RUOLO DELLA RM E DELLA TC NELLA VALUTAZIONE DEI PAZIENTI CON CARDIOMIOPATIA.

Come orientare la scelta tra i due esami.
Quali risultati attendersi.

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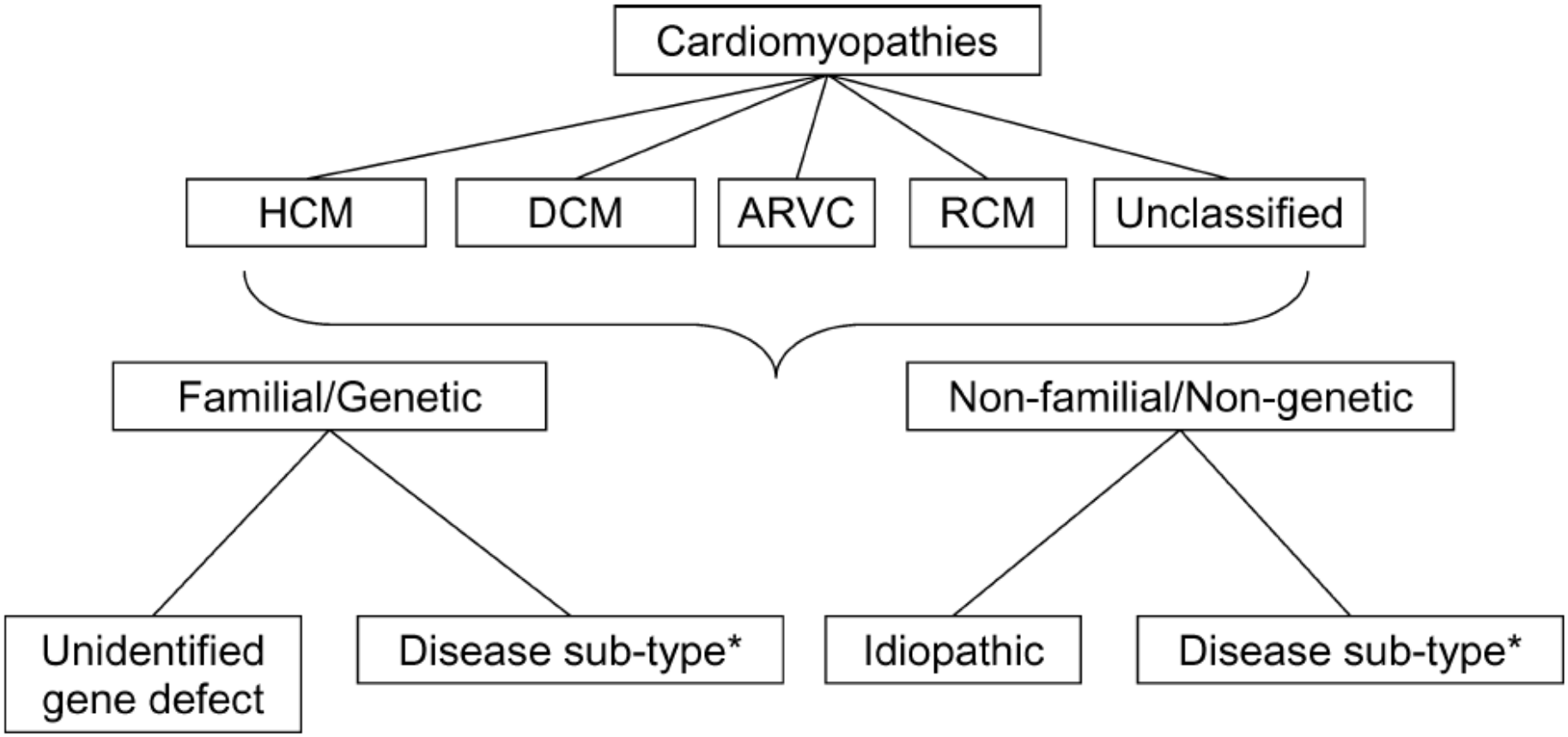
Table 1 Examples of different diseases that cause cardiomyopathies

	HCM	DCM	ARVC	RCM	Unclassified
Familial	Familial, unknown gene Sarcomeric protein mutations β myosin heavy chain Cardiac myosin binding protein C Cardiac troponin I Troponin-T α-tropomyosin Essential myosin light chain Regulatory myosin light chain Cardiac actin α-myosin heavy chain Titin Troponin C Muscle LIM protein Glycogen storage disease (e.g. Pompe; PRKAG2, Forbes', Danon) Lysosomal storage diseases (e.g. Anderson–Fabry, Hurler's) Disorders of fatty acid metabolism Carnitine deficiency Phosphorylase B kinase deficiency Mitochondrial cytopathies Syndromic HCM Noonan's syndrome LEOPARD syndrome Friedreich's ataxia Beckwith–Wiedermann syndrome Swyer's syndrome Other Phospholamban promoter Familial amyloid	Familial, unknown gene Sarcomeric protein mutations (see HCM) Z-band Muscle LIM protein TCAP Cytoskeletal genes Dystrophin Desmin Metavinculin Sarcoglycan complex CRYAB Epicardin Nuclear membrane Lamin A/C Emerin Mildly dilated CM Intercalated disc protein mutations (see ARVC) Mitochondrial cytopathy	Familial, unknown gene Intercalated disc protein mutations Plakoglobin Desmoplakin Plakophilin 2 Desmoglein 2 Desmocollin 2 Cardiac ryanodine receptor (RyR2) Transforming growth factor-β3 (TGFβ3)	Familial, unknown gene Sarcomeric protein mutations Troponin I (RCM +/− HCM) Essential light chain of myosin Familial amyloidosis Transthyretin (RCM + neuropathy) Apolipoprotein (RCM + nephropathy) Desminopathy Pseuxanthoma elasticum Haemochromatosis Anderson–Fabry disease Glycogen storage disease	Left ventricular non-compaction Barth syndrome Lamin A/C ZASP α-dystrobrevin
Non-familial	Obesity Infants of diabetic mothers Athletic training Amyloid (AL/prealbumin)	Myocarditis (infective/toxic/immune) Kawasaki disease Eosinophilic (Churg Strauss syndrome) Viral persistence Drugs Pregnancy Endocrine Nutritional — thiamine, carnitine, selenium, hypophosphataemia, hypocalcaemia Alcohol Tachycardiomyopathy	Inflammation?	Amyloid (AL/prealbumin) Scleroderma Endomyocardial fibrosis Hypereosinophilic syndrome Idiopathic Chromosomal cause Drugs (serotonin, methysergide, ergotamine, mercurial agents, busulfan) Carcinoid heart disease Metastatic cancers Radiation Drugs (anthracyclines)	Tako Tsubo cardiomyopathy

ARVC, arrhythmogenic right ventricular cardiomyopathy; DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; RCM, restrictive cardiomyopathy.



We define a cardiomyopathy as a myocardial disorder in which the heart muscle is structurally and functionally abnormal, in the absence of coronary artery disease, hypertension, valvular disease and congenital heart disease sufficient to cause the observed myocardial abnormality.

Position paper ESC *European Heart Journal 2007 Oct 4*

Per la diagnosi di cardiomiopatia… Quali risultati attendersi

	ECO
Escludere CAD	SI , ma…
Vizi valvolari	SI gold standard
Cardiopatie congenite	SI + fisiopatologia
FE	SI
Volume VS e VD, spessori e massa	SI ma oper-dip, apice, massa inaff
Caratterizzazione strutturale	NI
Pericardio	SI ma…

ACCF/AHA Practice Guideline: Full Text

2009 Focused Update Incorporated Into the ACC/AHA 2005 Guidelines for the Diagnosis and Management of Heart Failure in Adults

CMR: 3 voci

A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Pr

Developed in Collaboration With the International Society fo

IMAGING:

misura dell'FE

ITTEE ME
William T.
M. Feldman
niats, MD;
Mancini, MD
CC, FAHA; M
Lynne Warner Stevenson, MD, FACC, FAHA; Clyde W.

phy. Magnetic resonance imaging or computed tomography may be useful in evaluating chamber size and ventricular mass, detecting right ventricular dysplasia, or recognizing the presence of pericardial disease, as well as in assessing cardiac function and wall motion.[36]

Magnetic resonance imaging may also be used to identify myocardial viability and scar tissue.[37] Chest radiography can with phlebotomy and chelating agents. Magnetic resonance imaging of the heart or liver may be needed to confirm the presence of iron overload. Screening for human immunode-

2009 FOCUSED UPDATE WRITING GROUP MEMBERS

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Il paziente con FE ridotta…

- Risonanza e MSCT hanno uguale dignità :

- MSCT esclude meglio della coronarografia la presenza di aterosclerosi coronarica (valore predittivo negativo = 100%).

- CMR caratterizza (funzione, volumi) la struttura.

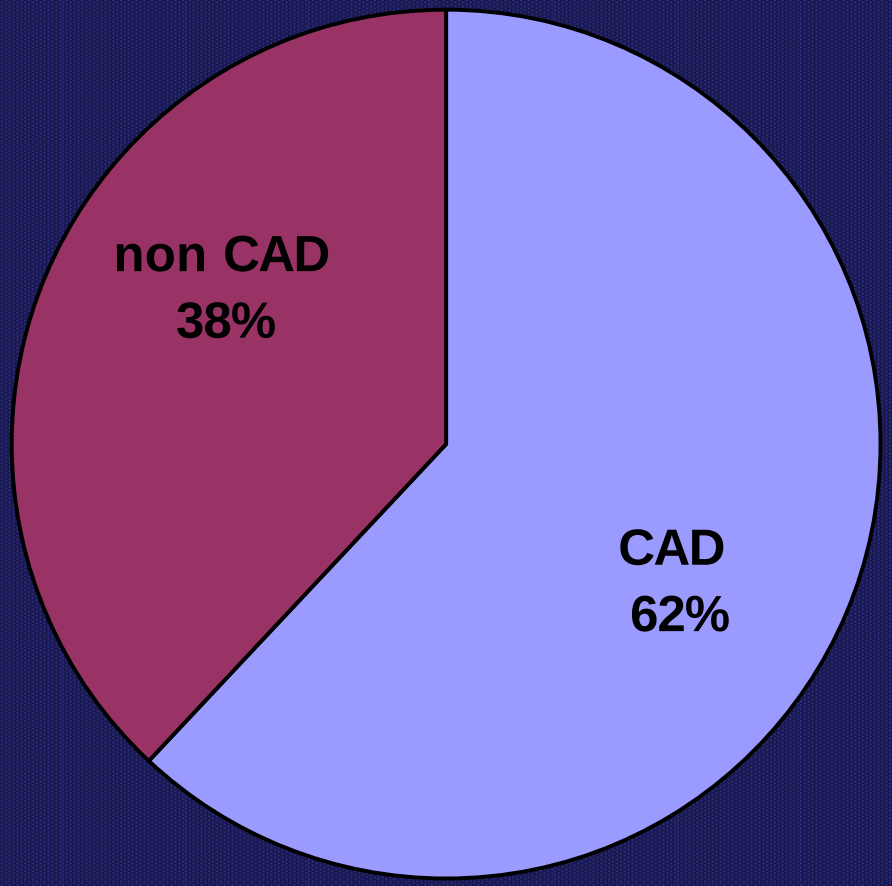
Serve sapere che le coronarie sono indenni?

Eziologia CHF nei trials sullo scompenso (FE ridotta)

24 trials consecutivi su CHF pubblicati su NEJM dal 1986

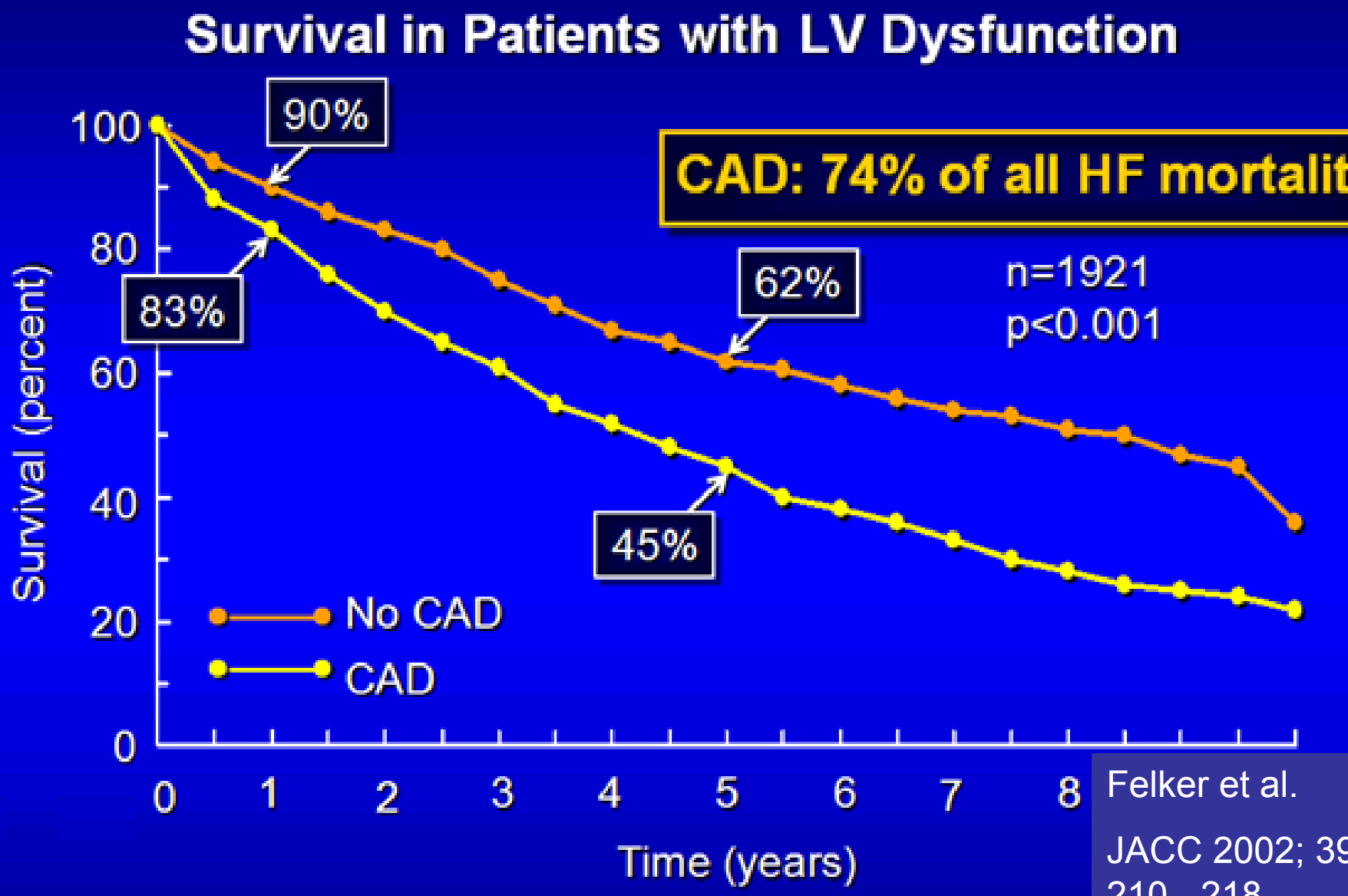
totale pz = 43568

CAD pz = 26877



Adattato da Georghiade et al
Circ 2006 114:202-213

La presenza di malattia coronarica ha conseguenze gestionali e prognostiche…



Felker et al.

JACC 2002; 39 – 210 - 218

Quando prescrivere la TC coronarica ad un pz con FE ridotta?

1. Profilo di rischio C-V intermedio (“45 – 65 anni”)
   Se ha un test da sforzo dubbio
   Se ha BBS o non è in grado di effettuare uno sforzo
2. Se è un pz valvolare (candidato a CCH)
3. Se c’è tempo di stabilizzarlo (FC<65)

Evitare >70 anni: elevata prevalenza Ca^{++}

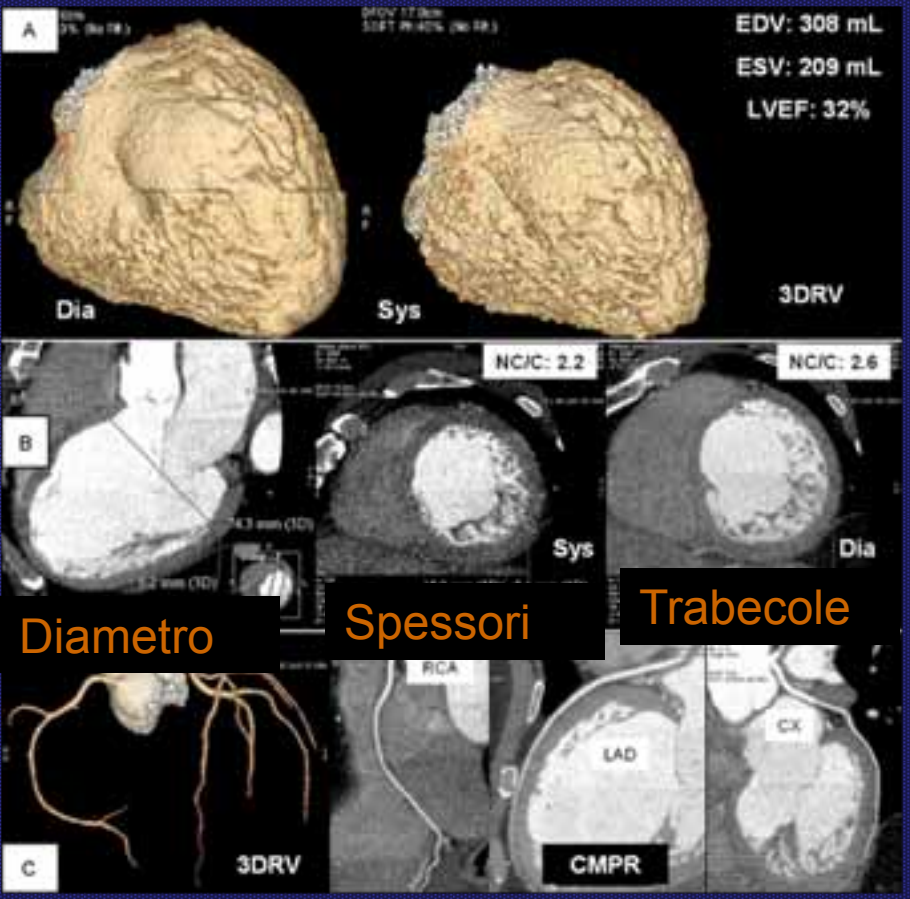
- Diabetici: elevata prevalenza Ca^{++}

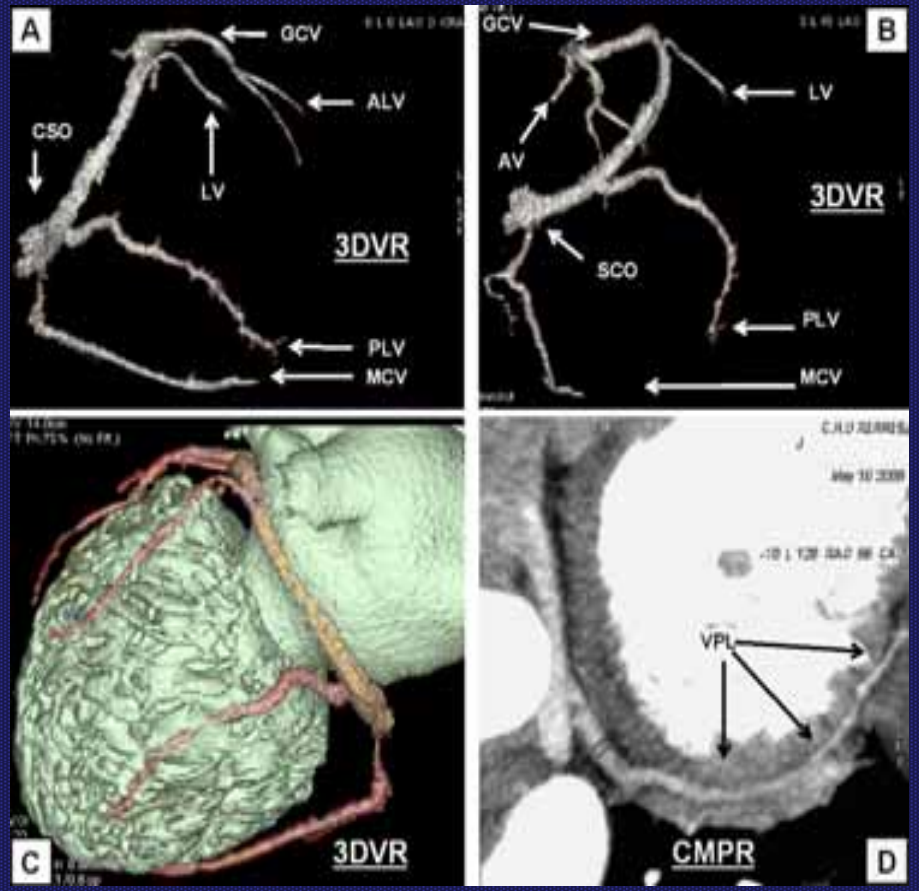
L’assenza di malattia coronarica evita una coronarografia normale.

MSCT in CMD: oltre le coronarie?

Esclude CAD

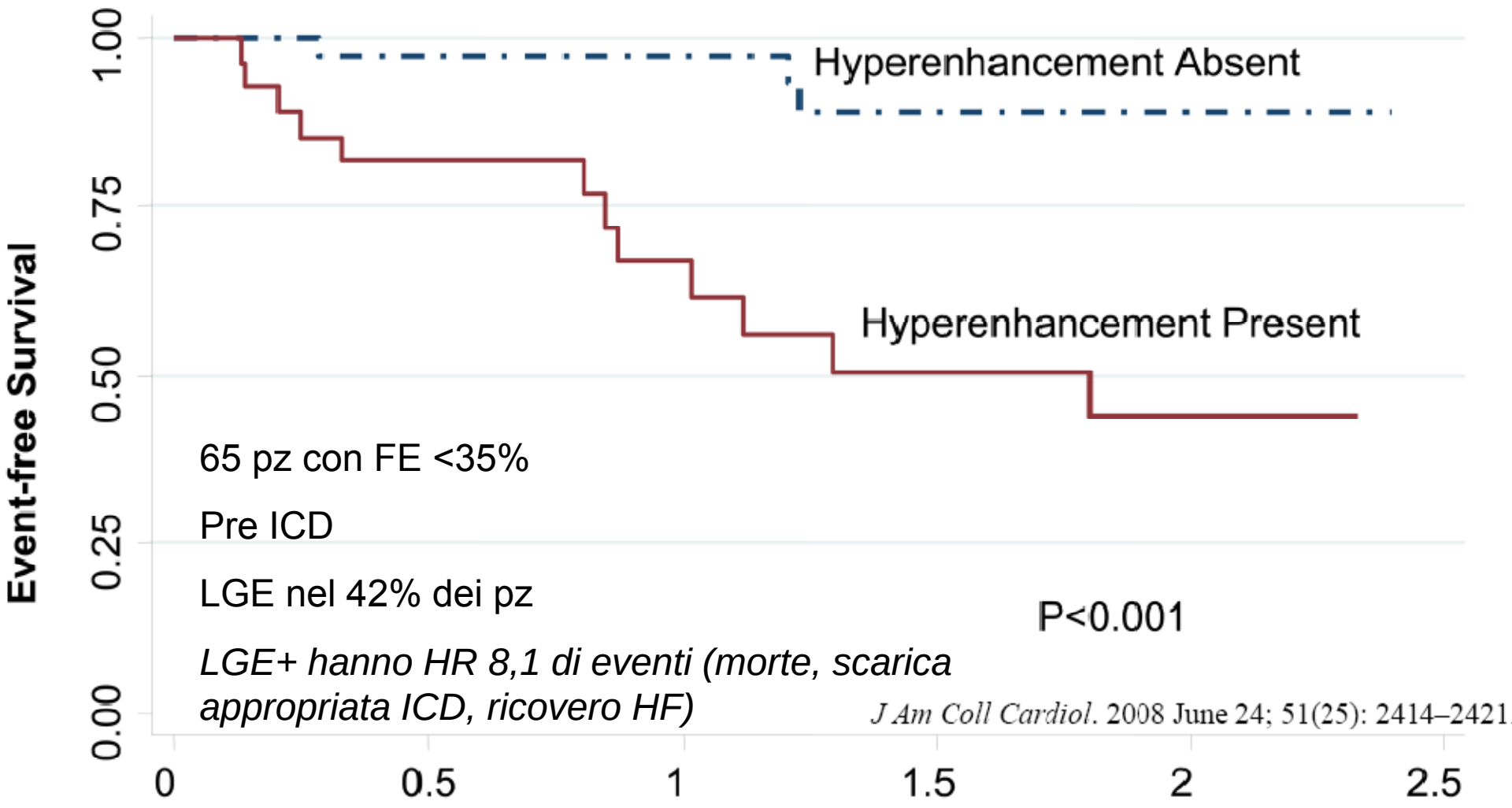
Descrive anatomia venosa





Arch Cardiovasc Dis. 2009 Oct;102(10):685-96

Cardiomiopatie dilatative e RM: fibrosi correla con prognosi



A quali pz con CMD fare la risonanza?

- Dati discordanti di FE, necessità di decisioni cliniche
- Prima dell'impianto di un device: stratificazione prognostica.
- MA NON indicazione all'impianto

Quando prescrivere la RM cardiaca ad un pz con FE ridotta?

1. FE ridotta + aritmie:
   - Cardiopatia aritmogena del VS (ICD)
   - Tachicardiomiopatie

2. Definizione diagnostica: non tutte le FE ridotte sono CMD:
   - Miocarditi
   - Tako Tsubo (risol. spont)
   - Emocromatosi – talassemici
   - Shunts (Botallo, etc..)

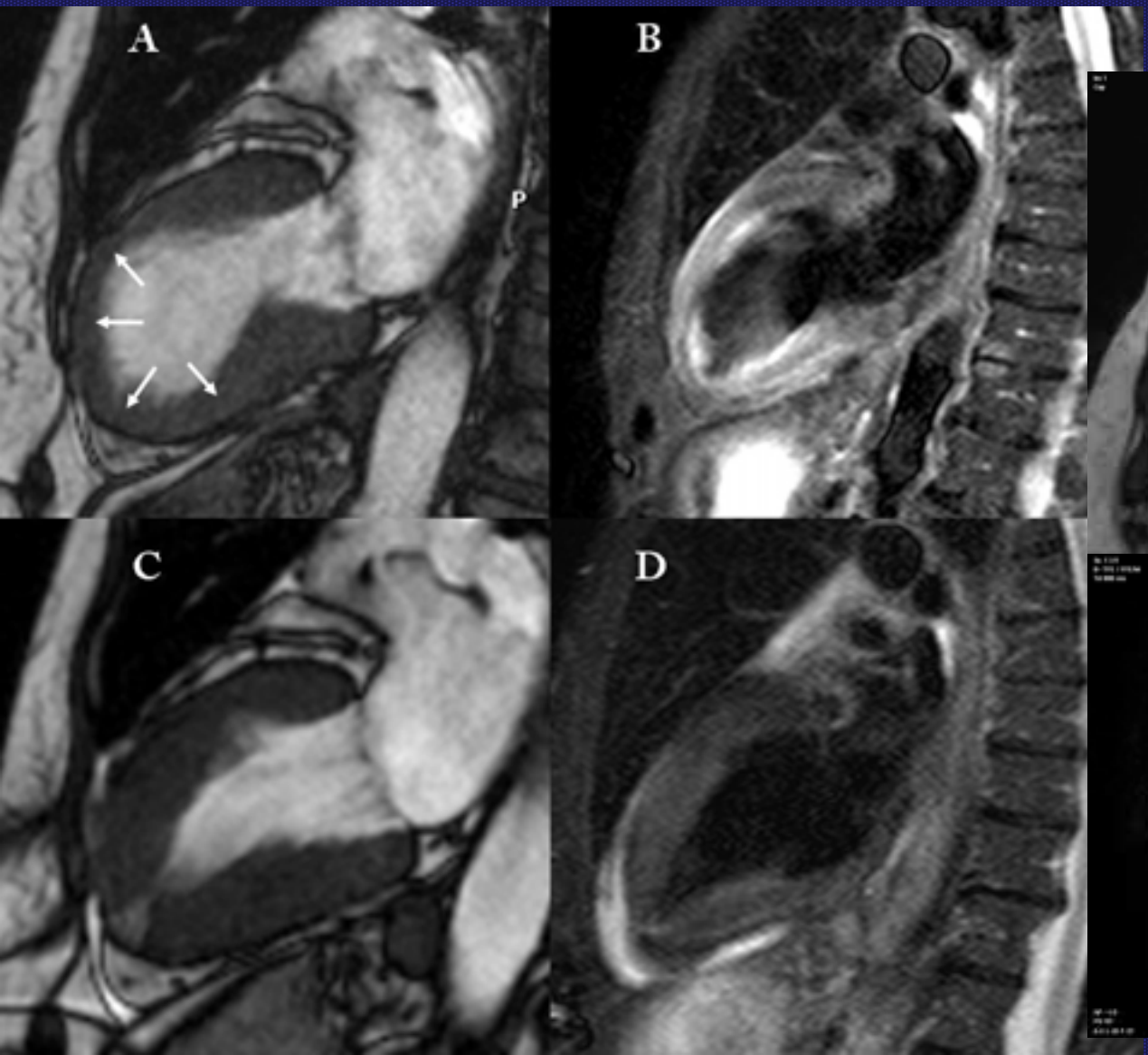
3. Follow-up – risposta a terapia farmacologica

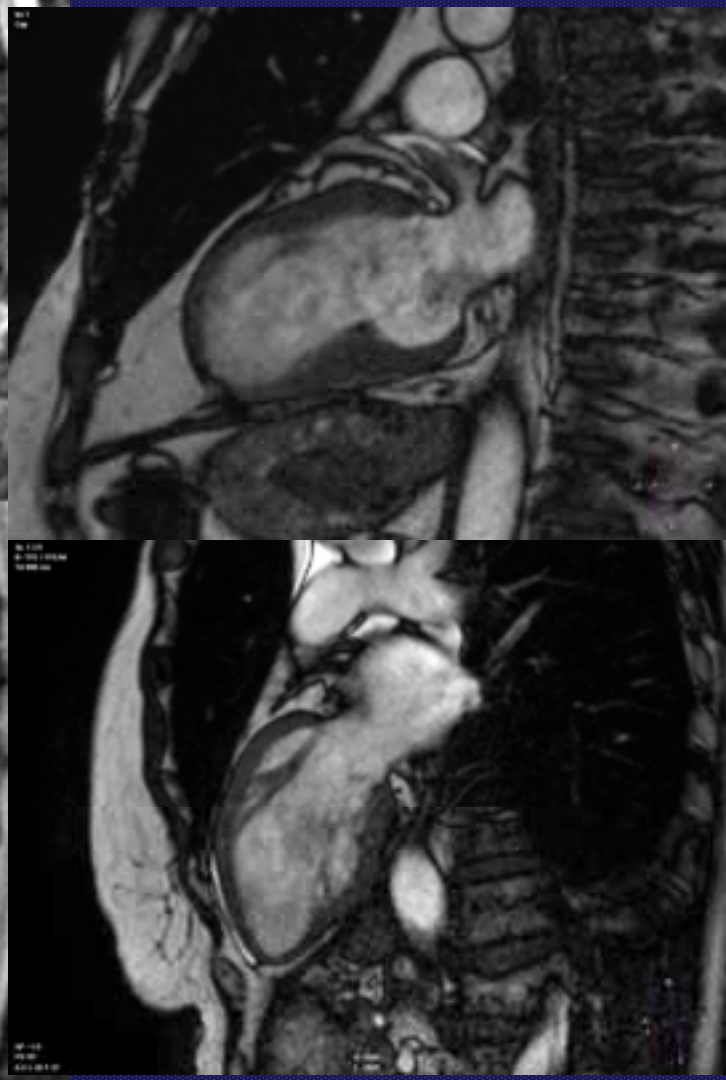
4. Stratificazione prognostica

Tako Tsubo

Stress + alterazioni ECG simil-infartuali + Troponina bassa.

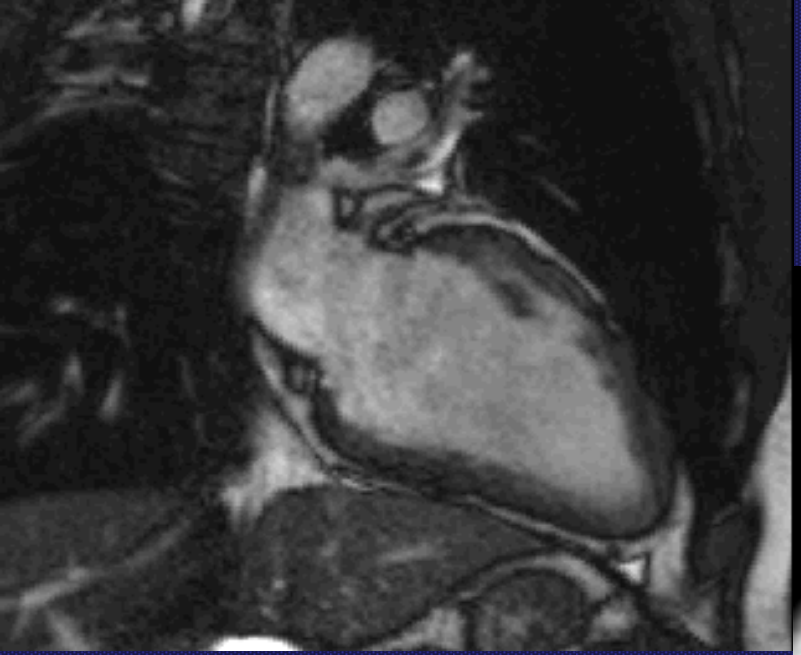
MR:acinesia con distribuzione non coronarica; edema +; LGE-

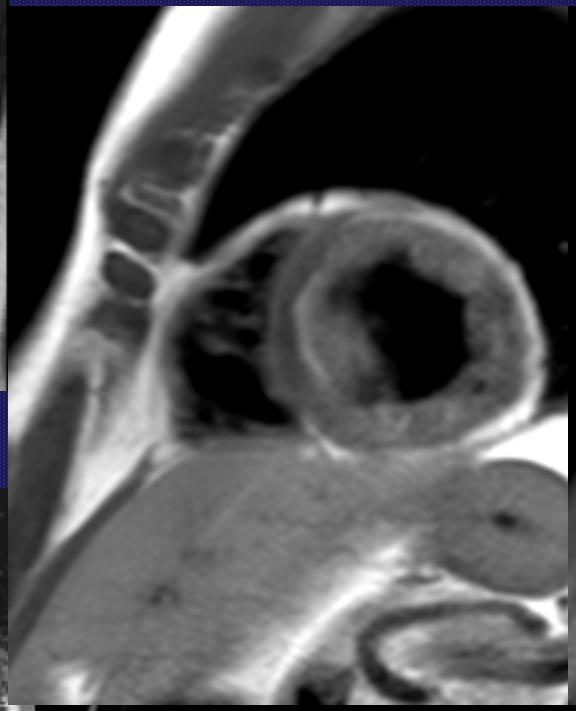


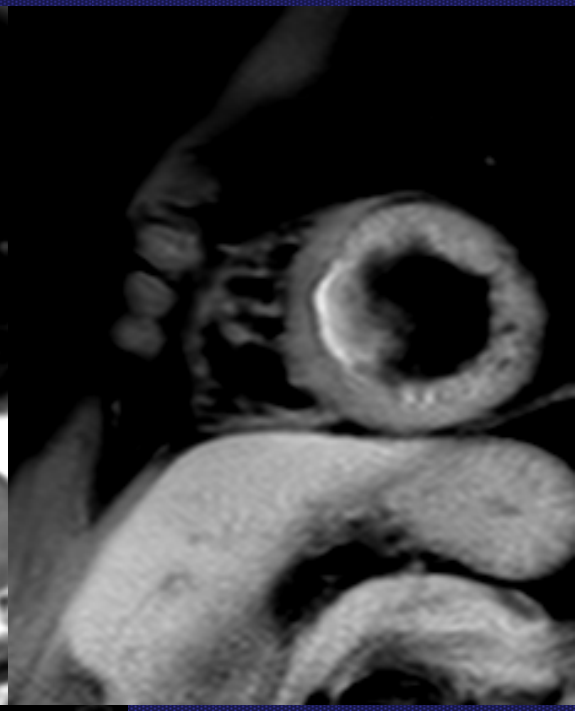


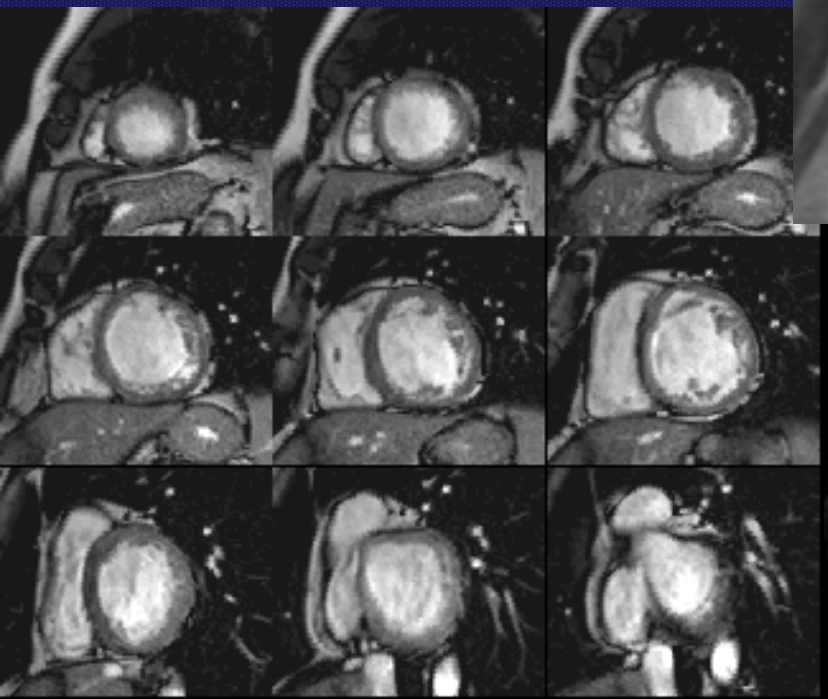
LV cardiomyopathy

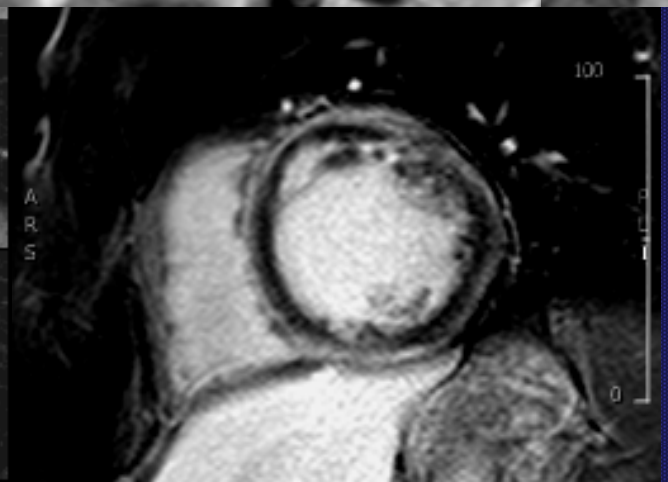
– caso 1: uomo 38 anni con pre-lipotimie









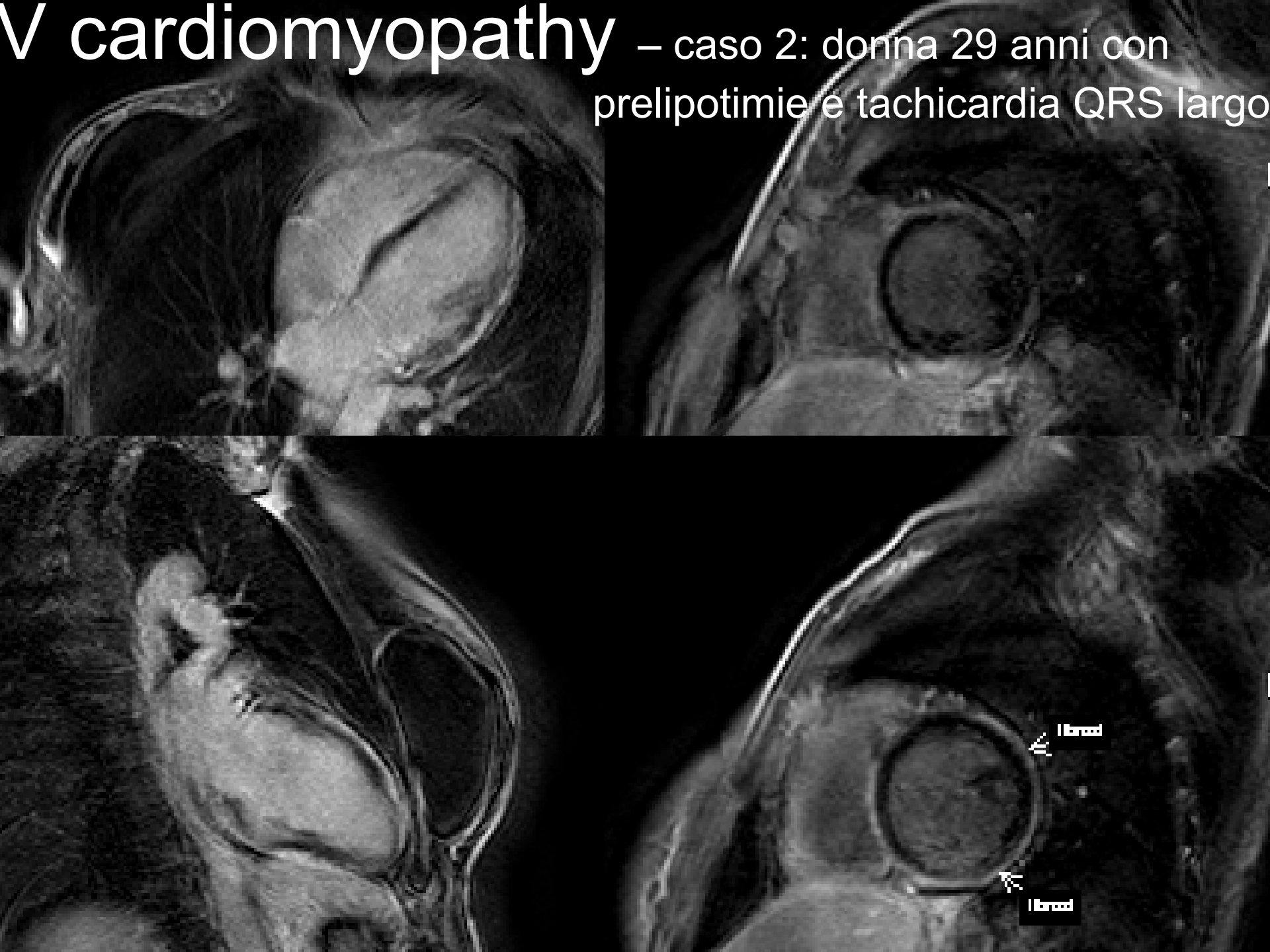


Coro normali

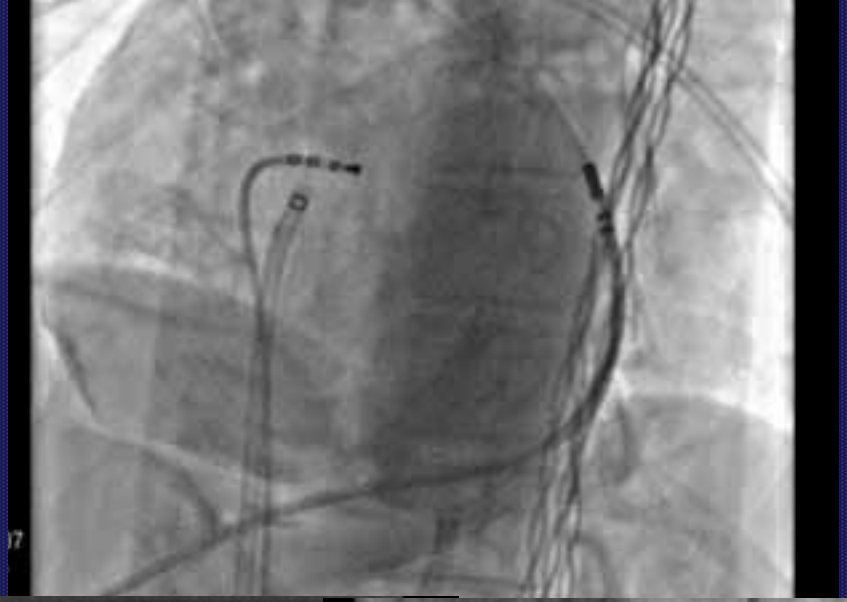
SEF: positivo

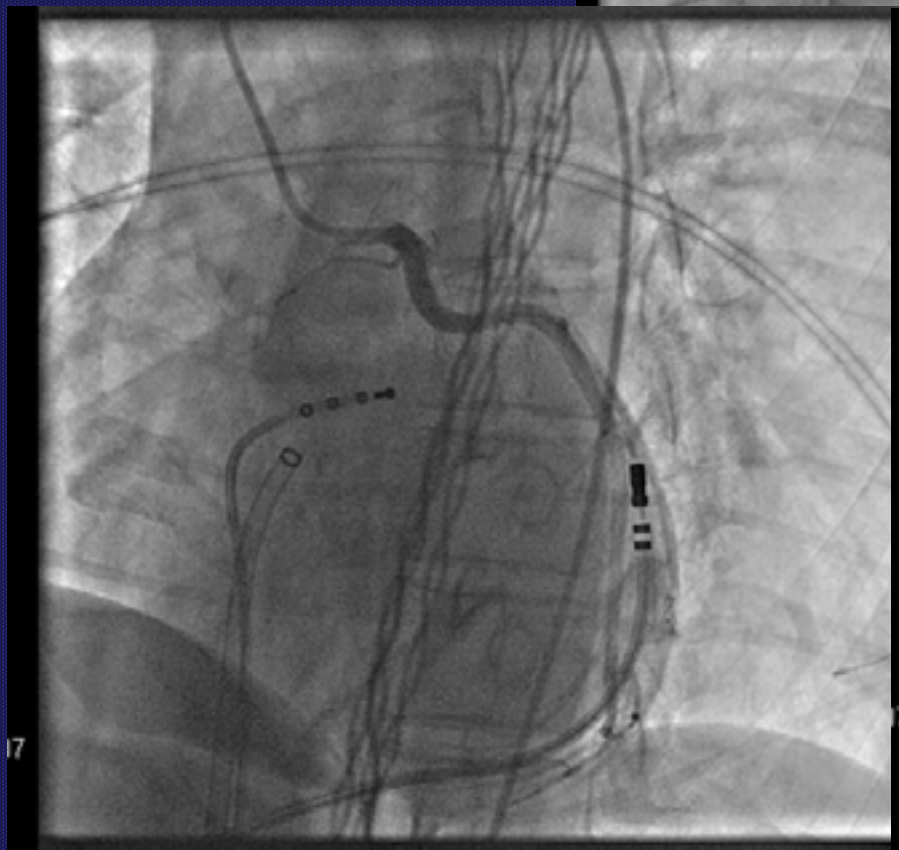
Impiantato ICD

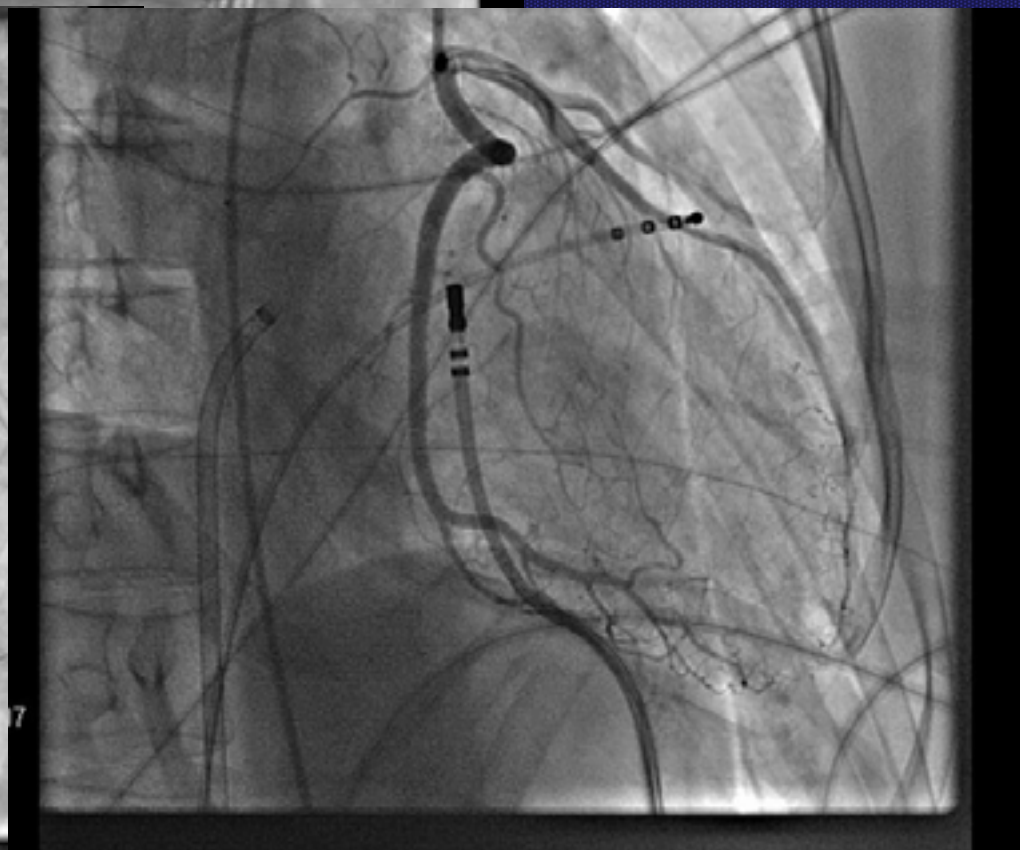
V cardiomyopathy – caso 2: donna 29 anni con prelipotimie e tachicardia QRS largo

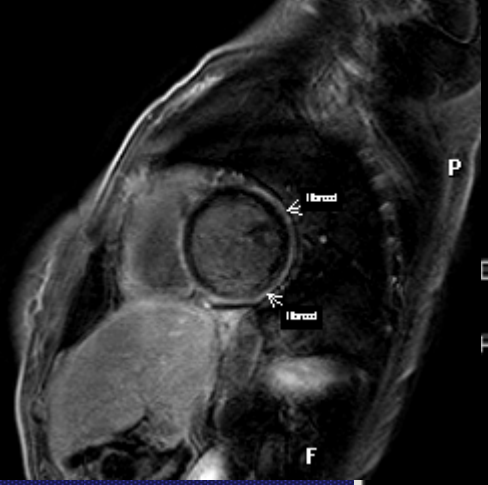


Effettuata ablazione epicardica della TV





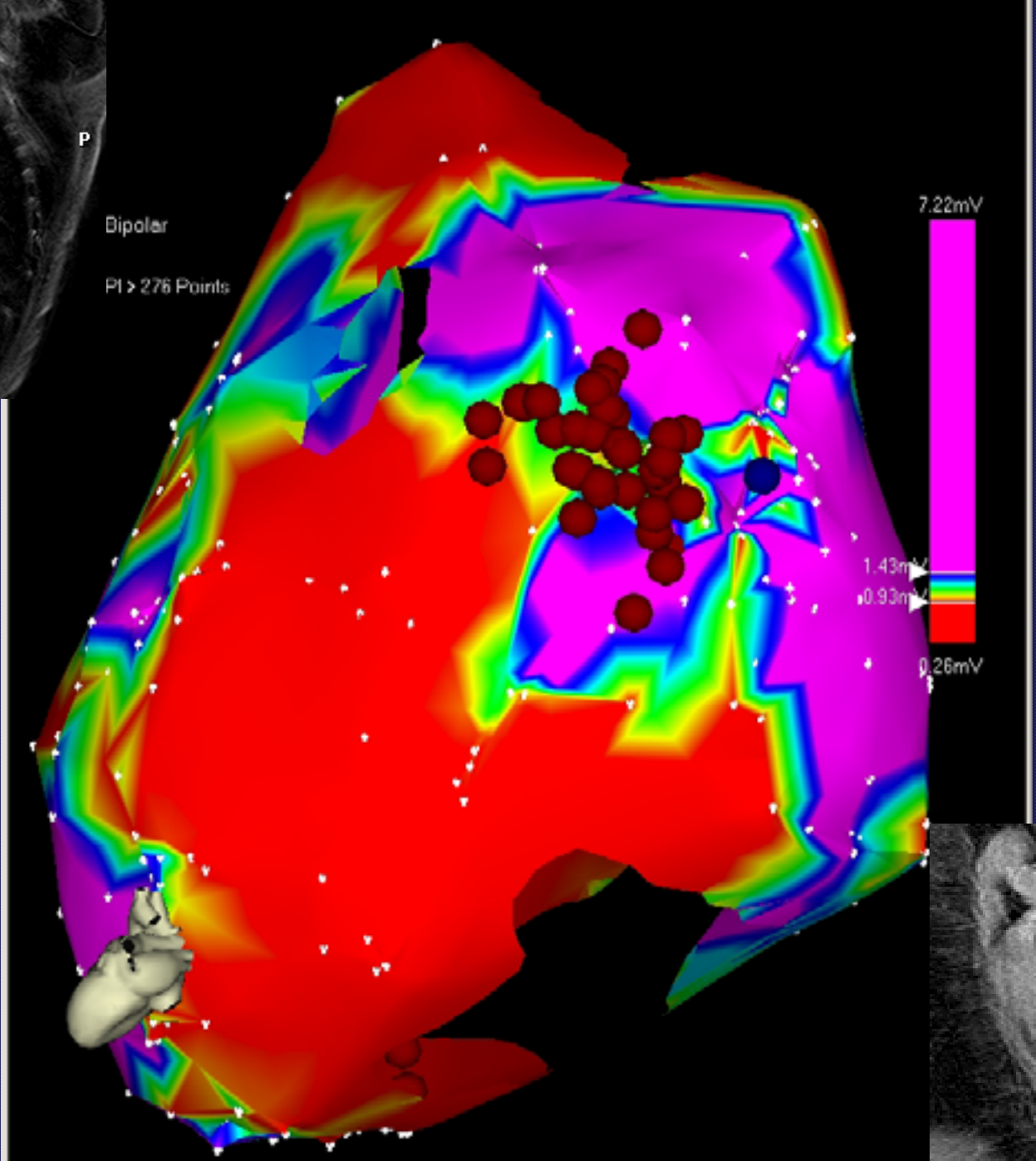


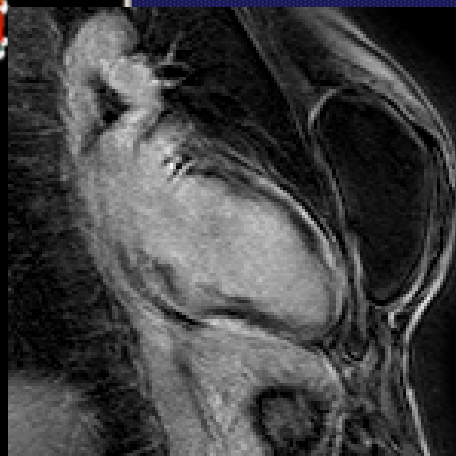


Recidiva clinica dopo pochi mesi

Impianto di ICD

Intervento appropriato dopo 1 anno

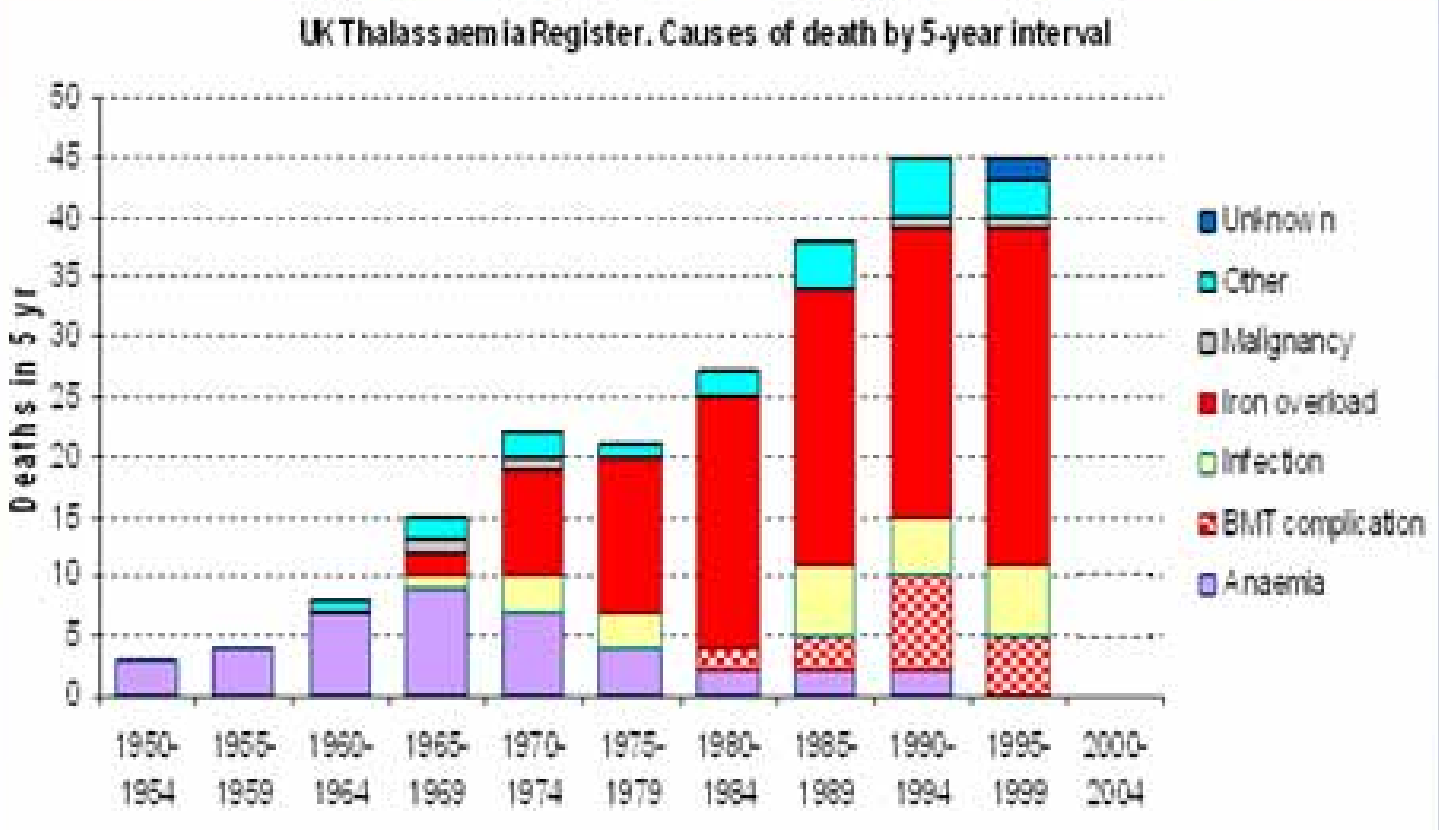




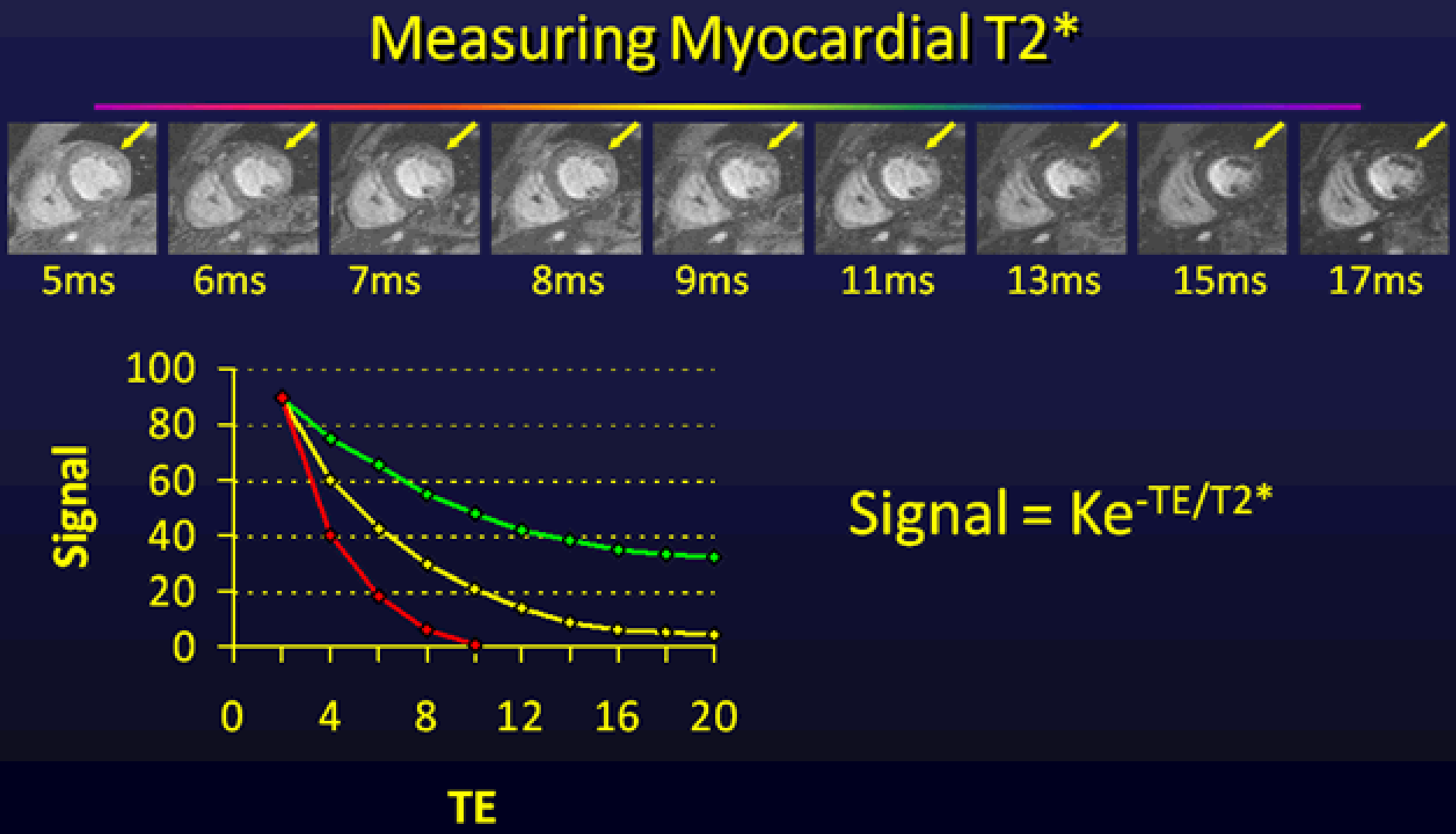
Talassemici

Thalassemia Mortality

Death by heart failure in 71%

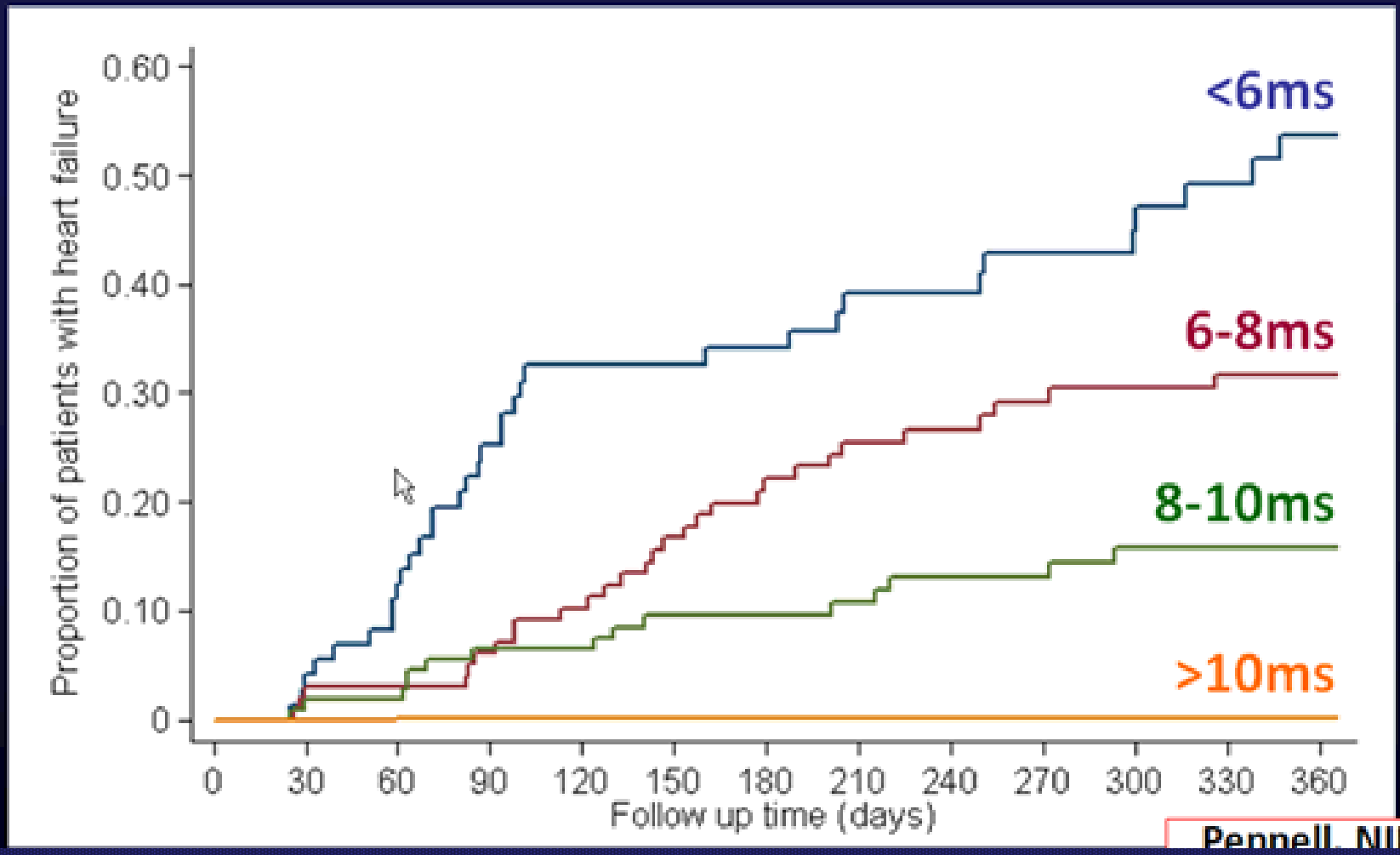


La misura del T2 * (star)



T2* normale > 40 msec,
T2* patologico < 20 msec → chelanti

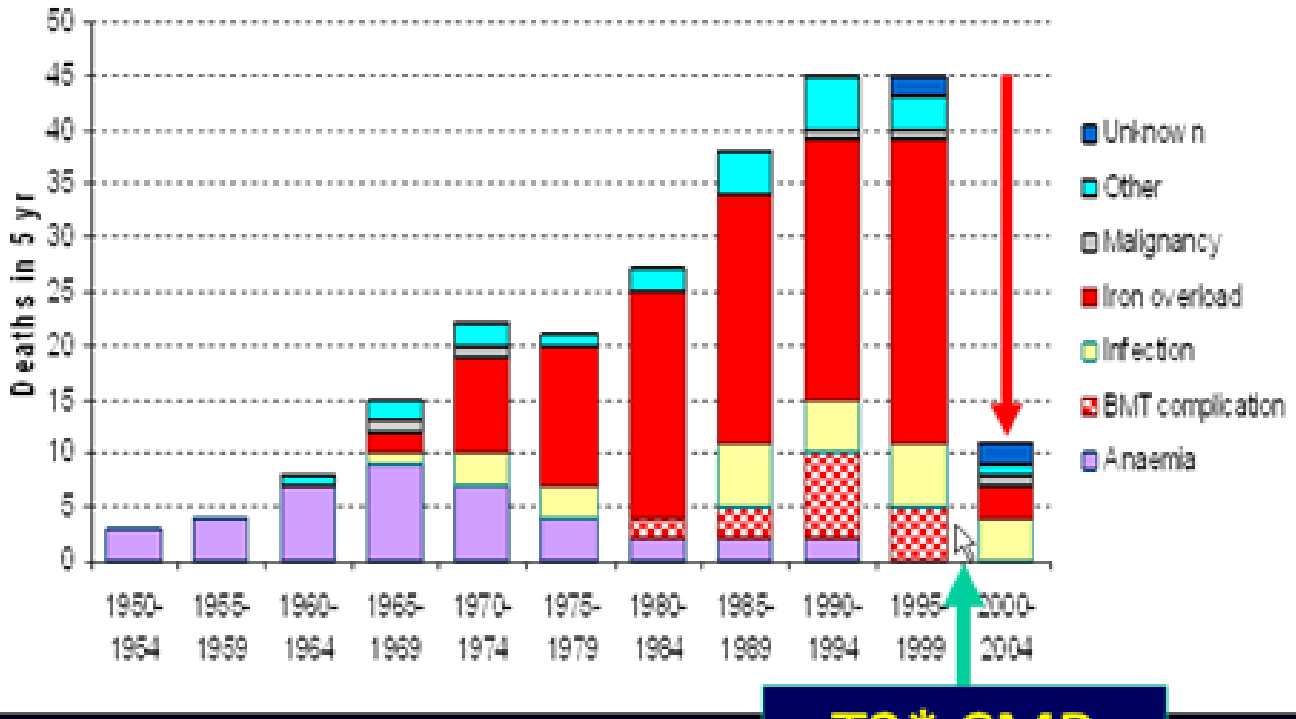
Kaplan Meier Curves: T2* and Heart Failure



Cosa è successo gestendo la terapia chelante in base al T2*

Change in Cardiac Death in UK Since 2000

Reduction in cardiac death by 71%

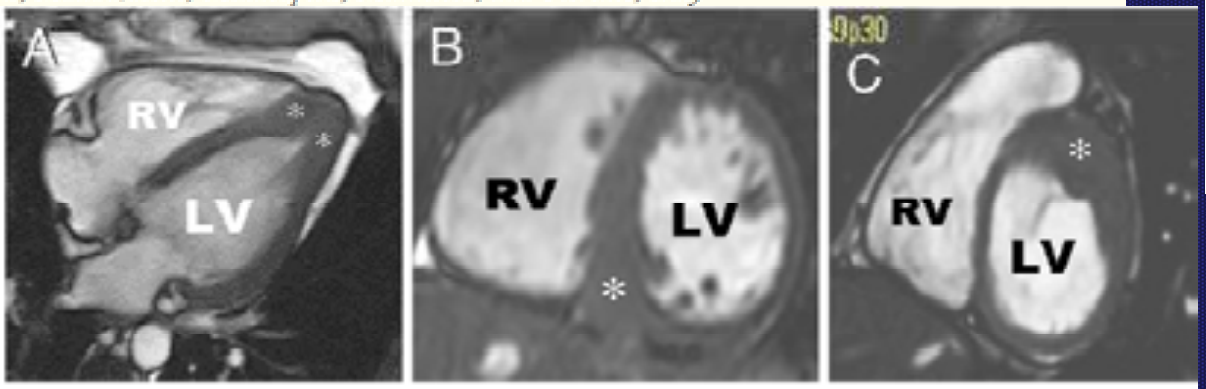


La cardiomiopatia ipertrofica.

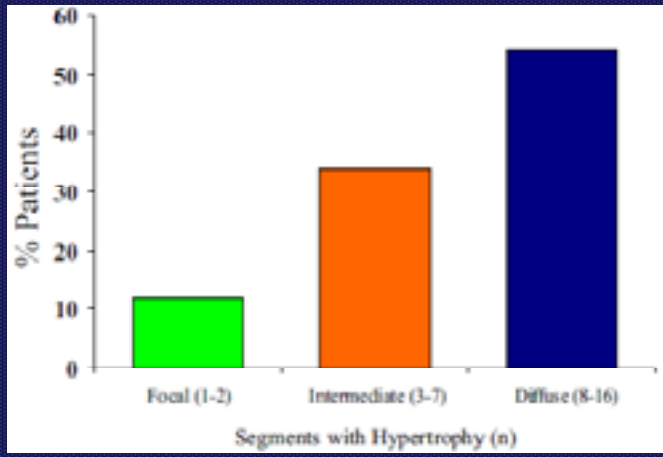
Hypertrophic Cardiomyopathy Phenotype Revisited After 50 Years With Cardiovascular Magnetic Resonance

Martin S. Maron, MD,* Barry J. Maron, MD,† Caitlin Harrigan, BA,* Jacki Buros, BA,‡
C. Michael Gibson, MD, MS,‡§ Iacopo Olivotto, MD,‖ Leah Biller, BA,† John R. Lesser, MD,†
James E. Udelson, MD,* Warren J. Manning, MD,‡§ Evan Appelbaum, MD‡§
Boston, Massachusetts; Minneapolis, Minnesota; and Florence, Italy

J Am Coll Cardiol 2009;54:220–8

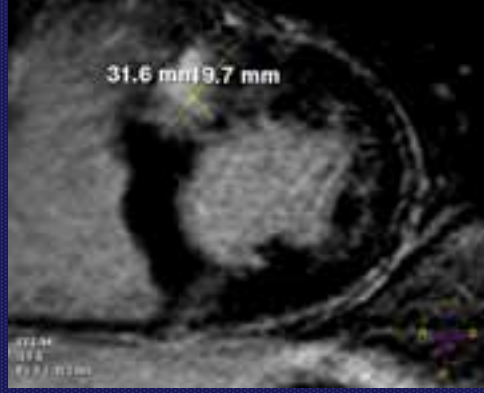


LV Hypertrophy Recognized by CMR But Not Reliably With 2-Dimensional Echocardiography

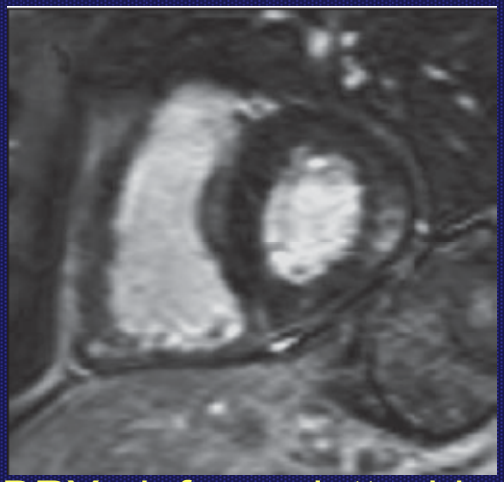


La RM permette di diagnosticare HCM nei casi difficili e in sedi atipiche.

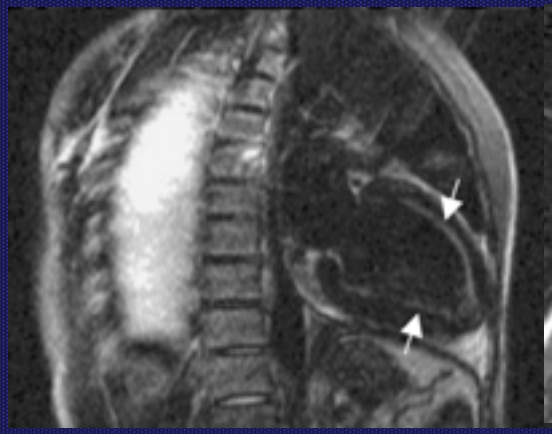
LA RM negli ipertrofici

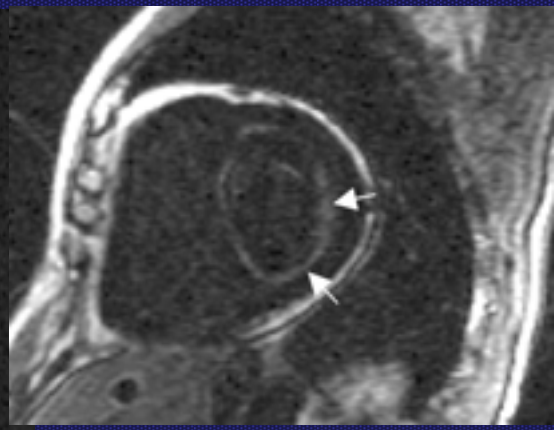


- Sempre se dubbio di ipertrofia/ discrepanza ECG – ecocardio.
- Casi selezionati di parenti per diagnosi precoce
- Se ipertrofia certa
  - Possibile ruolo prognostico di LGE
  - Diagnosi differenziale con altre cause di ipertrofia, ad es. Fabry, sarcoidosi e amiloidosi



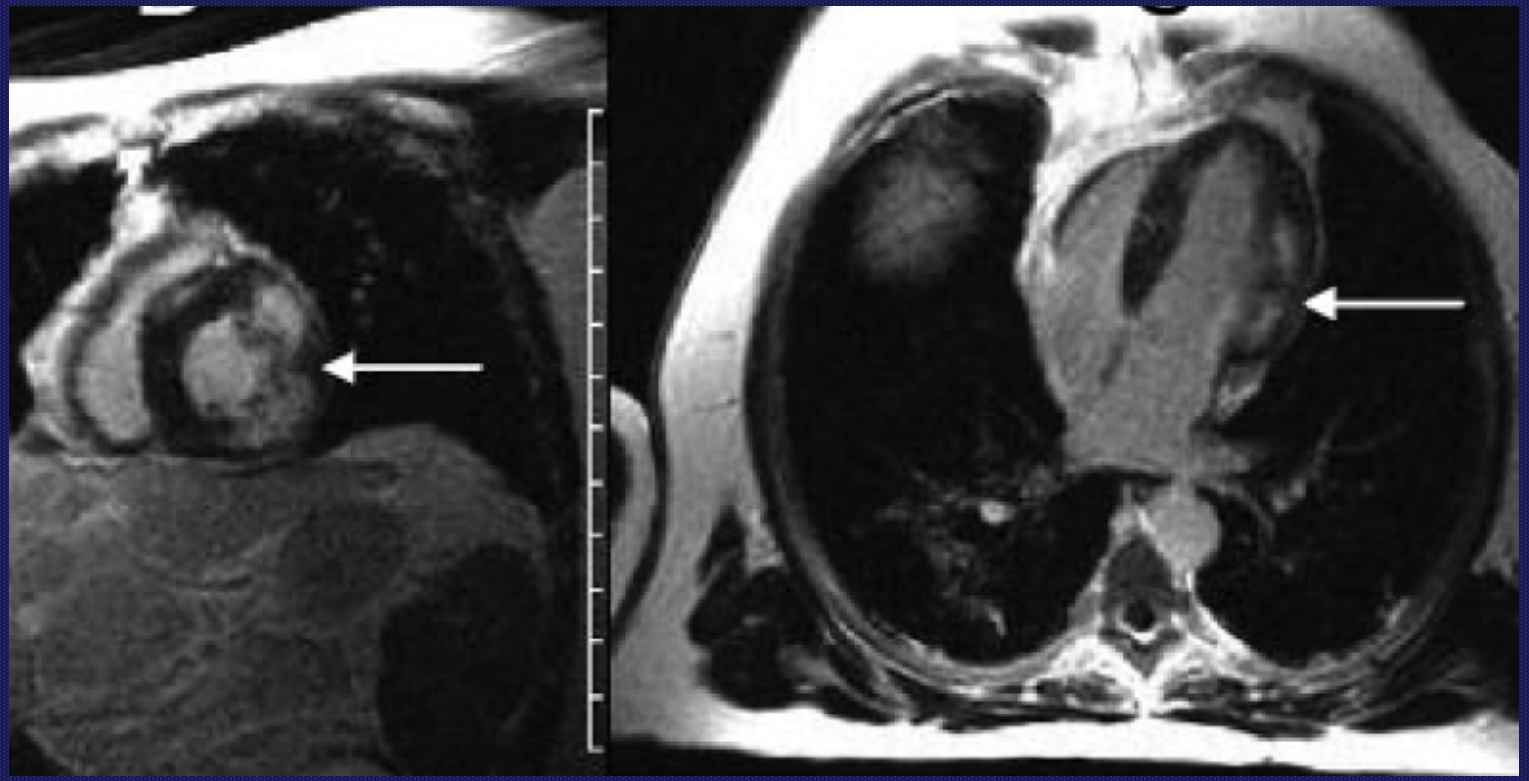
FABRY: def. a-galattosidasiA, X-linked





AMILOIDOSI AL

La RM nella sarcoidosi



Malattia granulomatosa multiorgano.

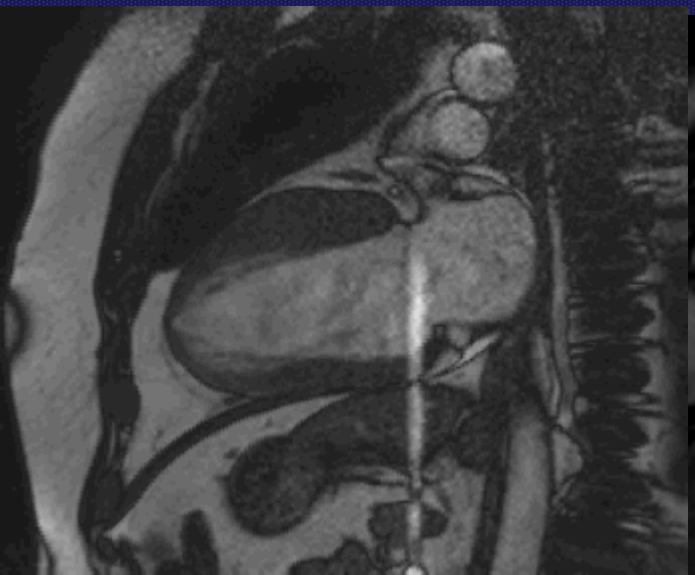
40% dei pz affetti ha coinvolgimento cardiaco all'autopsia.

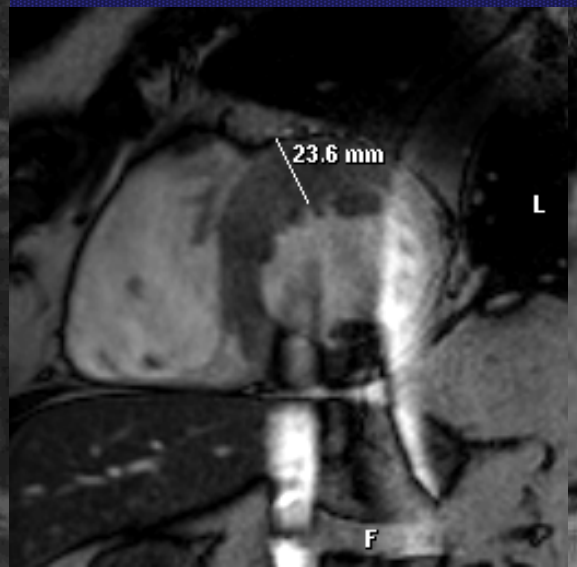
10% dei pz ha diagnosi di sarcoidosi cardiaca in vita.

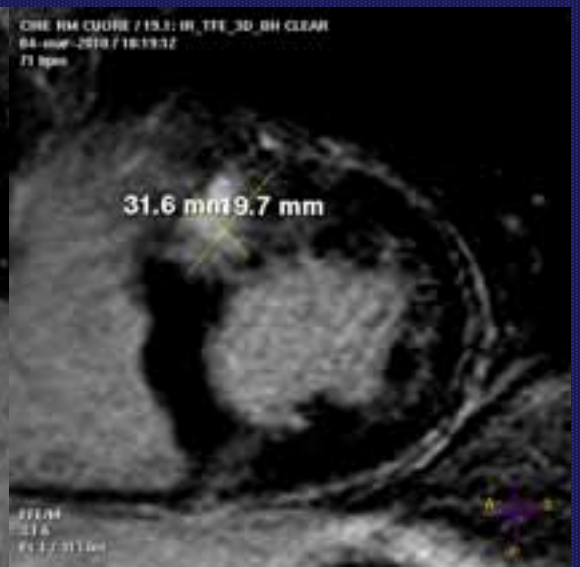
Un caso personale…

Sportivo 43 anni >100Kg; epigastralgie.
riscontro 5000 BEV e triplette
ecocardio: setto 14 mm. FE normale (cattiva finestra)-

RM parete anteriore 23 mm, estesa fibrosi



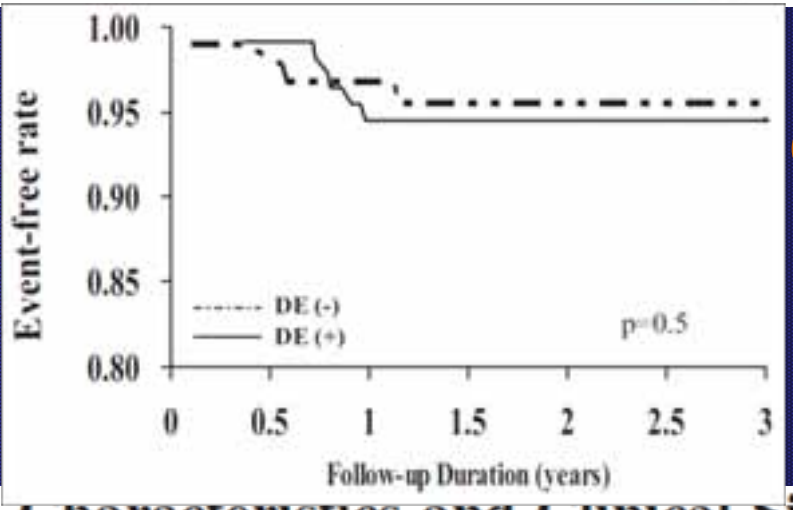




Clinical Profile and Significance of Delayed Enhancement in Hypertrophic Cardiomyopathy

Martin S. Maron, MD; Evan Appelbaum, MD; Caitlin J. Harrigan, BA; Iacki Buros, BA; C. Michael Gibson, MD, MS; Connie Hanna, RN; John R. Lesser, MD; James E. Udelson, MD; Warren J. Manning, MD; Barry J. Maron, MD

Circ Heart Fail. 2008;1:184-191



202 HCM pz

Prevalenza LGE = 55%

Media 10% LV mass

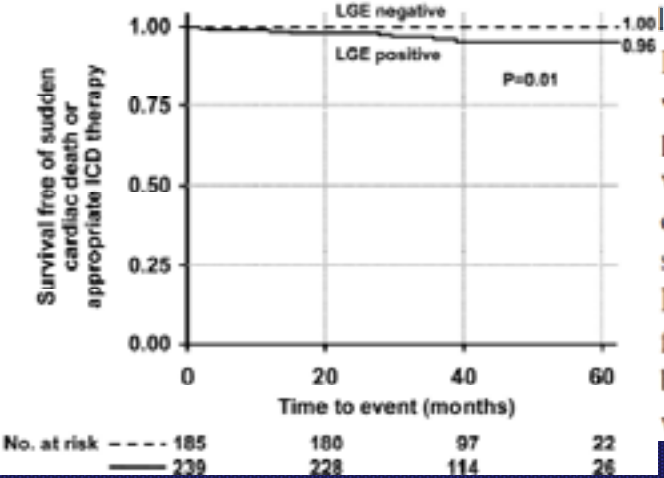
> 25% nel 10% pz

così è già evidente all'eco?

2008: "la presenza di fibrosi non predice progn

Characteristics and Clinical Significance of Late Gadolinium Enhancement by Contrast-Enhanced Magnetic Resonance Imaging in Patients With Hypertrophic Cardiomyopathy

Ronen Rubinshtein, MD; James F. Glockner, MD, PhD; Steve R. Ommen, MD; Philip A. Araoz, MD; Sorajja, MD; J. Martijn Bos, MD; A. Jamil Tajik, MD;



In conclusion, the presence of LGE on CE-MRI in patients with HCM was common but was not associated with higher rates of severe angina or dyspnea. However, LGE was strongly associated with surrogates of arrhythmia, and our data are the first to demonstrate that LGE remained a significant associate of subsequent SCD or appropriate ICD therapies after controlling for other factors. If these findings are confirmed in independent cohorts, LGE may be considered an additional risk factor for SCD in patients with HCM.

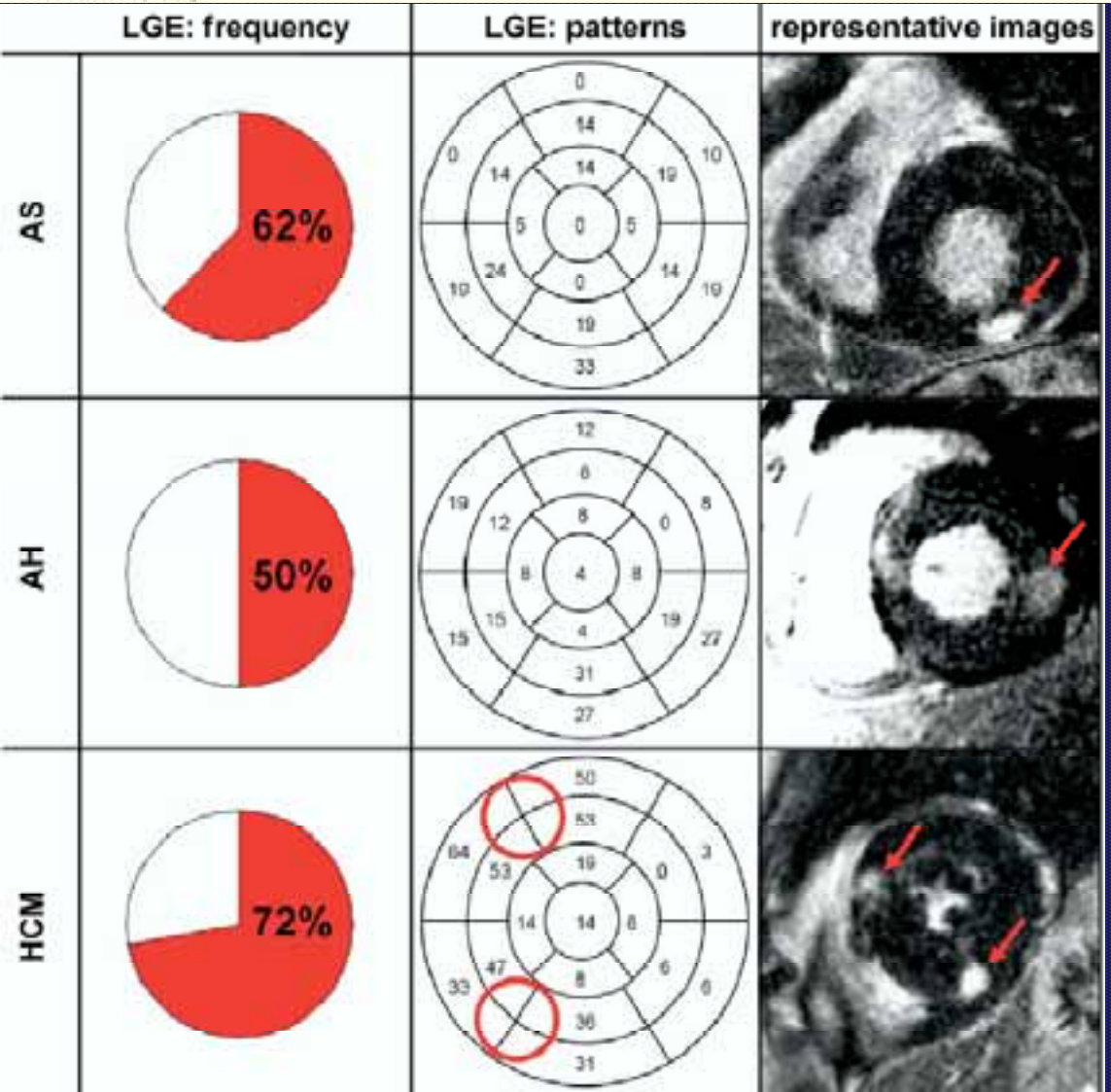
2010: LGE predice gli eventi aritmici.

Noninvasive Detection of Fibrosis Applying Contrast-Enhanced Cardiac Magnetic Resonance in Different Forms of Left Ventricular Hypertrophy

Relation to Remodeling

Andre Rudolph, MD, Hassan Abdel-Aty, MD, Steffen Bohl, MD, Philipp Boyé, MD, Anja Zagrosek, MD, Rainer Dietz, MD, Jeanette Schulz-Menger, MD
Berlin, Germany

J Am Coll Cardiol 2009;53:284–91



440 pz con ipertrofia VS all'eco

83 pz con ***aumento LVMI***
>1.06g/cm uomini
>0.8 g/cm donne

Quando la massa VS è patologica, la prevalenza di fibrosi ventricolare è elevata indipendentemente dallo stimolo che ha prodotto l'ipertrofia

La MSCT negli ipertrofici

- Se proprio non si può fare RM
  - per claustrofobia (…)
  - per ICD impiantato.

Displasia aritmogena del VD.

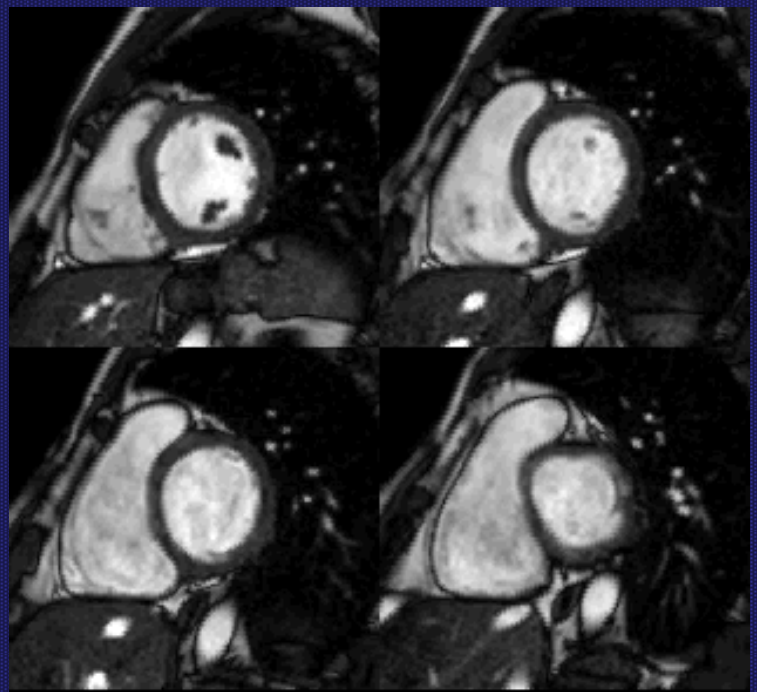
Non ha bisogno di sponsorizzazioni: indicazione certa a RM cuore

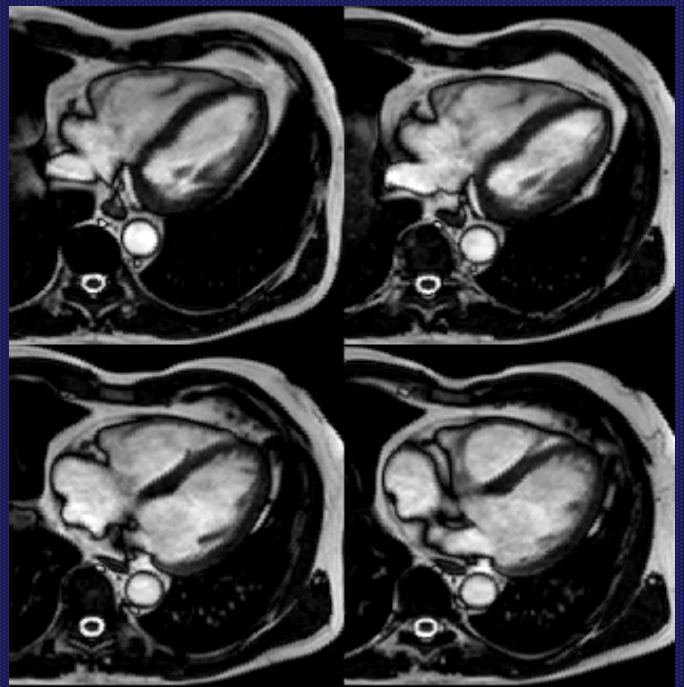
Original Task Force Criteria	Revised Task Force Criteria
I. Global or regional dysfunction and structural alterations*	
Major	
• Severe dilatation and reduction of RV ejection fraction with no (or only mild) LV impairment • Localized RV aneurysms (akinetic or dyskinetic areas with diastolic bulging) • Severe segmental dilatation of the RV	**By 2D echo:** • Regional RV akinesia, dyskinesia, or aneurysm • *and* 1 of the following (end diastole): — PLAX RVOT ≥32 mm (corrected for body size [PLAX/BSA] ≥19 mm/m²) — PSAX RVOT ≥36 mm (corrected for body size [PSAX/BSA] ≥21 mm/m²) — *or* fractional area change ≤33% **By MRI:** • Regional RV akinesia or dyskinesia or dyssynchronous RV contraction • *and* 1 of the following: — Ratio of RV end-diastolic volume to BSA ≥110 mL/m² (male) or ≥100 mL/m² (female) — *or* RV ejection fraction ≤40% **By RV angiography:** • Regional RV akinesia, dyskinesia, or aneurysm
Minor	
• Mild global RV dilatation and/or ejection fraction reduction with normal LV • Mild segmental dilatation of the RV • Regional RV hypokinesia	**By 2D echo:** • Regional RV akinesia or dyskinesia • *and* 1 of the following (end diastole): — PLAX RVOT ≥29 to <32 mm (corrected for body size [PLAX/BSA] ≥16 to <19 mm/m²) — PSAX RVOT ≥32 to <36 mm (corrected for body size [PSAX/BSA] ≥18 to <21 mm/m²) — *or* fractional area change >33% to ≤40% **By MRI:** • Regional RV akinesia or dyskinesia or dyssynchronous RV contraction • *and* 1 of the following: — Ratio of RV end-diastolic volume to BSA ≥100 to <110 mL/m² (male) or ≥90 to <100 mL/m² (female) — *or* RV ejection fraction >40% to ≤45%

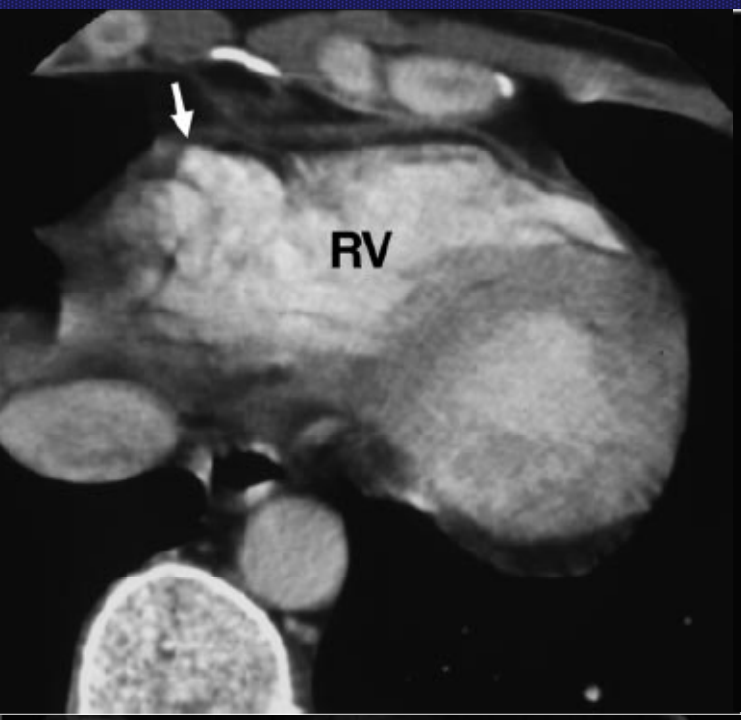
Circulation published online Feb 19, 2010

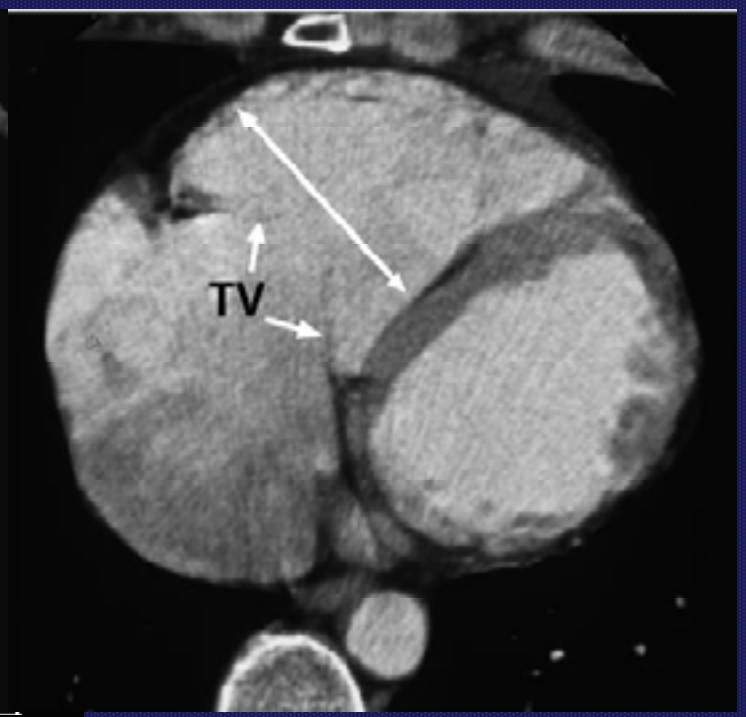
Misurare dilatazione e disfunzione del VD

- VOLUMI ed FE: il limite in questa valutazione è l'ampio movimento base-apice del piano tricuspidalico: utile modificare l'approccio standard usando proiezioni SSFP assiali.

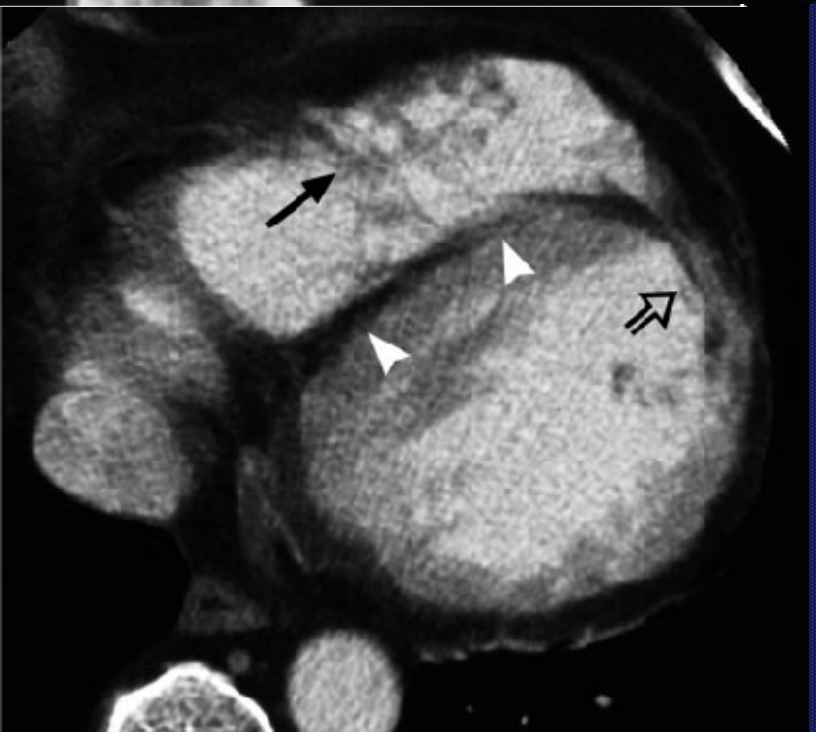








MSCT?

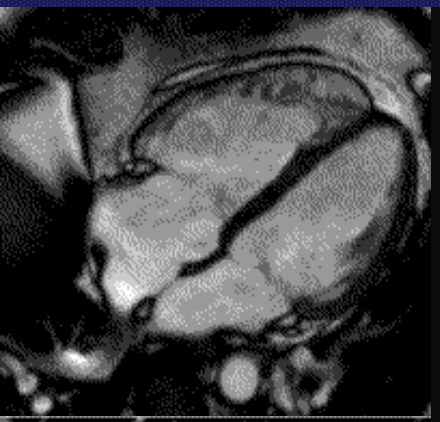


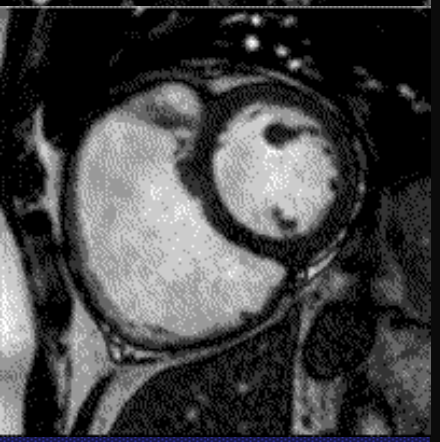
Morfologia VD

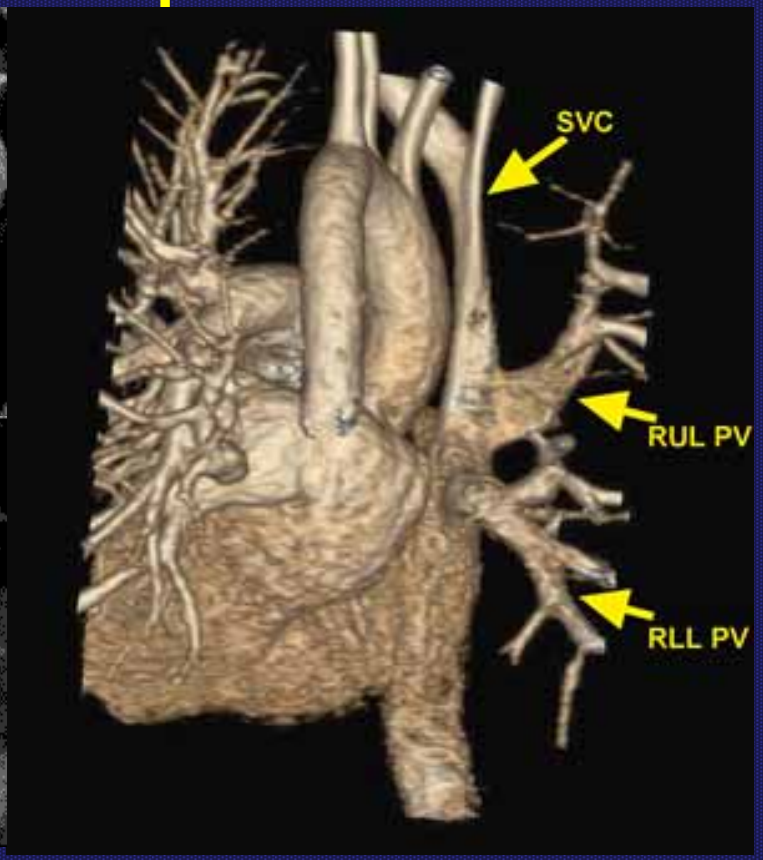
Aneurismi VD

Infiltrazione adiposa VD e VS

La dilatazione del VD: sempre misurare QP/QS







Sospetta DAVD

Qp/Qs 2.4:1

PAPVR

Non tutte le dilatazione del VD sono DAVD, anzi…

Escludere DIA, specie tipo seno venoso – ritorno venoso polmonare anomalo.

T1 assiali a tutti x anatomia. Ricerca vene polmonari e QP/QS a tutti

CONCLUSIONE

CMR e MSCT creano opportunità uniche per migliorare la diagnosi e la gestione terapeutica dei pz con cardiomiopatia. RM sta entrando nelle linee guida,

MA…..

Rappresentano oggi una sfida nella selezione dei pazienti, nell'educazione dei medici, nell'integrazione con le metodiche già disponibili, e soprattutto nell'utilizzo (costo) efficace delle risorse disponibili

GRAZIE
per l’attenzione