Les atteintes cardiaques des sarcoïdoses

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CARDIOCONNECT

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SALONS VARENNE, NOISY-LE-GRAND

Faire équipe

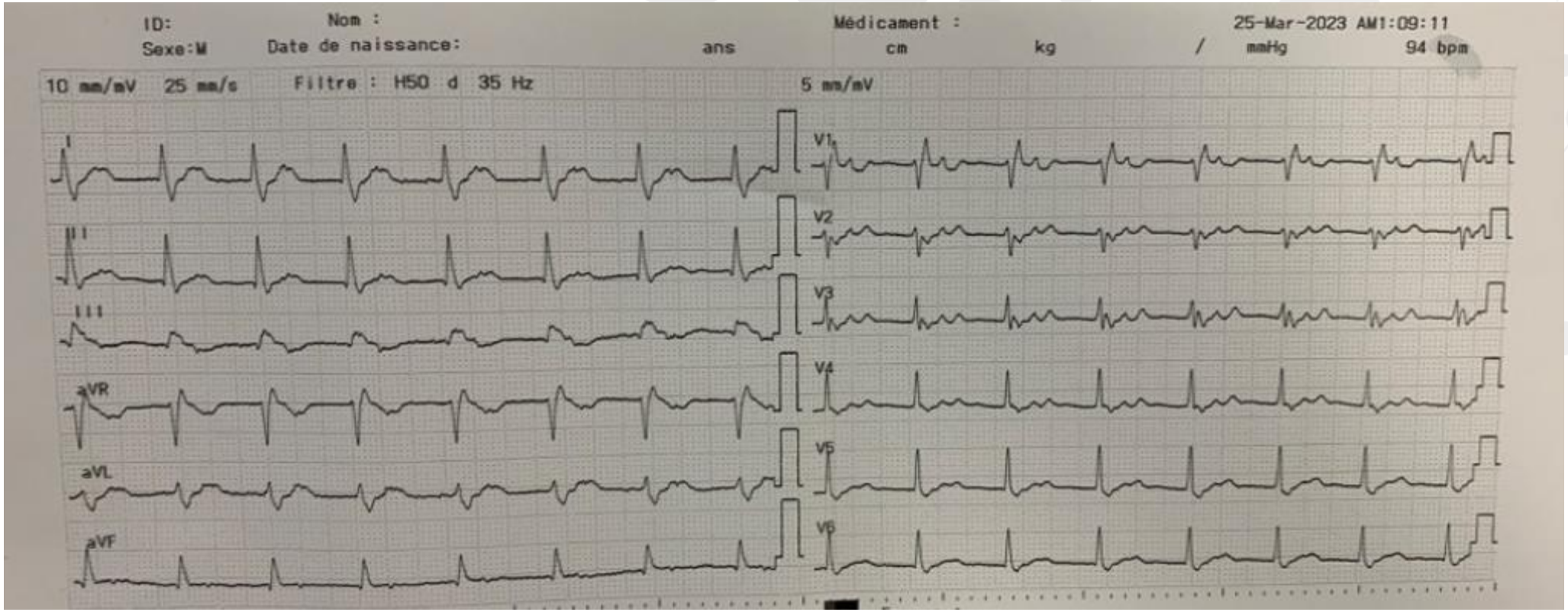
Les atteintes cardiaques de la sarcoïdose

Insuffisance cardiaque

Dr Silvia Oghina

Cas clinique

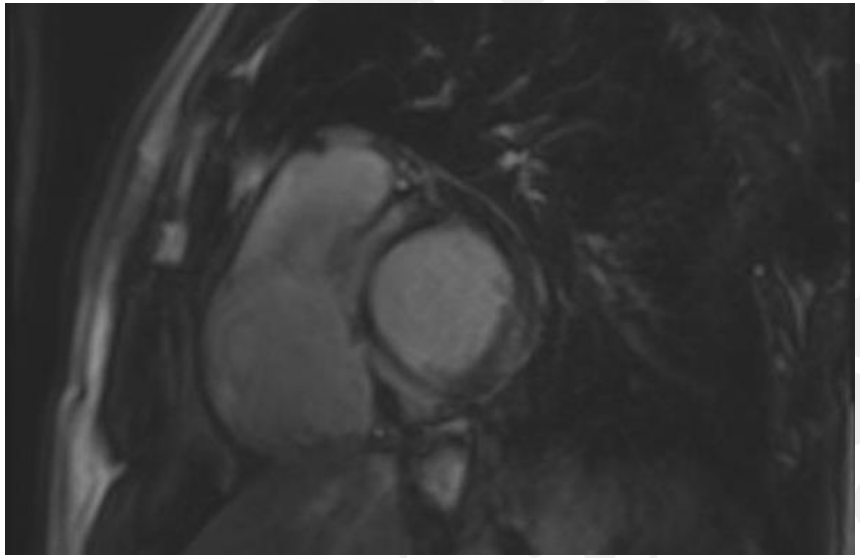
- Jeune homme de 37 ans, très sportif
- Baisse des performances sportives => Cs Cardiologie en ville

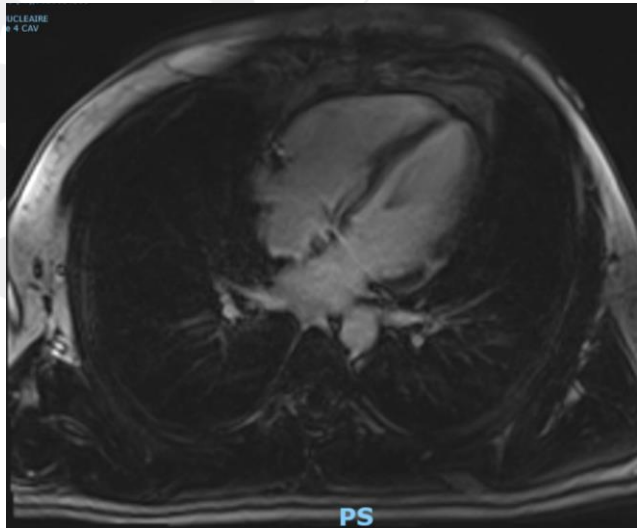


Cas clinique

- Jeune homme de 37 ans, très sportif
- Baisse des performances sportives => Cs Cardiologie en ville

- HolterECG :
  - Rythme sinusal permanent (52 - 137/mn), BAV 1 et 2 avec 440 périodes longues (max 1930 ms)
  - Absence d'hyperexcitabilité supraventriculaire significative.
  - Absence d'hyperexcitabilité ventriculaire significative (38 ESV, un doublet, un trigéminisme)





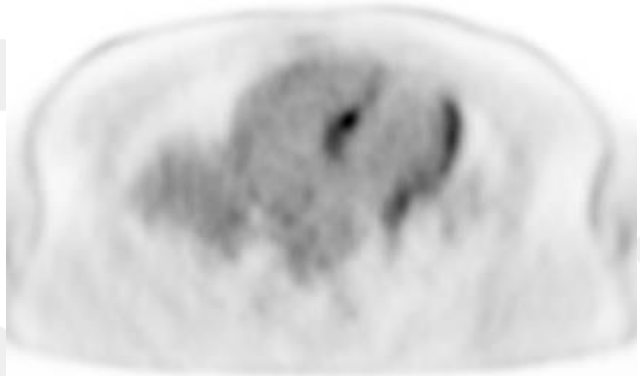
Cas clinique

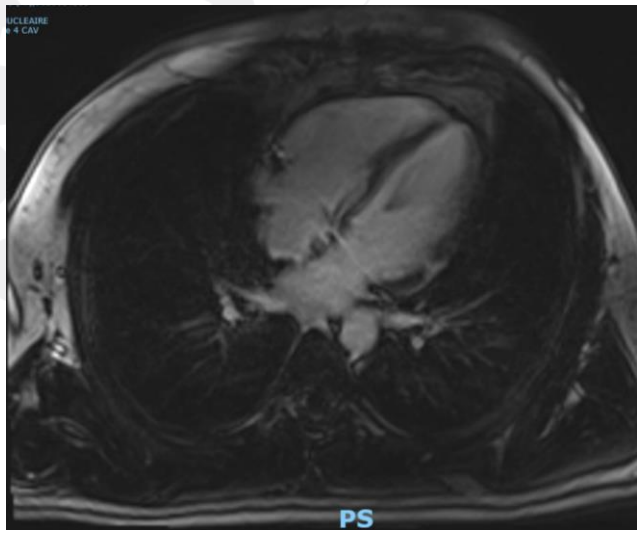
- ETT : VG non dilaté non hypertrophié de bonne fonction systolo-diastolique, FEVG 55 % en SBP, discrète hypokinésie en antéro-septo-basal. Absence de valvulopathie significative. VD non dilaté de bonne fonction longitudinale. Absence d'argument pour une HTP.

- IRM cardiaque : Ventricule gauche non hypertrophié (épaisseur maximale à 12 en antéro septo basal), avec FEVG conservée à 55%. RT à point de départ sous-épicardique, multifocal , plus étendu sur le septum basal et moyen. Œdème en T2. Dysfonction ventriculaire droite avec FEVD à 30%, et prise de contraste de sa paroi latérale évoquant une atteinte du VD associée.

- TEP: Hypermétabolisme intense en mottes du myocarde évoquant en premier lieu une atteinte sarcoïdosique, étendue sur environ 9 segments.

- Scanner TAP : Aspect de bronchopneumopathie avec multiples foyers d'infiltrats micronodulaires infracentimétriques péri-bronchiques de distribution centrale et périphérique prédominant aux lobes supérieurs, confluents par endroit donnant un aspect pseudo-nodulaire.





Qu'est-ce que la sarcoïdose ?

Granulomatose systémique de cause inconnue caractérisée par la présence de granulome épithélioïde et gigantocellulaire sans nécrose caséeuse liée à une **réponse immune exagérée**

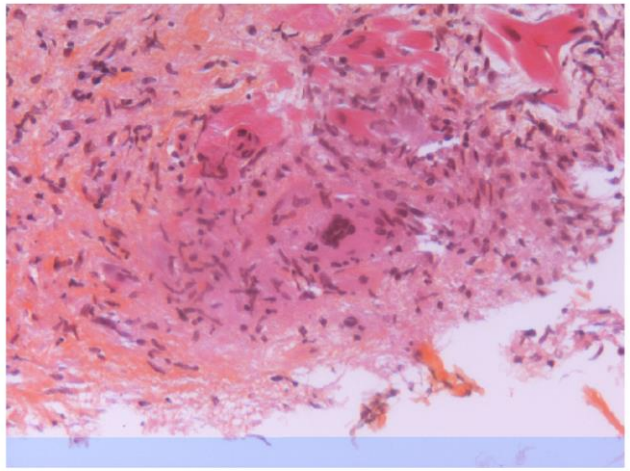
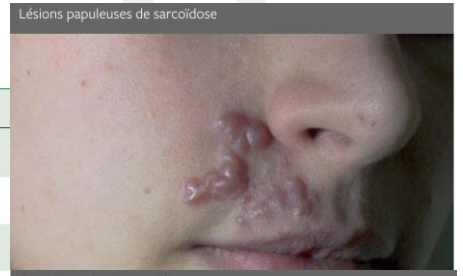


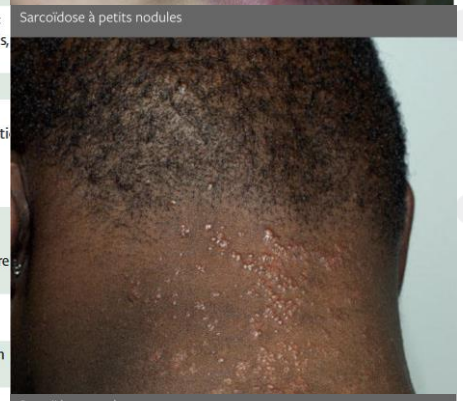
Fig. 1. Lésion histologique de granulome épithélioïde et gigantocellulaire obtenu sur la biopsie endomyocardique d'un patient ayant une insuffisance cardiaque et un aspect échographique de cardiopathie hypertrophique : granulome inflammatoire sans nécrose, composé de cellules épithélioïdes et une cellule géante, au contact de cardiomyocytes dystrophiques (×20).

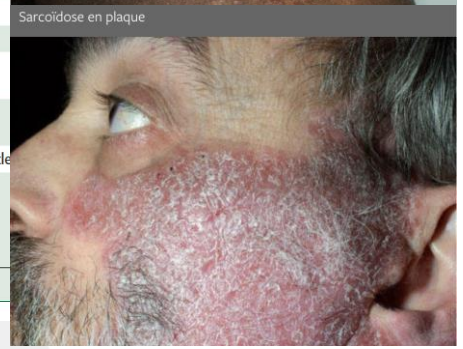
	Prevalence (%)	Symptoms
Skin[56]	~15	Papules, nodules, plaques, scar sarcoidosis, lupus pernio, subcutaneous sarcoidosis
Peripheral lymphadenopathy[1,57]	10–20	Mostly cervical or supraclavicular; inguinal, axillary, epitrochlear, or submandibular lymph node sites also possible; painless and mobile
Eye[58,59]	10–30	Anterior, intermediate, or posterior uveitis; retinal vascular change; conjunctival nodules; lacrimal gland enlargement
Liver[60]	20–30	Often symptom-free; abnormal liver function tests in 20–30% of patients; hepatomegaly; rarely hepatic insufficiency, chronic intrahepatic cholestasis, or portal hypertension
Spleen[57]	~10	Splenomegaly; rarely, pain or pancytopenia; very rarely, splenic rupture
Heart[48,49,61,62]	2–5	Atrioventricular or bundle branch block; ventricular tachycardia or fibrillation; congestive heart failure; pericarditis; impairment of sympathetic nerve activity; sudden death
Nervous system[63-65]	~5	Facial nerve palsy, optic neuritis, leptomeningitis, diabetes insipidus, hypopituitarism, seizures, cognitive dysfunction, deficits, hydrocephalus, psychiatric manifestations, spinal cord disease, polyneuropathy, small-fibre neuropathy
Kidney[66]	0·5–2	Rare symptoms; increased creatininemia sometimes associated with hypercalcaemia; nephrocalcinosis; kidney stones
Parotitis[1]	4%	Symmetrical parotid swelling, Heerfordt's syndrome when associated with uveitis, fever, and facial palsy
Nose[57]	0·5–6	Nasal stuffiness, nasal bleeding, crusting, anosmia
Larynx[57]	0·5–1	Hoarseness, breathlessness, stridor, dysphagia
Bones[67]	<5	Often asymptomatic; hands and feet classically most involved, also large bones and axial skeleton
Skeletal muscles[67]	1	Proximal muscle weakness, amyotrophy, myalgia, intramuscular nodules
Genitourinary tract[1,57]	..	All organs can be involved, including breast, uterus, epididymis, and testicle
Gastrointestinal tract[1]	1	Most often symptom-free, but the oesophagus, stomach, small intestine, and colon can be involved

CSF=cerebrospinal fluid. ^{18}F-FDG=^{18}F-fluorodeoxyglucose.

Table 1: **Extrapulmonary localisations of sarcoidosis**







Valeyre D et al. Lancet. 2014 Mar 29;383(9923):1155-67
Cohen Aubart et al. Rev Med Interne. 2017 Jan;38(1):28-35

Quelles sont les atteintes cardiaques de la sarcoïdose ?

- **Physiopathologie :**

  infiltration hétérogène du myocarde par les granulomes

  ➔ ischémie

  ➔ fibrose myocardique irréversible

- **Localisations préférentielles :**
  - Paroi libre ventriculaire gauche
  - Septum inter-ventriculaire dont voies de conduction
  - **Atteinte du VD pronostique** (peut mimer une DVDA !!)
- Signes cliniques, électriques ou échocardiographiques :
  - pas proportionnels au degré d'infiltration
  - **dépendent de la localisation des granulomes et de la formation éventuelle de cicatrices fibreuses**
- **Manifestations :**

  Asymptomatique (découverte histologique ou morphologique)

  Troubles du rythme

  Insuffisance cardiaque

  Troubles de la conduction

Table I. Prevalence of various cardiac findings in CS during course of disease

	Prevalence in study series
AV block	26%-62%
BBB	12%-61%
SVT	0%-15%
V-Tach	2%-42%
CHF	10%-30%
SD	12%-65%

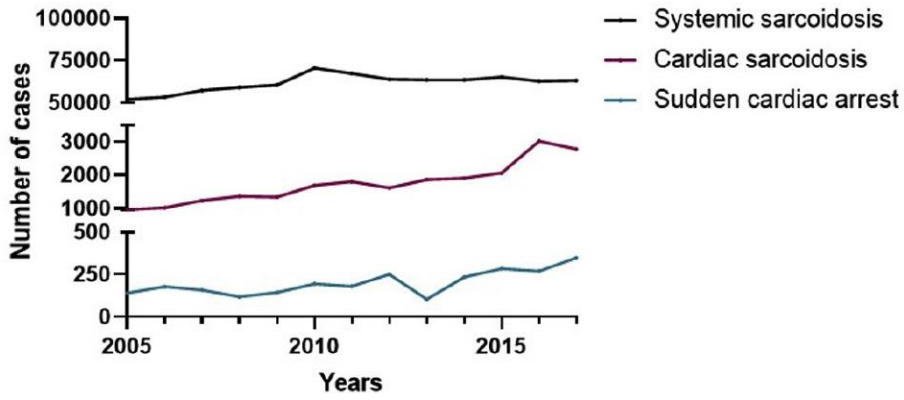
Data from references 1,26,29-32.
AV, Atrioventricular; *BBB*, bundle-branch block; *SVT*, supraventricular tachycardia; *V-Tach*, ventricular tachycardia; *CHF*, congestive heart failure; *SD*, sudden death.
See text for details.

Kim et al. Am Heart J. 2009 Jan;157(1):9-21.
Cohen Aubart et al. Rev Med Interne. 2017 Jan;38(1):28-35

Quelle est la prévalence de l'atteinte cardiaque ?

2 à 75 % selon les critères utilisés

FIGURE 2 Temporal Trends in Sarcoidosis and Sudden Cardiac Arrest



Over a 13-year period, a relatively stable number of systemic sarcoid patients were noted with a progressive increase in the rates of cardiac sarcoidosis and sudden cardiac arrest.

Tableau 1
Prévalence de la sarcoïdose cardiaque.

	Origine	Critères	Prévalence (%)	Nombre de sujets
Baughman et al., 2001 [58]	États-Unis (multicentrique)	Critères ACCESS	2,3	736
Johns, 1999	États-Unis (monocentrique)	Cohorte de sarcoïdoses chroniques	7	181
Smedema et al., 2005 [38]	Pays-Bas (multicentrique)	Dépistage systématique dans une cohorte de sarcoïdoses pulmonaires	19	101
Silverman et al., 1978 [15]	États-Unis (monocentrique)	Autopsies consécutives de sarcoïdoses	27	84
Sharma et al., 1993 [13]	États-Unis (monocentrique)	Autopsies consécutives de sarcoïdoses	20	123
Iwai et al., 1993 [4]	Japon (données nationales)	Autopsies consécutives de sarcoïdoses	66	320
Perry et Vuitch, 1995 [14]	États-Unis (monocentrique)	Autopsies consécutives de sarcoïdoses	75	38

Narasimhan et al. JACC Clin Electrophysiol. 2021 Sep;7(9):1087-1095
Cohen Aubart et al. Rev Med Interne. 2017 Jan;38(1):28-35

Quand suspecter une sarcoïdose cardiaque ?

Découverte de granulomes sur une biopsie myocardique ou un cœur explanté

En cas de sarcoïdose extracardiaque :

- ECG (Se 25%)
- HolterECG (Se 50%)
- ETT
- IRM
- TEP TDM (ou TEP IRM)

Section A: Diagnosis of CS *Expert Consensus Recommendations on* ***Criteria for the Diagnosis of CS***

There are two pathways to a diagnosis of CS:

1. Histological diagnosis from myocardial tissue CS is diagnosed in the presence of noncaseating granuloma on histological examination of myocardial tissue with no alternative cause identified (including negative organismal stains if applicable).
2. Clinical diagnosis from invasive and noninvasive studies

   It is probable* that CS is present if:

   a) There is a histological diagnosis of extracardiac sarcoidosis

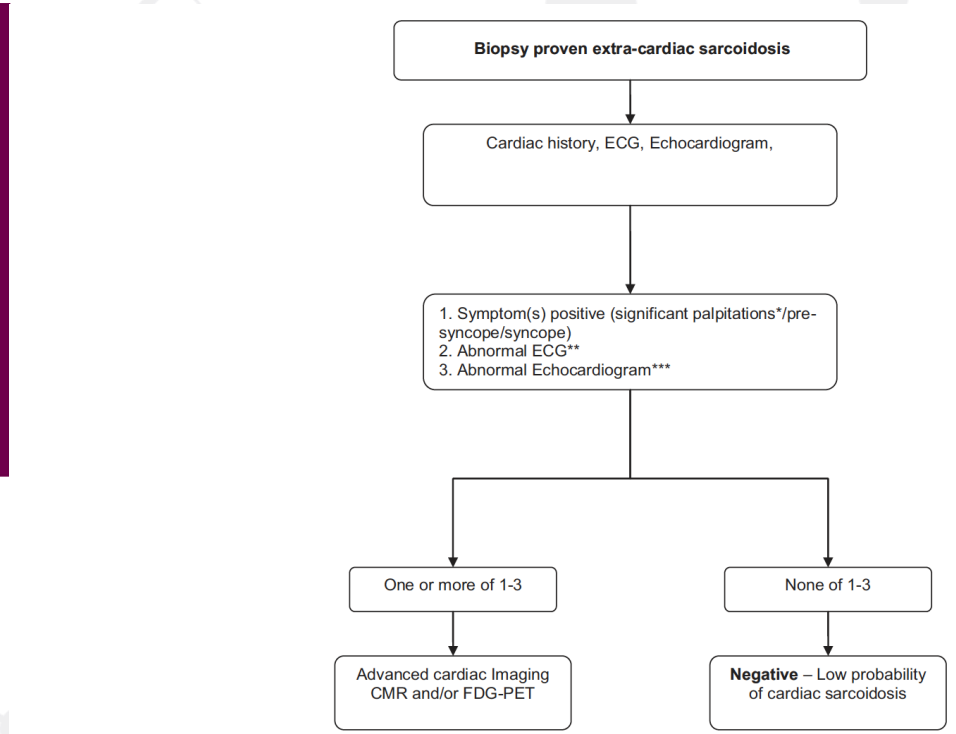
   and

   b) One or more of the following is present:
   - Corticosteroid- and/or immunosuppressant-responsive cardiomyopathy or heart block
   - Unexplained reduced LVEF (<40%)
   - Unexplained sustained (spontaneous or induced) ventricular tachycardia
   - Mobitz type II second-degree heart block or third-degree heart block
   - Patchy uptake on dedicated cardiac positron emission tomography (PET; in a pattern consistent with CS)
   - Late gadolinium enhancement (LGE) on cardiovascular magnetic resonance (CMR; in a pattern consistent with CS)
   - Positive gallium uptake (in a pattern consistent with CS)

   and

   c) Other causes for the cardiac manifestation(s) have been reasonably excluded.

*In general, "probable involvement" is considered adequate to establish a clinical diagnosis of CS.[33]



* palpitations were defined as "prominent patient complaint lasting > 2 weeks[25]"
** abnormal ECG defined as complete left or right bundle branch block and/or presence of unexplained pathological Q waves in 2 or more leads and/or sustained 2nd or 3rd degree AV block and/or sustained or non-sustained VT[25]
*** abnormal echocardiogram defined as RWMA and/or wall aneurysm and/or basal septum thinning and/or LVEF < 40%*[25]

Figure 3 Suggested algorithm for the investigation of patients with biopsy-proven extracardiac sarcoidosis. AV = atrioventricular; CMR = cardiovascular magnetic resonance; ECG =electrocardiogram; FDG-PET = ^{18}F-fluorodeoxyglucose–positron emission tomography; LVEF = left ventricular; RWMA = regional wall motion abnormality; VT = ventricular tachycardia.

Cohen Aubart et al. Rev Med Interne. 2017 Jan;38(1):28-35
Birnie et al. Heart Rhythm. 2014 Jul;11(7):1305-23.

Guidelines for Diagnosis of Cardiac Sarcoidosis from the Japanese Ministry of Health and Welfare.

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En l'absence de sarcoïdose connue si tableau cardiologique évocateur :

- BAV < 55 ans
- « myocardite » récidivante ou d'évolution atypique
- dysplasie arythmogène du ventricule droit
- myocardiopathie non ischémique et non familiale
- Eléments cliniques d'orientation : examen clinique avec peau (cicatrices), adénopathies et TDM thoracique (micronodules de topographie lymphatique ou adénopathies médiastinales de localisation hilaire souvent bilatérales et symétriques, normal dans < 10% des cas)

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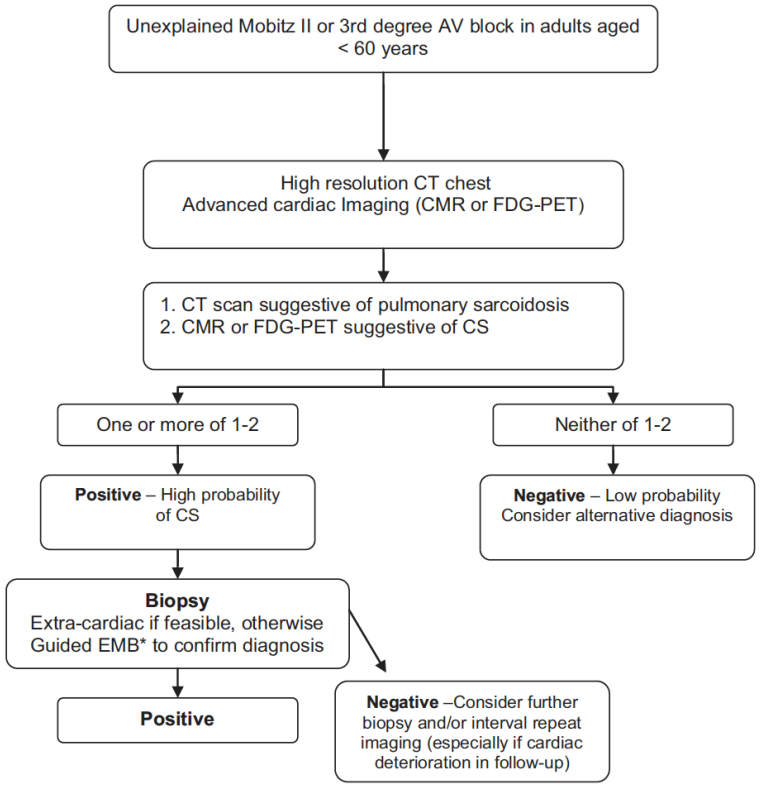

*voltage guided or advanced imaging guided endomyocardial biopsy (see text in Section B for details)

Figure 4 Suggested algorithm for the investigation of patients with unexplained Mobitz II or third-degree AV block who are younger than 60 years. AV = atrioventricular; CMR = cardiovascular magnetic resonance; CS = cardiac sarcoidosis; CT = computed tomographic; ECG =electrocardiogram; EMB = endomyocardial biopsy; FDG-PET = ^{18}F-fluorodeoxyglucose–positron emission tomography.

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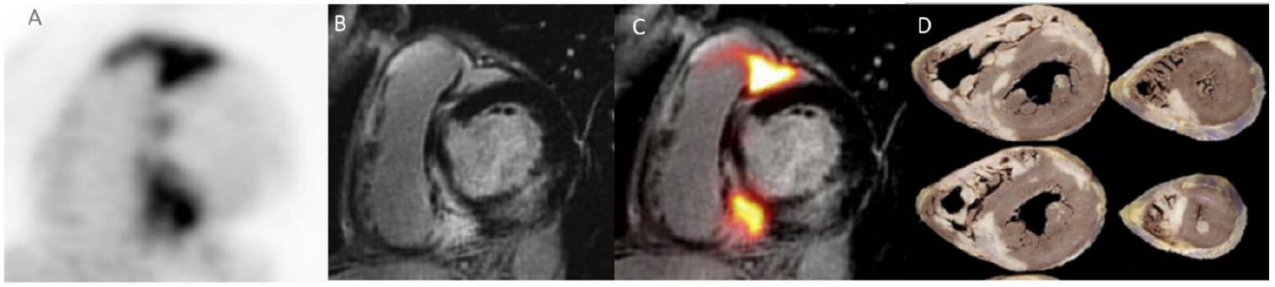
Cohen Aubart et al. Rev Med Interne. 2017 Jan;38(1):28-35
Birnie et al. Heart Rhythm. 2014 Jul;11(7):1305-23.

Guidelines for Diagnosis of Cardiac Sarcoidosis from the Japanese Ministry of Health and Welfare.

Comment diagnostiquer une sarcoïdose cardiaque ?

- **ETT :** VG dilaté, anomalies segmentaires, amincissement de paroi
- **IRM cardiaque peut montrer 3 types d'anomalies :**
  - Anomalies cinétiques et morphologiques (épaississement ou amincissement focal, dilatation ventriculaire)
  - Œdème intra-myocardique (hyper-signal T2) : infiltration granulomateuse potentiellement réversible
  - Prises de contraste tardives (typiquement sous-épicardiques linéaires ou nodulaires, transmurales, hétérogènes au sein de la paroi) : fibrose non réversible
- **18FDG-TEP TDM :**
  - après un régime pauvre en glucides et saturé en lipides permettant de supprimer l'hypermétabolisme physiologique myocardique
  - sensibilité de 89 % (79–100 %)et une spécificité de 78 % (38–100 %)



Enzyme de conversion ?

- Se 22-83%, Sp faible
- Peu utilisé en pratique car faibles performances diagnostiques

Cohen Aubart et al. Rev Med Interne. 2017 Jan;38(1):28-35

Trivieri et al. J Am Coll Cardiol. 2020 Oct 20;76(16):1878-1901.

Quelle prise en charge étiologique ?

- **Stratégie thérapeutique limitée par l'absence d'étude contrôlée**
- **Principes du traitement :**

1. L'immunosuppression (CTC 30 mg ou 1mg/kg/j pendant 24 mois +/- MTX +/- antiTNF dans les formes réfractaires) :
L'objectif de l'immunosuppression est de supprimer les lésions granulomateuses actives mais également d'empêcher l'évolution vers des cicatrices fibreuses.

- Efficacité sur BAV / FEVG stabilisée ou améliorée surtout si > 50%
- **Mais** troubles du rythme liés aux cicatrices fibreuses donc peu sensibles aux CTC

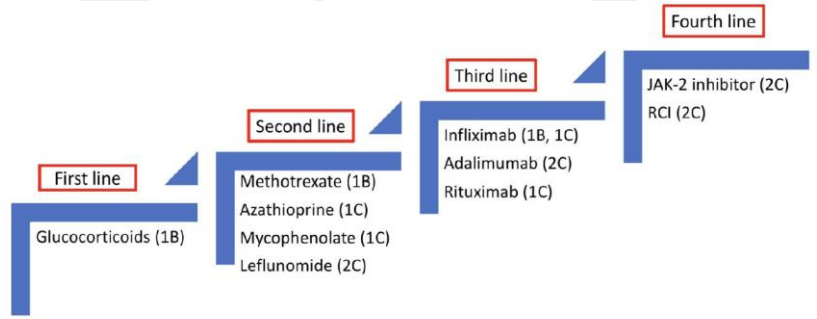


Figure 9: Stepwise approach to the therapy for CS.
Treatments were scored based on reported effectiveness for cardiac (1) or extra-cardiac (2) sarcoidosis. Level of evidence: A for randomized trials, B for case series or C for case reports. There is presently no randomized trial in CS. Initiation and escalation of treatment is commonly guided by PET.

2. La prise en charge de l'insuffisance cardiaque ou des troubles du rythme.

L'indication du DAI doit se faire indépendamment du traitement immunomodulateur et sans délai lorsque l'indication est retenue

Takaya et al. Am JCardiol 2015;115(4):505–9.
Birnie et al. Heart Rhythm 2014;11(7):1305–23.
Schuller et al. JCardiovasc Electrophysiol 2012;23(9):925–9

Quel est le risque rythmique de la sarcoïdose cardiaque ?

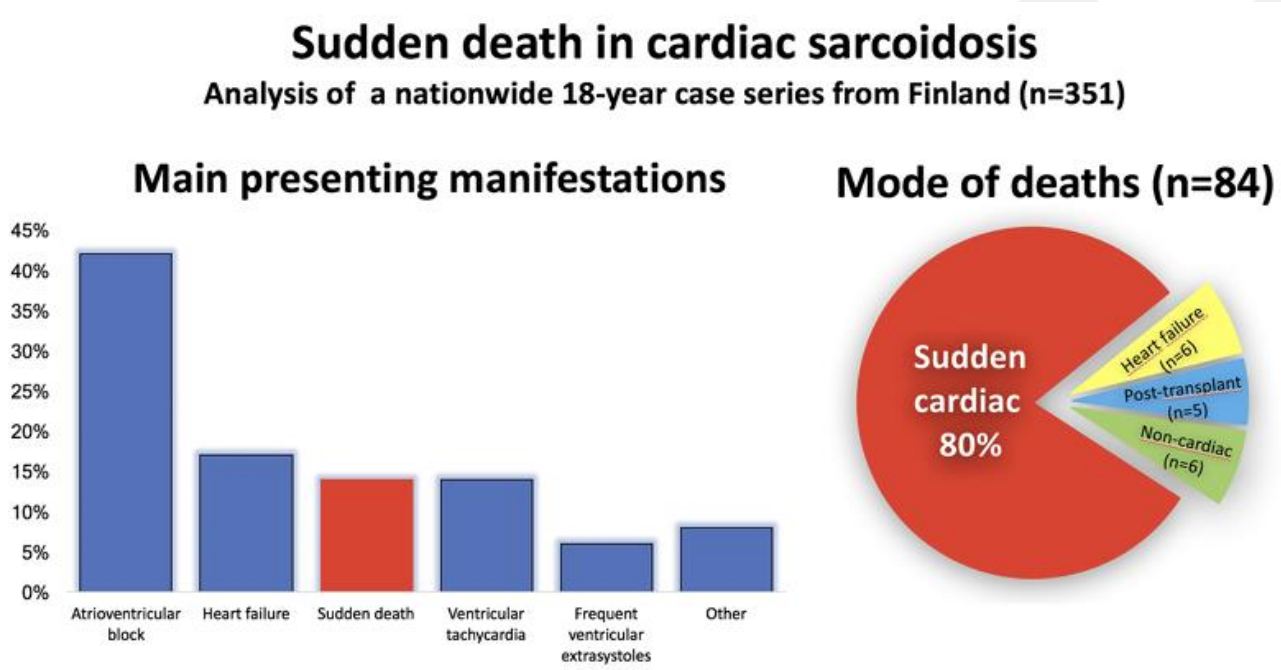


TABLE 2 Predictors of SCA in Sarcoidosis

Characteristic	Multivariate OR (95% CI)	p Value
Age		
<40	1.012 (1.006–1.02)	0.001
>80	0.74 (0.57–0.96)	0.03
Female	0.79 (0.68–0.92)	0.003
Smoking	0.83 (0.69–0.99)	0.046
African American	1.63 (1.39–1.92)	0.001
HFpEF	1.88 (1.48–2.39)	0.001
HFrEF	2.37 (1.77–3.17)	0.001
Atrial fibrillation	1.69 (1.39–2.06)	0.001
AV disturbances	3.82 (2.59–5.61)	0.001
1st degree AVB	-	-
2nd degree AVB	3.22 (1.32–7.86)	0.01
3rd degree AVB	7.12 (4.38–10.57)	0.001
Conduction disturbances	5.25 (3.45–7.97)	0.001
RBBB	2.59 (1.65–4.08)	0.001
LBBB	2.13 (1.13–4.02)	0.02
Fascicular block	2.75 (1.35–5.59)	0.005

Multivariate analysis: Adjusted for age, sex, race, insurance and hospital size, Charlson Comorbidity Index.

AVB = atrioventricular block; CI = confidence interval; HFpEF = heart failure with preserved ejection fraction; HFrEF = heart failure with reduced ejection fraction; LBBB = left bundle branch block; OR = odds ratio; RBBB = right bundle branch block; SCA = sudden cardiac arrest.

Ekström et al. Eur Heart J. 2019 Oct 1;40(37):3121-3128.
Narasimhan et al. JACC Clin Electrophysiol. 2021 Sep;7(9):1087-1095.

Comment stratifier le risque rythmique de la sarcoïdose cardiaque ?

IRM cardiaque: rehaussement tardif = fibrose non réversible

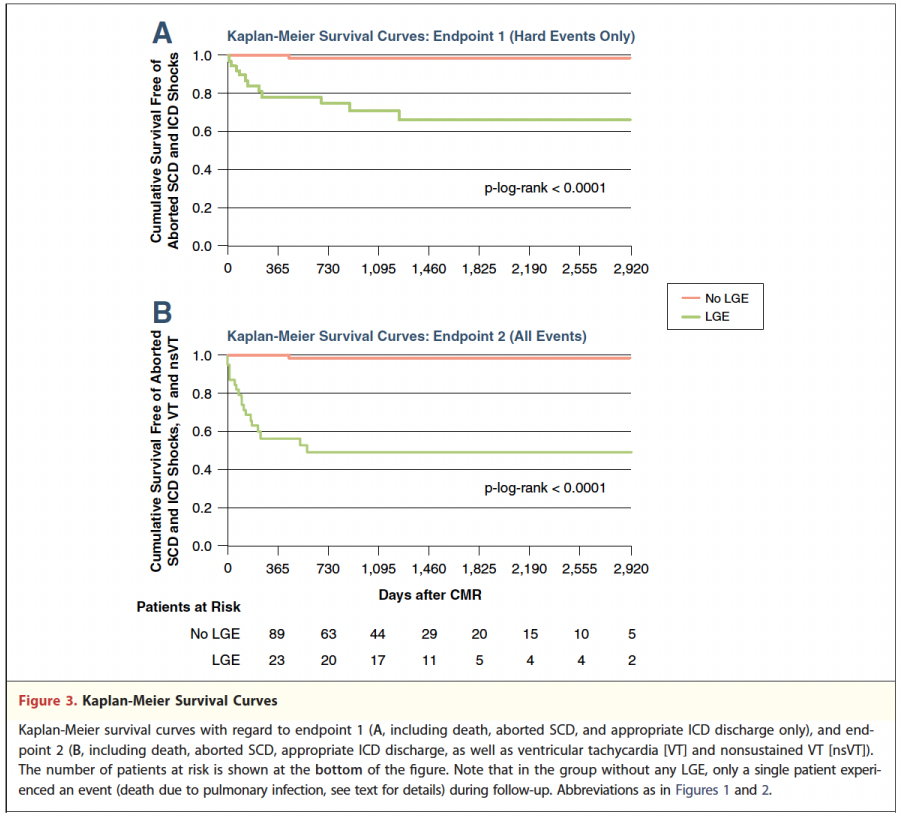


Figure 3. Kaplan-Meier Survival Curves

Kaplan-Meier survival curves with regard to endpoint 1 (**A**, including death, aborted SCD, and appropriate ICD discharge only), and endpoint 2 (**B**, including death, aborted SCD, appropriate ICD discharge, as well as ventricular tachycardia [VT] and nonsustained VT [nsVT]). The number of patients at risk is shown at the **bottom** of the figure. Note that in the group without any LGE, only a single patient experienced an event (death due to pulmonary infection, see text for details) during follow-up. Abbreviations as in Figures 1 and 2.

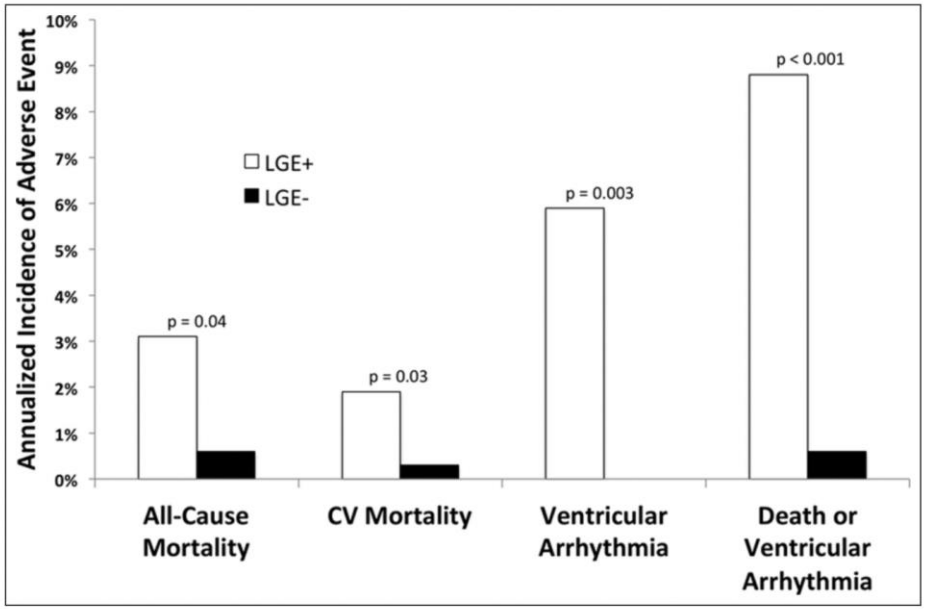


Figure 2. Annualized event rates according to late gadolinium enhancement (LGE) positive vs negative. CV indicates cardiovascular.

Trivieri et al. J Am Coll Cardiol. 2020 Oct 20;76(16):1878-1901.
Greulich et al. JACC Cardiovasc Imaging. 2013 Apr;6(4):501-11.

Comment stratifier le risque rythmique de la sarcoïdose cardiaque ?

PET 18 FDG [= activité inflammatoire] et 82 Rubidium [défaut de perfusion = séquelle]

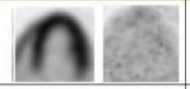
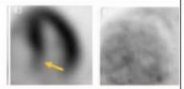
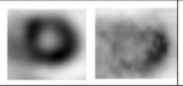
Rest Perfusion	FDG	Frequency	Example		Interpretation / Comment
			Perfusion	FDG	
Normal perfusion and metabolism					
Normal	Normal (negative)	32 (27%)			Normal
Normal	Diffuse (non-specific)	15 (12%)			Diffuse FDG most likely due to failure to suppress FDG from normal myocardium,.
Abnormal perfusion or metabolism					
Normal	Focal	20 (17%)			Nonspecific pattern ; focal increase in FDG may represent early disease vs. normal variant
Positive	Negative	17 (14%)			Rest perfusion defect may represent scar from cardiac sarcoidosis or other etiologies
Abnormal perfusion and metabolism					
Positive	Focal increase ("mismatch pattern")	23 (19%)			Presence of active inflammation ± scar in the same location
Positive	Focal on diffuse	6 (5%)			Similar to above but also areas of inability to suppress FDG from normal myocardium vs. diffuse inflammation
Positive	Focal increase (different area)	5 (4%)			Presence of both scar and inflammation but in different segments

Figure 1. Classification of cardiac PET/CT perfusion and metabolism imaging
Normal perfusion and metabolism (Category 1), abnormal perfusion or metabolism, (Category 2), abnormal perfusion and metabolism (Category 3).

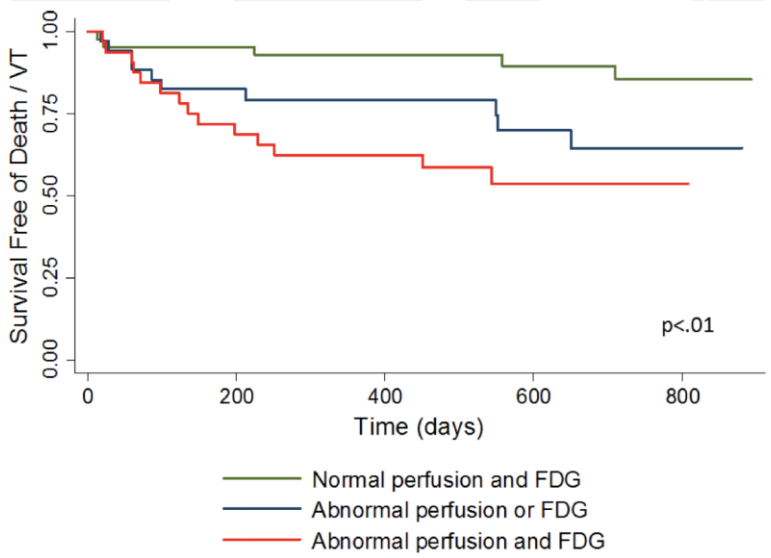


Figure 4. Survival free of death or VT stratified by cardiac PET exam results
Survival free of death or VT stratified by cardiac PET exam results.

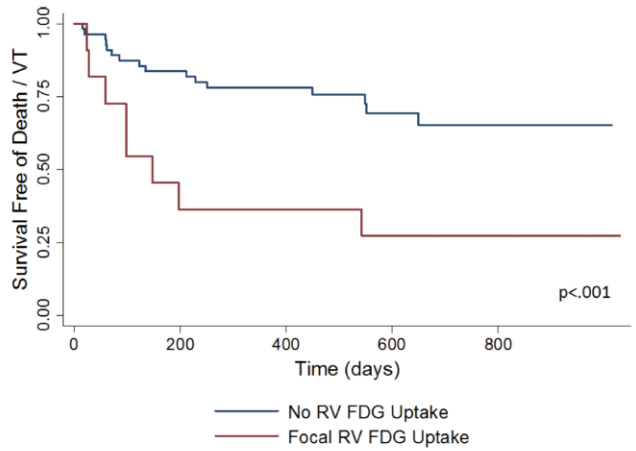


Figure 5. Survival free of death or VT stratified by focal RV inflammation
Survival free of death or VT stratified by the presence or absence of focal right ventricular FDG uptake among individuals with abnormal cardiac PET exam.

Blankstein et al. J Am Coll Cardiol. 2014 Feb 4;63(4):329-36.

Comment stratifier le risque rythmique de la sarcoïdose cardiaque ?

TEP IRM

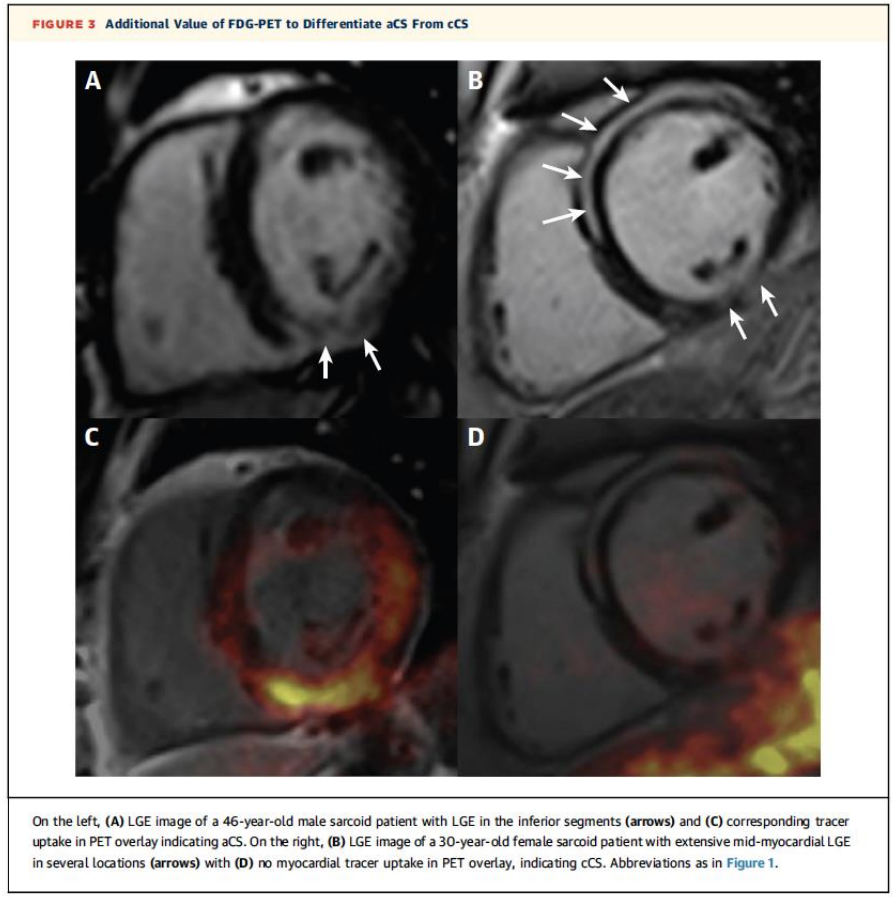
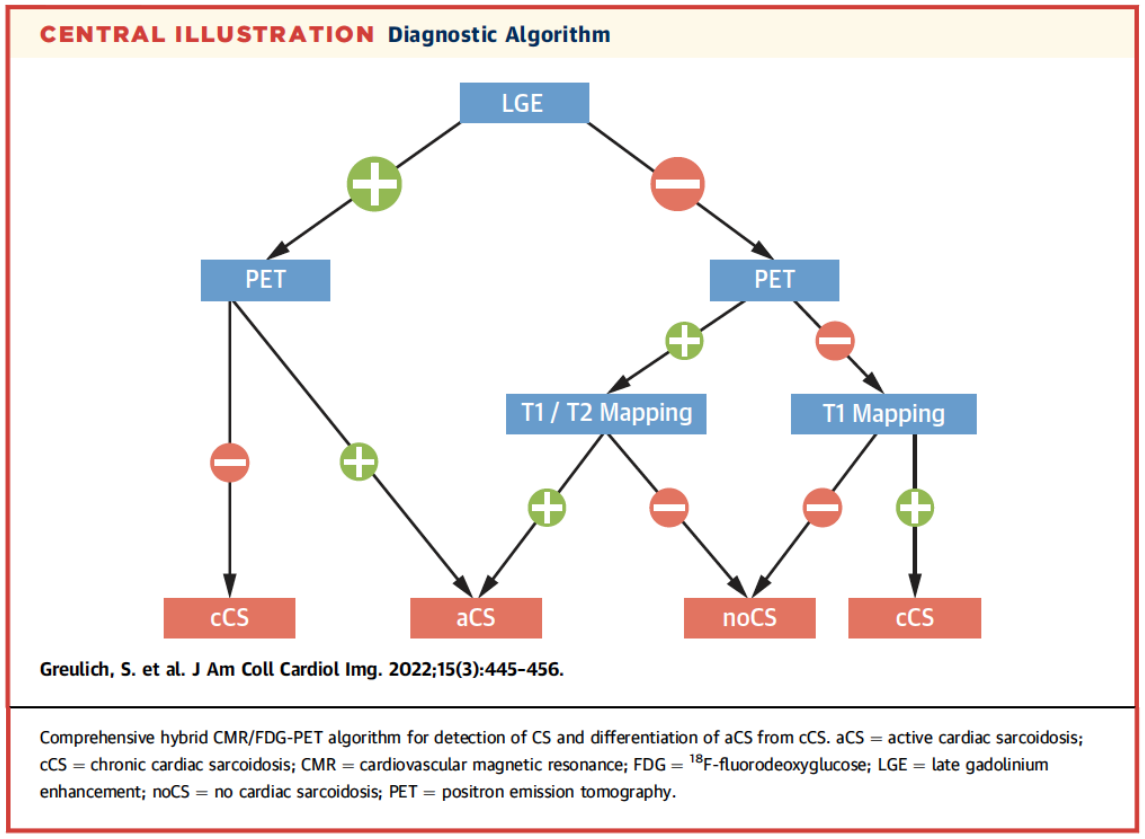


FIGURE 3 Additional Value of FDG-PET to Differentiate aCS From cCS

On the left, **(A)** LGE image of a 46-year-old male sarcoid patient with LGE in the inferior segments **(arrows)** and **(C)** corresponding tracer uptake in PET overlay indicating aCS. On the right, **(B)** LGE image of a 30-year-old female sarcoid patient with extensive mid-myocardial LGE in several locations **(arrows)** with **(D)** no myocardial tracer uptake in PET overlay, indicating cCS. Abbreviations as in Figure 1.



Greulich et al. JACC Cardiovasc Imaging. 2022 Mar;15(3):445-456.

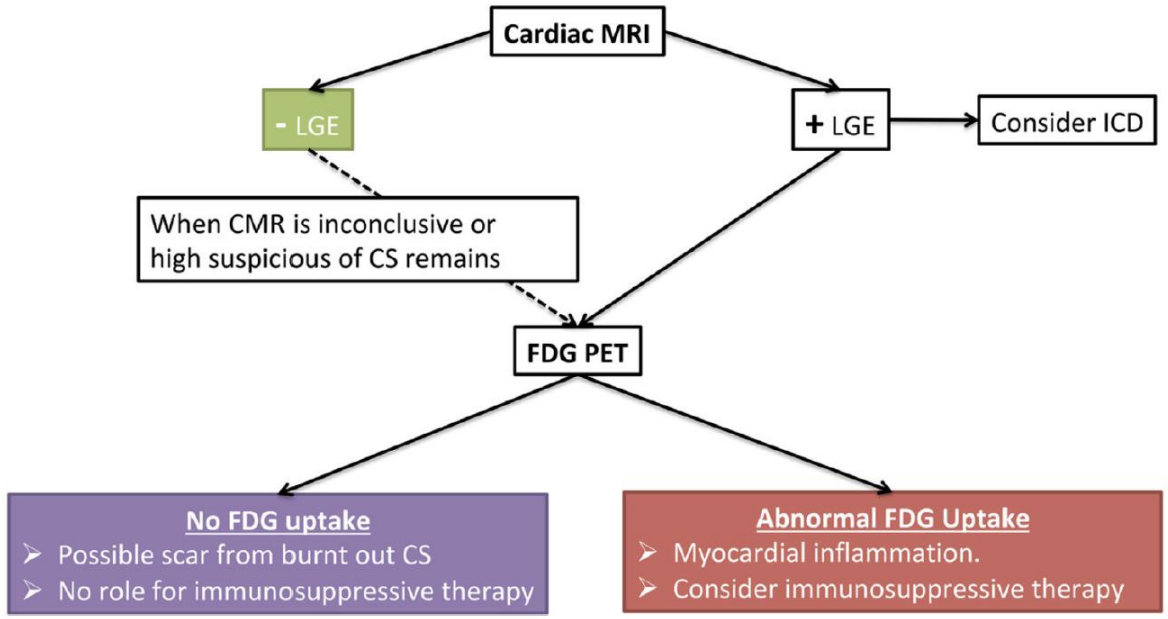
Comment stratifier le risque rythmique de la sarcoïdose cardiaque ?

TEP IRM

Observed Findings

	No LGE	Abnormal LGE No FDG Uptake	Abnormal LGE and FDG Uptake
Treatment:	▪ Δ Immunosup Tx = 1 ▪ ICD = 7 ▪ Pacemaker = 2	▪ Δ Immunosup Tx = 1 ▪ ICD = 13 ▪ Pacemaker = 3	▪ Δ Immunosup Tx = 18 ▪ ICD = 45 ▪ Pacemaker = 9
Annual Event Rate:	5%	13%	12%

Combine Use of Cardiac MRI and PET for Suspected Cardiac Sarcoidosis



Vita et al. Circ Cardiovasc Imaging. 2018 Jan;11(1):e007030.

Que nous disent les recommandations ?

ESC European Society of Cardiology
European Heart Journal (2022) 43, 3997–4126
https://doi.org/10.1093/eurheartj/ehac262

ESC GUIDELINES

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

…ation Table 35 — Recommendations for …tion, sudden cardiac death prevention, …t of ventricular arrhythmias in cardiac

Recommendations	Class[a]	Level[b]
Risk stratification and primary prevention of SCD		
ICD implantation is recommended in patients with cardiac sarcoidosis who have a LVEF ≤35%.[812,828–830,832]	I	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.[816]	IIa	C
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.[817–819,821,833,834]	IIa	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.	IIa	C
In patients with cardiac sarcoidosis, LVEF 35–50% and inducible SMVT at PES, ICD implantation should be considered.[823–825]	IIa	C

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

ESC European Society of Cardiology
European Heart Journal (2021) 00, 1–94
doi:10.1093/eurheartj/ehab364

ESC GUIDELINES

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%

Questions non résolues

- **Temporalité entre introduction du traitement médical IC et implantation du DAI ?**
  - Mais, si rehaussement tardif = séquelle de fibrose irréversible (lit du risque rythmique) : risque rythmique indépendant de la FEVG post titration ?
- **Efficacité du traitement immunosuppresseur sur :**

1. Les troubles conductifs ?

17 BAV complet traités par CTC
Récupération pour 7/17
Mais pas de modification du pronostic CV

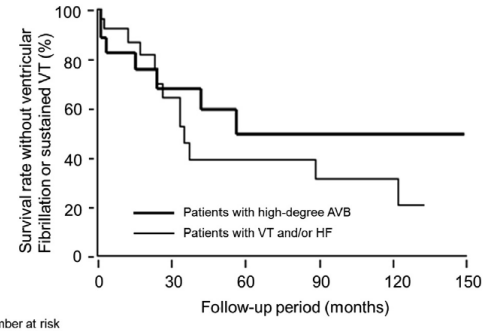


Figure 3. Survival rate without ventricular fibrillation or sustained VT in patients with high-degree AVB and in those with VT and/or HF, among patients treated with prednisone.

2. La FEVG ?

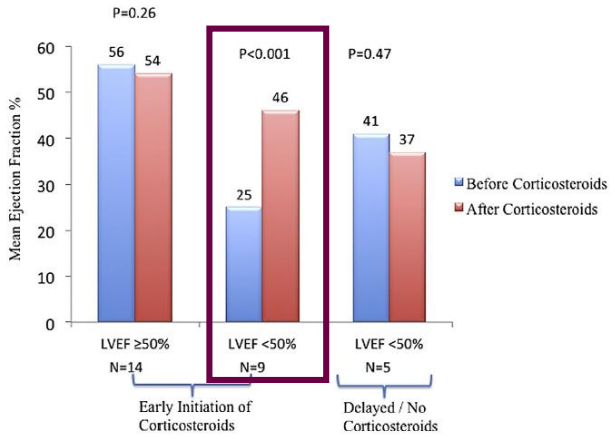


Fig. 1. Impact of corticosteroid therapy on left ventricular ejection fraction in patients with cardiac sarcoidosis.

Amélioration de la dysfonction VG si traitement précoce

3. Le risque rythmique ?

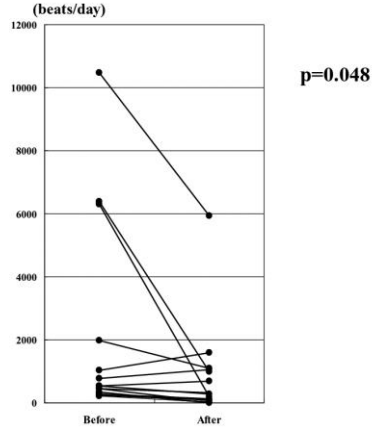


Figure 1. In the less advanced LV dysfunction group, the number of PVCs was significantly decreased after corticosteroid therapy.

Moins de complexes ventriculaires prématurés si FEVG > 35% après CTC (pas de différence si FEVG <35%)

Takaya et al. Am J Cardiol. 2015 Feb 15;115(4):505-9.
Padala et al. Int J Cardiol. 2017 Jan 15;227:565-570.
Yodogawa K, et al. Ann Noninvasive Electrocardiol. 2011.

Questions non résolues

- **Temporalité entre introduction du traitement médical IC et implantation du DAI ?**
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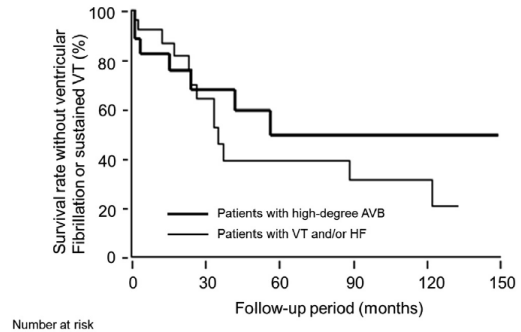

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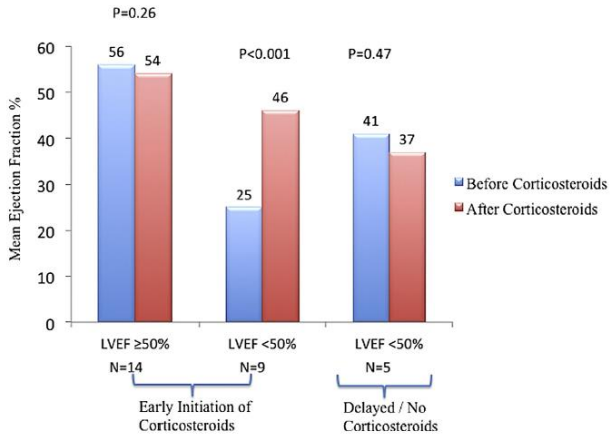

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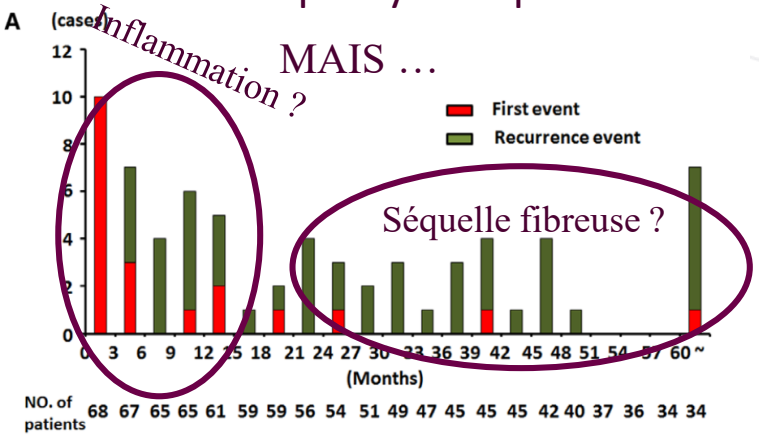

Risque d'évènement rythmique ventriculaire à long terme !!

Takaya et al. Am J Cardiol. 2015 Feb 15;115(4):505-9.
Padala et al. Int J Cardiol. 2017 Jan 15;227:565-570.
Segawa et al. Circ Arrhythm Electrophysiol. 2016 Jun;9(6):e003353.

Quelle place pour le gilet de défibrillation ?

10 choc sur 46
Survie post choc : 100%

Table 1. Baseline characteristics of cardiac sarcoidosis patients.

	All Patients (N = 46)	Shocked Patients (N = 10)	Not Shocked Patients (N = 36)	p-value
Demographic				
Male, N (%)	24 (52%)	6 (60%)	18 (50%)	0.73
Age, median [IQR]	48 [41–55]	47 [41–52]	48 [42–56]	0.79
Co-Morbidity, N (%)				
Congestive Heart Failure	22 (48%)	7 (70%)	15 (42%)	0.16
Ventricular Arrhythmia	19 (41%)	4 (40%)	15 (42%)	1
Syncope	15 (33%)	2 (20%)	13 (36%)	0.46
Heart Block	8 (17%)	1 (10%)	7 (19%)	0.66
Coronary Artery Disease	5 (11%)	2 (20%)	3 (8%)	0.30
LVEF, median [IQR]				
LVEF (%)	30 [21–58]	21 [20–29]	48 [25–60]	$7.0x10^{-3}$

Abbreviations: Interquartile Range, IQR; Left Ventricular Ejection Fraction, LVEF; Number, N.

Table 2. Wearable cardioverter defibrillator experience.

	All Patients (N = 46)	Shocked Patients (N = 10)	Not Shocked Patients (N = 36)	p-value
Wear Summary				
Total Days of Wear, median [IQR]	33 [23–66]	42 [14–66]	33 [23–64]	0.81
Daily Use (Hours), median [IQR]	23.6 [22.4–23.9]	23.8 [23.6–23.9]	23.3 [21.4–23.9]	0.09
End of Use Reason, N (%)				
Received ICD	23 (50%)	7 (70%)	16 (44%)	0.28
LVEF Improved	7 (15%)	0 (0%)	7 (19%)	0.32
Deceased	3 (7%)	1 (10%)	2 (6%)	0.53
Other Non-Cardiac	13 (28%)	2 (20%)	11 (31%)	0.70

Abbreviations: Implantable Cardioverter Defibrillator, ICD; Interquartile Range, IQR; Left Ventricular Ejection Fraction, LVEF; Number, N.

Une solution pour les patients sans rehaussement tardif ayant une dysfonction VG en attente d'une réévaluation après titration et IS ?

Skowasch et al. PLoS One. 2018 Mar 22;13(3):e0194496.

Take Home Messages

- **Sarcoïdose cardiaque : pathologie grave**
- **Attention aux atteintes cardiaques isolées** (27–54% dans la littérature)
- **Y penser** si sarcoïdose connue mais aussi si BAV < 55 ans, « myocardite » récidivante ou d'évolution atypique, dysplasie arythmogène du ventricule droit, CMD non ischémique non familiale
- **Imagerie multimodale** : TEP et IRM, idéalement TEP IRM
- **DAI si indication de « pacing »**
- **Stratification du risque rythmique : FEVG ++, rehaussement tardif IRM, atteinte VD, SVP**
- **Rehaussement tardif = fibrose non réversible = lit du risque rythmique**
- **Prévention de la mort subite à discuter :**
  - **De manière collégiale**
  - **Indépendamment du traitement étiologique**

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