

Right heart test for the right HF patient // Mexico, October 2016



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DISCLOSURE

No Relevant Financial Relationship(s)

No Off Label Usage

Case

- 50 year old white female, dilated CMP
- Hx Mv repair, severe functional MR
- Presented with syncopal event
- Noted to have 3rd Degree Heart Block
 - PPM implanted
 - Father died at age 59 of “heart disease”
 - Needed AICD for VT later

Case

- Blood work
 - Hgb 14.8 WBC 5.7 Plt 190
 - NA 142 K 4.2 Cr 1.1
- Physical exam
 - Normotensive, Normal Weight
 - Paradoxically Split S2, otherwise NI

Case

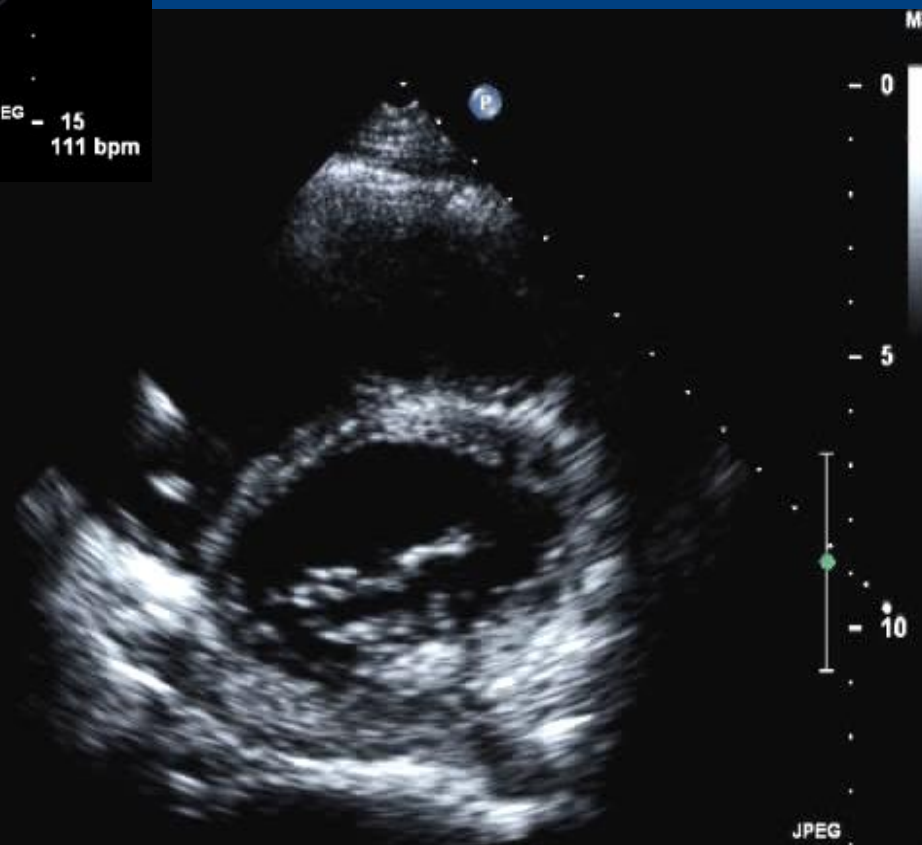


PR 50Hz
15cm

2D
58%
C 50
P Low
HGen



P Low
HGen



Case

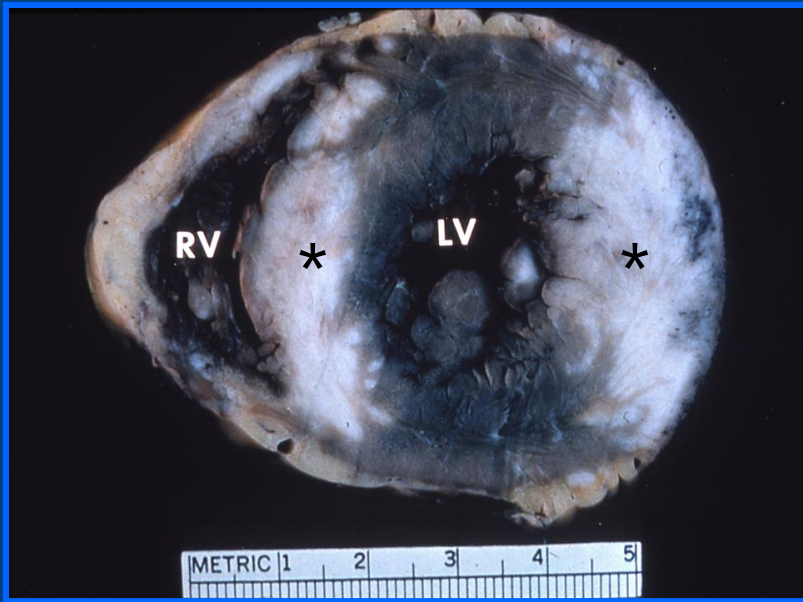
- A. Coronary artery Disease
- B. Cardiac Sarcoid
- C. Hypertrophic Cardiomyopathy
- D. Dilated Cardiomyopathy

Case

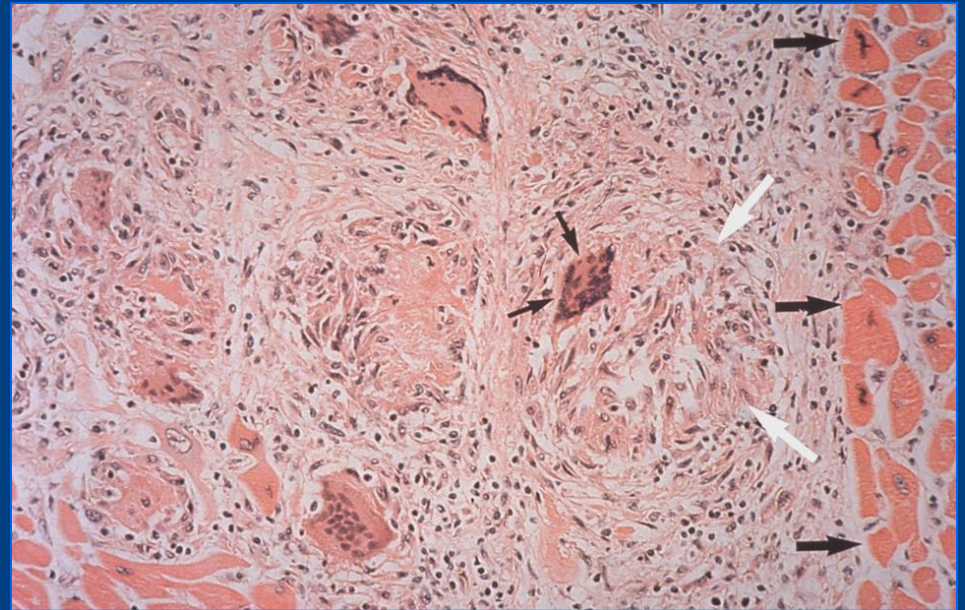
- A. Coronary artery Disease
- B. Cardiac Sarcoid (Isolated)
- C. Hypertrophic Cardiomyopathy
- D. Dilated Cardiomyopathy

Left atrial wall from Mv repair surgery: pathologic confirmation

Cardiac Sarcoidosis



*Extensive Transmurular
fibrosis non-coronary distribution



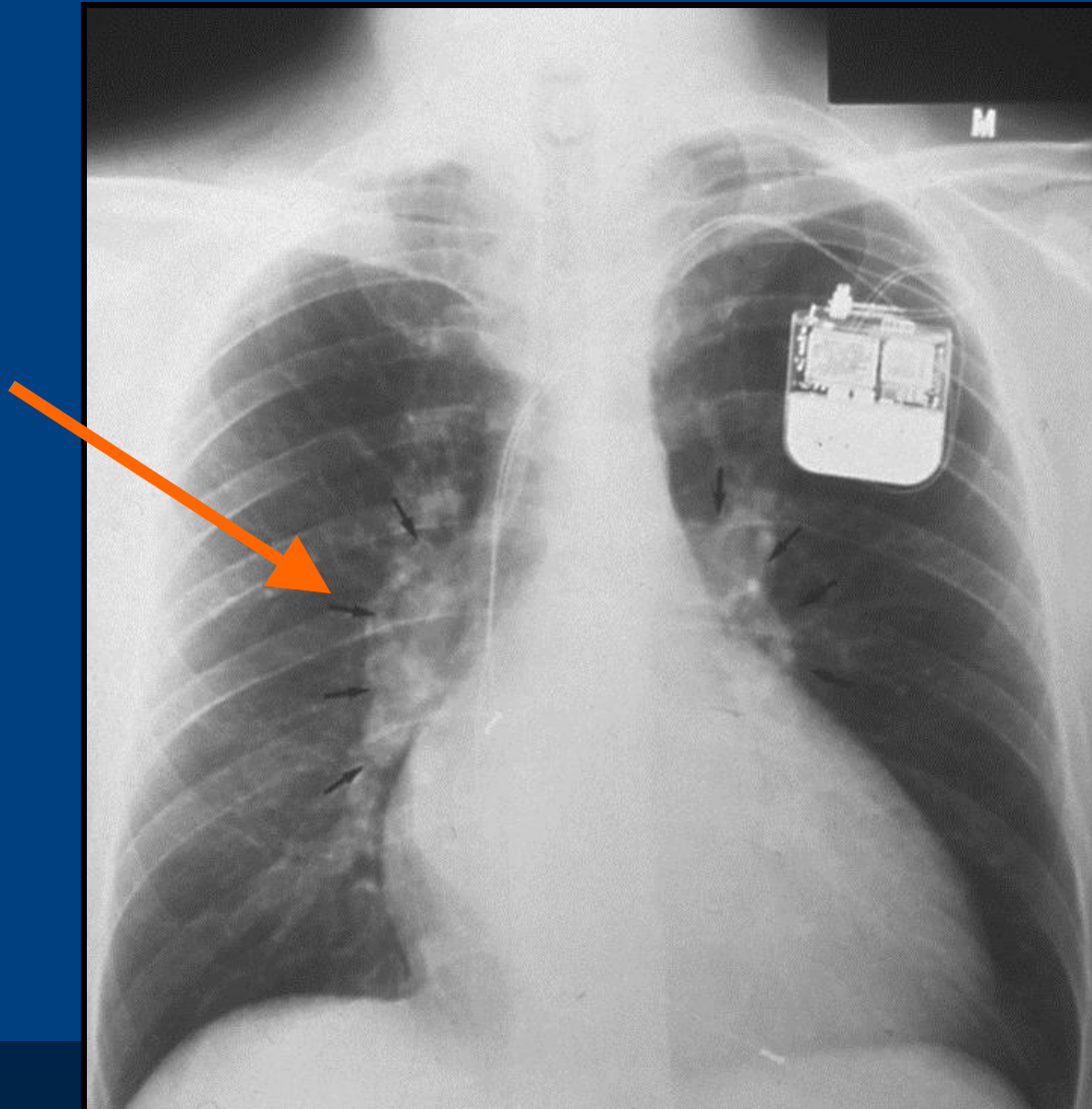
non-necrotizing granulomatous
inflammation, giant multinucleated cells,
and epithelioid histiocytes

Sarcoidosis

- Granulomatous inflammatory disease of unknown etiology involving multiple organs (immune system disorder)
- Main manifestations caused by pulmonary involvement
 - pulmonary fibrosis
 - right-sided heart failure
 - pulmonary hypertension

Sarcoid CXR

Pulmonary
Nodules



Cardiac Sarcoidosis

Clinical Cardiac Features

- Can affect any portion of the heart
- Can produce a myriad clinical problems that may simulate other more common disorders (palpitations, atypical chest pain, **syncope**, etc)
- Syncope or sudden death
 - **Heart Block, VT, VF**
- CHF
- Can also be asymptomatic

Cardiac Sarcoidosis

Echocardiographic Features

- Early: LV walls may be thick
- Late LV findings:
 - Segmental thinning
 - Regional wall motion abnormalities
 - LV aneurysms
 - LV enlargement, reduced function
- Diastolic dysfunction
- Pericardial effusion
- Valvular dysfunction(MR)
- With pulmonary involvement
 - RVE, decreased RV function and RVH

Cardiac Sarcoidosis

In Who??

- <55 yo ECG BLOCK
- Known extracardiac sarcoid with heart symptoms
- <55 yo
 - Monomorphic VT
 - Dilated CMP
 - HCM
 - ARVD

MRI

Cardiac Sarcoidosis

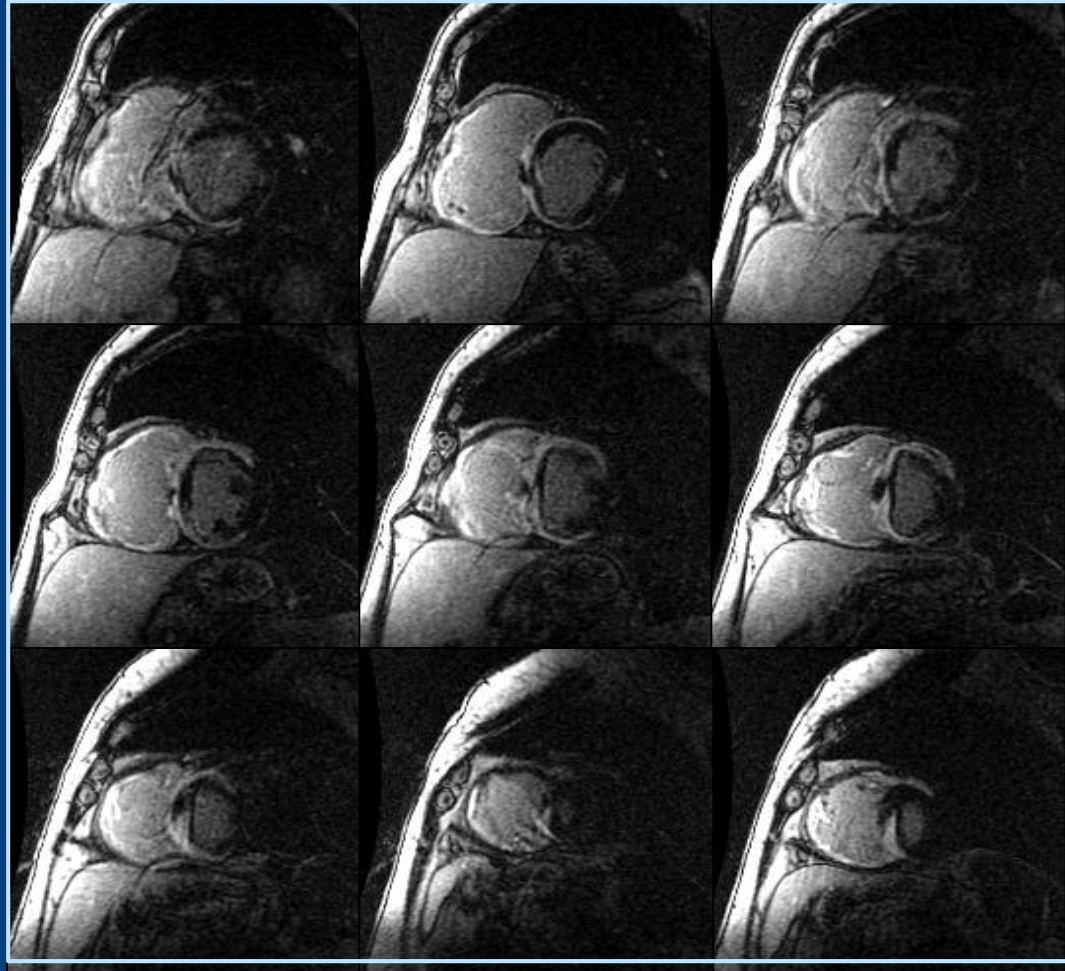
- Findings can overlap with other causes of myocarditis
- Direct cardiac involvement:
 - Clinical – 5%
 - Subclinical – about 70%
- Typical findings:
 - Wall thinning
 - Wall motion abnormalities
 - Edema in acute phase
 - Nonischemic delayed enhancement



MRI

Cardiac Sarcoidosis

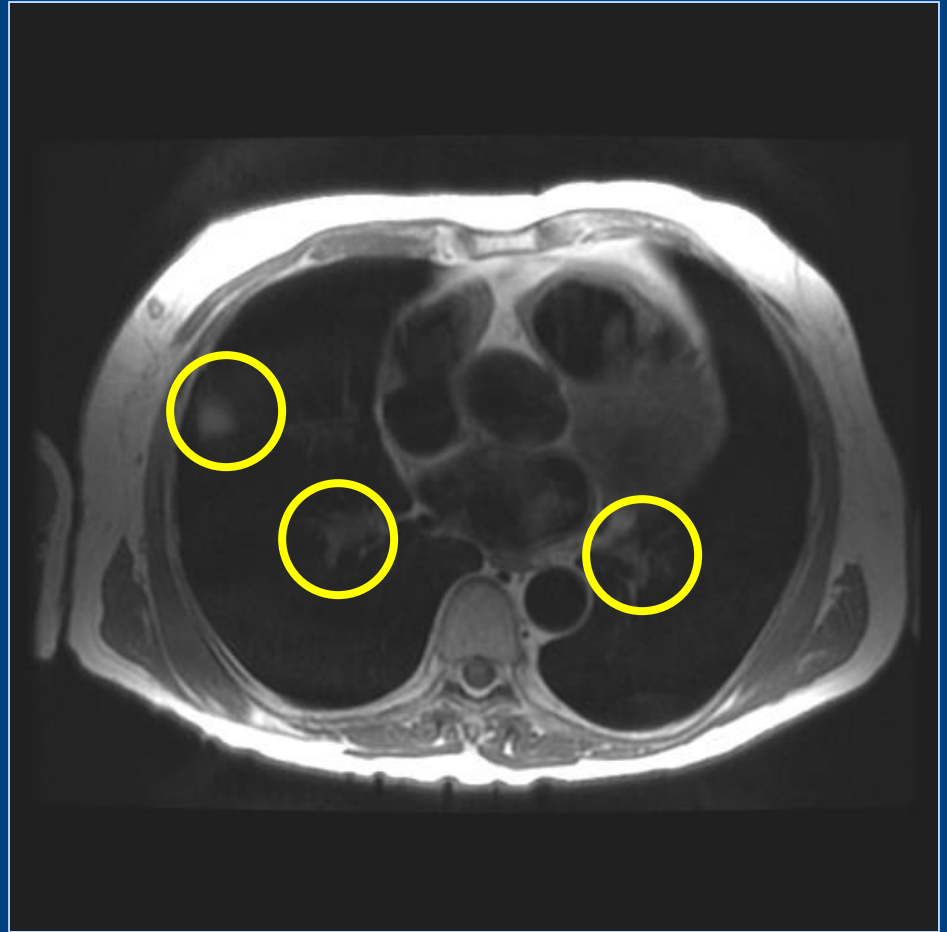
- Nonischemic delayed enhancement
 - Basal and mid septal predilection
 - RV enhancement common
 - Amount of delayed enhancement correlates inversely with EF



MRI

Cardiac Sarcoidosis

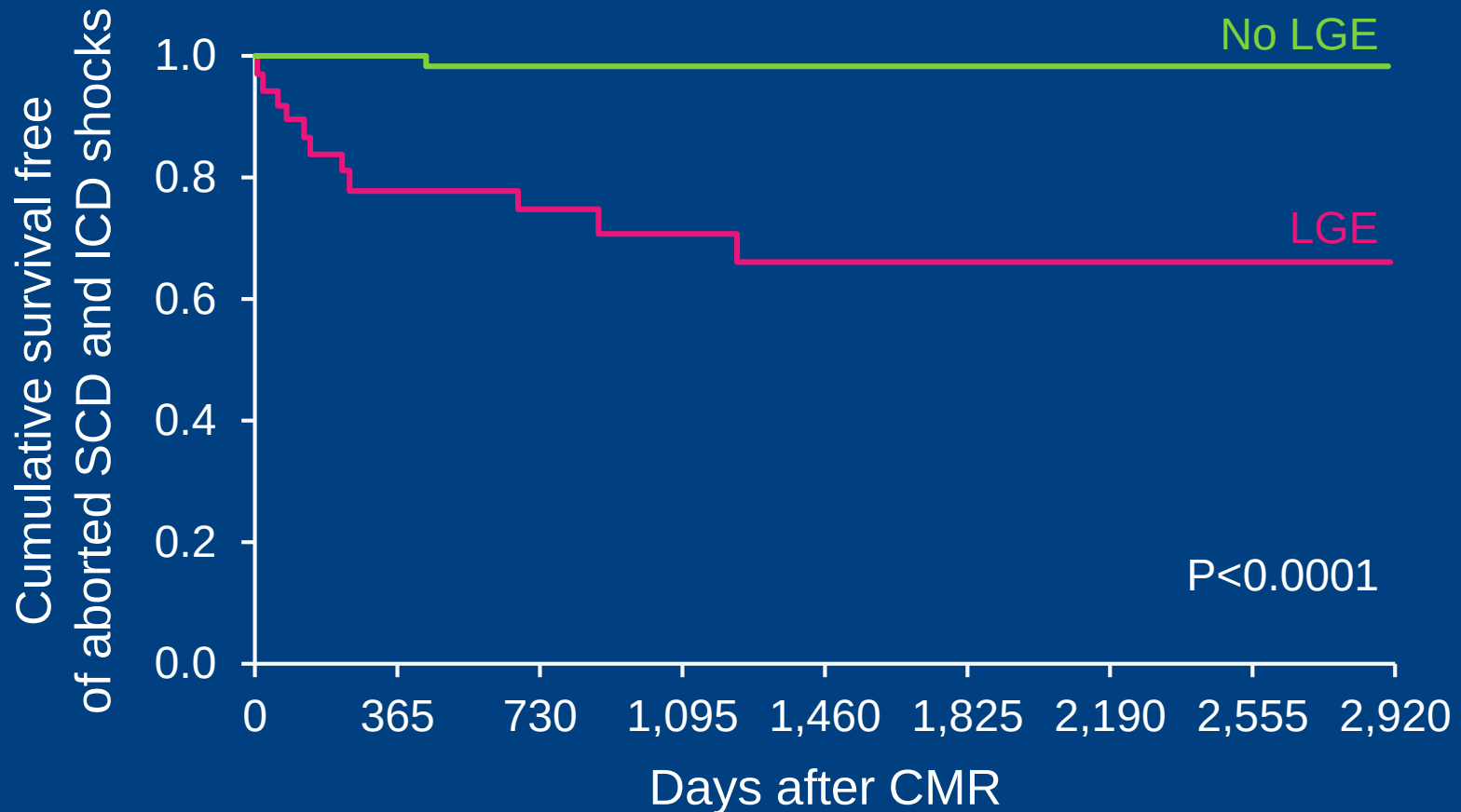
- May see pulmonary findings
 - Helps distinguish from other causes of myocarditis



CMR Imaging Predicts Death and Other Adverse Events in Suspected Cardiac Sarcoidosis

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Andreas Wahl, MD,‡ Christine Zürn, MD,§ Ulrich Kramer, MD,|| Detlev Nothnagel, MD,¶
Helmut Bültel, MD,# Julia Schumm, MD,* Stefan Grün, MD,* Peter Ong, MD,*
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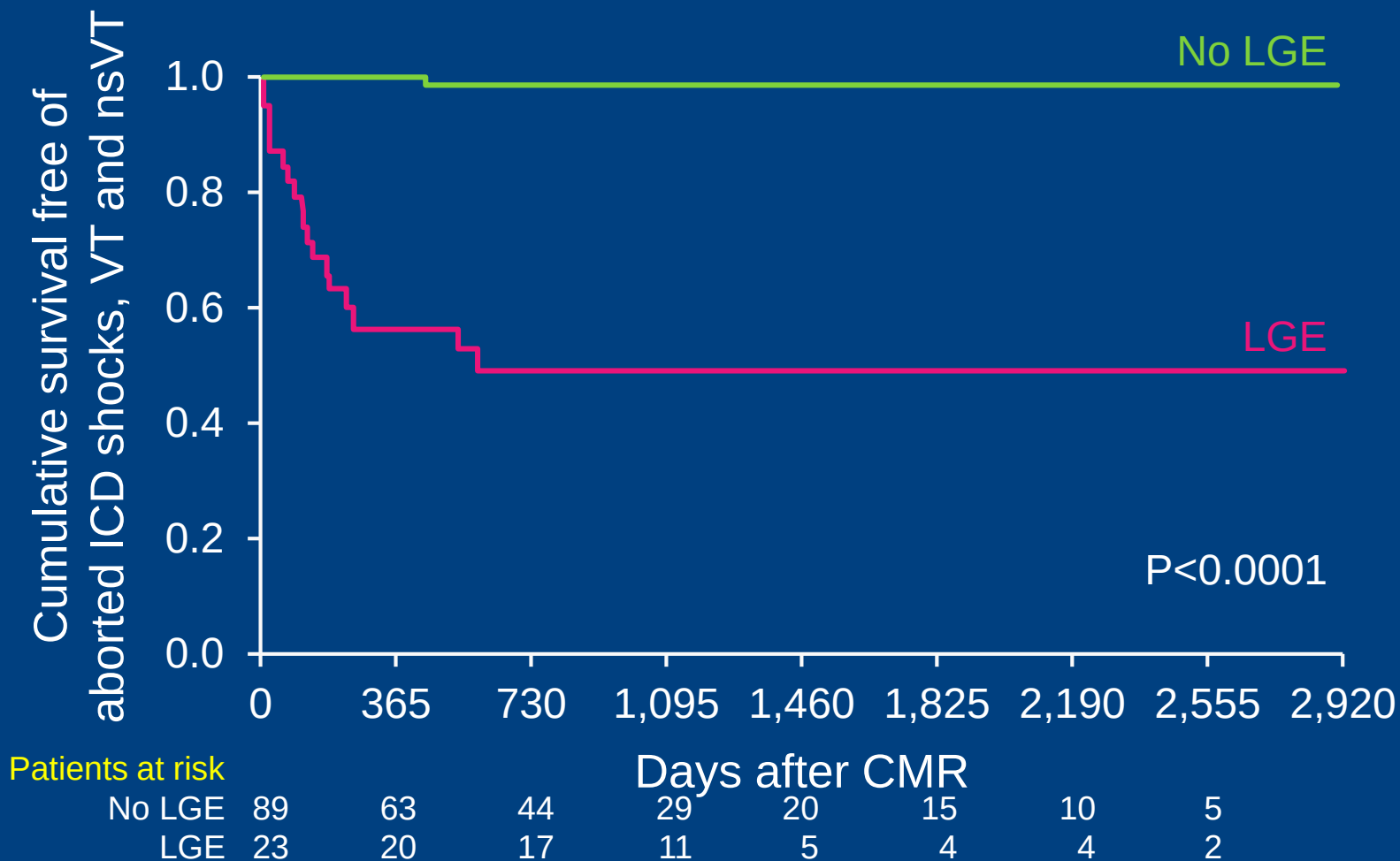
Cardiac MRI in Sarcoid Death, AICD Discharge



Greulich et al: J Am Coll Cardiol Img 6:501, 2013

Cardiac MRI in Sarcoid

Death, AICD Discharge, VT

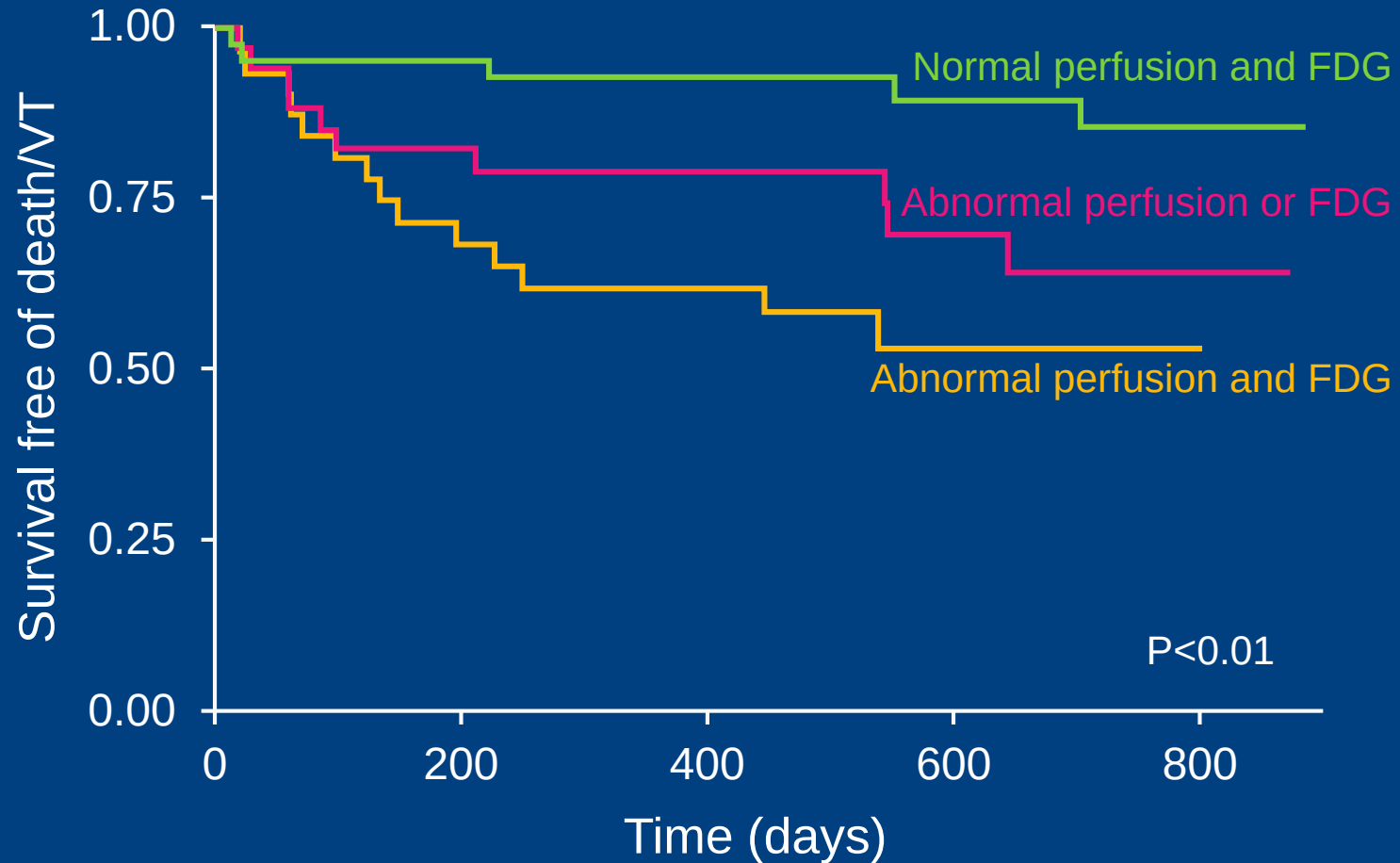


Cardiac Positron Emission Tomography Enhances Prognostic Assessments of Patients With Suspected Cardiac Sarcoidosis

Ron Blankstein, MD,^{*†} Michael Osborne, MD,[‡] Masanao Naya, MD,^{*} Alfonso Waller, MD,^{*}
Chun K. Kim, MD,[§] Venkatesh L. Murthy, MD,^{*†} Pedram Kazemian, MD,[†]
Raymond Y. Kwong, MD, MPH,^{*†} Michifumi Tokuda, MD,[†] Hicham Skali, MD,^{*†}
Robert Padera, MD, PhD,^{||} Jon Hainer, BS,^{*} William G. Stevenson, MD,[†]
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Boston, Massachusetts

Survival Free of Death or VT Stratified by Cardiac PET Examination Results



Blankstein et al: J Am Coll Cardiol 63:329, 2014

Reviews

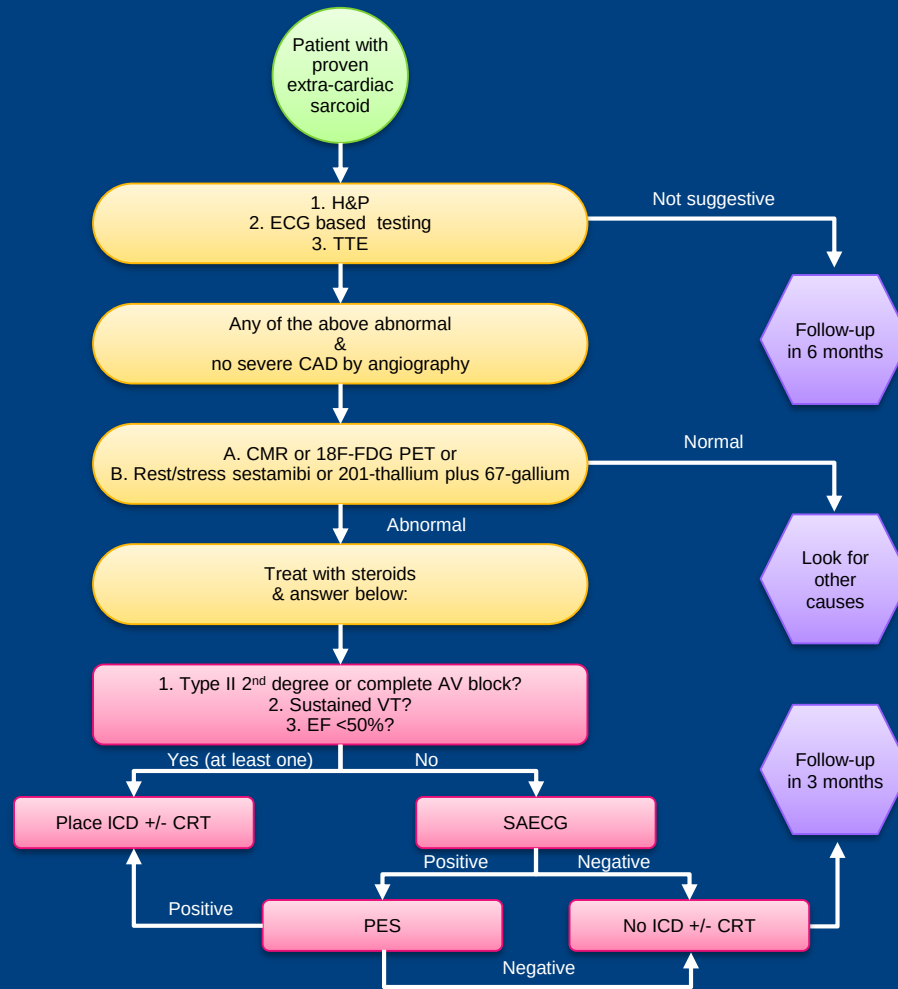


Cardiac Sarcoid: A Clinician's Review on How to Approach the Patient With Cardiac Sarcoid

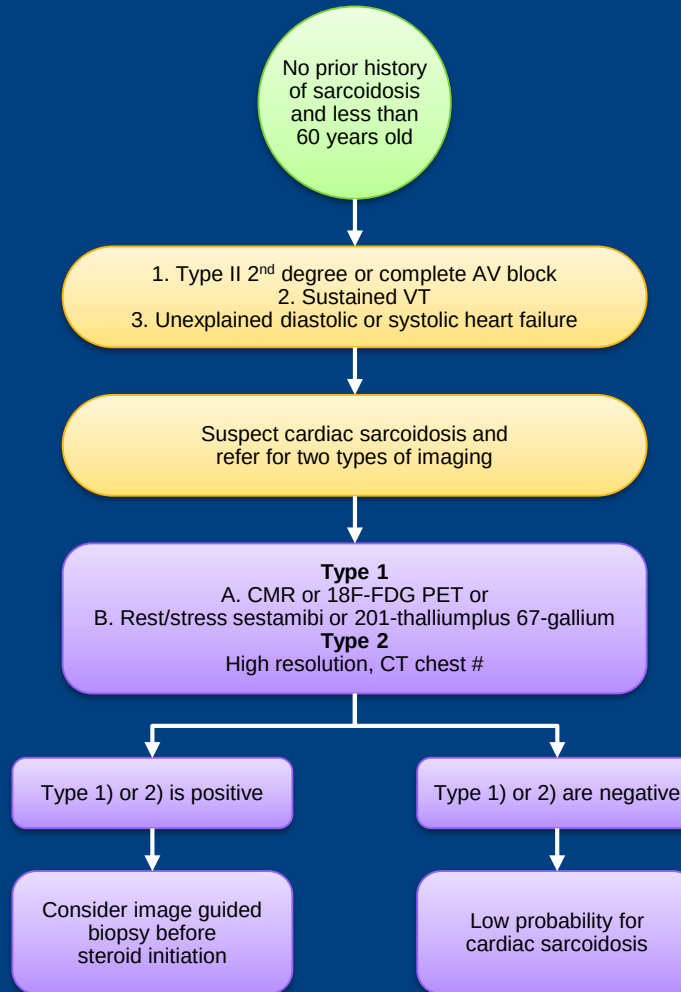
Nicholas Mantini, MD; Byron Williams, Jr. MD; Jim Stewart, MD; Leon Rubinsztain, MD; Andro Kacharava, MD, PhD

Department of Internal Medicine (Mantini), Emory University; Department of Cardiology (Kacharava), Atlanta VA Medical Center, Emory University; Department of Cardiology (Williams, Stewart), Emory University; Department of Radiology (Rubinsztain), Atlanta VA Medical Center, Emory University, Atlanta, Georgia.

Diagnostic and Treatment Algorithm for a Patient With Extracardiac Sarcoid



Diagnostic Algorithm for a Patient With Suspicion for Isolated Cardiac Sarcoidosis



Cardiac Sarcoid

- CMR may have a higher specificity for diagnosis of cardiac sarcoid
- PET appears to be more sensitive
- Both PET and CMR appear predictive of outcomes
- Local expertise and patient factors will determine optimal testing
- RV Biopsy is gold standard
 - Low sensitivity in 20% range (Patchy distribution)---Guided

Cardiac Sarcoid Treatment

- Steroids remain treatment of choice
- No Randomized Trials
- Steroid therapy
 - Appears to decrease mortality (non randomized)
 - Appears to maintain LV function if initiated in patients with normal function at time of diagnosis
 - LVEF improves in patients with mild to moderate LV dysfunction at time of diagnosis (our case)
 - No improvement in LVEF in patients with severe LV dysfunction (<30%) at time of diagnosis



Questions & Discussion