

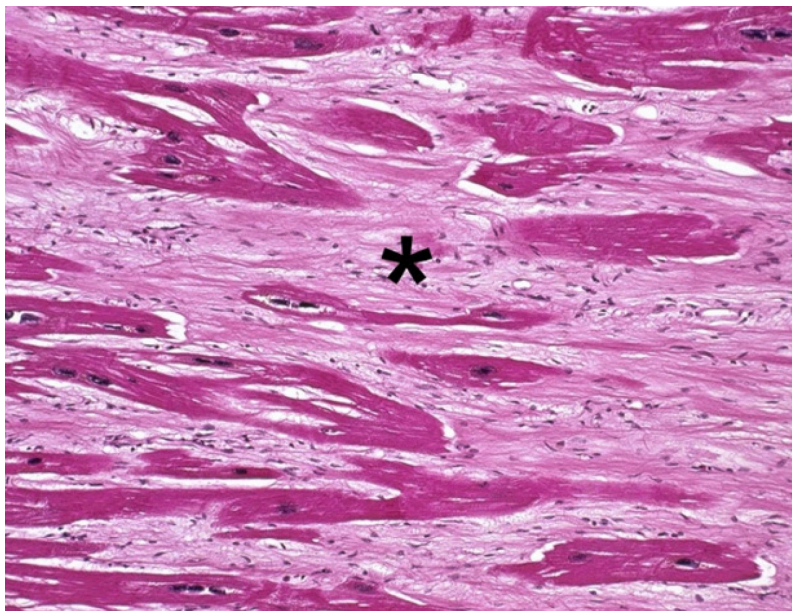
Fenotipo ipertrofico: Update 2026

Fibrosi miocardica e aritmogenesi nella cardiomiopatia ipertrofica

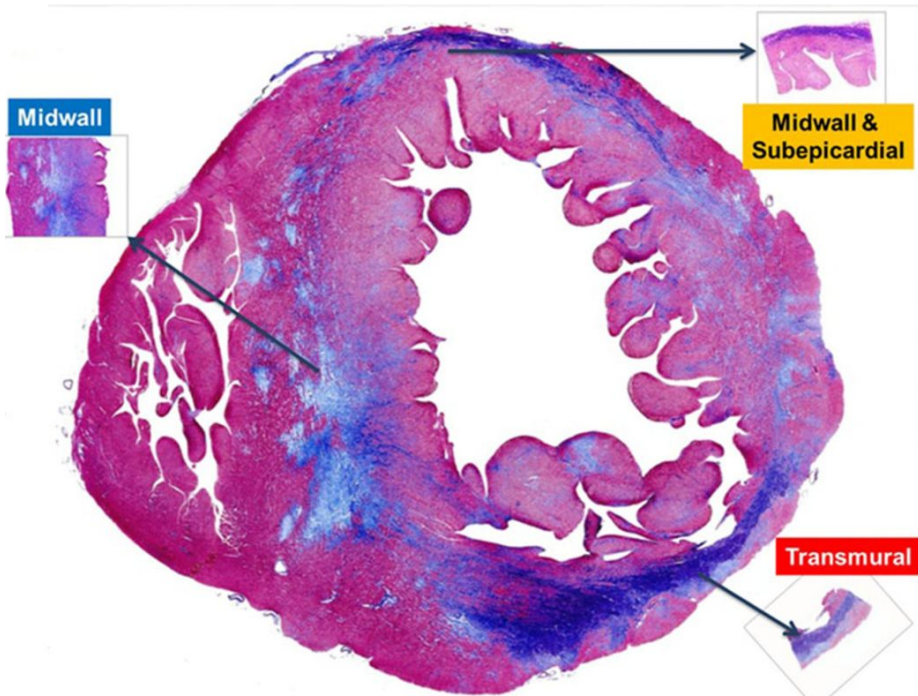
Edoardo Bertero
IRCCS Policlinico San Martino, AOM Genova



Fibrosi interstiziale



Fibrosi sostitutiva



ORIGINAL ARTICLE

Myocardial Fibrosis as an Early Manifestation of Hypertrophic Cardiomyopathy

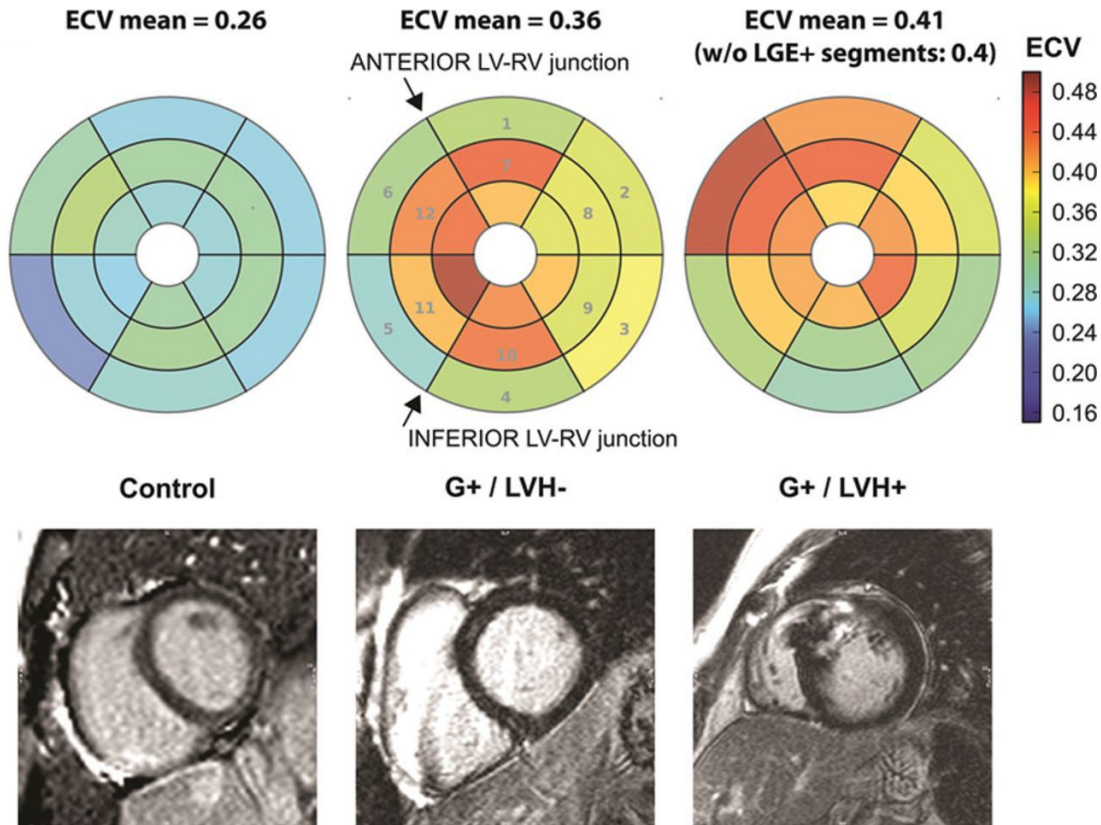
Table 2. Serum Biomarkers of Collagen Metabolism, Hemodynamic Stress, and Myocardial Injury.*

Variable	Control (N=30)	Mutation-Positive, LVH-Negative (N=39) [†]	P Value, Control vs. Mutation-Positive, LVH-Negative
PICP ($\mu\text{g/liter}$)	82.16 $\pm$ 3.03	107.73 $\pm$ 4.65	<0.001

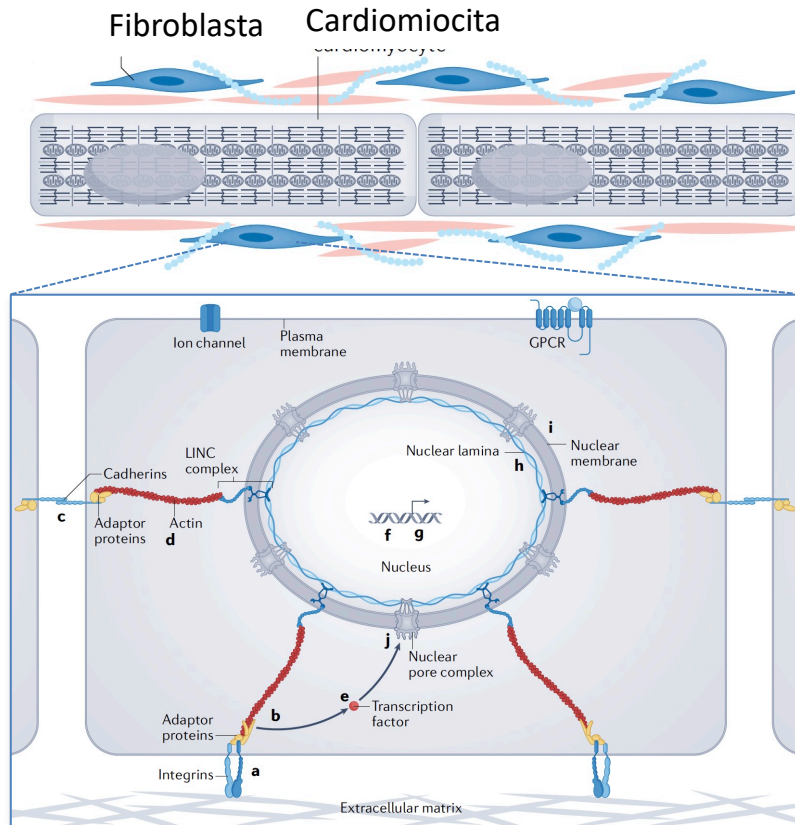
PICP: C-terminal propeptide of type I procollagen



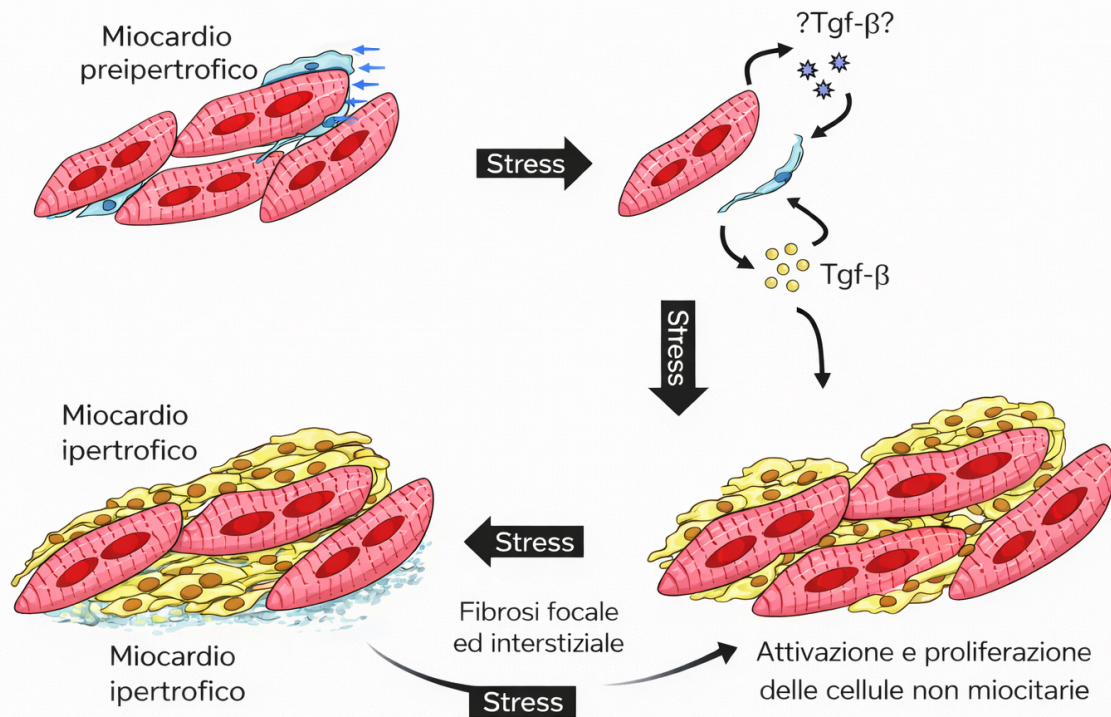
Fibrosi interstiziale nella CMI



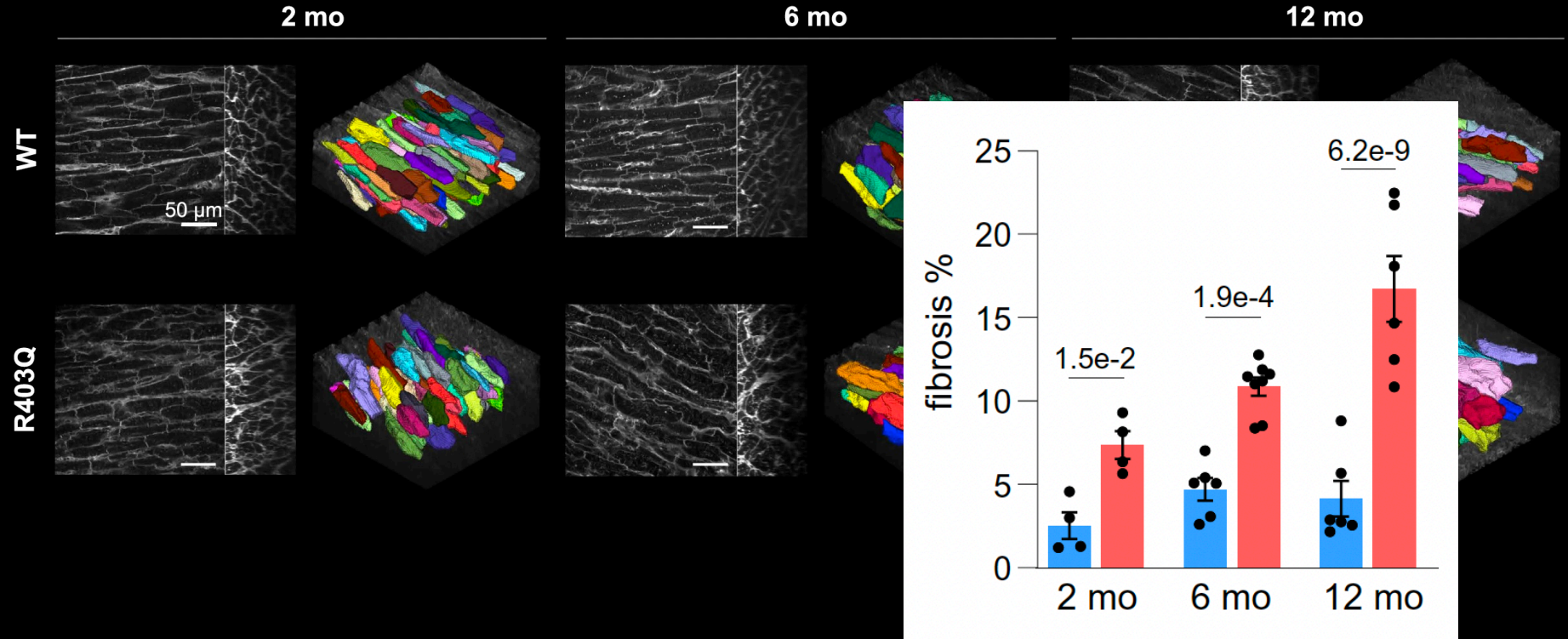
Fibrosi interstiziale nella CMI



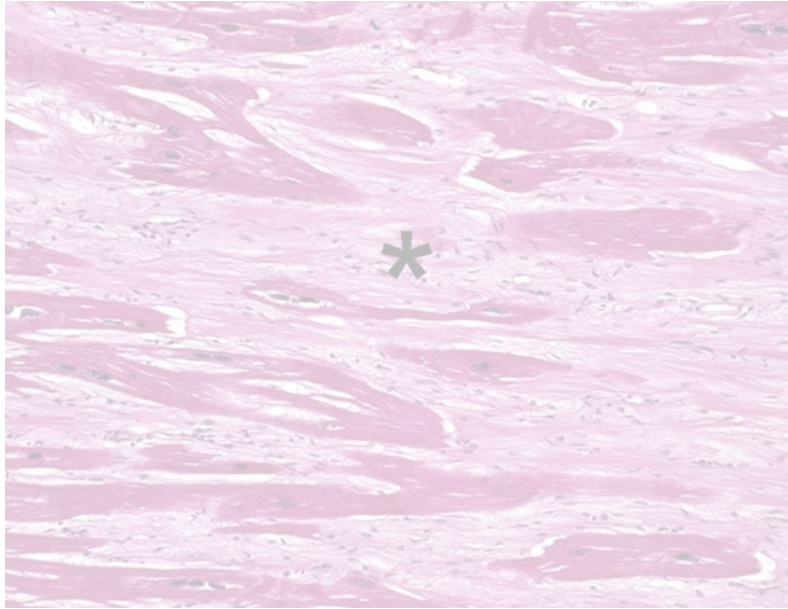
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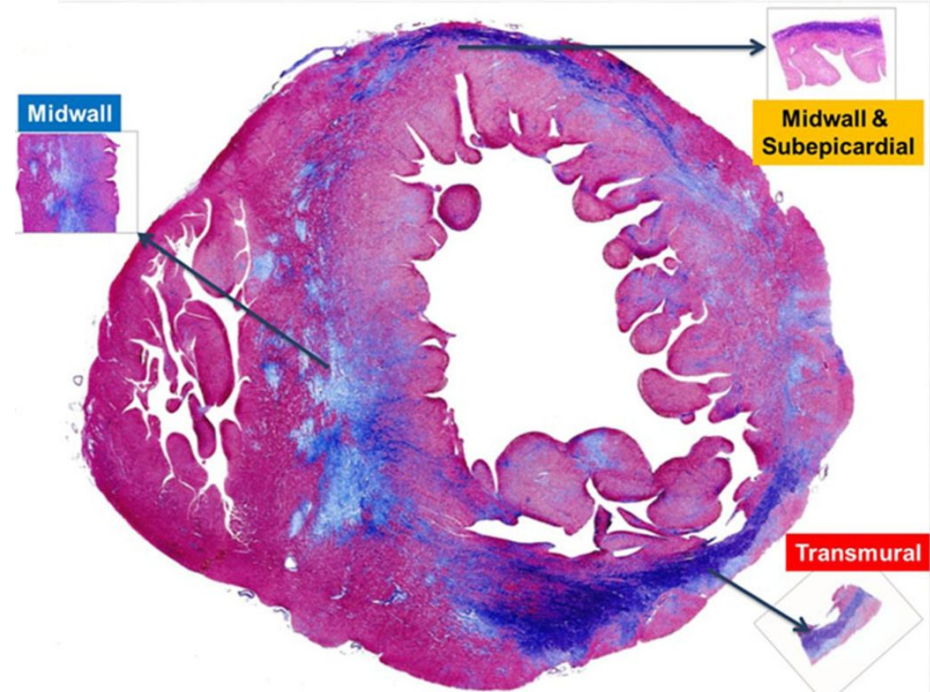
Fibrosi interstiziale nella CMI



Fibrosi interstiziale



Fibrosi sostitutiva



↑ **domanda**

Ipercontrattilità miocitaria

Ipertrofia miocardica

Disfunzione diastolica

Ostruzione all'efflusso

↓ **apporto**

Malattia dei piccoli vasi

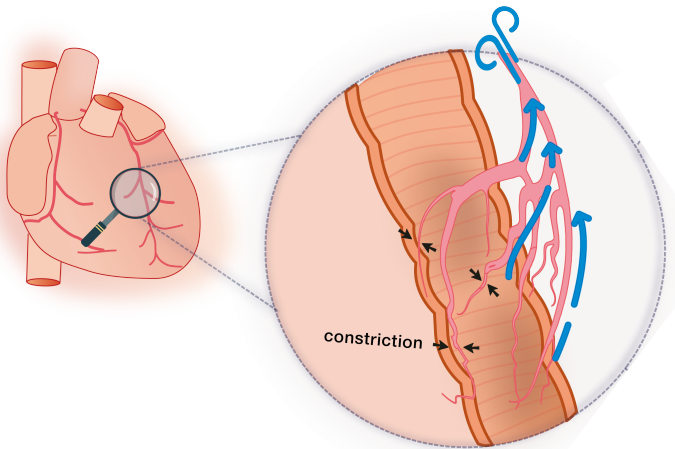
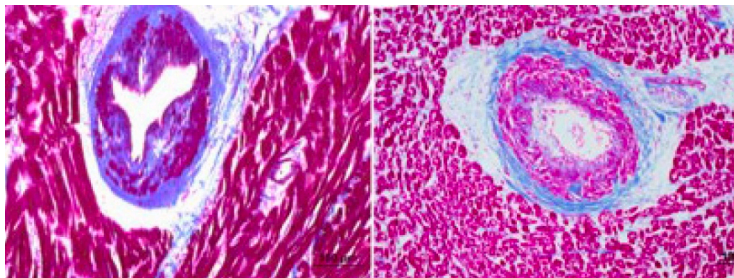
Ridotta densità capillare

Ridotta riserva di

perfusione coronarica



Cause di ischemia nella CMI



↓ **apporto**

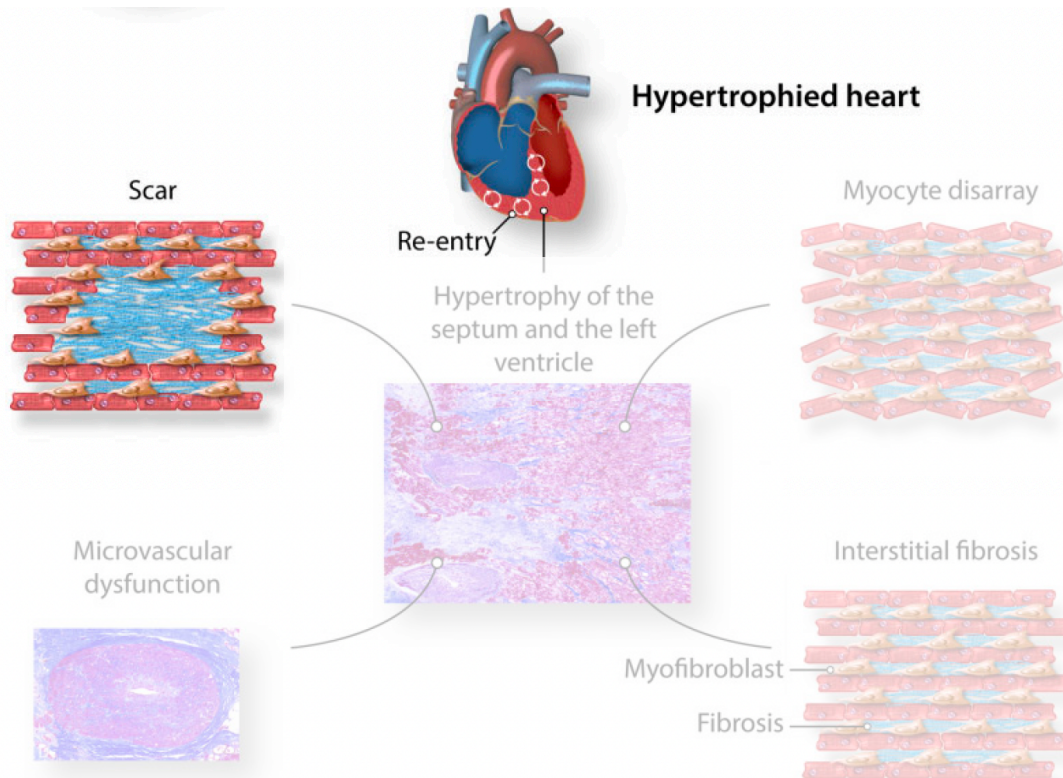
Malattia dei piccoli vasi

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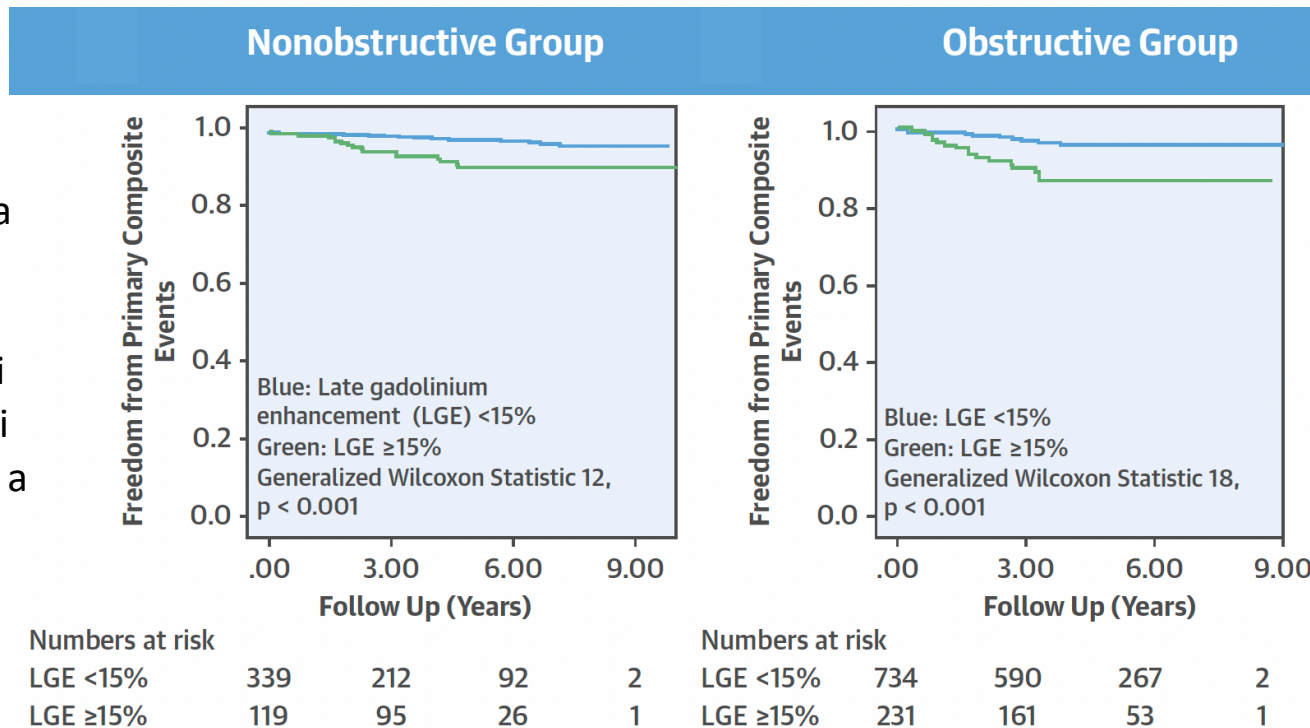


Aritmogenesi nella cardiomiopatia ipertrofica



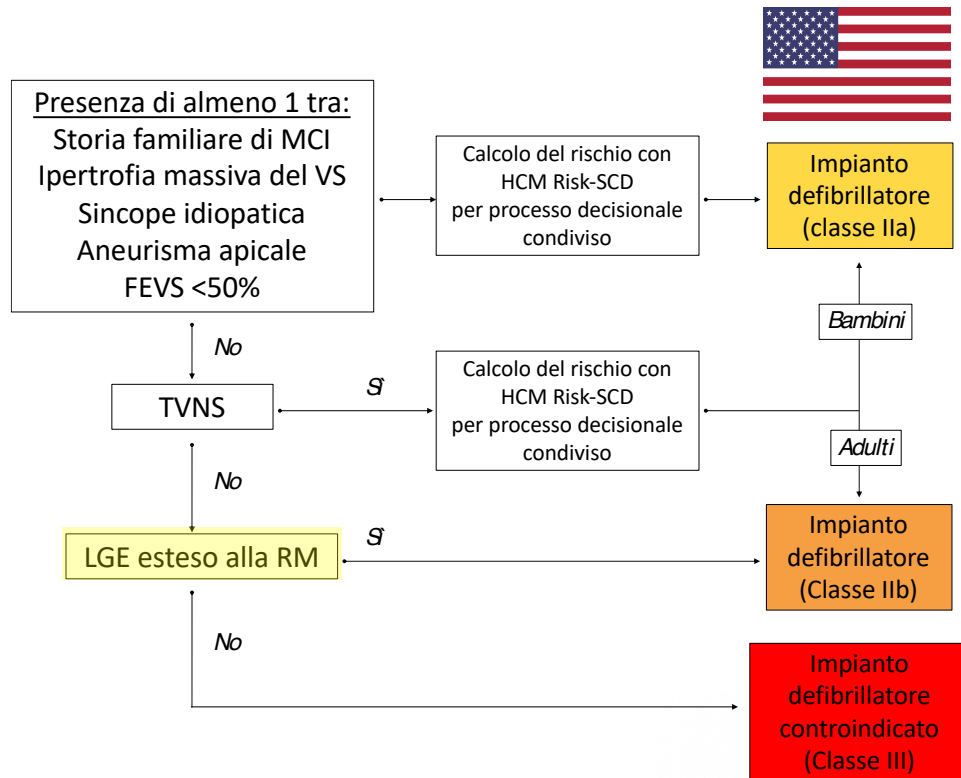
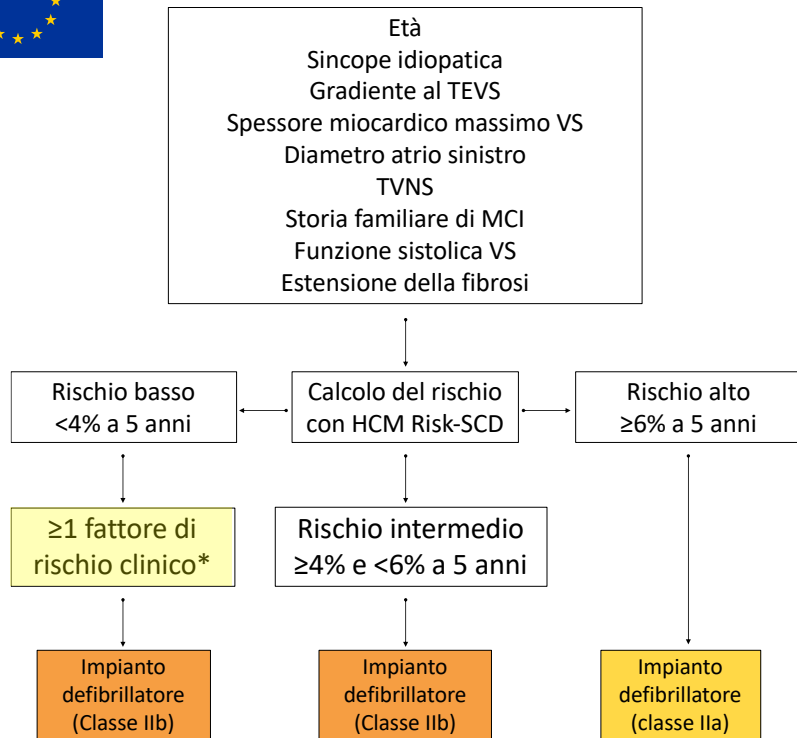
LGE e rischio aritmico nella CMI

1423 pazienti a
rischio basso-
intermedio
Età 56 ± 14 anni
FUP $4,7 \pm 2$ anni
686 sottoposti a
miectomia

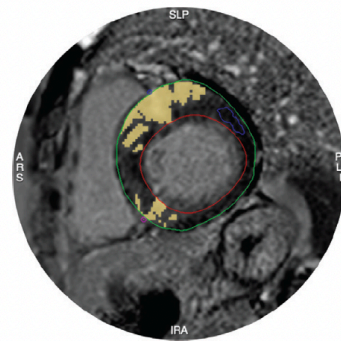
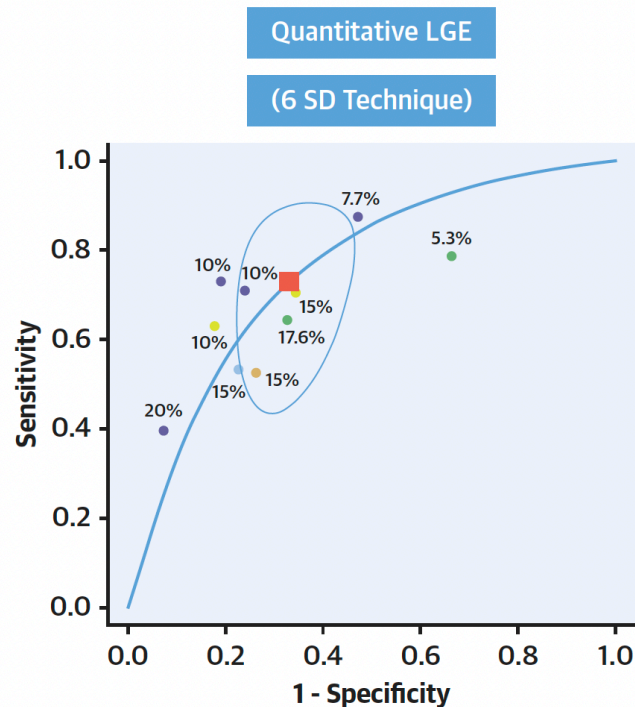
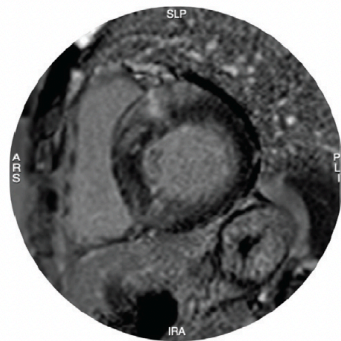


40 SCD
20 interventi
appropriati ICD





LGE e rischio aritmico nella CMI



Metanalisi di 11 studi
5550 pazienti
FUP mediano 5,2 anni
7 studi utilizzano 6 SD

— SROC Curve — Confidence Ellipse ■ Optimal Cutoff LGE 10%

JAMA | Original Investigation

Predictors of Long-Term Outcomes in Hypertrophic Cardiomyopathy The NHLBI HCM Registry

The HCMR Investigators

2698 pazienti, tutti con
RM e biomarcatori (ma
esclusi portatori di ICD!)
Età 50 anni
FUP 6,9 anni

Sudden cardiac death events ^c		
Variables	Hazard ratio per 10-unit change (95% CI) ^d	P value
Late gadolinium enhancement %	1.92 (1.55-2.38)	<.001
LV mass index	1.12 (1.04-1.21)	.004
LV ejection fraction	0.68 (0.52-0.89)	.004
Log(NTProBNP)	1.39 (1.12-1.73)	.003

34 SCD
43 aritmie
ventricolari non
fatali

C index 0,76

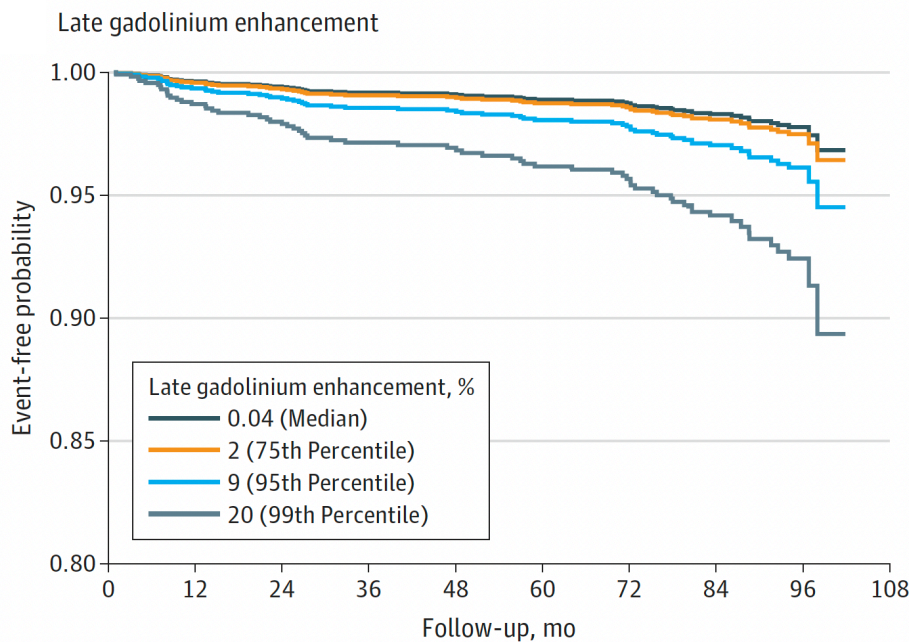


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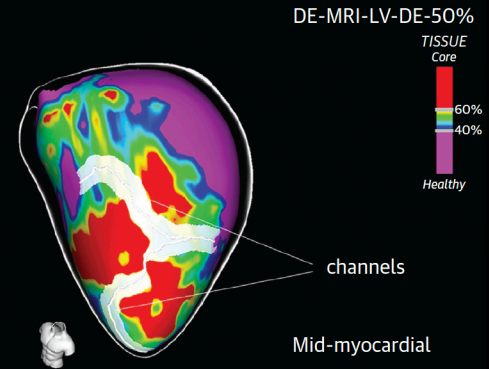
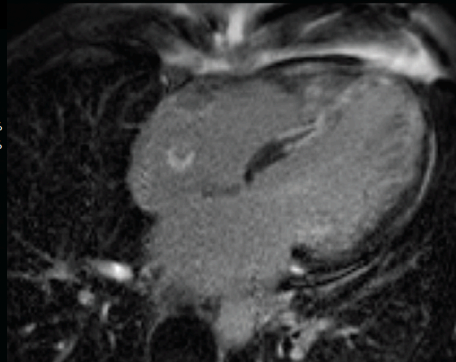
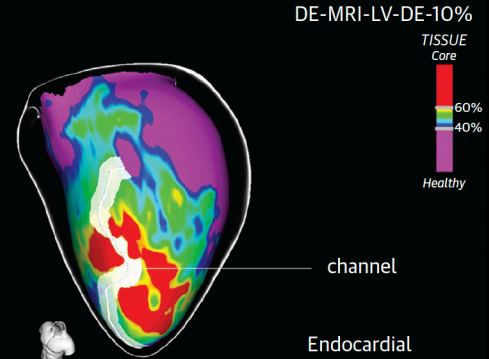
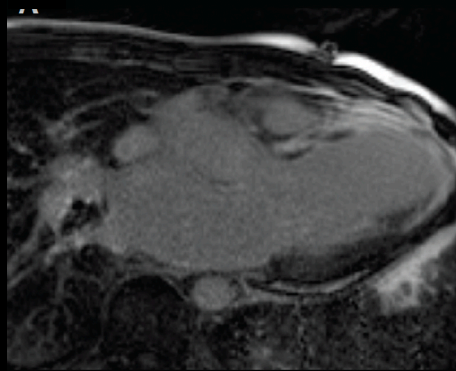
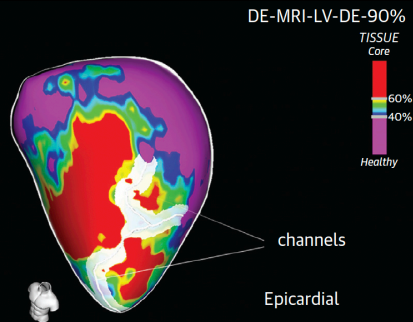
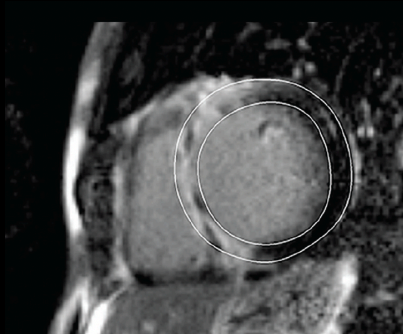


34 SCD
43 aritmie
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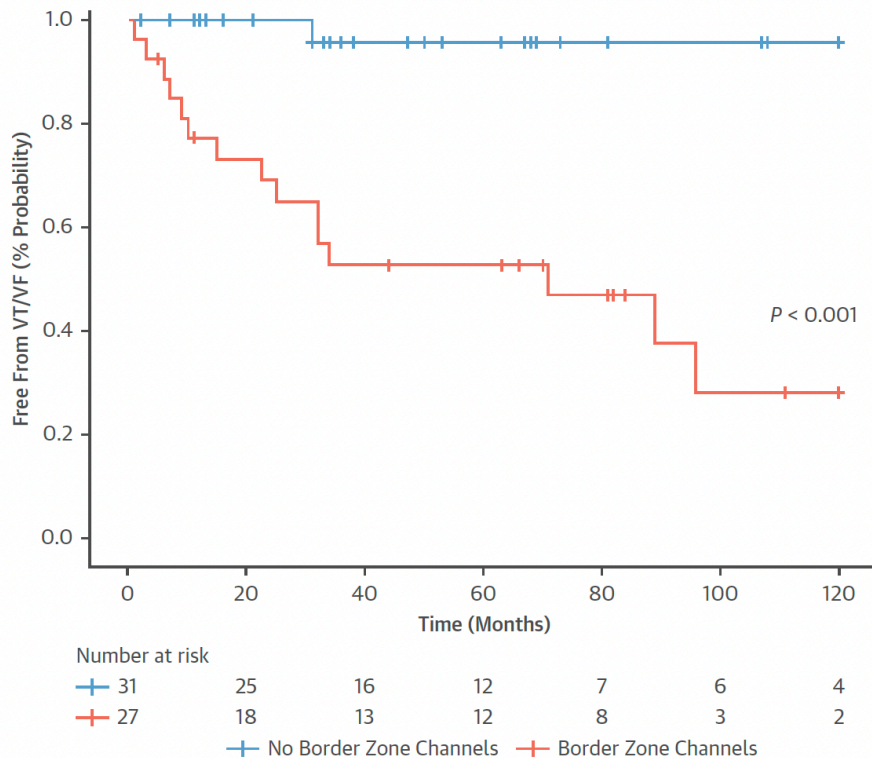
Scar architecture and arrhythmias

58 pazienti sottoposti ad
impianto di ICD
Età 44 ± 14 anni
HCM Risk-SCD: $5,3 \pm 3,2\%$
26% end stage



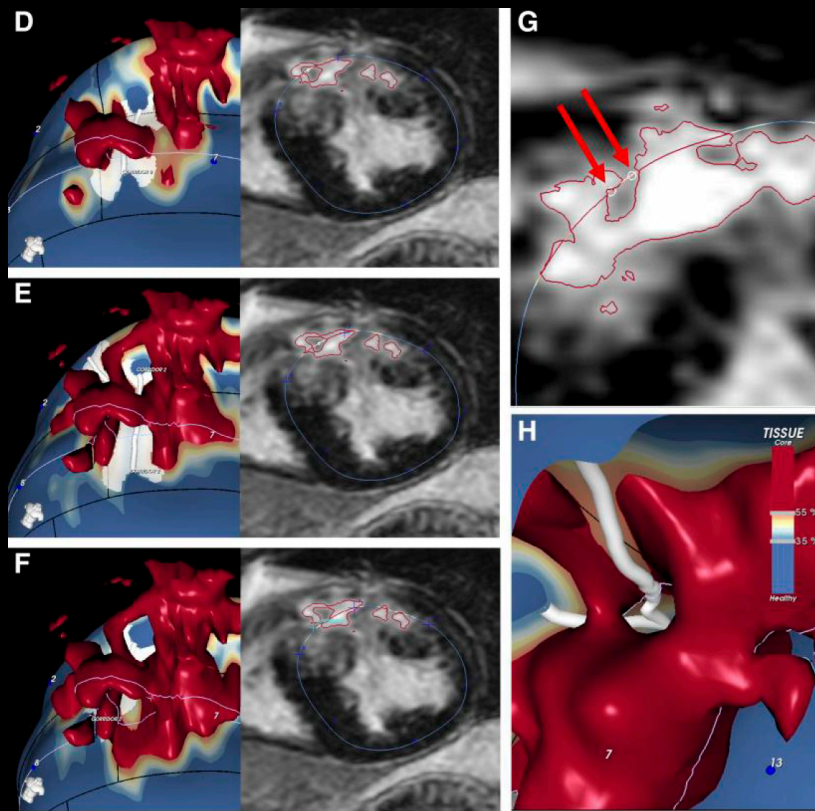
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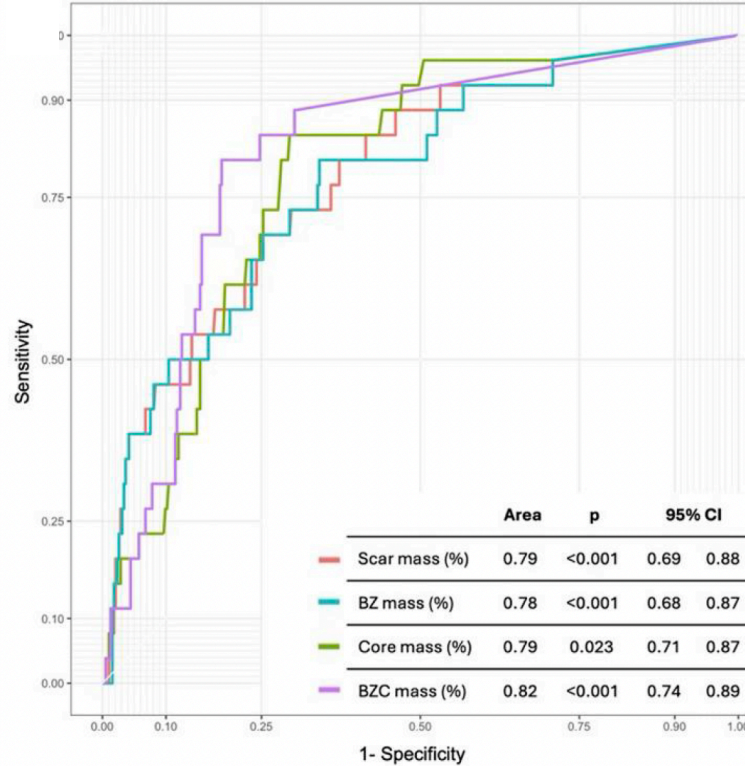
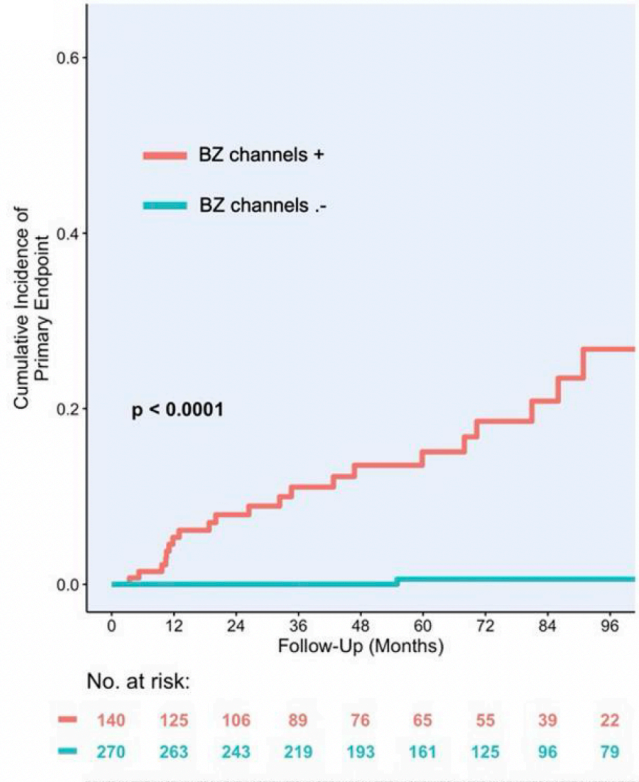


Scar architecture and arrhythmias

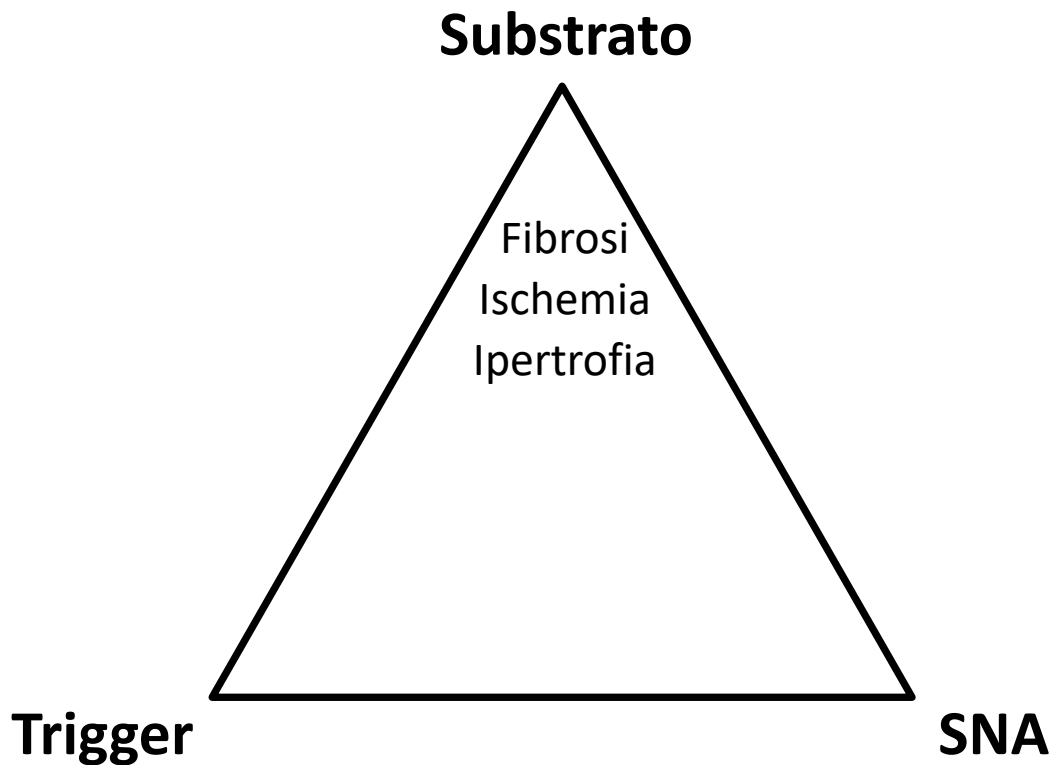
410 pazienti
Età 52 ± 17 anni
26 (6,3%) LVEF < 50%
298 (72,6%) LGE+
140 (47,0%) BZ channels+



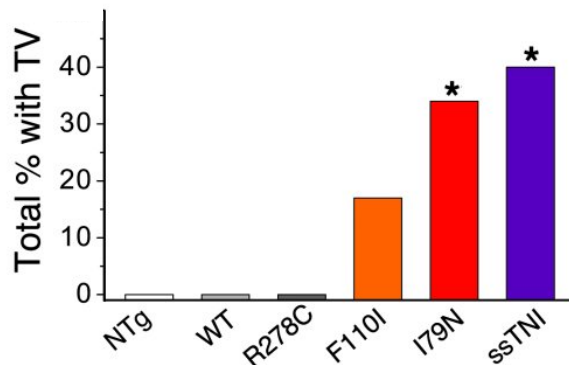
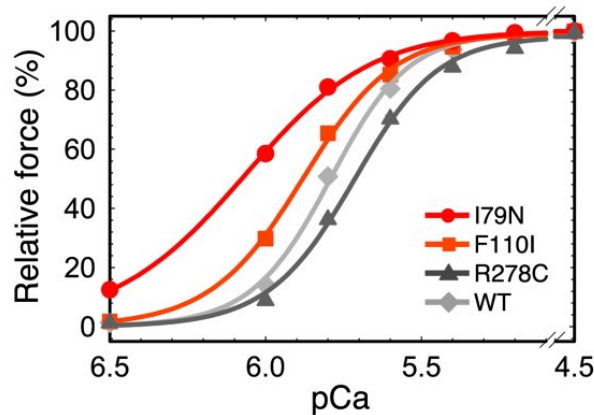
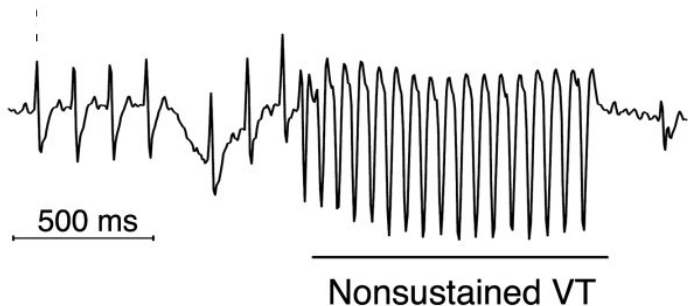
Scar architecture and arrhythmias



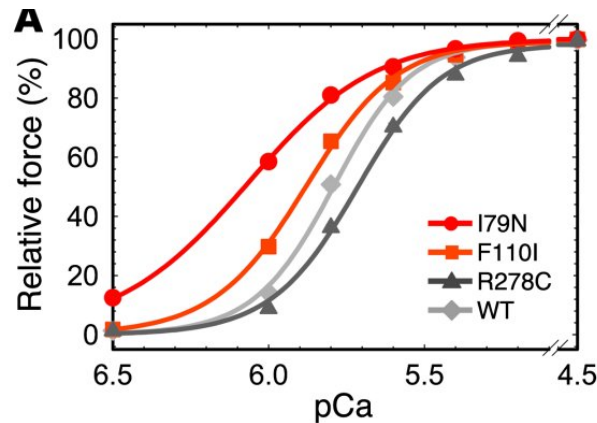
Aritmogenesi nella cardiomiopatia ipertrofica



Modello murino che esprime varianti della **troponina T umana** associate ad HCM



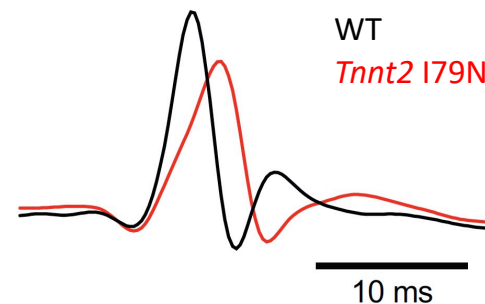
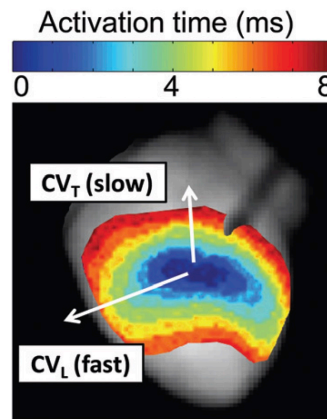
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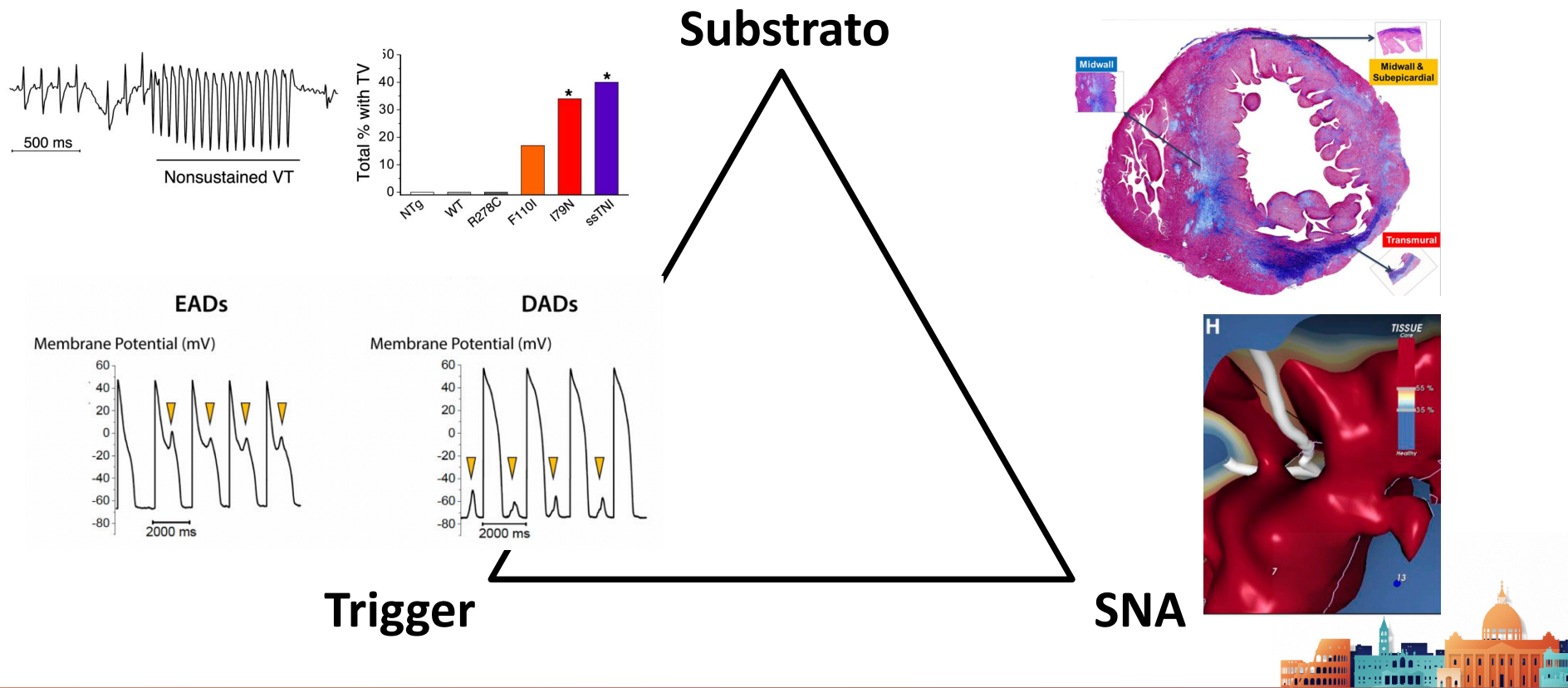


↑ consumo ATP



→ ↓ Connexin 43 →







OSPEDALE POLICLINICO SAN MARTINO
Sistema Sanitario Regione Liguria



UNIVERSITÀ DEGLI STUDI
DI GENOVA

Grazie per l'attenzione!



Ambulatorio Cardiomiopatie

Marco Canepa – Edoardo Bertero - Marianna Eleonora Labate - Pier Filippo Vianello