



## Controverses de la prise en charge cardiologique

# Défibrillateur dans les amyloses AL

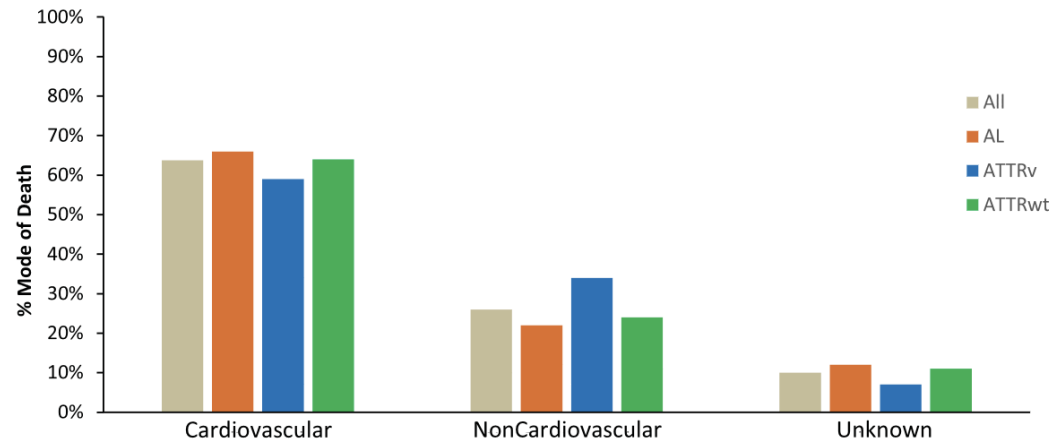


**Dr Thibaut MOULIN**

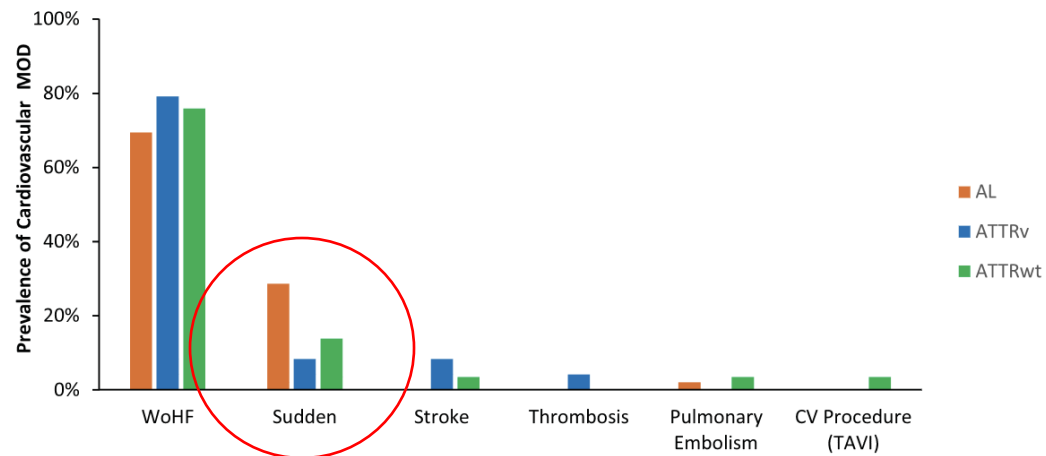
Centre de référence des Amyloses Cardiaques – Filière Cardiogen  
Service de Cardiologie - Unité de rythmologie  
CHU Henri Mondor - Créteil



A: Prevalence of the Mode of Death (Cardiovascular, Non-Cardiovascular, Unknown ) in CA according to amyloidosis type



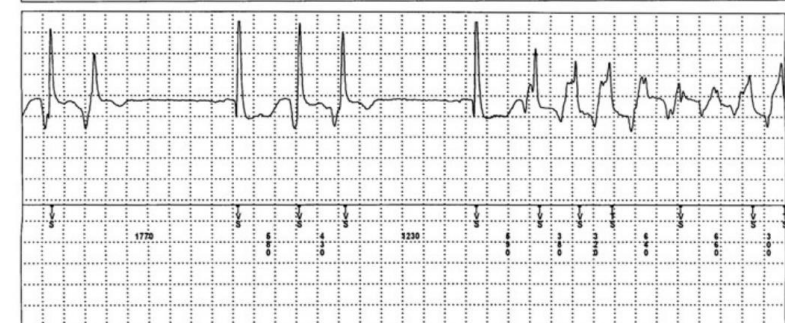
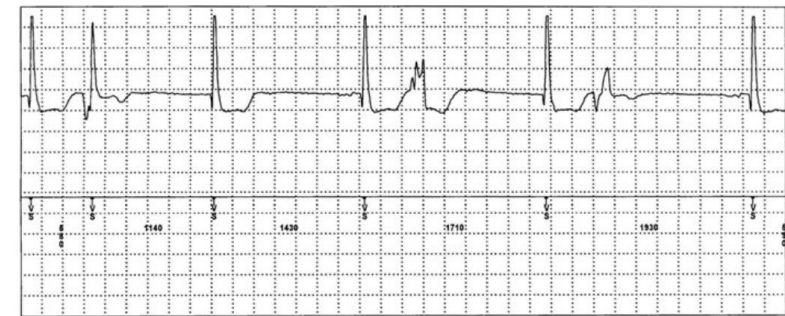
B : Prevalence of Cardiovascular Mode of Death in CA according to amyloidosis type



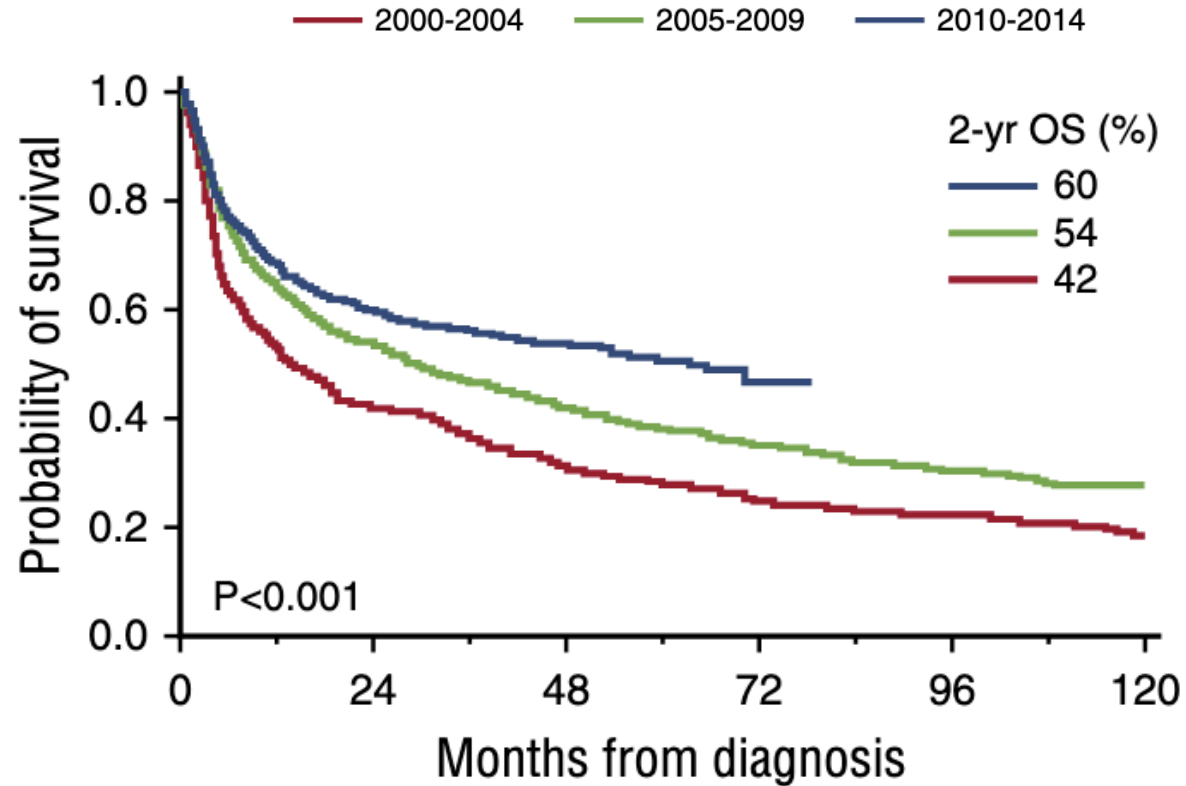
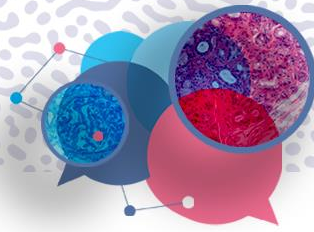
Kharoubi, Amyloid 2022

## Causes of sudden cardiac death (SCD)

- Electromechanical dissociation
- Severe bradycardia (SND or AVB)
- Ventricular arrhythmia (VT/VF)

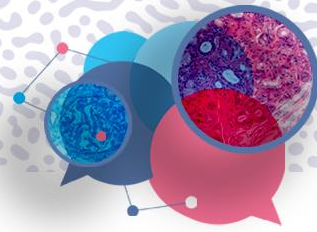


Sayed, Eur Heart J 2022



No. at Risk

— 422	177	131	106	94	78
— 604	321	248	205	121	45
— 525	257	132	19	0	0



## ICD in AL Cardiac Amyloidosis – Observational Studies

Year	Center	n	Primary prev (%)	Follow-up (years)	Appropriate ICD therapies - n (%)
2008	Heidelberg (Germany)	19	100	2.2 ± 0.4	2 (11)
2013	Mayo Clinic (US)	33	77	1.0	12 (36)
2014	Stanford (US)	15	73	-	5 (33)
2016	APHP (France)	12	84	1.4 ± 1.1	4 (33)
2020	Nashville (US)	7	100	3.4 ± 2.3	2 (29)

Kristen, Heart Rhythm 2008

Lin, J Cardiovasc Electrophysiol 2013

Varr, Heart Rhythm 2014

Hamon, Int J Cardiol 2016

Kim, Europace 2020



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### Limitations

- Small sample sizes
- High incidence of non-arrhythmic death
- Uncertain candidate selection.
- Inclusion of both AL and TTR

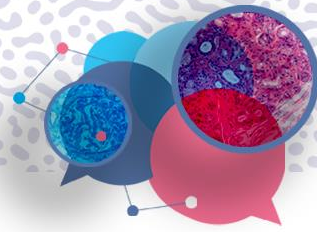
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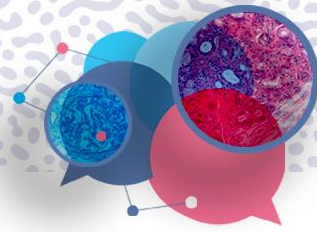
Kim, Europace 2020



## Recommendation Table 32 — Recommendations for implantable cardioverter defibrillator implantation in patients with cardiac amyloidosis

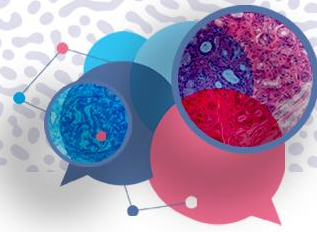
Recommendations	Class <sup>a</sup>	Level <sup>b</sup>
An ICD should be considered in patients with light-chain amyloidosis or transthyretin-associated cardiac amyloidosis and haemodynamically not-tolerated VT.	<b>IIa</b>	<b>C</b>

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## Henri Mondor practice – Assessment of SCD risk before chemotherapy

- 1) Assessment of life expectancy
  
- 2) Consider primary prevention ICD in European Stage III patients if :
  - Altered LVEF < 35% **or** LV GLS < -15%
  - NSVT
  - Non-postural syncope
  - Indication for permanent pacing (AVB, progressive PR/QRS, SND)



## Monocentric study

Inclusion: AL CA patients implanted with ICD (Nov 2014 – May 2024)

N = 150

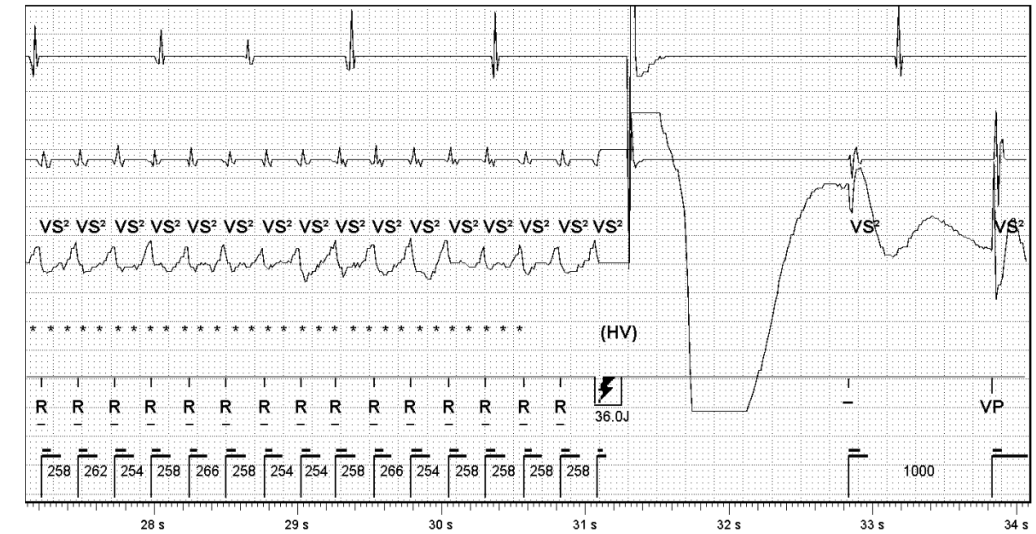
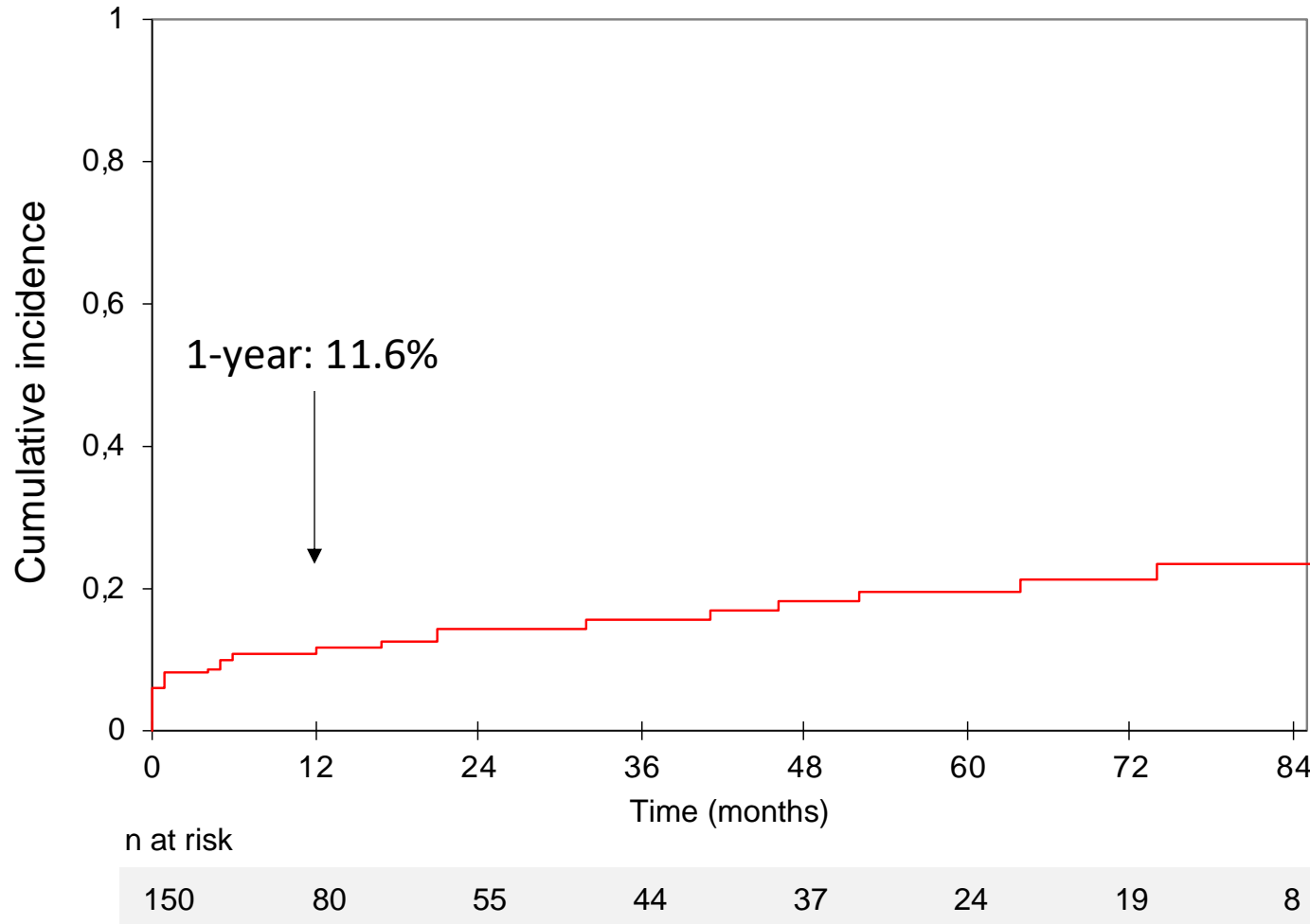
Median follow-up: 1.4 years [IQR, 0.5-4.7]

Baseline characteristics	Mean ± SD or n (%)
Primary prevention	150 (100%)
Age, years	63 ± 9
Male	98 (65%)
LVEF, %	50 ± 11
LV GLS, %	-9.7 ± 2.9





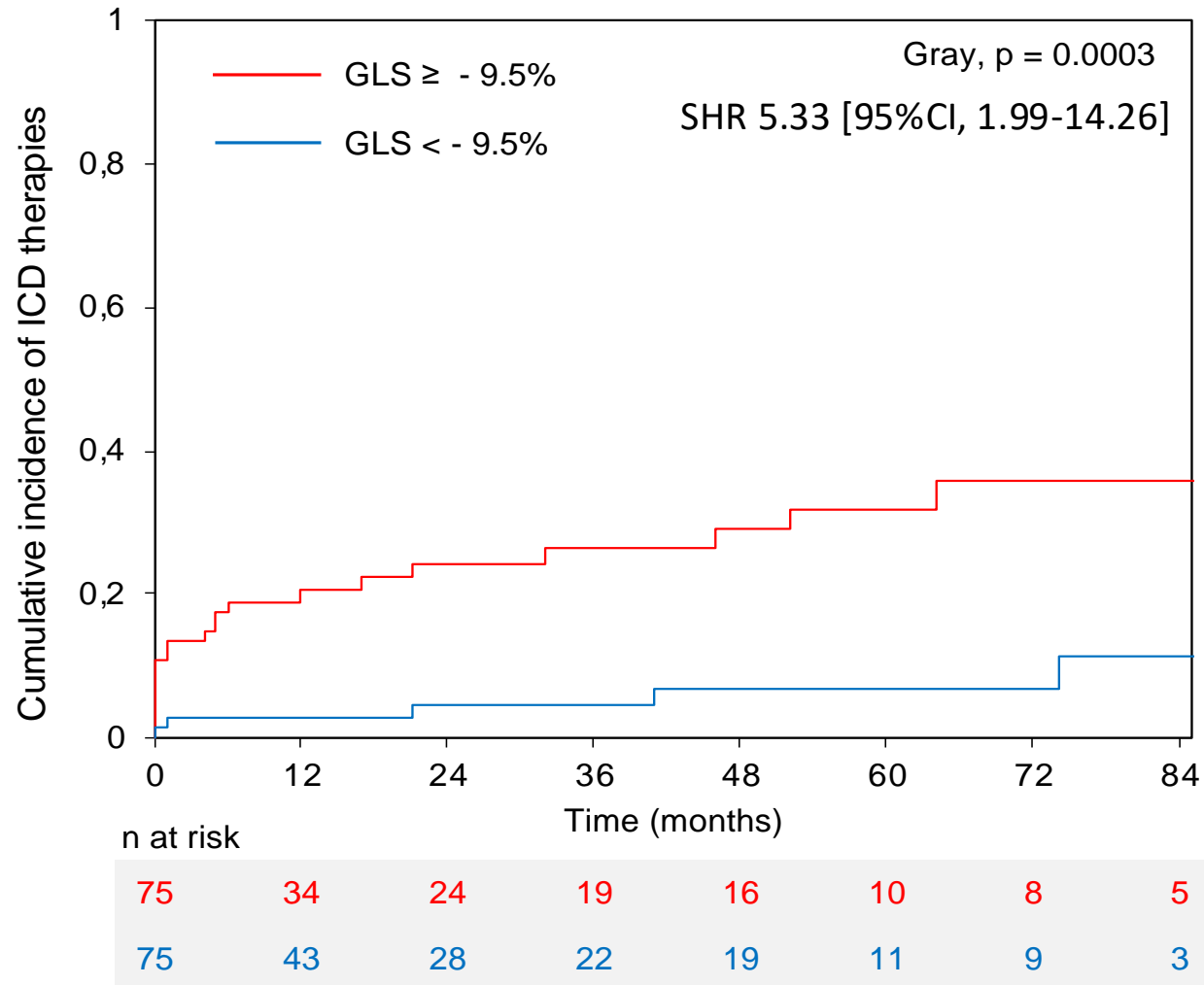
## Appropriate ICD therapies: n = 27 (18%)



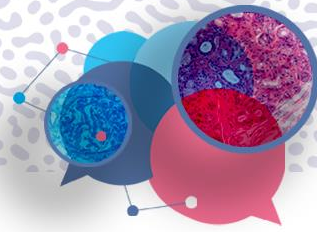
In preparation



## LV GLS = predictor of ventricular arrhythmias



In preparation



## Conclusions

- High rates of sudden death
- High rates of ventricular arrhythmias and ICD therapies
- LV GLS might improve risk stratification and ICD candidate selection
- Predictive value of CMR and Holter monitoring (NSVT) ?
- **Further studies needed to assess impact on survival**
- ICDs also prevent fatal brady-arrhythmias

## 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  
**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

**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 Clinial Epidemioloy 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

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