Atteintes cardiaque de la sarcoïdose

Matthieu Mahévas, Service de Médecine Interne Mondor,

- Faire le diagnostic
- Etablir le pronostic
- Traitement
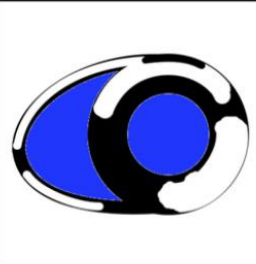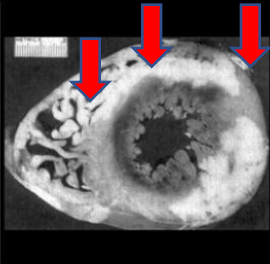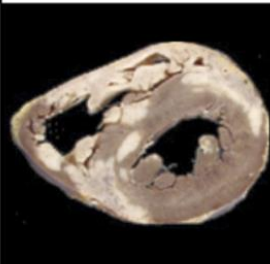
Pourquoi faire le diagnostic ?
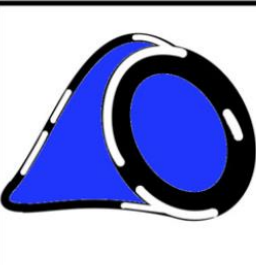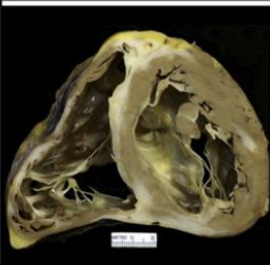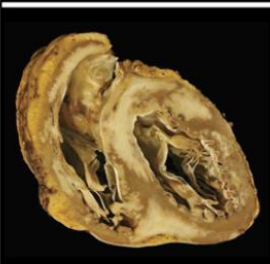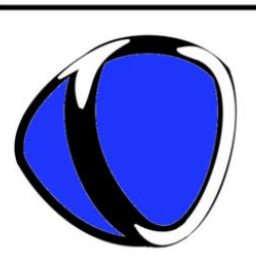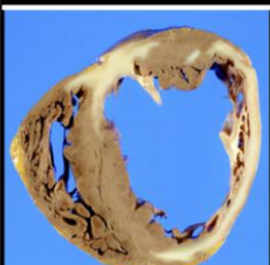
- Première cause de mortalité de la sarcoïdose au Japon
- Deuxième cause de mortalité aux USA

Prévalence de la sarcoidose cardiaque

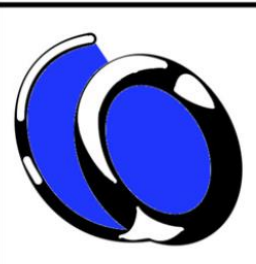|	Origine	Critères	Prévalence (%)	Nombre de sujets
Baughman et al., 2001	États-Unis (multicentrique)	Critères ACCESS	2.3	736
Johns, 1999	États-Unis (monocentrique)	Cohorte de sarcoïdoses chroniques	7	181
Smedema et al., 2005	Pays-Bas (multicentrique)	Dépistage systématique dans une cohorte de sarcoïdoses pulmonaires	19	101
Silverman et al., 1978	États-Unis (monocentrique)	Autopsies consécutives de sarcoïdoses	27	84
Sharma et al., 1993	États-Unis (monocentrique)	Autopsies consécutives de sarcoïdoses	20	123
Iwai et al., 1993	Japon (données nationales)	Autopsies consécutives de sarcoïdoses	66	320
Perry et Vuitch, 1995	États-Unis (monocentrique)	Autopsies consécutives de sarcoïdoses	75	38

dépend du contexte !

Cohen, RMI 2018

Clinical data	Guide	Pathology images	Involvement
A 35-year-old man; sudden death while driving a bus			Epicardial ✔ Multifocal ✔ Septal ✔ RV free wall ✔
B Age and sex unknown; sudden death			Epicardial ✔ Multifocal ✔ Septal ✔ RV free wall ✔
C 53-year-old man; heart transplantation due to cardiac sarcoidosis			Epicardial ✔ Multifocal ✔ Septal ✔ RV free wall ✔
D 53-year-old man; heart transplantation due to cardiac sarcoidosis			Epicardial ✔ Multifocal ✔ Septal ✔ RV free wall ✔
E 53-year-old man; end-stage heart failure due to cardiac sarcoidosis, died of hemorrhagic shock			Epicardial ✔ Multifocal ✔ Septal ✔ RV free wall ✔

Granulome
→
Ischémie locale
→
Fibrose

Septum

Voies de conduction

VD = défavorable

Okasha, JAHA, 2019
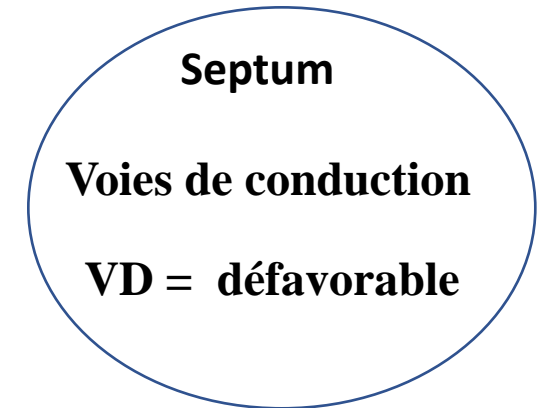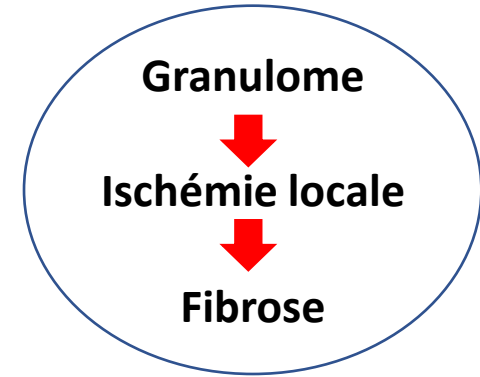
CENTRAL ILLUSTRATION: Clinical Features of Cardiac Sarcoidosis

Small patches of basal involvement, usually clinically silent
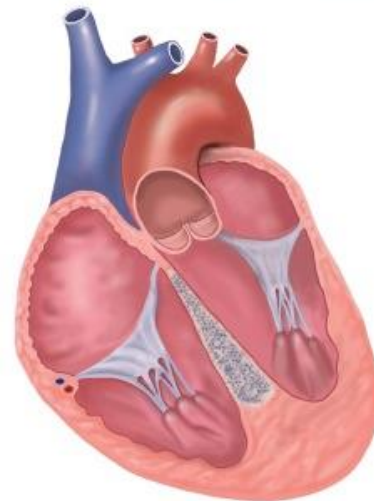
Large area of septal involvement, often clinically manifest as heart block

Re-entrant circuit involving area of granuloma/fibrosis leading to VT

Extensive areas of LV and RV involvement, often clinically manifest as heart failure +/- heart block +/- VT

Troubles de la conduction

Normal

Oreillette gauche

Fibrose

Oreillette droite

Noeud sinusal qui commande le rythme cardiaque

Noeud auriculo-ventriculaire
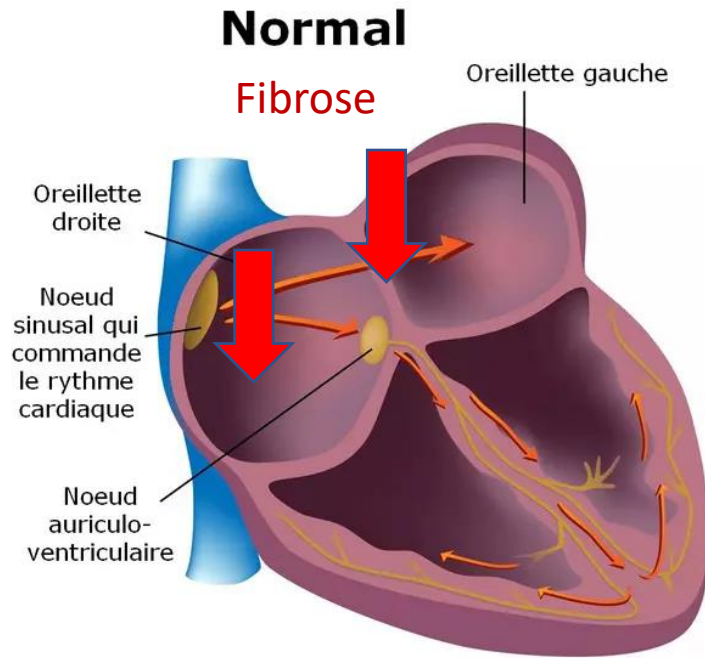
➢ Blocs auriculo-ventriculaires
BAV, Mobitz II, ou complets

➢ Parfois révélateurs de la sarcoïdose
30 % des BAV inexpliqués de l'adulte de < 60 ans

P-R constant
P bloquée parfois

BAV II Mobitz 2
(infranodal*)

PP réguliers
≥ 2 P bloquées

BAV haut degré
(infranodal*)

Dissociation entre P et complexes QRS

BAV III
(infranodal*)

Kandolin et al, Circ Arrhythm Electrophysiol 2011
Nery, CJE 2014

Tachychardie ventriculaire

Normal

- Oreillette gauche
- Oreillette droite
- Noeud sinusal qui commande le rythme cardiaque
- Noeud auriculo-ventriculaire

Fibrose et inflammation Septum /Purkinje
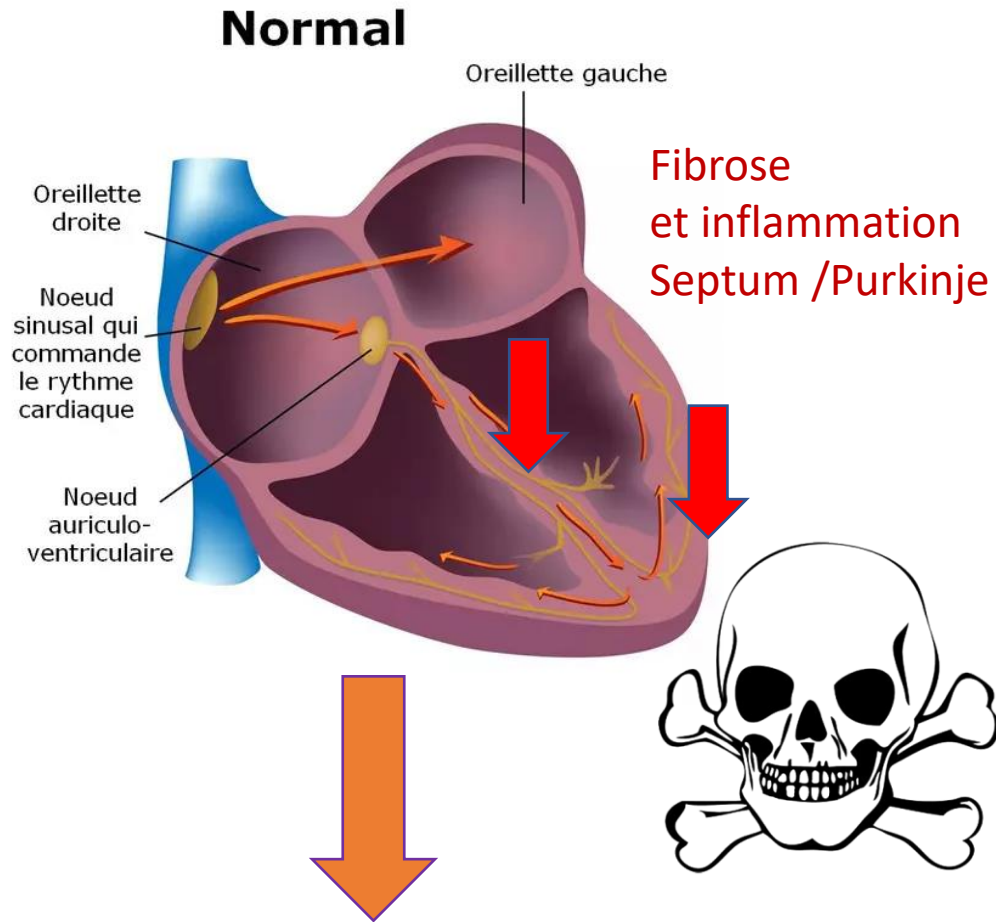
Très rarement fibrillation auriculaire

➢ Orages Rythmiques
➢ Parfois révélateurs de la sarcoïdose
➢ Précédés ESV, Bloc de branche biphasiculaire
➢ Syncope, mort subite

Okada, Circulation 2018

Insuffisance cardiaque

Oreillette gauche

Fibrose

Oreillette droite

Noeud sinusal qui commande le rythme cardiaque
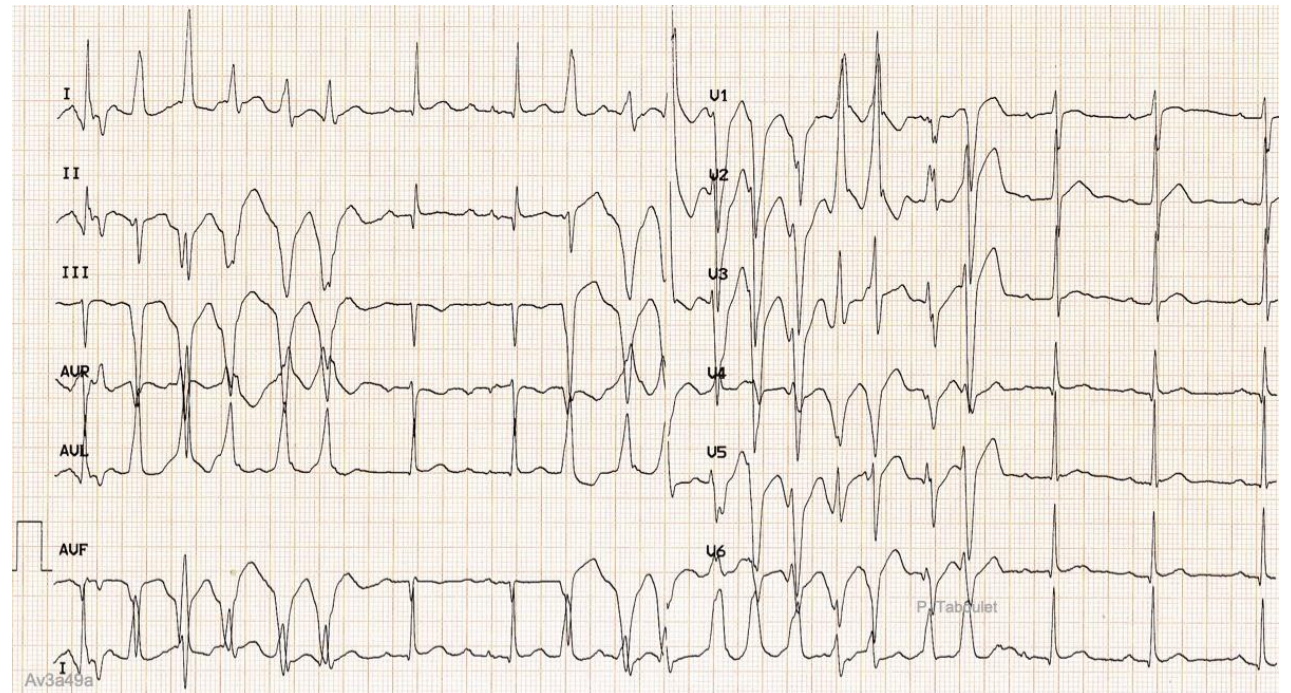
Noeud auriculo-ventriculaire
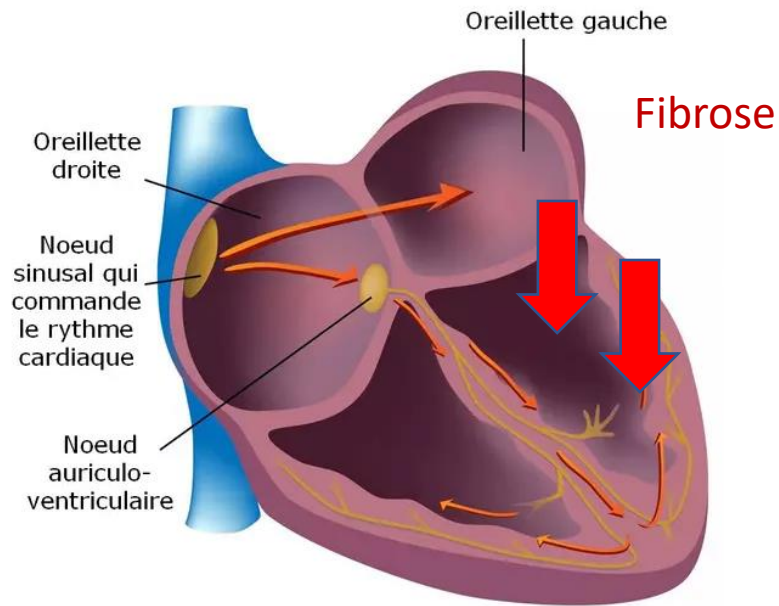
➢ Cardiopathie hypertrophique, dilatée
➢ Hypertrophie du septum
➢ Associations avec des troubles du rythme
➢ Décompensation cardiaque gauche+++

Okada, Circulation 2018
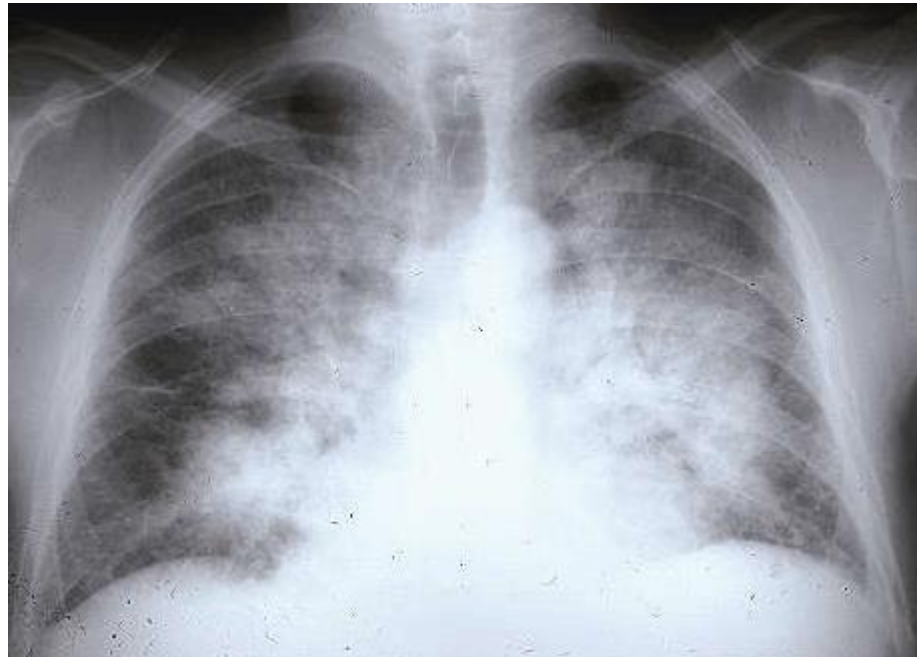
Deux grands modes de présentation
BAV/TV et insuffisance cardiaque

	Yazaki 2001, n=95	Blankstein, 2014	Greulich 2013	Kandolin 2015
BAV haut degré	45%	37%	14%	44%
TV	20%	20%	13%	33%
Insuffisance cardiaque	26%	14%	4%	18%

TEP

IRM
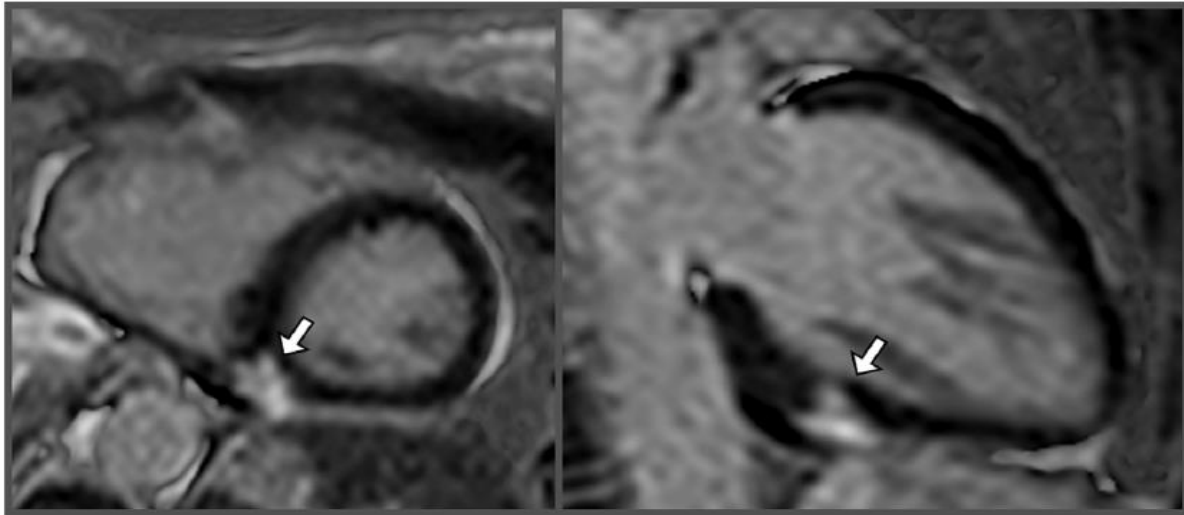
Activité métabolique de la maladie
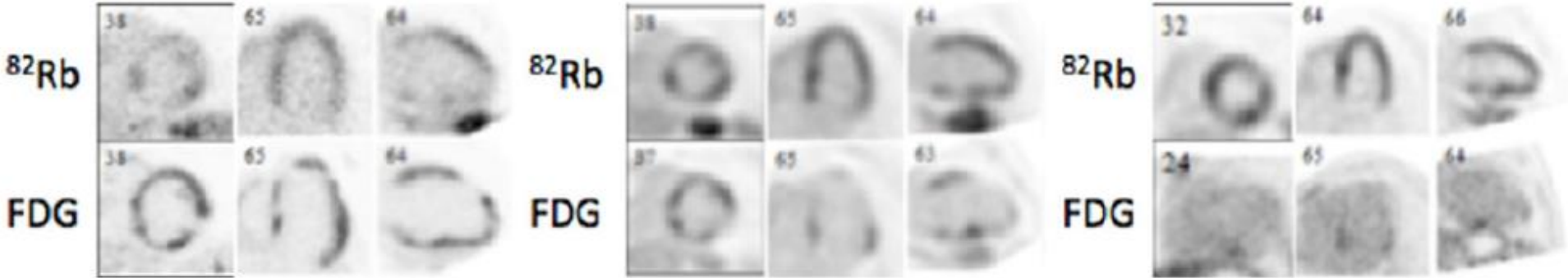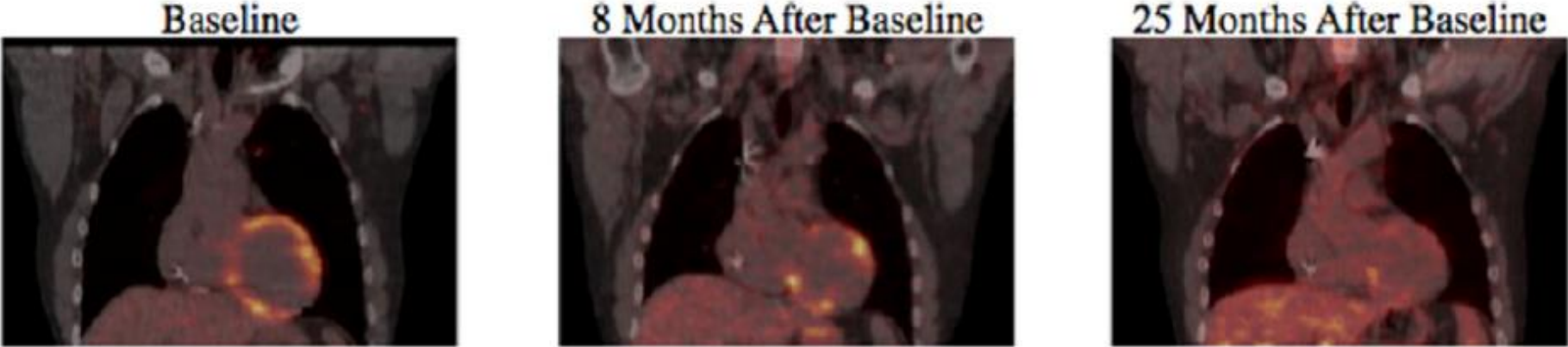
TROIS ASPECT: FOCAL, DIFFUS, FOCAL + DIFFUS

Ventricule droit = défavorable

Bernie, HRS 2014

Authors	Year	Subjects studied	JMHW guidelines	No. of patients ()	Fasting time (h)	Sensitivity (%)	Specificity (%)	Comments
Okumura et al.	2004	With sarcoidosis	1993	22	>12	100	91	PET is more sensitive than ^{67}Ga scintigraphy
Ishimaru et al.	2005	With sarcoidosis	1993	32	>6	100	82	Pre-administered heparin
Ohira et al.	2008	With suspected CS	1993	21	>12	88	39	**Comparing ^{18}F-FDG PET and MRI**
Langah et al.	2009	With suspected CS	1993	76	>18	85	90	PET CT with prolonged fasting >18 h
Tahara et al.	2010	With suspected CS	2006	24	>12	100	46→97	Analysis using the COV improved specificity
Manabe et al.	2013	With suspected CS	1993	67	>6	96	62	^{18}F-FDG uptake was related to ECG abnormalities
McArdle et al.	2013	With suspected CS	2006	134	>12	100	83	With a high-fat, low-carbohydrate diet on the day before PET
Blankstein et al.	2013	With suspected CS	1993	118	>3	71	45	With a high-fat, high protein, low-carbohydrate diet
Soussan et al	al 2013	With sarcoidosis	2006	58	>12	84	78	**Comparing ^{18}F-FDG PET avec IRM**

TEP/TDM: tomographie par émission de positons couplée à une tomodensitométrie; ^{18}F-FDG: fluorodésoxyglucose marqué au fluor-18.

Régime cardiaque: suppression de l'apport en hydrates de carbone 12 h avant l'examen

± régime riche en lipides

Période de jeûne

± héparine non fractionnée

Injection de ^{18}F-FDG

60-90 minutes

Acquisition TEP/TDM

Images

C'EST VRAIMENT TROP INJUSTE!
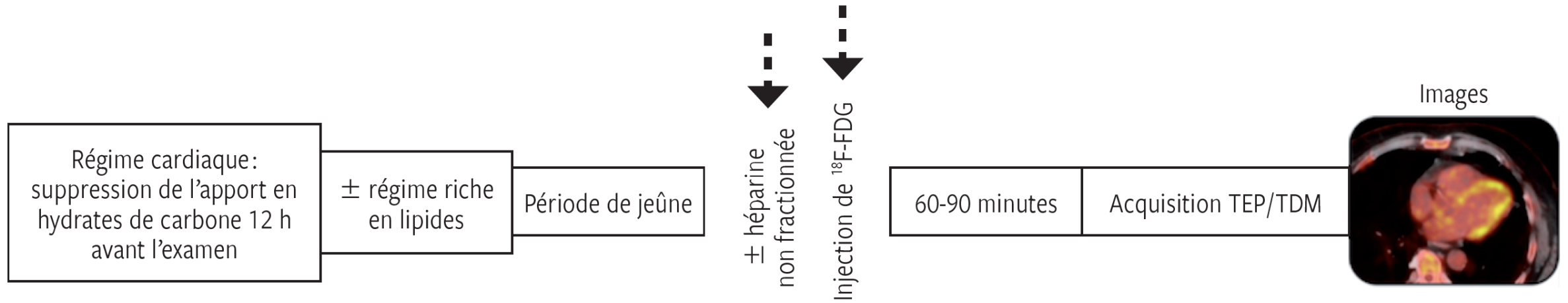
IRM cardiaque avec injection de Gadolinium

Non spécifique de la sarcoïdose
Aspect « Patchy » évocateur

Transmural   Focal   Diffus
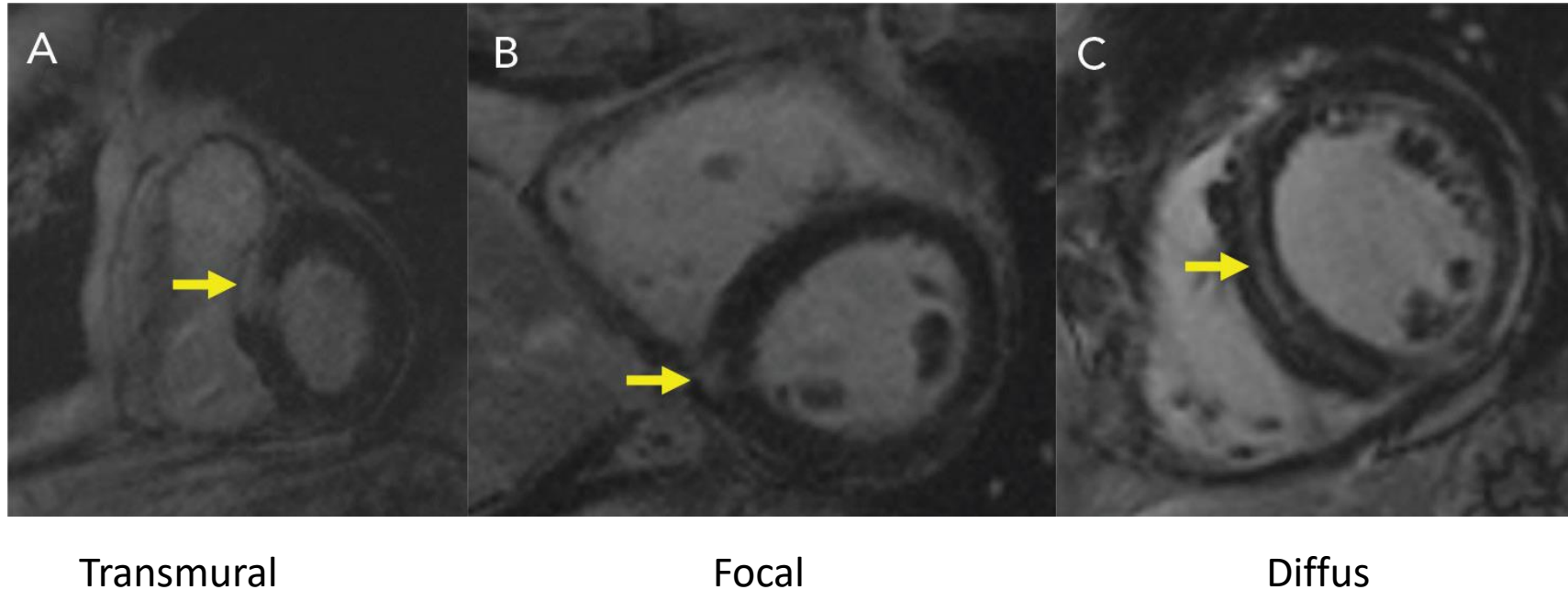
Anomalies cinétiques, morphologiques et surtout **prises de contraste tardives = Fibrose myocardique**
Anomalies non systématisés par rapport à la vascularisation des artères coronaires

Patient A

- Subendocardial DE in LV anterolateral wall
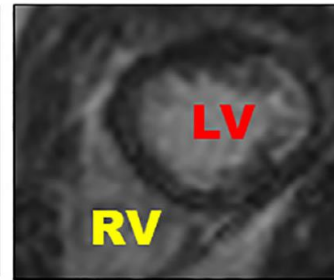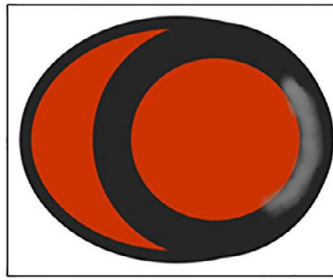
Patient B

- Midmyocardial DE in basal septum
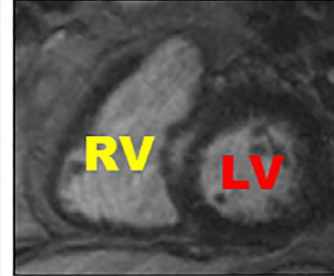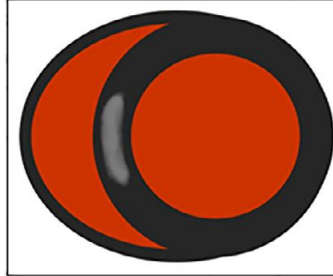
Patient C

- Midmyocardial to epicardial DE in LV inferoseptal wall and inferior LV wall
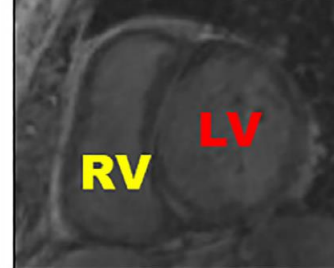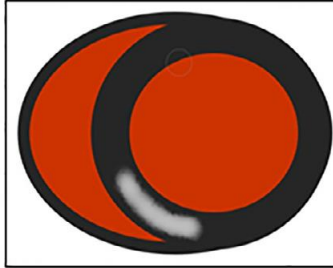
Patient D

- Transmural DE in LV inferolateral, inferior and inferoseptal wall
- RV free wall DE
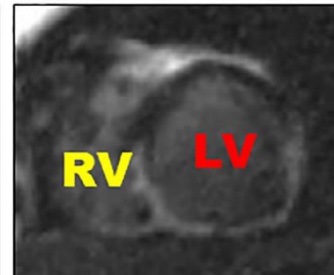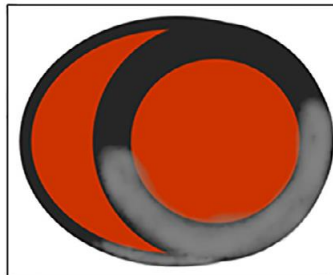
Patient E

- Epicardial DE in LV inferior, inferoseptal, and anteroseptal walls
- Midmyocardial DE in LV inferolateral and RV anterolateral walls

Tan, Am J Cardiol 2019
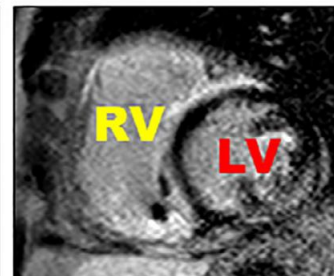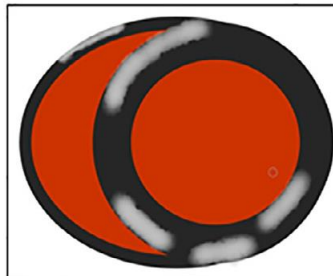
Rehaussement tardif après injection de Gadolinium du VG, intérêt pronostic

Kaplan-Meier Survival Curves: Endpoint 1 (Hard Events Only)

p-log-rank < 0.0001

Kaplan-Meier Survival Curves: Endpoint 2 (All Events)

p-log-rank < 0.0001

Days after CMR

Patients at Risk

No LGE	89	63	44	29	20	15	10	5
LGE	23	20	17	11	5	4	4	2
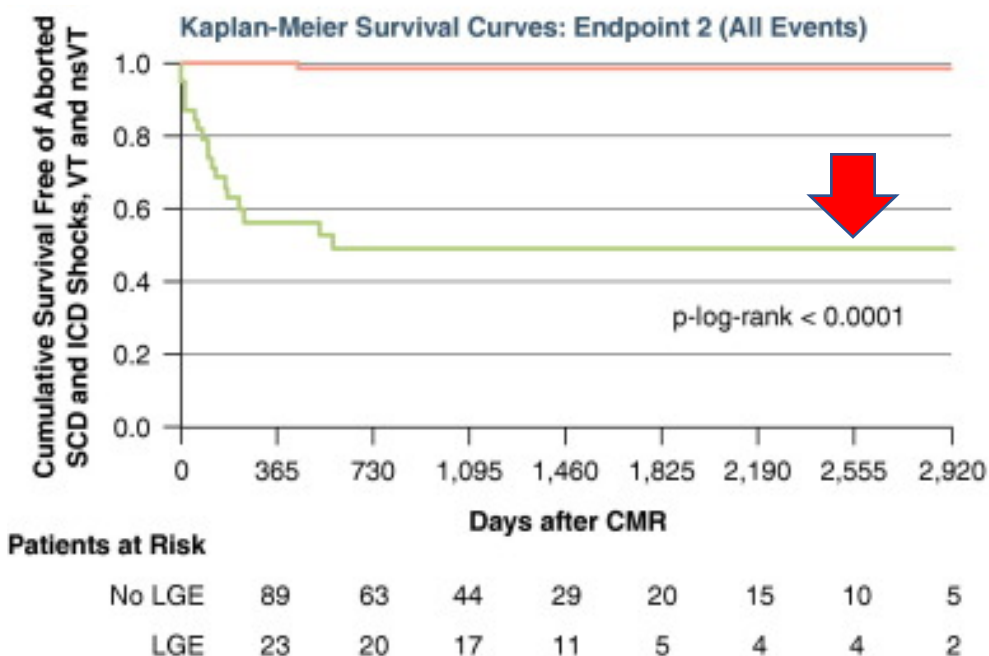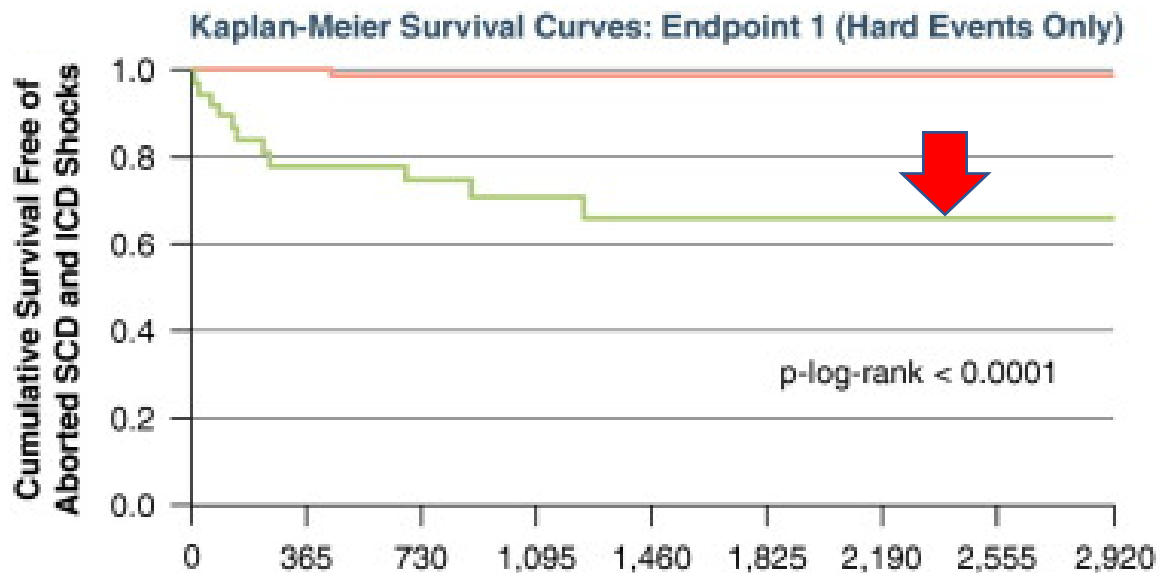
155 patients consécutifs, suivi 2.6 ans en médiane
Rehaussement tardif 39/155 (25.9%).
11/39 = Décés ou trouble du rythme grave
1/133 = Décés ou trouble du rythme grave

Patel. Circulation 2009, Greulich JACC 2013

IRM = stratifier le risque

TEP = Evaluer l'activité

Specific locations of myocardial inflammation and fibrosis are associated with higher risk of events in cardiac sarcoidosis ℮

Ana Devesa, MD, PhD,[1,2,3] Philip M. Robson, PhD,[1] Busra Cangut, MD, MS,[1] Ravi Vazirani, MD,[1,2] Vittoria Vergani, MD,[4] Gina LaRocca, MD,[3] Angelica M. Romero-Daza, MD,[5] Steve Liao, MD,[3] Lévi-Dan Azoulay, MD,[1] Renata Pyzik, MS, MA,[1] Rima A. Fayad, MPH,[1] Adam Jacobi, MD,[6] Ronan Abgral, MD, PhD,[7] Adam S. Morgenthau, MD,[8] Marc A. Miller, MD,[9] Zahi A. Fayad, PhD,[1] Maria Giovanna Trivieri, MD, PhD[1,3]

Longitudinal monocentric study

- **Clinical suspicion of active CS due to established extra-CS and either clinical presentation or previous imaging suggestive of cardiac sarcoidoisis**

- Exclusion criteria included insulin-dependent diabetes mellitus, blood sugar >200 mg/dL before scanning, claustrophobia, pregnancy/nursing, presence of an implanted device contraindicated forMR imaging before the study, and impaired renal function (estimated glomerular filtration rate <40mL/min/ 1.73 m2).

- **The primary endpoint included VA (defined as sustained ventricular tachycardia, ventricular fibrillation, or implantable cardioverter defibrillator appropriate discharge); heart failure hospitalization; and death of any cause.**

Qualitative analysis to assign a category

Coregistered short-axis hybrid LGE MR and ^{18}F-FDG PET images were assessed, and patients were categorized into the following 4 groups: MR(+)PET(+), MR(+)PET(−), MR(−)PET(+), and MR(−)PET(−), similar to our previous study.[7]:

1. MR(+)PET(+) is defined by the presence of LGE and a pattern of increased focal or focal on diffuse ^{18}F-FDG uptake
2. MR(−)PET(+) if there was increased focal or focal on diffuse ^{18}F-FDG uptake in the absence of LGE
3. MR(+)PET(−), defined by LGE presence with no focal ^{18}F-FDG uptake, is representative of inactive CS with residual scar
4. MR(−)PET(−) if there was neither LGE nor focal ^{18}F-FDG

	All	No events	Events	P value
n	124	102	22	
Age, y	57.1 (8.9)	57.3 (8.8)	56.5 (9.9)	.700
Femal	49 (39.5)	44 (42.7)	6 (27.3)	.200
Cardiovascular history				
Hypertension	58 (46.8)	45 (44.1)	13 (59.1)	.300
Diabetes	25 (20.2)	16 (15.7)	9 (40.9)	.019
Dyslipidemia	39 (31.5)	29 (28.4)	10 (45.5)	.200
Smoker	7 (5.6)	3 (2.9)	4 (18.2)	.019
Ex-smoker	39 (31.5)	30 (29.4)	9 (40.9)	.300
Previous myocardial infarction	1 (0.8)	1 (1.0)	0 (0.0)	1
Previous PCI	5 (4.0)	4 (3.9)	1 (4.5)	1
Previous CABG	2 (1.6)	2 (2.0)	0 (0.0)	1
Previous stroke	3 (2.4)	3 (2.9)	0 (0.0)	1
History of arrhythmia				
AF	21 (16.9)	16 (15.7)	5 (22.7)	.300
Flutter	9 (7.3)	7 (6.9)	2 (9.1)	.600
Supraventricular tachycardia	10 (8.1)	6 (5.9)	4 (18.2)	.090
Second-degree Mobitz I AV block	2 (1.6)	1 (1.0)	1 (4.5)	.300
Second-Degree Mobitz II AV block	0 (0.0)	0 (0.0)	0 (0.0)	NA
Complete heart block	0 (0.0)	0 (0.0)	0 (0.0)	NA
Sustained ventricular tachycardia	2 (1.6)	0 (0.0)	2 (9.1)	.030
Nonsustained ventricular tachycardia	3 (2.4)	3 (2.9)	0 (0.0)	1
Pacemaker	0 (0.0)	0 (0.0)	0 (0.0)	NA
Implantable cardioverter-defibrillator	0 (0.0)	0 (0.0)	0 (0.0)	NA

	All	No events	Events	P value
Syncope	5 (4.0)	4 (3.9)	1 (4.5)	1
Palpitation	20 (16.1)	16 (15.7)	4 (18.2)	1
Dyspnea	54 (43.5)	42 (41.2)	12 (54.5)	.700
Clinical heart failure	8 (6.5)	4 (3.9)	4 (18.2)	.059
Blood parameters				
White blood cell count (n = 95)	7.1 (2.1)	6.9 (2.1)	7.8 (1.9)	.090
Brain natriuretic peptide, pg/mL (n = 68)	33.3 (17.2-109.0)	24.0 (12.0-41.0)	186.0 (59.0–557.6)	<.001
C-reactive protein, mg/dL (n = 26)	2.8 (1.4-5.4)	2.6 (1.3-5.4)	3.8 (2.5–6.1)	.400
CMR findings				
LVEDVI, mL/m^2	79.9 (70.1-92.7)	78.9 (68.0-91.9)	86.7 (74.6–127.1)	.098
LVESVI, mL/m^2	32.1 (24.6-41.7)	30.4 (24.3-38.6)	41.3 (37.6–81.3)	<.001
LVEF, %	59.0 (51.0-66.0)	60.0 (54.5-66.0)	46.0 (32.0–55.5)	<.001

	All	No events	Events	P value
PET+/MR+	16 (12.9)	8 (7.8)	8 (36.4)	.001
PET+/MR−	62 (50.0)	54 (52.9)	8 (36.4)	.239
PET−/MR+	6 (4.8)	6 (5.9)	0 (0.0)	.590
PET−/MR−	40 (32.3)	34 (33.3)	6 (27.3)	.802
SUVmax	2.2 (1.4-4.7)	2.0 (1.3-4.7)	3.0 (1.7–6.4)	.400
TBRmax	1.7 (1.1-3.6)	1.6 (1.0-3.6)	2.2 (1.3–4.6)	.200
Free RV wall ^{18}F-FDG uptake	8 (6.5)	6 (5.9)	2 (9.1)	.631

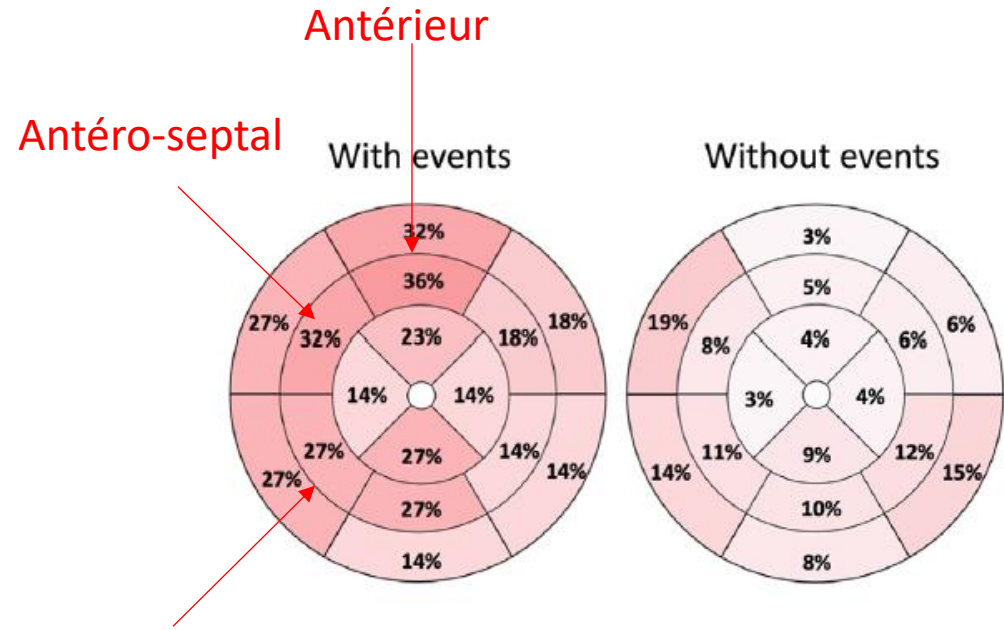Compared with patients without events, presence of LGE in patients with events was significantly more frequent in all anterior segments, mid- to apical inferior segments, and mid-anteroseptum (Figure 3 and Supplemental Table S3). When adjusted by LVEF and global relative enhanced volume (model 1), presence of LGE in the basal anterior segment remained an independent predictor for events (odds ratio [OR] 10.5 [1.2–92.4], P = .034), and the most frequent pattern in this segment was the intramyocardial scar. This finding remained significant when adjusted for LVEF, global relative enhanced volume, and immunosuppressive therapy and when adjusted for LVEF, global relative enhanced volume, and beta-blockers (Supplemental Table

273,0 mm

Modèle à 17 segments
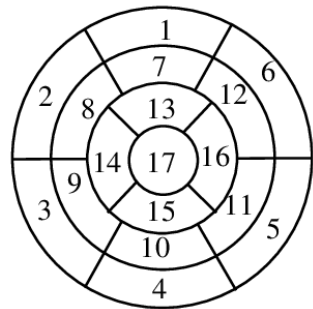
1. Base. Antérieur
2. Base. Antéro-septal
3. Base. Inféro-septal
4. Base. Inférieur
5. Base. Inféro-latéral
6. Base. Antéro-latéral
7. Médian. Antérieur
8. Médian. Antéro-septal
9. Médian. Inféro-septal
10. Médian. Inférieur
11. Médian. Inféro-latéral
12. Médian. Antéro-latéral
13. Apical. Antérieur
14. Apical. Septum
15. Apical. Inférieur
16. Apical. Latéral
17. Apex
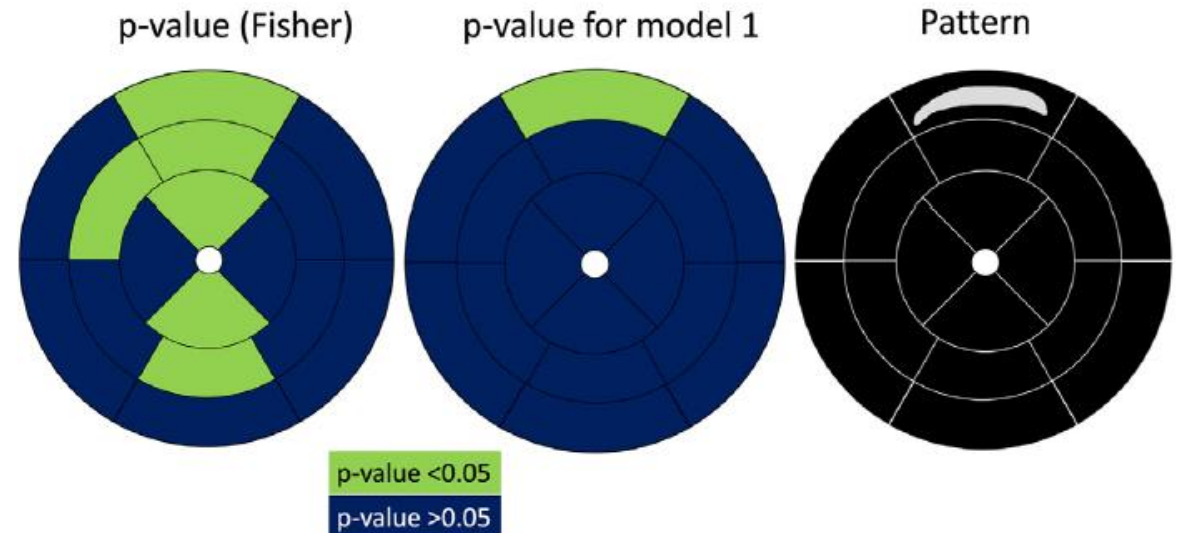
Similarly, focal ^{18}F-FDG uptake in patients with events was significantly more frequent in all anterior segments as well as in the mid-inferior segment (Figure 4 and Supplemental Table S4). After adjusting by model 1, the presence of focal ^{18}F-FDG in the basal anterior segment was an independent predictor for events (OR 5.5 [1.1–27.5], $P = .038$).

Compared with patients without VA, LGE presence in patients with VA was significantly more frequent in basal to mid-anterior and inferoseptal segments, as well as in the mid-anteroseptum (Supplemental Figure S2 and Supplemental Table S5). When adjusted by model 1, the presence of LGE in basal to mid-anterior segments, as well as in the mid-septum, was an independent predictor for VA (basal anterior: OR 50.6 [4.0–636.9], $P = 0.002$; mid-anterior: OR 17.2 [2.1–142.7], $P = .008$; mid-anteroseptum: OR 21.6 [3.5–131.9], $P = .001$; mid-inferoseptum: OR 9.2 [1.6–54.7], $P = .014$). The pattern most frequently found in these segments was an intramyocardial scar. In line with these findings,
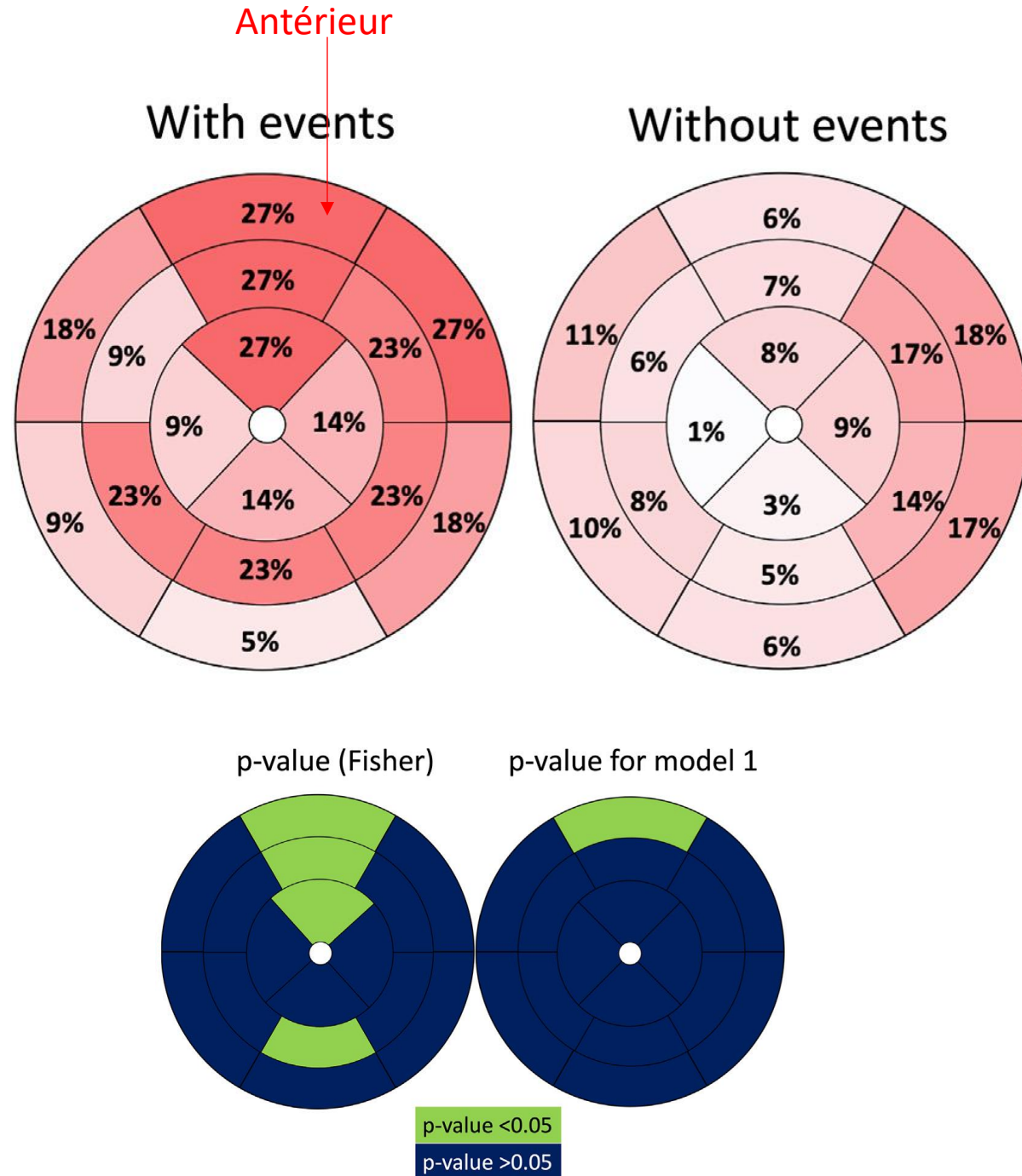
The results of this work suggest

(1) the concurrence of inflammation (ie, focal 18F-FDG uptake) and fibrosis (ie, LGE) increases the risk of VA, heart failure hospitalization, and all-cause death in patients with suspected CS.

2) thepresence of LGE and/or focal 18F-FDG uptake in the basal anterior myocardial segment is an independent predictor of adverse events;

(3) the presence of LGE and/or focal 18F-FDG in the basal to mid-anterior segments and in the mid-septum significantly increases the risk of VA in patients with suspected CS;

(4) the presence of inflammation and/or scar in the lateral wall (a frequent pattern) is not associated with a greater risk of events.

Intérêt de la TEP/IRM dans la sarcoïdose cardiaque

- **148 patients with clinical suspicion of CS due to established extracardiac involvement and/or clinical presentation suggestive of the disease based on cardiac-related symptoms or prior imaging were recruited at Mount Sinai Hospital**

- **The primary endpoint included cardiac-related death, aborted cardiac arrest, sustained ventricular arrhythmia, or secondary prevention implantable cardioverterdefibrillator**

TABLE 3 Patients Meeting the Composite Clinical Endpoint

|	MR(+) PET(+)$_{FOCAL}$ (n = 34)	MR(+) PET(+)$_{DIFFUSE}$ (n = 14)	MR(+) PET(−) (n = 23)	MR(−) PET(+)$_{FOCAL}$ (n = 18)	MR(−) PET(+)$_{DIFFUSE}$ (n = 21)	MR(−) PET(−) (n = 38)	Total (N = 148)	MR(+)$_{5.7\%}$ PET(+)$_{FOCAL}$ (n = 19)	Total[a] (N = 142)
VT with/without ICD shock	5	0	0	2	2	0	9	4	8
Secondary prevention ICD	4	0	1	1	1	2	9	4	9
Heart failure	2	2	0	0	1	4	9	1	9
New heart block	1	0	0	0	0	0	1	0	1
Total meeting endpoint									
Primary[b]	9 (26)	0 (0)	1 (4)	3 (17)	3 (14)	2 (5)	18 (12)	8 (42)	17 (12)

Patients designated MR(+)$_{5.7\%}$PET(+)$_{FOCAL}$ had increased odds of meeting the primary clinical endpoint compared to those with all other imaging classifications (unadjusted OR: 9.2 [95% CI: 3.0-28.7]; P = 0.0001),

Trivieri, JACC 2024

- Sarcoïdose connue, symptômes cardiologiques

- Symptômes cardiologiques, sarcoïdose inconnue

Les examens diagnostiques

Table 2 Prevalence of abnormalities, sensitivity, and specificity of diagnostic criteria

Abnormality on baseline testing	Prevalence[*]	Sensitivity (95% CI) (%)	Specificity (95% CI) (%)
History of cardiac symptoms	12 (19)	46 (26–27)	95 (82–99)
Electrocradiogram	3 (50)	8 (1–27)	97 (86–100)
Holter	13 (21)	50 (29–71)	97 (86–100)
Echocardiogram	8 (13)	25 (10–47)	95 (82–99)
Any screening variable	29 (47)	100 (88–100)	87 (72–96)
Two or more screening variables	7 (11)	25 (10–47)	97 (86–99)
Three or more screening variables	1 (2)	4 (1–21)	100 (92–100)

Mehta, Chest 2008
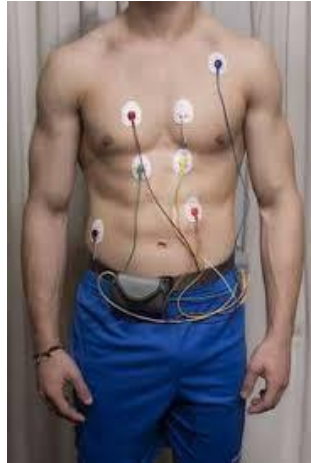
Stratégie de diagnostic histologique
Confirmation histologique d'une infiltration granulomatose lors d'une biopsie myocardique, en l'absence d'autres causes identifiées
Stratégie de diagnostic clinique

Diagnostic de probable sarcoïdose cardiaque (SC) en cas de:

• Confirmation histologique d'une infiltration granulomateuse lors d'une biopsie extracardiaque

• Positivité en présence d'au moins un critère suivant:
 – une cardiomyopathie ou un bloc atrioventriculaire (BAV) répondant au traitement stéroïdien et/ou immunosuppresseur
 – une baisse inexpliquée de la FEVG < 40 %
 – une tachycardie ventriculaire soutenue (spontanée ou induite) inexpliquée
 – un BAV II de type Mobitz II ou de type III
 – la présence d'une captation myocardique de type « patchy » à la TEP/TDM au ^{18}F-FDG (selon un pattern compatible avec la SC)
 – la présence d'un foyer de rehaussement tardif à l'IRM cardiaque (selon un pattern compatible avec la SC)
 – la présence d'une captation à la scintigraphie au gallium 67 (selon un pattern compatible avec la SC)

• Exclusion d'autres causes de manifestation cardiaque

HRS, 2014

Japanese Circulation Society for the Diagnosis of CS (2017)

1. **Histological diagnosis group (those with positive myocardial biopsy findings)**
2. **Clinical diagnosis group (those without a positive myocardial biopsy)**
   Granulomas are found in organs other than the heart, and clinical findings are strongly suggestive of cardiac involvement; or clinical findings are strongly suggestive of pulmonary or ophthalmic sarcoidosis and at least 2 of the 5 characteristic laboratory† and clinical findings of sarcoidosis are strongly suggestive of CS

Clinical findings that strongly suggest the presence of cardiac involvement.
   1. 2 or more of the 5 major criteria are satisfied.
   2. 1 of the 5 major criteria and 2 or more of the 3 minor criteria are satisfied.

1. **Major criteria**
   a) High-grade atrioventricular block or fatal ventricular arrhythmia (i.e., VT or VF)
   b) Basal thinning of the ventricular septum or abnormal ventricular wall anatomy (ventricular aneurysm, thinning of the middle or upper ventricular septum, regional ventricular wall thickening)
   c) Left ventricular contractile dysfunction (left ventricular ejection fraction <50%)
   d) Abnormalities of ^{67}Ga citrate scintigraphy or ^{18}F-FDG PET
   e) Gadolinium-enhanced CMR with delayed contrast enhancement of the heart

2. **Minor criteria**
   f) Abnormal ECG findings: ventricular arrhythmias (NSVT, multifocal PVCs), bundle branch block, axis deviation, or abnormal Q waves
   g) Perfusion defects on myocardial perfusion scintigraphy (SPECT)
   h) Endomyocardial biopsy: monocyte infiltration and myocardial fibrosis

Mme D. 51 ans

- **Sarcoïdose Médiastino-pulmonaire + atteinte rénale** au stade d'insuffisance rénale terminale évoluant depuis 5 ans.

- Liste de greffe

- Bilan cardiologique:
  - ➢ ETT normale
  - ➢ ECG bloc de branche incomplet gauche
  - ➢ Holter ECG ESV
  - ➢ IRM normale

3 mois plus tard

- Holter de contrôle car palpitations
- Très nombreuses ESV et épisode de tachychardie supraventriculaire non soutenue
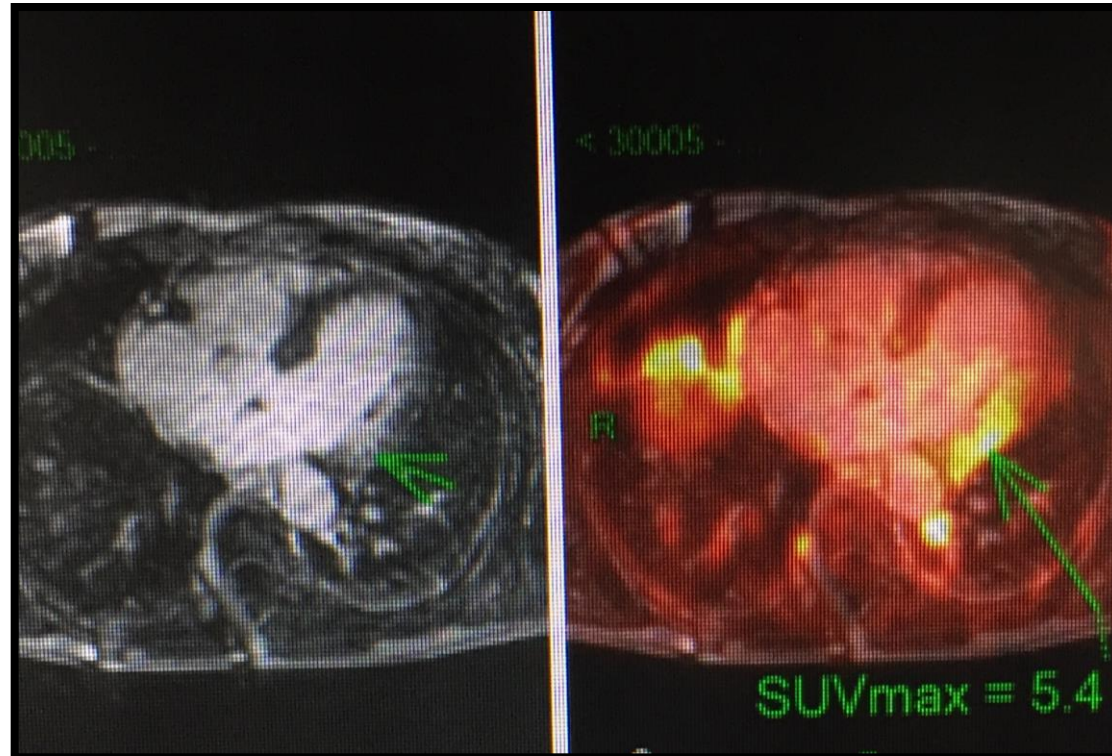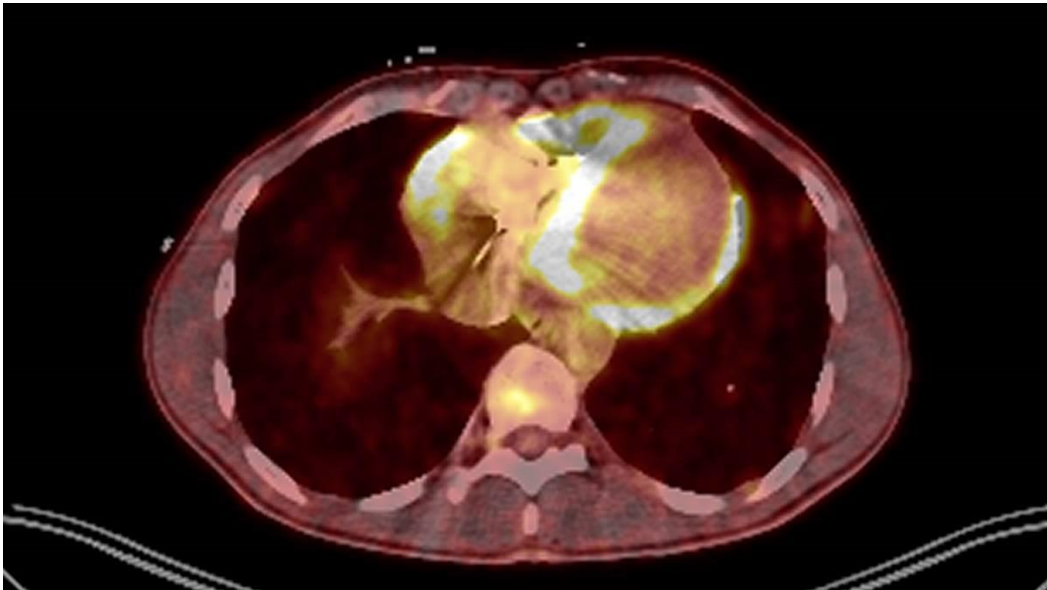
Mr C, 55 ans

- Originaire de Guadeloupe

- Asthénie, amaigrissement, dyspnée, toux, Ganglions médiastinaux

- BAV II lucciani wenckeback

ETT: Cardiopathie non dilaté hypokinétique, FEVG: 25%

IRM: Réhaussement sous épicardique global

- Orage rythmique = TV

- DAI double chambre

- Biopsies endobronchiques = Granulomes

Devant toute sarcoïdose cardiaque

POSEZ VOUS LA QUESTION !

FAUT IL METTRE UN DAI OU UNE LIFEVEST ?

Pour éviter

QUAND METTRE UN DAI

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

1. **DAI si FEVG < 35%**
2. **DAI si FEVG > 35% mais RT significatif après traitement de l'inflammation (>9/22)**
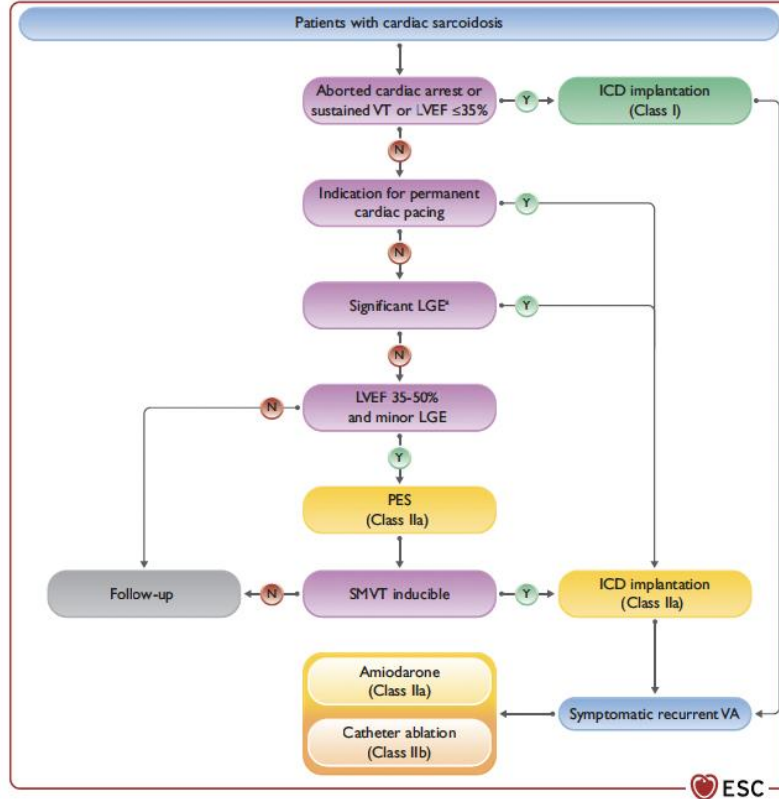3. **SVP si FEVG > 35% et RT minime. Si + : DAI**

Figure 24 Algorithm for sudden cardiac death prevention and treatment of ventricular arrhythmia in patients with cardiac sarcoidosis. ICD, implantable cardioverter defibrillator; LGE, late gadolinium enhancement; LVEF, left ventricular ejection fraction; N, No; PES, programmed electrical stimulation; SMVT, sustained monomorphic ventricular tachycardia; VA, ventricular arrhythmia; VT, ventricular tachycardia; Y, Yes. aLGE affecting ≥9/22 segments or ≥22% of the LV mass has been associated with arrhythmic endpoints.

2021 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy

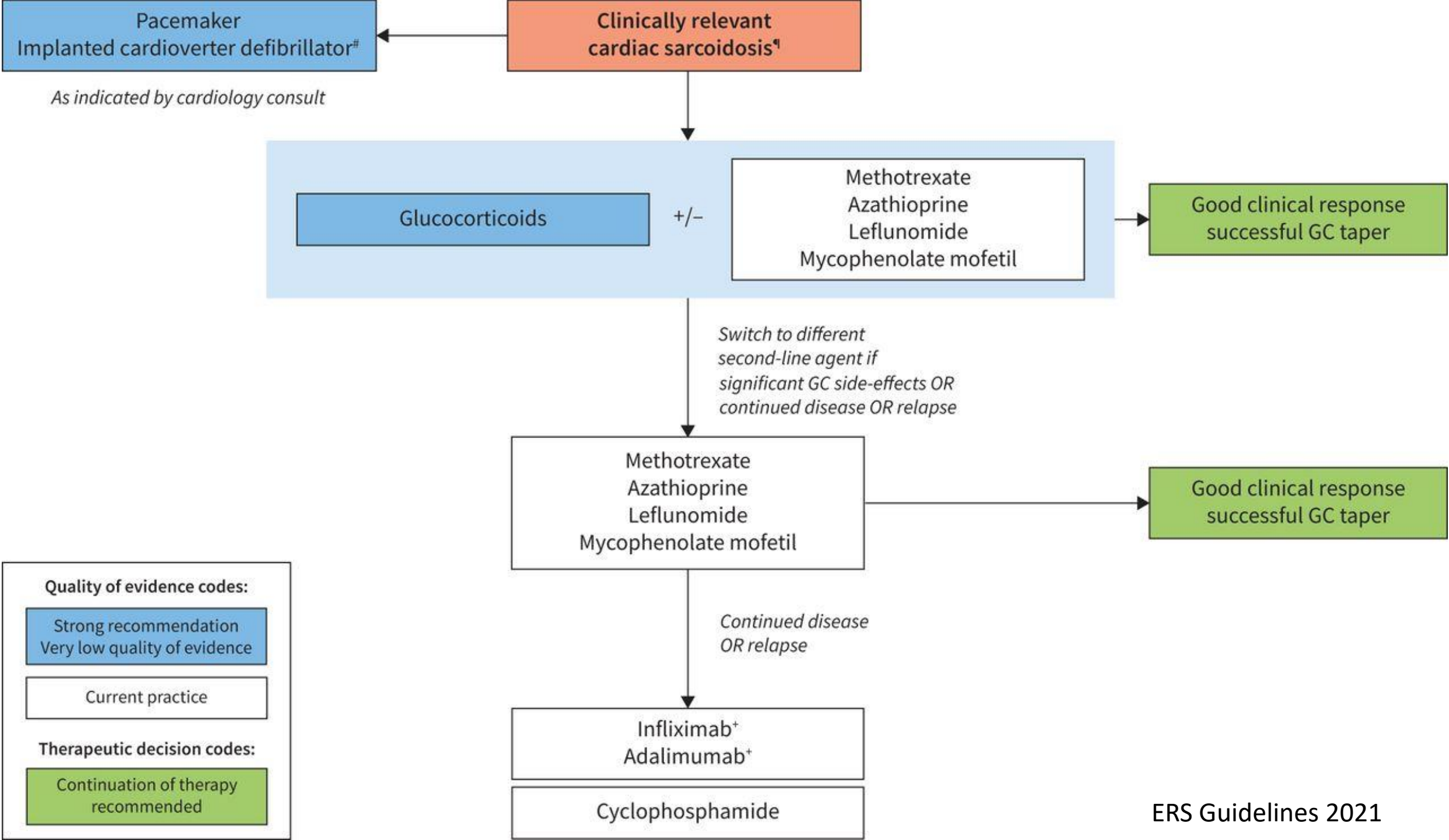| Sarcoidosis		
In patients with cardiac sarcoidosis who have permanent or transient AVB, implantation of a device capable of cardiac pacing should be considered.i	IIa	C
In patients with sarcoidosis and indication for permanent pacing who have LVEF <50%, implantation of a CRT-D should be considered.	IIa	C

4. « Pacing » si troubles conductifs de haut degré.
Et CRT-D >> PM si FEVG < 50%

Pacemaker
Implanted cardioverter defibrillator[#]

As indicated by cardiology consult

Clinically relevant
cardiac sarcoidosis[¶]

Glucocorticoids +/− Methotrexate
Azathioprine
Leflunomide
Mycophenolate mofetil

Good clinical response
successful GC taper

*Switch to different
second-line agent if
significant GC side-effects OR
continued disease OR relapse*

Methotrexate
Azathioprine
Leflunomide
Mycophenolate mofetil

Good clinical response
successful GC taper

*Continued disease
OR relapse*

Infliximab[+]
Adalimumab[+]

Cyclophosphamide

Quality of evidence codes:

Strong recommendation
Very low quality of evidence

Current practice

Therapeutic decision codes:

Continuation of therapy
recommended

ERS Guidelines 2021

Poor Prognostic Indicators in Patients with Cardiac Sarcoidosis[2]

Variable	Prognostic association
Age at diagnosis > 50 years	Increased mortality risk
LVEF < 40% Increased LV end-diastolic diameter Abnormal longitudinal strain on Echocardiography Interventricular septal thinning	Increased mortality risk
Complete heart block and high grade AVB	Increased mortality and as a mechanism of sudden deaths. Increased risk of relapse and less favorable course of CS.
Sustained ventricular tachycardia	Increased mortality risk
Late Gadolinium Enhancement (LGE) on cMRI	Increased risk of cardiovascular death & ventricular arrythmia. Extensive LGE has been associated with lack of improvement in LV function after steroid therapy.
Cardiac inflammation and abnormal perfusion defect identified by positive 18F FDG-PET	Increased risk of ventricular tachycardia & mortality.
Elevated troponin or brain natriuretic peptide (BNP)	Reduced event free survival and increased risk of adverse outcomes.
NYHA Functional class 3 or 4	Increased mortality risk

Baughman 2022

Effect of Immunosuppressive Therapy and Biopsy Status in Monitoring Therapy Response in Suspected Cardiac Sarcoidosis

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Mahesh K. Vidula, MD,[c] Senthil Selvaraj, MD,[c] Jacob Dubroff, MD, PhD,[a] Eliot Peyster, MD,[c] Caitlin B.
Karen Patterson, MD,[d] Francis E. Marchlinski, MD,[c] Milton Rossman, MD,[d] Lee Goldberg, MD,[c] Paco E
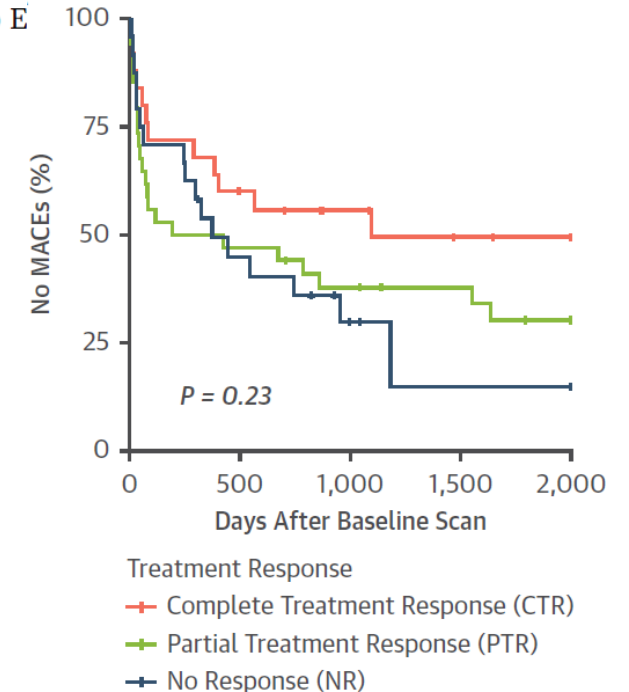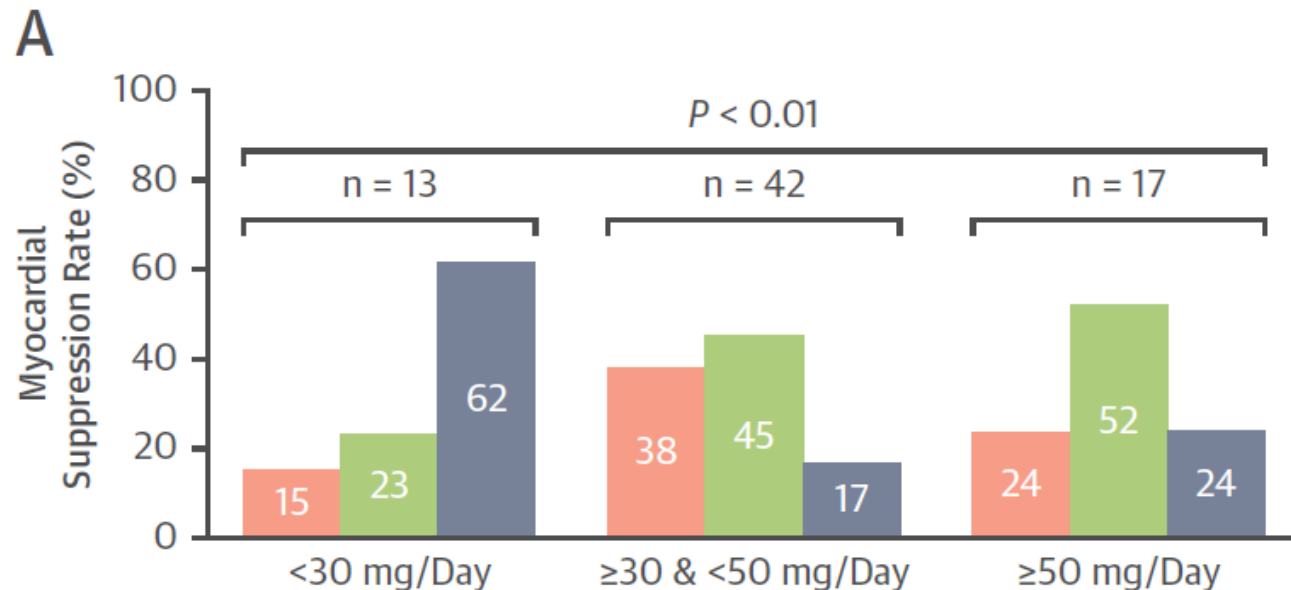
84 patients sans traitement avant le TEP
INTERER DU TEP DANS LE SUIVI +++