

Azərbaycan
Kardiologiya
Cəmiyyəti



SARKOİD ÜRƏK: NECƏ TANIYAQ VƏ KİMİNLƏ TƏQİB EDƏK?

HAZIRLADI: FESC. DR. CƏMİL BABAYEV



WHO
ARE
YOU?

Tarixçə:

It was first described in 1877 by Dr. [Jonathan Hutchinson](#), a [dermatologist](#) as a condition causing red, raised rashes on the face, arms, and hands.^[15]

In 1889 the term [lupus pernio](#) was coined by Dr. [Ernest Besnier](#), another dermatologist.^[170]

Later in 1892 lupus pernio's [histology](#) was defined.^[170] In 1902 bone involvement was first described by a group of three doctors.^[170] Between 1909 and 1910 uveitis in sarcoidosis was first described, and later in 1915 it was emphasised, by Dr. [Jörgen Nielsen Schaumann](#), that it was a systemic condition.^[170]

This same year lung involvement was also described.^[170] In 1937 [uveoparotid fever](#) was first described and likewise in 1941 [Löfgren syndrome](#) was first described.^[170]

In 1958 the first international conference on sarcoidosis was called in London, likewise the first USA sarcoidosis conference occurred in Washington, D.C., in the year 1961.^[170] It has also been called [Besnier–Boeck](#) disease or [Besnier–Boeck–Schaumann](#) disease.^[171]

First Heart Sarcoidosis was identified in 1929 by M.Bernstein.

Cardiac Sarcoidosis

Epidemiology, Characteristics, and Outcome Over 25 Years in a Nationwide Study

Riina Kandolin, MD; Jukka Lehtonen, MD; Juhani Airaksinen, MD; Tapani Vihinen, MD

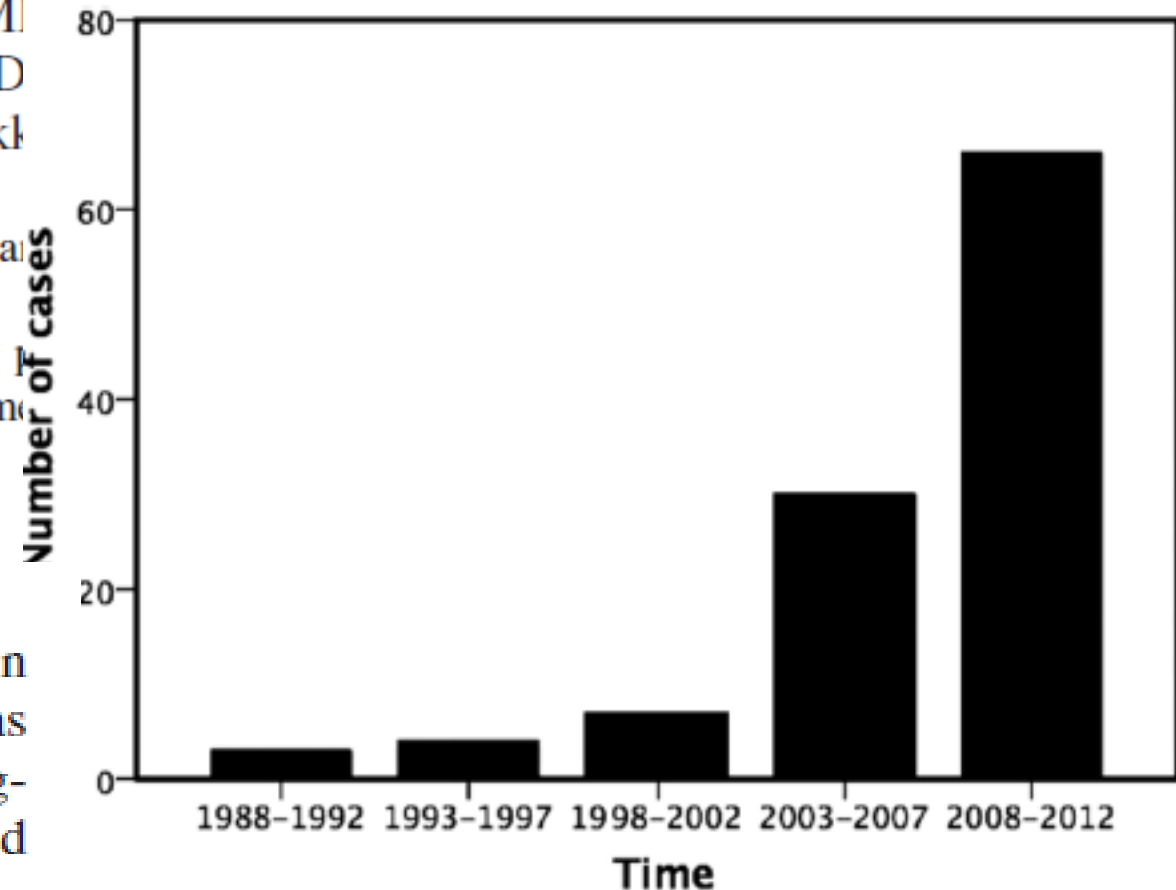
Heikki Miettinen, MD; Kari Ylitalo, MD; Kari Kaikkonen, MD
Petri Haataja, MD; Tuomas Kerola, MD; Jorma Kokkonen, MD
Päivi Pietilä-Effati, MD; Seppo Utrianen, MD; Marko

Background—This study was designed to assess the epidemiology, characteristics, and outcome of cardiac sarcoidosis (CS) in Finland.

Methods and Results—We identified in retrospect all adult (>18 years of age) patients with manifest CS in Finland between 1988 and 2012. A total of 110 patients (71 women)

Conclusions

The number of patients with manifest CS seen annually in Finland increased >20-fold from 1988 to 2012, most likely as a result of improved diagnostic methods and heightened diagnostic activity. The majority of patients had clinically isolated



Nervous system:

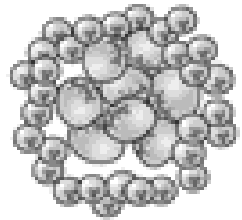
- Meningitis
- Encephalitis

Hilar and mediastinal lymph node granulomas

Lung granulomas

Liver:
• Granulomatous hepatitis

Histopathology
• Non-caseating granuloma



Lacrimal gland granuloma

Heart:

- Pericarditis
- Arrhythmias

Spleen:

- Splenomegaly

Lymph nodes

Joints:

- Arthritis

Skin:

- Erythema nodosum
- Subcutaneous nodules

Effects of Sarcoidosis

15% of cases affects the nervous system

25% of cases affects the eyes

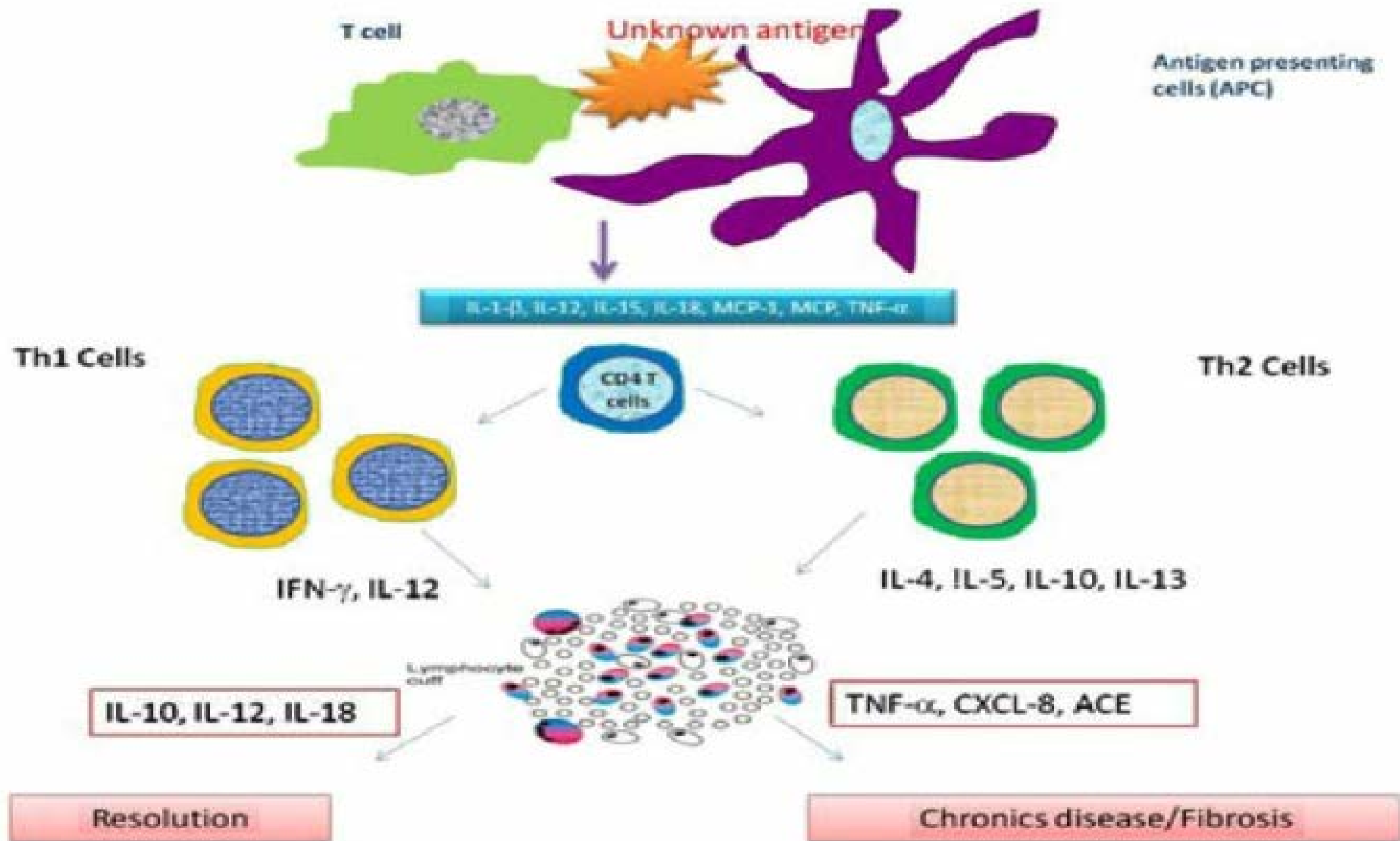
90% of cases affects the lungs and lymph nodes

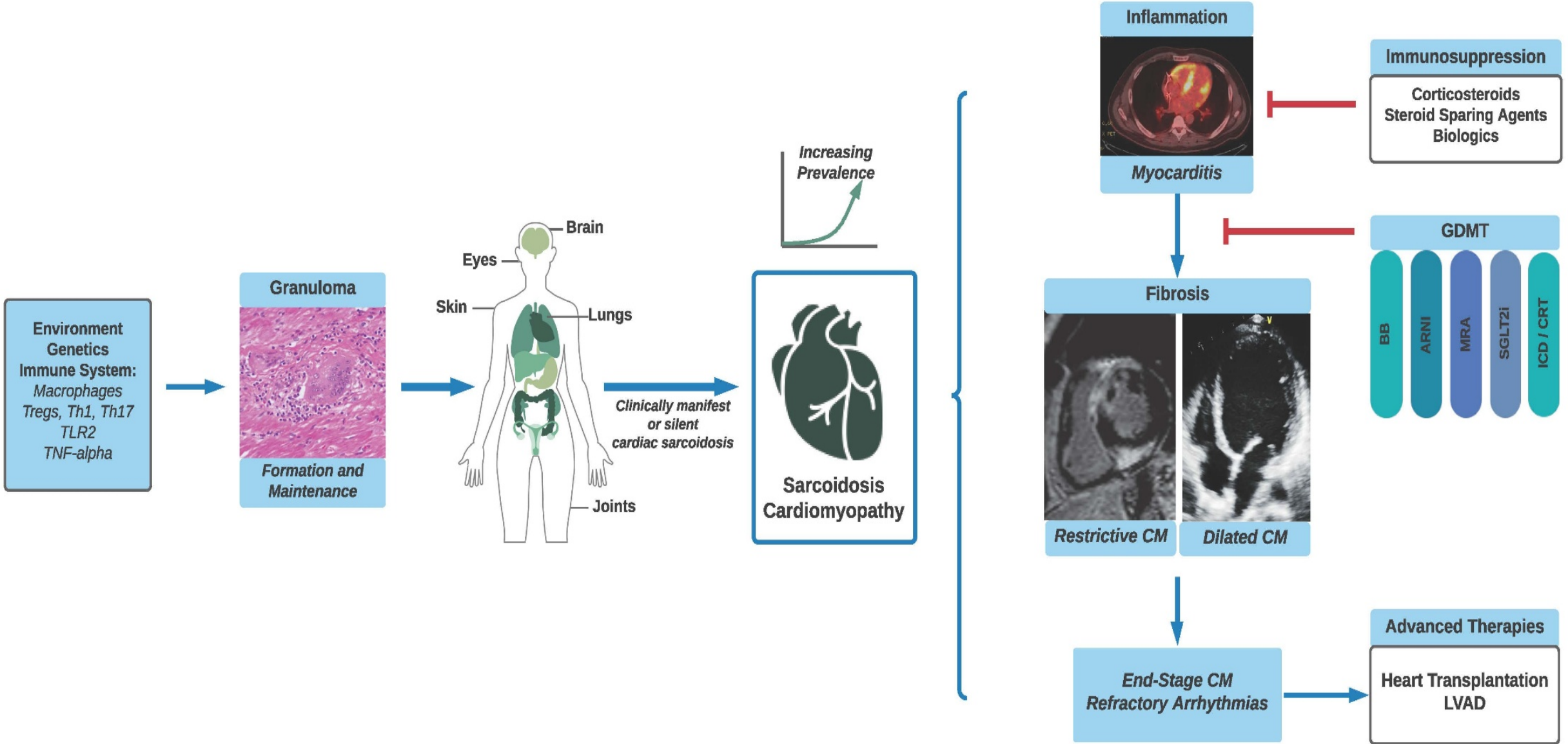
10% of cases affects the heart

65% of cases affects the liver

33% of cases affects the bones and joints

25% of cases affects the skin





Cardiac sarcoidosis occurs in up to a **THIRD** of all sarcoidosis patients

CARDIAC SARCOIDOSIS



DIZZINES AND/OR FAINTING SPELLS



IRREGULAR HEARTBEAT

Pounding or fluttering sensation or a 'skipping of beats'

CHEST PAIN



SHORTNESS OF BREATH

TESTS INCLUDE:

- Electrocardiogram (ECG)
- Echocardiogram (Echo)
- Magnetic Resonance Imaging (MRI)
- Nuclear Scans Holter monitor



SWELLING OF THE LEGS AND/OR ANKLES

In later stages



PULMONARY HYPERTENSION

This is when the heart is indirectly affected as a result of sarcoidosis in the lungs. This can affect up to 15% of patients with sarcoidosis

KLINIKA:

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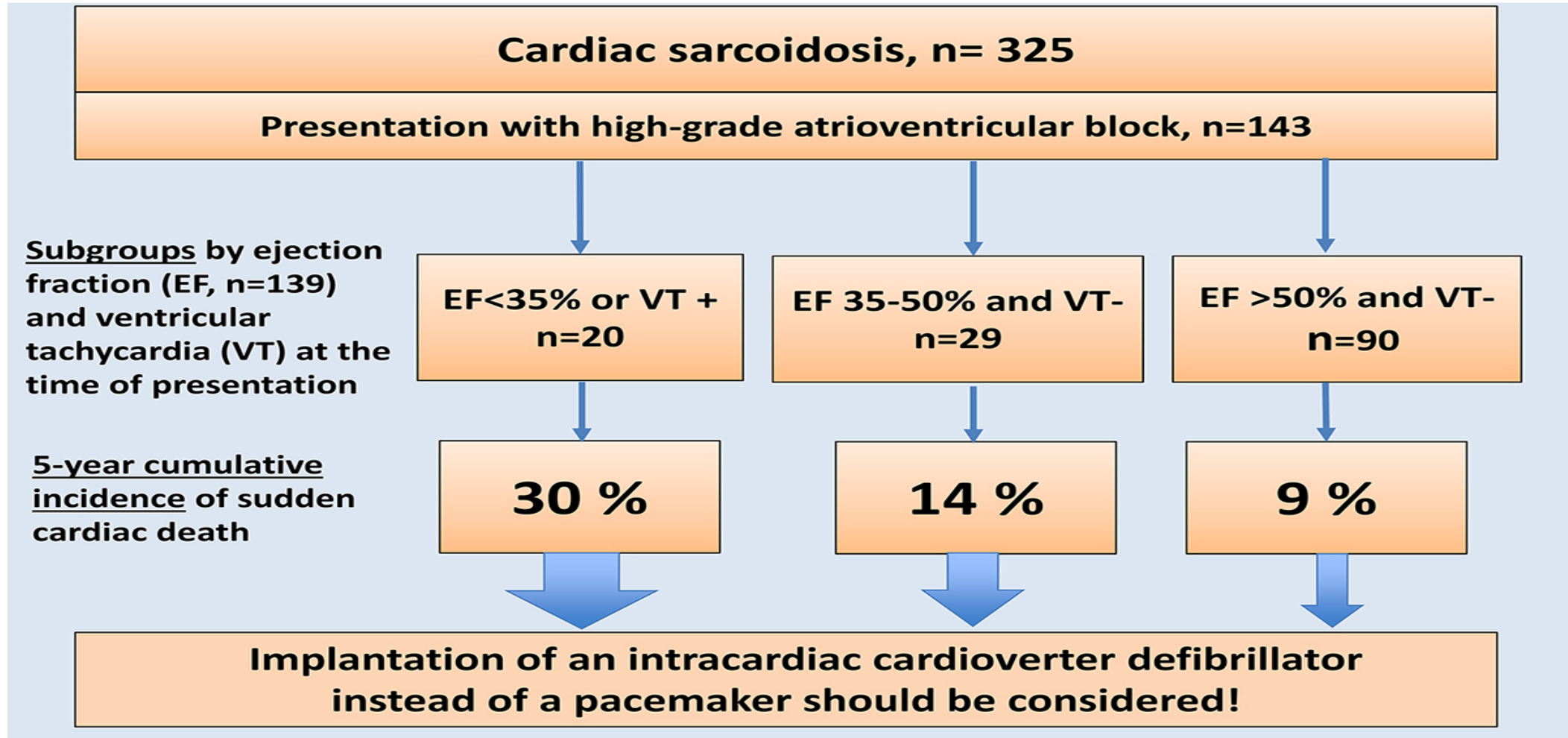
Cardiac Sarcoidosis: Clinical Manifestations, Imaging Characteristics, and Therapeutic Approach

Brian A. Houston¹ and Monica Mukherjee²

¹Division of Cardiology, Department of Medicine, Johns Hopkins Hospital, Baltimore, MD, USA. ²Division of Cardiology, Department of Medicine, Johns Hopkins Bayview Medical Center, Baltimore, MD, USA.

DIAGNOSTIC CATEGORY		Table 1. Clinical Manifestations of Cardiac Sarcoidosis.	
Histologic Diagnosis Group		CLINICAL MANIFESTATION	REPORTED PREVALENCE
Clinical Diagnosis Group*			
Major Clinical Criteria		Arrhythmias	
	AV block		26–62%
	Bundle Branch Block		12–61%
	Supraventricular Tachycardia		0–15%
	Ventricular Tachycardia		2–42%
	Sudden Cardiac Death		12–65%
Minor Clinical Criteria		Cardiomyopathy	
	Congestive heart failure <ul style="list-style-type: none"> • Left ventricular systolic failure • Heart failure with preserved ejection fraction or restrictive disease • Right ventricular failure secondary to pulmonary disease 		10–30%
	Pericardial		
	<ul style="list-style-type: none"> • Pericardial effusion detected by echo (common) • Pericarditis (rare) 		20%

Outcome of Cardiac Sarcoidosis Presenting with High-Grade Atrioventricular Block



Hanna-Kaisa Nordenswan. Circulation: Arrhythmia and Electrophysiology. Outcome of Cardiac Sarcoidosis Presenting With High-Grade Atrioventricular Block, Volume: 11, Issue: 8, DOI: (10.1161/CIRCEP.117.006145)

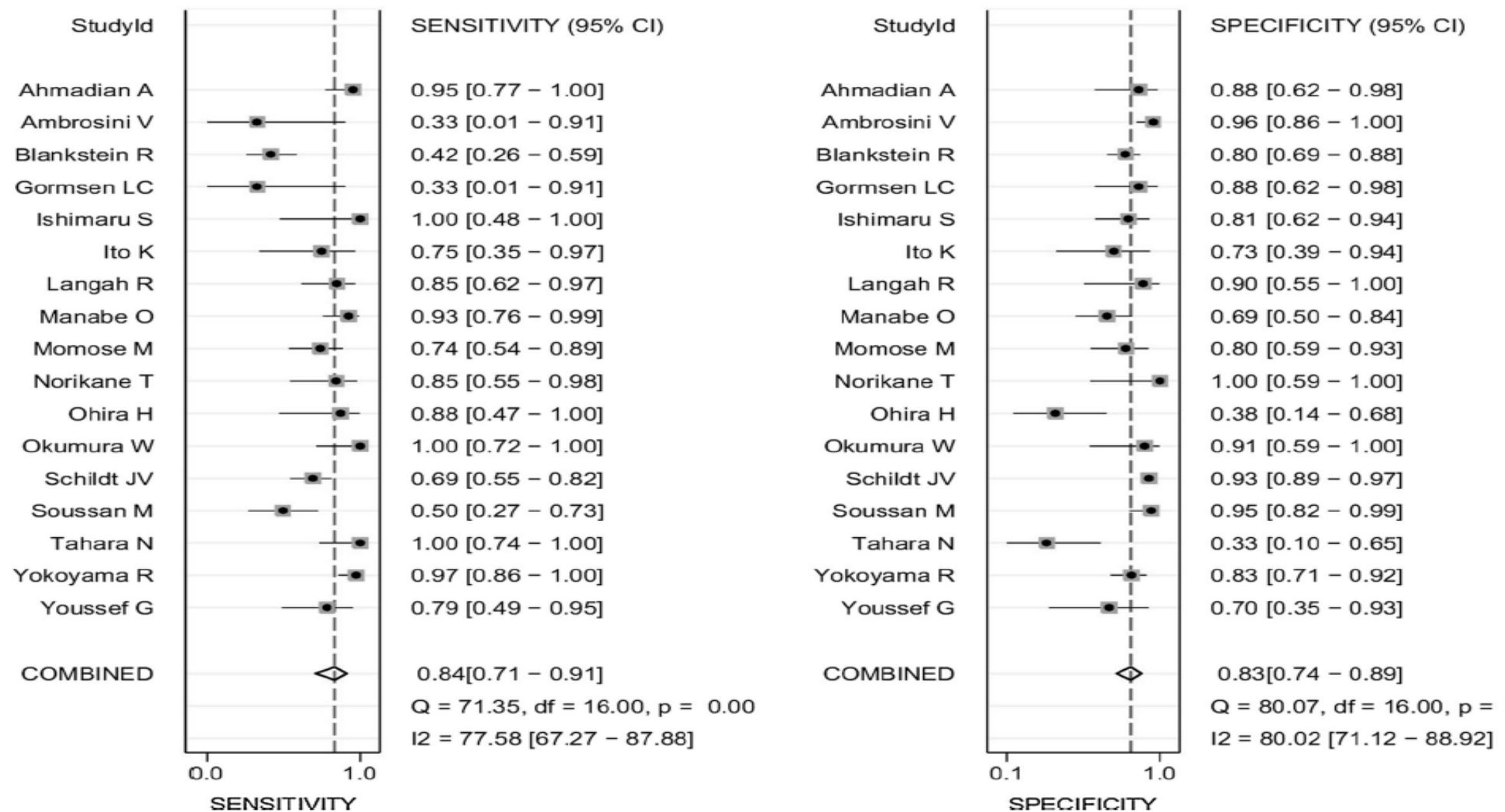
EXOKARDİOQRAM:



Figure 1. Echocardiogram, parasternal long axis view, of a 33-year-old patient with CS. Note the thinned, notched aneurysmal segment in the basal anteroseptal wall (red arrow).

- Sol mədəciyin atım fraksiyası
- Seqmentar divar hərəkət güsuru
- Sol və sağ mədəciyin 3 D strain
- Perikard
- Plevra
- Qulaqcıqlar
- Qapaqlar

Diagnostic performance of F-18 FDG PET for detection of cardiac sarcoidosis



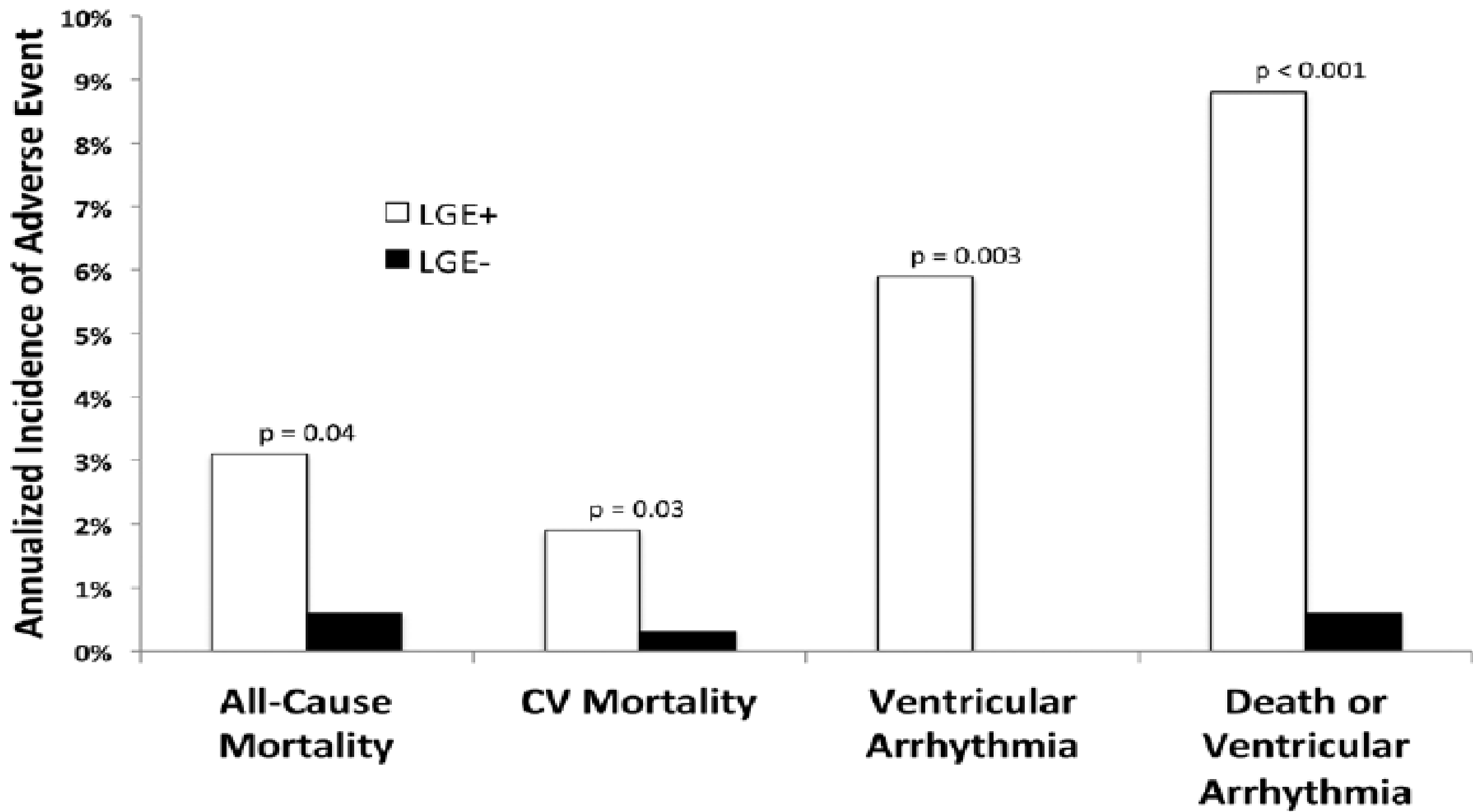
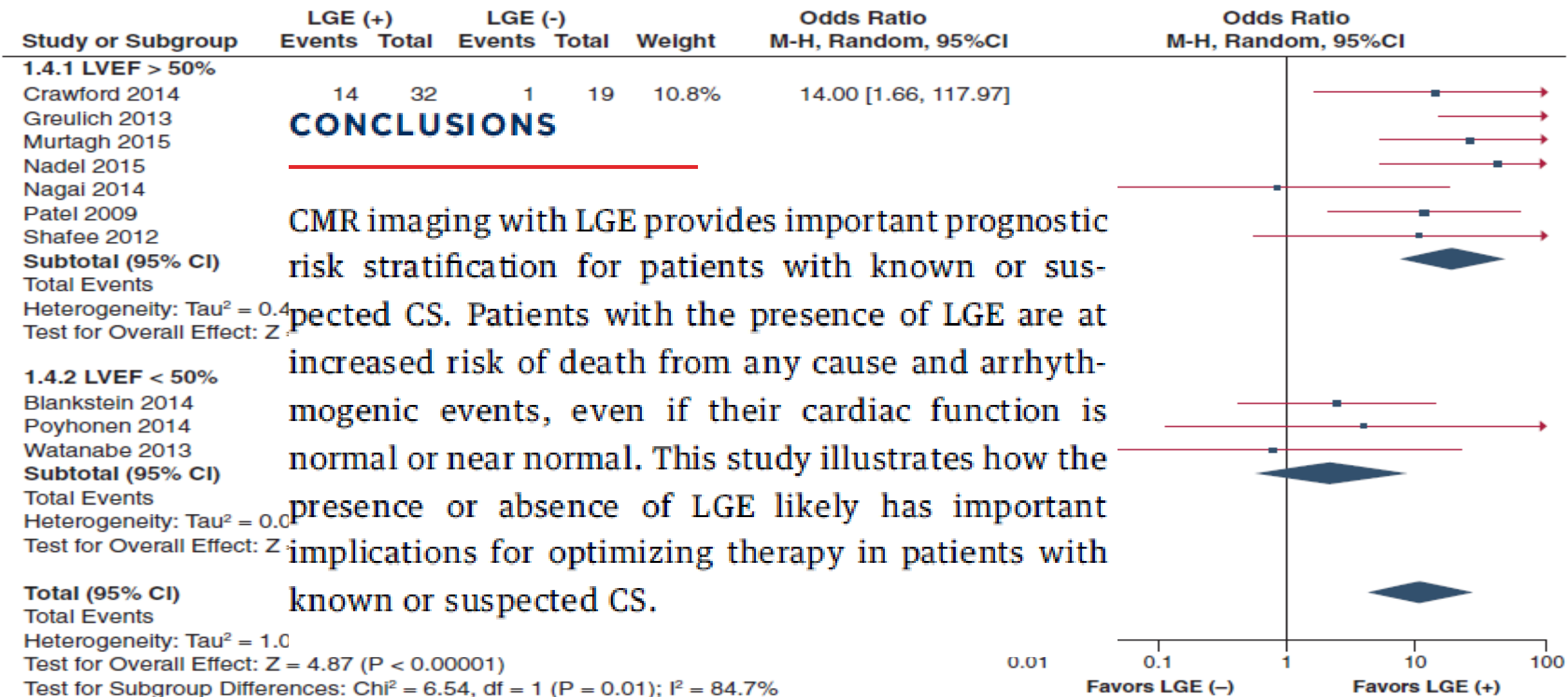


FIGURE 3 Forrest Plot for Composite Outcome



CONCLUSIONS

CMR imaging with LGE provides important prognostic risk stratification for patients with known or suspected CS. Patients with the presence of LGE are at increased risk of death from any cause and arrhythmogenic events, even if their cardiac function is normal or near normal. This study illustrates how the presence or absence of LGE likely has important implications for optimizing therapy in patients with known or suspected CS.

Clinical outcomes of patients with known or suspected cardiac sarcoid with the presence or absence of LGE on CMR. Composite outcome of all-cause mortality plus arrhythmogenic events stratified by LVEF; arrhythmogenic events defined as ventricular arrhythmias (ventricular tachycardia/ventricular fibrillation), sudden cardiac death, and appropriate implantable cardioverter-defibrillator discharge/aborted sudden cardiac death. CI = confidence interval; M-H = Mantel-Haenszel odds ratio; other abbreviations as in Figures 1 and 2.

Diagnostic Accuracy of Cardiac MRI versus FDG PET for Cardiac Sarcoidosis: A Systematic Review and Meta-Analysis

 Matthew Aitken,  Michael Vi
Matthew D. F. McInnes, 
 Paaladinesh Thavendiranatha



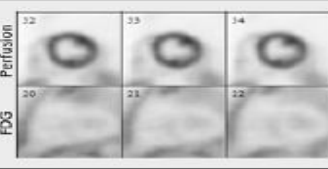
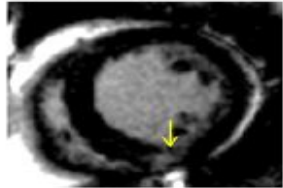

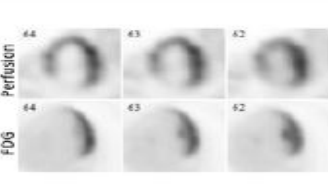
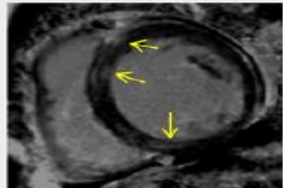

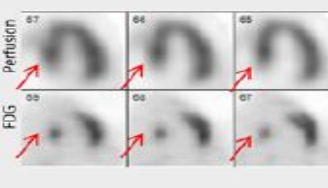
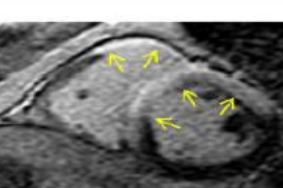

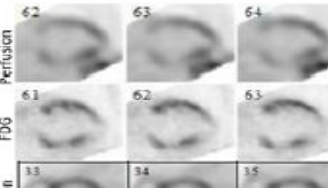
Results

Thirty-three studies were included (1997 patients, 687 with cardiac sarcoidosis); 17 studies evaluated cardiac MRI (1031 patients) and 26 evaluated FDG PET (1363 patients). Six studies directly compared cardiac MRI and PET in the same patients (303 patients). Cardiac MRI had higher sensitivity than FDG PET (95% vs 84%; $P = .002$), with no difference in specificity (85% vs 82%; $P = .85$). In a sensitivity analysis restricted to studies with direct comparison, point estimates were similar to those from the overall analysis: cardiac MRI and FDG PET had sensitivities of 92% and 81% and specificities of 72% and 82%, respectively. Covariate analysis demonstrated that sensitivity for FDG PET was highest with quantitative versus qualitative evaluation (93% vs 76%; $P = .01$), whereas sensitivity for MRI was highest with inclusion of T2 imaging (99% vs 88%; $P = .001$). Thirty studies were at risk of bias.

Conclusion

Cardiac MRI had higher sensitivity than fluorodeoxyglucose PET for diagnosis of cardiac sarcoidosis but similar specificity. Limitations, including risk of bias and few studies with direct comparison, necessitate additional study.

ÜRƏK-MRT VƏ PET

Likelihood Probability	MRI Likelihood	MRI Example	MRI Example Illustrated	PET Likelihood	PET Example
No CS (<10%)	<ul style="list-style-type: none"> -No LGE - LGE present but clear alternative diagnosis (e.g ARVC) 			- No FDG uptake <u>and</u> no perfusion defect	
Possible CS (50-90%)	<ul style="list-style-type: none"> - One focal areas of LGE but alternative diagnosis was more likely. (e. g. Pulmonary hypertension) 			<ul style="list-style-type: none"> - no FDG uptake but a small perfusion defect. - Non-specific FDG uptake and no perfusion defects.* 	
Probable CS (50-90%)	<ul style="list-style-type: none"> - Multifocal LGE in a pattern that is likely consistent with CS but cannot rule out other diagnosis (e. g. myocarditis) 			<ul style="list-style-type: none"> - Multiple non-contiguous areas of scar with no FDG uptake. - Focal or focal on diffuse FDG uptake associated with resting perfusion defect. 	
Highly Probable (>90%)	<ul style="list-style-type: none"> - Multifocal LGE in a pattern strongly consistent with CS with no alternative diagnosis. - The following features were used to identify high likelihood: <ul style="list-style-type: none"> → Intense signal of LGE. → Prominent involvement of insertion points with direct and contiguous extension across the septum into RV. ("hook sign") 			<ul style="list-style-type: none"> - Multiple areas of focal FDG uptake <u>AND</u> extra cardiac FDG. - Multiple areas of both FDG uptake and perfusion defect. 	

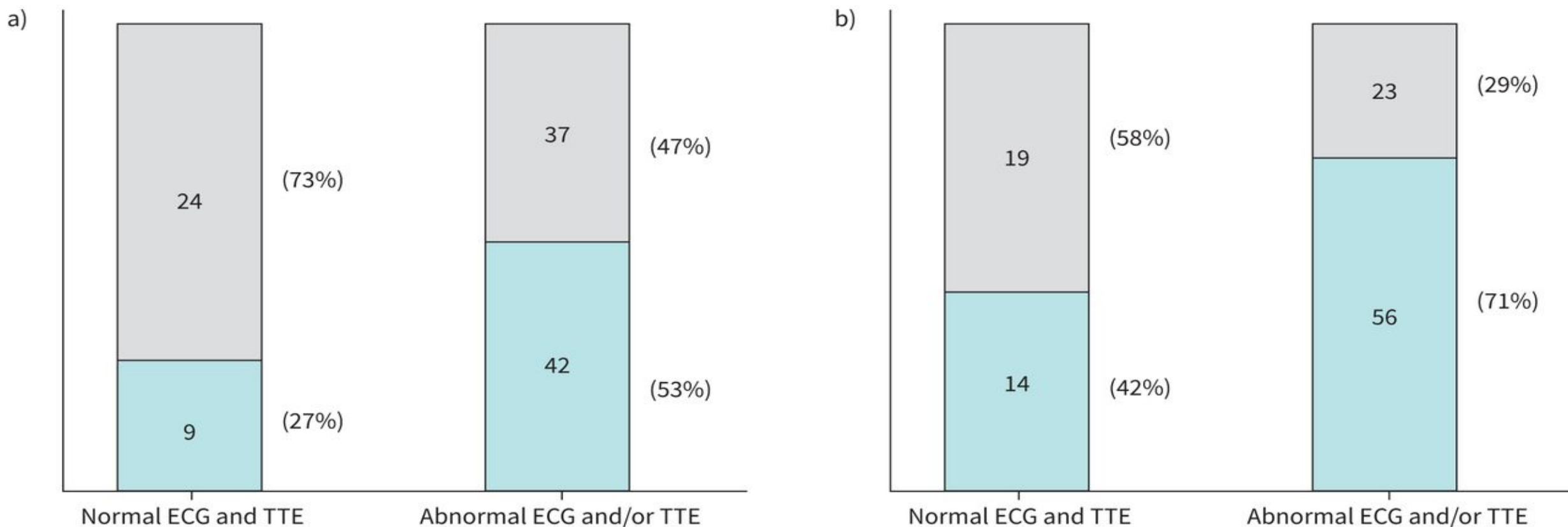


Underdiagnosis of cardiac sarcoidosis by ECG and echocardiography in cases of extracardiac sarcoidosis

Hiroshi Ohira, Takahiro Sato, Osamu Manabe, Noriko Oyama-Manabe, Akiko Hayashishita, Toshitaka Nakaya, Junichi Nakamura, Naoko Suzuki, Ayako Sugimoto, Sho Furuya, Satonori Tsuneta, Taku Watanabe, Ichizo Tsujino, Satoshi Konno

ERJ Open Research 2022 8: 00516-2021; DOI: 10.1183/23120541.00516-2021

Positive Negative



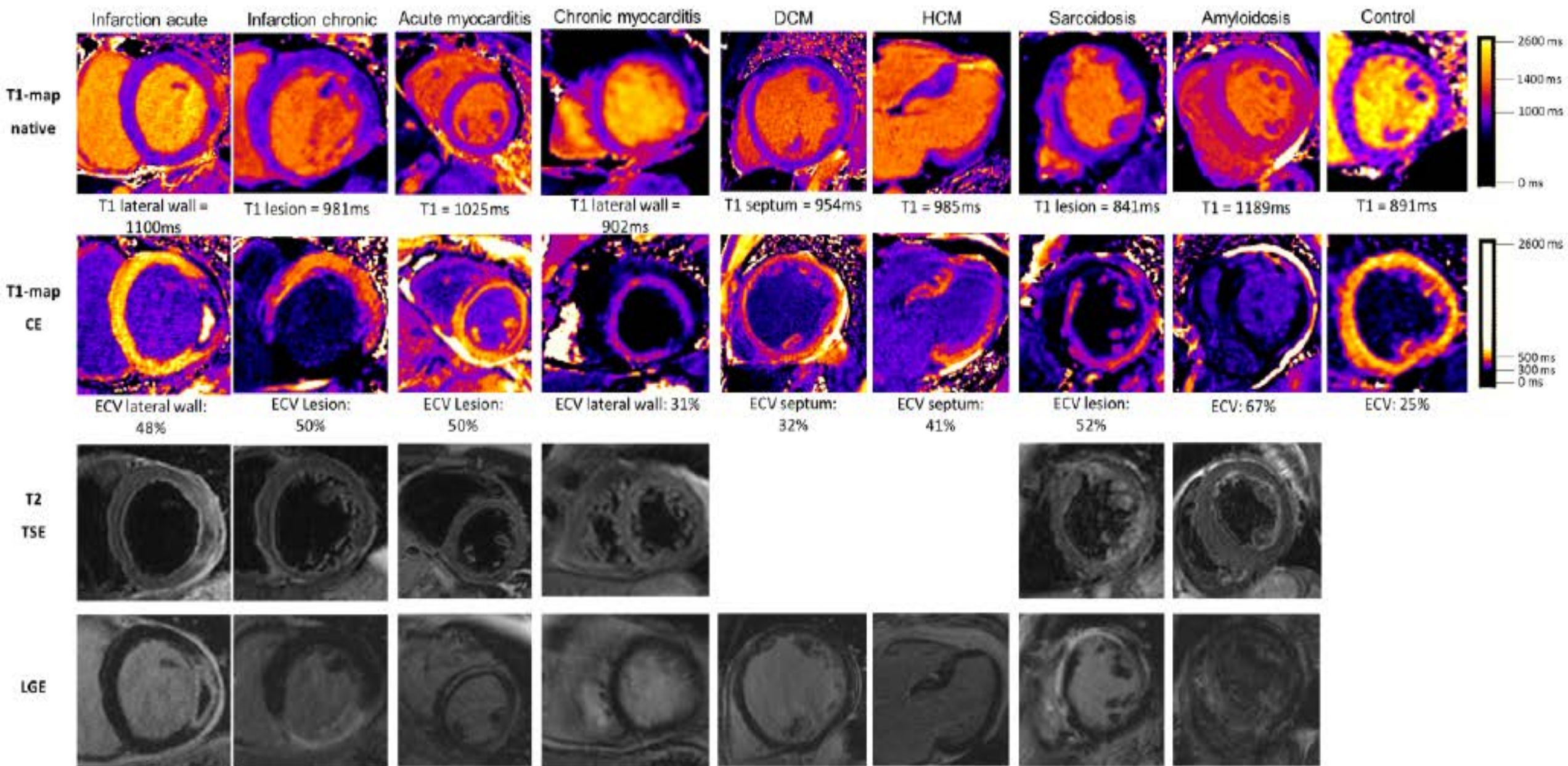
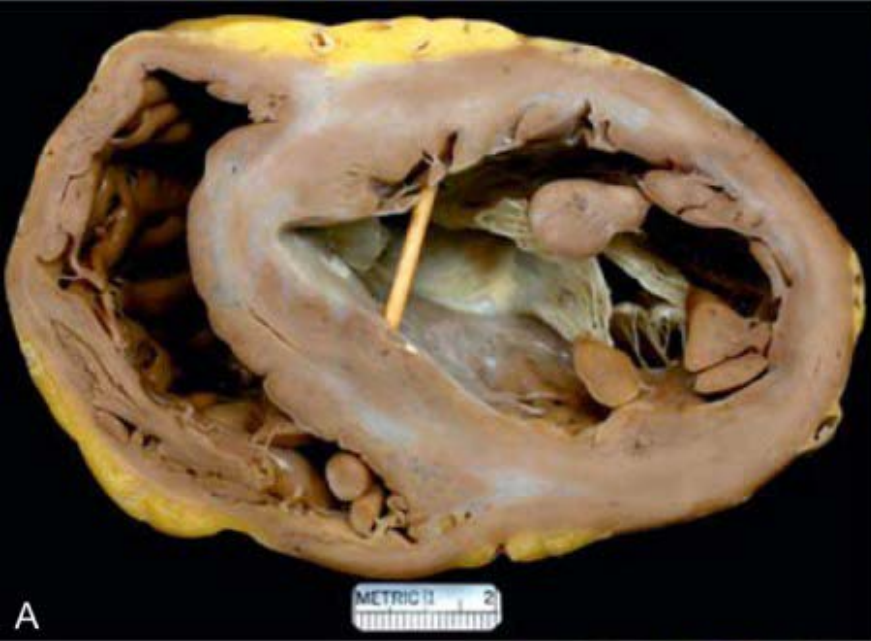
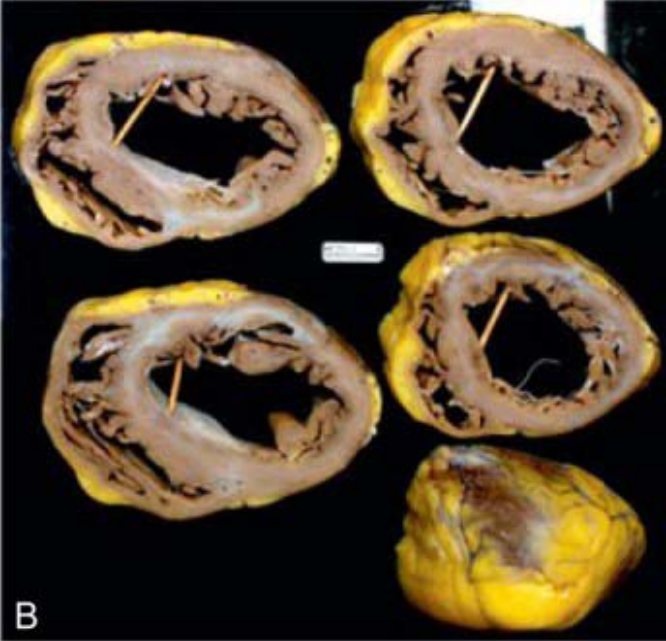


Figure 1: Common cardiac pathologies in native and contrast enhanced T1-mapping, T2w TSE and LGE.



A

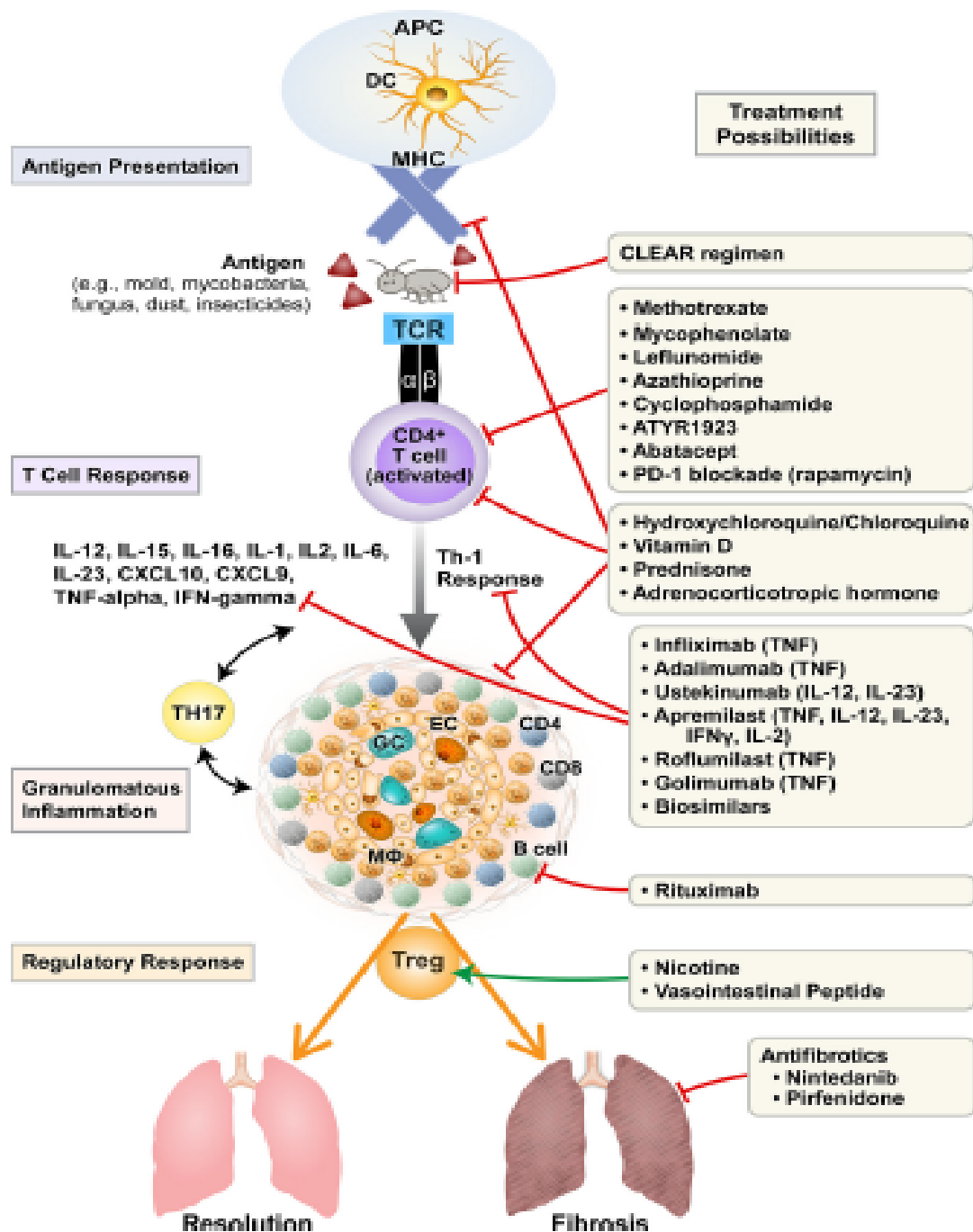


B



C

BIOPSIYA:



Etioloqiya və Patoqenez əsaslı müalicə

Müalicə Prinsipləri:

PRIMER

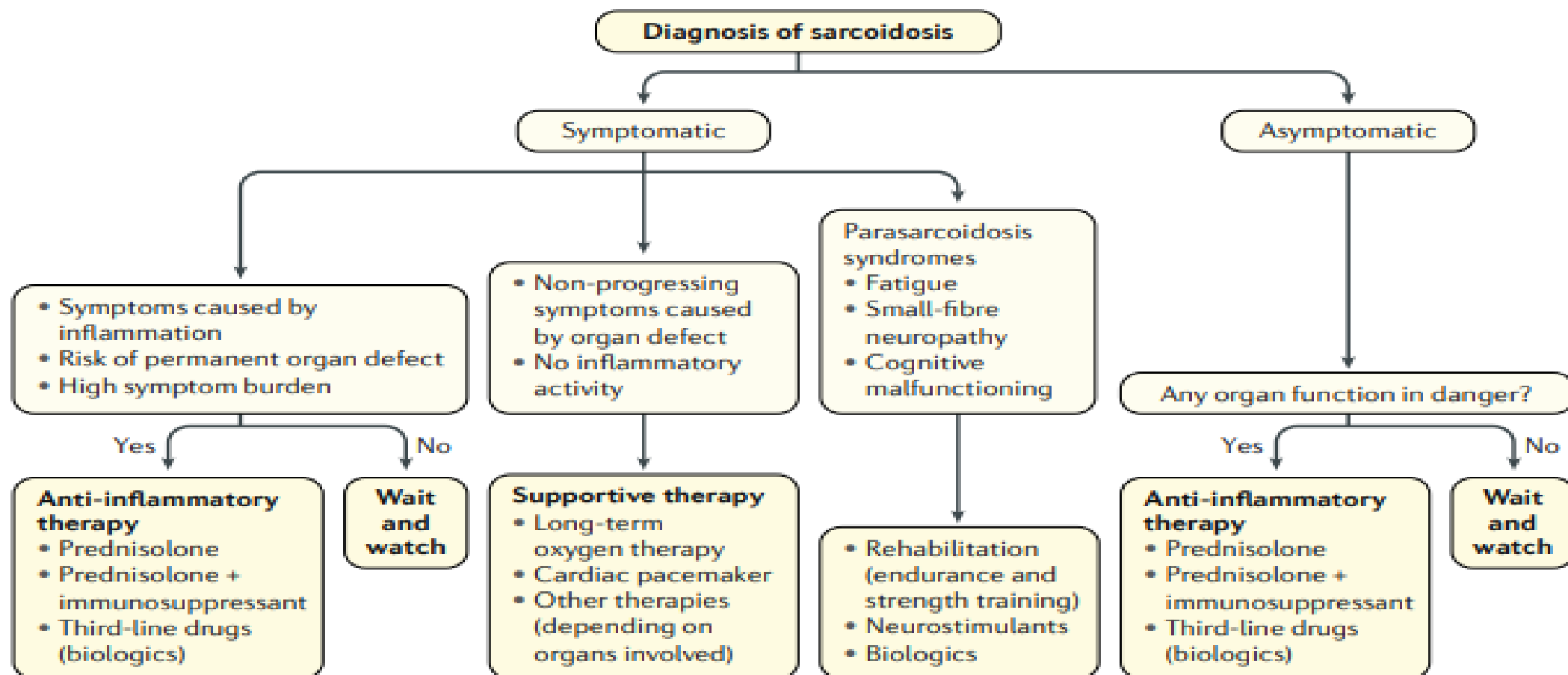


TABLE 1 | Common therapeutics for treatment of sarcoidosis*.

	Drug name	Suggested dose range	Special treatment issues/monitoring
First-Line Agents	Corticosteroids (Prednisone)	20–40 mg/day initial Dose, tapered to 7.5–15 mg/day	Bone density Eye exams (glaucoma and cataracts) Body Mass Index
Second-Line Agents	Methotrexate	7.5–25 mg/week orally or subcutaneously	Concurrent need for folic acid. Liver function, kidney function, CBC. Can cause hepatotoxicity, GI distress, pneumonitis, mouth ulcers, bone marrow suppression.
	Hydroxychloroquine	200–400 mg/day	Eye exams for retinopathy. Rarely associated with QT elongation (consider drug interactions).
	Leflunomide	10–20 mg/day	Liver function, kidney function, CBC. Can cause neuropathy, hepatotoxicity, GI distress, pneumonitis, bone marrow suppression. In cases of toxicity, can clear more urgently with cholestyramine.
	Azathioprine	50–200 mg/day	Liver function, kidney function, CBC. Consider TPMT level. Can cause hepatotoxicity, GI distress, hypersensitivity reaction, bone marrow suppression.
	Mycophenolate	500–3,000 mg/day	Liver function, kidney function, CBC. Associated with GI distress, bone marrow suppression. Enteric coated option available (different dose range).
Third-Line Agents	Infliximab	3–5 mg/kg intravenously at weeks 0, 2, and every 4–8 weeks thereafter	Tuberculosis Testing Caution in heart failure. Allergic reactions possible with injections. Associated with demyelination syndrome, malignancy, and sarcoid-like reactions.
	Adalimumab	40 mg subcutaneous every 1–2 weeks	Similar precautions and adverse reactions as infliximab.

Kim təqib etsin?

Cardiac Sarcoidosis Clinic Overview

At Mayo Clinic, people with symptoms that indicate they might have cardiac sarcoidosis receive diagnosis and treatment from a team of experts in the Cardiac Sarcoidosis Clinic. These services are available at Mayo Clinic's campuses in Arizona, Florida and Minnesota.

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Sarcoidosis

Sarcoidosis is a condition that causes lumps or nodules (granulomas) to form in your lungs, lymph nodes, skin, eyes and other parts of your body. Symptoms include cough, shortness of breath, tender sores on your shins, eye pain and redness. Many cases go away on their own or with treatment, but sometimes it becomes a chronic condition.

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1. Sarkoidoz, geniş yayılmış olmasada, ÜÇ və ciddi Ritm pozulmasının səbəblərindən biridir!
2. Diaqnostik metodlar təkmirləşdikcə xəstə populasiası artır!
3. Erkən diaqnostika, erkən müalicə deməkdir!
4. Exokardioqrafiya mütləkdir!
5. PET və ya MRT, seçim sizindir!
6. Zamanla ayaqlaşmaq sizin əlinizdədir!

W

WHO

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ARE

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YOU

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DİQQETİNİZƏ GÖRƏ TƏŞƏKKÜRLƏR!!!
