

Inflammation and Heart Disease- Myocarditis

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Myocarditis

- **Definition:** Inflammation of the myocardium leading to interstitial edema, myocyte necrosis, +/- fibrosis

Causes of Myocarditis

- **Infection**
 - **Viral *** (ie. coxsackie, herpes)
 - **Bacteria** (ie mycoplasma, pneumococcus)
 - **Spirochetal** (ie borrelia, syphilis)
 - **Protozoal** (ie. trypanosoma)
 - **Fungal** (ie histoplasma, candida)
 - **Rickettsial** (ie Q fever)
- **Giant Cell**
- **Hypersensitivity**
- **Systemic disease** (ie polymyositis, sarcoidosis)
- **Radiation**

Clinical Presentation: Myocarditis

- Congestive heart failure *
- Chest Pain
- Fatigue
- Palpitations
- Syncope
- Cardiogenic shock

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EKG: Myocarditis

- **Non-specific ST/T abnormalities ***
- **Arrhythmias**
- **Acute injury Pattern**
- **Conduction disease**

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Laboratory tests: Myocarditis

- ↑ WBC
- ↑ cardiac enzymes (acutely)
- +/- cardiac auto-antibodies

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Diagnosis of Myocarditis

- **No gold standard ****
- **Biopsy: ACC/AHA guidelines : Class IIa**
 - *Can be useful when a specific diagnosis is suspected that would influence therapy*
Circulation 2005: e154-e235.
 - **Sensitivity: 25% with 1 specimen, 66 % with 5 specimens**
Am Heart J. 2005: 7-12.
- **Antimyocin scan: Sensitivity 83%, Specificity 53%**
- **Gallium Scan: Sensitivity 36%, Specificity 98%**
- **Cardiac MRI: Sensitivity 100%, Specificity 90-100%**

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Dallas Histologic Classification for Myocarditis

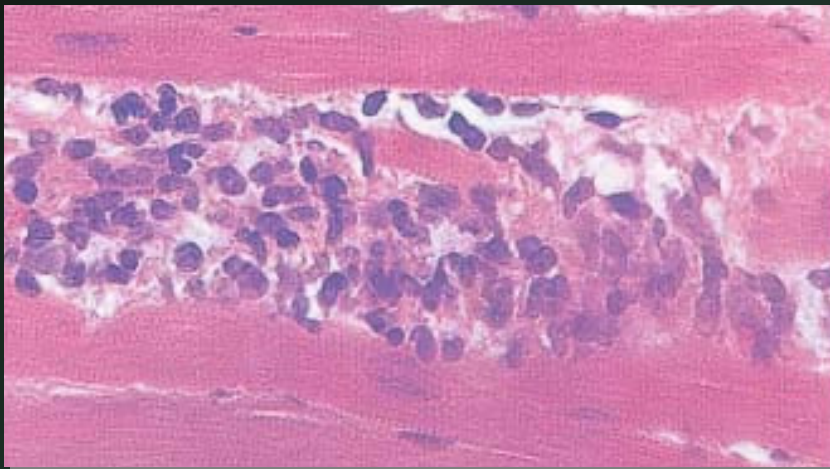
- **Myocarditis:** Inflammatory infiltrate + “damage” to adjacent myocytes
- **Borderline myocarditis:** “implies the inflammatory infiltrate too sparse and/or damage to myocyte is not demonstrable by light microscopy.

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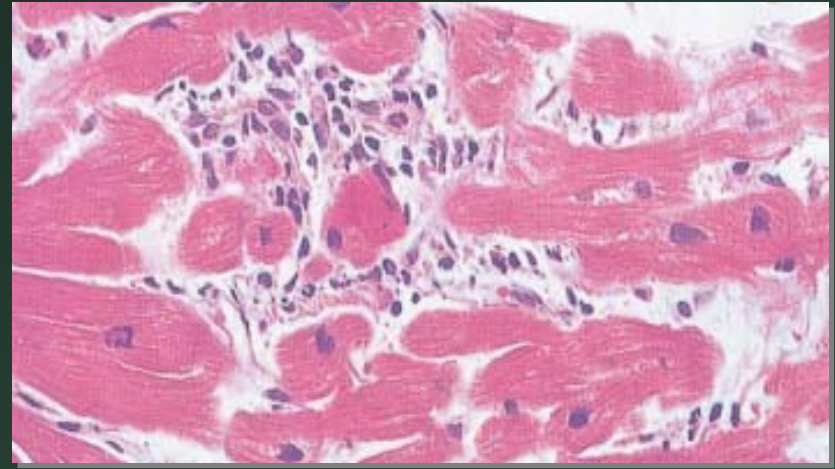
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Dallas Criteria for Myocarditis

**Active
Myocarditis**



**Borderline
Myocarditis**



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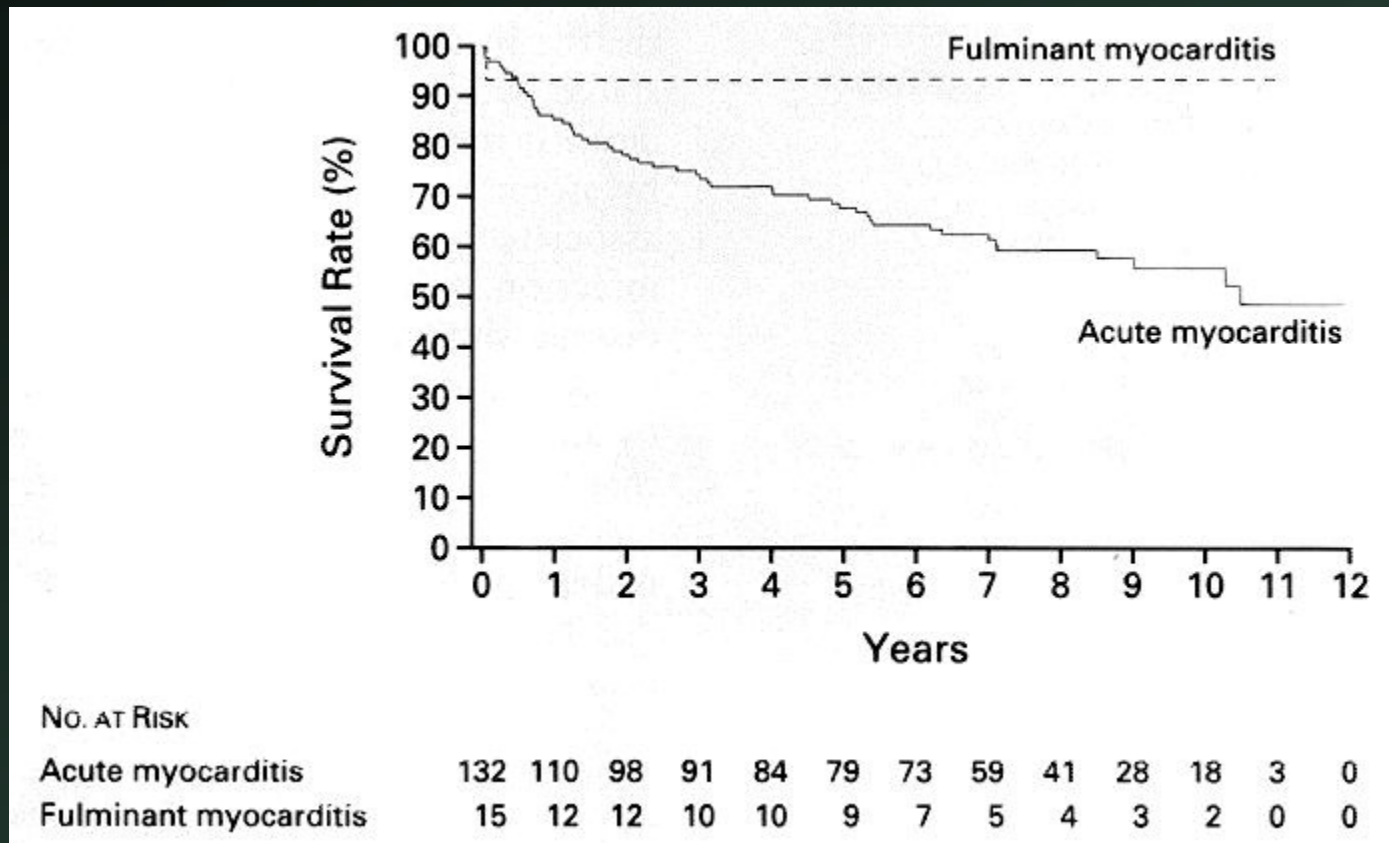
Clinicopathologic Classification of Myocarditis

	Fulminant	Subacute	Chronic Active	Chronic Persistent
Onset	Distinct	Indistinct	Indistinct	Indistinct
Biopsy +	Multi foci	Active or borderline	Active or borderline	Active or borderline
LV dysfxn	Severe	Moderate	Moderate	None
Prognosis	Recovery or death	Dilated cardio-myopathy	Restrictive cardio-myopathy	+Symptoms but no LV dysfxn

J Am Coll Cardiol. 1991; 1617-1626.

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Survival According to Clinicopathologic Classification



NEJM. 2000; 690-695.

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Therapy for Myocarditis

- **Limit physical activity**
- **Standard medical therapy for CHF**
- **No clear role for immunosuppression**
- **No clear role for immunoglobulins**

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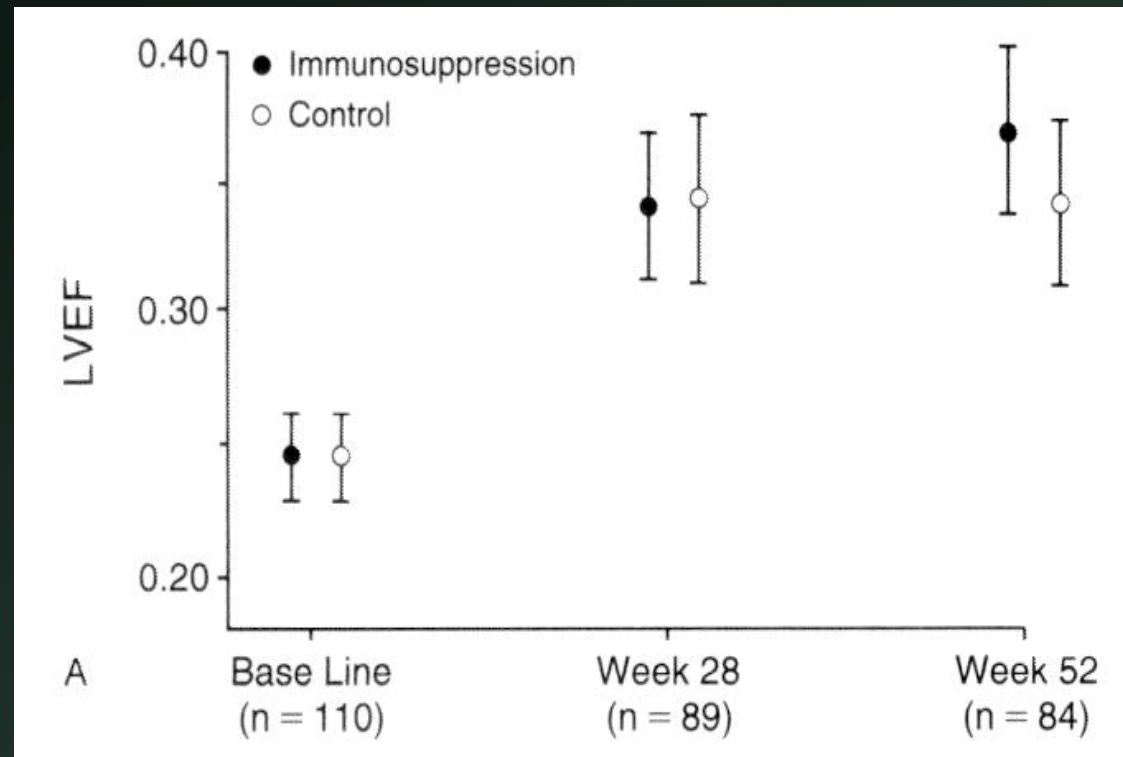
Task Force 4 Recommendations: For Athletes with Myocarditis

- Rest x 6 mo
- Can return to training/competition under the following circumstances:
 - Normal rest/exercise echo
 - No significant arrhythmias on holter or with graded exercise
 - Serum markers for heart failure and inflammation have returned to normal
 - Normal EKG

JACC 2005: 1340-1345

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Immunosuppressive Therapy for Myocarditis

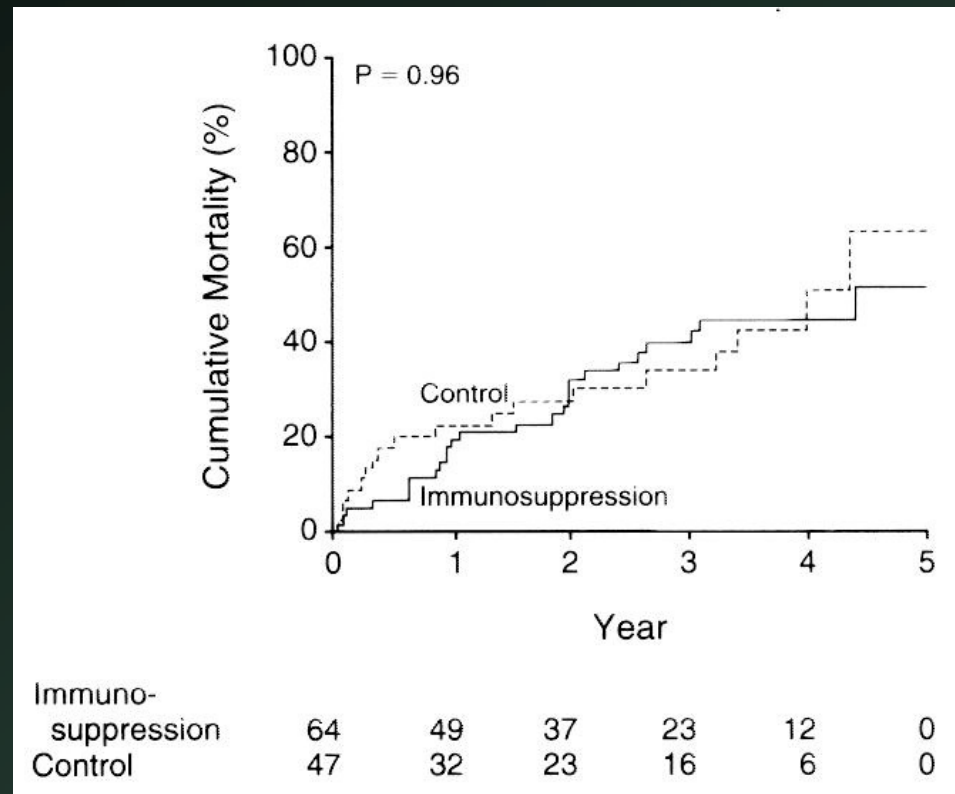


N=111 pts biopsy + myocarditis with LVEF<45%
Randomization: Placebo vs immunosuppression
(ie. prednisone + cyclosporine or azathioprine)
Result: no significant difference in LVEF

NEJM. 1995: 269-275.

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Immunosuppressive Therapy for Myocarditis



N=111 pts biopsy + myocarditis with LVEF<45%

Randomization: Placebo vs immunosuppression

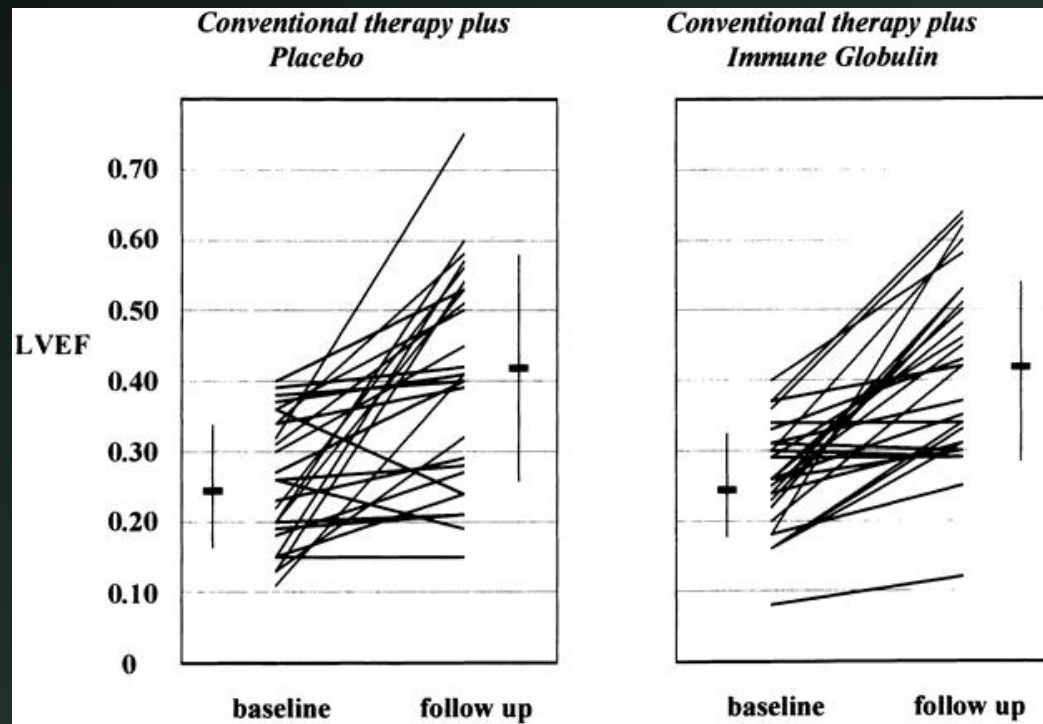
(ie. prednisone + cyclosporine or azathioprine)

Result: no significant difference in survival

NEJM. 1995; 269-275.

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Immunoglobulins for Myocarditis + Acute Cardiomyopathy



N=66 pts with recent onset LVEF<40%

Randomization: Placebo vs IVIG

Results: No significant difference in LVEF

Circulation. 2001; 2254-2259.

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Special Causes of Myocarditis



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Lyme Carditis

- **Incidence: 10% pts with Lyme disease**
- **Presentation: Syncope (CHF rare)**
- **Echocardiogram: Usually normal LV fxn**
- **EKG:**
 - AVB (all types)
 - VT rare
- **Therapy:**
 - Temporary PPM for high grade AVB
 - Disease is self limiting but abxs are often given.

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Chagas disease

- **Pathogen:** *Trypanosoma cruzi*
- **Prevalence:** Central and South America
- **Presentation:**
 - Usually 20 yrs after acute infection
 - CHF (esp, right sided)
 - Arrhythmias/Sudden Death
 - Chest pain
 - Thromboembolic events

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Chagas disease

- **Echocardiogram:** Cardiomegaly (R>L)
- **EKG:** RBBB *, LAFB *, atrial fibrillation, ventricular arrhythmias*
- **Diagnosis:** serologic or PCR
- **Prognosis:** poor
- **Therapy:** B-blockers, amiodarone, ICD, heart transplantation (esp, in Brazil)

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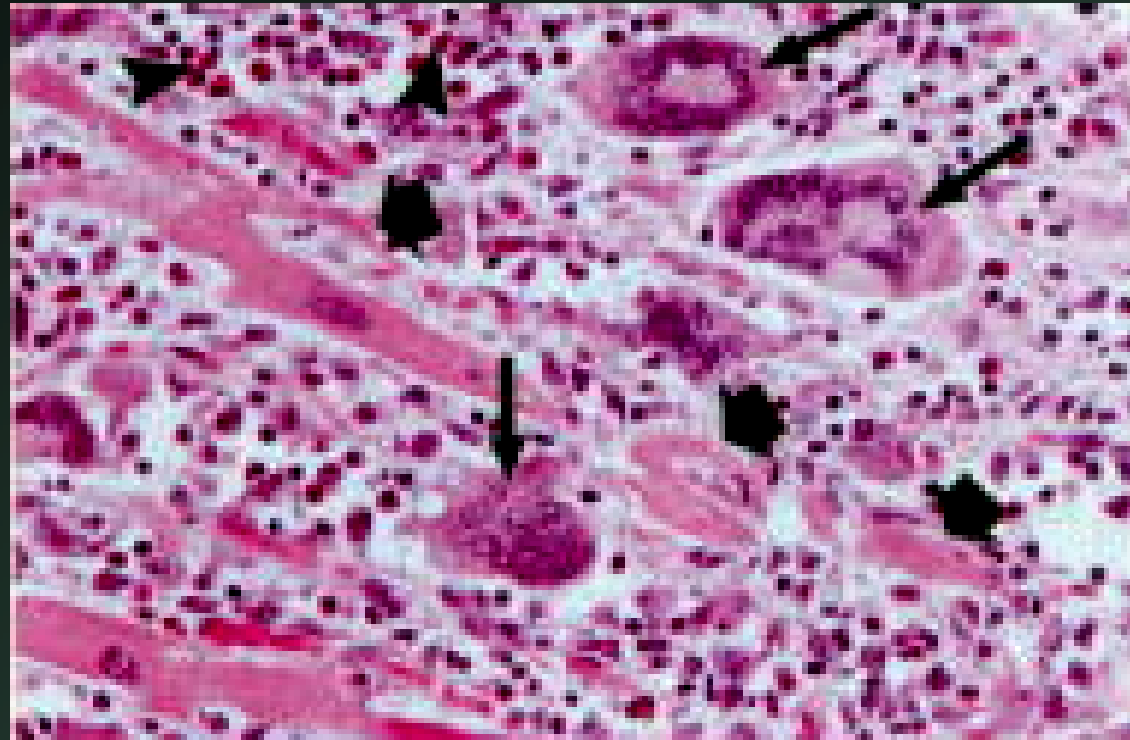
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Giant Cell Myocarditis

- **Presentation:** Abrupt onset of CHF with rapid progression + refractory ventricular arrhythmias
- **EKG:** Arrhythmias and conduction dz
- **Biopsy:** + Giant cells, inflammation, fibrosis
- **Prognosis:** Median Survival 5.5 mo
- **Therapy:** Immunosuppression, mechanical support, early heart transplantation

Am Heart J. 2005: 7-12.

Giant Cell Myocarditis



Multinucleated giant cells (long arrows), degenerating myocytes (short arrows), inflammatory cells (arrowheads).

NEJM . 1997: 1860-1866.

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Hypersensitivity Myocarditis

- **Symptoms/Signs: Fever, rash, eosinophilia**
- **EKG: Nonspecific**
- **Biopsy: + Eosinophils, other inflammatory cells, little necrosis**
- **Cause:**
 - 1) **Drugs**, 2) **Primary eosinophilic syndrome**

Am Heart J. 2005: 7-12.

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Hypersensitivity Myocarditis

- **Therapy:**
 - Discontinue offending drug
 - Corticosteroids +/- other immunosuppression
- **Prognosis: Variable**
 - Recovery
 - Severe LV dysfunction +/- valvular disease
 - Sudden death from arrhythmias
 - Arteritis

Am Heart J. 2005: 7-12.

Cardiac Sarcoidosis

- $\leq 5\%$ pts with systemic sarcoidosis
- Presentation: AVB, CHF, VT
- Biopsy: + non-caseating granulomas
- Therapy: ? Immunosuppressive therapy (*although ACC/AHA 2005 guidelines state that there is no conclusive evidence that treatment can affect the course of the disease*)

*Braunwald. 7th Edition
Circulation 2005: e154-e235.*

Conclusion: Myocarditis

- Most common cause is viral
- No adequate gold standard for diagnosis
- Biopsy is Class IIa indication but not indicated for routine evaluation (although essential for dx of giant cell)
- No clear role for immunosuppressive therapy in viral myocarditis
- Giant cell myocarditis is rapidly fatal and should be suspected if recurrent VT + rapid deterioration of CHF

Case #1

40 yo man had a viral like illness and then 2wks later had heart failure symptoms/signs with LVEF 20%

Does he need a RV biopsy?

A Yes

B No

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Case #2

36 yo man from El Salvador has megaesophagus, NSVT, and R > L heart failure. LVEF 20% with + LV apical aneurysm. LHC no CAD. EKG RBBB with LAFB. Serology c/w T cruzi

Are anti-protozoal antibiotics helpful?

A Yes

B No

Case #2

36 yo man from El Salvador has megaesophagus, NSVT, and R > L heart failure. LVEF 20% with + LV apical aneurysm. LHC no CAD. EKG RBBB with LAFB. Serology c/w T cruzi

Are anti-protozoal antibiotics helpful?

A Yes

B No

Case #3

40 yo previously healthy man now presents with refractory VT and a cardiomyopathy. He is deteriorating quickly. No preceding viral symptoms. No fever or rash.

What disease do you suspect?

- A Giant Cell Myocarditis
- B Hypersensitivity Myocarditis
- C Lyme disease

Case #3

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What disease do you suspect?

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- B Hypersensitivity Myocarditis
- C Lyme disease

Case #4

25 yo woman was hiking this summer and was bitten by a tick. She developed a rash and now in ER after a syncopal episode. Echo nl. EKG with complete heart block.

Does she need a permanent PPM?

A Yes

B No

Case #4

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A Yes

B No