# Anticoagulation au cours de la cardioversion chez le patient avec amylose cardiaque

Amyl'O.S.E.
Objectif Spécialisation & Expertise en Amylose

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# Généralités

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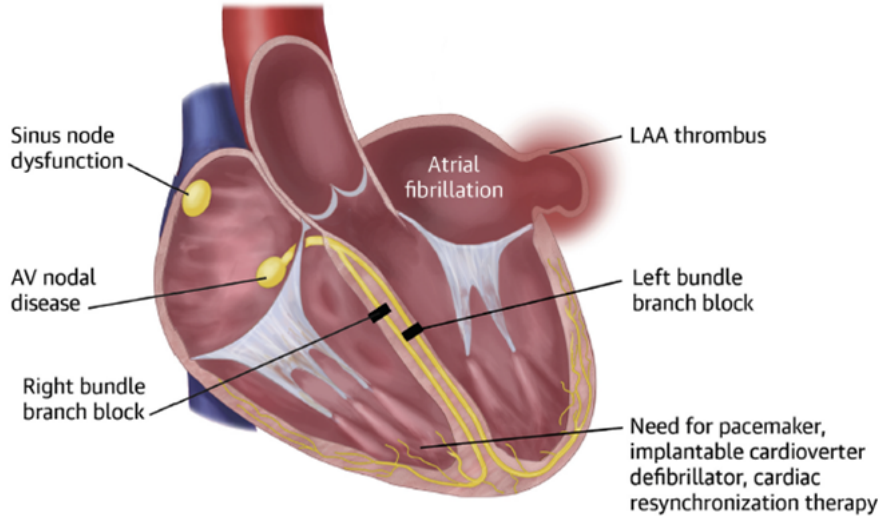
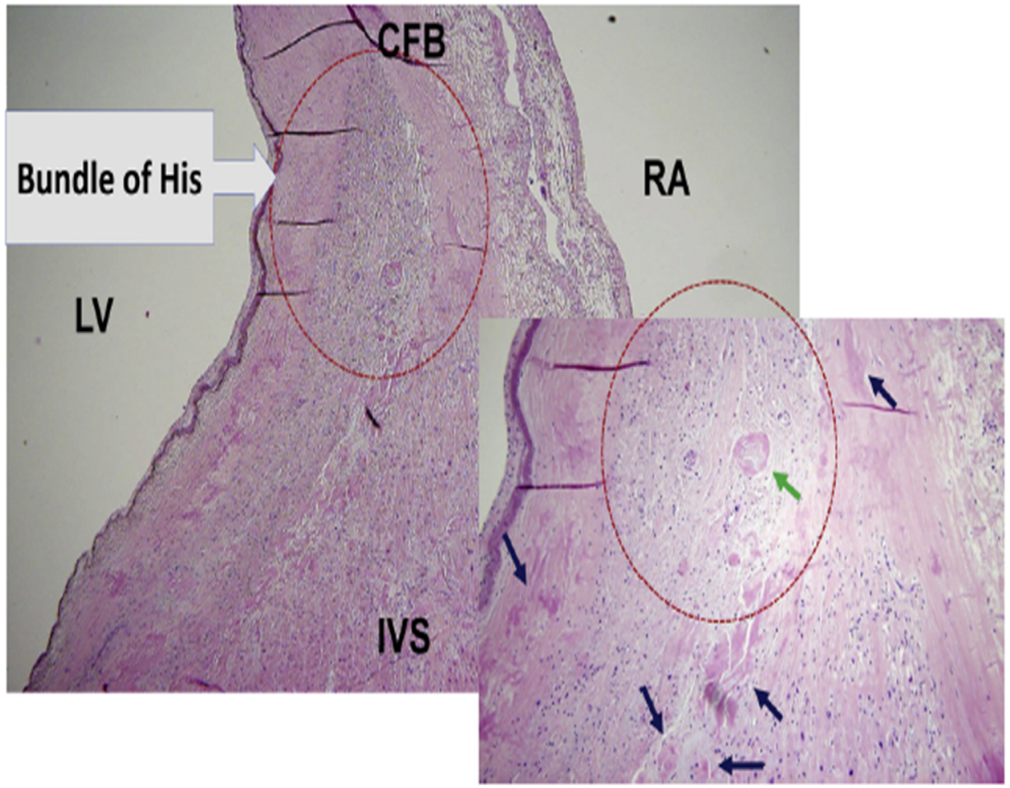
**CENTRAL ILLUSTRATION** Electrophysiologic Manifestations of Cardiac Amyloidosis




**Electrophysiological Manifestations of Cardiac Amyloidosis**

| Atrial Fibrillation | High Degree AV Block | Ventricular Arrhythmia | Dyssynchrony | Device Management |
|---|---|---|---|---|
| Anticoagulation<br>Rhythm or Rate Control<br>LAA Occlusion | AV Nodal Blocking Agents<br>Pacemaker | Role of ICD and Mortality | Cardiac Resynchronization Therapy | Pacemaker ICDs |

Hartnett, J. et al. J Am Coll Cardiol CardioOnc. 2021;3(4):506-515.




Troubles de la conduction
Rôle de la dysautonomie dans les ATTRm++

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*Harnett, J. et al. J Am Coll Cardiol CardioOnc. 2021;3 (4) 506–515*

# 1. Quand anticoaguler ?

- 1. En cas de FA
- 2. En rythme sinusal ?

# 1. Quand anticoaguler ?
# La fibrillation atriale est très prévalente dans l'AC

**Table 2** Prevalence of AF in AL and TTR amyloidosis in most recent studies

| Study, date | Population | Overall prevalence | Prevalence in TTR | Prevalence in AL |
|---|---|---|---|---|
| Longhi *et al.*, 2015[20] | *N* = 262 | | | |
| | 123 AL, 94 hTTR, 45 wtTTR | 15% | 35% | 9% |
| Mints *et al.*, 2018[23] | N = 146 wtTTR | 70% | 70% | — |
| Sanchis *et al.*, 2019[19] | *N* = 238, 115 AL, 97 wtTTR, 26 hTTR | 44% | 60% | 26% |
| Martinez-Naharro *et al.*, 2019[18] | *N* = 324, 166 TTR, 155 AL | | 46% | 14% |
| Mitrani *et al.*, 2020[21] | *N* = 290 TTR | 75% | 75% | — |
| Donellan *et al.*, 2020[22] | *N* = 265, 205 wt, 60 vTTR | 69% | 69% | — |

AL, immunoglobulin light chain amyloidosis; TTR, transthyretin; vTTR, variant transthyretin; wTTR: wild type transthyretin.

FA dans les séries « d'insuffisance cardiaque » : 13-27%

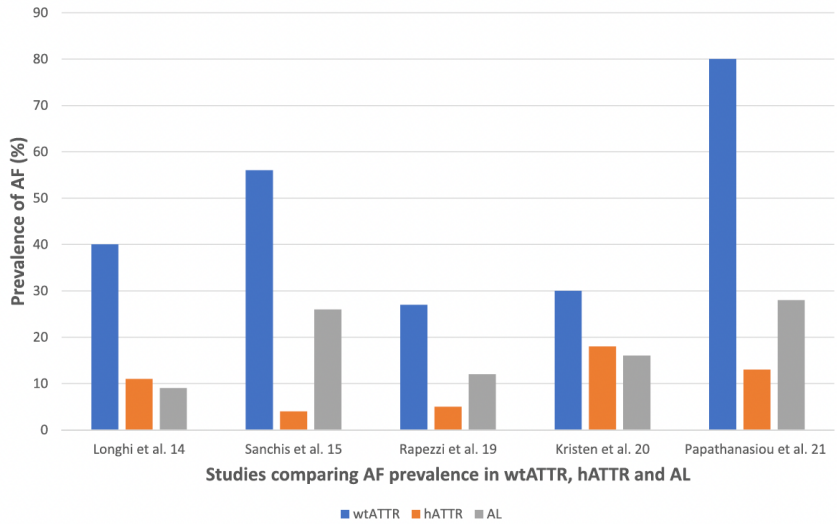


**Fig. 1.** Prevalence of atrial fibrillation in different types of cardiac amyloidosis. The prevalence of AF has consistently been seen to be higher in wtATTR compared with both hATTR and AL. AF, atrial fibrillation; AL, light-chain cardiac amyloidosis; hATTR, hereditary transthyretin cardiac amyloidosis; wtATTR, wild-type transthyretin cardiac amyloidosis.

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*Nicol et al. ESC Heart Fail. 2022 Feb;9(1):11-20*
*Bukhari et al. J Card Fail. 2023 Jan;29(1):76-86.*

# 1. Quand anticoaguler ?
## La fibrillation atriale est très prévalente dans l'AC

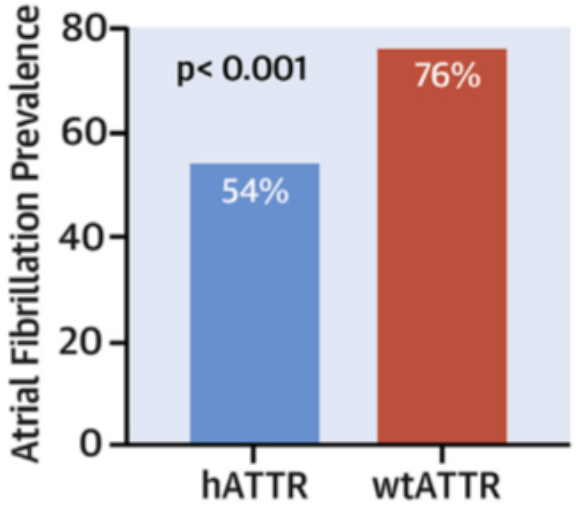
### Atrial Fibrillation in Transthyretin Cardiac Amyloidosis

**Predictors, Prevalence, and Efficacy of Rhythm Control Strategies**

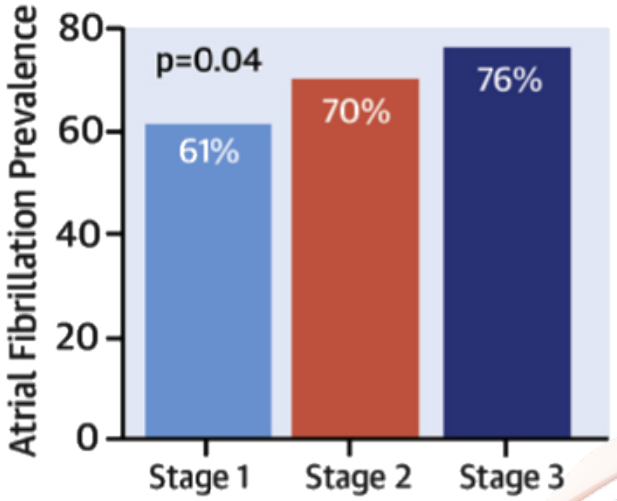
Eoin Donnellan, MD, Oussama M. Wazni, MD, Mazen Hanna, MD, Mohamed B. Elshazly, MD, Rishi Puri, MD, PhD, Walid Saliba, MD, Mohamed Kanj, MD, Sneha Vakamudi, MD, Divyang R. Patel, MD, Bryan Baranowski, MD, Daniel Cantillon, MD, Thomas Dresing, MD, Wael A. Jaber, MD

- 382 ATTR (111 ATTRv, 271 ATTRwt)
- 1 centre (Cleveland Clinic), 2004-2018
- Suivi médian = 35 mois
- Évaluation en fonction du NAC score

- **FA chez 265 patients (69 %) dont 33 % au diagnostic**
- **Temps médian entre dg d'ATTR et AFA : 15 mois**




NAC Score :
- ❑ Stade I : NT-proBNP ≤ 3000 ng/L et eGFR ≥ 45 ml/min
- ❑ Stade III : NT-proBNP >3000 ng/L et eGFR <45 ml/min
- ❑ Stade II : les autres

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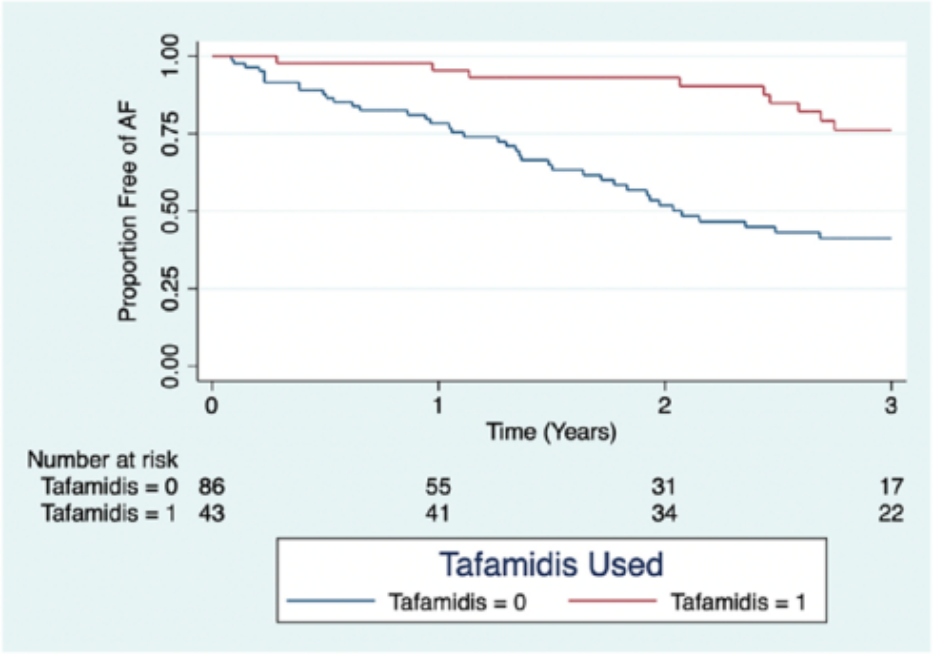
*Donnellan et al., J Am Coll Cardiol EP 2020, Gillmore et al., Eur Heart J 2018.*

# 1. Quand anticoaguler ?
## Le traitement étiologique ne neutralise pas le risque de FA

**FIGURE 1** Kaplan-Meier Atrial Fibrillation-Free Survival




AF = atrial fibrillation.

- 129 patients (43 % ATTRwt, 36 % ATTRv), sans ATCD de FA
- Monocentrique, Columbia, 2008-2019
- Suivi médian = 3.4 ± 2.4 ans

- Définition de la FA : enregistrement ≥ 30 sec
- Age médian 73,9 ans, FEVG médiane 49 %
- 33 % (n=43) des patients traités par Tafamidis

*Girvin et al. JACC Clin Electrophysiol. 2023 Apr;9(4):586-587.*

# 1. Quand anticoaguler ?
## La fibrillation atriale n'est pas corrélée à la mortalité

- **Pronostic incertain (patients très graves++)**

ARTICLE

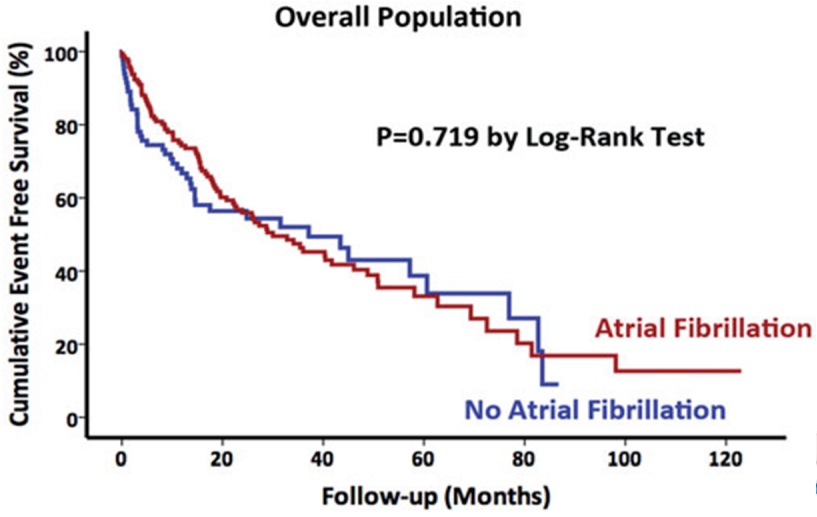
Check for updates

### Atrial fibrillation and subtype of atrial fibrillation in cardiac amyloidosis: clinical and echocardiographic features, impact on mortality

Kevin Sanchis[a,b], Eve Cariou[a,b,c], Magali Colombat[d], David Ribes[e,f], Antoine Huart[e,f], Pascal Cintas[g], Pauline Fournier[a,b], Anne Rollin[a], Didier Carrié[a,b,h], Michel Galinier[a,b,c], Philippe Maury[a,h,i], Alexandre Duparc[a] and Olivier Lairez[a,b,h,j], On behalf of the Toulouse Amyloidosis Research Network collaborators*

**Results:** One hundred and four (44%) patients had history of AF at the time of diagnosis: 62 (60%) permanent and 42 (40%) non-permanent. There were 30 (26%) and 74 (60%) patients with history of AF among patients with AL and ATTR (including 5 hereditary and 69 wild-type), respectively ($p<.0001$). During the follow-up, 48 new patients developed AF (29, 12 and 7 among patients with AL, wild-type ATTR and hereditary ATTR). After adjustment for age, survival was similar in patients with or without history of AF (HR 0.87 (95% CI, 0.60 to 1.27; $p=.467$). AF had no impact on cardiovascular mortality. Among the 152 patients with history of AF included in the whole study, there were 75 (49%) patients with permanent AF. After adjustment for age, survival was similar in patients with permanent and non-permanent AF: HR 1.29 (95% CI, 0.84 to 1.99; $p=.251$). The results were the same among patients with AL or wild-type amyloidosis. Subtype of AF had no impact on cardiovascular mortality.

**Conclusions:** AF is common in patients with CA. However, AF and clinical subtype of AF have no impact on all-cause mortality, whatever the type of amyloidosis.




*Sanchis et al. Amyloid. 2019 Sep;26(3):128-138.*

# 1. Quand anticoaguler ?
## Historiquement...

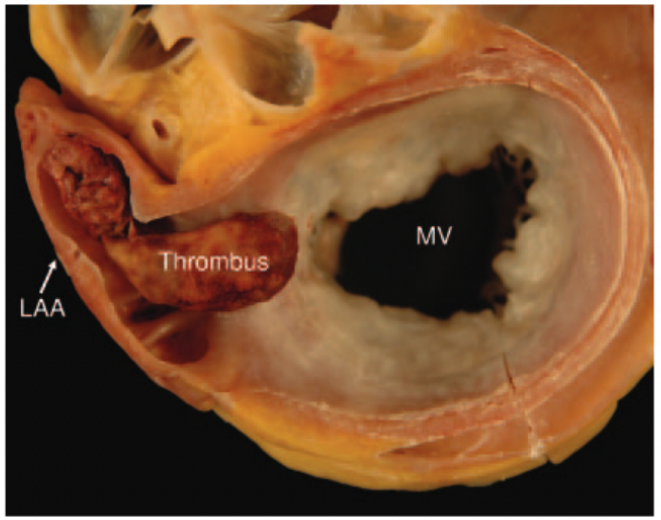



**Figure 1.** A representative intracardiac thrombus protruding from the LA appendage (LAA) in an AL heart. MV indicates mitral valve.

**Table 2. Characteristics in Patients With and Without Thromboembolism**

| | With (n=45) | Without (n=71) | P |
|---|---|---|---|
| Age, y | 65±2.2 | 77±1.8 | <0.0001 |
| Gender, % male | 58 | 62 | 0.67 |
| Body mass index, kg/m$^2$ | 26.0±0.9 | 26.7±0.8 | 0.54 |
| Hypertension, % | 17 | 41 | 0.008 |
| Diabetes mellitus, % | 7 | 19 | 0.08 |
| CHF, % | 65 | 71 | 0.55 |
| NYHA class (1–4) | 2.4±1.3 | 2.3±1.2 | 0.65 |
| AF, % | 37 | 33 | 0.64 |
| Anticoagulation, % | 36 | 28 | 0.42 |
| Cancer, % | 15 | 22 | 0.41 |
| Syncope, % | 34 | 17 | 0.08 |
| AL amyloidosis, % | 73 | 31 | <0.0001 |
| Stem cell transplant, % | 23 | 10 | 0.09 |
| Recent operation, % | 10 | 17 | 0.31 |
| History of thrombosis, % | 27 | 24 | 0.76 |
| Systolic BP, mm Hg | 111±18 | 128±24 | 0.0005 |
| Diastolic BP, mm Hg | 67±12 | 69±15 | 0.54 |
| Creatinine, mg/dL | 2.2±1.7 | 1.9±1.1 | 0.17 |
| CAD by autopsy, % | 29.3 | 55.4 | 0.008 |
| CAD severity score (0–4)* | 2.0±1.2 | 2.7±1.3 | 0.02 |

Data are shown as mean±SD unless otherwise indicated.
*CAD severity score: 0=no, 1=minimal, 2=mild, 3=moderate, and 4=severe.

**Table 3. TTE Characteristics in Subjects With and Without Thromboembolism**

| | With (n=35) | Without (n=47) | P |
|---|---|---|---|
| Time from TTE to death, d | 15 (1-55) | 43 (2-209) | 0.12 |
| HR at echocardiography, bpm | 86±17 | 79±14 | 0.02 |
| LV end-diastolic diameter, mm | 44.3±7.8 | 48.1±8.1 | 0.04 |
| LV end-systolic diameter, mm | 32.4±9.3 | 33.1±9.8 | 0.76 |
| LV septal thickness, mm | 14.2±3.9 | 12.9±3.3 | 0.12 |
| LV posterior wall thickness, mm | 13.8±3.6 | 12.2±3.1 | 0.03 |
| RV free wall thickness, mm | 8.3±3.6 | 5.9±2.1 | 0.0007 |
| LA volume index, mL/m$^2$ | 40.1±14.9 | 48.2±35.2 | 0.21 |
| RA enlargement (0–3) | 1.8±1.2 | 1.4±1.1 | 0.08 |
| Stroke volume, mL | 51.1±20.9 | 67.6±23.4 | 0.002 |
| RV systolic pressure, mm Hg | 44.3±9.4 | 51.1±15.9 | 0.045 |
| LVEF, % | 46±19 | 54±15 | 0.03 |
| LV diastolic function grade (0–4) | 3.1±1.1 | 1.9±0.8 | 0.0001 |
| Mitral deceleration time, ms | 160±37 | 193±60 | 0.006 |
| Mitral E velocity, m/s | 0.87±0.21 | 0.90±0.26 | 0.65 |
| Mitral A velocity, m/s | 0.27±0.29 | 0.52±0.33 | 0.0008 |
| E/A | 3.4±2.6 | 2.0±1.9 | 0.03 |
| Mitral annulus e′ velocity, cm/s | 4.4±2.3 | 5.9±2.7 | 0.06 |
| Mitral annulus a′ velocity, cm/s | 2.3±3.3 | 6.6±4.7 | 0.003 |
| E/e′ | 23±12 | 16±8 | 0.02 |
| PV systolic velocity, m/s | 0.32±0.18 | 0.48±0.20 | 0.002 |
| PV diastolic velocity, m/s | 0.65±0.20 | 0.60±0.18 | 0.37 |
| PV A velocity, m/s | 0.14±0.13 | 0.24±0.13 | 0.008 |
| PV diastolic/systolic ratio | 3.0±2.3 | 1.5±0.8 | 0.001 |

Data are shown as mean±SD. PV indicates pulmonary venous flow.

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*Feng et al. Circulation. 2007 Nov 20;116(21):2420-6.*

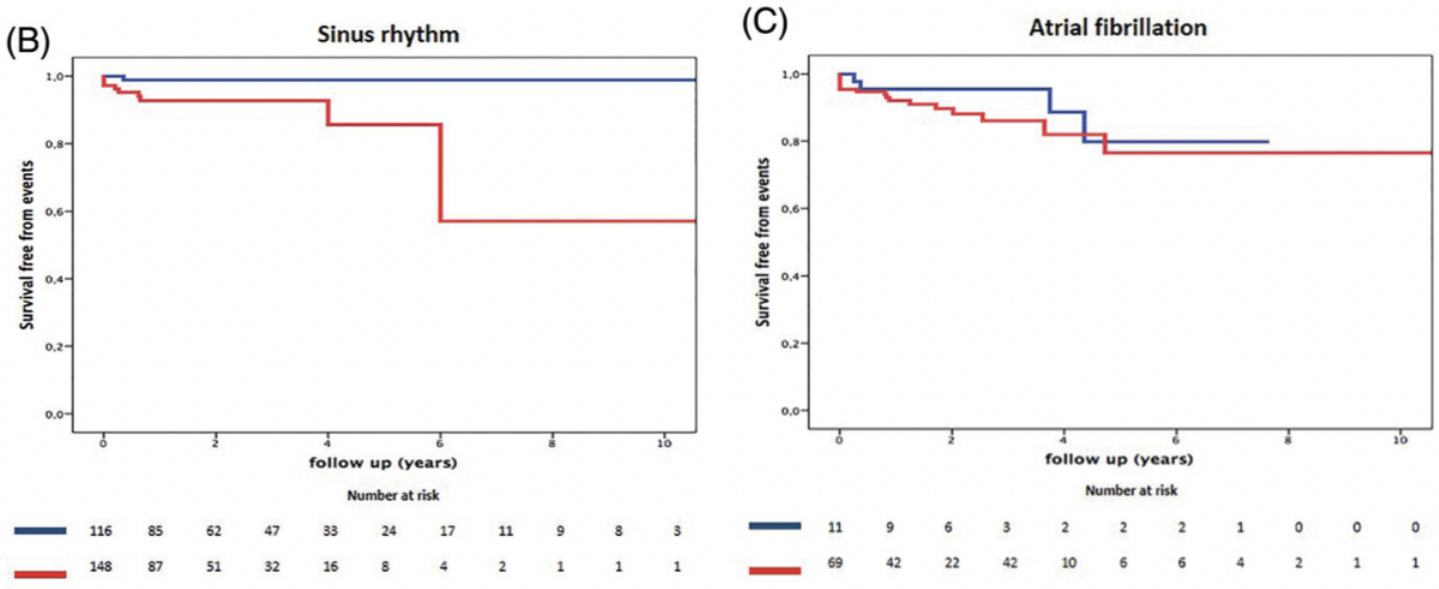
# 1. Quand anticoaguler ?
## Plus récemment

### Arterial thrombo-embolic events in cardiac amyloidosis: a look beyond atrial fibrillation

Francesco Cappelli[a,b], Giacomo Tini[c,d], Domitilla Russo[e], Michele Emdin[f,g], Annamaria Del Franco[f,g], Giuseppe Vergaro[f,g], Gianluca Di Bella[h], Anna Mazzeo[h], Marco Canepa[c,d], Massimo Volpe[e,i], Federico Perfetto[a], Camillo Autore[e], Carlo Di Mario[b], Claudio Rapezzi[j,k] and Maria Beatrice Musumeci[e]

**Table 2.** Relationship between clinical, morphologic and functional features at initial evaluation and occurrence of arterial thrombo-embolic events in patients with cardiac amyloidosis.

| | Univariate | | |
|---|---|---|---|
| | Hazard ratio | 95% CI | *p* |
| Female sex | 0.53 | 0.16–1.82 | .31 |
| Transthyretin cardiac amyloidosis | 0.50 | 0.13 – 1.92 | .31 |
| Age ≥75 years | 1.34 | 0.55–3.24 | .52 |
| Chronic kidney disease | 1.94 | 0.77–4.87 | .16 |
| Atrial fibrillation at first evaluation | 1.65 | 0.80–3.39 | .17 |
| $CHA_2DS_2$-VASc score ≥3 | 2.84 | 1.02–7.92 | .05 |
| Left ventricular ejection fraction ≤50% at first evaluation | 1.95 | 0.83–4.60 | .13 |
| Anticoagulation therapy | 1.23 | 0.52–2.92 | .64 |




Facteurs prédictifs d'évènements thrombo emboliques

Patient en FA :
- CHADS-VASc non prédictif d'AEs

Patients en RS :
- CHADS-VASC ≥ 3 prédictif d'AEs

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*Cappelli et al, Amyloid 2020.*

# 1. Quand anticoaguler ?
## Plus récemment

- **AL :**
  - **Moins de FA**
  - **Plus de thrombi**

**TABLE 2** Intracardiac Thrombus in Cardiac Amyloid Patients

| First Author (Year) Ref. # | Demographics | Patients With Intracardiac Thrombus | Patients With Multiple Thrombi | Location of Thrombus | Method of Diagnosis | AL Amyloid With Thrombus | ATTR Amyloid With Thrombus |
|---|---|---|---|---|---|---|---|
| Roberts et al. (1983) (35) | N = 54<br>49 AL<br>5 hATTR | 14 (26) | 5 | 10 RA<br>7 LA<br>2 RV<br>3 LV | Pathology | NR | NR |
| Feng et al. (2007) (20) | N = 116<br>55 AL<br>2 hATTR<br>55 wtATTR<br>4 AA | 38 (33) | 15 | 40 RA<br>19 LA<br>3 RV<br>1 LV | Pathology | 51 | 16 |
| Feng et al. (2009) (21) | N = 156<br>80 AL<br>17 hATTR<br>56 wtATTR | 42 (27) | 8 | 19 RA<br>32 LA | TEE | 67 | 18 |
| Mints et al. (2018) (13) | N = 39<br>39 wtATTR | 6 (15) | NR | 6 LA | TEE | N/A | 15 |
| El-Am et al. (2019) (33) | N = 46<br>24 AL<br>21 ATTR | 13 (28) | NR | 13 LA | TEE | 25 | 33 |
| Donnellan et al. (34) | N = 100<br>25 hATTR<br>75 wtATTR | 30 (30) | NR | 30 LA | TEE | N/A | 30 |

Values are n (%) or %, unless otherwise indicated.

LA = left atrium; LV = left ventricle; N/A = not applicable; RA = right atrium; RV = right ventricle; TEE = transesophageal echocardiogram; other abbreviations as in Table 1.

*Giancaterino S, et al. JACC Clin Electrophysiol. 2020 Apr;6(4):351-361.*

# 1. Quand anticoaguler ?
## Que nous disent les recommandations pour les patients en RS?

Position Paper ESC et SFC pour les patients en RS : aires d'incertitude

PNDS : Le risque élevé continu d'événements thromboemboliques chez les patients atteints d'une amylose cardiaque justifie de **discuter pour chaque patient des bénéfices** (prévention de la thrombose) et des **risques hémorragiques** (atteinte cutanée importante ou gastrique) d'une anticoagulation à dose efficace. Il est nécessaire de demander un avis au centre expert le plus proche.

| | Risques Thrombotiques | Risques Hémorragiquse |
|---|---|---|
| **AL** | -Profil transmitral restrictif<br>-Onde E unique avec onde A absente sur le flux transmitral<br>-Hyperexcitabilité atriale ou fibrillation atriale<br>-Dysfonction VG | -Lésions cutanées ou muqueuses importantes<br>-Déficit en facteur X |
| **ATTRwt** | | |
| **ATTRv** | | -Lésions digestives avec antécédents de saignement.<br>-Mutation ATTR Val30Met p.(Val50Met) avec antécédent de greffe hépatique |

*Tableau 7 : Anomalies associées au risque thrombotique ouhémorragique dans les amyloses cardiaques*

*PNDS Amyloses Cardiaques – 2021.*

ESC European Society of Cardiology
European Heart Journal (2024) **45**, 3314–3414
https://doi.org/10.1093/eurheartj/ehae176





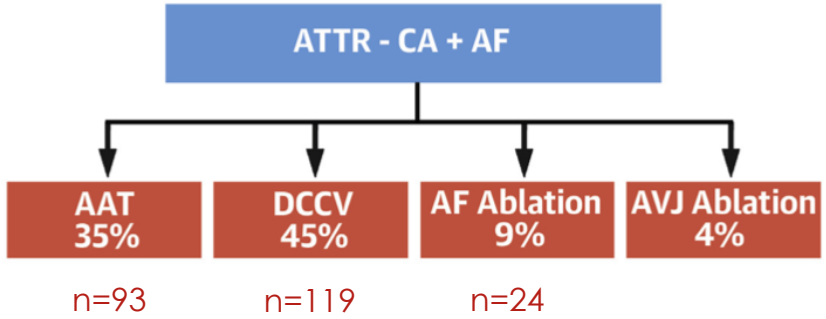
# 2024 ESC Guidelines for the management of atrial fibrillation developed in collaboration with the European Association for Cardio-Thoracic Surgery (EACTS)

**Developed by the task force for the management of atrial fibrillation of the European Society of Cardiology (ESC), with the special contribution of the European Heart Rhythm Association (EHRA) of the ESC.**
***Endorsed by the European Stroke Organisation (ESO)***

| | | |
|---|---|---|
| anticoagulation. | | |
| Oral anticoagulation is recommended in all patients with AF and hypertrophic cardiomyopathy or cardiac amyloidosis, regardless of $CHA_2DS_2$-VA score, to prevent ischaemic stroke and thromboembolism. | I | B |

# La Cardioversion

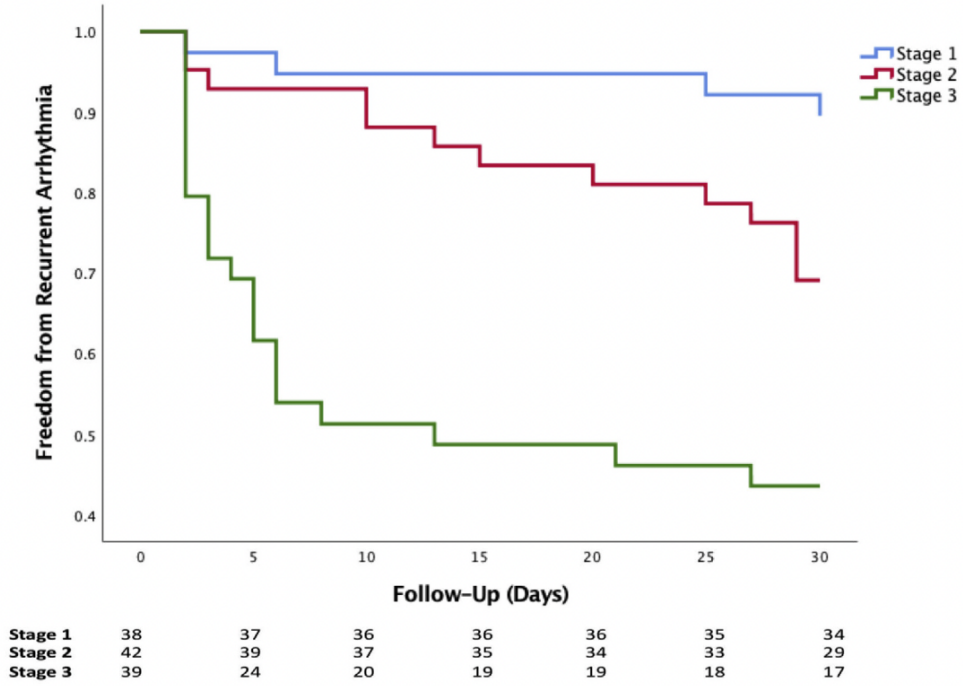
- **Cardioversion (taux de succès environ 50 % à 30 jours). Dépend de l'évolution de la pathologie**




**Après AAT (medt)** : 31% maintiennent le RS (53 % stade 1 *vs* 17 % stade 3)
**Après DCCV (CEE)** :

- Efficacité immédiate : 95 % (97 % stade 1; 95 % stade 2 ; 92 % stade 3 ; p 0.58).
- A 30 jours : 61 % en RS (90% stade 1 ; 60 % stade 2 ; 33 % stade 3)
- A 1 an : 41 % en RS (74% stade 1 ; 33 % stade 2 ; 18 % stade 3)

**Après ablation de FA** : Après un suivi médian de 40 mois : récidive chez 58 % (36 % stade 1 ou 2 ; 90 % stade 3 (p 0.005)




**30-day maintenance of NSR following DCCV**

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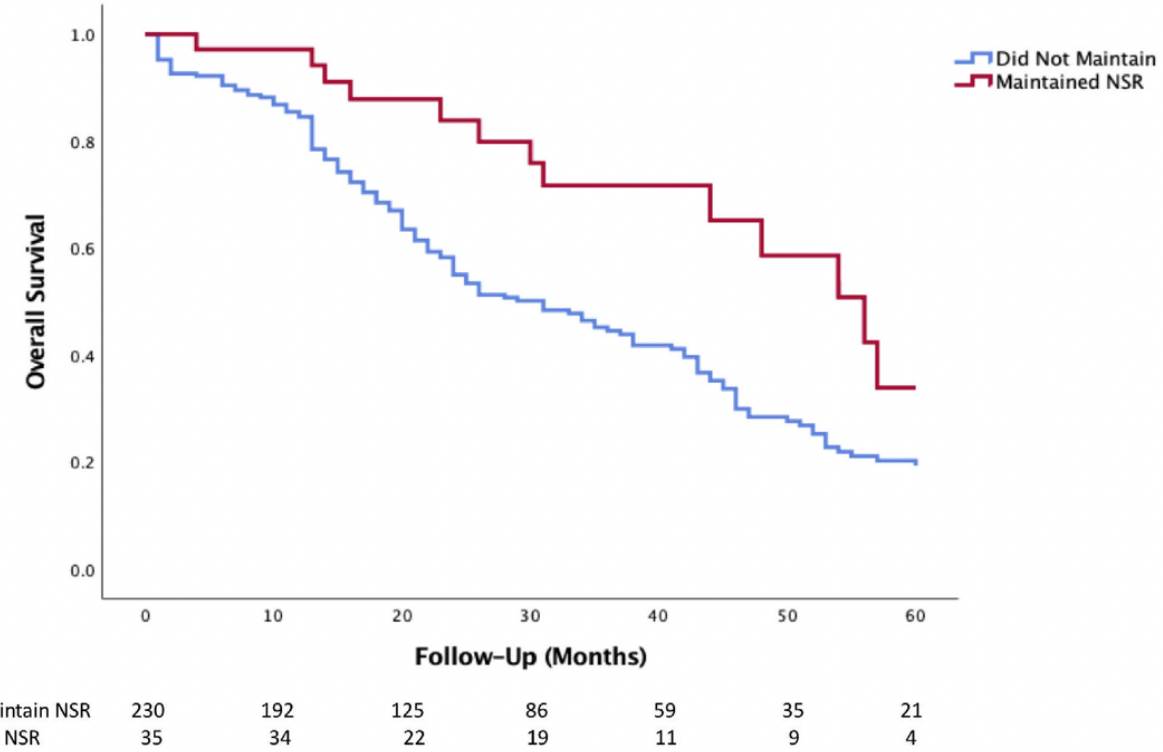
*Donnellan et al. JACC Clin Electrophysiol. 2020 Sep;6(9):1118-1127.*

# 2. Que faire des TSV ?
## Cardioversion ?

**Meilleur pronostic en cas de maintien du RS après cardioversion ++**




| | | | | | | | |
|---|---|---|---|---|---|---|---|
| Did Not Maintain NSR | 230 | 192 | 125 | 86 | 59 | 35 | 21 |
| Maintained NSR | 35 | 34 | 22 | 19 | 11 | 9 | 4 |

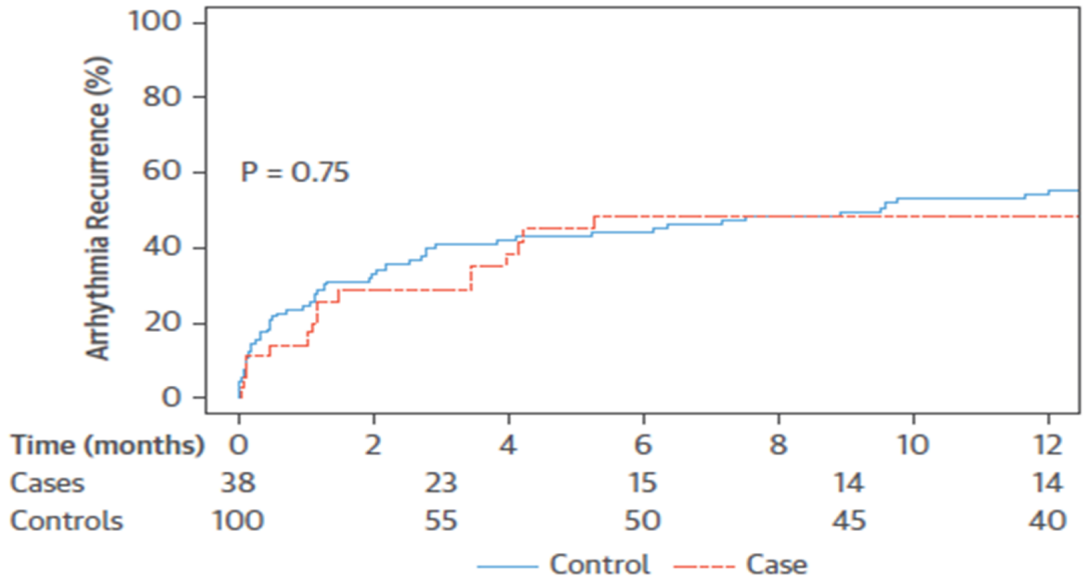
*Donnellan et al. JACC Clin Electrophysiol. 2020 Sep;6(9):1118-1127.*

# Cardioversion ? Oui, mais…

## Direct Current Cardioversion of Atrial Arrhythmias in Adults With Cardiac Amyloidosis

Edward A. El-Am, MD,[a] Angela Dispenzieri, MD,[b] Rowlens M. Melduni, MD, MPH,[a] Naser M. Ammash, MD,[a] Roger D. White, MD,[c] David O. Hodge, MS,[d] Peter A. Noseworthy, MD,[a] Grace Lin, MD,[a] Sorin V. Pislaru, MD, PHD,[a] Alexander C. Egbe, MBBS, MPH,[a] Martha Grogan, MD,[a] Vuyisile T. Nkomo, MD, MPH[a]

**FIGURE 1** Rate of Atrial Arrhythmia Recurrence Following Successful DCCV in Patients With CA Compared With Control Patients




- 58 AC comparé à une population sans amylose
- 28 % d'annulation de cardioversion chez les AC vs. 7 % ; p < 0.001 pour cause de thrombus malgré anticoagulation efficace ou FA < 48h
- Succès cardioversion identique (90 % *vs* 94 % ; p = 0.4)
- Plus de **complications** chez les AC : arythmie ventriculaire et **bradycardie nécessitant PM**
- ➔ Taux élevé de thrombus intra-OG ➔ ETO systématique avant CEE

**TABLE 3** TEE Data of CA Compared With Control Patients

| | Cardiac Amyloidosis (n = 46) | Control Group (n = 79) | p Value |
|---|---|---|---|
| Spontaneous echocardiogram contrast | 31 (67) | 34 (43) | 0.01 |
| Thrombus identified on echocardiogram | 13 (28) | 2 (2.5) | <0.001 |
| Echo LAA emptying velocity, cm/s | 20.6 ± 14.1 (n = 38) | 33.9 ± 18.4 (n = 65) | <0.001 |

Values are n (%) or mean ± SD.
LAA = left atrial appendage; other abbreviations as in Table 1.

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*El-Am et al. J Am Coll Cardiol. 2019 Feb 12;73(5):589-597.*

# 2. Que faire des TSV ?
## Éliminer thrombus intracardiaque+++

### Electrical cardioversion of atrial arrhythmias with cardiac amyloidosis in the era of direct oral anticogulants

Olivier Touboul[1], Vincent Algalarrondo[2], Silvia Oghina[1], Nathalie Elbaz[1], Segolene Rouffiac[1], David Hamon[1], Fabrice Extramiana[2], Estelle Gandjbakhch[3], Thomas D'Humieres[4], Eloi Marijon[5], Tarvinder S. Dhanjal[6], Emmanuel Teiger[1], Thibaud Damy[1] and Nicolas Lellouche[1]*

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- 67 patients
- Tous anticoagulés sauf 1 (AOD dans 74 % des cas)
- Imagerie à la recherche de thrombus systématique (ETO ou scanner)

- **14 % des cardioversions annulées pour cause de thrombus**

- Résolution complète du thrombus dans seulement 17 % des cas.
- Deux facteurs prédictifs de thrombus : créatinine, AAP

- 88 % de succès. Récidive (sous amiodarone) dans 51 % des cas après un suivi médian de 30 ± 27 mois



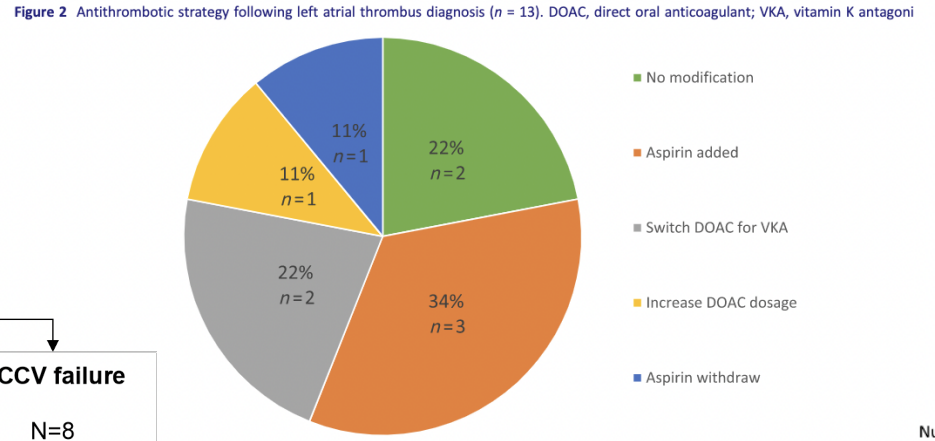
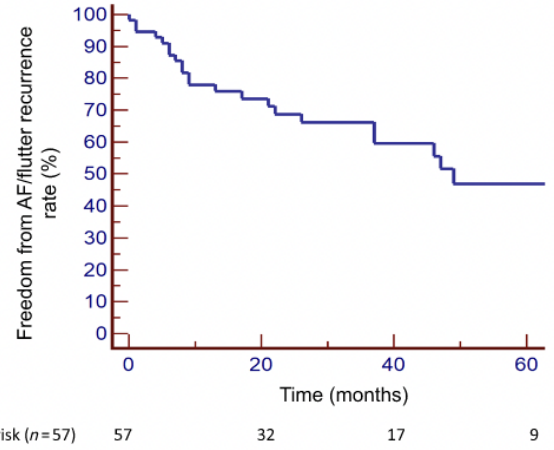




**Figure 2** Antithrombotic strategy following left atrial thrombus diagnosis ($n$ = 13). DOAC, direct oral anticoagulant; VKA, vitamin K antagoni

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*Touboul et al. ESC Heart Fail. 2022 Oct;9(5):3556-3564.*

**FIGURE 1** Breakdown of Outcomes in Scheduled DCCV Procedures

192 scheduled DCCV procedures (total number of CA patients = 108)

Pre-DCCV TEE

no

yes

132 (69%) procedures (n = 61)

60 (31%) TEE studies (n = 47)

48 TEE studies did not reveal LAAT

12 TEE studies revealed LAAT and DCCV was aborted

n = 132

n = 48

- Repeat pre-DCCV TEE was performed in 5 cases:
- 3 studies showed LAAT resolution
- 2 studies showed persistent LAAT and DCCV was again aborted
- DCCV without repeat TEE was performed in 4 cases
- Sinus restoration was not pursued in 3 cases

n = 7

187 DCCV procedures

- Procedural success in 185 (99%)
- Thromboembolic complications 0

The outcomes of 192 scheduled DCCV procedures as well as the findings of 60 TEE studies that were performed before DCCV. CA = cardiac amyloidosis; DCCV = direct current cardioversion; LAAT = left atrial appendage thrombus; TEE = transesophageal echocardiography.

# 2. Que faire des TSV ?
## FAG

- Fermeture de l'auricule gauche avec thrombus intra auriculaire (ici protection carotidienne ou non)

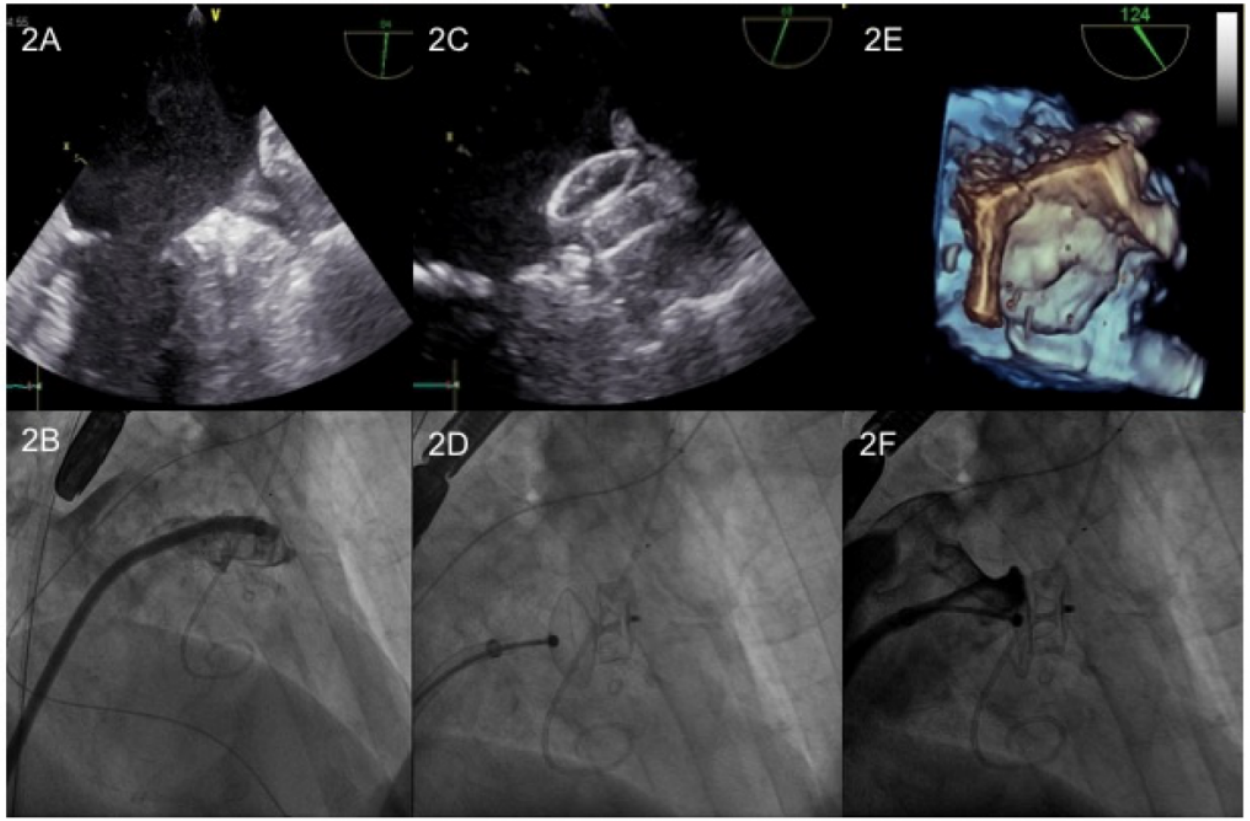


FIGURE 2. (A) Transesophageal echocardiography (TEE) showing thrombus in the left atrial appendage (LAA). (B) Angiography of the LAA. Amplatzer Amulet device was deployed and its stability was confirmed by Minnesota maneuver at (C) TEE and (D) fluoroscopy. (E) Three-dimensional TEE showing the Amplatzer Amulet device in place. (F) No leaks were documented at final angiography.

*Bordignon S, et al. J Cardiovasc Electrophysiol. 2019 Dec;30(12):2858-2863.*

# Quand/qui anticoaguler ? Que faire de la TSV ? Dans notre pratique

- Traquer la FA +++
  - HolterECG / 6 mois
  - Discuter Reveal si palpitations
  - Anticoagulation de la FA quel que soit le score CHA2DS2-VASc
- Anticoaguler si absence de systole atriale en ETT (onde A)
- Discuter anticoagulation en rythme sinusal au cas par cas (CHA2DS2-VASc élevé, profil mitral restrictif, dysfonction VG)

- Toujours vérifier l'absence du thrombus avant la cardioversion (ETO/scanner/IRM)

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# Indications de PM et DAI

# 3. Quelles surveillance conductive ?
## Prévalence élevée des troubles conductifs

ELSEVIER

The American Journal of Cardiology
Volume 128, 1 August 2020, Pages 140-146

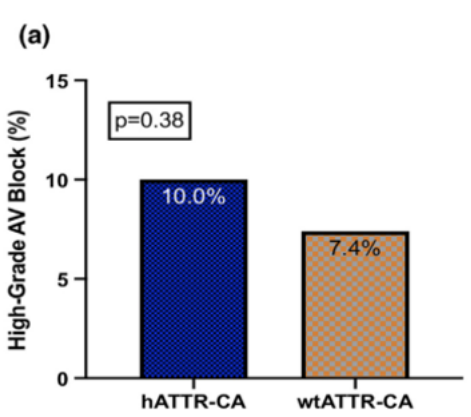
### Prevalence, Incidence, and Impact on Mortality of Conduction System Disease in Transthyretin Cardiac Amyloidosis

Eoin Donnellan MD, Oussama M. Wazni MD, Walid I. Saliba MD, Mazen Hanna MD, Mohamed Kanj MD, Divyang R. Patel MD, Bryan Wilner MD, Arshneel Kochar MD, Wael A. Jaber MD

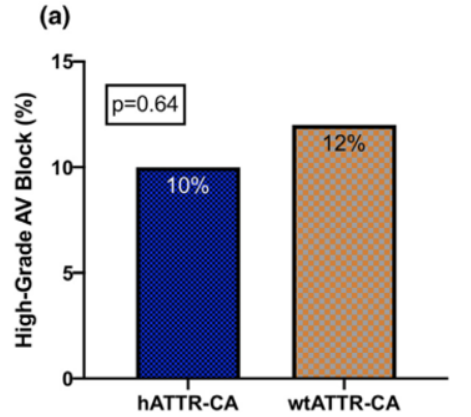
- 369 patients avec ATTR (261 ATTRwt and 108 hATTRv)
- 1 centre (Cleveland Clinic), 2004-2019
- Suivi médian = 28 mois
- **Au diagnostic :**
  - 35 patients (9.5 %) BAV haut degré
  - Environ 50 % ont QRS larges et BAV1
- **Pendant le suivi :** dysfonction sinusale dans 7 % des cas et BAV haut degré dans 11 % des cas
- Stade NAC élevé ou CPI associée associés à un excès de mortalité en cas de troubles conductifs de haut degré

Table 1
Baseline characteristics of patients with wild-type (wtATTR-CA) and hereditary (hATTR-CA) transthyretin cardiac amyloidosis

| Variable | wtATTR-CA (n = 261) | hATTR-CA (n = 108) | p-Value |
|---|---|---|---|
| Sinus rhythm | 169 (65%) | 94 (87%) | <0.001 |
| Atrial fibrillation | 87 (33%) | 14 (13%) | <0.001 |
| Junctional rhythm | 5 (2%) | 0 | 0.15 |
| PR interval (ms) | 212±65 | 201±50 | 0.14 |
| QRS duration (ms) | 127±35 | 118±31 | 0.02 |
| QTc duration (ms) | 488±51 | 478±42 | 0.11 |
| 1st degree AV block | 83 (49%) | 40 (43%) | 0.31 |
| Wide QRS complex | 133 (51%) | 52 (48%) | 0.62 |
| Left bundle branch block | 39 (15%) | 10 (9%) | 0.18 |
| Right bundle branch block | 48 (18%) | 19 (18%) | 0.86 |
| Interventricular conduction delay | 34 (13%) | 23 (21%) | 0.05 |
| Bifascicular block | 3 (1%) | 1 (1%) | 0.85 |
| Trifascicular block | 1 (0.4%) | 1 (0.9%) | 0.52 |
| High-grade AV block | 27 (10%) | 8 (7%) | 0.38 |




**PRÉVALENCE**




**INCIDENCE**

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*Donnellan et al., Am J Cardiol. 2020 Aug 1;128:140-146.*

# 3. Quelles surveillance conductive ?
## Prévalence élevée des troubles conductifs

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- Facteurs prédictifs de survenue de bloc de haut degré : durée du QRS de plus de 120 ms.
- Facteurs plutôt « protecteurs » : QRS fin < 100 ms et rythme sinusal.

➔ Donc QRS large = surveillance rapprochée (holterECG)

Table 3
Univariable and multivariable Cox regression analyses for the development of high-grade AV block

| | Univariable | | Multivariable | |
|---|---|---|---|---|
| Variable | HR (95% CI) | p-Value | HR (95% CI) | p-Value |
| Age | 0.99 (0.96-1.03) | 0.61 | | |
| ATTR-CA stage | 1.18 (0.79-1.77) | 0.41 | | |
| eGFR | 1.01 (0.99-1.04) | 0.26 | | |
| Obstructive CAD | 1.58 (0.82-3.03) | 0.17 | | |
| Digoxin use | 0.28 (0.04-2.02) | 0.21 | | |
| Beta blocker use | 1.26 (0.68-2.34) | 0.46 | | |
| hATTR-CA | 0.81 (0.41-1.61) | 0.55 | | |
| Diabetes mellitus | 1.18 (0.58-2.4) | 0.65 | | |
| Normal sinus rhythm | 0.34 (0.19-0.63) | 0.001 | 0.39 (0.21-0.73) | 0.003 |
| Atrial fibrillation | 2.27 (0.89-5.8) | 0.09 | 1.97 (0.77-5.05) | 0.16 |
| PR interval ≥200 msec | 0.73 (0.28-1.9) | 0.52 | | |
| QRS duration ≥120 ms | 5.2 (2.2-12.37) | <0.001 | 4.71 (1.97-11.26) | <0.001 |
| QRS duration <100 msec | 0.15 (0.055-0.43) | <0.001 | 0.17 (0.06-0.49) | 0.001 |
| Ejection fraction | 0.99 (0.97-1.01) | 0.3 | | |
| LV mass index | 1.0006 (0.997-1.02) | 0.2 | | |

CAD = coronary artery disease; eGFR = estimated glomerular filtration rate; LV = left ventricle.

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*Donnellan et al., Am J Cardiol. 2020 Aug 1;128:140-146.*

# 3. Quelles surveillance conductive ?
## Prévalence élevée des troubles conductifs

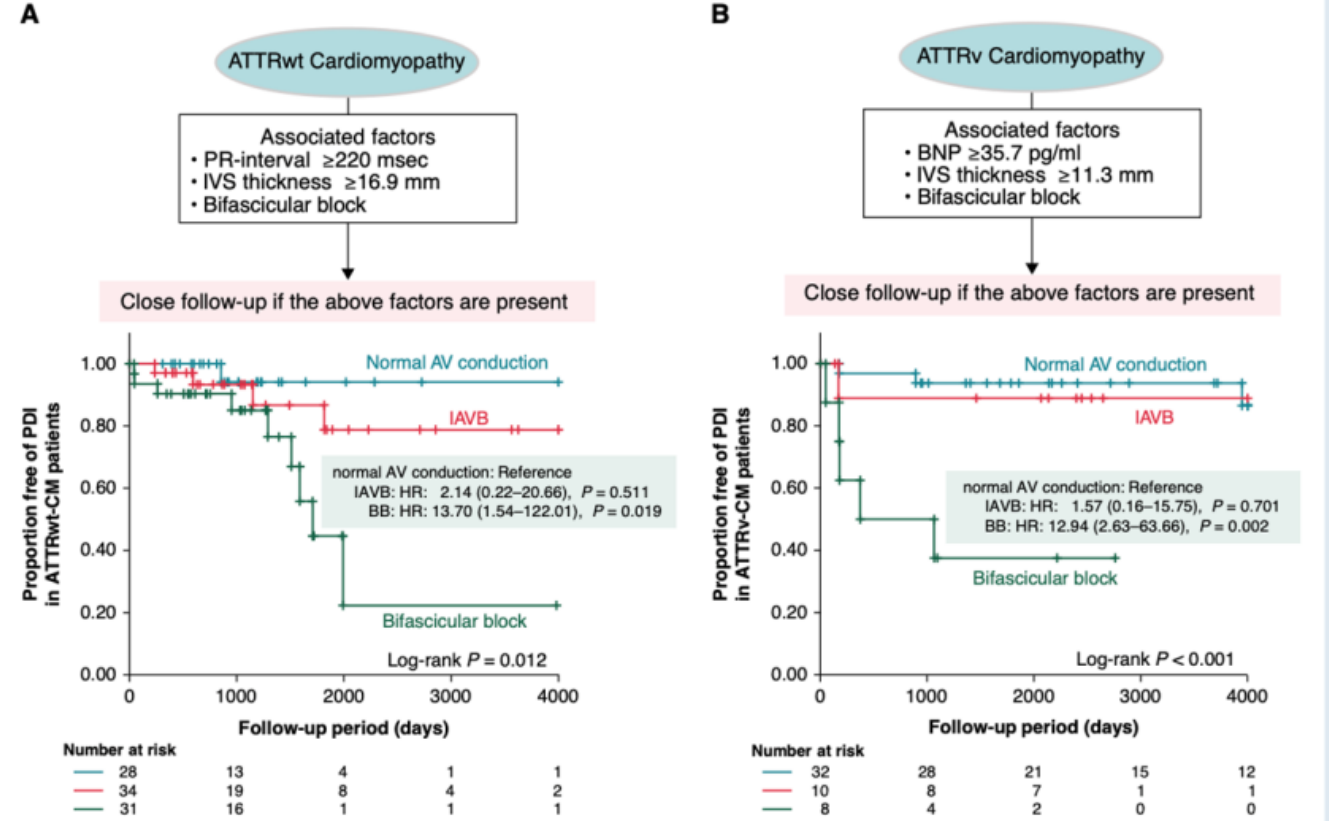
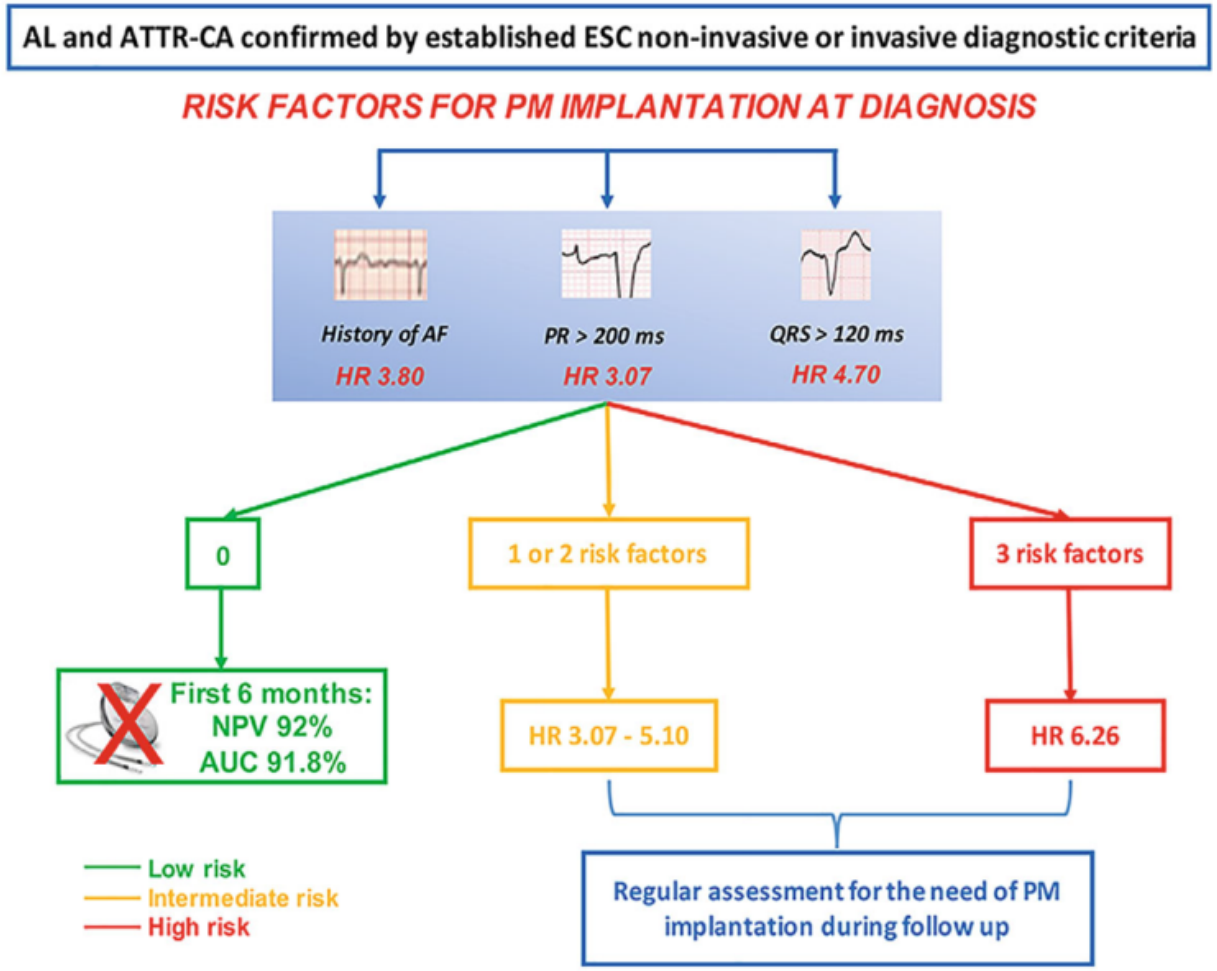



**Figure 3** Incidence of pacing device implantation in each conduction disturbance at diagnosis. Kaplan-Meier analysis of the comparison of pacing device implantation (PDI) between normal atrioventricular (AV) conduction, first-degree AV block (IABV) alone, and bifascicular block at the time of diagnosis are shown in patients with wild-type transthyretin amyloid cardiomyopathy (ATTRwt-CM) (*panel A*) and hereditary transthyretin amyloid cardiomyopathy (ATTRv-CM) (*panel B*). HR, hazard ratio.

- **114 patients** ATTRwt et **50** ATTRv)
- Suivi médian = non donné
- Rétrospectif monocentrique entre 1999 et 2018
- **Pendant le suivi :** 15 wtATTR (13%) et 14 hATTR (28%) ont nécessité un PM cliniquement indiqué selon les recos
- **En analyse multivariée** , les facteurs décrits dans les figures de droite étaient associés au rique de pacemaker au suivi
- Le risque de PM chez les patients ATTRwt et d'ATTRv présentant un bloc bifasciculaire (BBD+HB ou BBG) était significativement plus élevé que chez ceux présentant une conduction auriculo-ventriculaire normale, mais pas en cas de BAV1 isolé

*Kawahara et al Europace (2023) 25, 1–13*

# 3. Quelles surveillance conductive ?
## Prévalence élevée des troubles conductifs




- **405 patients** avec ATTR (56% ATTRwt and 14% hATTRv) et AL (29%)
- Suivi médian = 33 mois
- Rétrospectif multicentrique entre 2017 et 2020
- **Pendant le suivi :** 36 (9%) ont nécessité un PM cliniquement indiqué selon les recos
- **En analyse multivariée** , un antécédent de FA, l'intervalle PR et un QRS >120 ms sur l'ECG de base étaient indépendamment associés à l'implantation d'un PM
- L'absence de ces trois facteurs avait une valeur prédictive négative de 92 % avec une aire sous la courbe de 91,8 % à 6 mois.

*Kawahara et al Europace (2023) 25, 1–13*

# 3. Quelles surveillance conductive ?
## Prévalence élevée des troubles conductifs

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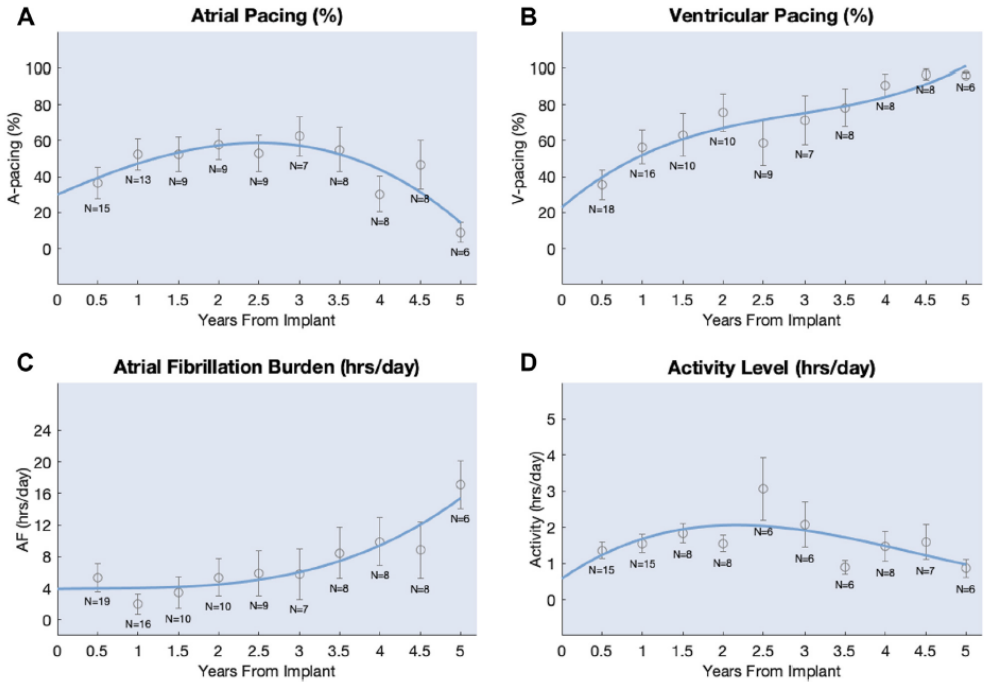
### Cardiac Implantable Electronic Devices

**A Window Into the Evolution of Conduction Disease in Cardiac Amyloidosis**

Michael R. Rehorn, MD, MS,* Rahul S. Loungani, MD,* Eric Black-Maier, MD, Amanda C. Coniglio, MD, Ravi Karra, MD, MHS, Sean D. Pokorney, MD, MBA, Michel G. Khouri, MD

| Native conduction findings | |
|---|---|
| Native PR (ms) | 259 ± 19 |
| Native QRS interval, ms | 125 ± 6 |
| Native QRS morphology† | |
| Narrow | 9 (33) |
| LBBB | 3 (11) |
| RBBB | 10 (37) |
| IVCD | 5 (19) |

- 22 (65%) patients implantés d'un pacemaker :
  - 6 pour dysfonction sinusale (27 %),
  - 14 pour BAV haut degré (64 %),
  - 2 pour MRO (9 %).
- 12 (35 %) patients implantés d'un DAI :
  - 9 en prévention primaire [7 ATTR, 2 AL]
  - 3 en prévention secondaire [2 ATTR, 1 AL]
- 10 patients (24%) implantés d'un PM ou DAI biventriculaire.
- L'implantation du PM/DAI a été réalisée en médiane 4.5 ± 5.4 ans avant le diagnostic chez 14 patients (41 %) et 1.2 ± 1.5 ans après le diagnostic chez 20 patients (59 %).




**Pacing ventriculaire :**

- **56 ± 9 % à 1 an et 96 ± 1 % à 5 ans** (p<0.03), avec la plupart des patients avec un pacing ventriculaire proche de 100 %

**Arythmie atriale :**

- ➔ 21 patients (62 %) dépistés d'une FA ou d'un flutter atriale au cours du suivi
- ➔ Après un suivi de 3.1 ± 4.0 ans, **28 patients (82 %) étaient en arythmie atriale**

AP-HP. Hôpitaux universitaires Henri-Mondor

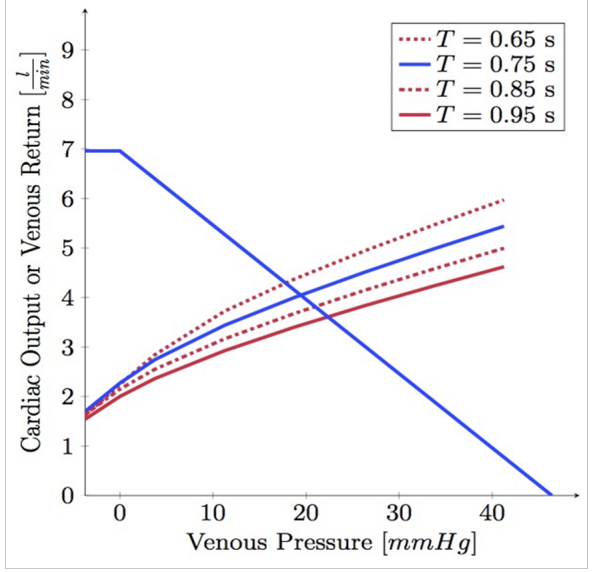
*Rehorn et al. JACC Clin Electrophysiol. 2020 Sep;6(9):1144-1154.*

# 3. Quid de l'incompétence chronotrope ?

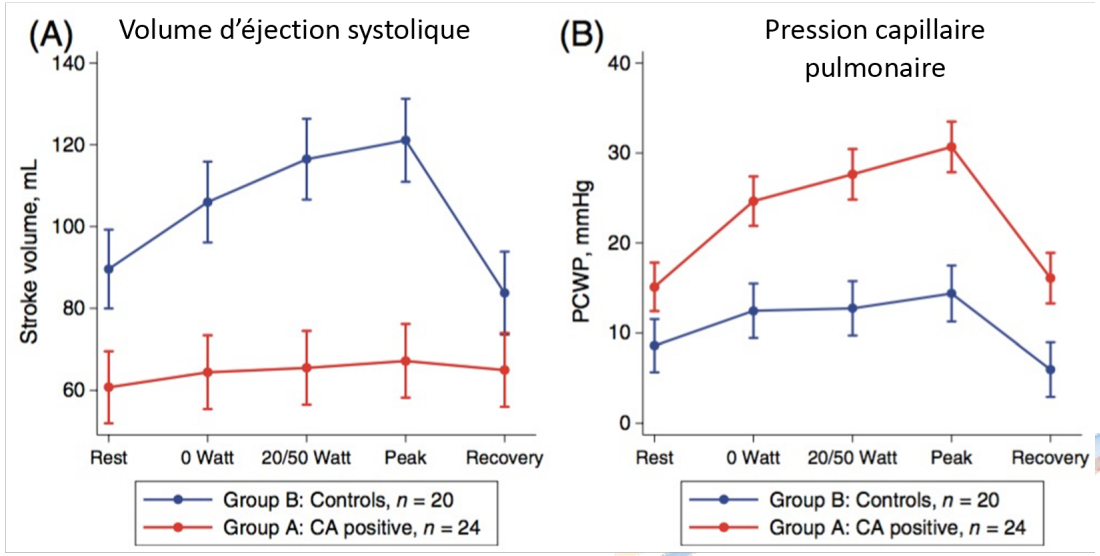
- Y penser si dyspnée d'effort avec NTproBNP bas.
- Très fréquente dans les ATTRv (neuro), fréquente dans les autres amyloses
- Mécanisme :

**Patient-Specific Biomechanical modeling of cardiac amyloidosis**
*D. Chapelle et al, 2015*

**Débit cardiaque = VES (fixé) × FC**




**Inotropic myocardial reserve deficiency is the predominant feature of exercise haemodynamics in cardiac amyloidosis**
*T. Clemmensen et al, Eur Jof Heart failure, 2017*



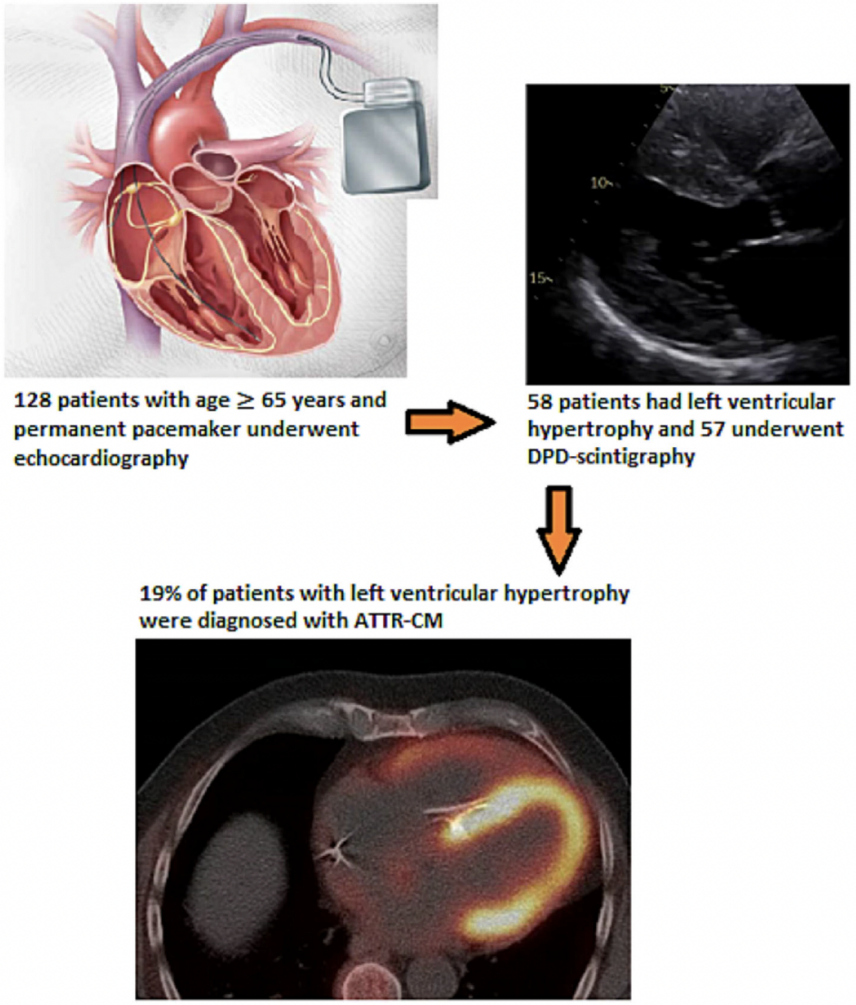
# 3. Quid de l'incompétence chronotrope ?

- Mécanisme :
- Faible élévation du VES à l'effort et incompétence chronotrope expliquée par…
  - Infiltration myocardique amyloïde : responsable d'une cardiomyopathie hypertrophique et restrictive mais aussi d'une atteinte conductive
  - Dysautonomie cardiaque
    - Comment évaluer la dysautonomie ?

# 3. Quelle prévalence de l'ATTR chez les patients porteurs de PM ?

- 128 patients porteurs de PM (indications : BAV haut degré, BSA, MRA) ≥ 65 ans
- ETT > scintigraphie osseuse si SIV ≥ 12 mm.
- 11/128 patients (9%) et 11/58 patients avec HVG fixaient à la scintigraphie osseuse.
- Comparé aux patients sans amylose, les ATTR :
  - Biomarqueurs plus élevés
  - + de dysfonction diastolique
  - SGL plus bas
  - Plus d'IC clinique

**Figure 1** Study design and major findings. We report a 19% prevalence of ATTR-CM in our cohort of pacemaker patients with left ventricular hypertrophy. Echocardiographic image showing left ventricular hypertrophy defined as septum wall thickness ≥12 mm. SPECT-CT image reviles high uptake of DPD tracer in the myocardium of the left ventricle. Both pictures are from a study patient diagnosed with ATTR-CM. ATTR-CM, transthyretin amyloid cardiomyopathy; DPD, $^{99m}$Technesium 3,3-diphosphono-1,2-propanodicarboxylicacid; SPECT, single-photon emission computerized tomography.

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*Aaseth et al. ESC Heart Failure 2024; 11: 871–876*

# 3. Quelle prévalence de l'ATTR chez les patients âgés en BAV de haut degré?

- 39 patients avec indication de PM pour BAV de haut degré et âgés de 70 à 85 ans
- > scintigraphie osseuse systématique
- 3/39 patients (7,7%, 100% hommes) Perugini 2 ou 3.
- Comparé aux patients sans amylose, les ATTR :
  - Avaient des signes extra-cardiaques : canal lombaire (100%), canal carpien (2/3)
  - Etaient plus hypertrophiés (19+/-3,6 vs 11,4+/- 2,7 mm).

**Table 2** Features of patients with Perugini grade 2 or 3 on DPD scan

| | Patient 1 | Patient 2 | Patient 3 |
|---|---|---|---|
| Sex, n (%) | Male | Male | Male |
| Comorbidities | Hypothyroidism | MI, T2DM, prostate cancer | Hypertension |
| Age at device implantation (years) | 84.8 | 78.6 | 78.1 |
| LV internal diameter (mm) | 35 | 55 | 50 |
| Left atrial diameter (mm) | 37 | 48 | 39 |
| LV ejection fraction (%) | 62.5 | 22.5 | 39 |
| Maximum LV wall thickness (mm) | 20 | 15 | 22 |
| Previous arrhythmia | NSVT | Nil | Nil |
| NTproBNP (ng/L) | 983 | 3462 | 626 |
| Perugini grade | 2 | 2 | 3 |
| Degree of heart block | Fixed 2:1 | Third degree | Third degree |
| Light chain amyloidosis screen | Negative | Negative | Negative |
| Genotype | Wild-type | Wild-type | Wild-type |
| Carpal tunnel syndrome | Yes | No | Yes |
| Spinal canal stenosis | Yes | Yes | Yes |
| Biceps tendon rupture | No | No | No |

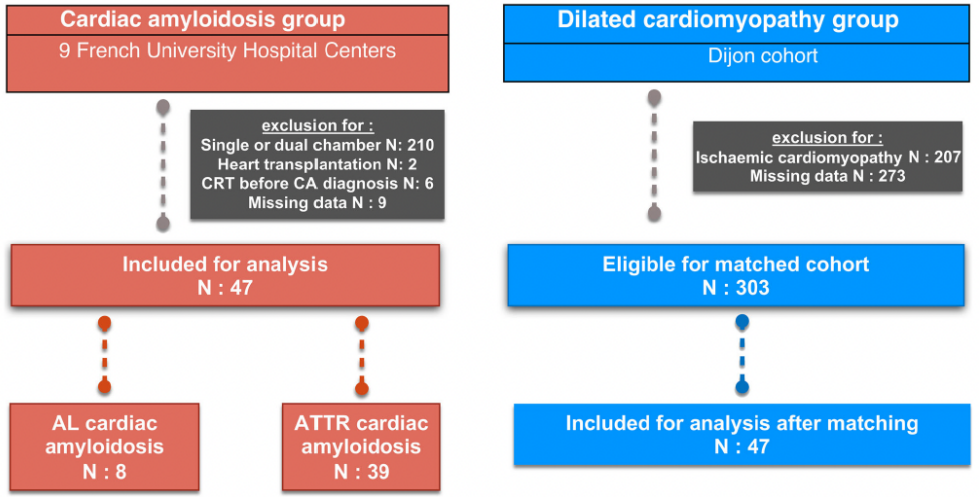
DPD, 3,3-diphosphono-1,2-propanodicarboxylic acid; LV, left ventricular; MI, myocardial infarction; NSVT, non-sustained ventricular tachycardia; NTproBNP, N-terminal pro-B-type natriuretic peptide.; T2DM, type 2 diabetes mellitus.

*Cannie D, Open Heart 2024;11:e002606.*

# 4. Quelles indications de CRT ?

## Cardiovascular outcomes after cardiac resynchronization therapy in cardiac amyloidosis

Kilian Fischer[1], Nicolas Lellouche[2], Thibaud Damy[2], Raphaël Martins[3], Nicolas Clementy[4], Arnaud Bisson[4], François Lesaffre[5], Madeline Espinosa[5], Rodrigue Garcia[6], Bruno Degand[6], Guillaume Serzian[7], François Jourda[8], Olivier Huttin[9], Jean-Baptiste Guichard[10], Hervé Devilliers[11], Jean-Christophe Eicher[1], Gabriel Laurent[1] and Charles Guenancia[1,12]*



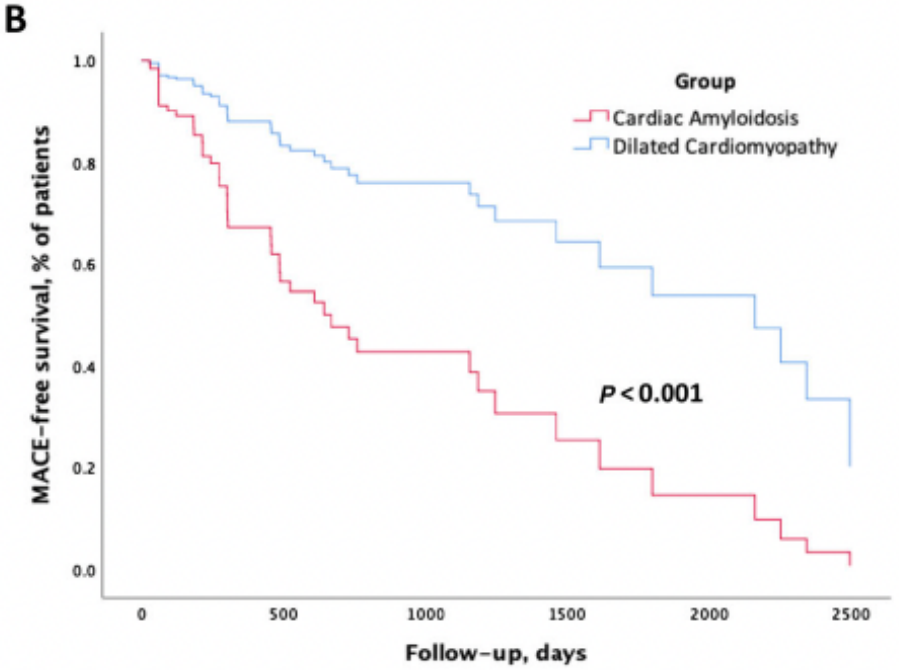

| | Cardiac amyloidosis N = 47 | DCM N = 47 | P |
|---|---|---|---|
| Population | | | |
| Age at implantation (years) | 77.2 ± 5.9 | 76.3 ± 5.3 | 0.64 |
| Age at implantation >75 years | 29 (62) | 28 (60) | 1 |
| Male sex | 43 (92) | 31 (66) | 0.002 |
| NYHA stage | | | 0.73 |
| I | 5 (11) | 5 (11) | |
| II | 26 (55) | 29 (62) | |
| III | 15 (32) | 13 (28) | |
| IV | 1 (2) | 0 | |
| Cardiovascular history | | | |
| Coronary artery disease | 10 (21) | 0 | <0.001 |
| Previous atrial fibrillation | 32 (68) | 21 (45) | 0.04 |
| Previous hospitalization for heart failure | 41 (87) | 22 (47) | 0.004 |
| Chronic medications | | | |
| Beta-blocker | 8 (17) | 46 (98) | <0.001 |
| ACE inhibitor/ARBs | 15 (32) | 41 (87) | <0.001 |
| Diuretic | 42 (89) | 35 (75) | 0.11 |
| MRA | 22 (47) | 19 (40) | 0.68 |
| Valsartan/sacubitril | 2 (4) | 3 (6) | 1 |
| Digoxin | 2 (4) | 5 (11) | 0.44 |
| Calcium channel blocker | 0 | 1 (2) | 1 |
| Amiodarone | 14 (30) | 6 (13) | 0.08 |
| Anticoagulation therapy | 43 (92) | 24 (51) | 0.004 |
| Amyloidosis treatment | | | |
| Tafamidis | 15 (32%) | | |
| Chemotherapy for AL | 7 (15%) | | |
| Echocardiography | | | |
| LVEF at baseline (%) | 30 (25–35) | 30 (25–34) | 0.89 |
| LVEF at follow-up after implantation (%) | 37 (31–43) | 45 (40–50) | <0.001 |
| Time between CRT and TTE (days) | 273 (182–365) | 306 (182–458) | 0.20 |
| Absolute delta LVEF ≥ 10% | 17 (36) | 33 (70) | 0.002 |
| Indication for implantation | | | 0.20 |
| LBBB + LVEF ≤ 35% | 27 (54) | 32 (68) | |
| Non-LBBB enlarged QRS + LVEF ≤ 35% | 6 (13) | 1 (2) | |
| Upgrading | 10 (21) | 8 (17) | |
| 'BLOCK-HF like' indication | 4 (12) | 6 (13) | |
| Biventricular stimulation rate (%) | 98 (95–99) | 99 (97–99) | 0.002 |
| Biventricular stimulation rate >95% | 36 (77) | 43 (92) | 0.09 |

*Fischer et al. ESC Heart Fail. 2022 Feb;9(1):740-750.*

# 4. Quelles indications de CRT ?

## Cardiovascular outcomes after cardiac resynchronization therapy in cardiac amyloidosis

Kilian Fischer[1], Nicolas Lellouche[2], Thibaud Damy[2], Raphaël Martins[3], Nicolas Clementy[4], Arnaud Bisson[4], François Lesaffre[5], Madeline Espinosa[5], Rodrigue Garcia[6], Bruno Degand[6], Guillaume Serzian[7], François Jourda[8], Olivier Huttin[9], Jean-Baptiste Guichard[10], Hervé Devilliers[11], Jean-Christophe Eicher[1], Gabriel Laurent[1] and Charles Guenancia[1,12]*




## Predictors of MACE in CA group

| Variable | Univariate HR | Univariate 95% CI | Univariate P | Multivariate HR | Multivariate 95% CI | Multivariate P |
|---|---|---|---|---|---|---|
| Male sex | 3.99 | 0.54–29.47 | 0.17 | | | |
| Age at implantation | 1.04 | 0.98–1.11 | 0.23 | | | |
| Previous atrial fibrillation | 1.29 | 0.59–2.82 | 0.53 | | | |
| Previous hospitalization for heart failure | 2.51 | 0.75–8.40 | 0.14 | 2.79 | 0.984–9.33 | 0.095 |
| LBBB + LVEF ≤ 35% | 0.78 | 0.36–1.64 | 0.52 | | | |
| Amyloidosis treatment | 0.69 | 0.24–1.40 | 0.30 | | | |
| CRT-P (vs. CRT-D) | 1.40 | 0.68–2.89 | 0.37 | | | |
| Biventricular stimulation rate <95% | 2.02 | 0.87–4.66 | 0.10 | | | |
| Delta LVEF ≥ 10% | 0.43 | 0.18–1.01 | 0.05 | 0.36 | 0.15–0.86 | 0.002 |

CI, confidence interval; CRT-D, cardiac resynchronization therapy—defibrillator; CRT-P, cardiac resynchronization therapy—pacemaker; HR, hazard ratio; LBBB, left bundle block branch; LVEF, left ventricular ejection fraction; MACE, major cardiovascular event.
Multivariate analysis adjusted on the delay between cardiac resynchronization therapy and follow-up echocardiography.

## Predictors of CRT response in CA group

| Variable | Univariate HR | Univariate 95% CI | Univariate P | Multivariate HR | Multivariate 95% CI | Multivariate P |
|---|---|---|---|---|---|---|
| Male sex | 0.54 | 0.07–4.19 | 0.55 | | | |
| Age at implantation >75 years[a] | 0.33 | 0.10–1.08 | 0.068 | 0.45 | 0.21–0.99 | 0.047 |
| Previous atrial fibrillation | 1.20 | 0.33–4.36 | 0.78 | | | |
| LVEF at implantation ≤35% | 0.68 | 0.34–1.39 | 0.29 | | | |
| Creatinine at implantation | 0.997 | 0.991–1.003 | 0.36 | | | |
| Previous hospitalization for heart failure | 0.93 | 0.19–4.50 | 0.93 | | | |
| LBBB | 1.60 | 0.47–5.47 | 0.45 | | | |
| Amyloidosis treatment | 1.02 | 0.31–3.35 | 0.98 | | | |
| AL | 1.07 | 0.22–5.17 | 0.93 | | | |
| Biventricular stimulation rate <95%[a] | 0.64 | 0.33–1.24 | 0.19 | | | |

AL, amyloidosis with immunoglobulin light chains; CA, cardiac amyloidosis; CI, confidence interval; CRT, cardiac resynchronization therapy; HR, hazard ratio; LBBB, left bundle block branch; LVEF, left ventricular ejection fraction.
[a]Included in the bivariate analysis.

*Fischer et al. ESC Heart Fail. 2022 Feb;9(1):740-750.*

# 4. Stimulation branche gauche ?

22 patients

Pas de changement sur FEVG ou NTproBNP à 6 mois (mais délai court)

**TABLE 1** Patient baseline characteristics, pacing indication, treatment.

| Variable | |
|---|---|
| Age (years) | 78.6 ± 11.7 |
| Male sex (%) | 19 (82.6) |
| HTN (%) | 16 (69.6) |
| DM Type 2(%) | 7 (30.4) |
| Chronic kidney disease (%) | 5 (21.7) |
| ATTR Type (%) | 20 (87.0) |
| AL Type (%) | 3(13) |
| Paroxysmal atrial fibrillation (%) | 3 (13.0) |
| Persistent atrial fibrillation (%) | 3 (13.0) |
| Permanent atrial fibrillation (%) | 11 (47.8) |
| LBBB (%) | 9 (39.1) |
| RBBB (%) | 8 (34.8) |
| **Pacing indication** | |
| Sick Sinus Disease (%) | 5 (21.7) |
| AV conduction disorder (%) | 4 (17.3) |
| CRT (%) | 8 (34.7) |
| Atrial fibrillation with slow ventricular response (%) | 6 (26.1) |
| **Treatment** | |
| Beta blocker (%) | 6 (26.1) |
| Aldosterone antagonist (%) | 2 (8.7) |
| ACE/ARB (%) | 6 (26.1) |
| Diuretic (%) | 21 (91.3) |
| Digoxin (%) | 1 (4.3) |

**TABLE 2** Echocardiographic, biochemical and electrical parameters evolution.

| Variable | Baseline | Follow up |
|---|---|---|
| Echocardiographic and biochemical parameters | | |
| LVEF (%) | 45.5 ± 16.2 | 41.9 ± 13.6 ($p = .962$) |
| LVESV (mL) | 47.6 ± 23.8 | 50.1 ± 15.5 ($p = .251$) |
| LVEDV (mL) | 85.4 ± 32.6 | 88.3 ± 30.9 ($p = .467$) |
| LA volume (mL/m$^2$) | 46.8 ± 12.8 | 57.3 ± 17.8 ($p = .098$) |
| IVS thickness (mm) | 18.4 ± 4.4 | |
| Nt ProBNP (pg/dL) | 5093 ± 4306 | 4808 ± 4251($p = .990$) |
| Electrical parameters | | |
| R wave (mV) | 9.6 ± 6.3 | 10.6 ± 5.1 ($p = .985$) |
| Lead impedance (Ohm) | 612 ± 213 | 370 ± 94 ($p = .021$) |
| Threshold (V) | 1.1 ± 0.6 | 0.8 ± 0.1 ($p = .210$) |

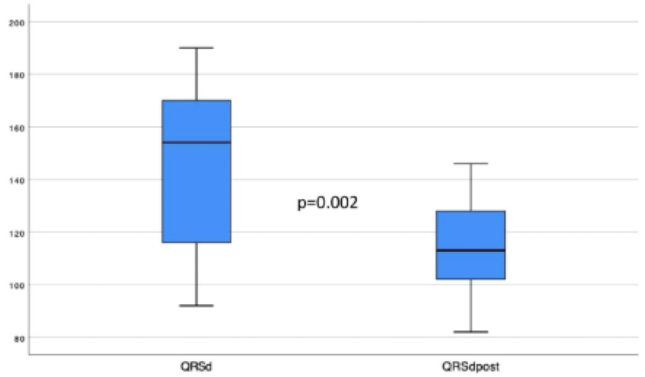
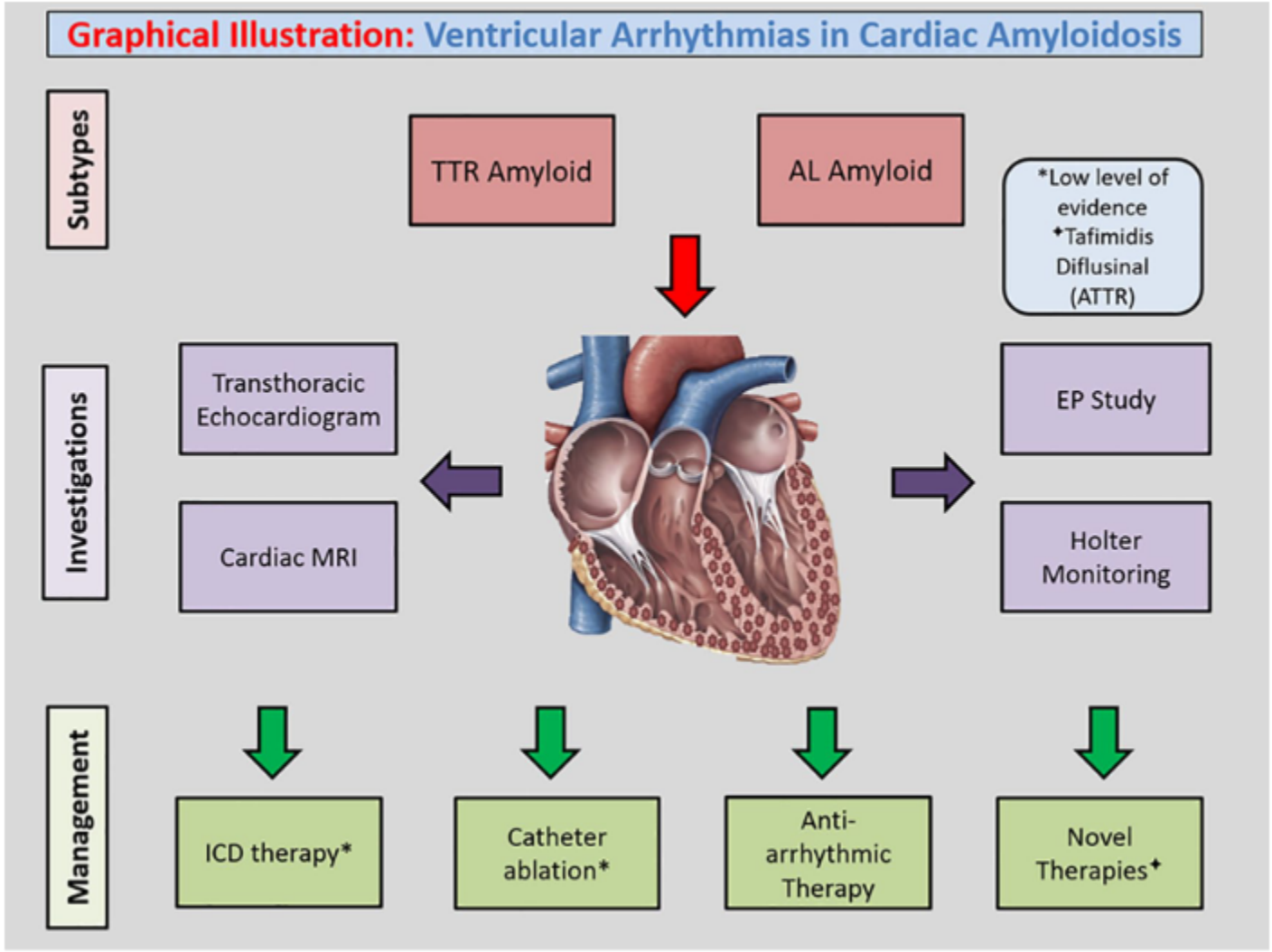



**FIGURE 3** QRSd: intrinsic QRS duration (msec); QRSdpost: Paced QRS duration (ms). [Color figure can be viewed at wileyonlinelibrary.com]

*Pham Trung et al. Pacing Clin Electrophysiol. 2024 Jan;47(1):149-155.*






*Khanna S et al. Clin Med Insights Cardiol. 2020 Sep 29:14:1179546820963055*

| STUDY SUMMARY | NUMBER OF PATIENTS | TYPE OF CARDIAC AMYLOID | METHOD OF MONITORING | NON-SUSTAINED VT IN % OF PATIENTS | OTHER RELATED FINDINGS |
|---|---|---|---|---|---|
| Palladini et al[7] | 51 | AL | 24-hour Holter monitoring | Non-sustained VT in 18% of patients. | Ventricular tachycardia was a significant prognostic determinant for survival ($P=0.04$). |
| Sayed et al[8] | 20 | AL with symptoms of pre-syncope or syncope | Implantable loop recorder | Non-sustained VT in 5% of patients | Terminal syncopal event was marked by bradycardia, not tachycardia, in every available recording. |
| Murtagh et al[9] | 127 | AL | Electrocardiogram | Non-sustained VT in 1% of patients | Premature ventricular contractions were noted in 13% of patients. |
| Goldsmith et al[10] | 24 | AL monitored peri autologous stem cell transplantation | Telemetry, average of 24 days | Non-sustained VT in 100% of patients | In the deceased patients (n=3), VT/VF events were the highest of all 24 patients. There was a correlation between VT and serum BNP levels before SCT ($r=0.47$, $P=0.019$) and during admission for SCT ($r=0.62$, $P=0.0012$), serum creatinine before SCT ($r=0.62$, $P=0.001$), and inverse relationship with cardiac output ($r=-0.72$, $P<0.001$) |
| Dubrey et al[12] | 232 | AL | 24-hour Holter monitoring | Non-sustained VT in 26.7% of patients | Ventricular tachycardias were not associated with increased risk of sudden cardiac death. |
| Hörnsten et al[11] | 30 | ATTR (ATTR Val30Met trait), before liver transplant | 24-hour Holter monitoring | Non-sustained VT in 16.7% of patients | – |
| Varr et al[45] | 31 | AL 77%, ATTR 23% (V122I mutation 10%, wild type 13%) | ICD, permanent pacemaker, telemetry | VT in 74% of patients | ICD therapy was successful in most patients, with therapy in 4 of 5 (80%) patients resulting in the termination of the arrhythmia. |
| Kristen et al[13] | 19 | AL with ICD insitu | ICD | VT/VF in 11% of patients | Ventricular extra beats (grade IVa or higher) were present more often in non-survivors than in survivors ($P<0.05$). |
| Hamon et al[49] | 45 | Familial ATTR 60%, AL 27%, senile ATTR 13%. All with ICD in situ | ICD | VT/VF in 27% of patients | Inappropriate shocks was uncommon and occurred in 2 patient (4.4%). |

*Khanna S et al. Clin Med Insights Cardiol. 2020 Sep 29:14:1179546820963055*

# Mode de décès au cours de l'amylose cardiaque

## Describing mode of death in three major cardiac amyloidosis subtypes to improve management and survival

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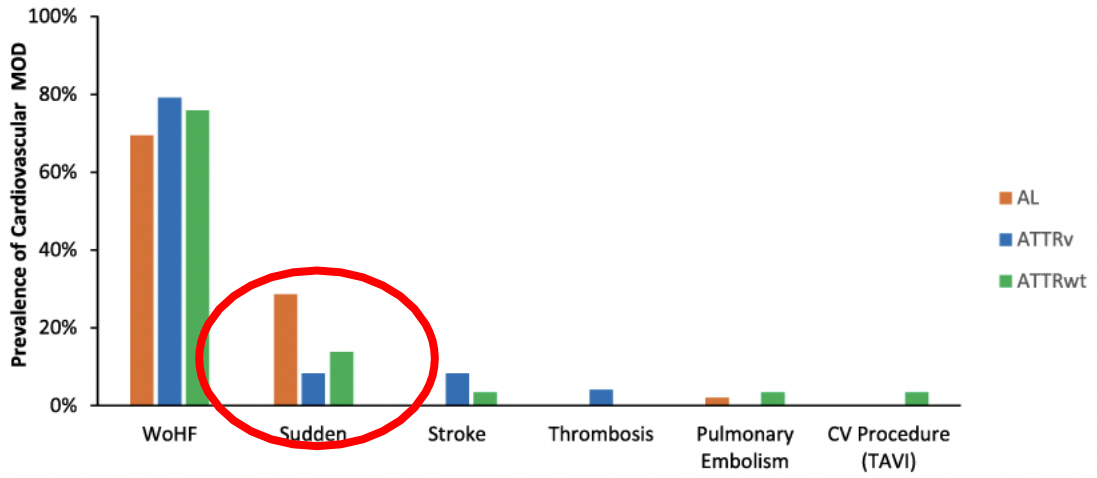
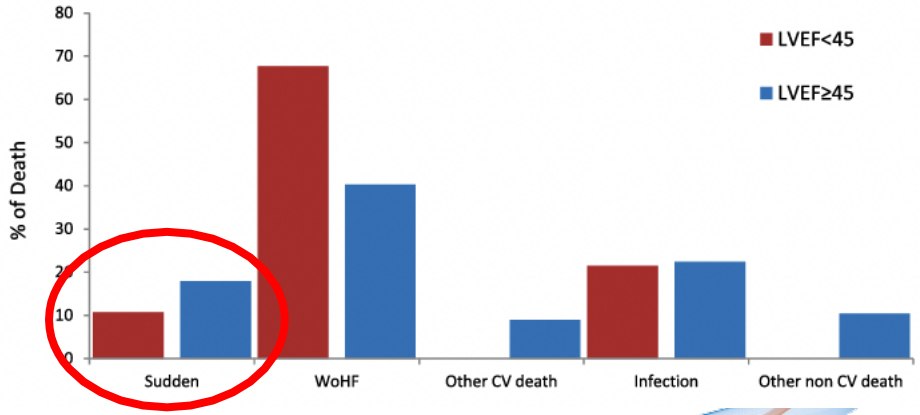
- **Mort subite = 2e cause de décès CV dans l'AC**
- **Risque mal stratifié par la FEVG**

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




**B: Mode of Death of Cardiac Amyloidosis Population depending on LVEF (<45% or ≥ 45%)**




AP-HP. Hôpitaux universitaires Henri-Mondor

*Kharoubi et al. Amyloid. 2022 Jun;29(2):79-91.*

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CREATIVE CONCEPTS

# Implantable cardioverter-defibrillator placement in patients with cardiac amyloidosis

Brandon C. Varr, MD,[*] Shirin Zarafshar, MD,[†] Terra Coakley, MAT,[†] Michaela Liedtke, MD,[‡] Richard A. Lafayette, MD,[§] Sally Arai, MD,[‖] Stanley L. Schrier, MD,[‡] Ronald M. Witteles, MD[†]

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- 31 patients avec PM ou DAI amylose TTR ou AL
- TVNS 74 % et TVS ou FV 19 % des patients.
- 19 % des patients avec DAI ont reçu une thérapie appropriée à 18 mois

**Table 1** Patient characteristics (n = 31)

| | |
|---|---|
| Age (y) | 68 (51–87) |
| Sex: male | 84% |
| AL | 77% |
| ATTR | |
| Wild type | 13% |
| Familial V122I mutation | 10% |
| Pacemaker | 32% |
| ICD | 61% |
| NYHA class 1 | 6% |
| NYHA class 2 | 52% |
| NYHA class 3 | 42% |
| NYHA class 4 | 0% |
| LVEF <55% | 39% |
| NT-proBNP (pg/mL) | 6635 (193–30,000) |
| Device indication | |
| Bradycardia/heart block | 34% |
| Primary prevention SCD | 52% |
| Secondary prevention SCD | 14% |

AL = light-chain amyloidosis; ATTR = transthyretin amyloidosis; ICD = implantable cardioverter-defibrillator; NT-proBNP = N-terminal pro–brain natriuretic peptide; NYHA = New York Heart Association; SCD = sudden cardiac death; V122I = valine to isoleucine substitution at position 122.

**Table 3** Results of ICD therapy

| Age (y), sex, amyloid type | Reason for implantation | NYHA class | Rhythm | ICD therapy | Defib energy (J) | Success therapy | Survival |
|---|---|---|---|---|---|---|---|
| 70, M, AL | Primary prevention | 2 | VT and VF | ATP/Defib | 35 | Yes | 6 mo, Alive |
| 60, M, AL | Primary prevention | 2 | VF | Defib | 35 | No | Decd |
| 59, M, AL | Secondary prevention | 2 | VT | Defib | 41 | Yes | 6 wk, Decd |
| 67, M, AL | Secondary prevention | 2 | VT | ATP | n/a | Yes | 18 mo, Decd |
| 62, F, AL | Secondary prevention | 3 | VT | Defib | 35 | Yes | 19 mo, Decd |
| 60, M, ATTR (V122I mutation) | Primary prevention | 2 | VT | Shock aborted owing to spontaneous termination | | | 23 mo, Alive |

AL = light-chain amyloidosis; ATP = antitachycardia pacing; ATTR = transthyretin amyloidosis; Decd = deceased; Defib = defibrillation; ICD = implantable cardioverter-defibrillator; NYHA = New York Heart Association; VF = ventricular fibrillation; VT = ventricular tachycardia; V122I = valine to isoleucine substitution at position 122.

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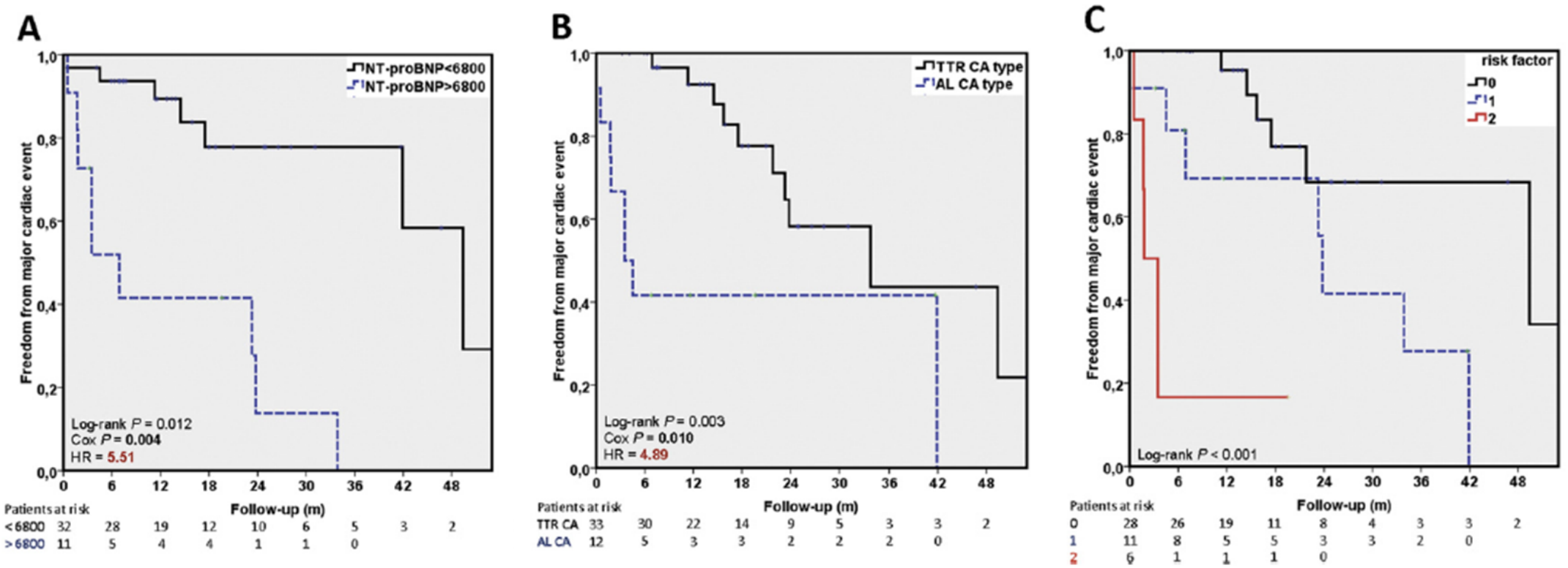
*Varr et al. Heart Rhythm. 2014 Jan;11(1):158-62.*




Facteurs de mauvais pronostic: taux de Nt pro-BNP élevé, amylose AL (mais les nouveaux traitements améliorent le pronostic), évolutivité de la pathologie

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*Hamon, D. et al. Int J Cardiol. 2016;222:562-568.*

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**Table 3. Ventricular tachyarrhythmia and implantable cardioverter-defibrillator (ICD) studies in cardiac amyloidosis**

| Study | Population | Sample size | Prevalence of bradyarrhythmia |
|---|---|---|---|
| Varr et al. (2014)[33] | AL or ATTR amyloidosis | 31 patients with CA and implanted devices or ambulatory monitoring | Nonsustained VT in 74%, sustained VT or VF in 19% |
| Kristen et al. (2008)[16] | Cardiac amyloidosis (unspecified) | 19 patients with CA and previous syncope or PVCs with an ICD | Two patients (11%) developed sustained VT successfully treated by ICD |
| Lin et al. (2013)[36] | AL, wtATTR, hATTR, and AA amyloidoses | 53 patients with CA with ICD for primary (77%) and secondaryn (23%) preventio | Appropriate ICD shocks in 32% during first year, mostly in patients with AL amyloidosis and secondary prevention devices |
| Hamon et al. (2016)[37] | AL, wtATTR, and hATTR amyloidoses | 55 patients with CA with ICD for primary (84%) and secondary prevention (16%). | Appropriate ICD therapy in 27% after an average of 4.7 ± 6.6 months; no independent predictors of ICD therapy; 27% patients died in follow-up |

AA, amyloid A protein; AL, amyloidogenic light chain; ATTR, amyloidogenic transthyretin; CA, cardiac amyloidosis; hATTR amyloidosis, hereditary amyloidogenic transthyretin; PVC, premature ventricular complexes; VF, ventricular fibrillation; VT, ventricular tachycardia; wtATTR, wild-type amyloidogenic transthyretin.

# Indications du DAI dans l'amylose reco US

| Implantable cardioverter-defibrillator | | | |
|---|---|---|---|
| I | C-EO | In individuals with CA who have survived a cardiac arrest, an ICD is recommended if meaningful survival > 1 year is expected. | 34 |
| IIb | B-NR | In individuals with AL-type cardiac amyloidosis with nonsustained ventricular arrhythmias, a prophylactic ICD may be considered if meaningful survival > 1 year is expected. | 29 |

AL, amyloidogenic light chain; AV, atrioventricular; CA, cardiac amyloidosis; ICD, implantable cardioverter-defibrillator.

*Class of recommendation: I: strong, benefit > > > risk; IIa: moderate, benefit > > risk; IIb: weak, benefit > risk.

†Level of evidence: B-NR: level B, nonrandomized: moderate-quality evidence from ≥ 1 well designed, well executed nonrandomized studies, observational studies, or registry studies, or meta-analysis of such studies. C-EO: level C, expert opinion: consensus of expert opinion based on clinical experience.

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*John RM, Stern DL. Can J Cardiol. 2020;36(3):408-415.*

ESC European Society of Cardiology
Europace (2020) 22, 1216–1223
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# Outcomes in patients with cardiac amyloidosis and implantable cardioverter-defibrillator

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**Table I** **Baseline characteristics in all groups**

| | | Group 1 (CA + ICD) | Group 2 (CA without ICD) | Group 3 (non-CA with ICD) |
|---|---|---|---|---|
| *N* | | 23 | 68 | |
| Type of amyloidosis | AL | 7 (30.0%) | 42 (62.0%) | 0.04 |
| | ATTRm | 6 (26.0%) | 13 (19.0%) | |
| | ATTRwt | 10 (43.0%) | 12 (18.0%) | |
| | Unknown | 0 (0%) | 1 (1.0%) | |
| Kappa | | 1 (4.0%) | 12 (18.0%) | 0.12 |
| Lambda | | 6 (26.0%) | 30 (44.0%) | 0.13 |
| Elevated FLC-diff (>18 mg/dL) | | 1 (14.0%) | 27 (64.0%) | 0.01 |
| Renal amyloidosis | | 7 (30.0%) | 23 (34.0%) | 0.77 |
| GI system amyloidosis | | 4 (17.0%) | 9 (13.0%) | 0.62 |
| Pulmonary amyloidosis | | 0 (0.0%) | 6 (9.0%) | 0.14 |
| Autonomic dysfunction | | 3 (13.0%) | 7 (10.0%) | 0.72 |
| Peripheral neuropathy | | 7 (30.0%) | 10 (15.0%) | 0.09 |
| Hepatic amyloidosis | | 3 (13.0%) | 2 (3.0%) | 0.07 |
| Dermatological involvement | | 1 (4.0%) | 3 (4.0%) | 0.99 |
| Others | | 2 (8.0%) | 10 (9.9%) | 0.04 |
| Mayo Staging (in AL) | 1 | 0 | 0 | 0.05 |
| | 2 | 0 | 3 (7.0%) | |
| | 3 | 6 (86.0%) | 15 (36.0%) | |
| | 4 | 1 (14.0%) | 24 (57.0%) | |

AL, light chain amyloidosis; ATTRm, transthyretin mutant amyloidosis; ATTRw, transthyretin wild-type amyloidosis; FLC, free light chains.



*Kim et al. Europace (2020) 22, 1216-1223*

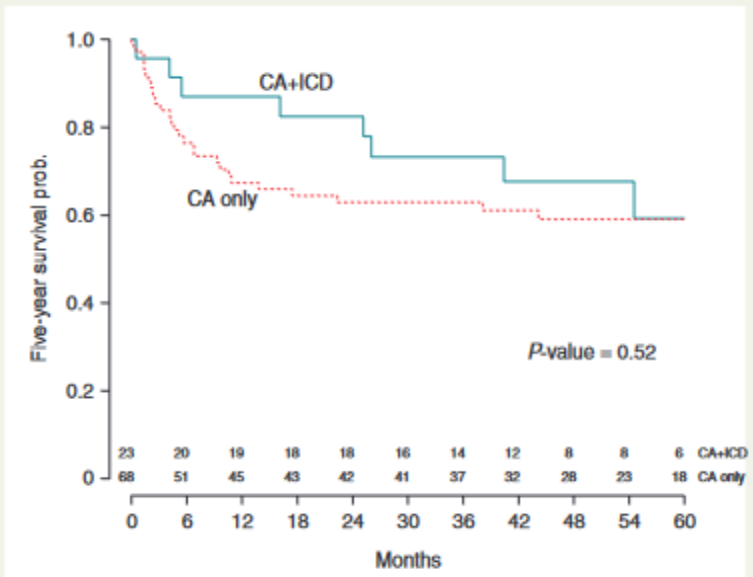



**Figure 2** The Kaplan–Meier plot of survival probabilities from CA diagnosis to death in patients with CA. The figure compared the survival of Group 1 (CA and ICD) with Group 2 (CA and no ICD) utilizing the Kaplan–Meier curve. There is no significant survival difference to 5 years after the diagnosis ($P = 0.52$). CA, cardiac amyloidosis; ICD, implantable cardioverter-defibrillator.

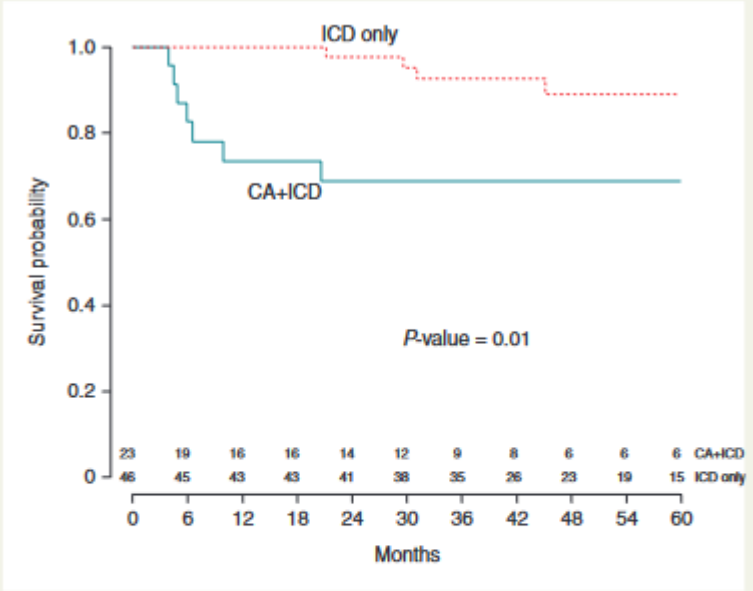



**Figure 3** The Kaplan–Meier plot of survival probabilities from ICD to death in patients with ICD. This figure compared the survival of Group 1 (CA and ICD) with Group 3 (non-amyloid and ICD) utilizing the Kaplan–Meier curve. ($P = 0.01$). Survival was significantly better in the non-amyloid population with primary prevention ICDs [HR=6.98 (Group 1 vs. Group 3), 95%CI = 1.53–31.7, $P = 0.01$]. CA, cardiac amyloidosis; ICD, implantable cardioverter-defibrillator.



*Kim et al. Europace (2020) 22, 1216-1223*

-La FEVG semble peu prédictive du risque de TDR ventriculaire (implantation trop tardive?)

-l'EEP ne semble pas prédictive des TDR ventriculaires

-Espérance de vie > 1an

-Peu d'intérêt d'implanter les patients trop tôt ou trop tard

Table 2. Univariate Analysis of Characteristics of All 23 Patients Who Died During Follow-Up

| | Sudden Death Group (n = 10) | Nonsudden Death Group[a] (n = 13) | p Value |
|---|---|---|---|
| Age at time of EP study (yr) | 52.6 ± 10.1 | 60.1 ± 7.3 | 0.066 |
| Male | 6 (60%) | 10 (77%) | 0.38 |
| HV interval (ms) | 86 ± 14 | 71 ± 18 | 0.038 |
| HV interval ≥80 ms | 7 (70%) | 3 (23%) | 0.024 |
| LAFB or LPFB | 3 (30%) | 4 (31%) | 0.97 |
| QRS duration ≥120 ms | 2 (20%) | 3 (23%) | 0.86 |
| Positive SAECG[b] | 4/6 (67%) | 3/10 (30%) | 0.15 |
| Inducibility of monomorphic VT | 2 (20%) | 2 (15%) | 0.77 |
| History of syncope or presyncope | 6 (60%) | 2 (15%) | 0.026 |
| Septal thickness (cm) | 1.6 ± 0.3 | 1.6 ± 0.3 | 0.94 |
| LVEF (%) | 49 ± 13 | 48 ± 12 | 0.82 |
| Heart failure | 9 (90%) | 12 (92%) | 0.85 |

Reisinger et al. JACC 1997

# Toxicité des chimios dans l'AL ?

- Analyse rétrospective de 66 AL
- Age moyen 67 (+/-10) ans, 44 (67%) hommes.
- 6 (+/-3) mois après le diagnostic, 8 patients (12%) ont présenté une mort subite, :
  - 4 (6%) réanimés
  - 4 sont décédés
- Pas de différences statistiques sur les principales caractéristiques cliniques (SIV, FEVG).

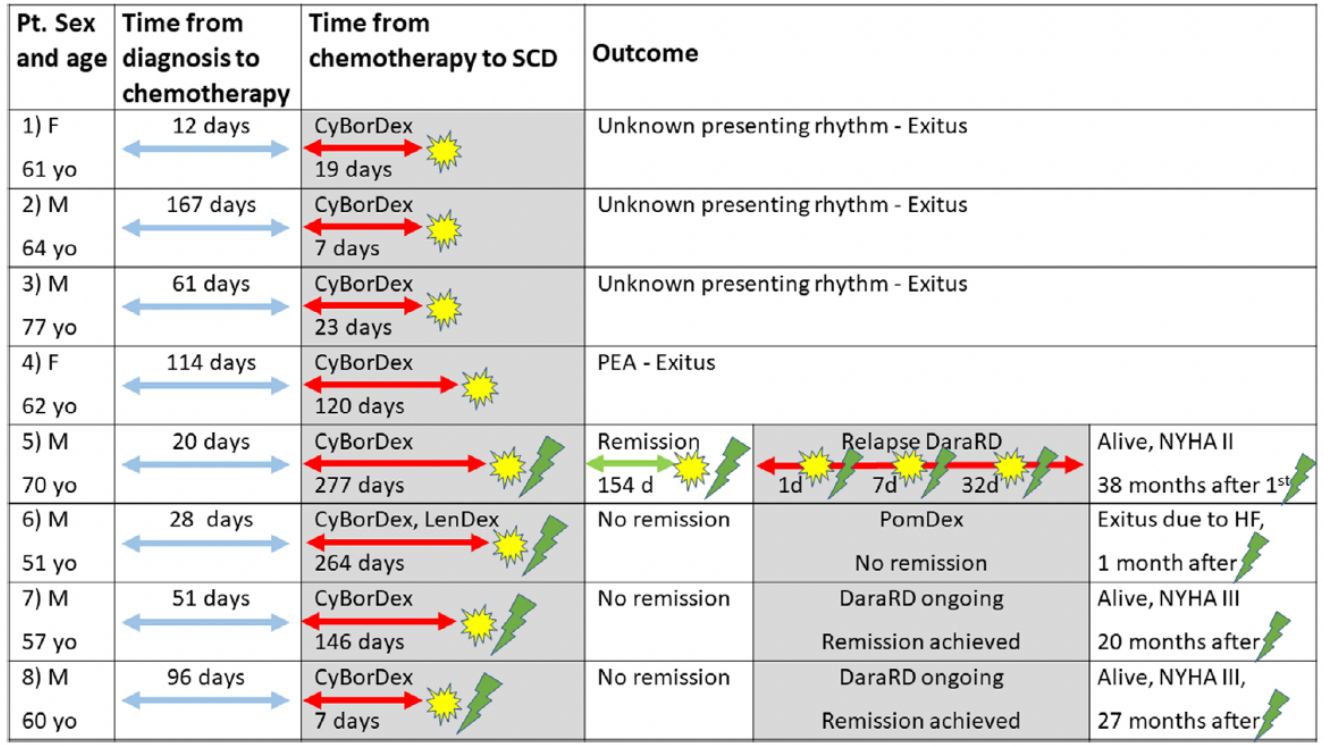



Fig 1. Time interval from chemotherapy start to sudden cardiac death/cardiac arrest (SCD) and patients outcomes. In our cohort eight patients experienced SCD. In three of the eight patients with SCD, the presenting rhythm was unknown (Patients 1, 2, 3), pulseless electrical activity (PEA) was the presenting rhythm in Patient 4 and documented ventricular tachyarrhythmia was the presenting rhythm in Patients 5, 6, 7, 8. All patients with SCD underwent cyclophosphamide/bortezomib/dexamethasone (CyBorDex) as first-line chemotherapy. All patients experienced SCD during chemotherapy. In only one case an appropriate shock occurred during the remission period (Patient 5). All patients with ventricular tachyarrhythmias had resuscitated cardiac arrest; three of them received an implantable cardioverter defibrillator (ICD), whereas one did not because of advanced heart failure (Patient 6). Patient 5 had multiple appropriate ICD shocks during disease relapse while receiving daratumumab/lenalidomide/dexamethasone (DaraRD). All patients with an ICD were alive at the census date and classified as functional New York Heart Association (NYHA) Class II or III after >20 months from resuscitated cardiac arrest (Patients 5, 7, 8). F, female; LenDex, lenalidomide/dexamethasone; M, male; PomDex, pomalidomide/dexamethasone; UNK, unknown; yo, years old; , sudden cardiac death/cardiac arrest; , external or implantable cardiac defibrillator shock following ventricular tachyarrhythmias.

*Zampieri et al. British Journal of Haematology, 2021, 193, e27–e38*

# COHORTE MONDORIENNE DAI ET AMYLOSE AL

- **Inclusion** : DAI + Amylose AL (Nov 2014 – Mars 2024)
- **Exclusion** : 1 patient perdu de vue < 6 mois
- **145 patients**
- Age moyen 63,4 ± 9,2 ans
- 64% Homme
- Prévention primaire 100%
- 18 (81,4%) DR
  - dont 2 échecs CRT
- 26 (17,9%) CRT
- 1 (0,7%) VR

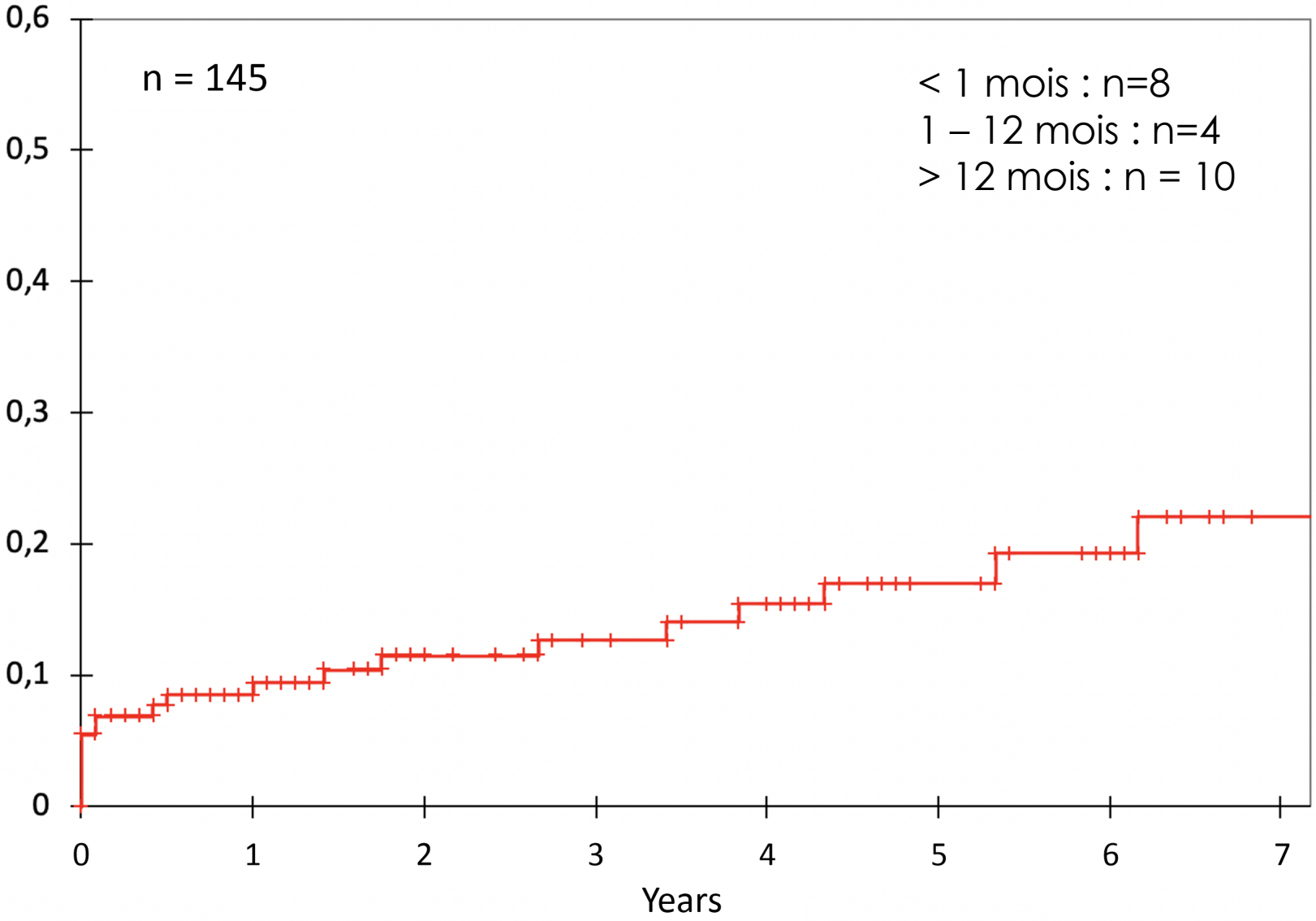
# Critère primaire

- **Incidence cumulée 1ère thérapie appropriée**
  - **Risque compétitif :** décès
  - **Censure :** perdu de vue (n = 9), transplantation (n = 10), explantation définitive (n = 3)

- **Suivi médian** : 14,9 mois (IQR 4,2 - 51,9)

- **<u>22 patients (15,2%)</u>** ont présenté au moins 1 thérapie appropriée

## Cumulative incidence of first appropriate ICD therapy



# Thérapies

- 37 thérapies chez 22 patients
- 5 patients (3,5%) avec > 1 thérapie
- 10 FV/TV > 220 bpm (8 patients)
- 20 TV < 220 bpm
- 7 TV de cycle inconnu
- 3 « orages rythmiques »
- 13 CEI (35%)
  - 10 efficaces
  - 2 FV non réduites
  - 1 DEM

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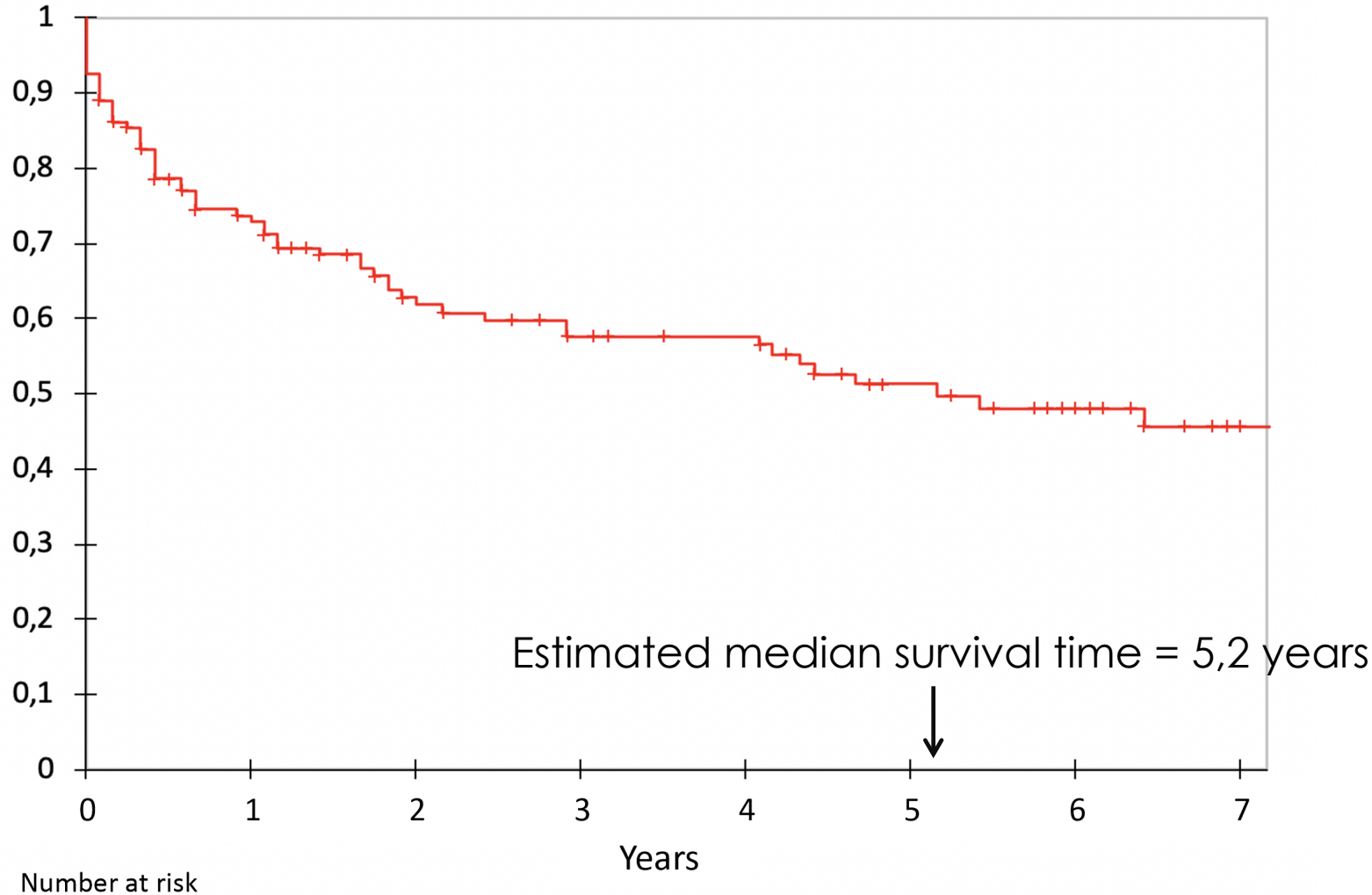
## Kaplan-Meier curve of survival in AL-CA patients with ICD




Number at risk

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| 145 | 87 | 64 | 53 | 50 | 33 | 25 | 13 |

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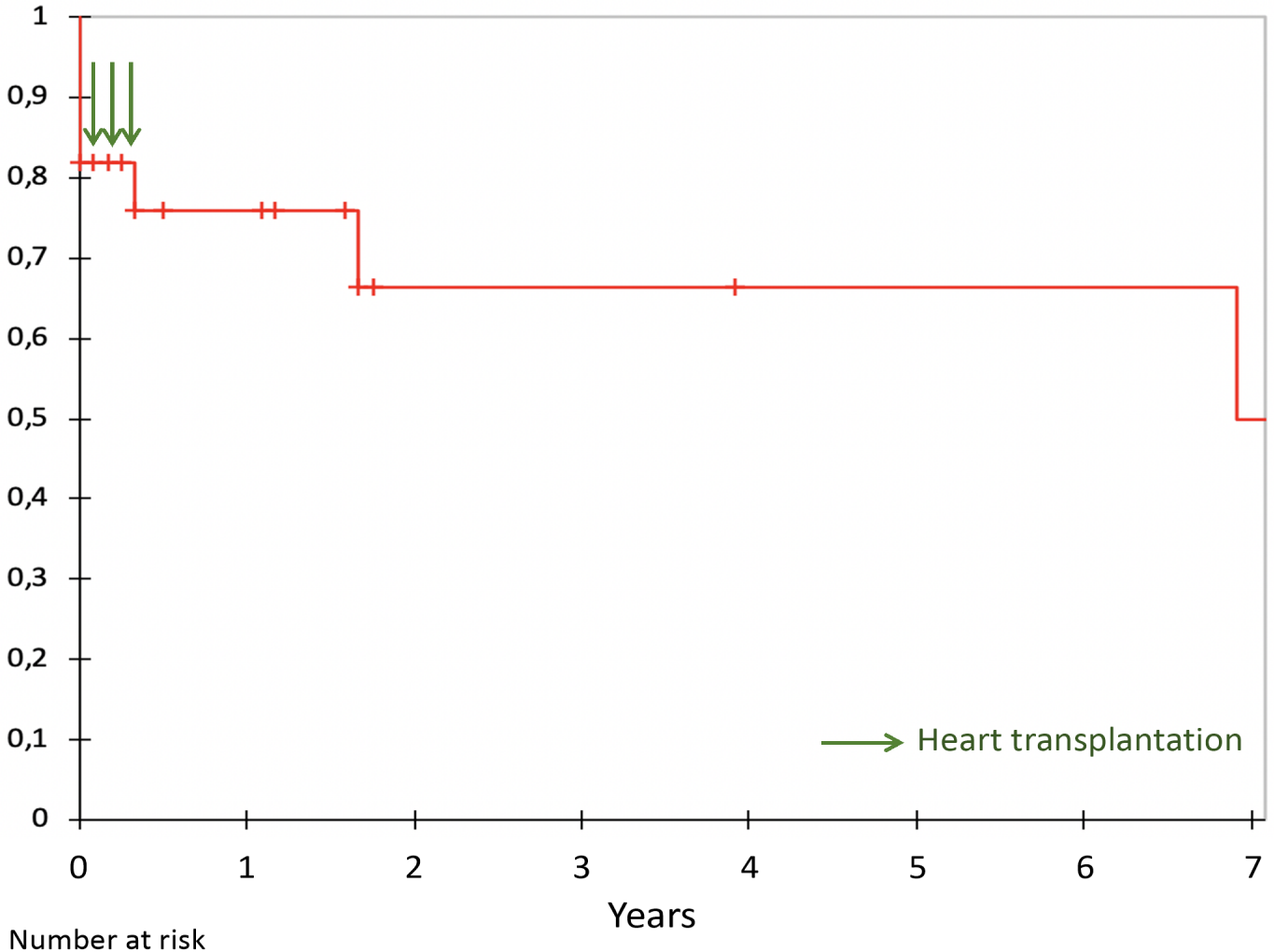
## Kaplan-Meier curve of survival after first appropriate ICD therapy




Number at risk

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| 22 | 11 | 5 | 5 | 4 | 4 | 4 | 3 |

# Limites

- Rétrospectif, recueil sur compte-rendu avant télésuivi
  - Risque de sous-estimation des thérapies
- Certaines morts subites extra-hospitalières inexpliquées
- Pas de groupe contrôle
- Thérapie = pas nécessairement mort subite avortée
  - Impact sur mortalité?
- Evolution des chimiothérapies et programmation DAI sur 10 ans?
- Monocentrique

# Nos indications de PM / DAI dans l'amylose AL

| Gravité | Prévention primaire (dysfonction VG SGL < -14%, TVNS) | Si indication de pacing formelle (dysfonction sinusale, BSA ou BAV de haut degré) | Indication non formelle de pacing mais spécifique à l'amylose AL : PR > 210 ms, BAV 2 M1 | Indication non formelle de pacing et discutée dans l'amylose AL : bloc de branche (BBG ou trouble de conduction intraventriculaire aspécifique si > 120 ms) |
|---|---|---|---|---|
| **Stade I** | Rien | PM | Si évolutivité | Si évolutivité |
| **Stade II** | Rien | PM | Si évolutivité | Si évolutivité |
| **Stade IIIa** | DAI (sauf exception sur l'âge > 75 ans ou les comorbidités) | DAI | DAI | DAI |
| **Stade IIIb** | Life Vest (ou rien) | PM + Life Vest (sauf exception jeune et bon état général : DAI) | PM + Life Vest (sauf exception jeune et bon état général : DAI) | PM + Life Vest (sauf exception jeune et bon état général : DAI) |

| Type d'amylose | double | Triple ou stim branche gauche |
|---|---|---|
| **AL** | Si PR < 250 ms et QRS fins<br><br>Et SGL > -14% | Si PR > 250 ms et / ou QRS > 120 ms<br><br>Si SGL < -14% |

# Take Home Messages

- **Indication PM et DAI** : surveiller l'évolution des troubles conductifs++
- La cause de décès est souvent un trouble conductif d'autant plus que la maladie est évoluée
- Mauvaise réponse à la CRT avec indications classiques : intérêt d'une implantation **précoce** ? (PHRC)
- **FA fréquente** (60 % des cas) : taux élevé de **thrombus OG** (15-30 %) : **ETO systématique si CEE**.
- Intérêt des ATC ± fermeture auricule gauche: systématique en FA, discuté en RS au cas par cas
- Les arythmies ventriculaires sont assez fréquentes mais ne semblent être responsables de la majorité des DC

# Take Home Messages

## PM

- Troubles conductifs de haut degré symptomatique ou non (BAV 3, BAV 2 mobitz II, BSA complet)
- Troubles conductifs de bas degré symptomatique ou non (BAV 1, BAV 2 mobitz I, BBDtc, BBGc, association hémi-bloc et BAV 1), **évolutifs au cours du suivi**
- Incompétence chronotrope symptomatique à l'effort (asthénie/dyspnée d'effort) ou au repos (bradycardie sinusale avec FC < 50/min en l'absence de ralentisseurs de la FC)
- FA avec cadence ventriculaire lente
- FA avec cadence ventriculaire rapide non ralenti par un traitement médical maximal et indication d'ablation du NAV
- Projet de transplantation hépatique (ATTRv)

## DAI

- Prévention primaire (FEVG < 35 %) + espérance de vie > 1 an
- Prévention primaire (FEVG > 35 % et critères de gravité à l'appréciation du cardiologue prescripteur : troponine élevée, nt-pro BNP élevé, altération sévère du SGL, histoire de syncope inexpliquée, épisodes de TVNS…) + espérance de vie > 1 an
- Amylose AL traitée par chimiothérapie et considéré comme à risque de mort subite par le prescripteur (Europe Staging IIIA) surtout si troubles conductifs

## LIFE vest

- Amylose AL traitée par chimiothérapie (Eur Staging IIIb) ou AL avec atteinte cutanée hors remboursement

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Expert consensus

# Management of conduction disease and arrhythmias in patients with cardiac amyloidosis: A position paper from the Working Group of Cardiac Pacing and Electrophysiology of the French Society of Cardiology

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# MERCI DE VOTRE ATTENTION!!

AP-HP.
Hôpitaux universitaires
Henri-Mondor