PLACE 2026
PLATFORM OF LABORATORIES FOR ADVANCES IN CARDIAC EXPERIENCE

CONGRESSO
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CARDIOMIOPATIE RARE, CARDIOPATIE NEUROMUSCOLARI E MALATTIA DI ANDERSON-FABRY

# Sarcoidosi


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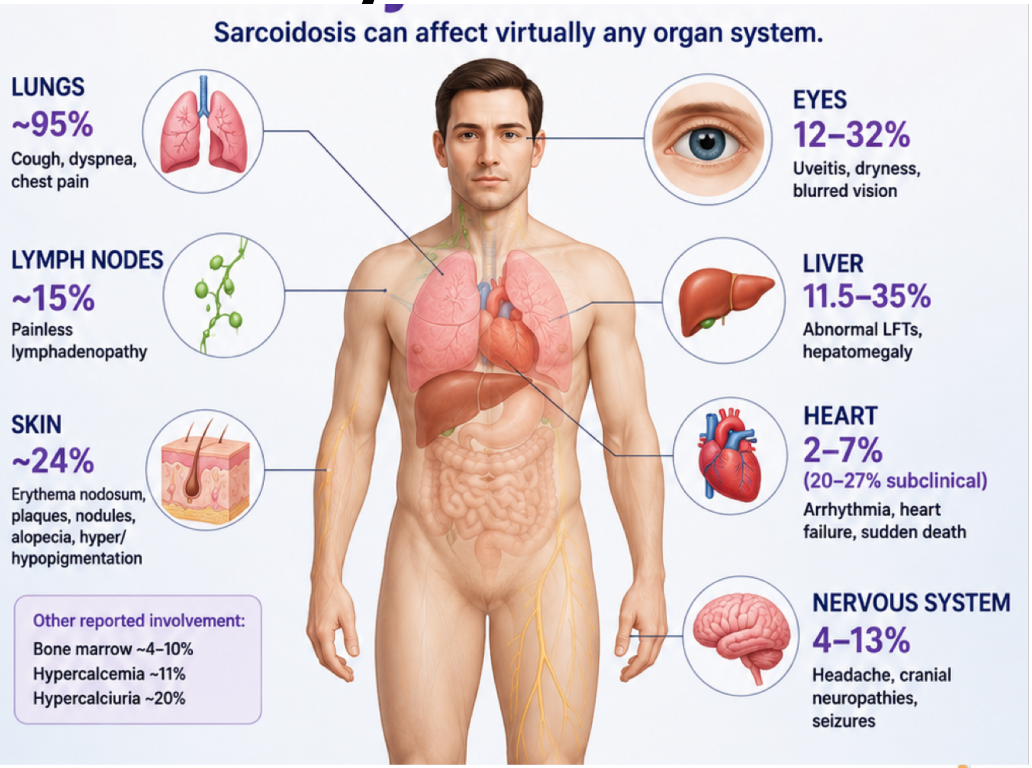
# Disclosure

No competing interests declared

# Sarcoidosis as a Multisystem Disease

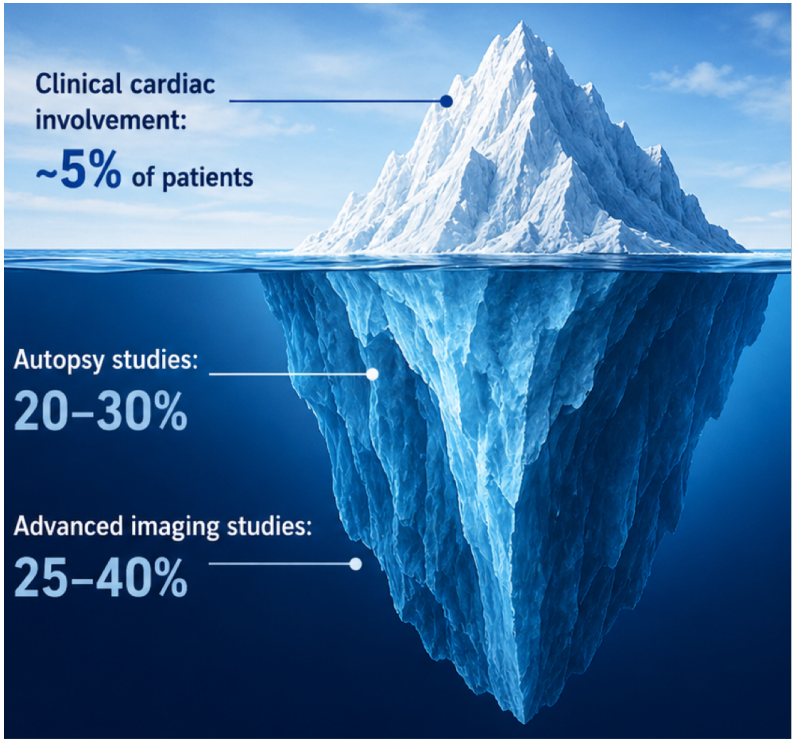
- Sarcoidosis is a systemic immune-mediated granulomatous disease of unknown cause.

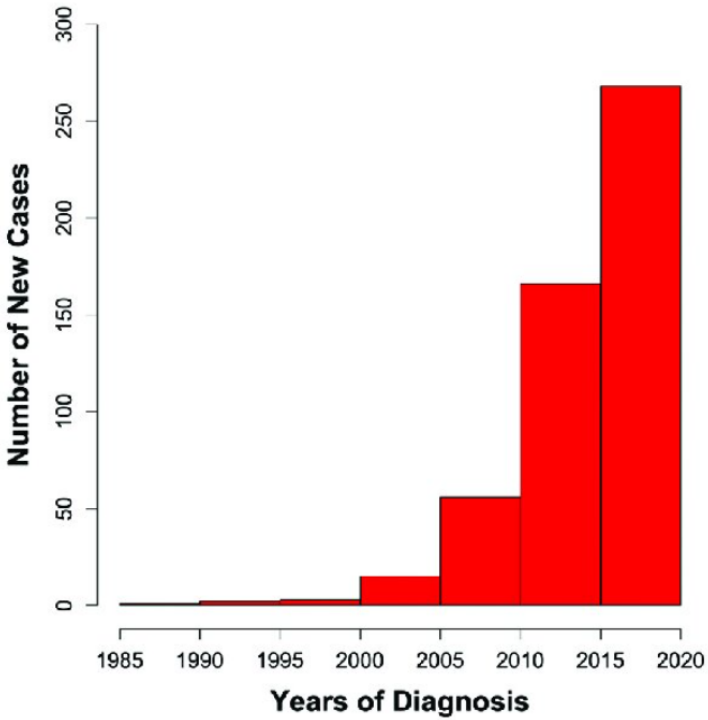
- Although pulmonary involvement dominates clinically, cardiac involvement often determines prognosis.




Lehtonen J et al. Eur Heart J 2023; Birnie DH et al. Heart Rhythm 2014

# Why Should Cardiologists Care?

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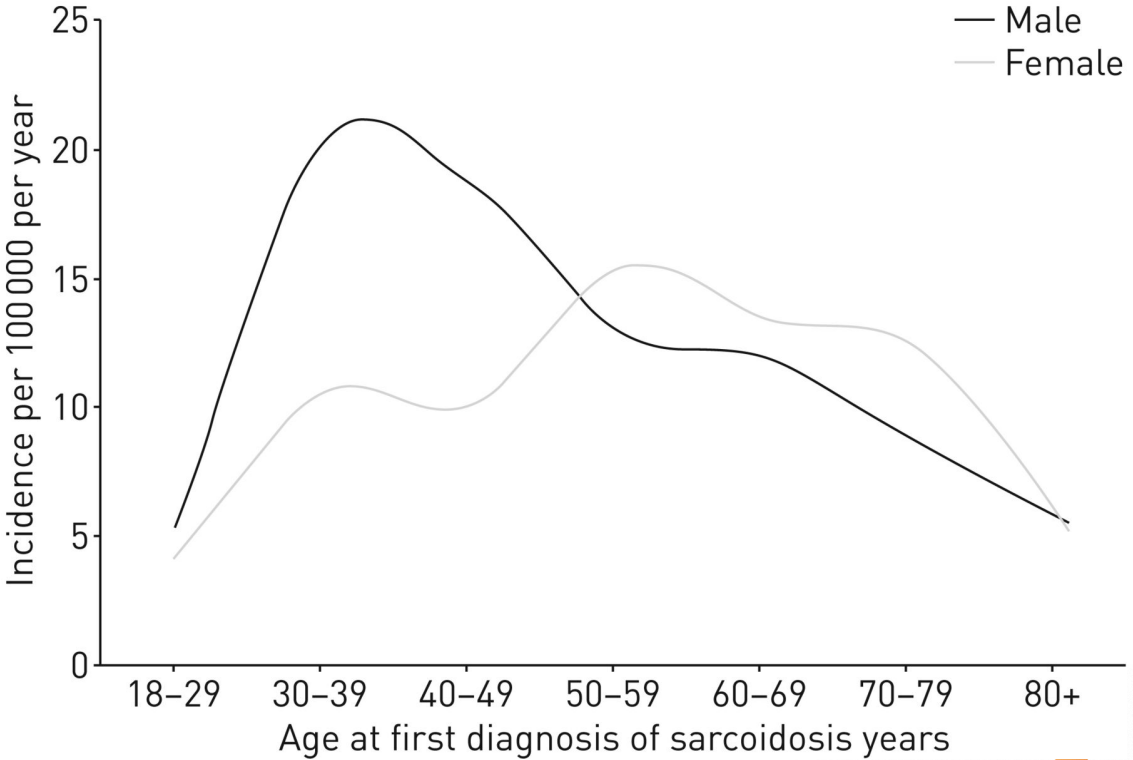
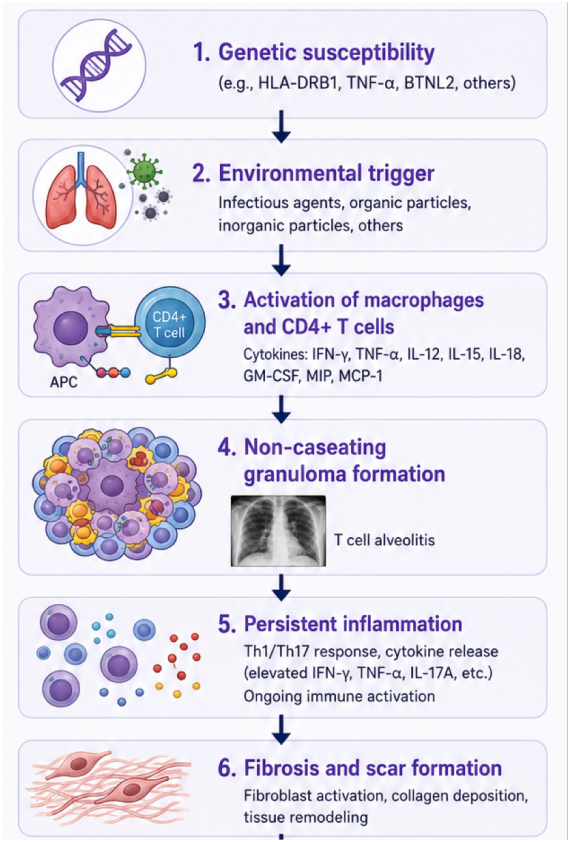
# Epidemiology

Incidence of sarcoidosis per 100 000 per year by age and sex in Sweden (2003–2012)




*Arkema E et al. Eur Respir J 2016*

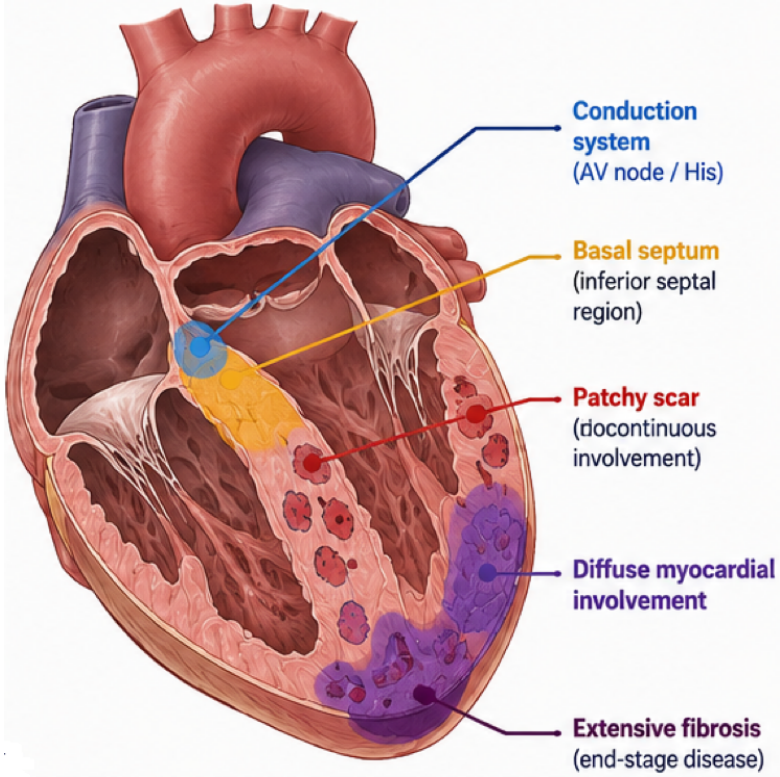
# Pathophysiology




- Disease activity and fibrosis may coexist simultaneously.
- Inflammation is potentially reversible. Scar is usually permanent.


Valeyre D. Lancet 2014; Iannuzzi MC. N Engl J Med 2007

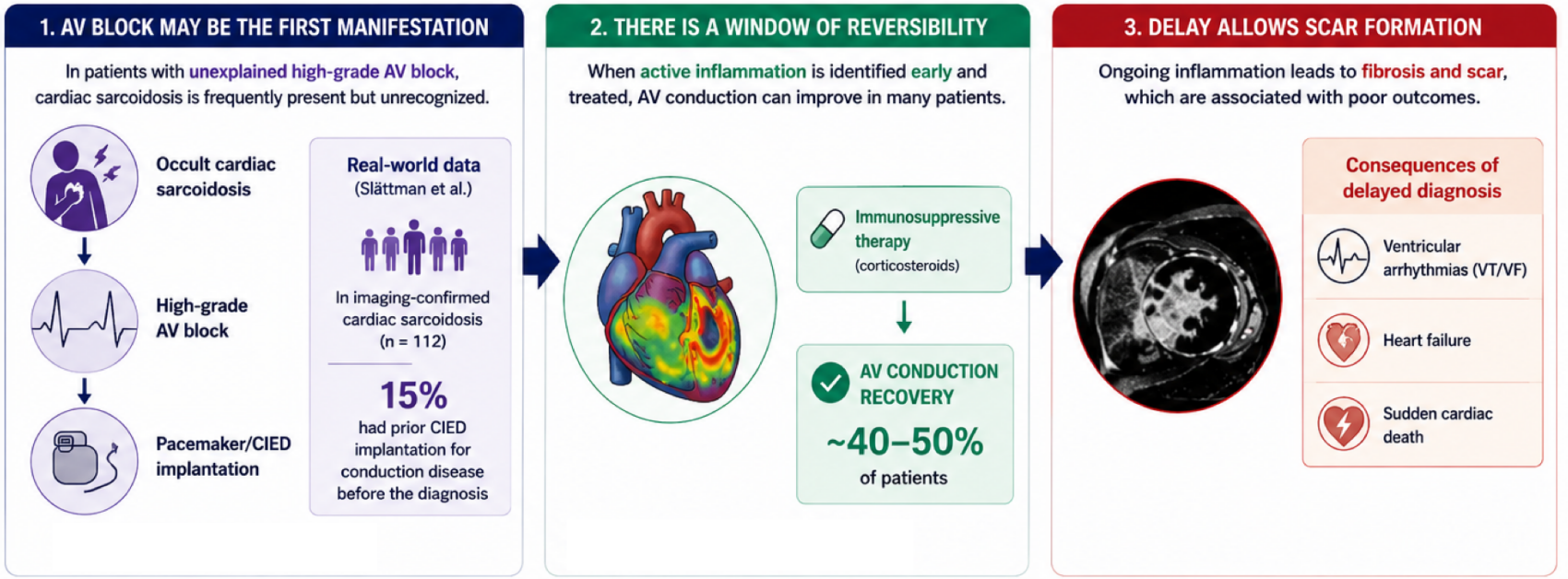
# Why Does Cardiac Sarcoidosis Present So Differently?




| LOCATION OF INJURY | PREDOMINANT CLINICAL PRESENTATION |
|---|---|
| Conduction system (AV node / His) bundle) | → AV block |
| Basal septal inflammation | → Bundle branch block |
| Patchy scar (non-ischemic distribution) | → Ventricular arrhythmias (VT / VF) |
| Diffuse myocardial involvement | → Heart failure |
| Extensive fibrosis (end-stage) | → End-stage cardiomyopathy |

Lehtonen J et al. Eur Heart J 2023

# Why Early Recognition Matters




**The greatest opportunity in cardiac sarcoidosis is recognizing inflammation before irreversible scar develops.**

Slättman J. et al. Eur Heart Cardiovasc Imaging 2021; Sadek MM. Et al. Can J Cardiol 2013

# Phenotypes of Cardiac Sarcoidosis

**1. CS detected only at autopsy**
- Unexpected sudden cardiac death (SCD) with no prior diagnosis.

**2. CS with known extracardiac sarcoidosis**
- Established sarcoidosis and no or minimal cardiac symptoms
- Myocardial involvement found in 25–30% of sarcoidosis patients

**3. Clinically manifest CS**
- Acute or significant cardiac symptoms
- Isolated/de novo CS ranges from 3% to 43% and is generally more severe.

# When to Suspect Cardiac Sarcoidosis

- **Suspect cardiac sarcoidosis** in patients with known sarcoidosis who develop **cardiac symptoms or ECG/echo abnormalities**.

- Also suspect CS in **any patient** with **unexplained**:

1. 2nd–3rd degree AV block
2. Sustained ventricular arrhythmias
3. New-onset heart failure

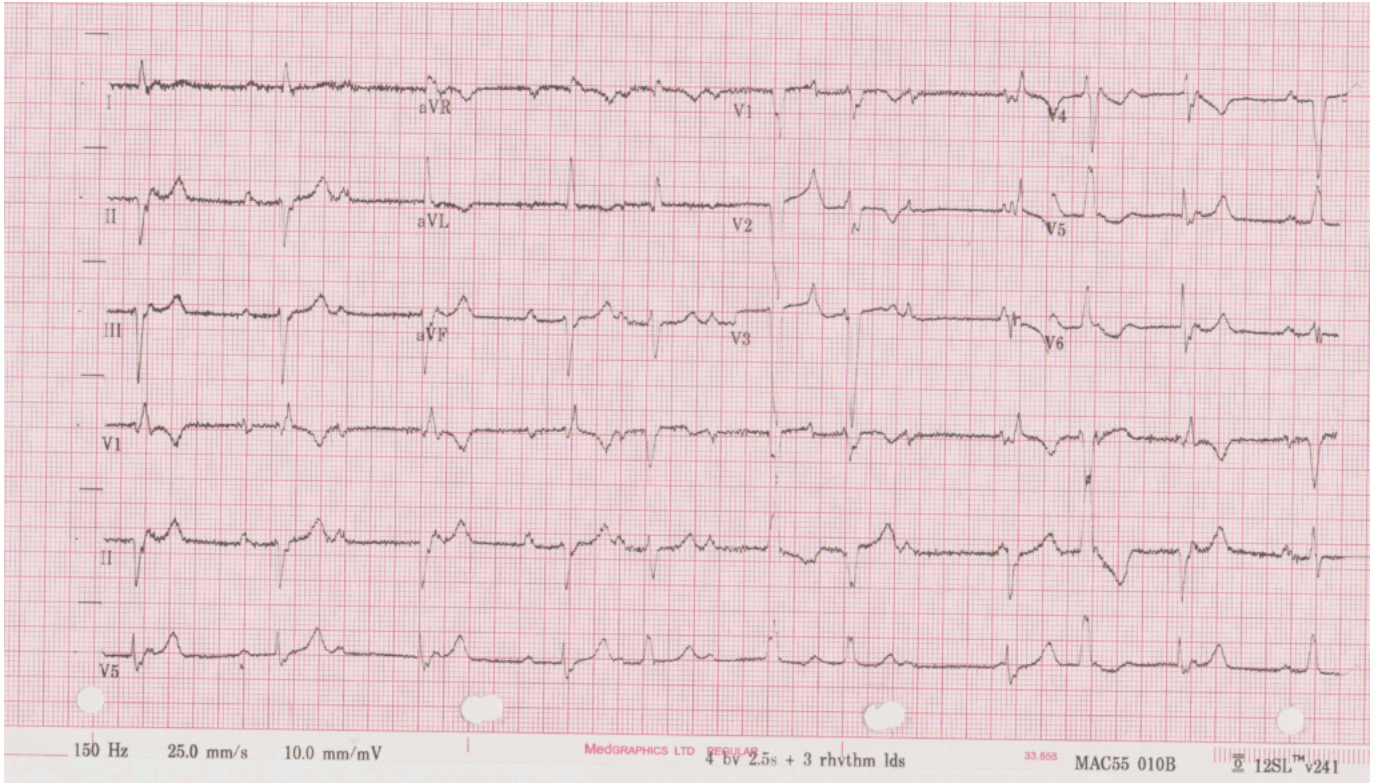
- In patients with sarcoidosis, **elevated troponin**, **natriuretic peptides**, and emerging **anti-heart / anti–**intercalated **disk antibodies** strengthen suspicion

**CS accounts for:**

- 20–34% of idiopathic high-grade AV block in middle-aged adults
- A similar share of idiopathic sustained VT
- About 5% of mixed ventricular arrhythmias (including frequent PVCs)

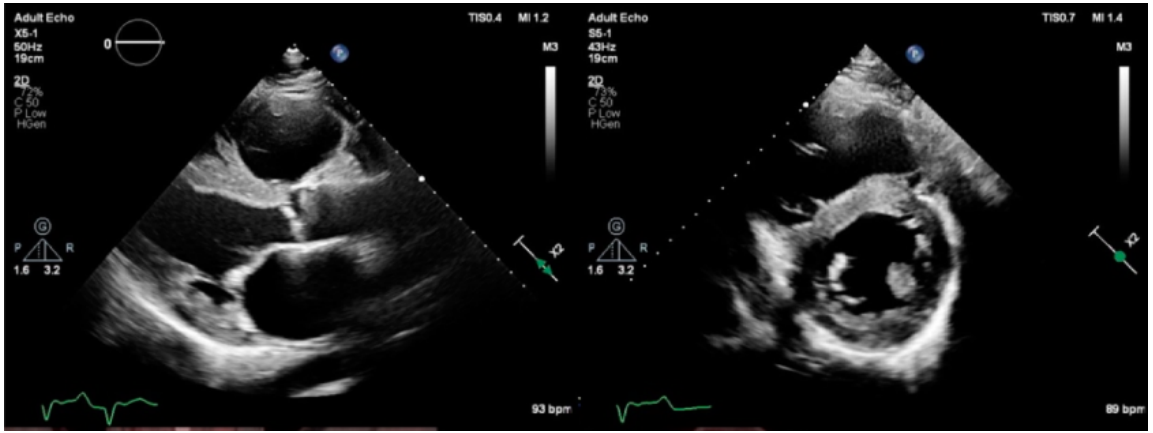
# ECG Red Flags

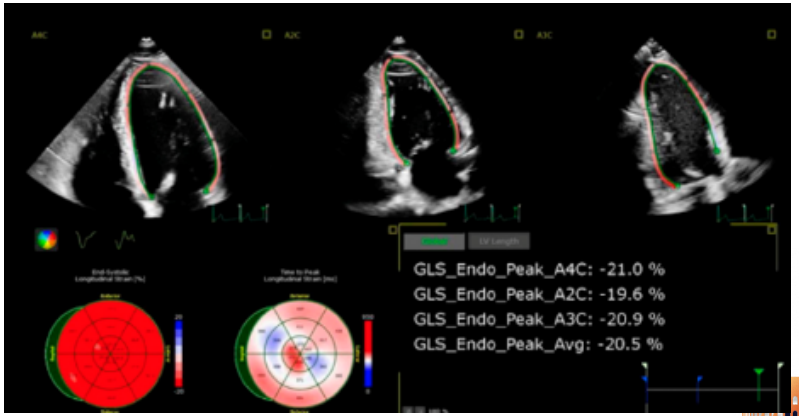
- AV block (II–III)
- New bundle-branch block
- QRS fragmentation
- PVCs, NSVT



# Echocardiographic Red Flags

| Echocardiogram |
| --- |
| LV dysfunction (EF < 50%) |
| LV wall thickening or thinning |
| Local akinesia or dyskinesia |
| LV aneurysm |
| Reduced global LV longitudinal strain |
| RV enlargement and reduced RV free wall strain |
| Pericardial effusion |

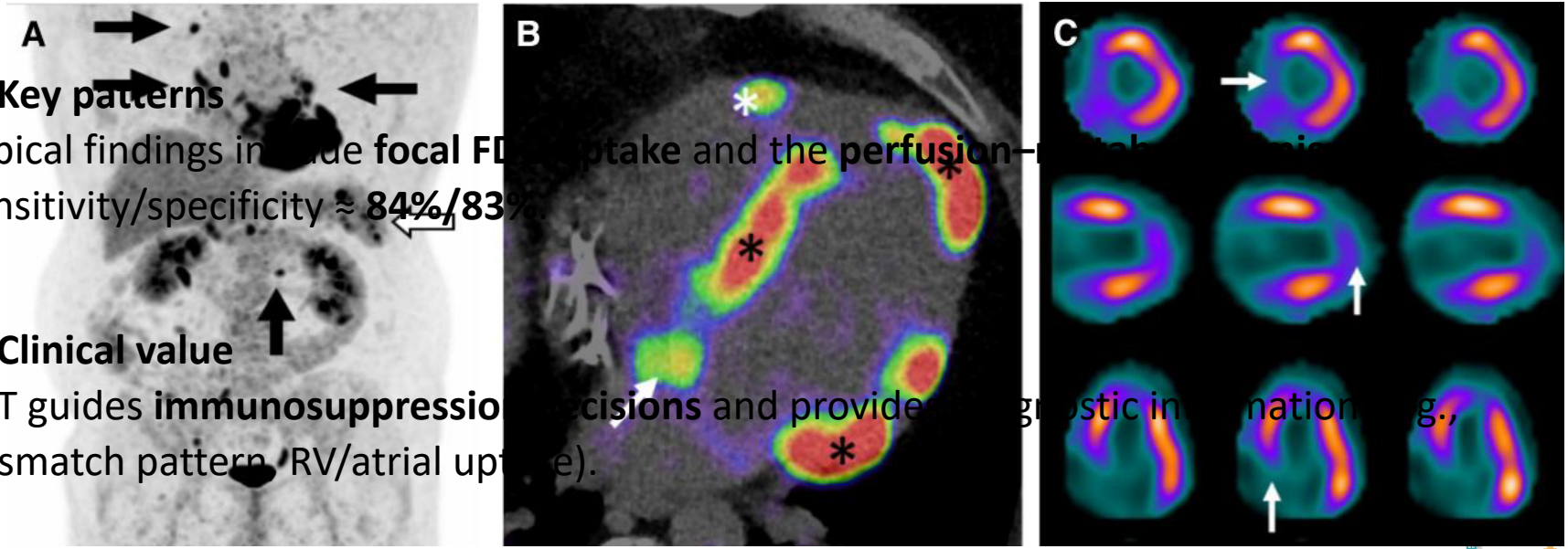


# Advanced imaging - $^{18}$F-FDG-PET

## 1. Diagnostic role

18F-FDG-PET detects **active cardiac inflammation** but requires metabolic preparation; ~10–15% of scans are non-diagnostic.




## 2. Key patterns

Typical findings in[illegible]ude **focal FD[illegible]**ptake and the **perfusion–[illegible]**
sensitivity/specificity ≈ **84%/83%**

## 3. Clinical value

PET guides **immunosuppressio[illegible]cisions** and provide[illegible]gnostic in[illegible]mation[illegible]g.,
mismatch pattern, RV/atrial upt[illegible]e).

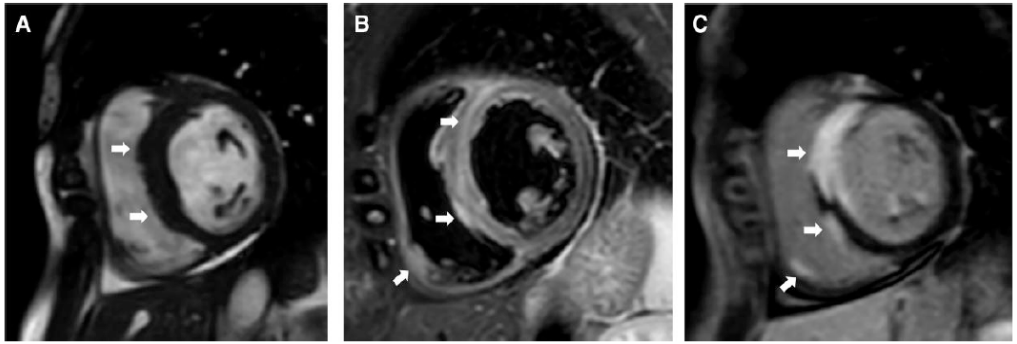
# Advanced Imaging - CMR

- CMR assesses **structure, function, edema, inflammation, and scarring**

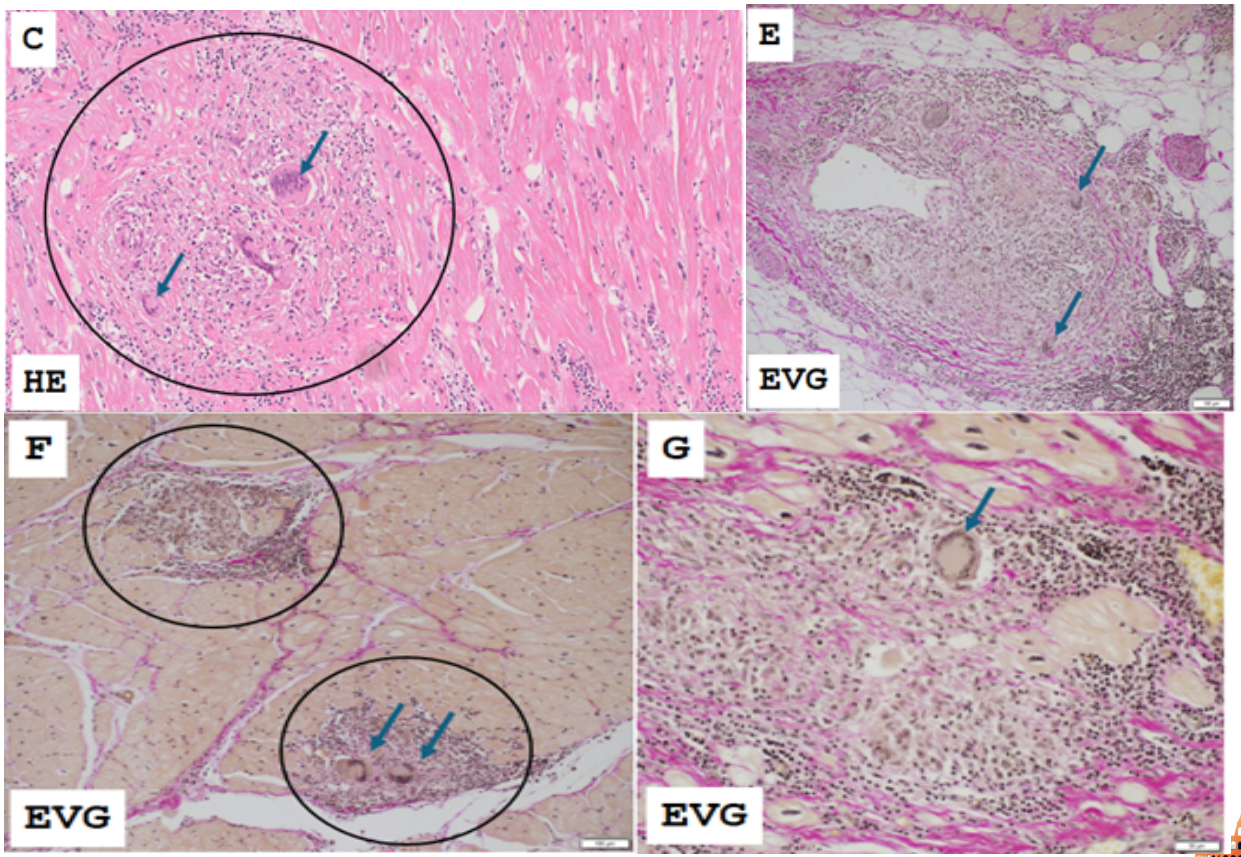
- **Late gadolinium enhancement (LGE)**: typically patchy, non-ischemic, often involving basal LV and the septal region

- Diagnostic performance: **sensitivity 75–100%**, **specificity 77–85%**

- LGE burden (extent/segments involved) is strongly associated with **risk of adverse outcomes**



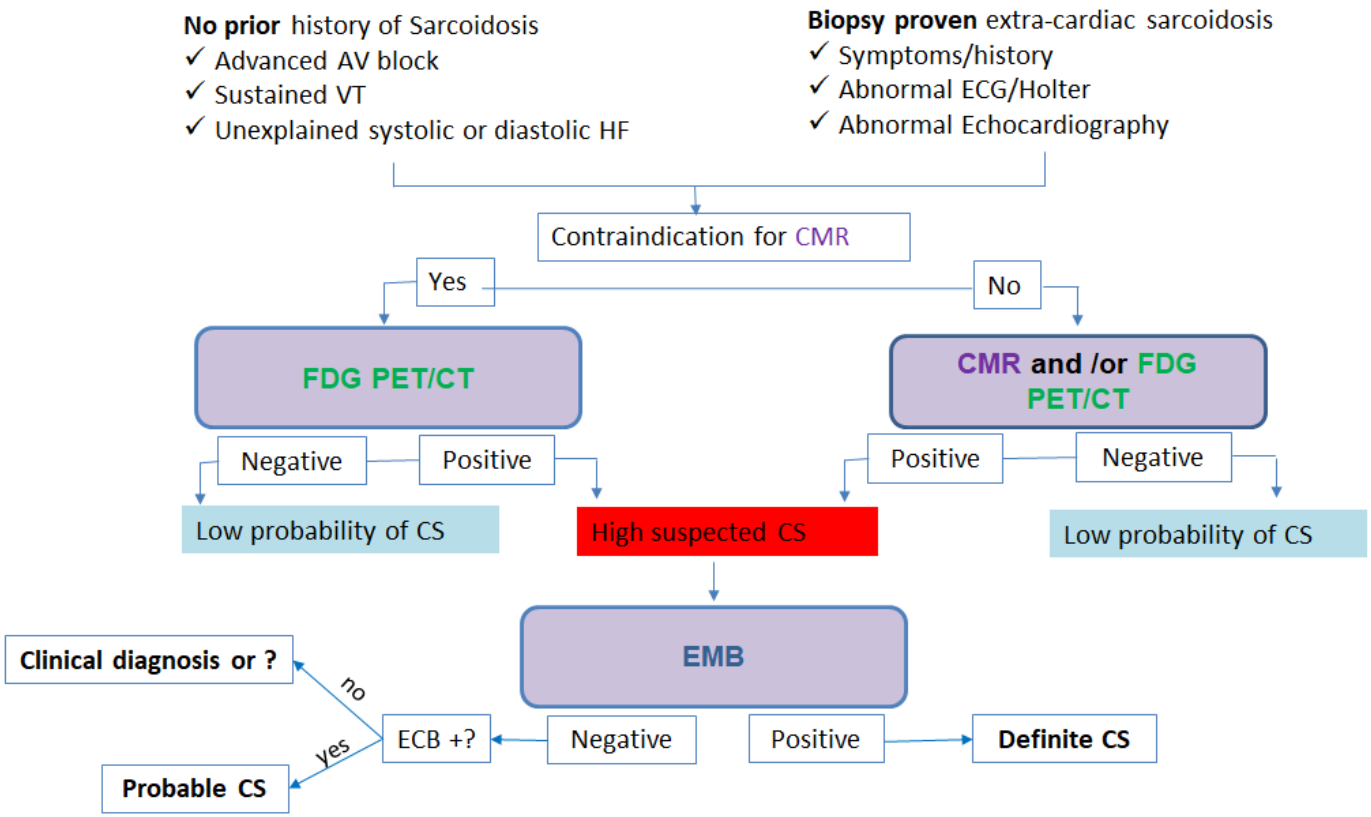
# Endomyocardial Biopsy



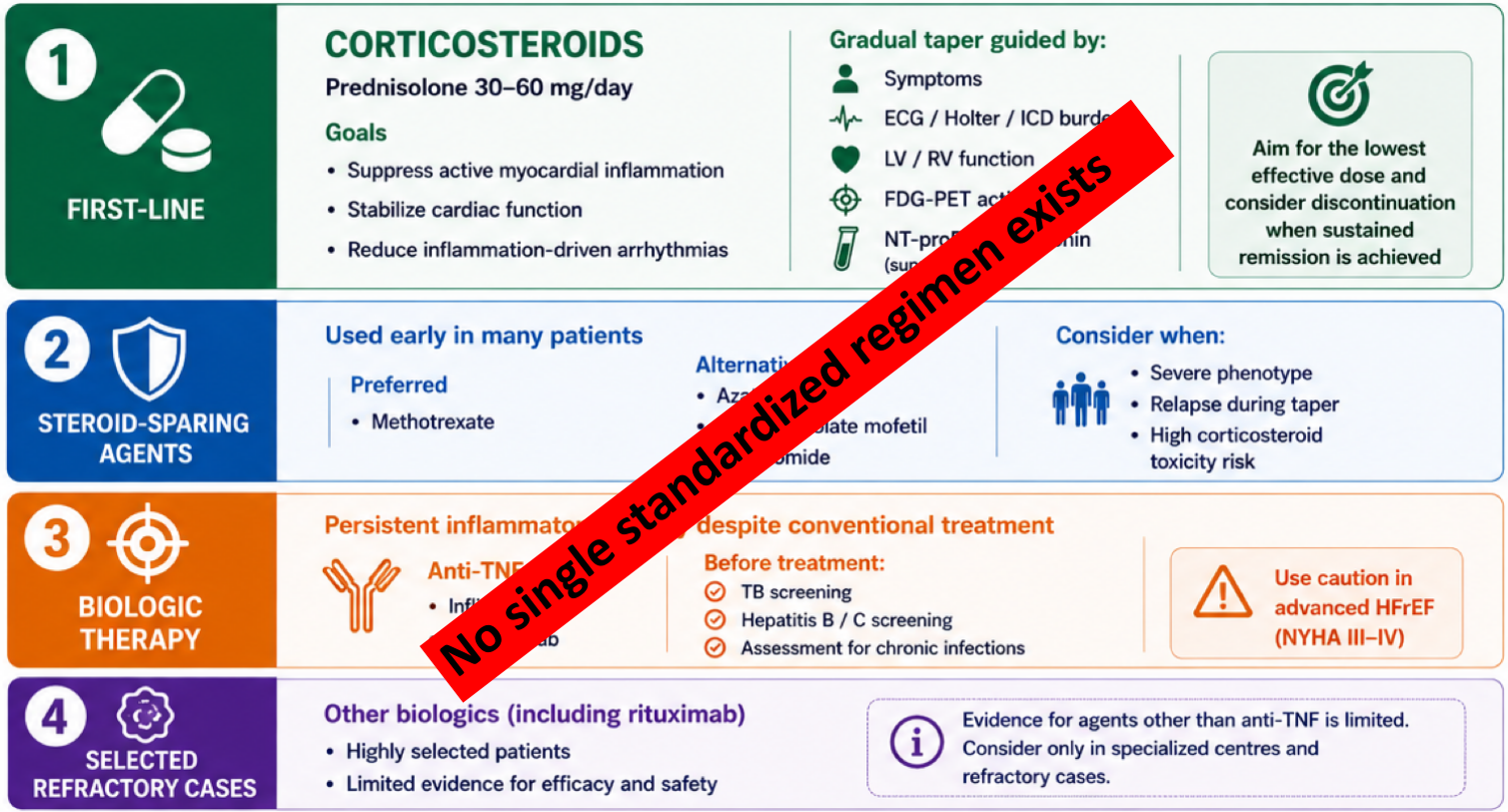
# Diagnostic Criteria

| | JCS[9] | HRS[7] | WASOG[8] |
|---|---|---|---|
| Histological diagnosis of CS | | | |
| EMB with noncaseating granulomas | x | x | X |
| Clinical diagnosis of CS | | | |
| Histological diagnosis of extracardiac sarcoidosis mandatory for clinical diagnosis | | X | x |
| Mobitz type II 2nd-degree heart block or 3rd-degree heart block | x | X | x |
| Unexplained sustained (spontaneous or induced) VT | x | X | x |
| Patchy uptake on dedicated cardiac PET (pattern consistent with CS) | x | X | x |
| Late gadolinium enhancement on CMR (pattern consistent with CS) | x | X | x |
| Positive gallium uptake (pattern consistent with CS) | x | X | x |
| Steroid ± immunosuppressant-responsive cardiomyopathy or heart block | | X | x |

# Diagnosis: definitive/probable/presumable?



# Immunosuppression in Cardiac Sarcoidosis



# Device Therapy in Cardiac Sarcoidosis

## ICD INDICATIONS IN CARDIAC SARCOIDOSIS*

*Based on 2017 AHA/ACC/HRS Guidelines and 2022 ESC Guidelines*

### SECONDARY PREVENTION

| | |
|---|---|
| Survived cardiac arrest (aborted SCD) | Class I |
| Documented sustained VT | Class I |

### PRIMARY PREVENTION

| | |
|---|---|
| LVEF ≤ 35% | Class I |
| Indication for permanent pacing for high-grade AV block with LVEF > 35% | Class IIa |
| Unexplained syncope with LVEF > 35% | Class IIa |
| Myocardial scar on CMR or PET with LVEF > 35% | Class IIa |

* Meaningful survival > 1 year is expected and reversible causes have been excluded.

## ADDITIONAL RISK STRATIFICATION

**Electrophysiological study (EPS)**

Reasonable for additional risk stratification in selected patients with LVEF > 35% who have:

- Uncertain arrhythmic risk
- Preserved or mildly reduced ventricular function
- Myocardial scar on CMR or PET
- No prior sustained VT/VF

↓

**Inducible sustained VT at EPS supports ICD implantation**

Class IIa

## CLINICAL APPROACH

High-grade AV block or other CS presentation

↓

Need for permanent pacing

↓

**Assess arrhythmic risk**

LVEF, scar (CMR/PET), syncope, prior VT/VF, EPS (selected)

↓

**ICD is generally preferred over pacemaker alone in CS**

# Take-home Messages

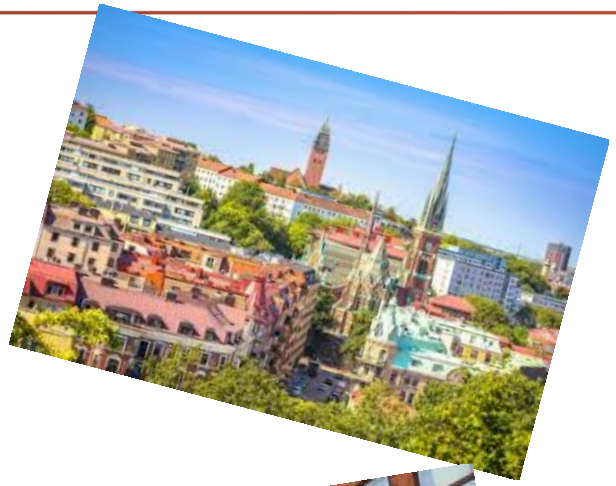
1. **Cardiac sarcoidosis is often under-recognized**
   Clinical disease represents only the tip of the iceberg; myocardial involvement is substantially more common when assessed by advanced imaging.

2. **Think beyond the heart**
   Cardiac sarcoidosis is the cardiac manifestation of a systemic immune-mediated disease and requires a multidisciplinary approach.

3. **Early recognition matters**
   Unexplained AV block, ventricular arrhythmias, heart failure, or characteristic CMR/PET findings should prompt consideration of cardiac sarcoidosis.

4. **Treatment requires two parallel strategies**
   - Control **inflammation** with immunosuppression
   - Manage **consequences**:
     - heart failure therapy, ICDs, catheter ablation when indicated

5. **Risk stratification goes beyond EF**
   LGE burden, FDG-PET activity, RV involvement, arrhythmias, and conduction disease are key determinants of prognosis and therapeutic decisions.



# Thank you!