



CAS CLINIQUE N°4

Une amylose obstructive



Dr Soulef Guendouz

Centre de référence des Amyloses Cardiaques

Filière Cardiogen

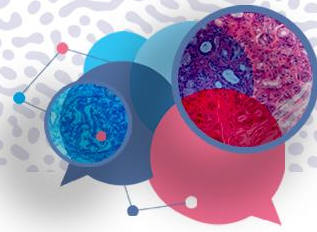
G.H.U. Henri Mondor - Créteil



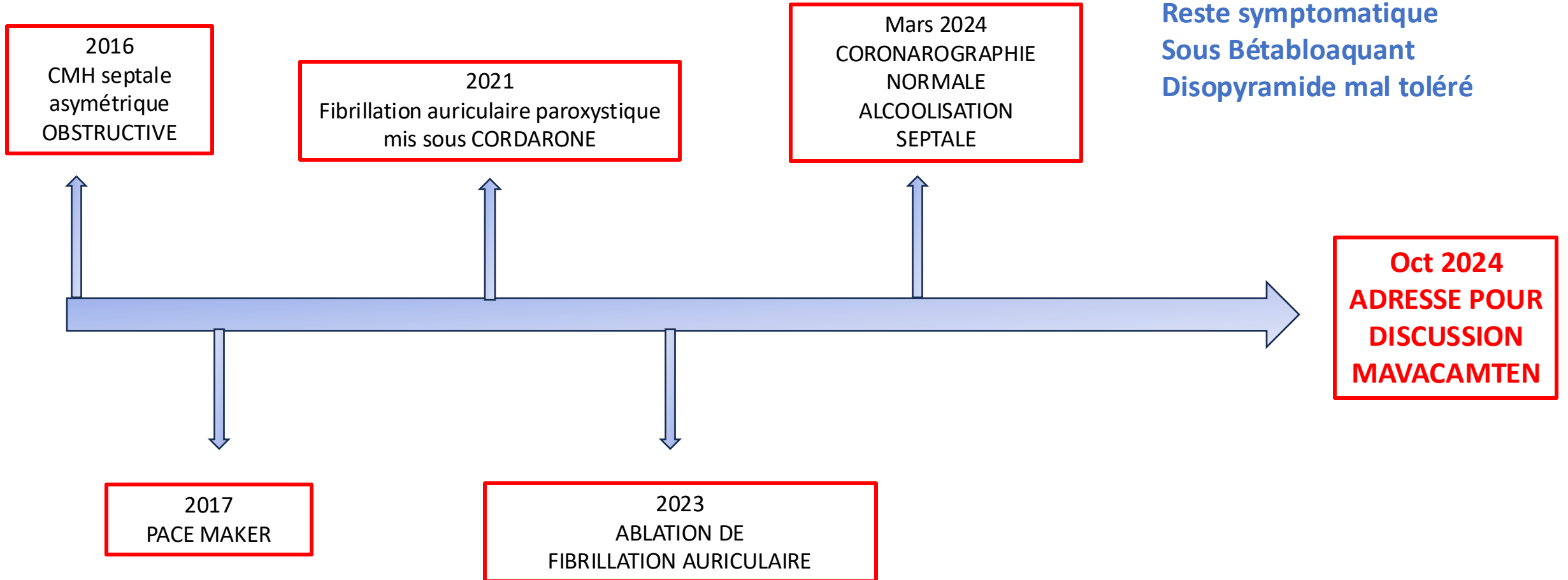
AP-HP.
Hôpitaux universitaires
Henri-Mondor

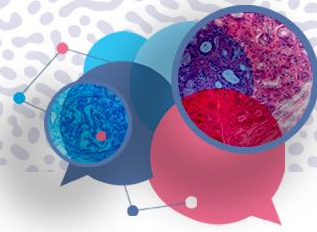


**Réseau Amylose
Mondor** Centre de Référence National
des Amyloses Cardiaques

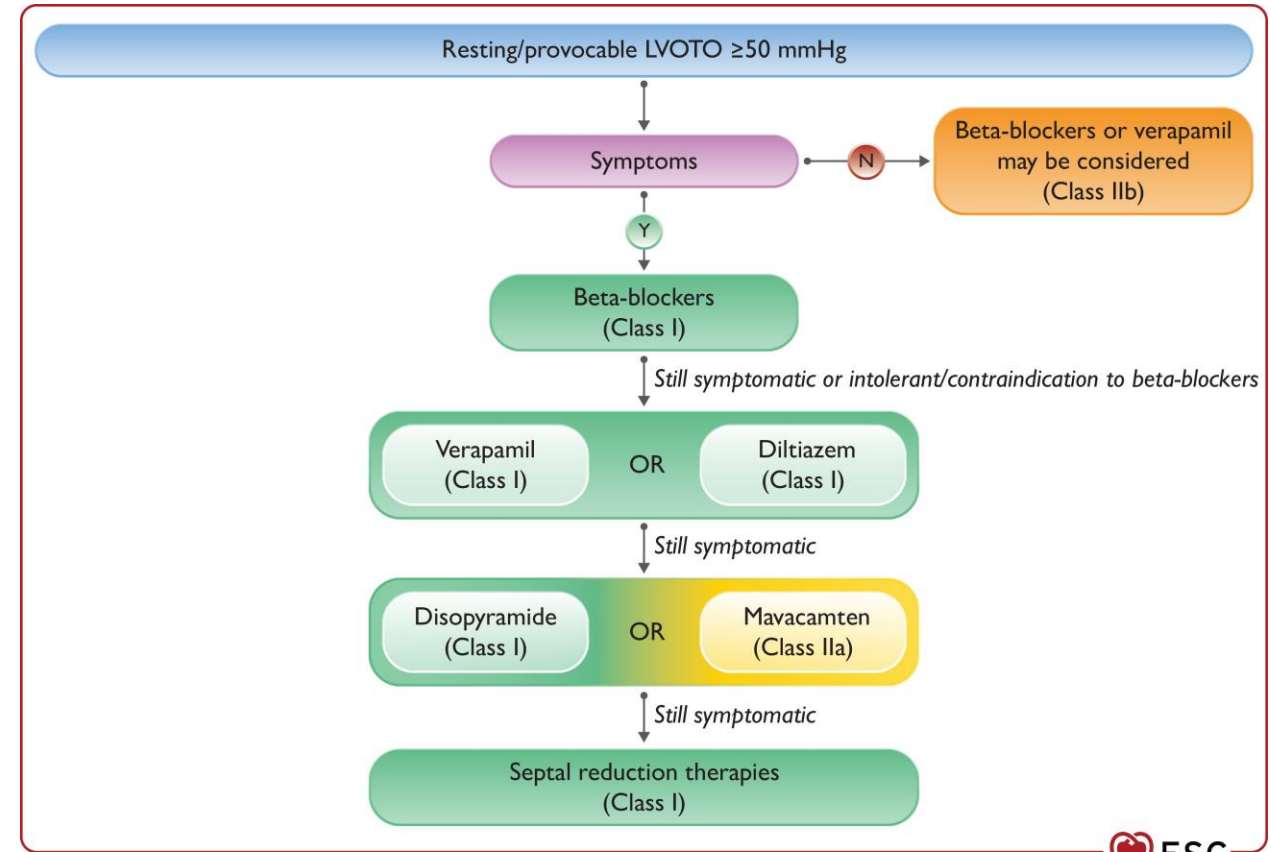
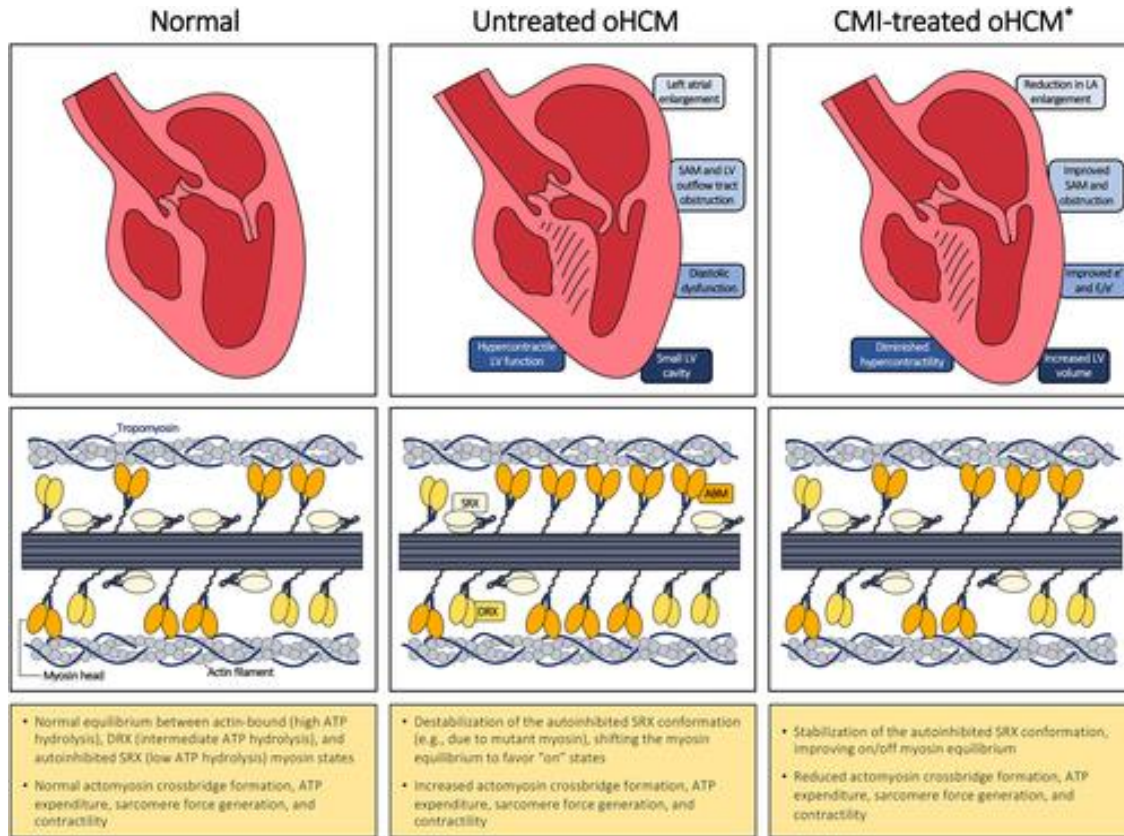


M.A. 80 ANS



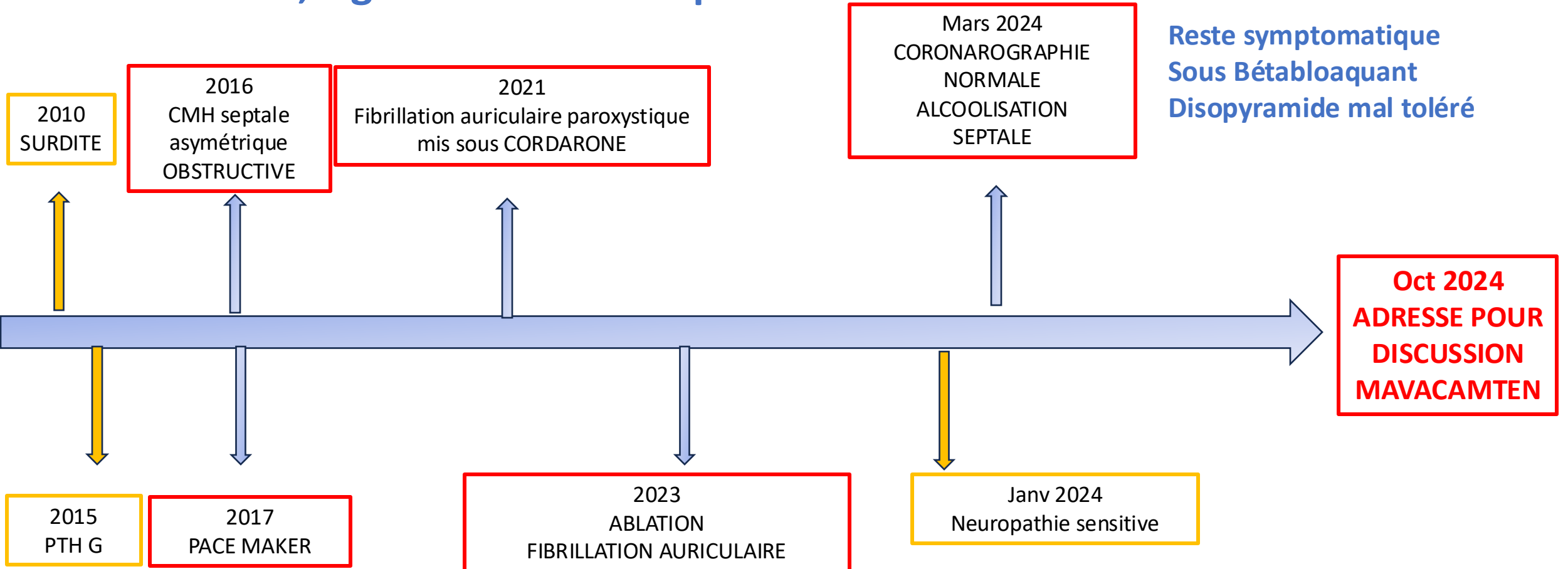


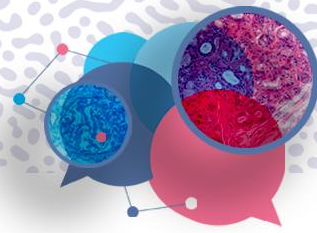
MAVACAMTEN





M.A. 80 ANS, signes extracardiaques

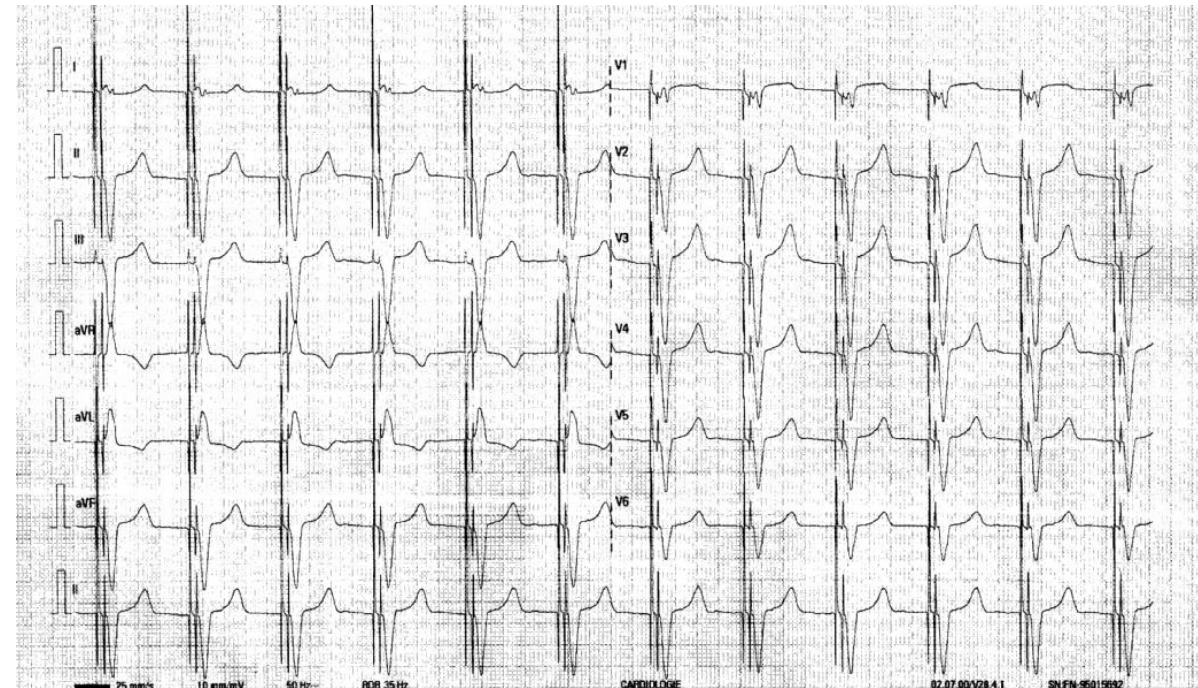




M.A. 80 ANS

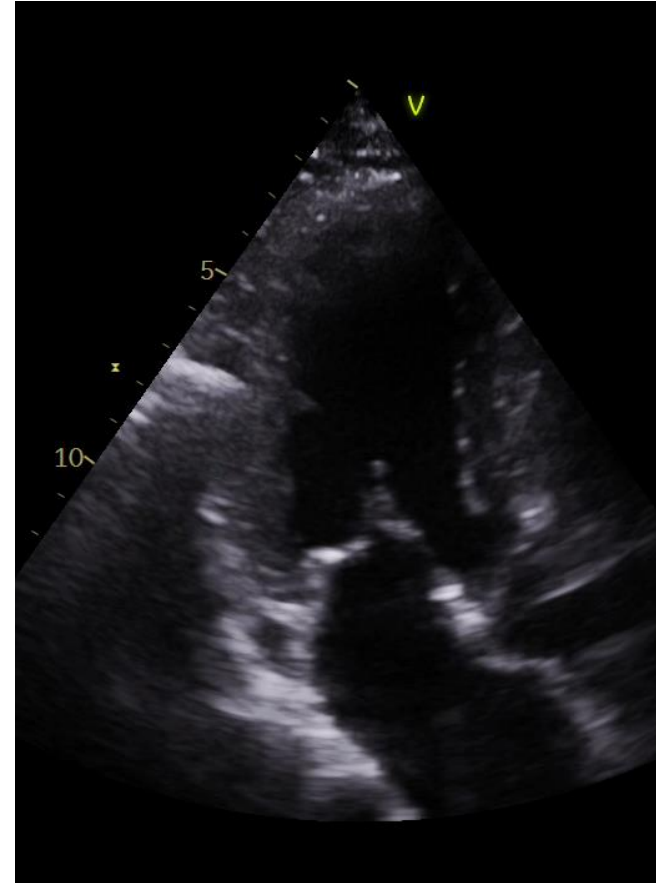
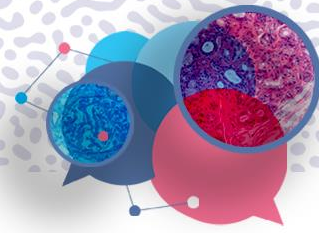
Bilan neuropathie sensitive des membres inférieurs :

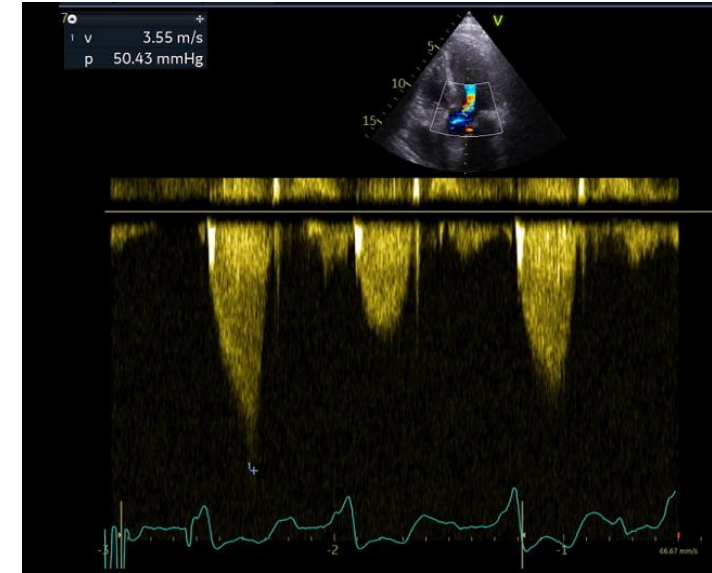
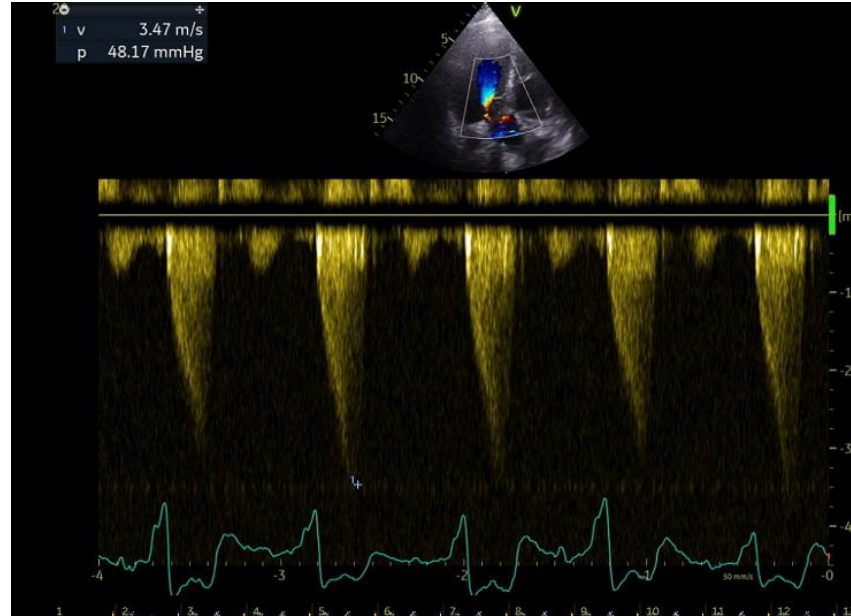
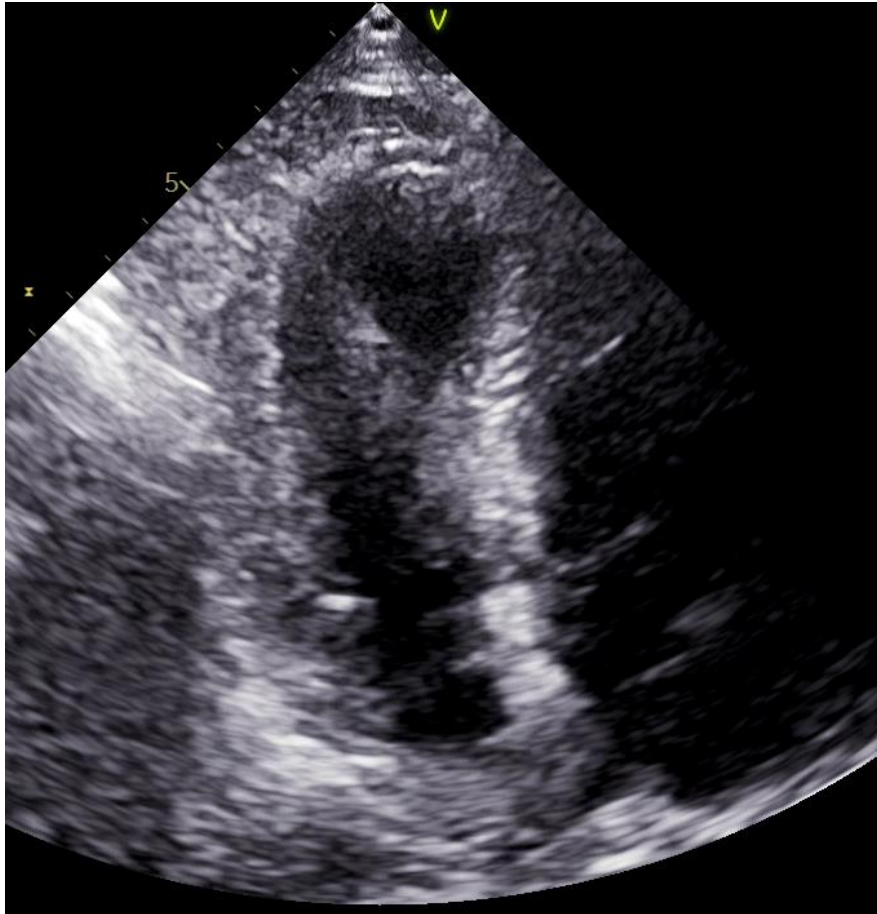
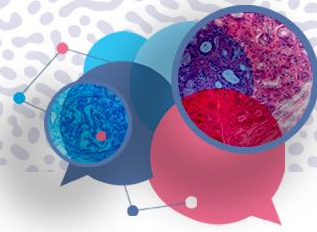
- Absence de gammopathie
 - EPP : absence de pic, présence d'une hypogammaglobulinémie
 - IF : absence d'anomalie
 - EPU + IEPU : absence de protéinurie de Bence Jones
 - Taux de kappa : 14 mg/L, Taux de Lambda : 13 mg/L ; soit un rapport de 1.1
- Mutation TTR : négative
- BGSA : discrets remaniements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



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

➔ attribuée à l'amiodarone

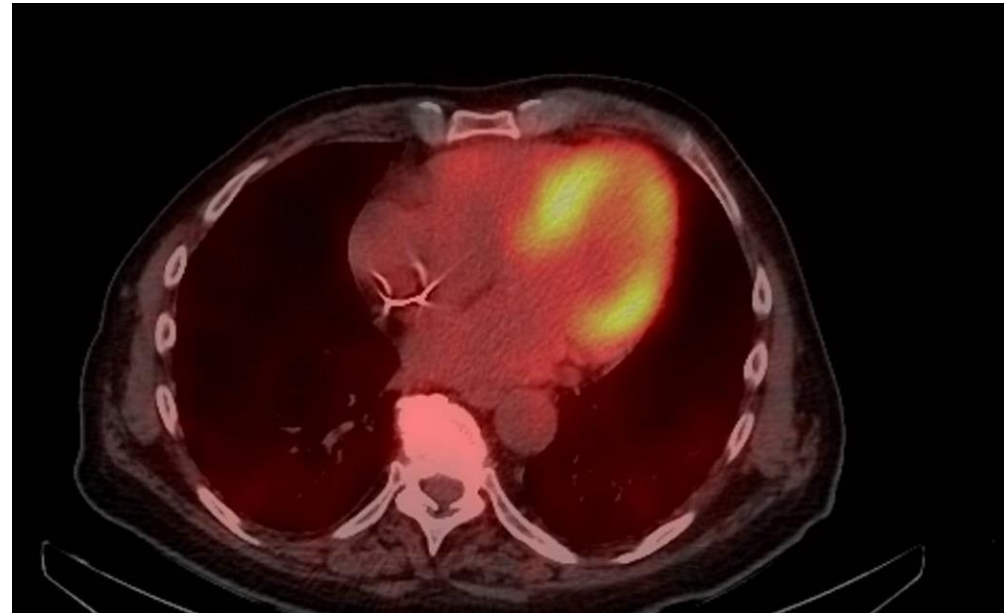
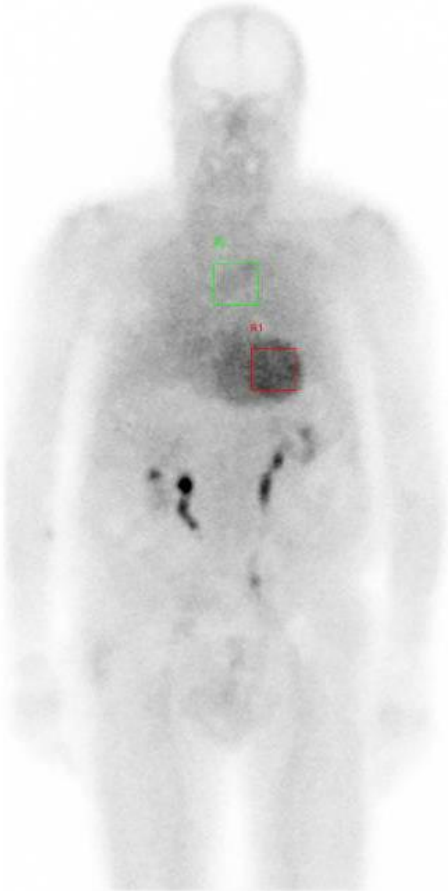




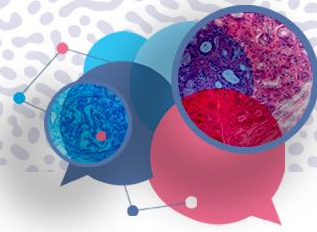
Gradient max à 50mmHg



Scintigraphie osseuse



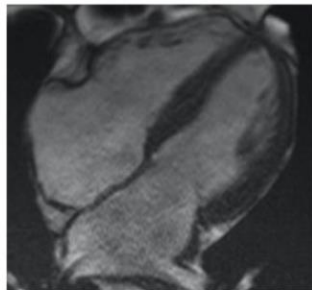
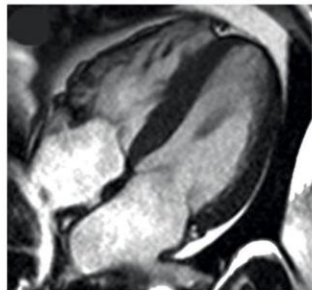
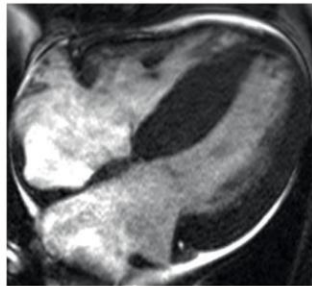
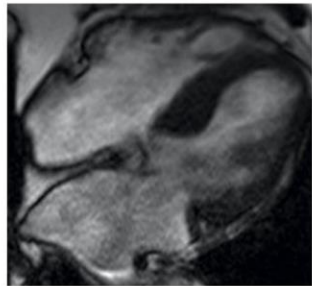
Rapport cœur/médiastin = 1,76.



Asymmetric Hypertrophy

Sigmoid Septal
Contour
55%

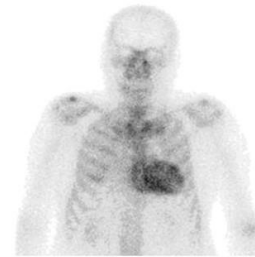
Reverse Septal
Contour
24%



Symmetric
Hypertrophy
18%

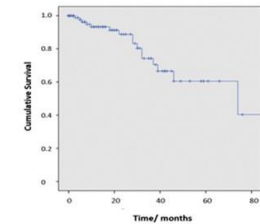
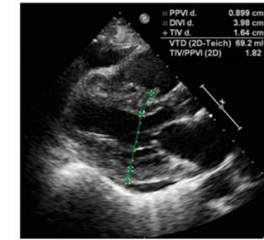
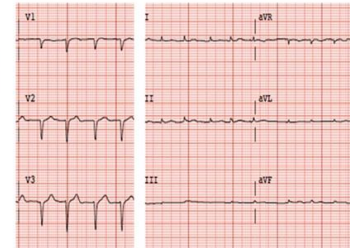
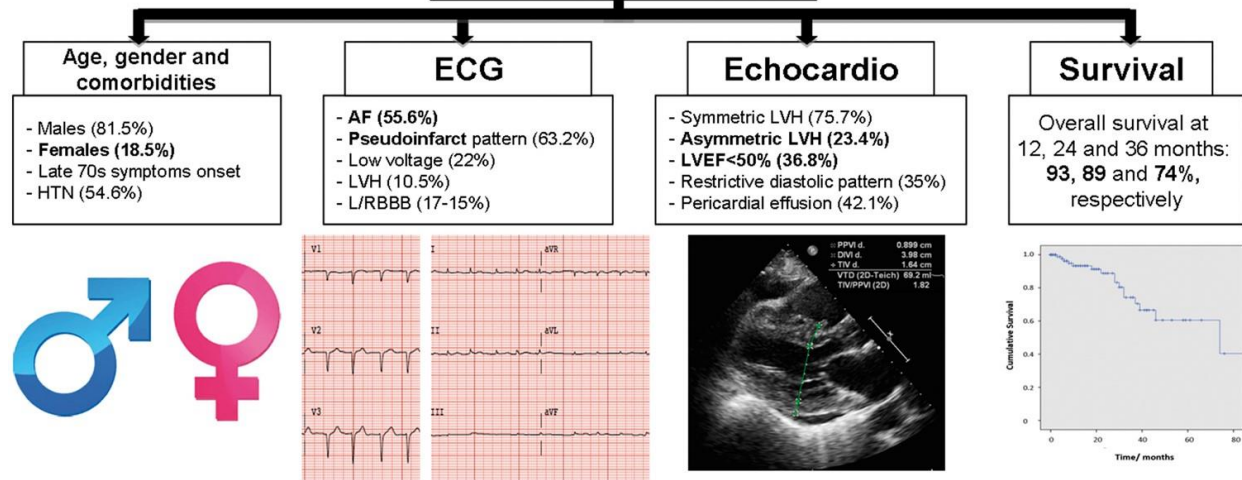
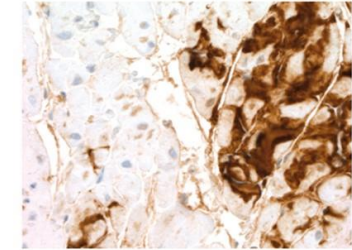
No LVH
3%

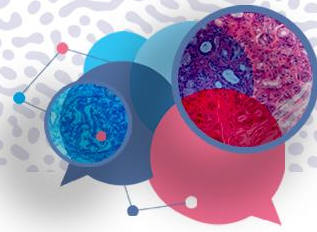
ATTRwt's clinical spectrum



Mode of presentation:

- HF (67.6%)
- AV block (7.4%)
- Stroke
- HCM or RCM (13.9%)
- Degenerative AS
- Incidental (11.1%)





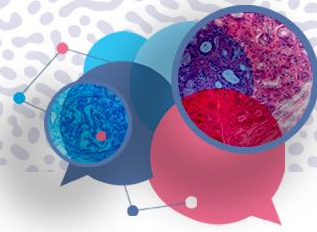
AMYLOSE ATTR sauvage obstructive

Pas de prescription de Mavacamten

Mais prescription de Tafamidis

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

→ Sous une CMH asymétrique peut se cacher une amylose



Merci à l'ensemble du RÉSEAU AMYLOSE Mondor !

Cardiac Amyloidosis Referral Center (Rare Disease Network)

Cardiologists Team

Cardiologist: T Damy, S Oghina, A Zaroui, S Guendouz, A Galat, S Mallet, GDS Chadha, M Hentati, E Charbonneau, S Odouard, A Copie, E Teiger
Rythmologist: N Lellouche, T Moulin, K Ramoul, N Elbaz, S Rouffiac, V Ouazana

Coordination - Quality of Life

Healthcare pathway: C Henrion, Anaïs
Referral center secretariat: I Vallat
IDE amyloidosis coordination: S Maupou
Psychology: J Pompougnac



Clinical Research Team/HEAR

Study engineer: M Kharoubi,
Research assistant: Ani, Dilan, Saafa, Sarah, Benoît, Lola



Medicine Multidisciplinary Network

Neurology: V Planté-Bordeneuve, T Gendre
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
Geriatrics: 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

Electrophysiology: JP Lefaucheur
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

INSERM U955 Clinical Epidemiology in Aging

Florence Canoui-Poitrine
Etienne Audureau
Charlotte Lafont

HF Telemonitoring

Coordination: E Sarre, A Duchenne
Nurses: A Gauchard, M Frelat, S Dias, C Lecerf
Cardiologist : L Hittinger

UMR CNRS 8256, INSERMU1164

Onnik Agbulut
Ekaterinii Kordeli
France Lam