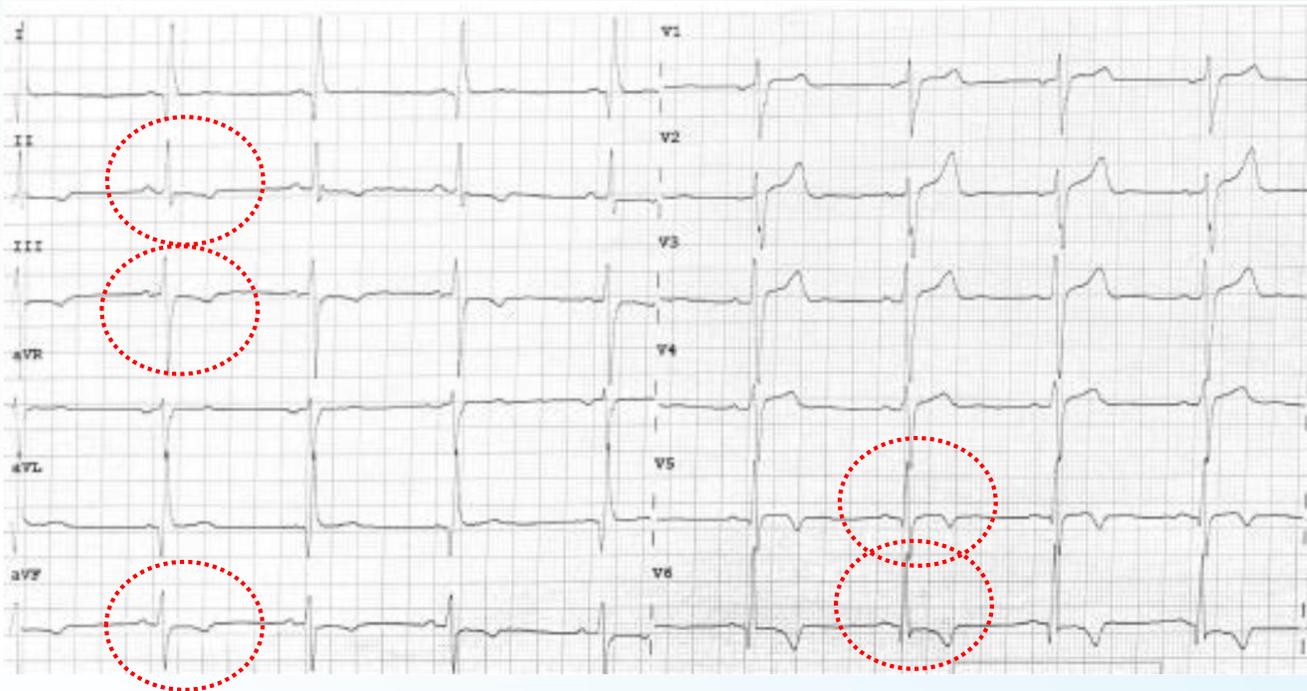
Miocarditi
Silvana Cicala

C.S., maschio 23 anni

Fumatore

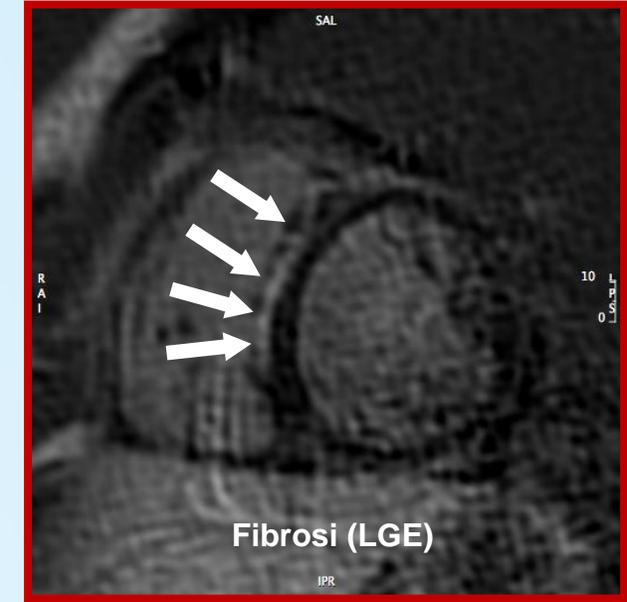
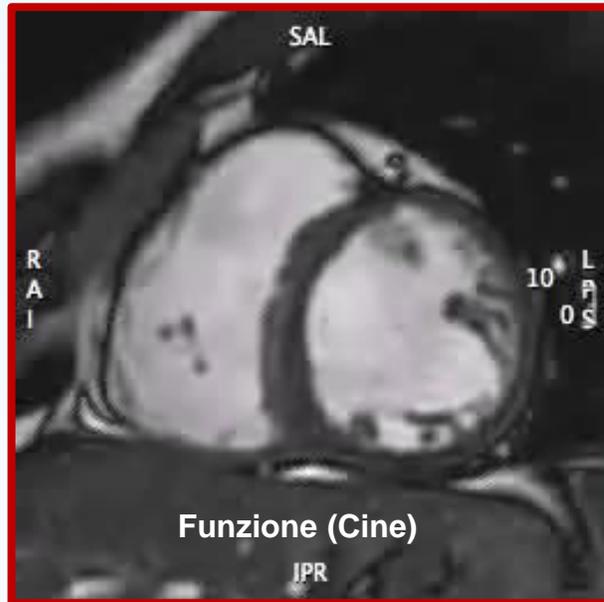
Anamnesi familiare negativa per patologie cardiache e/o morte improvvisa

Anamnesi personale positiva per 2 episodi di dolore toracico acuto con ammissione in PS
(5 anni prima con diagnosi di miocardite; 4 mesi prima con diagnosi di MINOCA)



- **Dolore toracico acuto**
(non accompagnato a sintomi di tipo infettivo)
- **Segni di danno miocardico acuto**
(picco hs-TnI 327 ng/L)
- **Nulla di rilevante alla valutazione ecocardio**
- **Non sottoposto a rivalutazione coro**

RM Cardiaca n. 1



CONCLUSIONI

- Ventricolo non dilatato, con funzione sistolica globale ai limiti inferiori della norma
- Immagini di caratterizzazione tissutale in sede ventricolare sinistra compatibili con processo miocarditico in fase attiva ad interessamento settale

Biopsia endomiocardica rifiutata dal paziente
Dimesso con diagnosi di recidiva di miocardite acuta
e raccomandazione a stretto follow-up clinico

Le recidive di miocardite acuta sono possibili, ma non frequenti

Circulation

ORIGINAL RESEARCH ARTICLE



Clinical Presentation and Outcome in a Contemporary Cohort of Patients With Acute Myocarditis

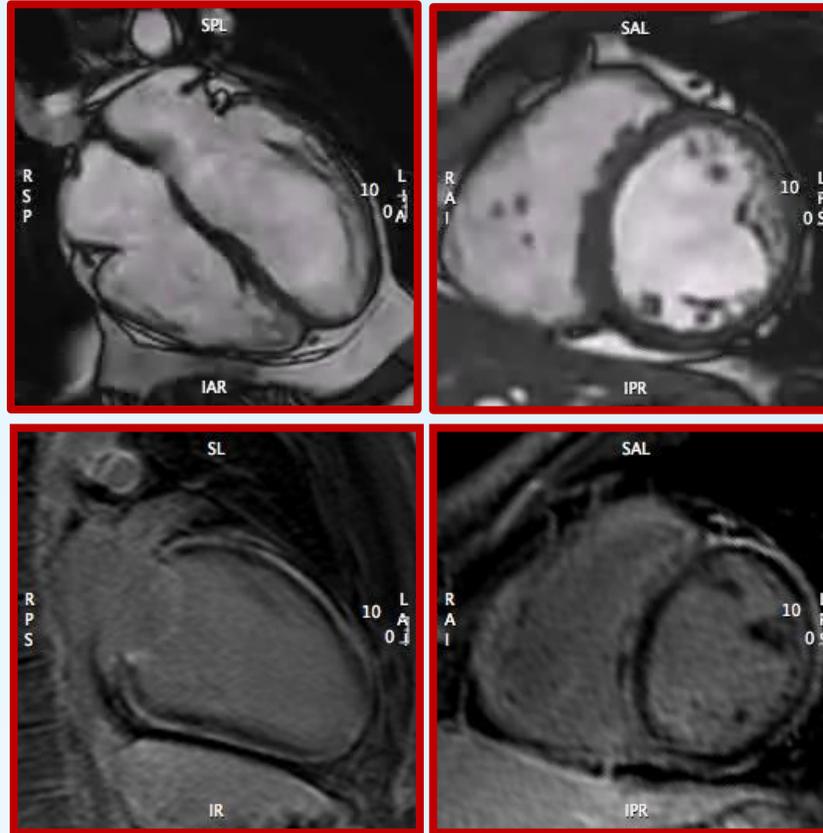
Multicenter Lombardy Registry *Circulation*. 2018;138:1088–1099

Tot = 443 pz. con miocardite acuta → 11 con recidiva (**2,6%**; FU mediana = 35 mesi)

C.S.

Follow-up a 1 anno

Holter ECG: 1198 BEVs/24h con 2 coppie



RM Cardiaca n. 2



CONCLUSIONI

- Ventricolo sinistro non dilatato, con lieve disfunzione contrattile in sede laterale e con iniziale riduzione della funzione sistolica globale (FE = 52%).
- Ventricolo destro non dilatato e con normale funzione sistolica regionale e globale.
- Immagini di caratterizzazione tissutale compatibili con la presenza di estesa fibrosi miocardica, coinvolgente in maniera diffusa i segmenti basali e medi del ventricolo sinistro con pattern di distribuzione di tipo non ischemico («ring-like pattern»).



CONTEMPORARY REVIEW

Evolving Diagnostic Criteria for Arrhythmogenic Cardiomyopathy

Domenico Corrado ¹, MD, PhD; Alessandro Zorzi ², MD, PhD; Alberto Cipriani ³, MD; Barbara Baucé ⁴, MD, PhD; Riccardo Bariani ⁵, MD; Giorgia Beffagna, BS, PhD; Manuel De Lazzari, MD, PhD; Federico Migliore ⁶, MD, PhD; Kalliopi Pillichou ⁷, BS, PhD; Alessandra Rampazzo ⁸, BS, PhD; Ilaria Rigato, MD, PhD; Stefania Rizzo ⁹, MD, PhD; Gaetano Thiene ¹⁰, MD; Martina Perazzolo Marra, MD, PhD; Cristina Basso ¹¹, MD, PhD

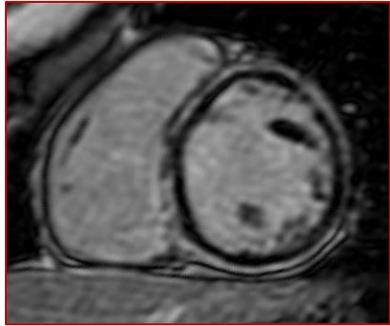


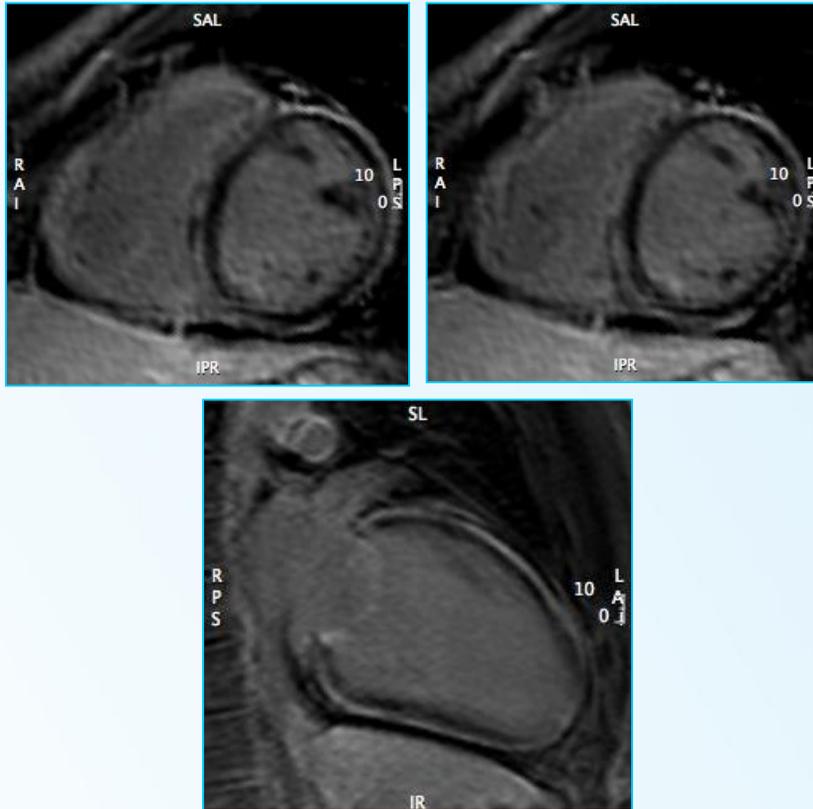
Table 1. Comparison of 2010 TF Criteria and 2020 International Criteria for Diagnosis of ARVC

Category	2010 TF criteria	2020 International criteria
I. Global or regional dysfunction and structural alteration	<p><i>Major</i></p> <p>By 2D echocardiogram:</p> <ul style="list-style-type: none"> Regional RV akinesia, dyskinesia, or aneurysm and one of the following (end diastole) PLAX RVOT ≥ 32 mm (corrected for body size [PLAX/BSA] ≥ 19 mm/m²) 	<p><i>Major</i></p> <p>By 2D echocardiogram, CMR, or angiography:</p> <ul style="list-style-type: none"> Regional RV akinesia, dyskinesia, or bulging plus 1 of the following: Global RV dilatation (increase of RV EDV according to the imaging test specific nomograms for age, sex, and BSA)

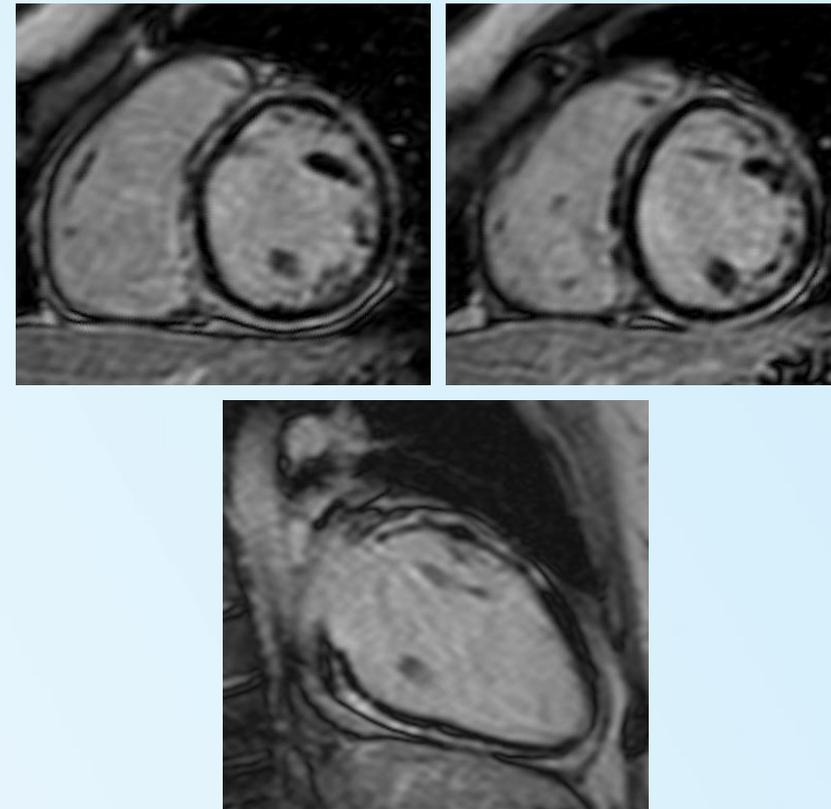
Table 4. The 2020 International Criteria for Diagnosis of ALVC

Category	Diagnostic criteria
I. Morpho-functional ventricular abnormalities	<p><i>Minor</i></p> <ul style="list-style-type: none"> Global LV systolic dysfunction* (depression of LV EF or reduction of echocardiographic global longitudinal strain), with or without LV dilatation (increase of LV EDV according to the imaging test specific nomograms for age, sex, and BSA) <p><i>Minor</i></p> <ul style="list-style-type: none"> Regional LV hypokinesia or akinesia of LV free wall, septum, or both
II. Structural myocardial abnormalities	<p><i>Major</i></p> <ul style="list-style-type: none"> LV LGE (stria pattern) of ≥ 1 Bull's Eye segment(s) (in 2 orthogonal views) of the free wall (subepicardial or midmyocardial), septum, or both (excluding septal junctional LGE)
III. Repolarization abnormalities	<p><i>Minor</i></p> <ul style="list-style-type: none"> Inverted T waves in left precordial leads (V₄-V₆) (in the absence of complete LBBB)
IV. Depolarization abnormalities	<p><i>Minor</i></p> <ul style="list-style-type: none"> Low QRS voltages (<0.5 mV peak to peak) in limb leads (in the absence of obesity, emphysema, or pericardial effusion)
V. Ventricular arrhythmias	<p><i>Minor</i></p> <ul style="list-style-type: none"> Frequent ventricular extrasystoles (>500 per 24 h), nonsustained or sustained ventricular tachycardia with a RBBB morphology (excluding the "fascicular pattern")
VI. Family history/genetics	<p><i>Major</i></p> <ul style="list-style-type: none"> ACM confirmed in a first-degree relative who meets diagnostic criteria ACM confirmed pathologically at autopsy or surgery in a first-degree relative Identification of a pathogenic or likely pathogenic ACM mutation in the patient under evaluation <p><i>Minor</i></p> <ul style="list-style-type: none"> History of ACM in a first-degree relative in whom it is not possible or practical to determine whether the family member meets diagnostic criteria Premature sudden death (<35 y of age) because of suspected ACM in a first-degree relative ACM confirmed pathologically or by diagnostic criteria in second-degree relative

C.S. ♂ 23 anni



C. A. ♀ 29 anni



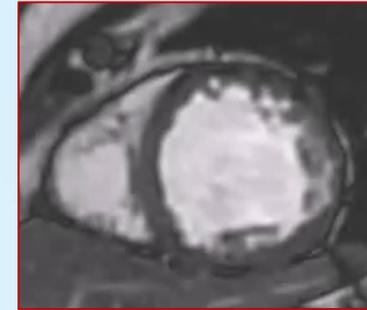
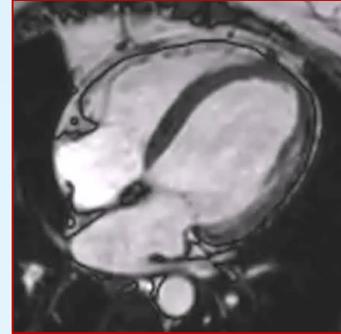
La sorella

P. P. Femmina 49 anni

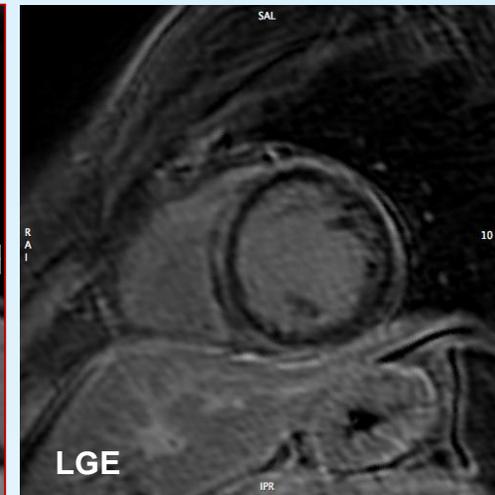
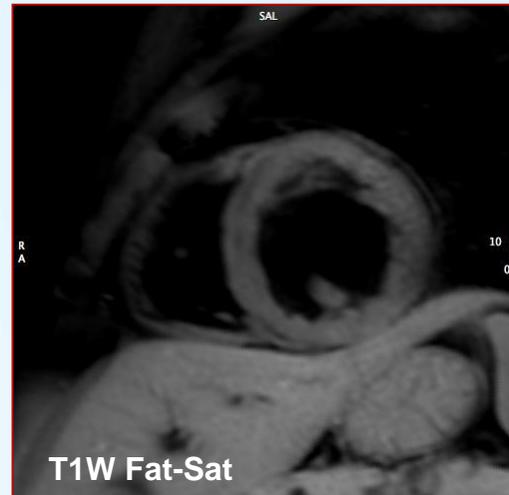
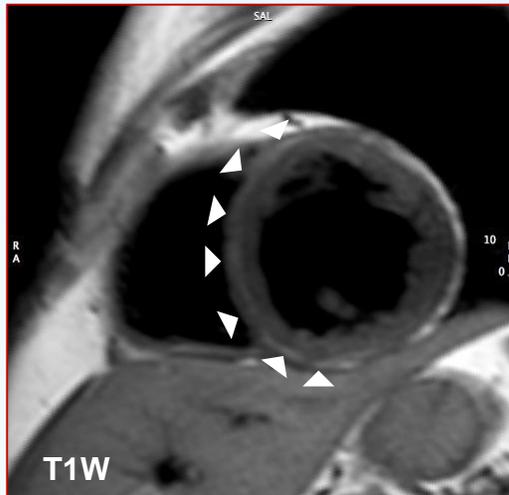
Episodio sincopale (2011) mai indagato

Holter ECG: 6730 BEVs/24h con 4 triplette

ECG: T neg DII, DIII, aVF, V4-V6



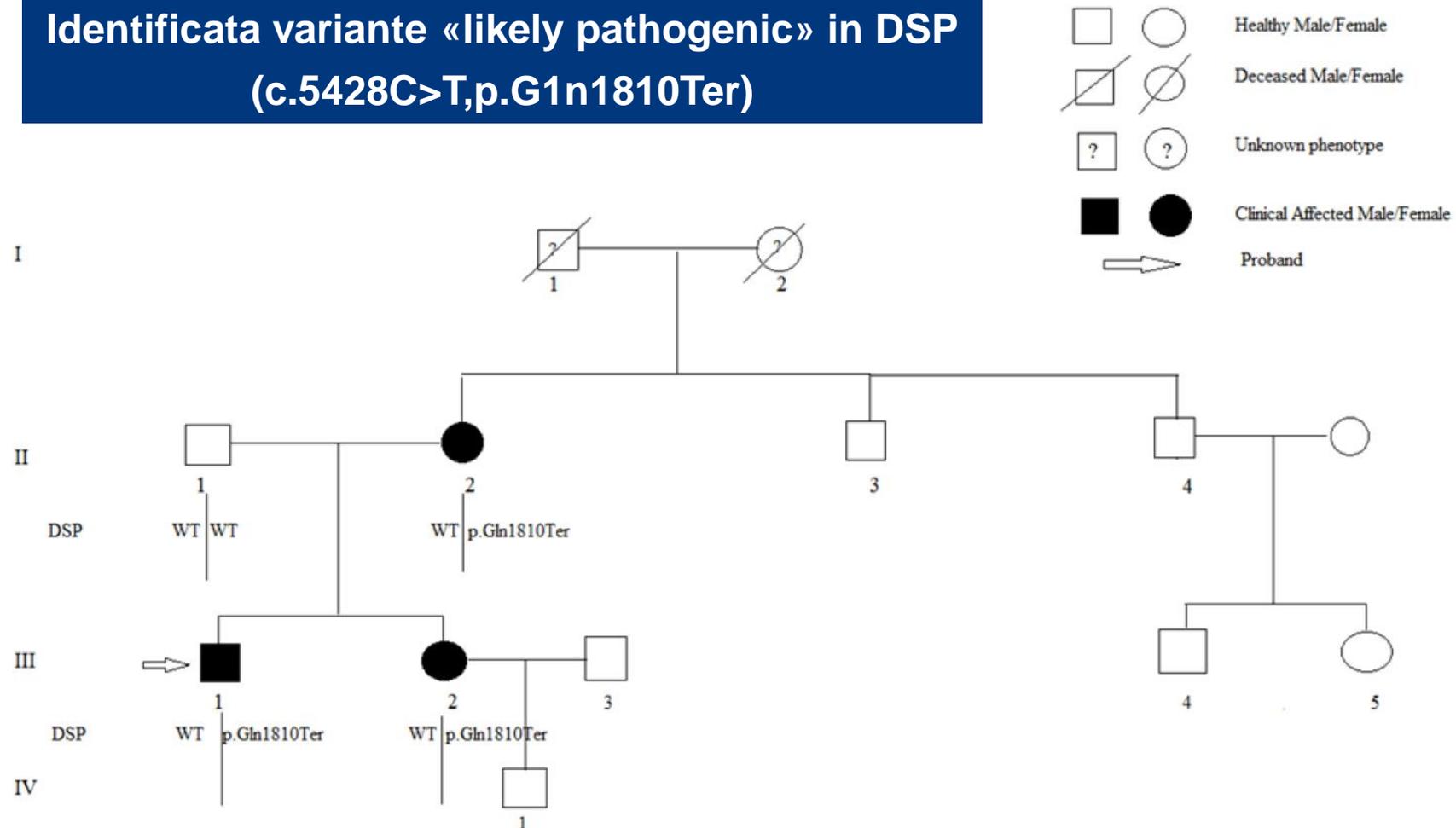
La madre



CONCLUSIONI

- Ventricolo sinistro d'aspetto globoso, lievemente dilatato, con funzione sistolica globale lievemente ridotta (FE = 46%).
- Ventricolo destro non dilatato e con normale funzione sistolica regionale e globale.
- Immagini di caratterizzazione tissutale compatibili con la presenza di estesa infiltrazione fibro-adiposa coinvolgente in maniera diffusa i segmenti basali e medi del ventricolo sinistro («ring-like pattern»).

**Identificata variante «likely pathogenic» in DSP
(c.5428C>T,p.G1n1810Ter)**



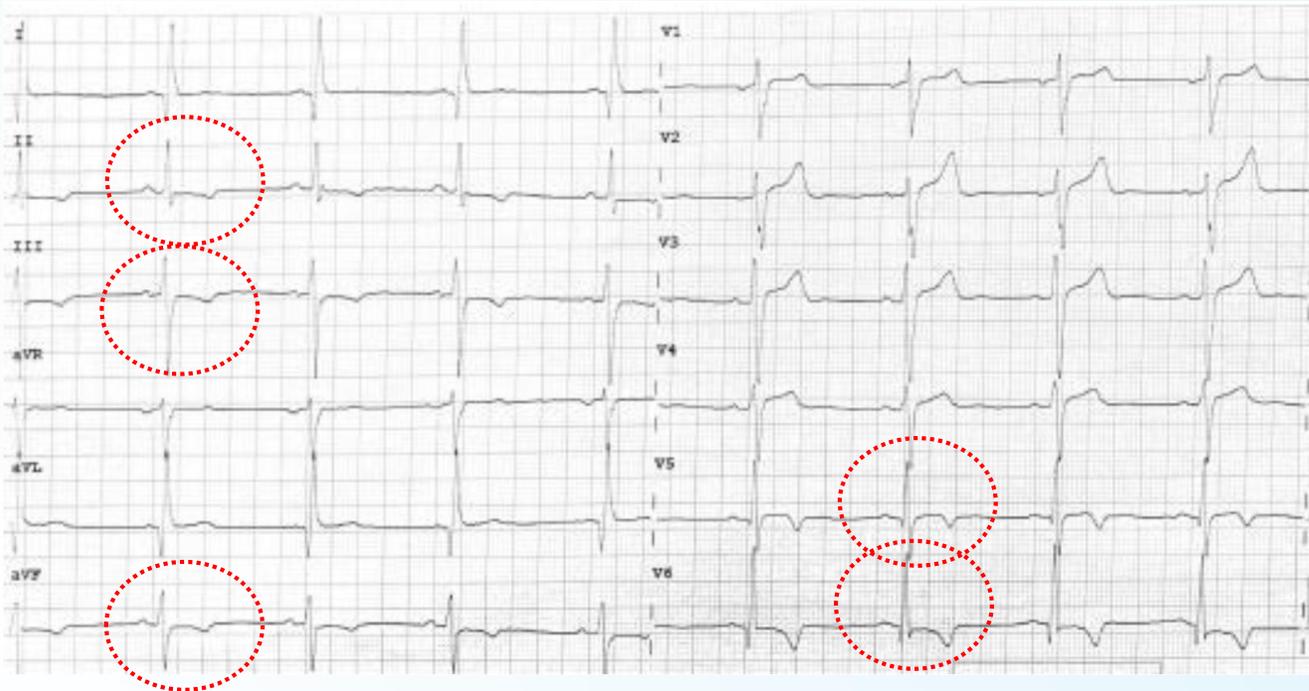
- **Terapia con β -bloccanti in tutti i portatori identificati**
- **Impianto s-ICD nel probando**

C.S., maschio 23 anni

Fumatore

Anamnesi familiare negativa per patologie cardiache e/o morte improvvisa

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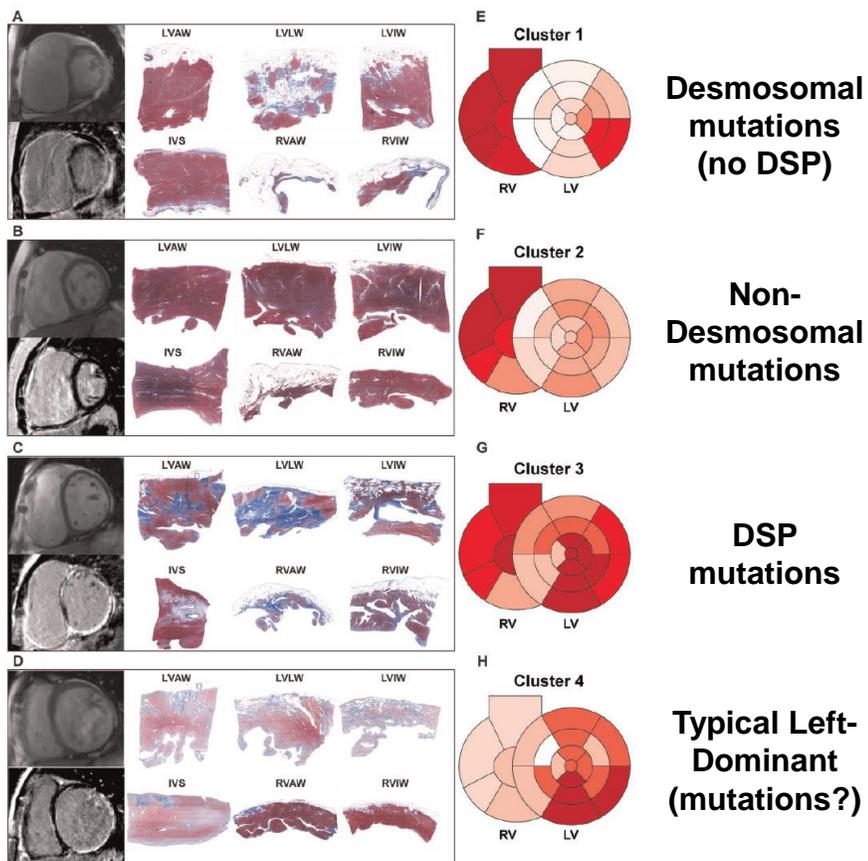


- **Dolore toracico acuto**
(non accompagnato a sintomi di tipo infettivo)
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Fase attiva di **cardiomiopatia desmosomiale**
con presentazione simil-miocarditica

A novel genotype-based clinicopathology classification of arrhythmogenic cardiomyopathy provides novel insights into disease progression

Liang Chen^{1†}, Jiangping Song^{1*†}, Xiao Chen^{1,2†}, Kai Chen¹, Jie Ren¹, Ningning Zhang¹, Man Rao¹, Zhenliang Hu¹, Yan Zhang³, Min Gu⁴, Hong Zhao², Hanwei Tang¹, Zhongfa Yang¹, and Shengshou Hu^{1*}

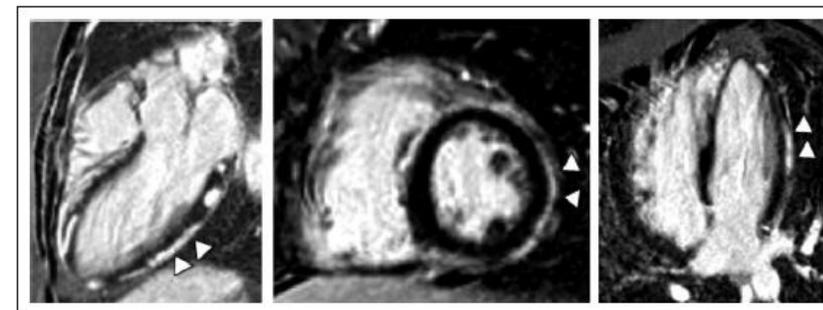


Circulation

ORIGINAL RESEARCH ARTICLE

Desmoplakin Cardiomyopathy, a Fibrotic and Inflammatory Form of Cardiomyopathy Distinct From Typical Dilated or Arrhythmogenic Right Ventricular Cardiomyopathy

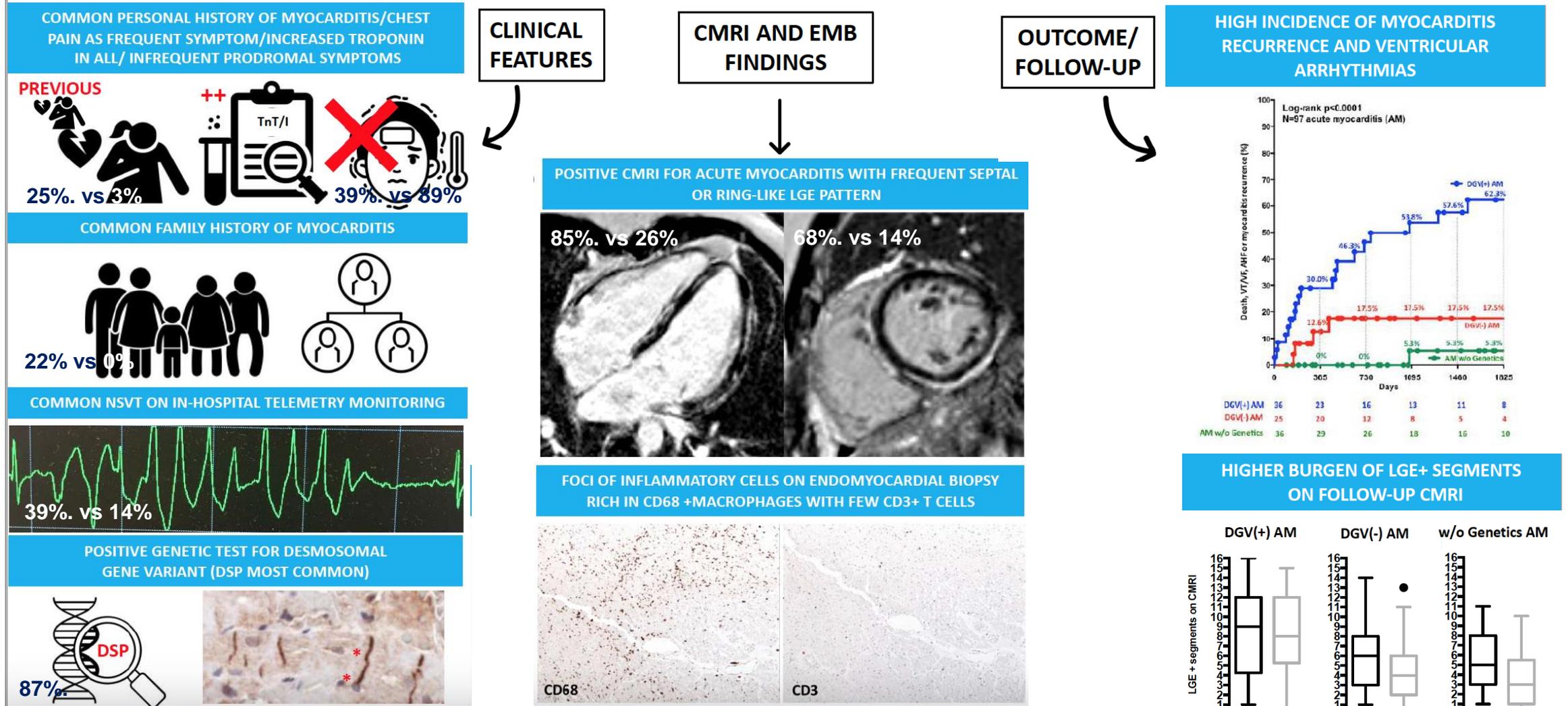
Eric D. Smith, MD
Neal K. Lakdawala, MD
Nikolaos Papoutsidakis, MD, PhD
Gregory Aubert, MD, PhD
Andrea Mazzanti, MD
Anthony C. McCanta, MD
Prachi P. Agarwal, MBBS
Patricia Arscott, MS
Lisa M. Dellefave-Castillo, MS
Esther E. Vorovich, MD
Kavitha Nutakki, MBBS
Lisa D. Wilsbacher, MD, PhD
Silvia G. Priori, MD, PhD
Daniel L. Jacoby, MD
Elizabeth M. McNally, MD, PhD
Adam S. Helms[Ⓢ], MD, MS



CONCLUSIONS: DSP cardiomyopathy is a distinct form of arrhythmogenic cardiomyopathy characterized by episodic myocardial injury, left ventricular fibrosis that precedes systolic dysfunction, and a high incidence of ventricular arrhythmias. A genotype-specific approach for diagnosis and risk stratification should be used.

Acute Myocarditis Associated with Desmosomal Gene Variants

Multicenter retrospective study (97 pts from 23 hospitals); 36 DGV (+) AM – 25 DGV (-) AM – 36 w/o genetics AM



Acute clinical presentation of nonischemic cardiomyopathies: early detection by cardiovascular magnetic resonance

Giancarlo Todiere^{a,*}, Andrea Barison^{a,*}, Anna Baritussio^b, Alberto Cipriani^b, Andrea Igoren Guaricci^c, Silvia Pica^{d,†}, Ciro Indolfi^e, Gianluca Pontone^f and Santo Dellegrottaglie^g, on behalf of the Working Group on Cardiac Magnetic Resonance of the Italian Society of Cardiology

J Cardiovasc Med. 2022, 23:1–11

