Diagnosi e Terapia delle Aritmie Cardiache: Ablazione transcatetere endo-epicardica di Tachicardia Ventricolare in paziente con Malattia di Anderson-Fabry

Martedì dell'Ordine
15 Aprile 2014

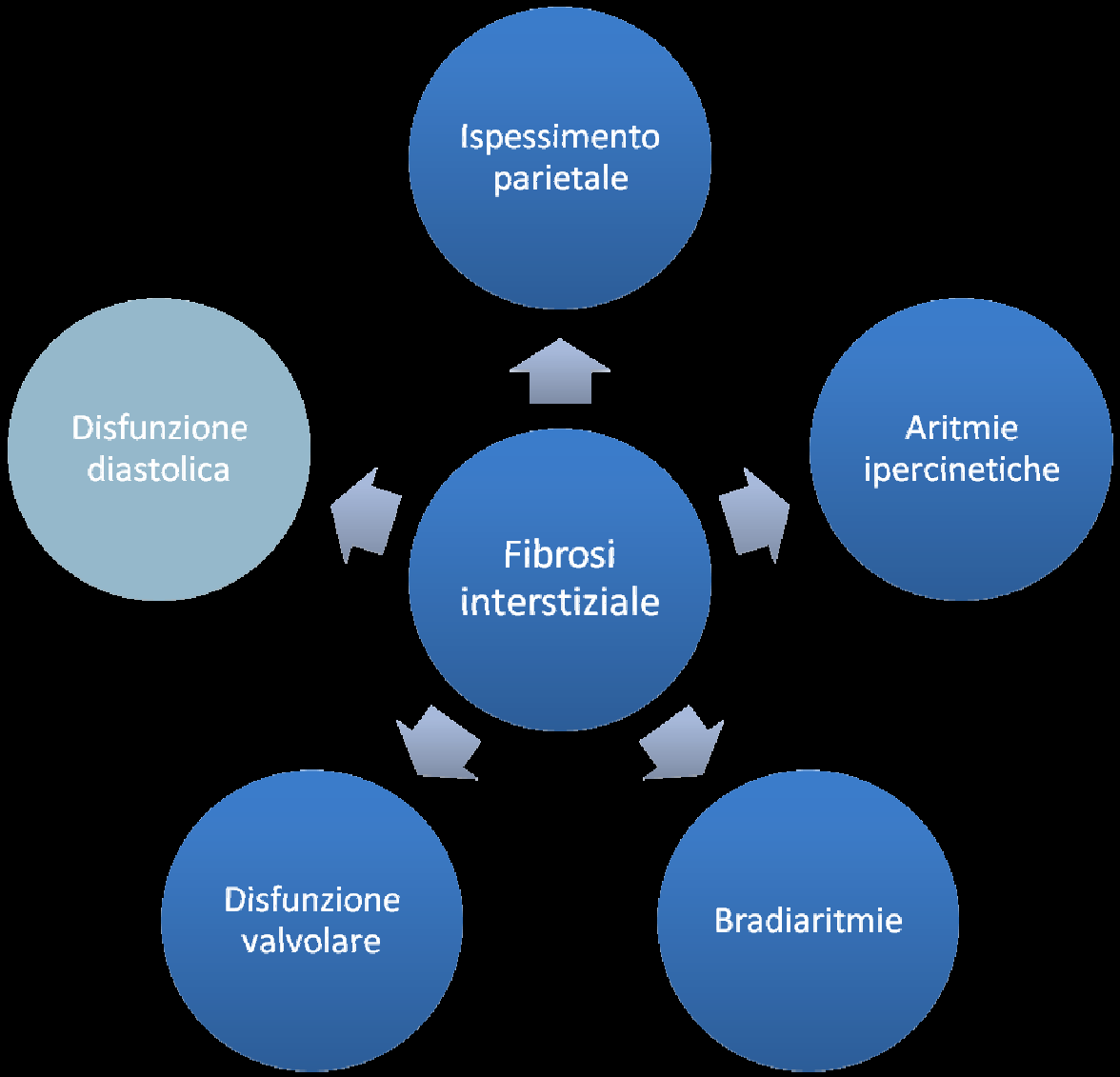
Laboratorio di Elettrofisiologia ed Elettrostimolazione, U.O. Cardiologia, A.O.U. di Parma

La malattia di Anderson-Fabry

Malattia da accumulo lisosomiale, recessiva, legata al cromosoma x, causata dal deficit dell'enzima alfa-galattosidasi A coinvolto nel catabolismo lipidico cellulare

Accumulo di glicosfingolipidi a livello cellulare con disfunzione cellulare, tessutale e danno d'organo (cuore, reni, cute, SN)

La malattia di Anderson-Fabry

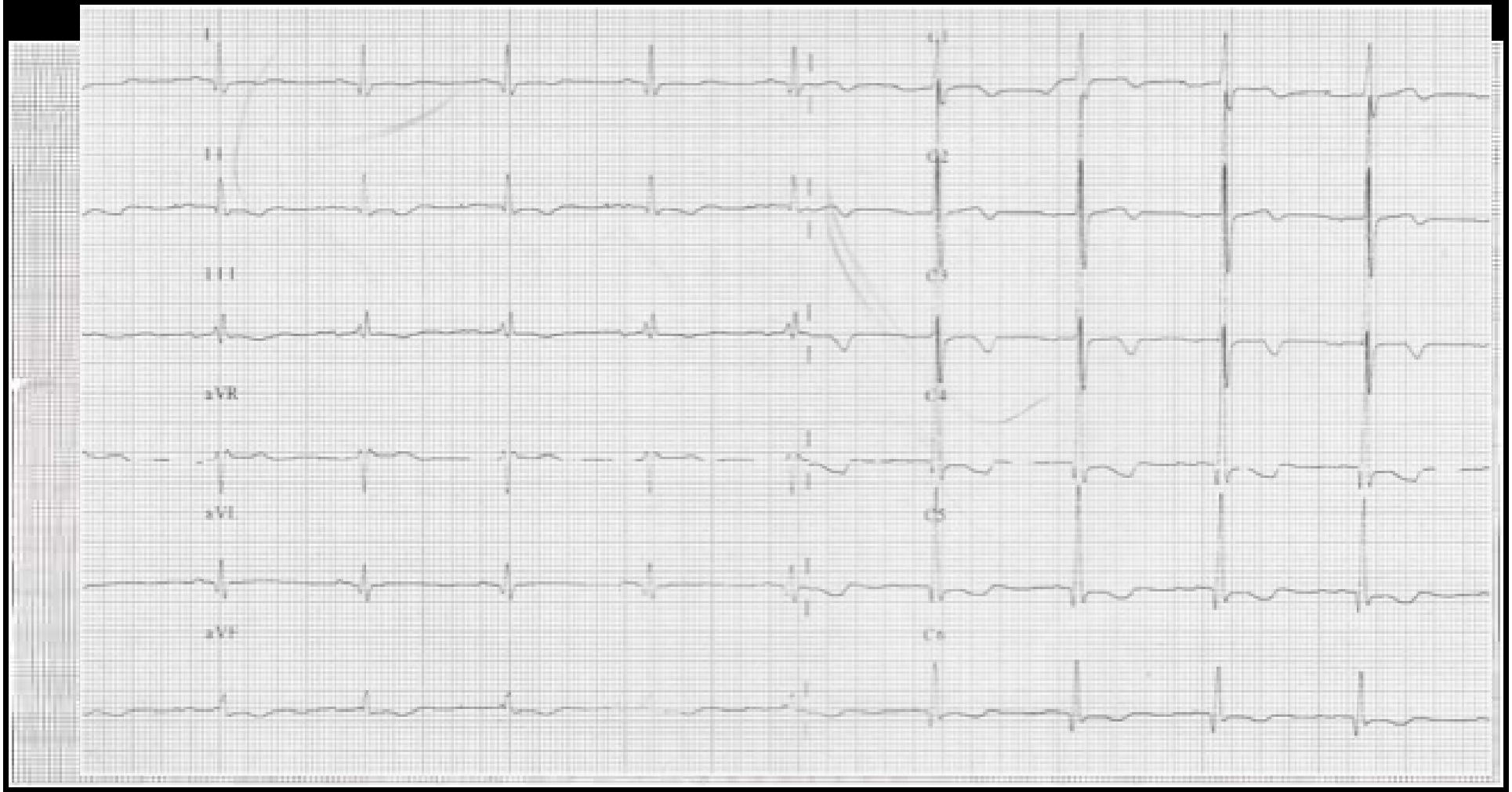


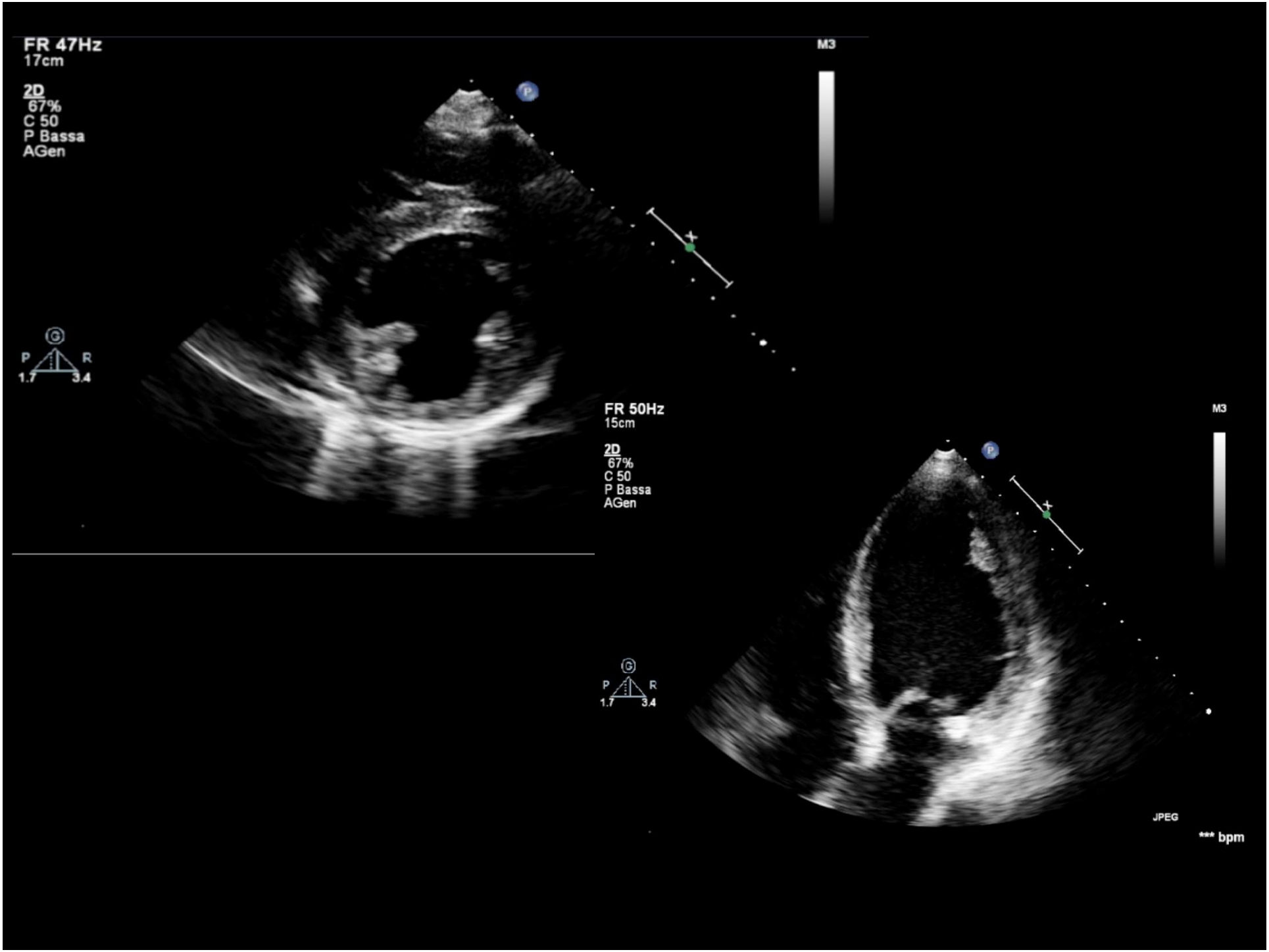
Case Report

- Maschio di 54 anni
- IRC nota da circa 15 anni
- “Cardiopatia ipertrofica” in F.U.
- Familiarità per morte cardiaca improvvisa
- Angiocheratoma dello scroto e natiche
- Ipoacusia neurosensoriale
- Deficit di Alfa-Galattosidasi A da mutazione del gene

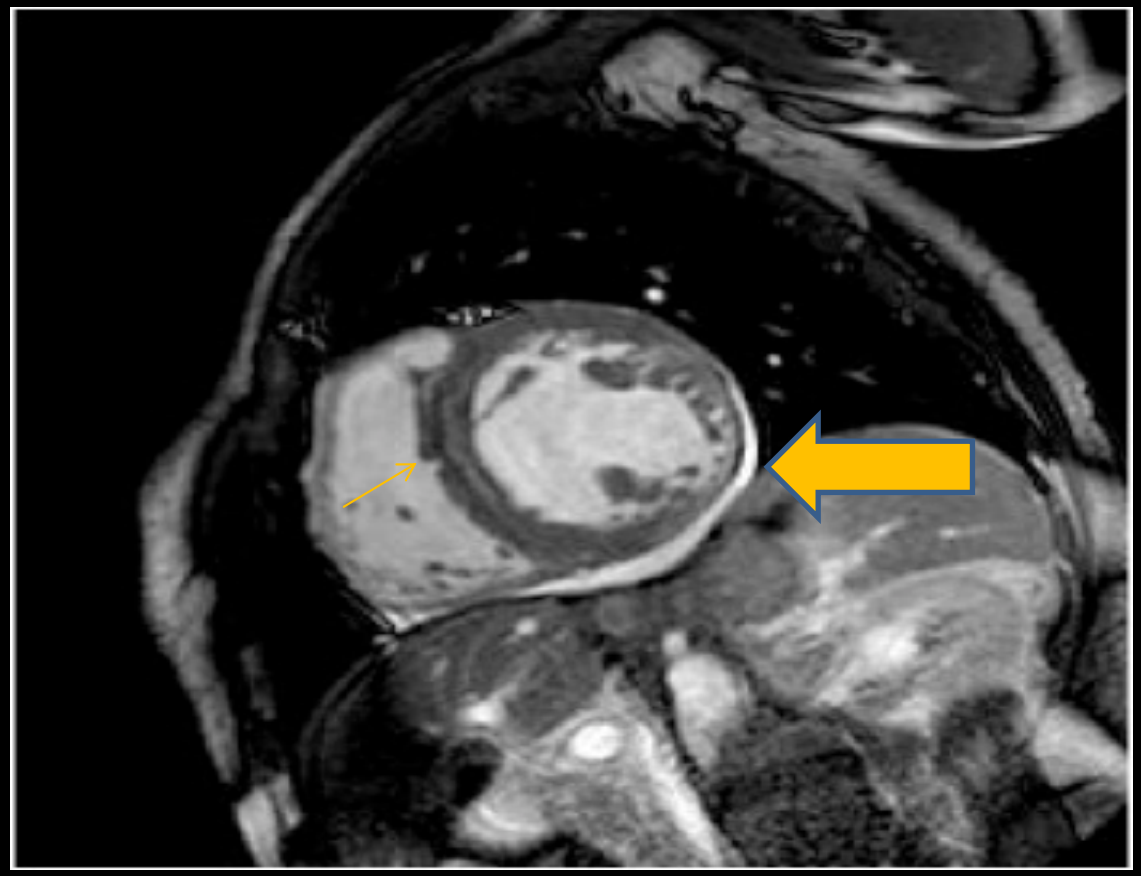
malattia di Anderson-Fabry

Giunge in PS sintomatico per cardiopalmo,
Instabilità emodinamica




FR 47Hz
17cm
2D
67%
C 50
P Bassa
AGen
M3
P
R
1.7
3.4
FR 50Hz
15cm
2D
67%
C 50
P Bassa
AGen
M3
P
R
1.7
3.4
JPEG
*** bpm

- Coronarografia: negativa
- RMN cuore

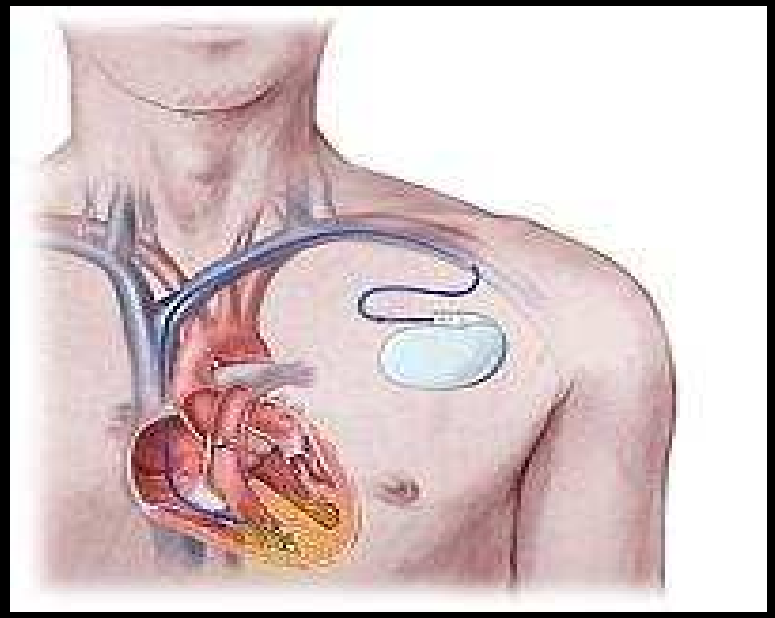


Quadro compatibile con malattia di Anderson Fabry

Recidive di TV → terapia con am[illegible]e e carvedilolo

↓

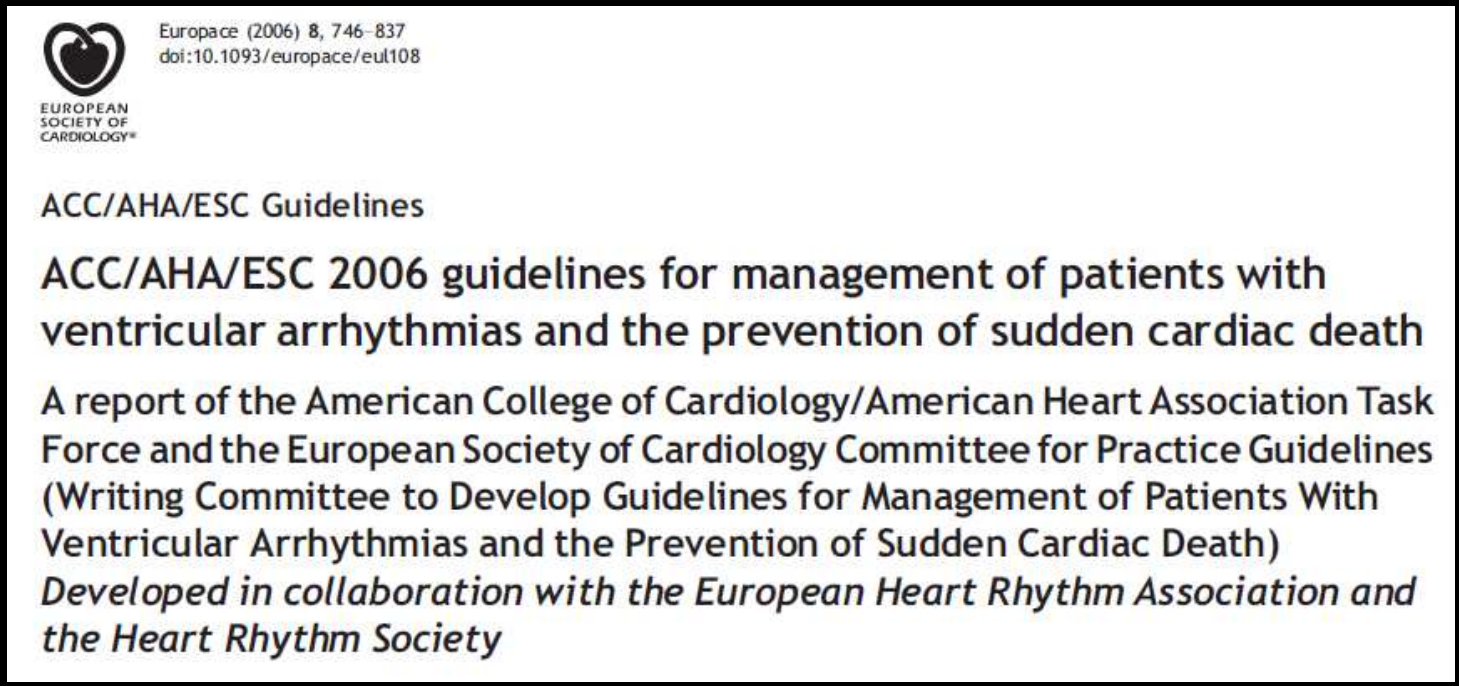
sotalolo



Impianto di defibrillatore bicamerale

- Nei mesi successivi numerosi ricoveri per recidive aritmiche trattate con overdrive pacing e/o DC shock

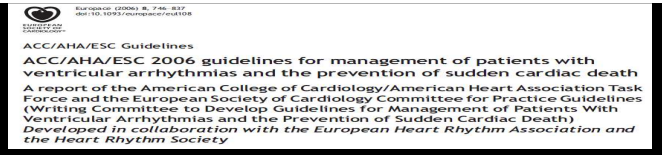
- Il paziente accetta la proposta della procedura di ablazione transcatetere dell'aritmia, precedentemente rifiutata



8.4.2. Infiltrative cardiomyopathies

In addition to managing the underlying infiltrative cardiomyopathy, life-threatening arrhythmias **should be treated in the same manner that such arrhythmias** are treated in patients with other cardiomyopathies, including the use of ICD and pacemakers in patients who are receiving chronic optimal medical therapy and who have reasonable expectation of survival with a good functional status for more than 1 y. (Level of Evidence: C)

6.6. Ablation Recommendations



Class I

(1) Ablation is indicated in patients who are otherwise at low risk for SCD and have sustained predominantly monomorphic VT that is drug resistant, who are drug intolerant, or who do not wish long-term drug therapy. (Level of Evidence: C)

(2) Ablation is indicated in patients with bundle-branch reentrant VT. (Level of Evidence: C)

(3) Ablation is indicated as adjunctive therapy in patients with an ICD who are receiving multiple shocks as a result of sustained VT that is not manageable by reprogramming or changing drug therapy or who do not wish long-term drug therapy (206,278). (Level of Evidence: C)

(4) Ablation is indicated in patients with WPW syndrome resuscitated from sudden cardiac arrest due to AF and rapid conduction over the accessory pathway causing
VF (279). (Level of Evidence: B)

Class Iia (1) Ablation can be useful therapy in patients who are otherwise at low risk for SCD and have symptomatic nonsustained monomorphic VT that is drug resistant, who are drug intolerant or who do not wish long-term drug therapy. (Level of Evidence: C)

(2) Ablation can be useful therapy in patients who are otherwise at low risk for SCD and have frequent symptomatic predominantly monomorphic PVCs that are drug
resistant or who are drug intolerant or who do not wish long-term drug therapy. (Level of Evidence: C)

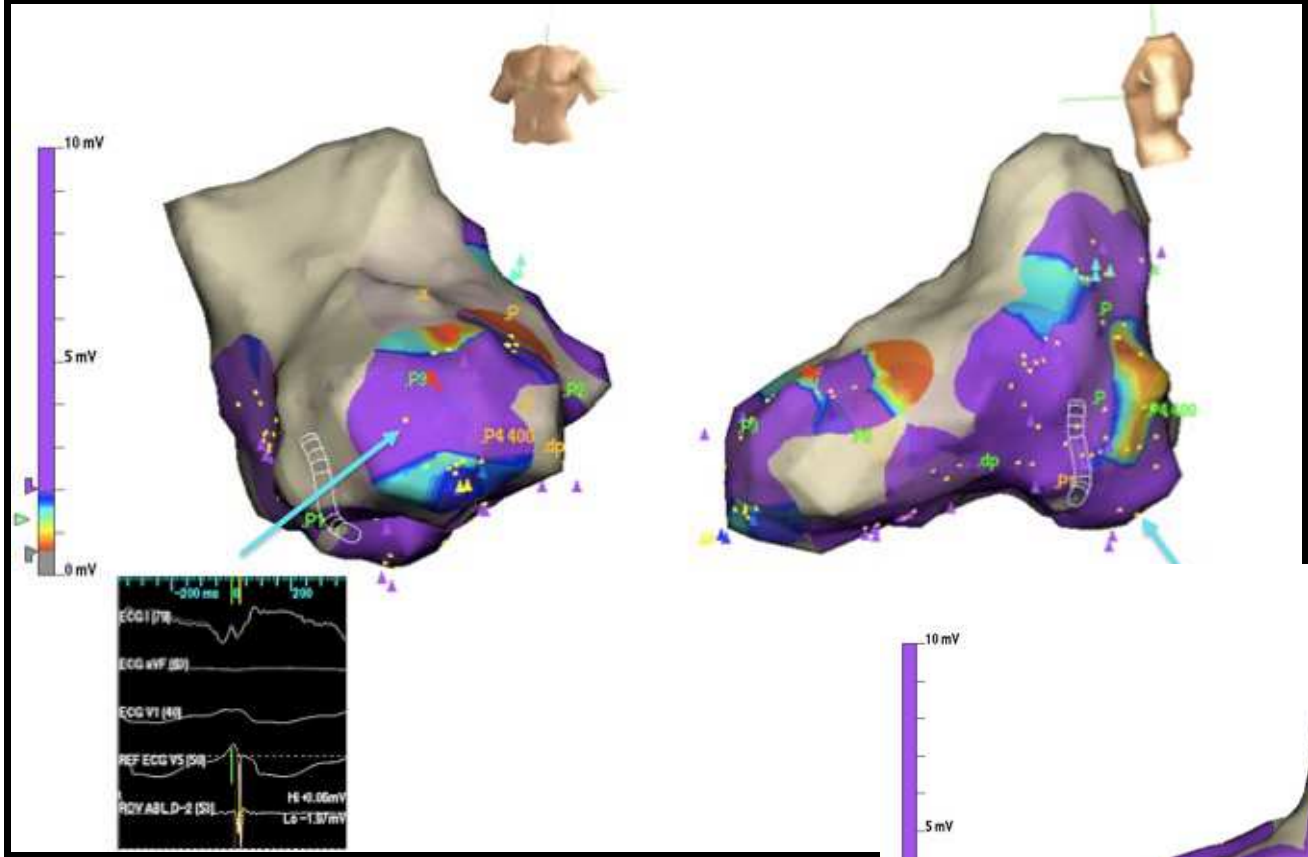
(3) Ablation can be useful in symptomatic patients with WPW syndrome who have accessory pathways with refractory periods less than 240 ms in duration (279).
(Level of Evidence: B)

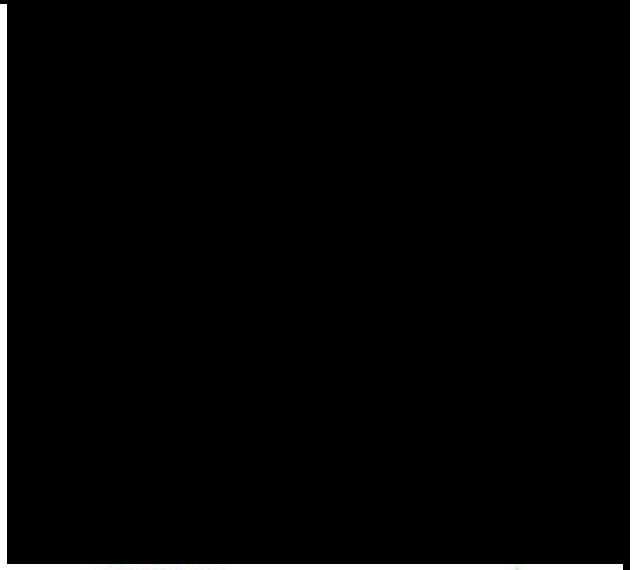
Class IIb

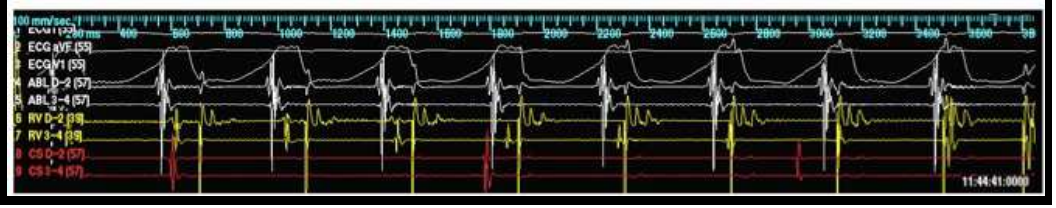
(1) Ablation of Purkinje fiber potentials may be considered in patients with ventricular arrhythmia storm consistently provoked by PVCs of similar morphology (280).
(Level of Evidence: C)

(2) Ablation of asymptomatic PVCs may be considered when the PVCs are very frequent to avoid or treat tachycardia-induced cardiomyopathy (281). (Level ofEvidence: C)

Mappaggio endocardico del substrato della TV







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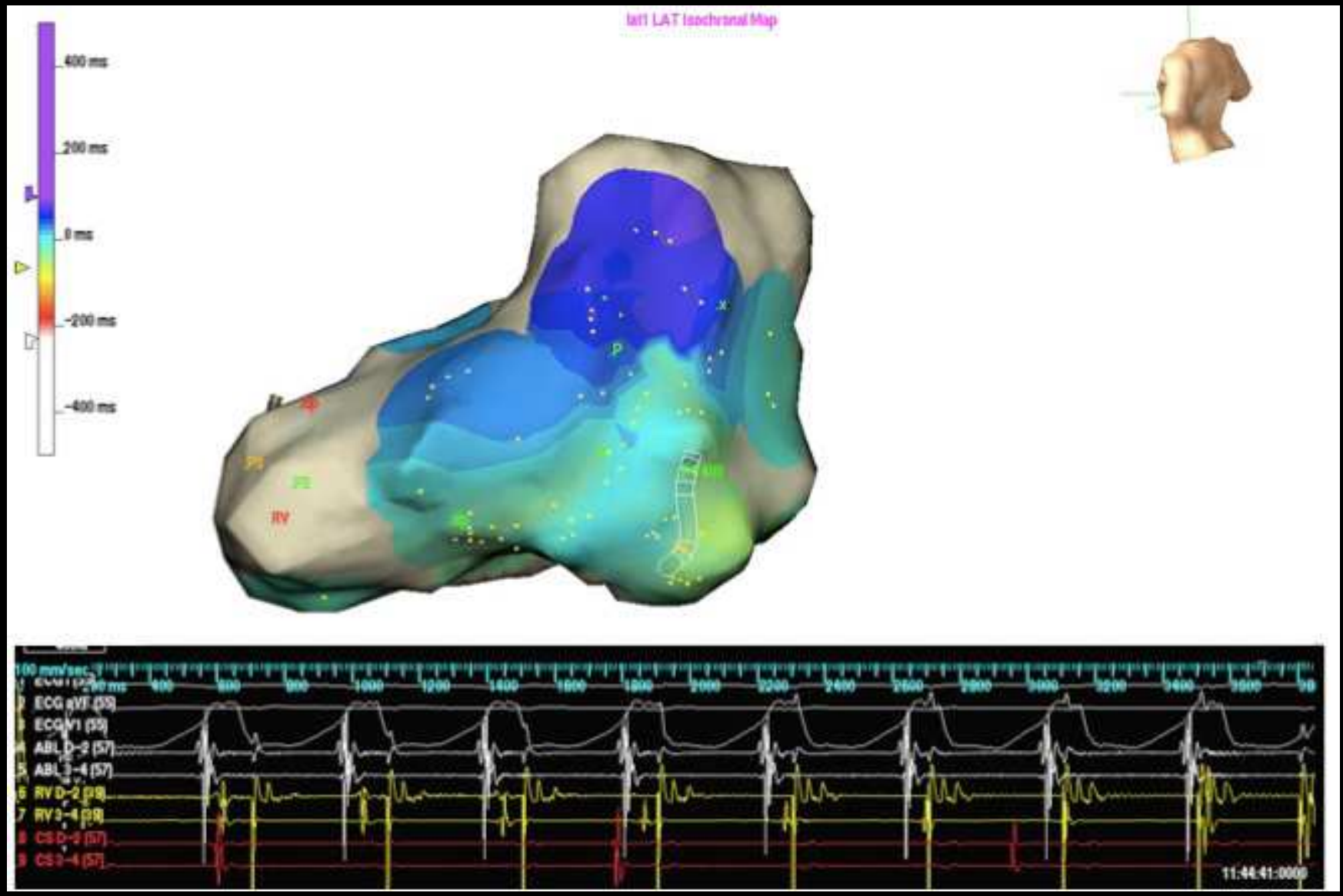
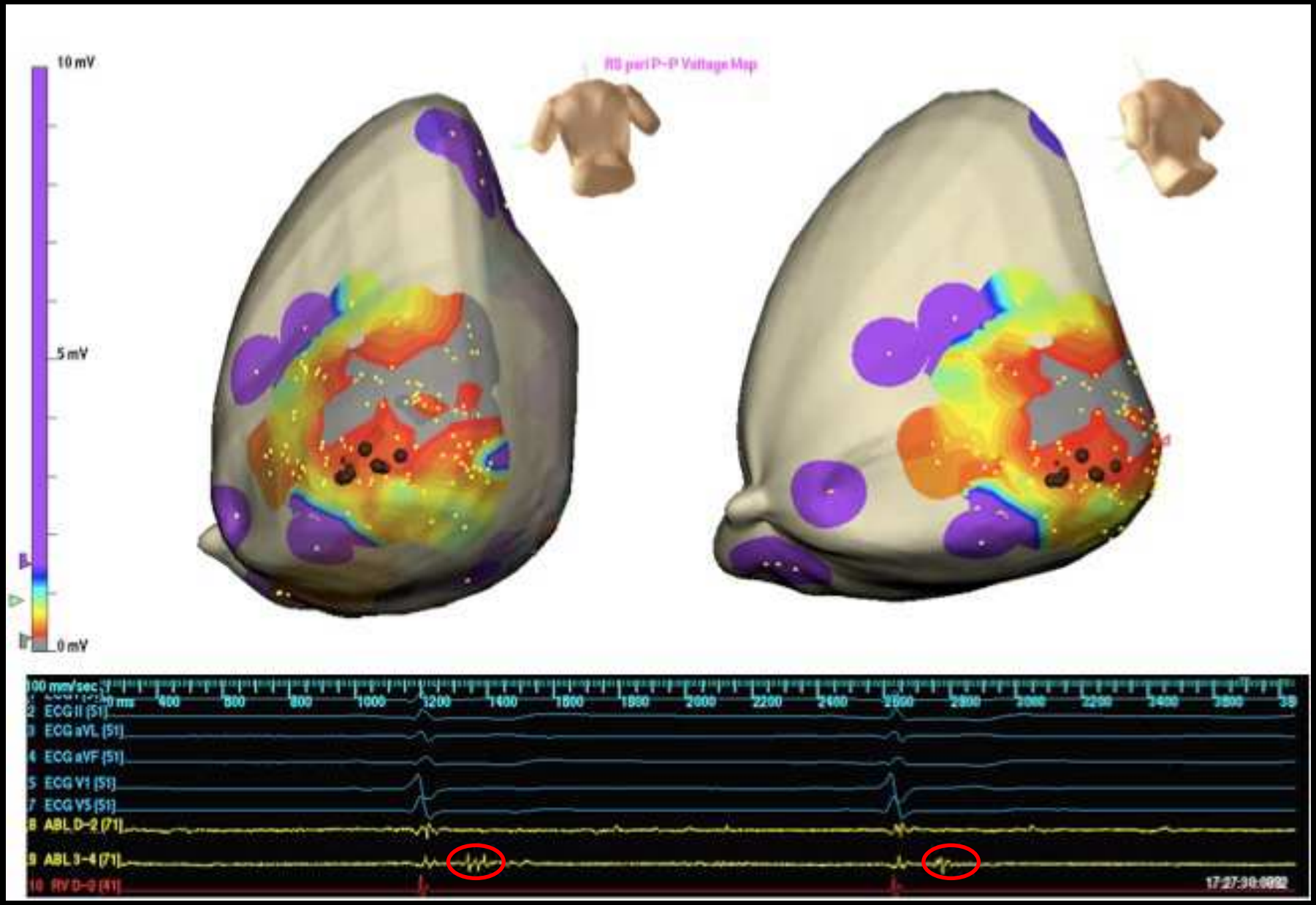
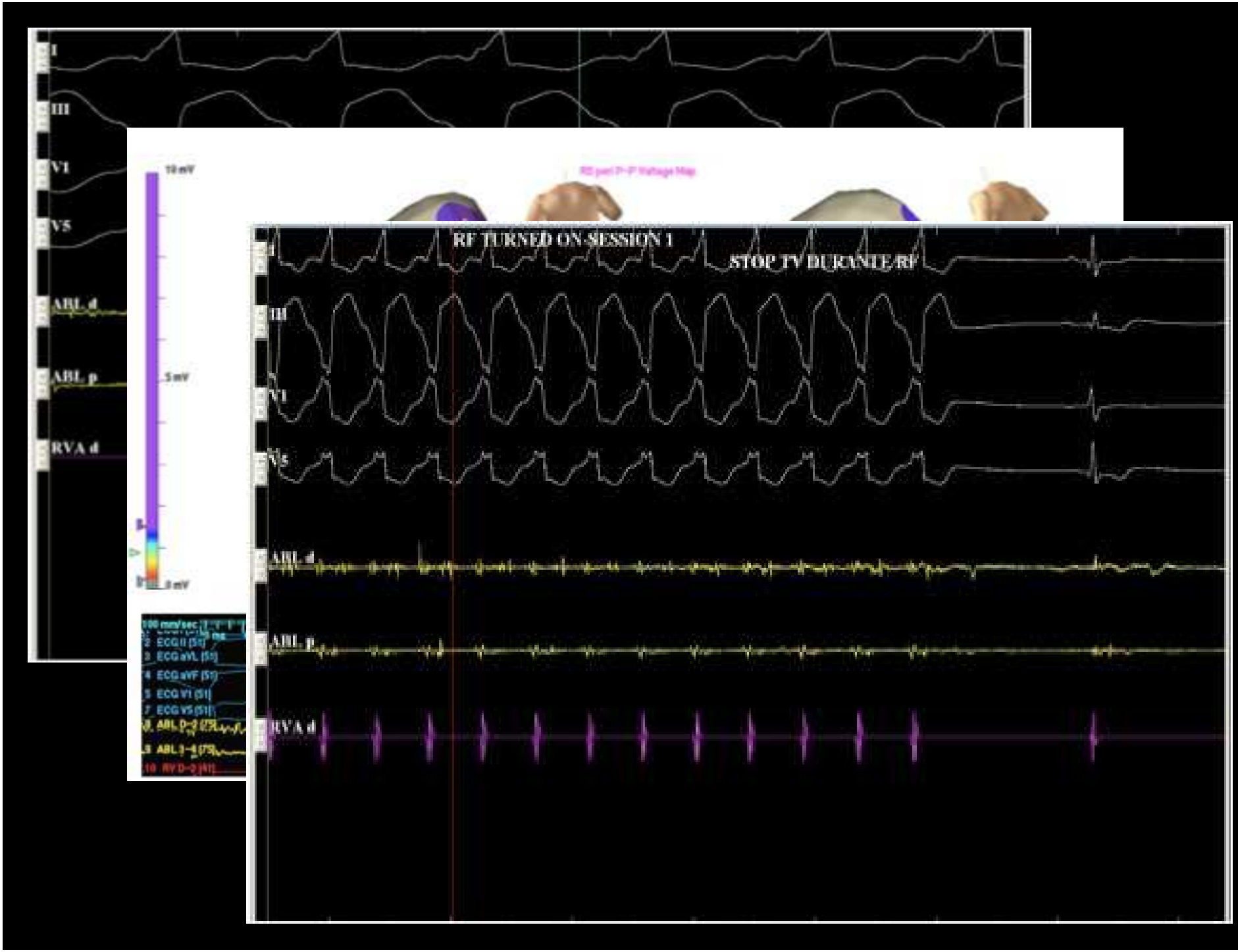
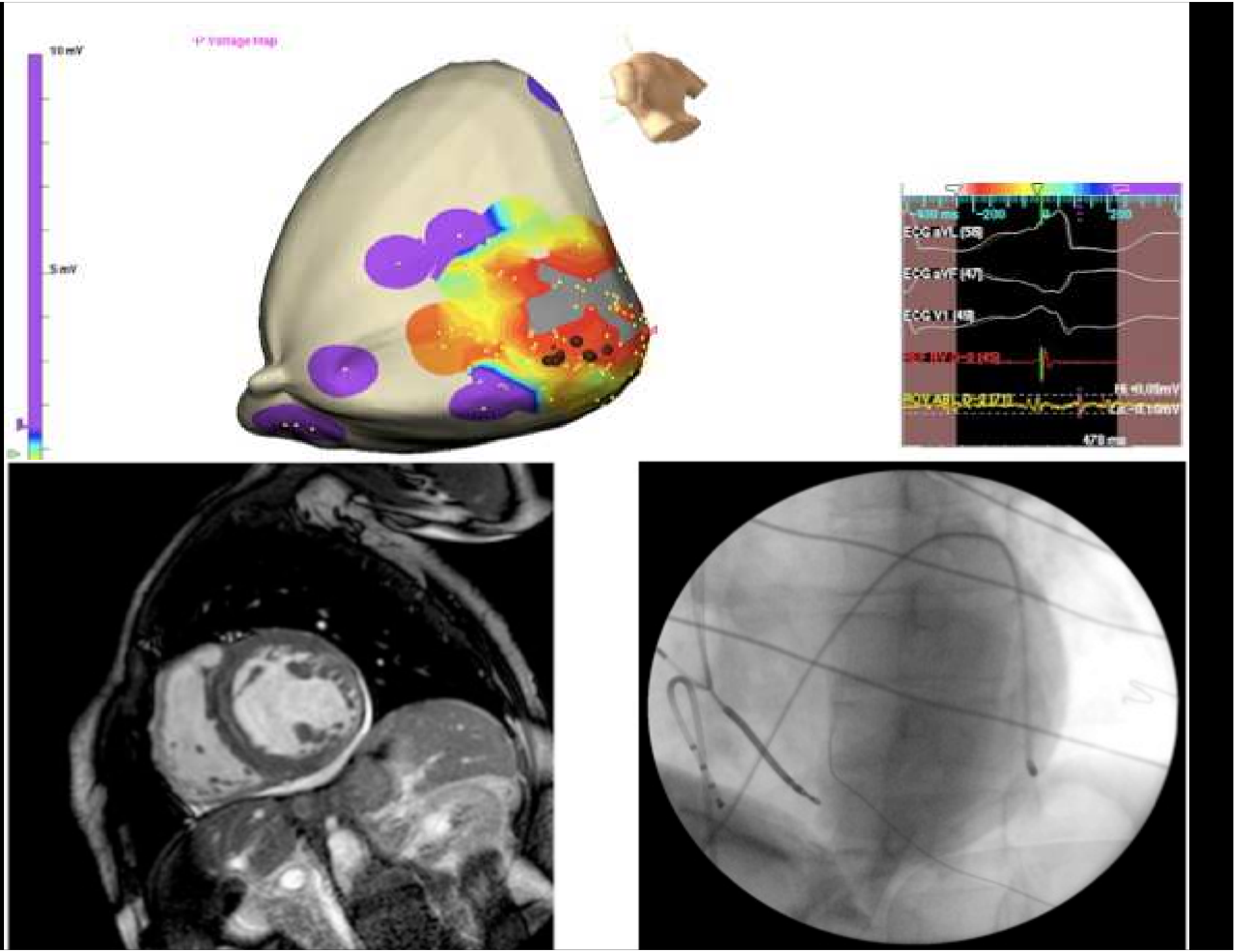


Mappaggio ed Ablazione epicardica della TV




I
III
V1
V5
ABL d
ABL p
RVA d
10 mV
5 mV
0 mV
RF TURNED ON-SESSION 1
STOP TV DURANTE RF
I
III
V1
V5
ABL d
ABL p
RVA d


10 mV
5 mV
ECG aVL
ECG aVF
ECG V1

- Procedura efficace in acuto
- Assenza di complicanze
- F.U. Clinico > 12 mesi: non ulteriori recidive cliniche in assenza di terapia antiaritmica
- Interrogazione dell'ICD: non registrazione di ulteriori episodi aritmici

Conclusioni

- L’ablazione della tachicardia ventricolare è indicata in classe I nel trattamento di TV refrattarie alla terapia farmacologica, in presenza o meno di cardiopatia strutturale, anche al fine di prevenire e/o ridurre il numero di interventi dell’ICD.
- Un approccio epicardico è indicato nei pazienti in cui il mappaggio endocardico non sia stato in grado di identificare il substrato aritmico, nei pazienti con doppia protesi valvolare meccanica mitro-aortica, nei pazienti con evidenza di trombosi ventricolare sinistra .

Conclusioni

- un approccio combinato endo-epicardico è possibile, ma in questo caso sarebbe preferibile predisporre l'accesso epicardico prima di iniziare il mappaggio endocardico e quindi prima di scoagulare il paziente