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18<sup>èmes</sup> journées françaises  
pratiques de rythmologie  
& de stimulation cardiaque

5-6 DÉCEMBRE 2024

HOTEL VILLA MASSALIA,  
MARSEILLE | FRANCE

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# Sarcoïdose cardiaque Focus, recommandations et consensus

Dr Rim EL BOUAZZAOUI

05/12/2024

Aucun conflit d'intérêt

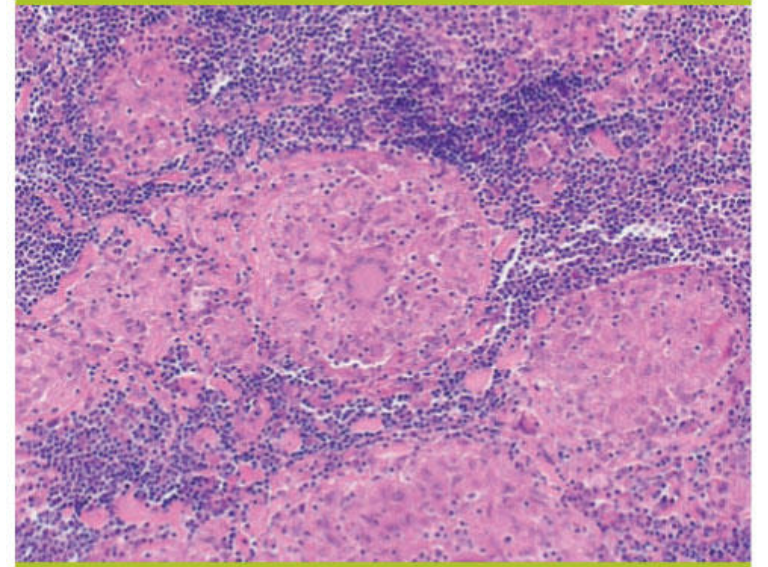
# Atteinte cardiaque de la sarcoïdose

La sarcoïdose est une pathologie infiltrative d'étiologie inconnue

L'atteinte cardiaque est secondaire à l'inflammation du myocarde liée à la présence de **granulomes épithélioïdes et géantocellulaires sans nécrose caséuse**

Incidence aux USA : 35.2 cas pour 100 000 habitants

Environ 20% des patients avec sarcoïdose systémique, adressés pour imagerie ont une atteinte cardiaque. 5% ont une atteinte clinique patente.



# Atteinte cardiaque de la sarcoïdose

Original Article

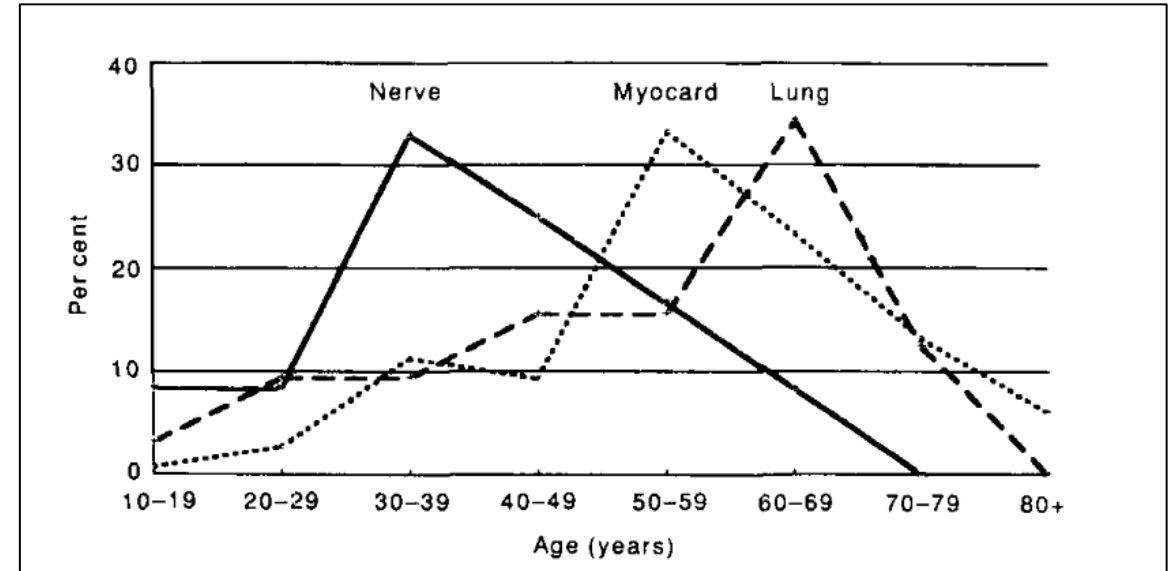
## Pathological studies on sarcoidosis autopsy. I. Epidemiological features of 320 cases in Japan

Kazuro Iwai,<sup>1</sup> Teruo Tachibana,<sup>2</sup> Tamiko Takemura,<sup>3</sup> Yasuo Matsui,<sup>3</sup> Masanori Kitaichi<sup>4</sup> and Yoshinori Kawabata<sup>1</sup>

*Acta Pathologica Japonica* 1993; **43**: 372–376

**Table 3** Causes of death in sarcoidosis autopsy

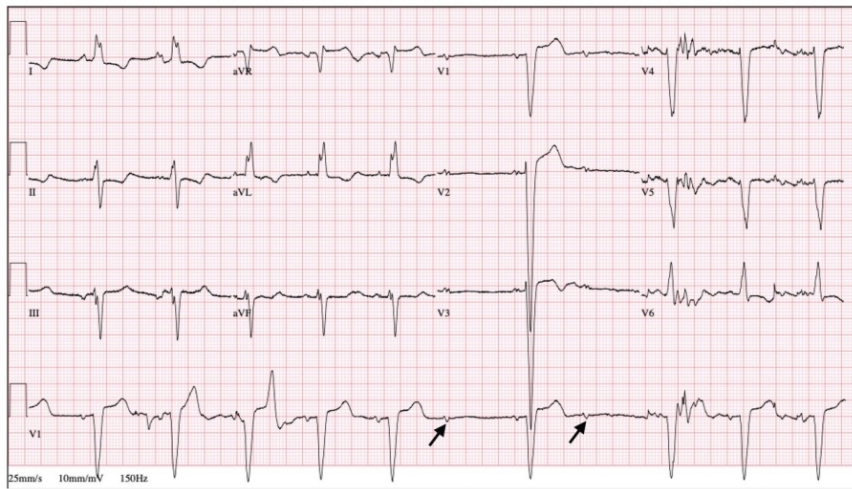
Cause of death	<i>n</i>	%
Non-sarcoidosis death	123	38.4
Sarcoidosis death	194	60.6
Related to cardiac sarcoidosis	150	46.9
Related to pulmonary sarcoidosis	32*	10.0
Related to nervous sarcoidosis	12†	3.8
Unknown	3	1.0
Total	320	100.0





# Atteinte cardiaque de la sarcoïdose

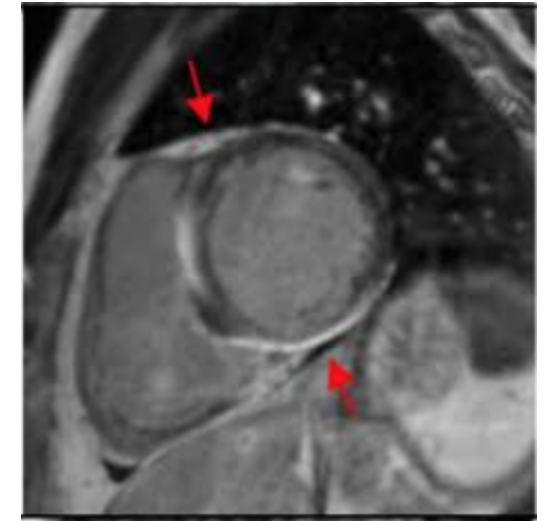
3 grands modes de présentation



troubles conductifs



Arythmies ventriculaires



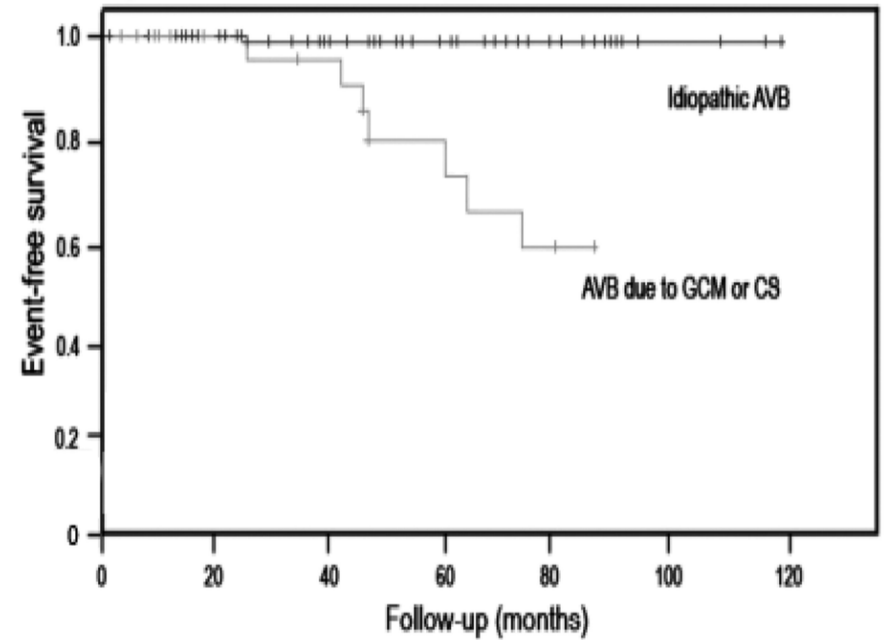
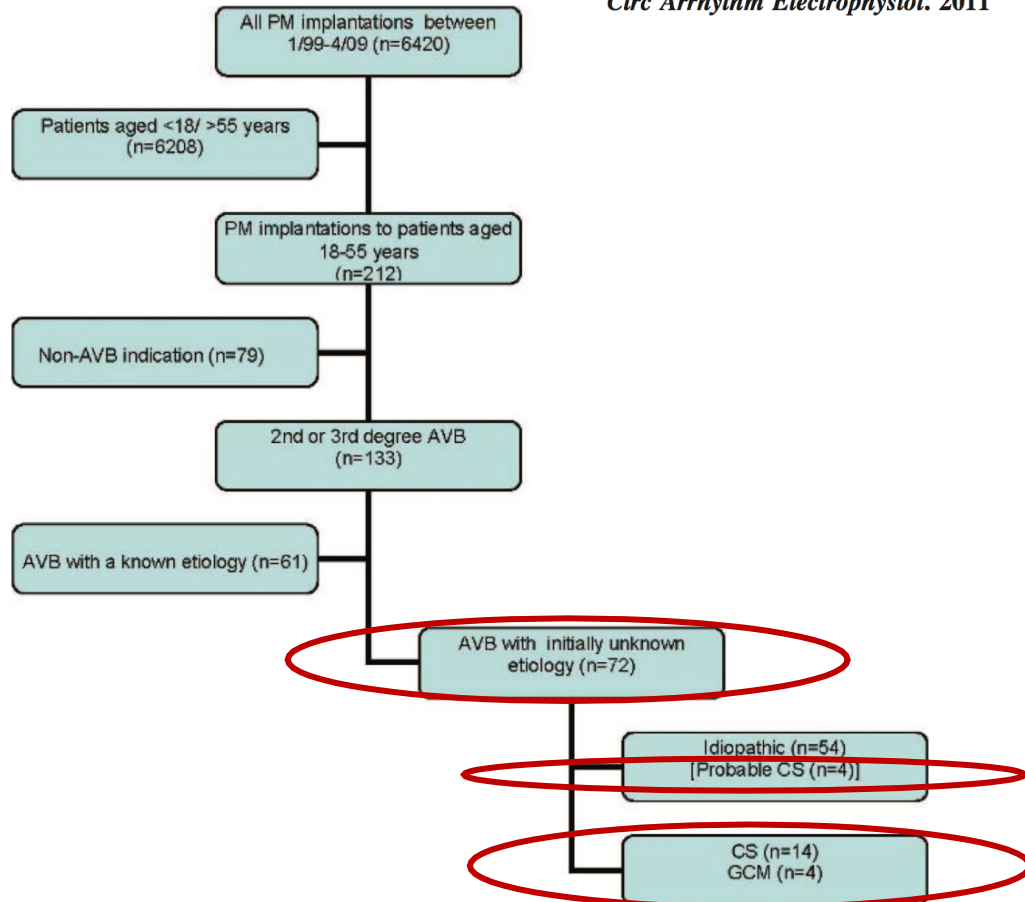
Dysfonction ventriculaire gauche

# Atteinte cardiaque de la sarcoïdose

## Cardiac Sarcoidosis and Giant Cell Myocarditis as Causes of Atrioventricular Block in Young and Middle-Aged Adults

Riina Kandolin, MD; Jukka Lehtonen, MD, PhD; Markku Kupari, MD, PhD

*Circ Arrhythm Electrophysiol.* 2011



Event	Cause of AVB		
	Idiopathic (n=54)	Confirmed CS/GCM (n=18)	Confirmed or Probable CS/GCM (n=22)
Ventricular fibrillation	0	4 (22)‡	4 (18)
Ventricular tachycardia			
Nonsustained*	9 (17)	4 (22)	6 (27)
Sustained†	1 (2)	6 (33)	7 (32)
Cardiac death	0	4 (22)	4 (18)
Cardiac transplantation	0	3 (17)	3 (14)
MACE	1 (2)	7 (39)	8 (36)

# Atteinte cardiaque de la sarcoïdose

## Ventricular tachyarrhythmia as a primary presentation of sarcoidosis

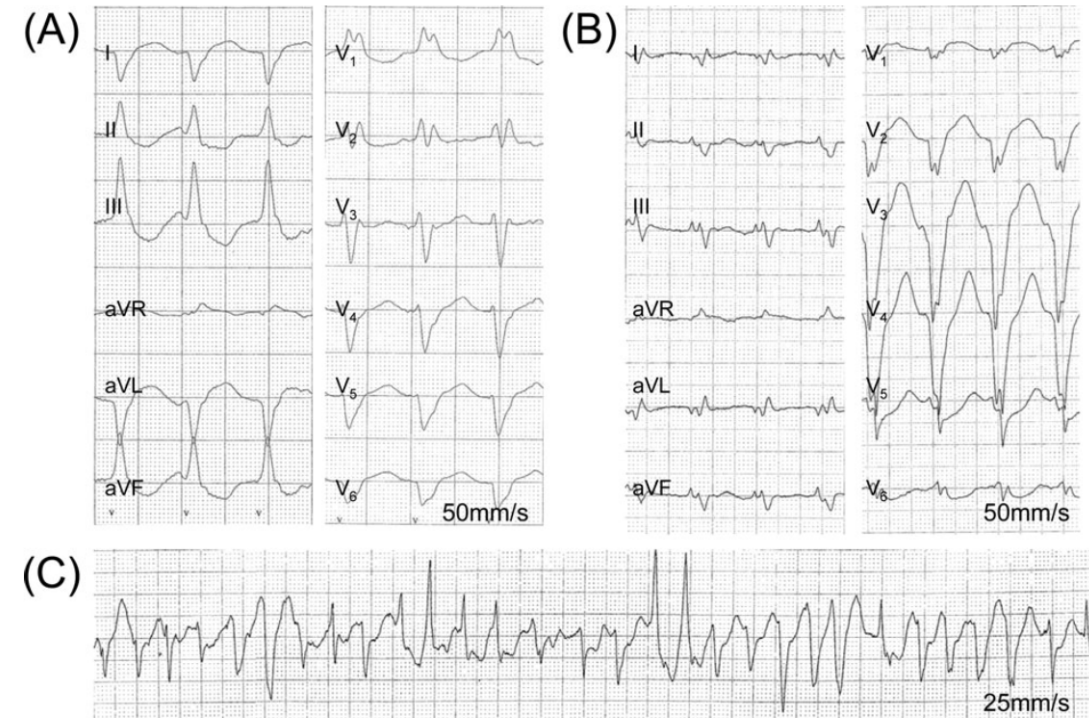
Uusimaa et al. Europace 2008

**Table 1** Clinical characteristics of the patients

Patient no.	Age	Sex	Symptoms	Other diseases	ESR (<16 mm/h)	ACE (9–65 U/L)	Hypercalcemia and/or hypercalciuria	Chest X-ray	Findings in endomyocardial biopsy
1	61	Male	Syncope	Hypertension, diabetes	5	31	Yes	Normal	Granulomas
2	43	Female	Presyncope, fatigue	None	8	32	No	Normal	Granulomas
3	50	Male	Palpitations	None	6	43	Yes	Normal	Fat <sup>a</sup>
4	59	Male	Cardiac arrest	Myeloma	48	18	No	Normal	Granulomas
5	52	Female	Palpitations, dyspnoea	None	14	24	Yes	Normal	Granulomas
6	58	Female	Presyncope	None	5	52	No	Normal	Granulomas
7	68	Female	Syncope	Hypertension, breast cancer	11	28	Yes	Normal	Granulomas
8	53	Female	Palpitations	None	3	36	No	Normal	Granulomas
9	33	Male	Palpitations, chest pain	None	3	24	No	Normal	Granulomas

ESR, erythrocyte sedimentation rate; ACE, angiotensin-converting enzyme.

<sup>a</sup>Endomyocardial biopsy contained fat suggesting arrhythmogenic right ventricular dysplasia but biopsy from mediastinal lymph nodes revealed granulomas years afterwards.



FU 50 +/- 34 months, 5 patients underwent appropriate ICD therapies  
 2 patients developed incessant VT treated by catheter ablation.  
 1 patient was referred for heart transplantation.



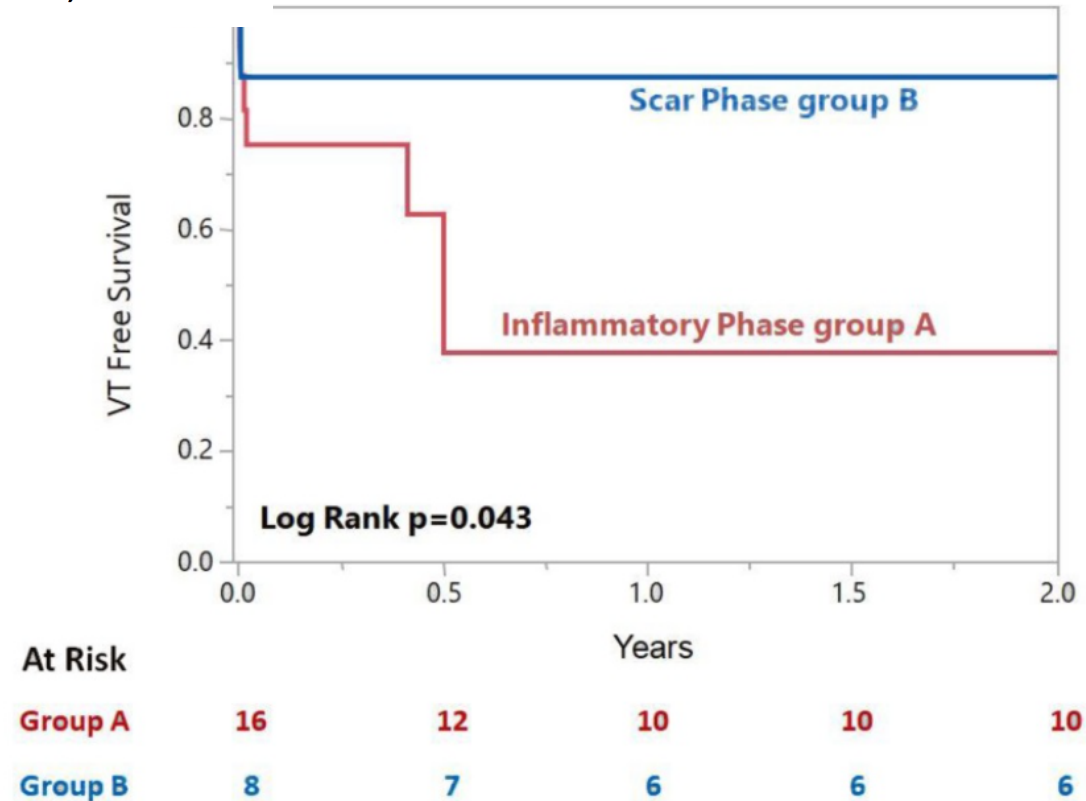
# Ablation de TV chez les patients avec sarcoïdose cardiaque

Impact of the Inflammation on the outcomes of catheter ablation of drug refractory ventricular tachycardia in Cardiac Sarcoidosis.

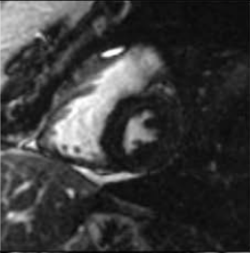
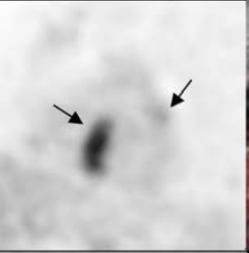
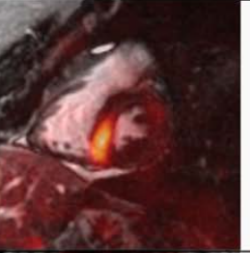
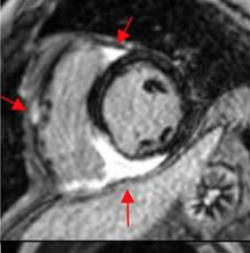
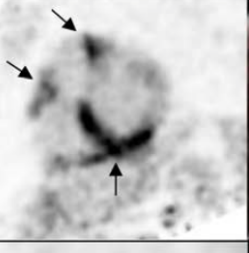
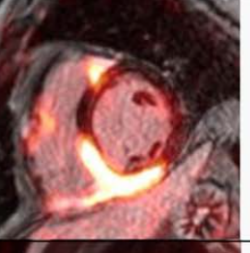
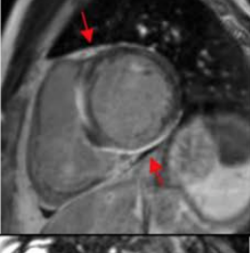
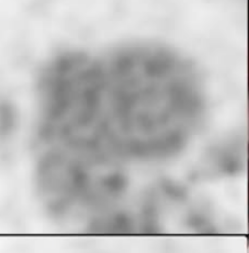
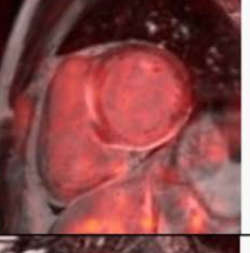
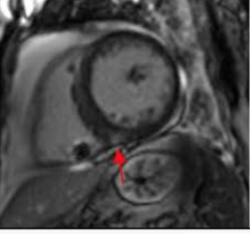
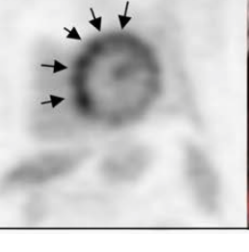
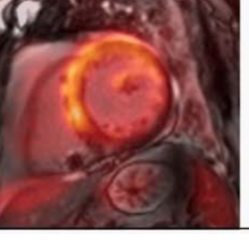
Short title: Catheter ablation of ventricular tachycardia in cardiac sarcoidosis

Daljeet kaur, MD\*; Henri Roukoz, MD†; Mandar Shah, MD\*; Sachin Yalagudri, MD\*;  
Ulhas Pandurangi, MD #; Sridevi Chennapragada, MD\* ; Narasimhan. C, MD\*

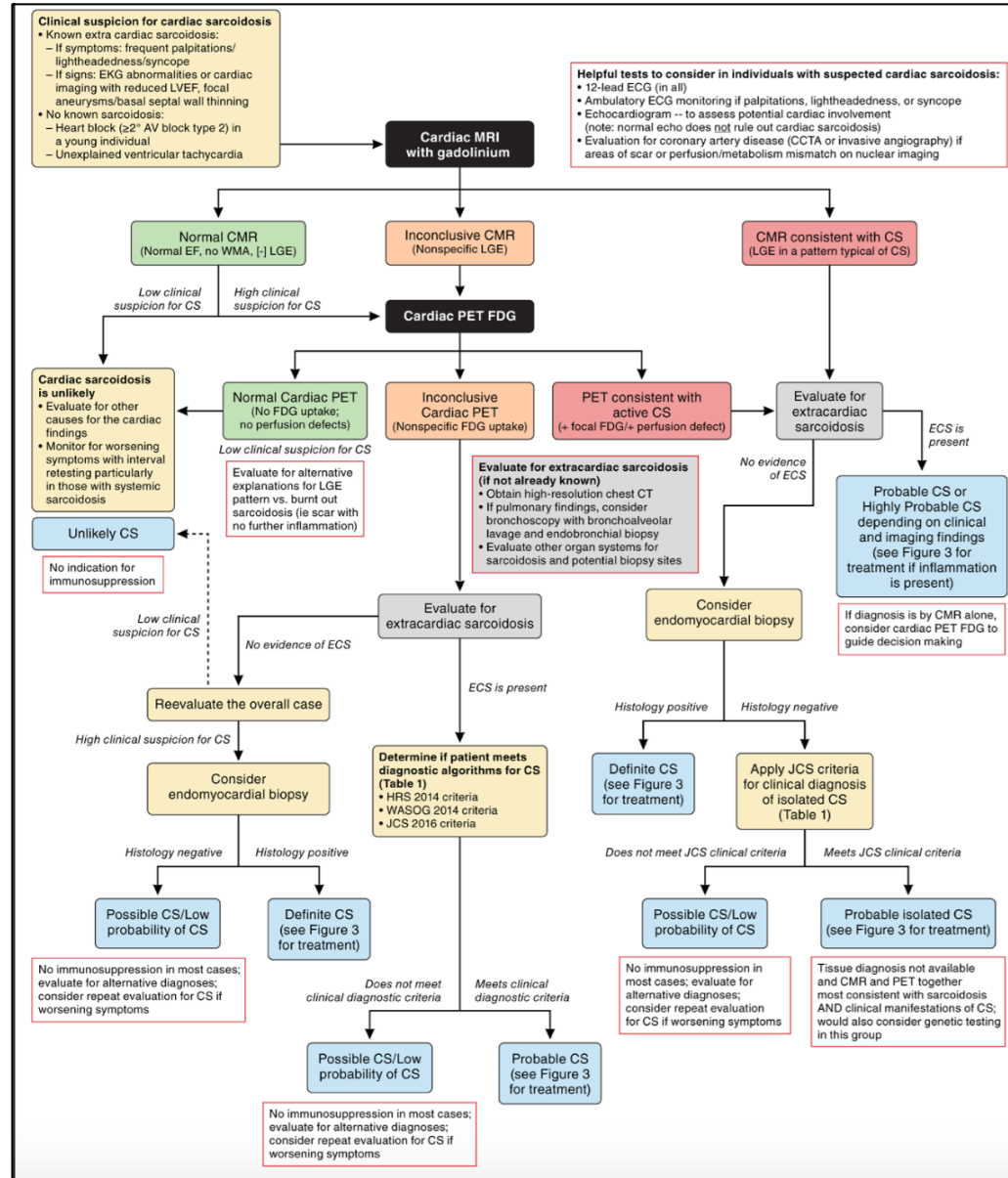
J Cardiovasc Electrophysiol . 2020



# Atteinte cardiaque de la sarcoïdose – intérêt de l'IRM et du TEP scanner

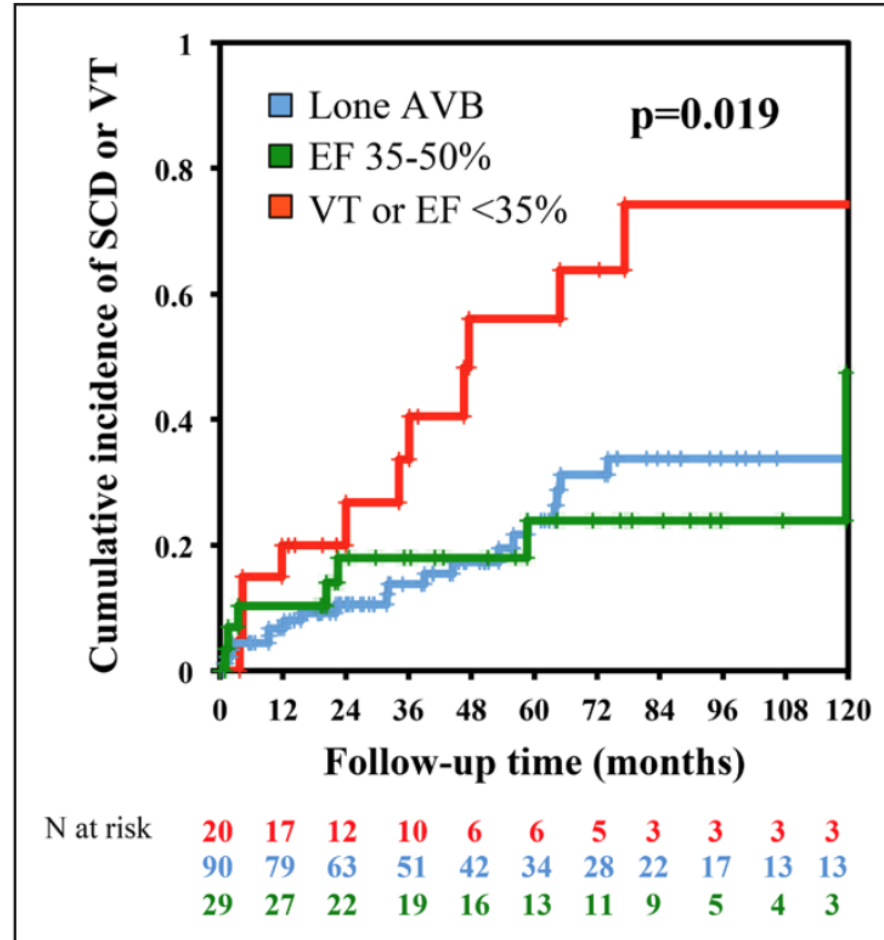
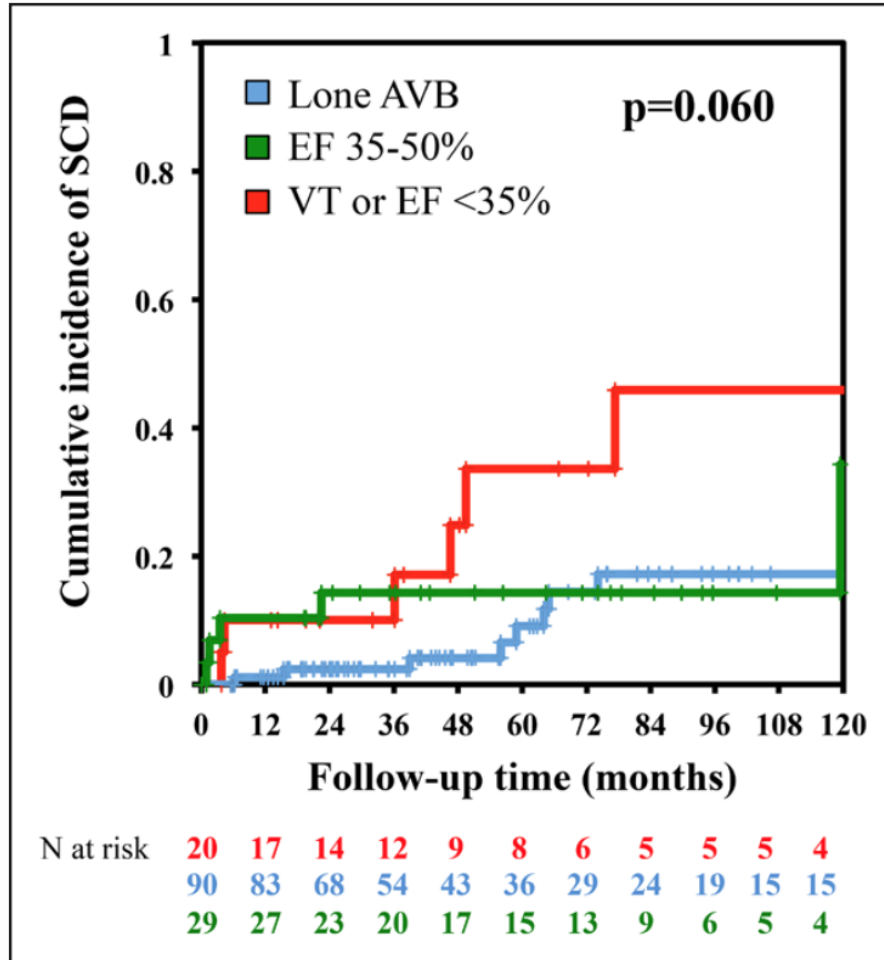
Phenotypes	CMR	FDG PET	PET-MR	Typical Presentation
<b>A</b> Focal septal FDG uptake with or without corresponding LGE				Heart block
<b>B</b> Multifocal LGE and FDG uptake in a pattern consistent with cardiac sarcoidosis				Heart block Ventricular arrhythmias LV systolic dysfunction
<b>C</b> Multifocal LGE in a pattern consistent with cardiac sarcoidosis without FDG uptake				Ventricular arrhythmias LV systolic dysfunction
<b>D</b> LGE or FDG uptake in a pattern <u>NOT</u> consistent with cardiac sarcoidosis				Miscellaneous, including other presentations, such as palpitations, dyspnea, dizziness, ventricular ectopy

# Atteinte cardiaque de la sarcoïdose et difficultés diagnostiques



# Outcome of Cardiac Sarcoidosis Presenting With High-Grade Atrioventricular Block

Nordenswan circ EP 2018

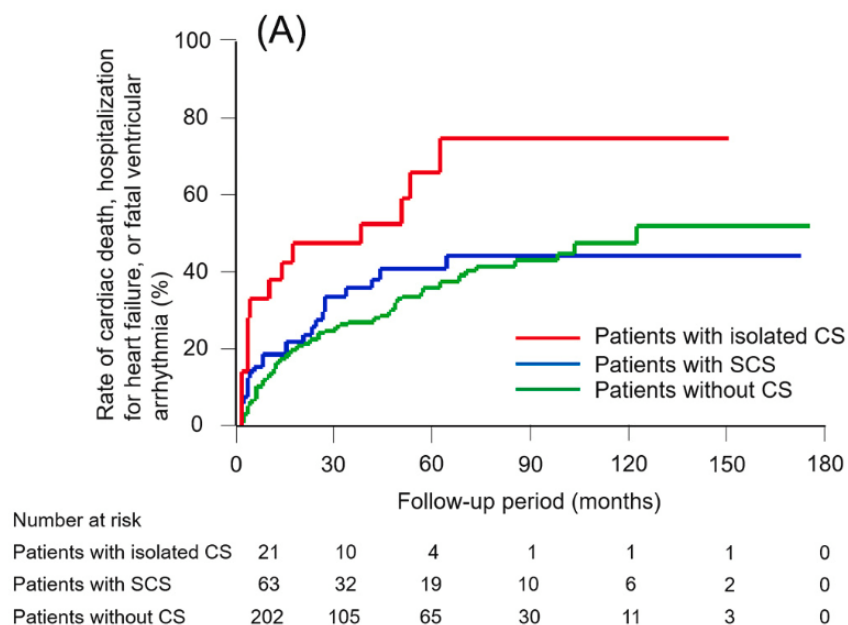




# Risque rythmique de la sarcoïdose cardiaque

Clinical outcomes of patients with isolated cardiac sarcoidosis confirmed by clinical diagnostic criteria

Yoichi Takaya <sup>\*,1</sup>, Kazufumi Nakamura <sup>1</sup>, Nobuhiro Nishii <sup>1</sup>, Hiroshi Ito <sup>1</sup>



Factors related to cardiac death, hospitalization for heart failure, or fatal ventricular arrhythmia.

Variables	Univariate analysis		Multivariate analysis	
	Hazard ratio (95% confidence interval)	<i>p</i>	Hazard ratio (95% confidence interval)	<i>p</i>
Age > 60 years	2.15 (1.43–3.29)	<0.01	2.23 (1.47–3.46)	<0.01
Male	1.15 (0.77–1.72)	0.50	1.09 (0.72–1.67)	0.68
New York Heart Association functional class III or IV	1.53 (1.03–2.28)	0.03	1.55 (1.02–2.38)	0.04
Left ventricular ejection fraction <35%	1.42 (0.96–2.15)	0.08	1.34 (0.86–2.12)	0.19
Isolated CS	2.36 (1.28–4.01)	<0.01	2.09 (1.12–3.62)	0.02

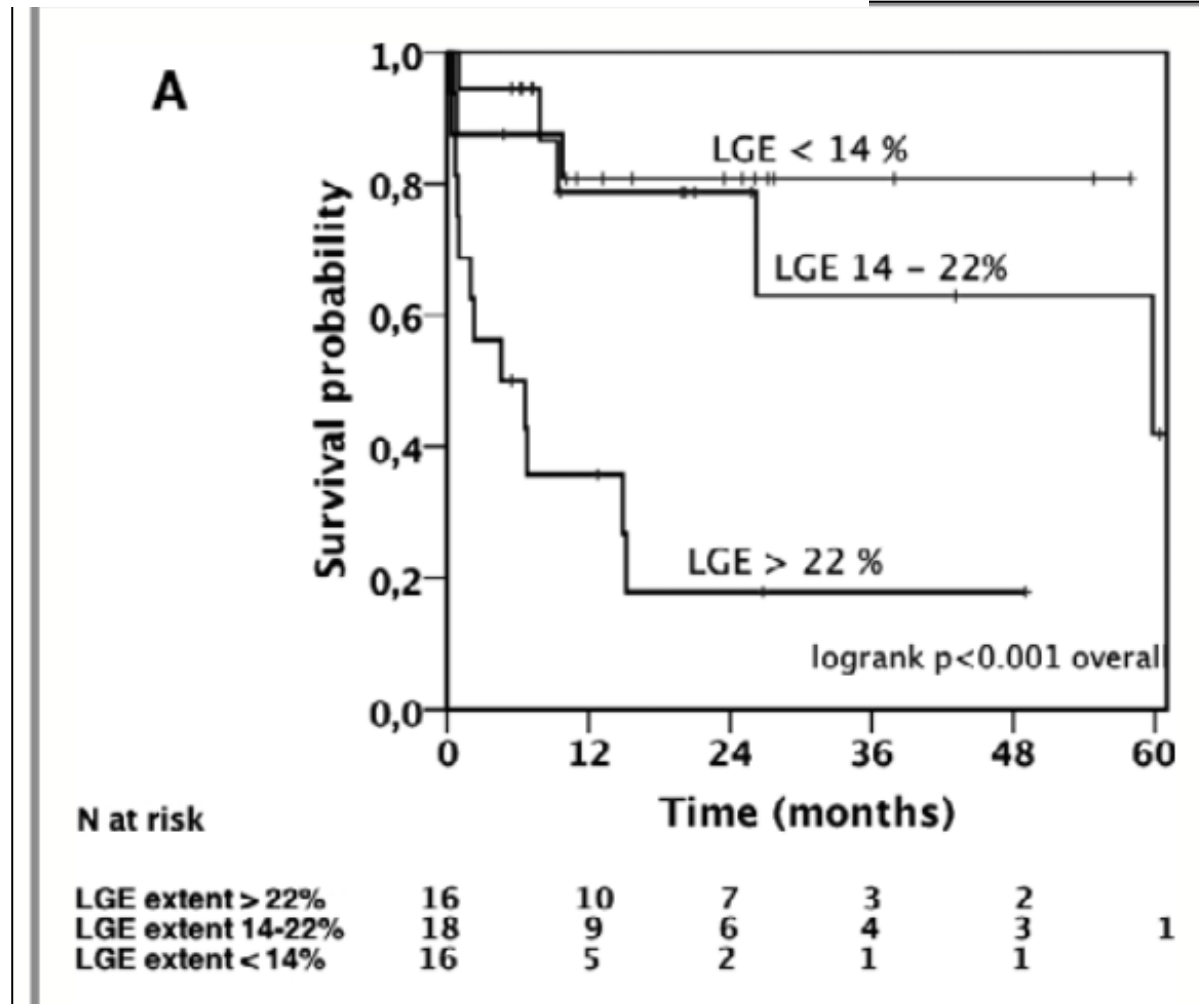
CS = cardiac sarcoidosis.

# Risque rythmique de la sarcoïdose cardiaque

Magnetic Resonance Imaging as a Predictor of Survival Free of Life-Threatening Arrhythmias and Transplantation in Cardiac Sarcoidosis

*J Am Heart Assoc.* 2016

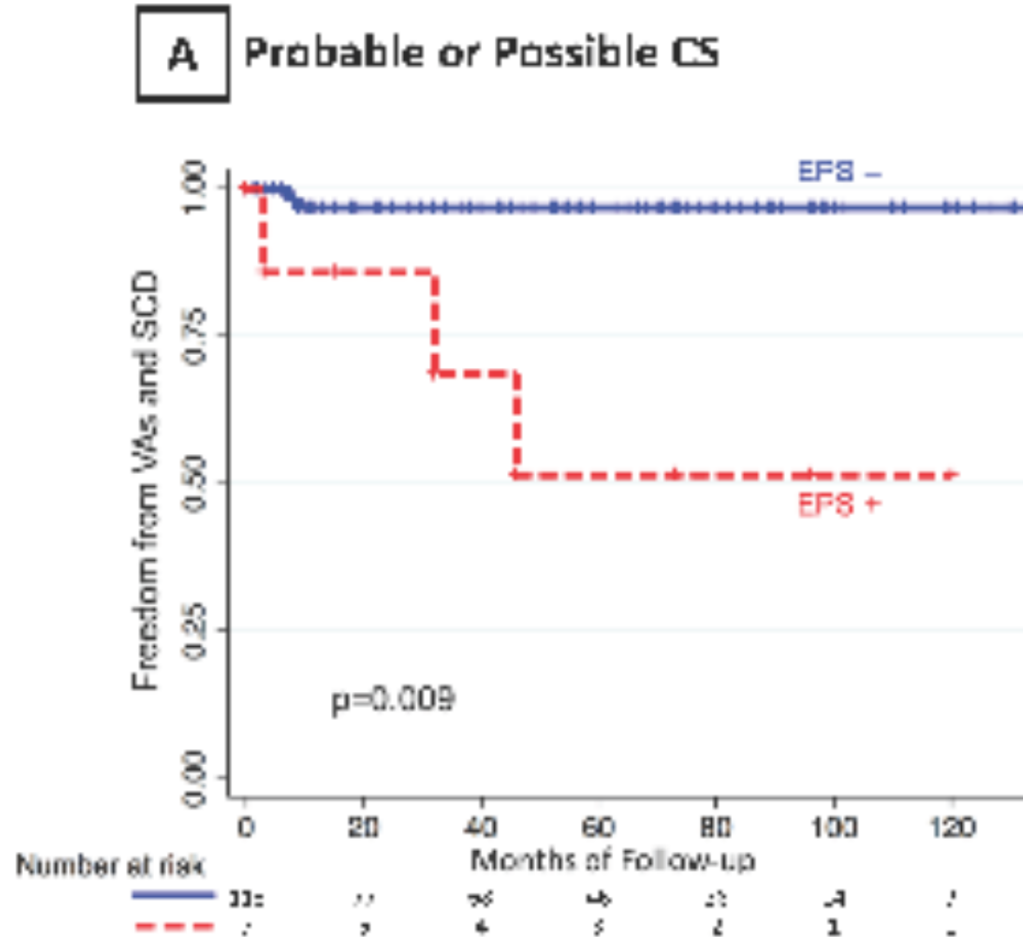
Kaj Ekström, MD; Jukka Lehtonen, MD; Helena Hänninen, MD; Riina Kandolin, MD; Sari Kivistö, MD; Markku Kupari, MD



# Risque rythmique de la sarcoïdose cardiaque

## Electrophysiologic Testing for Diagnostic Evaluation and Risk Stratification in Patients with Suspected Cardiac Sarcoidosis with Preserved Left and Right Ventricular Systolic Function

Matthew M. Zipse JCE 2019



120 consecutive patients with biopsy-proven extracardiac sarcoidosis and preserved LV/RV systolic function underwent EPS

Patients were followed for  $4.5 \pm 2.6$  years for SCD and VAs.

# Risque rythmique de la sarcoïdose cardiaque

Recommendations	Class <sup>a</sup>	Level <sup>b</sup>
<b>Risk stratification and primary prevention of SCD</b>		
ICD implantation is recommended in patients with cardiac sarcoidosis who have a LVEF $\leq 35\%$ . <sup>812,828–830,832</sup>	<b>I</b>	<b>B</b>
In patients with cardiac sarcoidosis who have an indication for permanent cardiac pacing related to high-degree AV block, ICD implantation should be considered, regardless of LVEF. <sup>816</sup>	<b>IIa</b>	<b>C</b>
In patients with cardiac sarcoidosis who have a LVEF $> 35\%$ but significant LGE at CMR after resolution of acute inflammation, ICD implantation should be considered. <sup>817–819,821,833,834</sup>	<b>IIa</b>	<b>B</b>
In patients with cardiac sarcoidosis who have a LVEF 35–50% and minor LGE at CMR, after resolution of acute inflammation, PES for risk stratification should be considered.	<b>IIa</b>	<b>C</b>
In patients with cardiac sarcoidosis, LVEF 35–50% and inducible SMVT at PES, ICD implantation should be considered. <sup>823–825</sup>	<b>IIa</b>	<b>C</b>

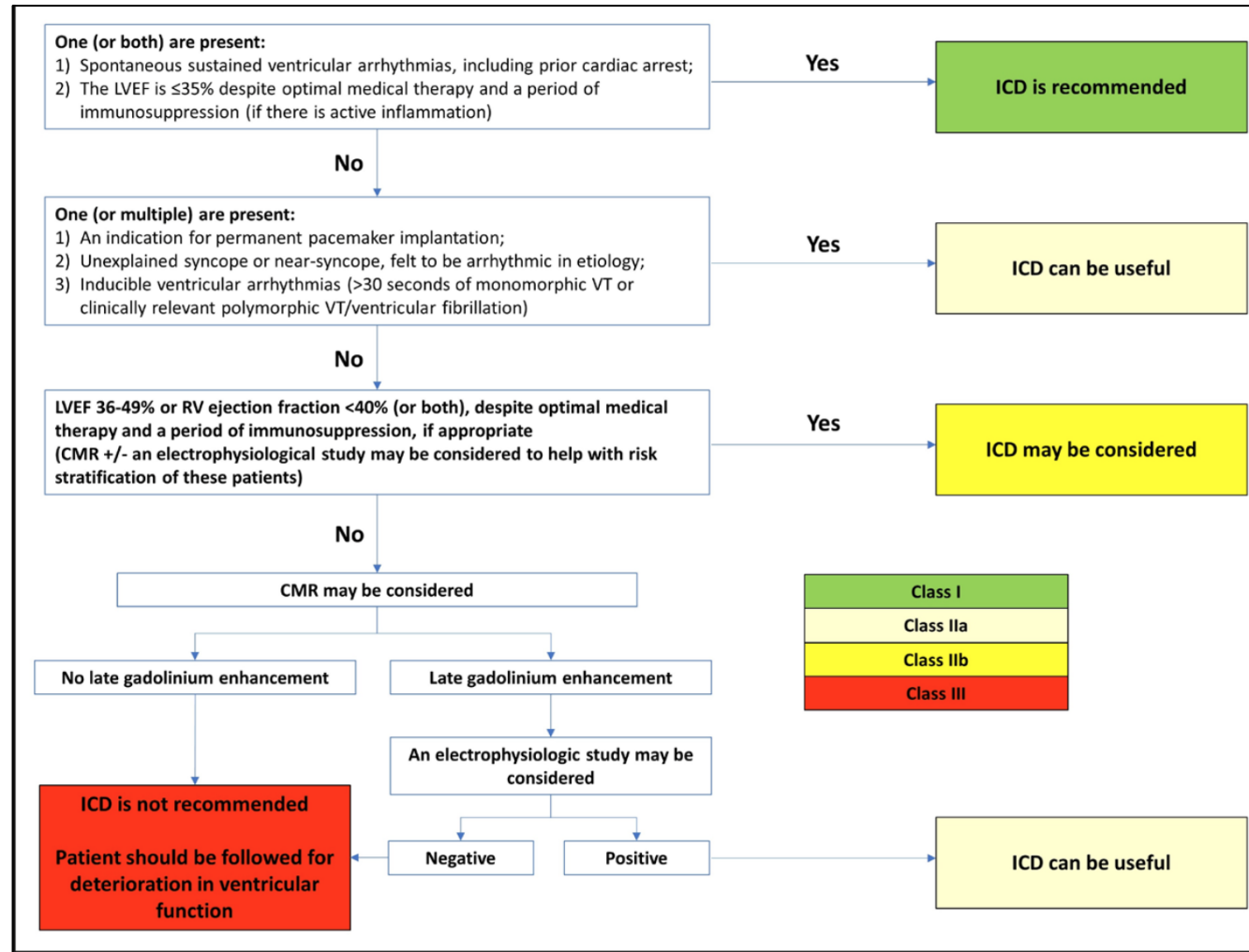
## 2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

<b>Secondary prevention of SCD and treatment of VAs</b>		
ICD implantation is recommended in patients with cardiac sarcoidosis who (1) have documented sustained VT, or (2) aborted CA. <sup>812,828–830,832</sup>	<b>I</b>	<b>B</b>

A widely accepted definition of *significant* LGE is not available.

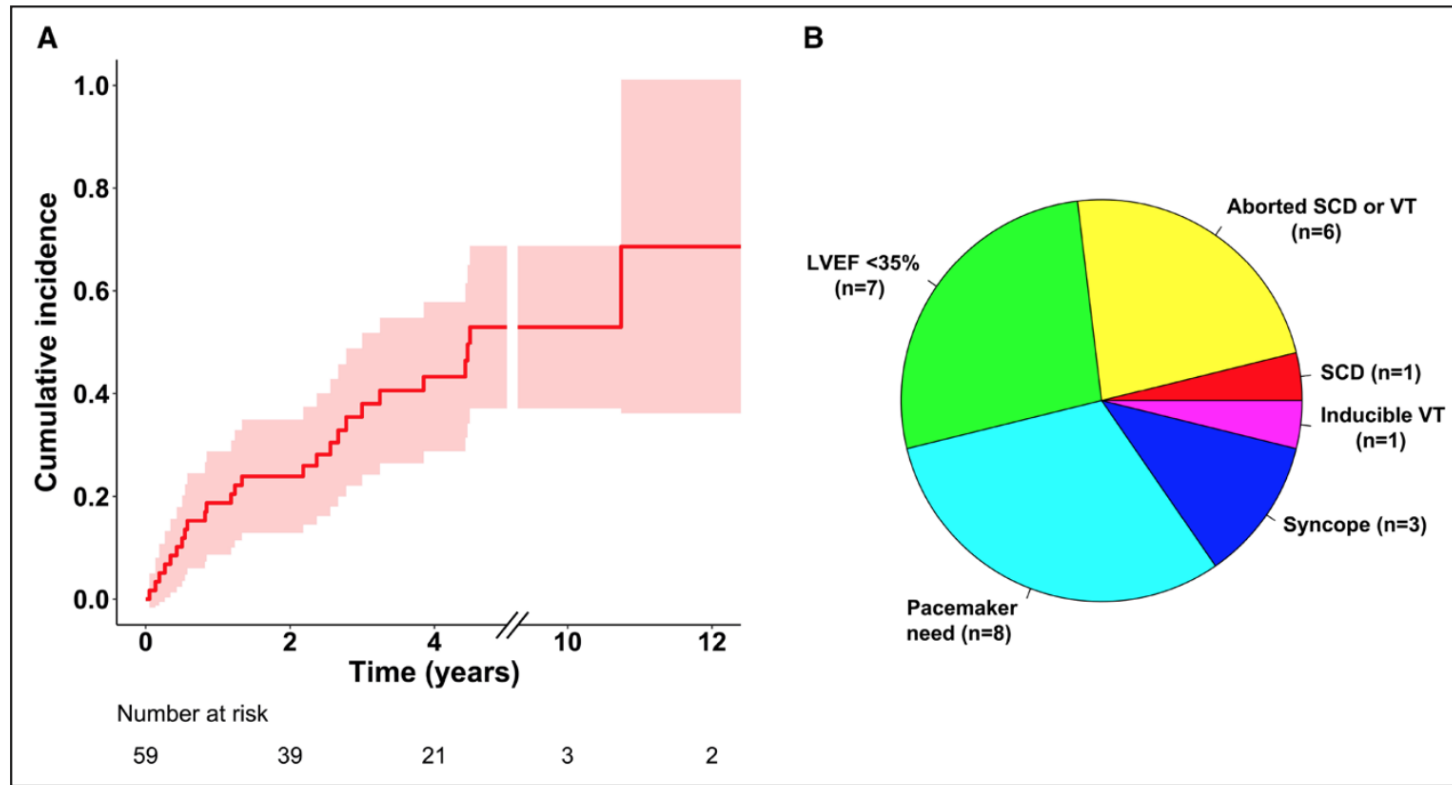


# Risque rythmique de la sarcoïdose cardiaque



# Incidence of Sudden Cardiac Death and Life-Threatening Arrhythmias in Clinically Manifest Cardiac Sarcoidosis With and Without Current Indications for an Implantable Cardioverter Defibrillator

Nordenswan et al. *circulation* 2022



Emergence of ICD indications during follow-up

## CONCLUSION

Diagnostic compliqué de sarcoïdose cardiaque difficile

Challenge diagnostique de la sarcoïdose cardiaque isolée

3 tableaux de présentations typiques : BAV, arythmies ventriculaires isolées, cardiomyopathie

Multiples éléments susceptibles d'orienter la stratégie

Extension des indications de défibrillateur

Merci pour votre attention

