7e MASTERCLASS AMYLOSES CARDIAQUES
ÉVÉNEMENT HYBRIDE
PRÉSENTIEL & DISTANCIEL
Mercredi 11 décembre 2024
Fondation Biermans-Lapôtre ■ PARIS
www.masterclass-amylose.com

CAS CLINIQUE N°4

Une amylose obstructive

Dr Soulef Guendouz

Centre de référence des Amyloses Cardiaques
Filière Cardiogen
G.H.U. Henri Mondor - Créteil

AP-HP. Hôpitaux universitaires Henri-Mondor

cardiogen
filière nationale de santé
maladies cardiaques héréditaires ou rares
www.filiere-cardiogen.fr

Réseau Amylose Mondor
Centre de Référence National des Amyloses Cardiaques

M.A. 80 ANS

2016
CMH septale asymétrique OBSTRUCTIVE

2017
PACE MAKER

2021
Fibrillation auriculaire paroxystique mis sous CORDARONE

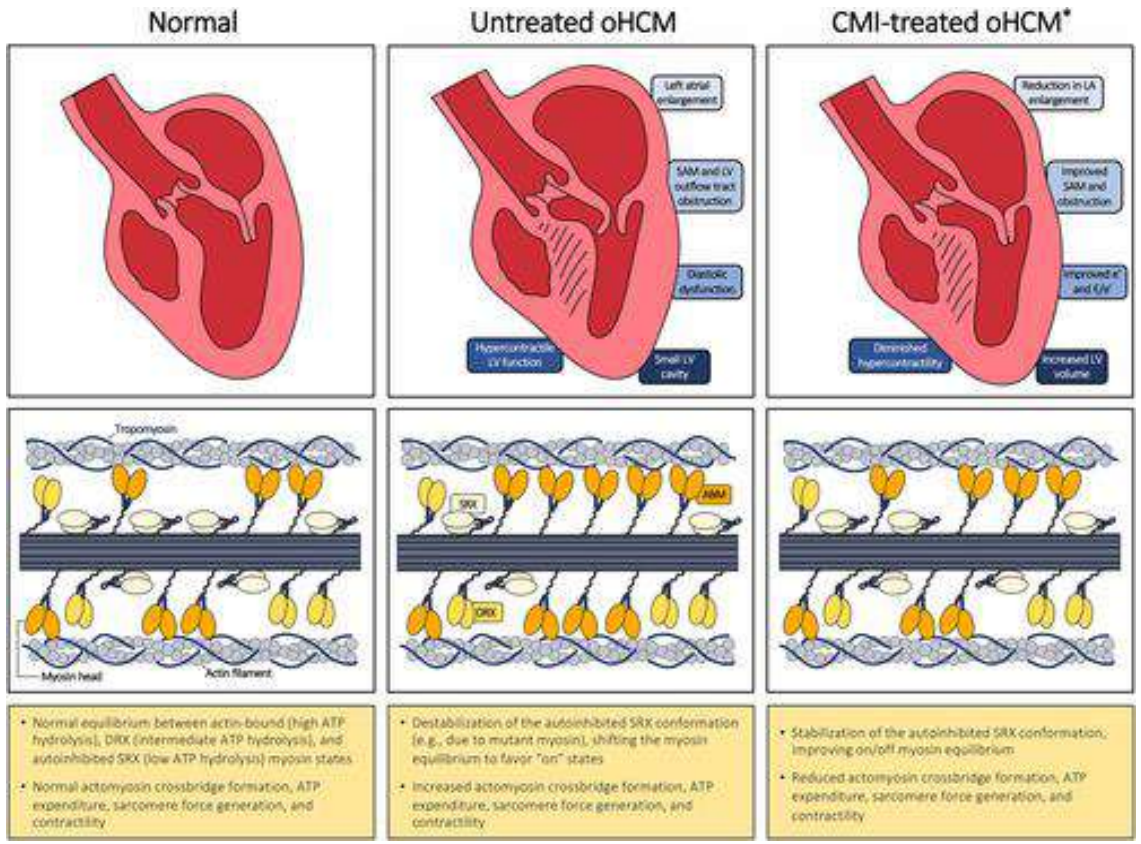
2023
ABLATION DE FIBRILLATION AURICULAIRE

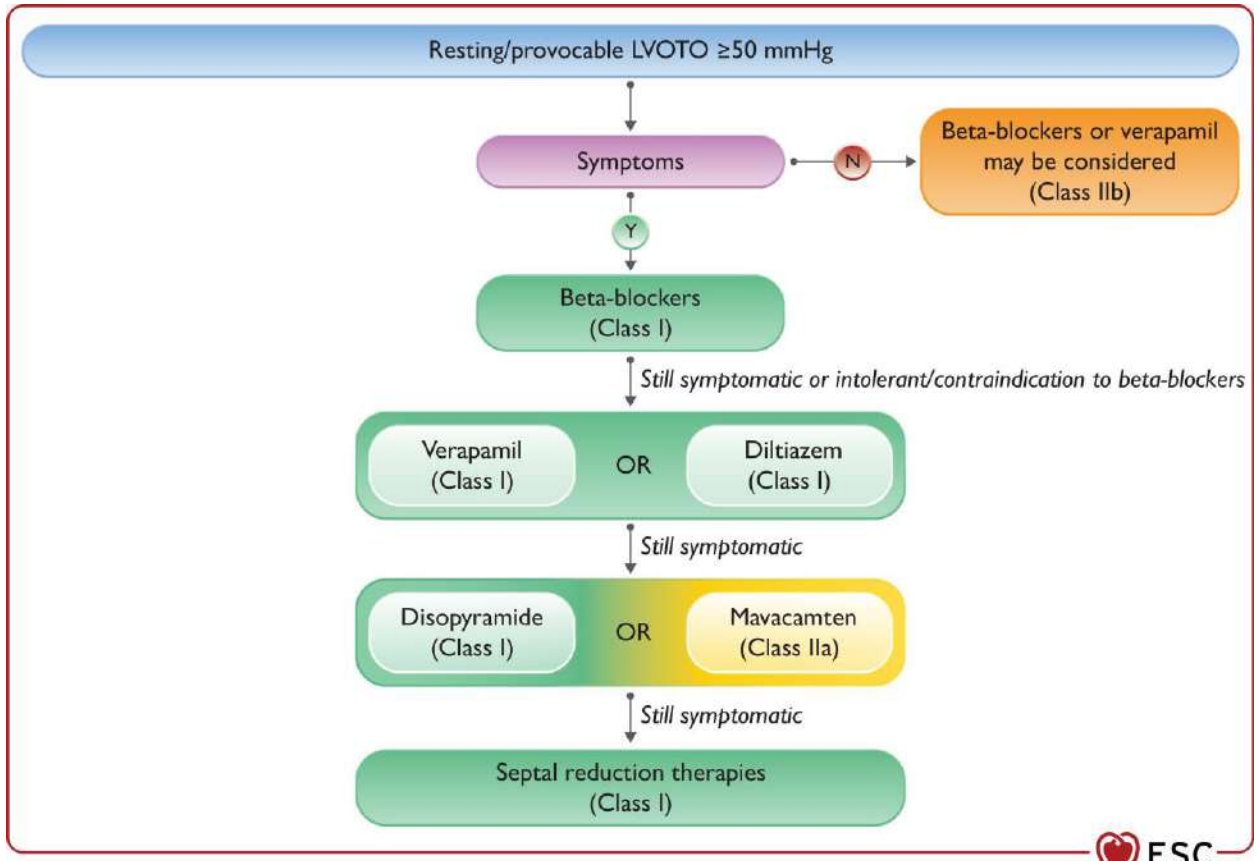
Mars 2024
CORONAROGRAPHIE NORMALE
ALCOOLISATION SEPTALE

Reste symptomatique
Sous Bétabloquant
Disopyramide mal toléré

Oct 2024
ADRESSE POUR DISCUSSION MAVACAMTEN

MAVACAMTEN

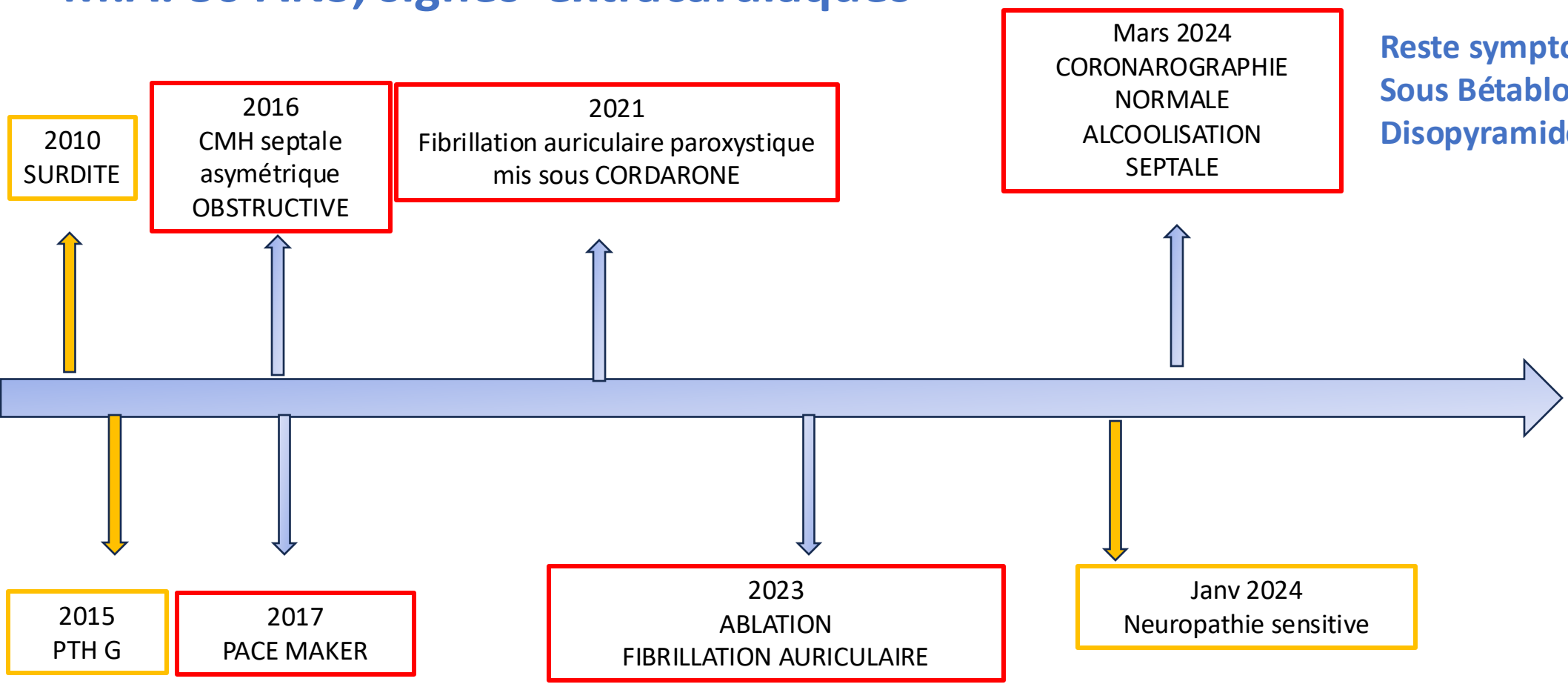




John W. Ostrominski et al. J Am Coll Cardiol HF 2023; 11:735-748.

Eur Heart J, Volume 44, Issue 37, 1 October 2023, Pages 3503–3626

M.A. 80 ANS, signes extracardiaques



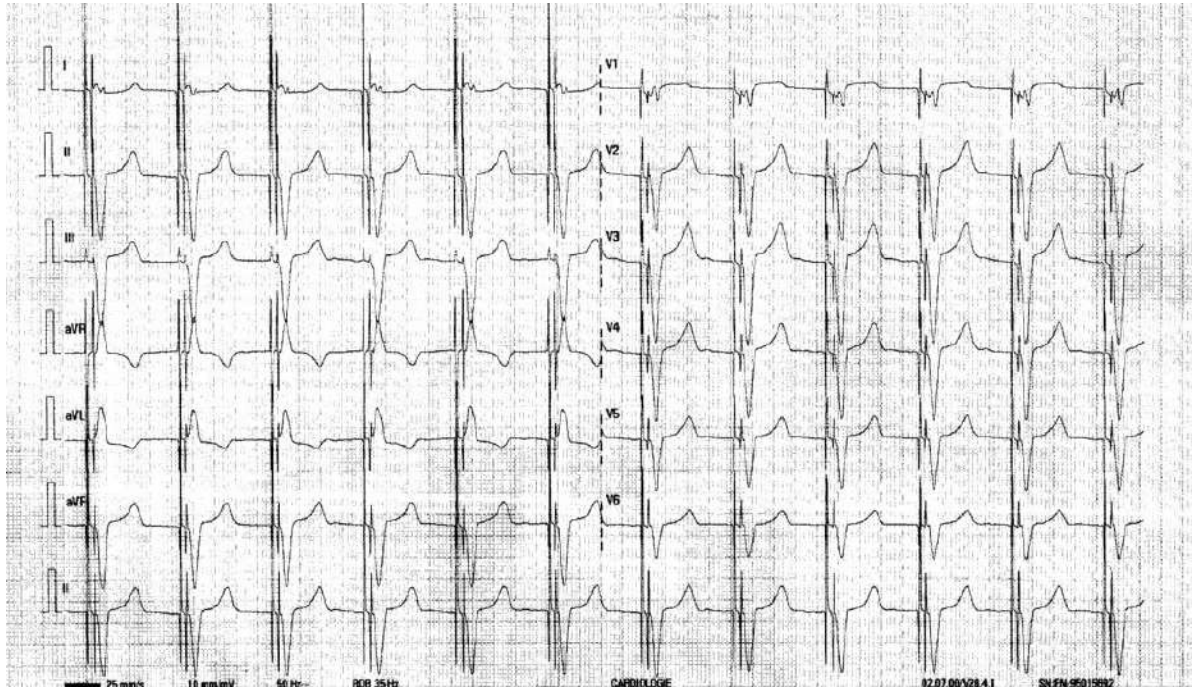
Reste symptomatique
Sous Bétabloaquant
Disopyramide mal toléré

M.A. 80 ANS

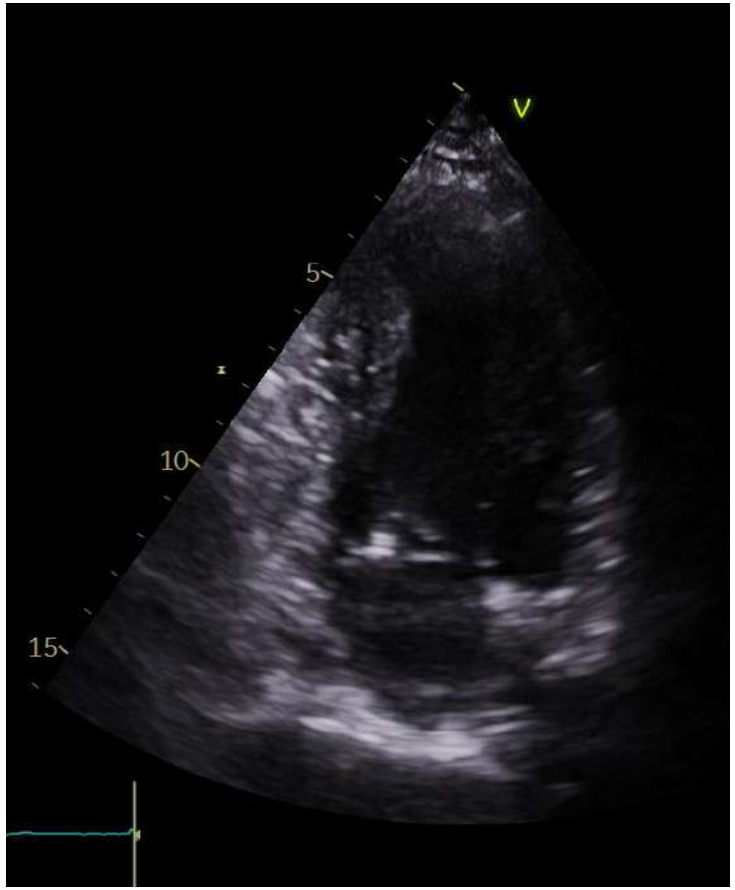
Bilan neuropathie sensitive des membres inférieurs :

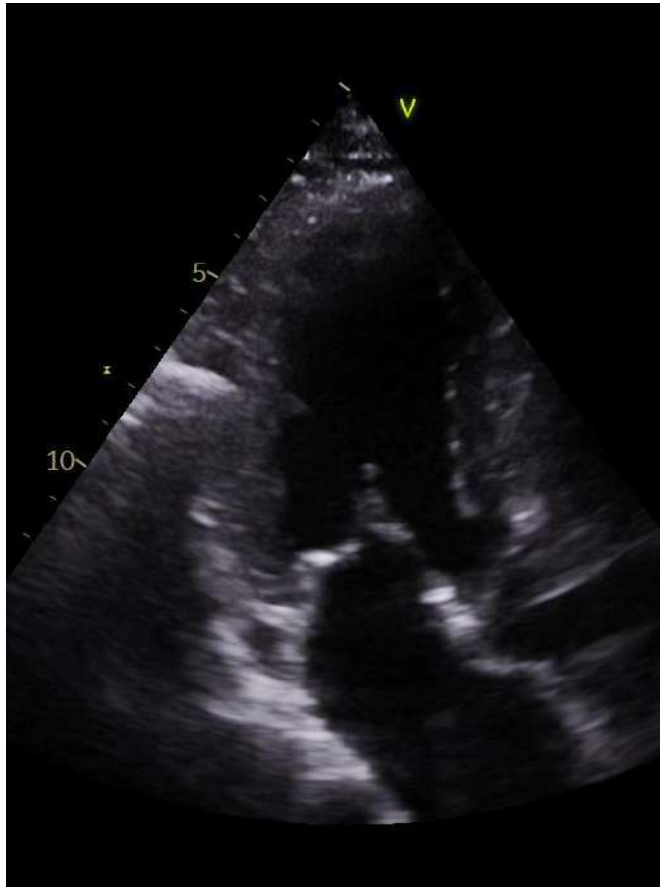
- Absence de gammapathie
  - EPP : absence de pic, présence d'une hypogammaglobulinémie
  - IF : absence d'anomalie
  - EPU + IEPU : absence de protéinurie de Bence Jones
  - Taux de kappa : 14 mg/L, Taux de Lambda : 13 mg/L ; soit un rapport de 1.1
- Mutation TTR : négative
- BGSA : discrets remaniements fibreux

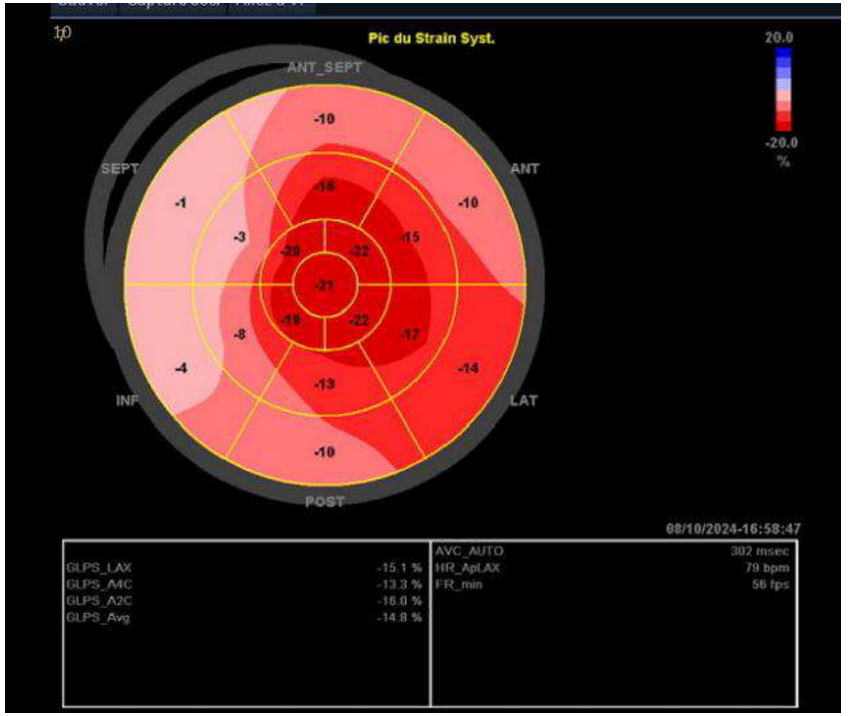
⇨ attribuée à l'amiodarone



NT pro BNP 1413 ng/ml
Troponine THS 30ng/l


V
5
10
15


V
5
10


Pic du Strain Syst.
20.0
-20.0
%
ANT_SEPT
SEPT
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INF
LAT
POST
-10
-1
-3
-20
-22
-15
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-17
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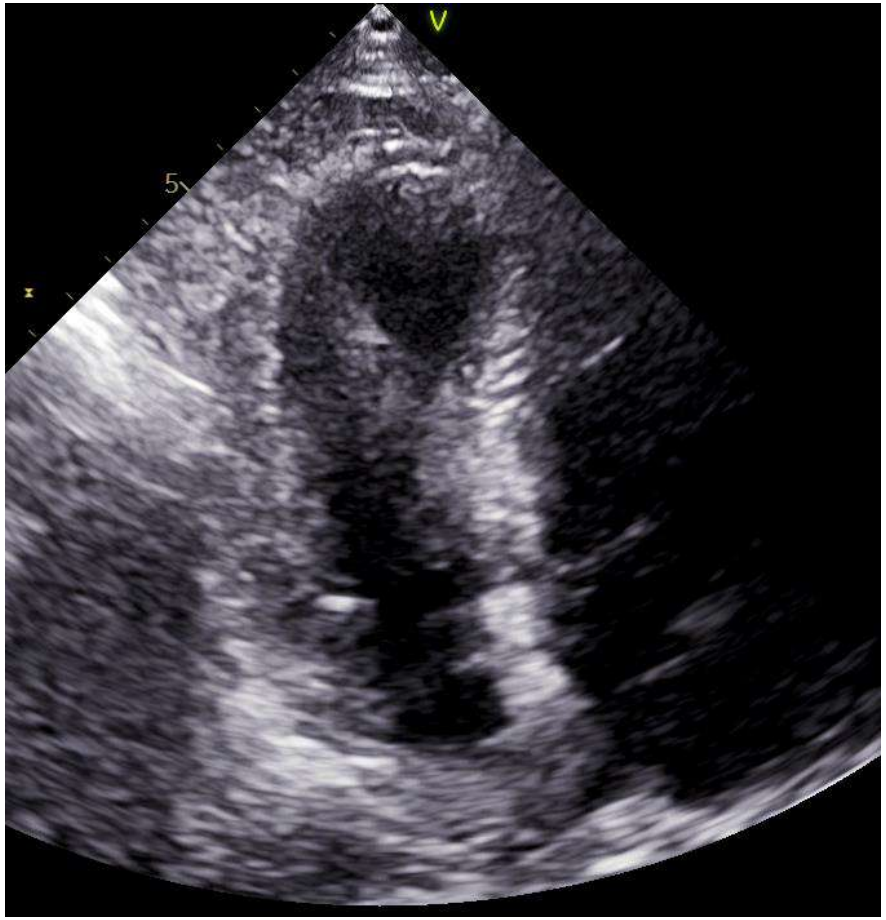
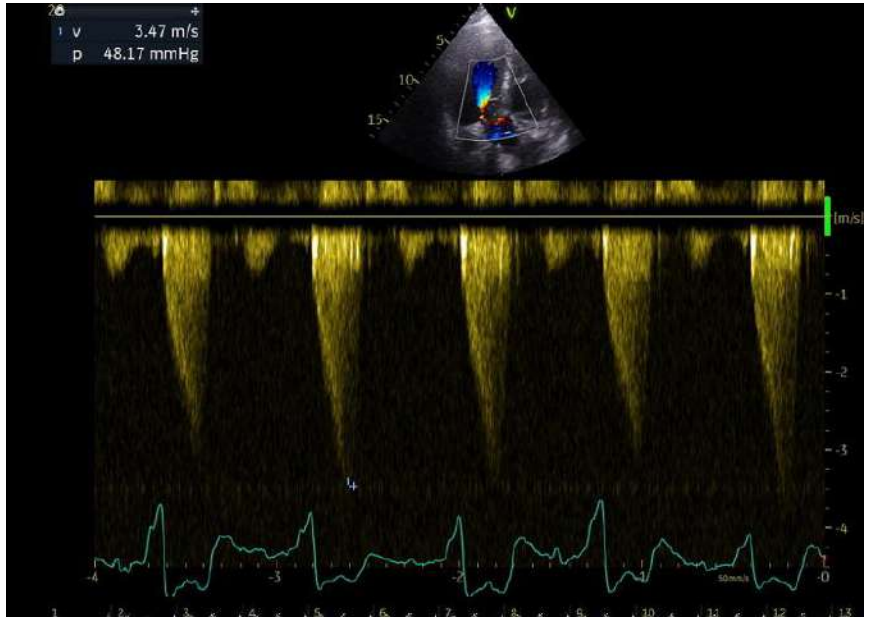
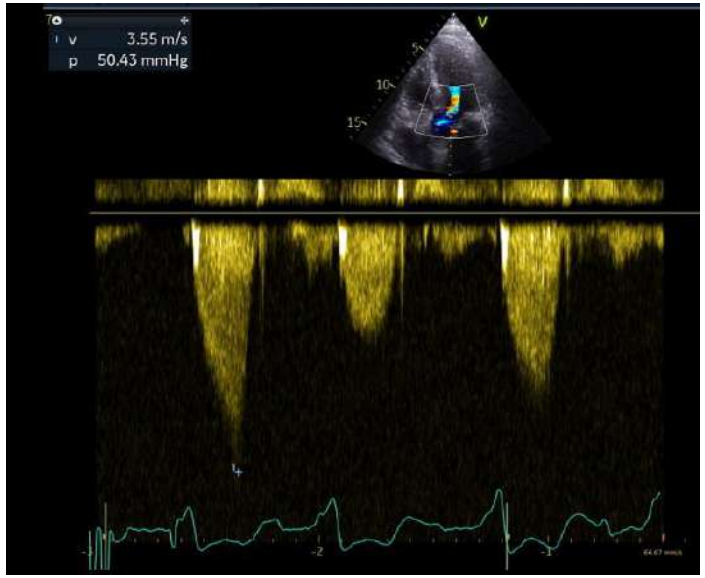
-13
-14
-10
08/10/2024-16:58:47
GLPS_LAX -15.1 %
GLPS_A4C -13.3 %
GLPS_A2C -16.0 %
GLPS_Avg -14.8 %
AVC_AUTO 302 msec
HR_ApLAX 79 bpm
FR_min 58 fps

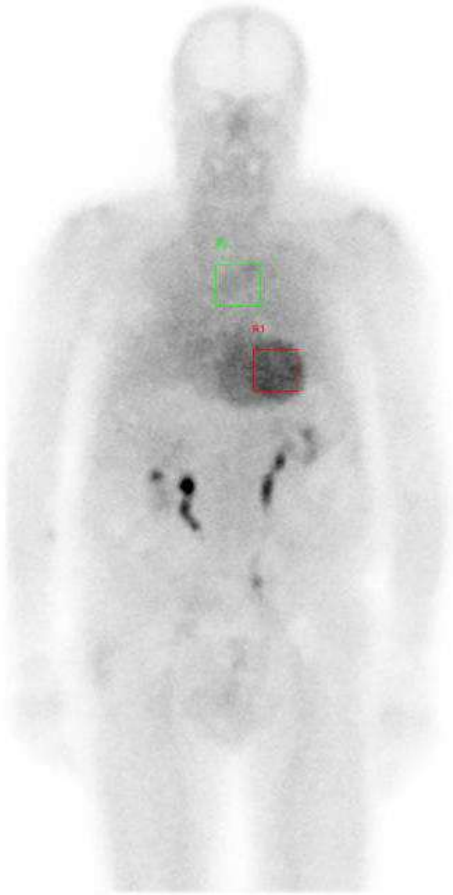


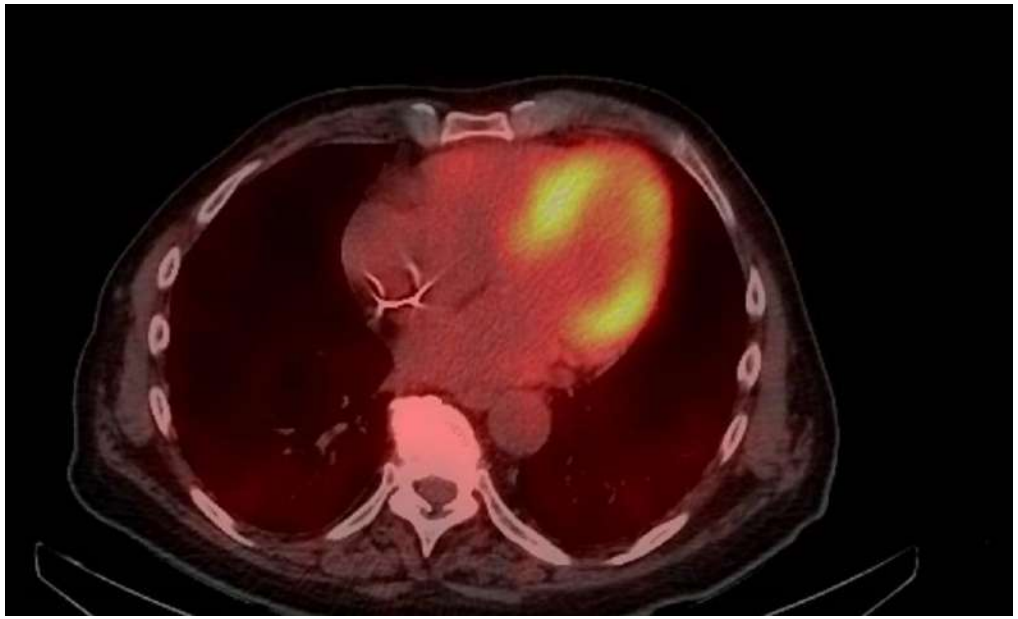




Gradient max à 50mmHg

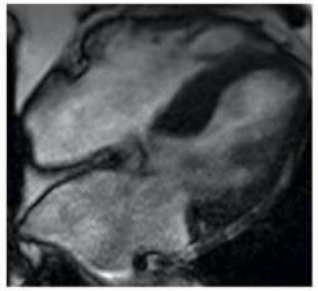
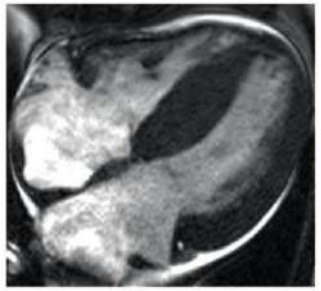
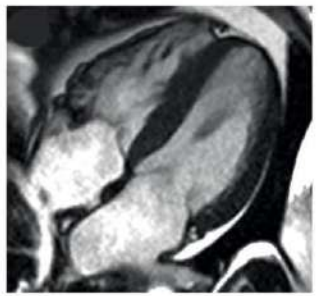
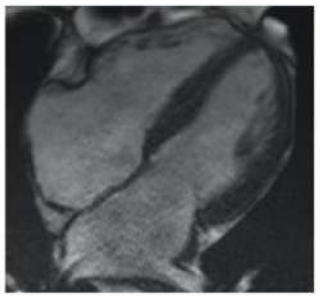
Scintigraphie osseuse





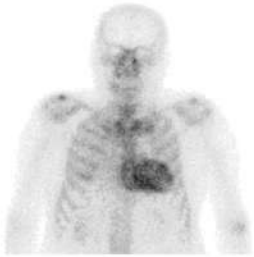
Rapport cœur/médiastin = 1,76.

Asymmetric Hypertrophy

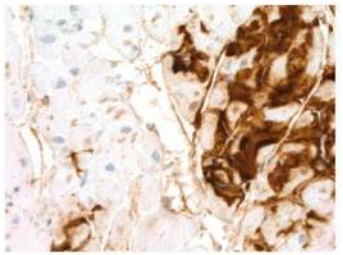
Sigmoid Septal Contour	Reverse Septal Contour
55%	24%
	
	
Symmetric Hypertrophy	No LVH
18%	3%

Martinez-Naharro, A. et al. J Am Coll Cardiol. 2017;70(4):466-77.

ATTRwt`s clinical spectrum

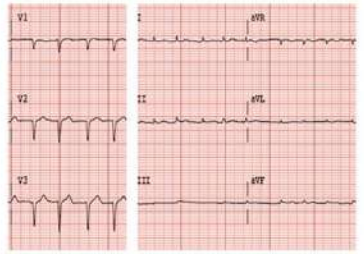


Mode of presentation:
- **HF** (67.6%)
- **AV block** (7.4%)
- **Stroke**
- **HCM or RCM** (13.9%)
- **Degenerative AS**
- **Incidental** (11.1%)

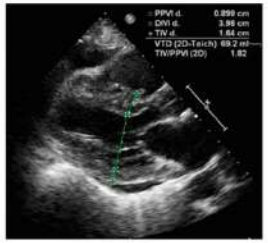


Age, gender and comorbidities
- Males (81.5%)
- **Females (18.5%)**
- Late 70s symptoms onset
- HTN (54.6%)

ECG
- **AF (55.6%)**
- **Pseudoinfarct** pattern (63.2%)
- Low voltage (22%)
- LVH (10.5%)
- L/RBBB (17-15%)

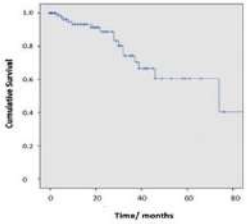


Echocardio
- Symmetric LVH (75.7%)
- **Asymmetric LVH (23.4%)**
- **LVEF<50% (36.8%)**
- Restrictive diastolic pattern (35%)
- Pericardial effusion (42.1%)



Survival

Overall survival at 12, 24 and 36 months: **93**, **89** and **74%**, respectively



Eur Heart J, Volume 38, Issue 24, 21 June 2017, Pages 1895–1904,

AMYLOSE ATTR sauvage obstructive

Pas de prescription de Mavacamten

Mais prescription de Tafamidis

Poursuite des bétabloquants avec tentative de désynchronisation PM et discussion d'une nouvelle alcoolisation septale

→ Sous une CMH asymétrique peut se cacher une amylose

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Merci à l'ensemble du RÉSEAU AMYLOSE Mondor !

Cardiac Amyloidosis Referal Center (Rare Disease Network)

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Referral center secretariat: I Vallat
IDE amyloidosis coordination: S Maupou
Psychology: J Pompougnac

cardiogen
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maladies cardiaques héréditaires ou rares

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Research assistant: Ani, Dilan, Saafa, Sarah, Benoît, Lola

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Neuromuscular disease: S Souvannanorath
Nephrology: V Audard, H Sakhi
Haematology: F Lemmonier, K Belhadj, J Dupuis, F Le Bras, R Gounot, M Van Den Akker
Internal medicine: M Michel
Hepatology: V Leroy, A Sessa
Geriatry: A Broussier, N Liu, N Marie Nelly
Genetic: B Funalot, B Hébrard, C Nativelle
Rhumato : S Guignard
Orthopédie : O Pidet

Amyloidosis Diagnosis and Monitoring Platforms

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Pathology: E Poullot, C Charpy, A Moktefi
Sequencing: P Fanen, M Konyukh
Immuno-biology: V Frenkel, H Abroud, A Beldi Ferichou
Radiology: V Tacher, I Sifaoui
Nuclear medicine: E Itti, L Lerman

HF Telemonitoring

Coordination: E Sarre, A Duchenne
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