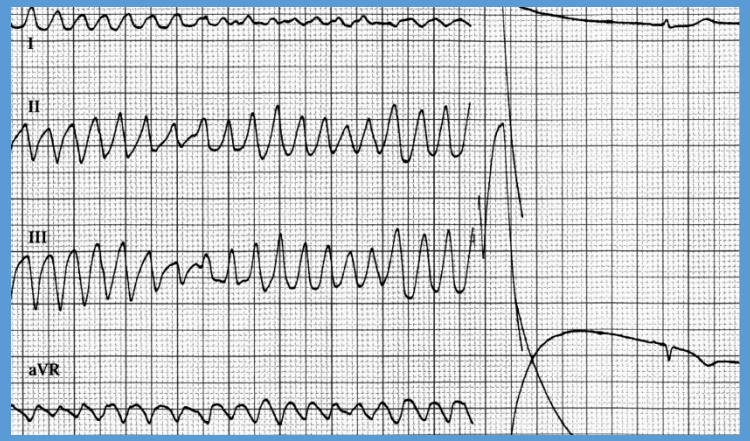
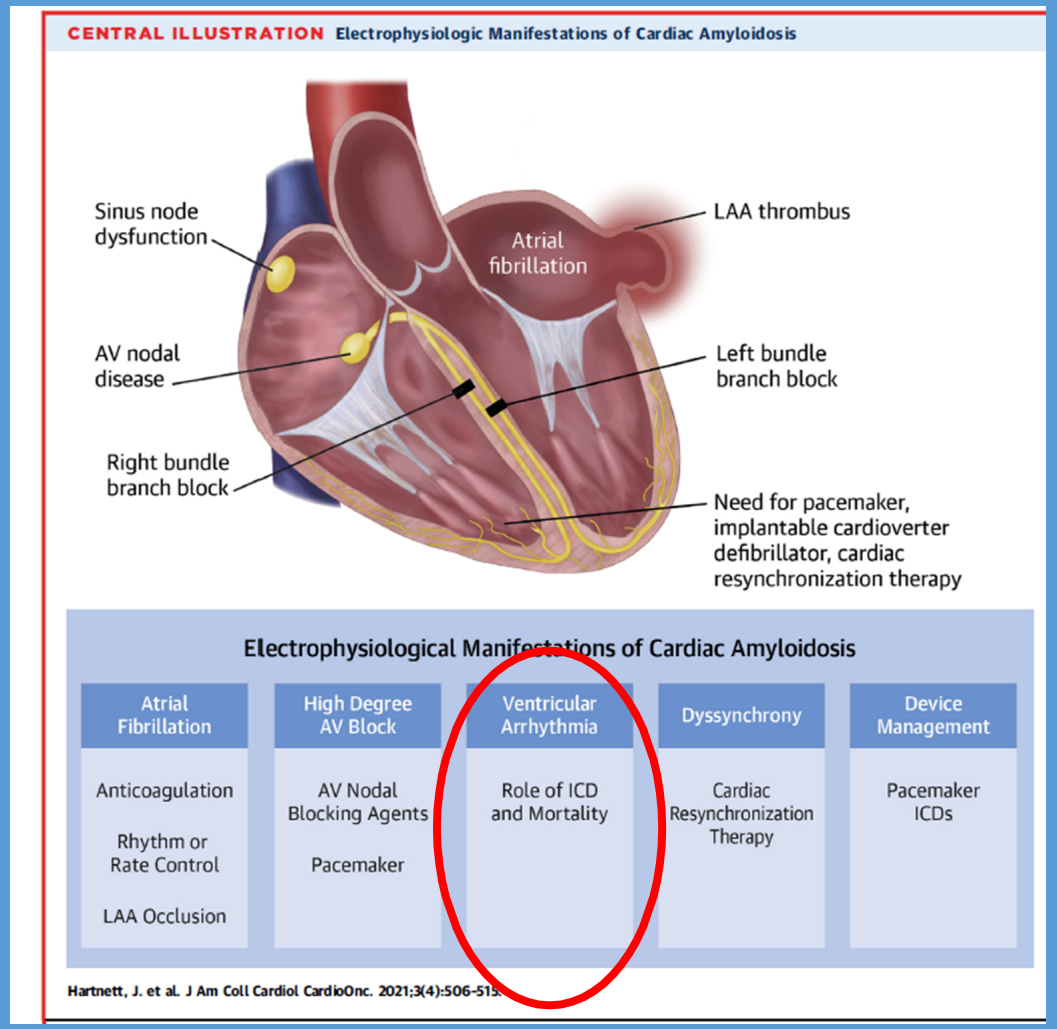
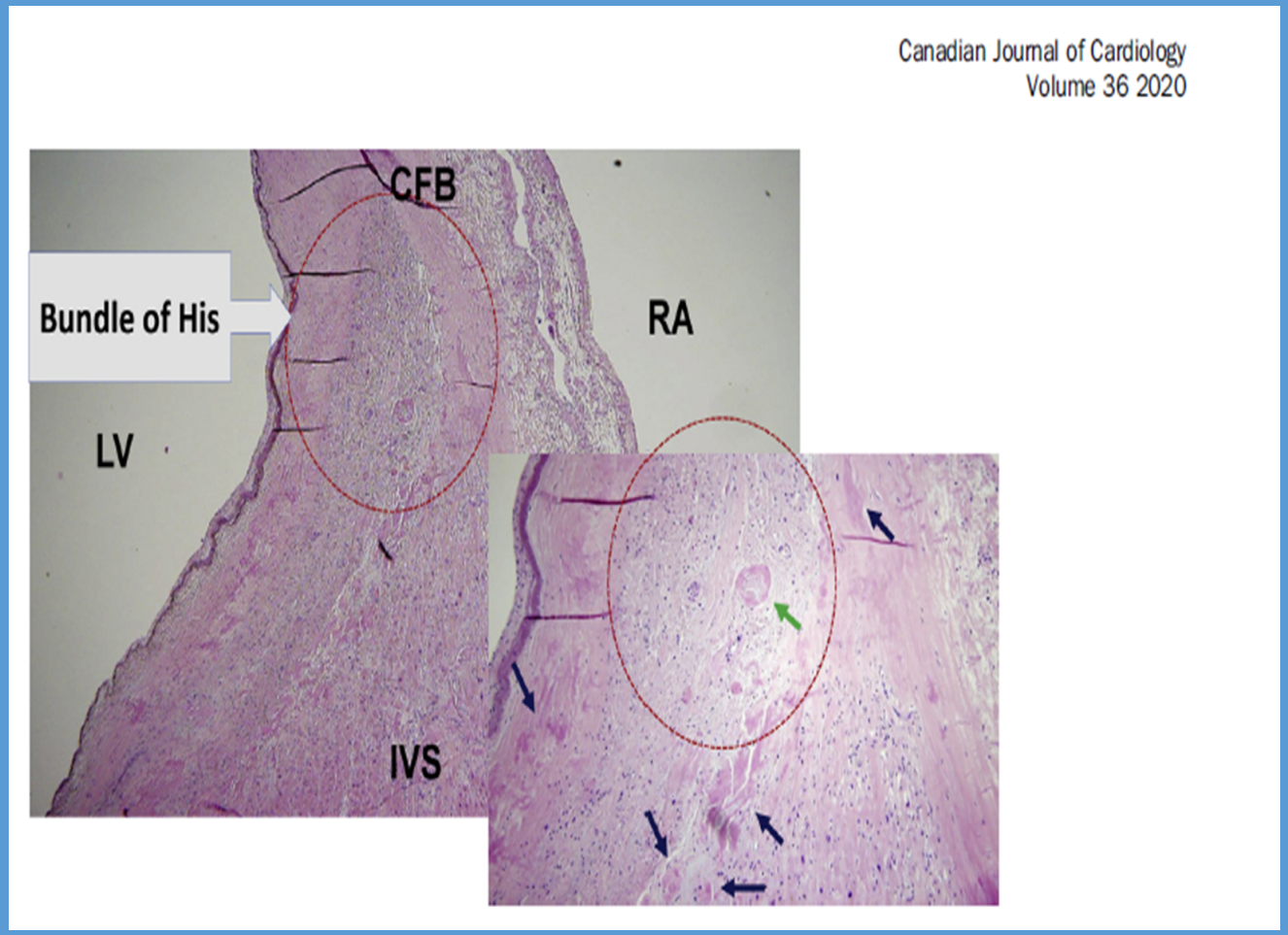
# Prise en charge des arythmies ventriculaires au cours de l'amylose cardiaque




Nicolas Lellouche
Rythmologie, Henri Mondor, Créteil, APHP

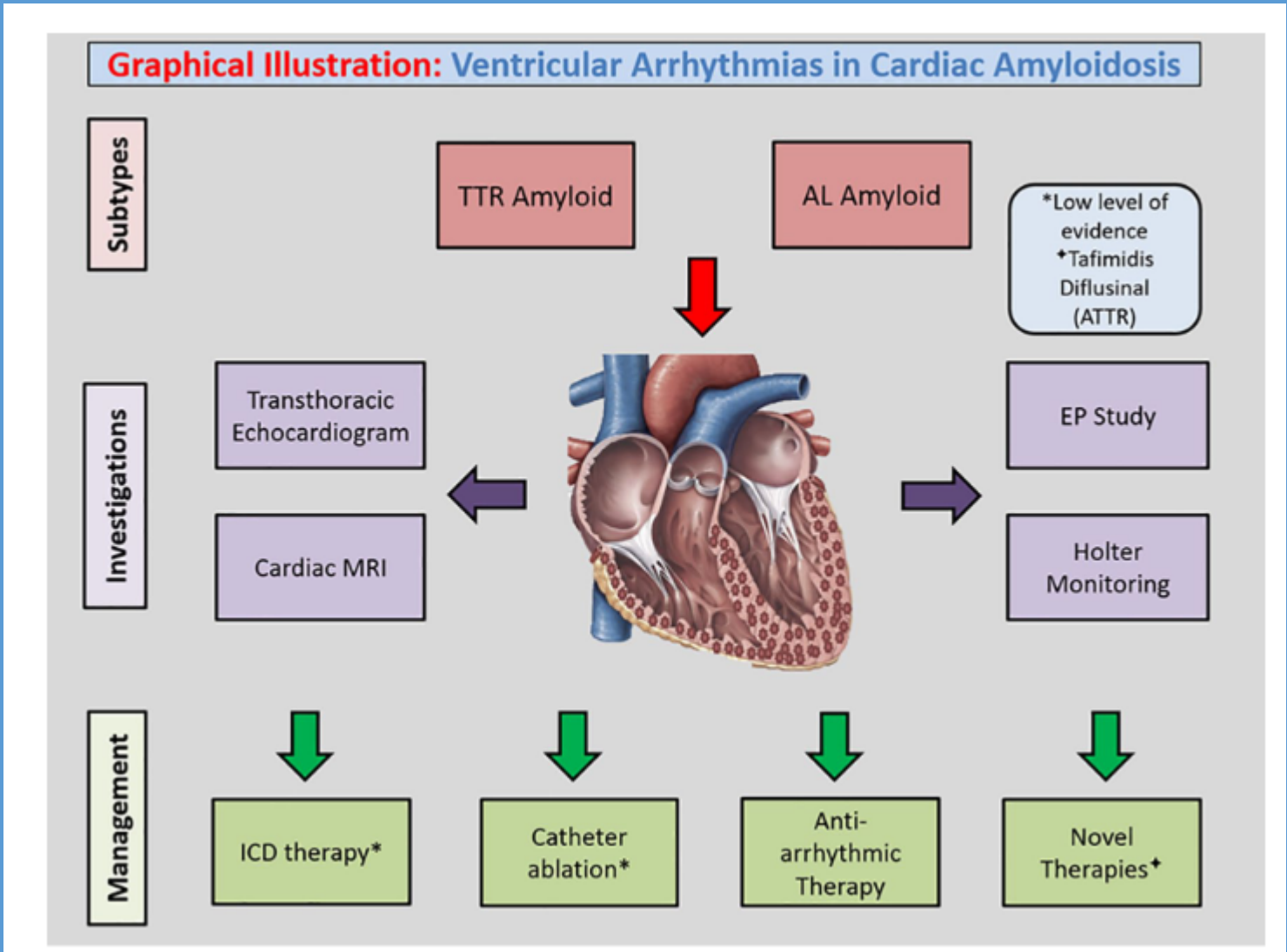
# Généralités




Arythmies atriales++
Peu d'arythmies ventriculaires + au cours de Amylose AL

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




# Toxicité cardiaque des chimiothérapies++

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# PREVALENCE DES TROUBLES DU RYTME VENTRICULAIRE

| 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%). |

TVNS: 20%/an

TV: 10%/an

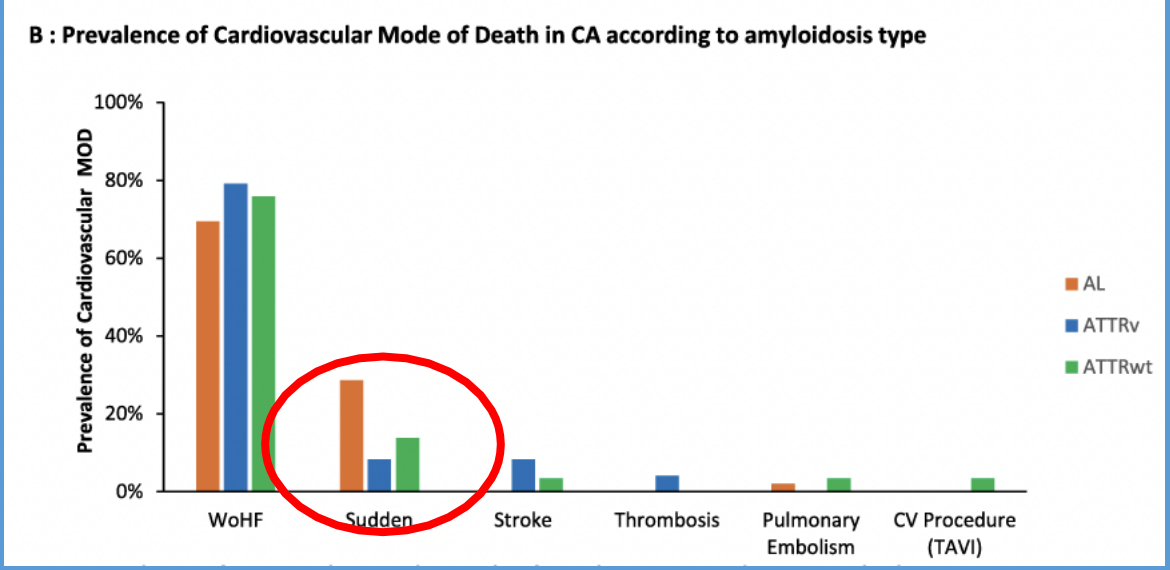
La prévalence dépend du type d'amylose et du mode de recueil:
DAI ou Monitoring continue (24h, 3 semaines ou 3 ans ILR)

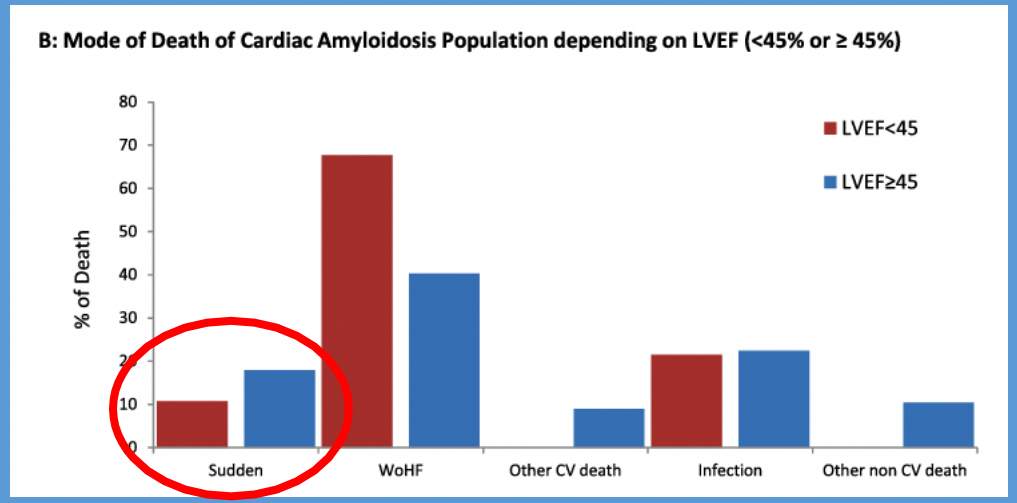
# Mode de décès au cours de l'amylose cardiaque

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

Mounira Kharoubi[a,b,c,d], Diane Bodez[a,b,c,d,e], Mélanie Bézard[a,b,c,d], Amira Zaroui[a,b,c,d], Arnault Galat[a,b,c,d], Soulef Guendouz[a,b,c,d], Thierry Gendre[a,b,f], Luc Hittinger[a,b,c,d], David Attias[e], Dania Mohty[g,h], Eric Bergoend[i], Emmanuel Itti[a,b,j], Fabien Lebras[a,b,k], David Hamon[a,b,c,d], Elsa Poullot[a,b,l], Valérie Molinier-Frenkel[a,b,l,m], Nicolas Lellouche[a,b,c,d], Jean-François Deux[a,b,n], Benoit Funalot[a,b,o], Pascale Fannen[a,b,o], Silvia Oghina[a,b,c,d], Raphael Arrouasse[a,b,p], Philippe Lecorvoisier[a,b,p], Sarah Souvannanorath[a,b,q], Aurelien Amiot[r], Emmanuel Teiger[a,b,c,d], Wulfran Bougouin[s,t] and Thibaud Damy[a,b,c,d,p]

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

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

# TRAITEMENTS

## A) Traitements anti-arythmiques

- **Amiodarone ++ mais attention au risque de TDP: drogues utilisées notamment dans l'amylose AL, hypoK+,..**
- **AA de classe I contre indiqués**
- **Béta-bloquants souvent mal tolérés**
- **Aucune série publiée sur l'ablation de TV (quelques cas cliniques)**
- **2 cas à Mondor avec bon succès**
- **Transplantation cardiaque en cas d'orage rythmique (très rare…)**

# B) Le DAI

-Prévention secondaire: Indication au DAI

-Prévention primaire: quels sont les FDR de mort subite?

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[†]

*From the [*]Division of Cardiology, Columbia University Medical Center, New York, New York, [†]Division of Cardiovascular Medicine, Stanford University Medical Center, Stanford University School of Medicine, Stanford, California, [‡]Division of Hematology, [§]Division of Nephrology; and [‖]Division of Blood and Marrow Transplantation, Stanford University School of Medicine, Stanford, California.*

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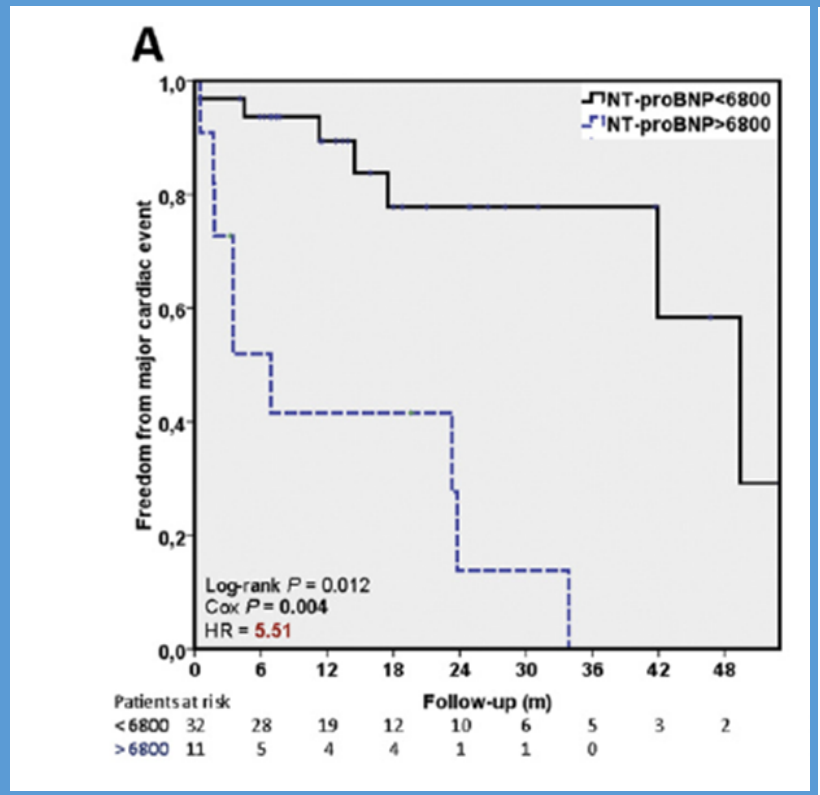
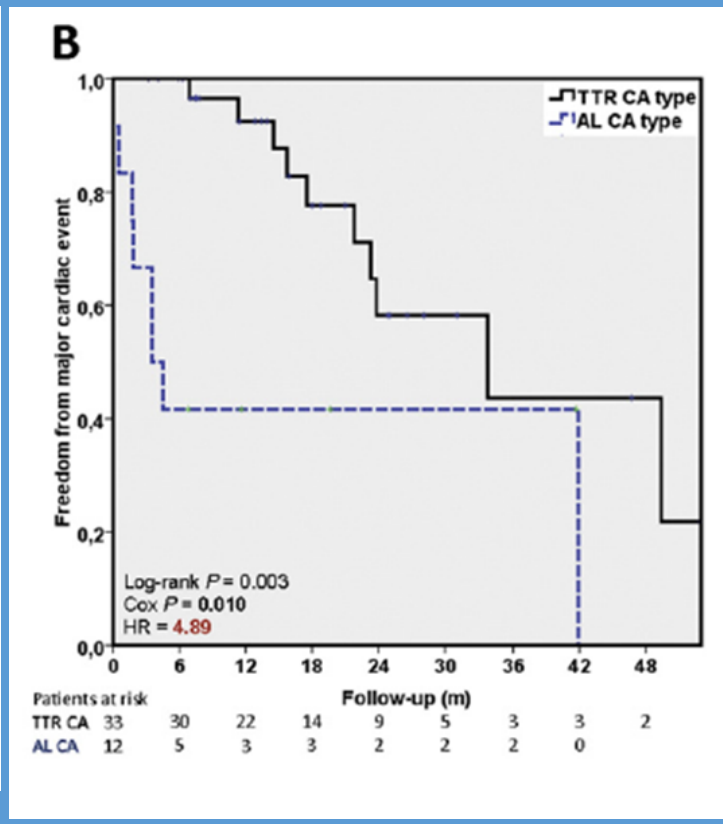
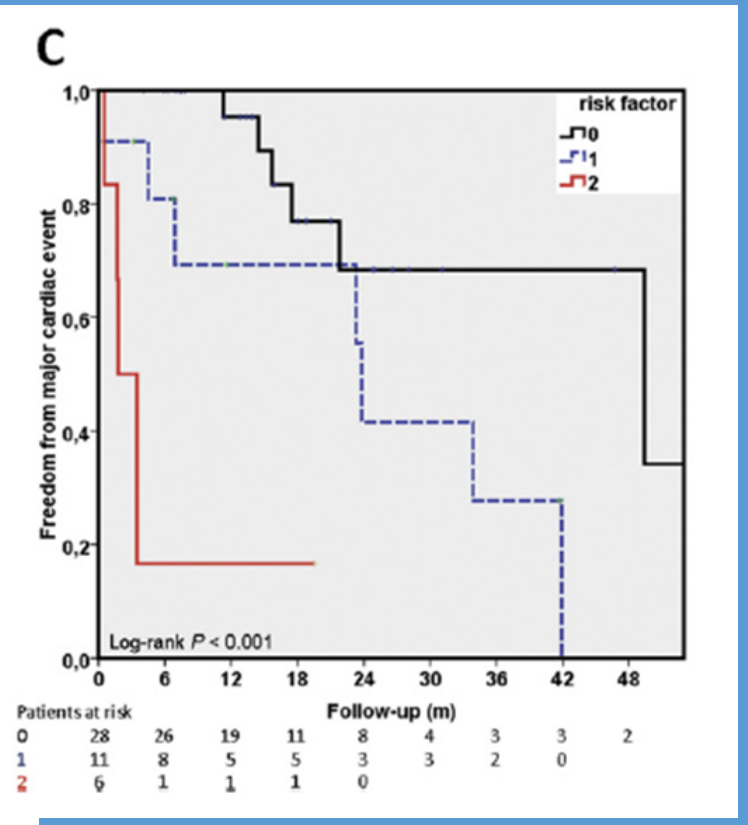
*Varr et al. Heart Rhythm. 2014 Jan;11(1):158-62.*




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.*

**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
doi:10.1093/europace/euaa094

CLINICAL RESEARCH
*Sudden death and ICDs*

# Outcomes in patients with cardiac amyloidosis and implantable cardioverter-defibrillator

**Eun-Jeong Kim [1], Benjamin B. Holmes[2], Shi Huang[3], Ricardo Lugo[2], Asad Al Aboud[2], Stacey Goodman[4], Rebecca R. Hung[2], David Slosky[2], William G. Stevenson[2], Gregory F. Michaud[2], and Roy M. John[5]***

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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) |
|---|---|---|---|

| | | Group 1 | Group 2 | *P*-value |
|---|---|---|---|---|
| *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.

FEVG basse chez les patients implantés

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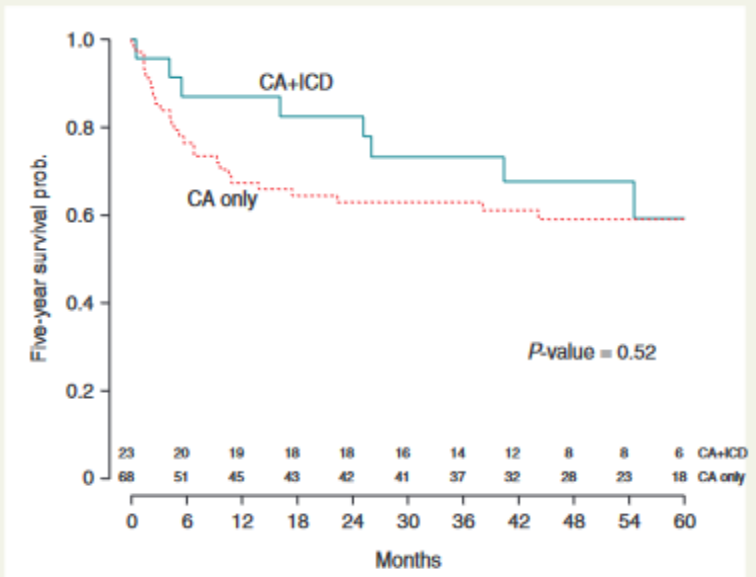



**Figure 2** The Kaplan–Meier plot of survival probabilities from CA diagnosis to death in patients with CA. The figure compared the survival of Group 1 (CA and ICD) with Group 2 (CA and no ICD) utilizing the Kaplan–Meier curve. There is no significant survival difference to 5 years after the diagnosis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

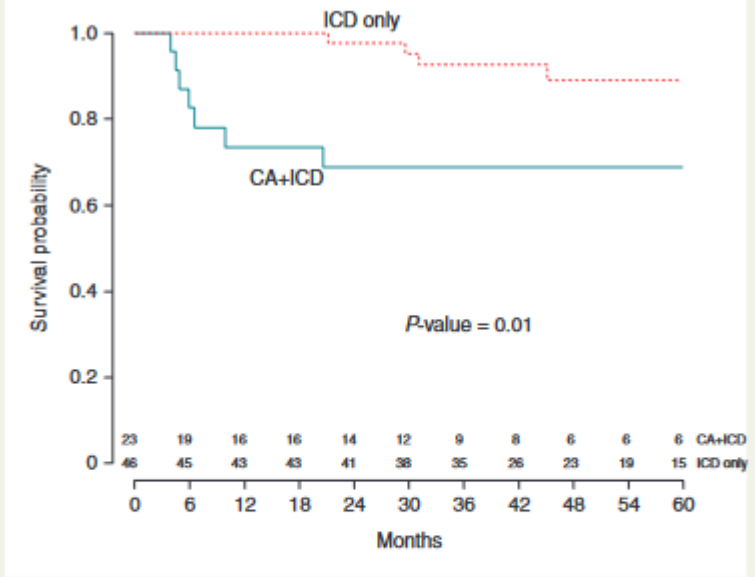
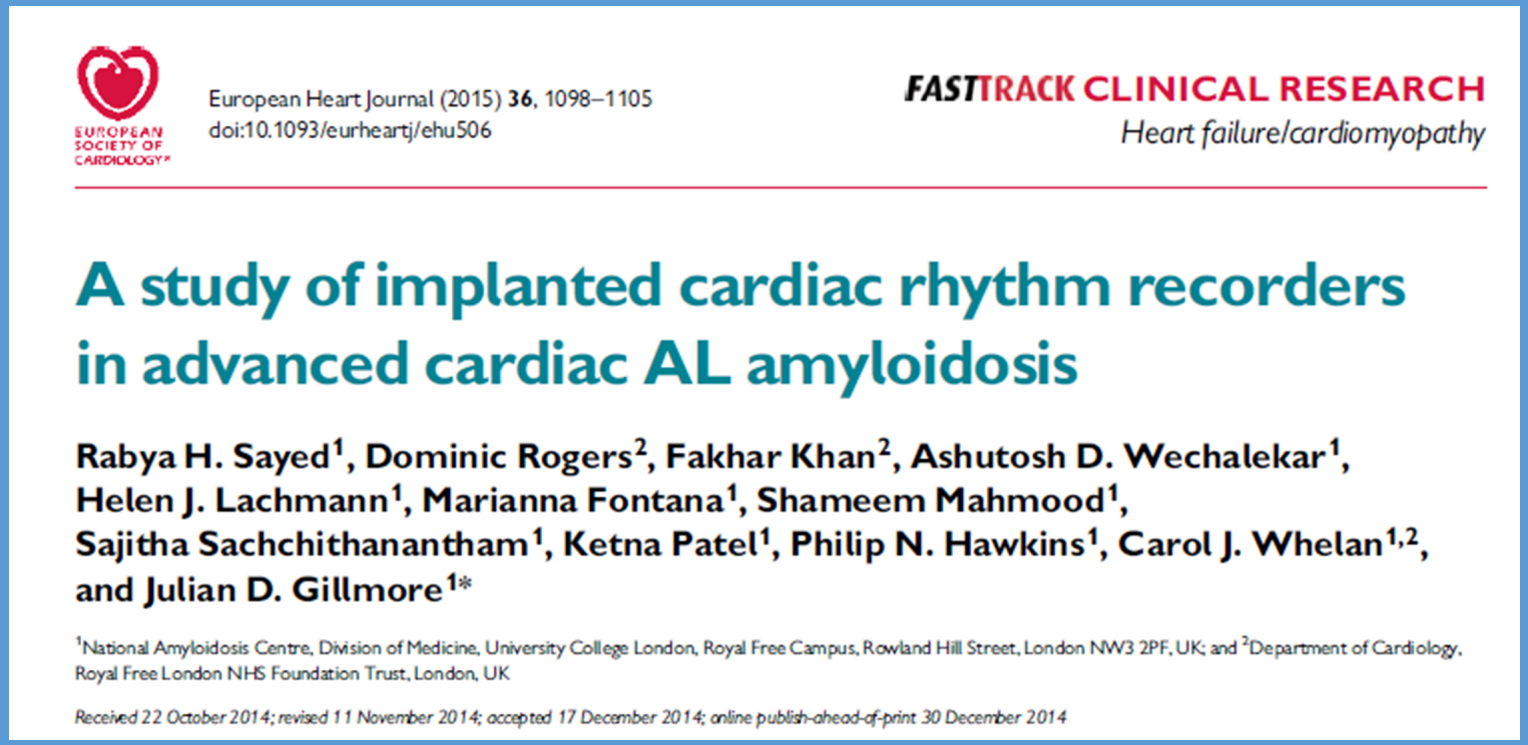



**Figure 3** The Kaplan–Meier plot of survival probabilities from ICD to death in patients with ICD. This figure compared the survival of Group 1 (CA and ICD) with Group 3 (non-amyloid and ICD) utilizing the Kaplan–Meier curve. ($P = 0.01$). Survival was significantly better in the non-amyloid population 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.



European Heart Journal (2015) **36**, 1098–1105
doi:10.1093/eurheartj/ehu506

**FASTTRACK CLINICAL RESEARCH**
*Heart failure/cardiomyopathy*

# A study of implanted cardiac rhythm recorders in advanced cardiac AL amyloidosis

**Rabya H. Sayed[1], Dominic Rogers[2], Fakhar Khan[2], Ashutosh D. Wechalekar[1], Helen J. Lachmann[1], Marianna Fontana[1], Shameem Mahmood[1], Sajitha Sachchithanantham[1], Ketna Patel[1], Philip N. Hawkins[1], Carol J. Whelan[1,2], and Julian D. Gillmore[1]***

[1]National Amyloidosis Centre, Division of Medicine, University College London, Royal Free Campus, Rowland Hill Street, London NW3 2PF, UK; and [2]Department of Cardiology, Royal Free London NHS Foundation Trust, London, UK

*Received 22 October 2014; revised 11 November 2014; accepted 17 December 2014; online publish-ahead-of-print 30 December 2014*

- Mais cause principale de DC dans amylose : trouble de conduction ou asystolie
- 20 patients implantés pour amylose AL sévère et syncope.
- 13 patients DCD dont 8 dus à un trouble de conduction de haut degré
- 1/3 DCD dans les 3 mois suivant l'implantation

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*Sayed, RH. et al. Eur Heart J. 2015;36(18):1098-105.*

**Table 2** Drug therapy including chemotherapy regimen, cardiac events, and outcome

| Pt | Baseline rhythm | Nature of cardiac rhythm change | Anti-arrhythmic medication prior to onset of cardiac rhythm change (or at censor[a]) | Chemotherapy regimen (days from last dose to cardiac rhythm change) | Intervention | Outcome (days from ILR to death/censor) |
|---|---|---|---|---|---|---|
| 1 | SR | Sinus bradycardia (HR 25–30 bpm), significant pauses followed by CAVB | None | CVD (2) | None (NFR) | Died, 12 |
| 2 | SR | Bradycardia (HR 30 bpm) | Bisoprolol 5 mg od | CVD (3) | None | Died, 22 |
| 3 | SR, 1st degree HB | CAVB with significant pauses (HR 20 bpm) | None | Lenalidomide/dexamethasone (14) | CPR, TP | Died, 57 |
| 4 | SR, 1st degree HB | CAVB (HR 25) | None | CVD (4) | CPR, PPM | Died, 10 |
| 5 | Atrial flutter | CAVB (HR 20 bpm) | Amiodarone 200 mg od | CVD (3) | None | Died, 44 |
| 6 | SR | Sinus bradycardia (HR 35 bpm) | Bisoprolol 3.75 mg od | CVD (7) | CPR | Died, 34 |
| 7 | SR | None detected up to 4 days prior to death | None | CVD (3) | None | Died, 75 |
| 8 | SR | CAVB (HR 30 bpm) | Bisoprolol 1.25 mg od | CVD (4) | CPR, TP | Died, 17 |
| 9 | SR, 1st degree HB | Atrial flutter 120–160 bpm | None | CVD (4) | Atrial ablation | Alive, 399 |
| 10 | SR | AF at >140 bpm | None | CVD (NA) | Amiodarone | Alive, 308 |
| 11 | SR | None detected | None[a] | CVD (NA) | NA | Alive, 353 |
| 12 | SR, 1st degree HB | None detected | None[a] | CVD (NA) | NA | Alive, 357 |
| 13 | SR, 1st degree HB | Sinus bradycardia, significant pauses and CAVB | None | None (CVD later) | PPM | Alive, 300 |
| 14 | SR, Wenckebach | CAVB (30 bpm) followed by NSVT | Bisoprolol 1.25 mg od | CVD (6) | None | Died, 45 |
| 15 | SR | None detected up to 6 days prior to death | None | CVD (6) | None | Died, 36 |
| 16 | SR, 1st degree HB | None detected | Bisoprolol 1.25 mg od | Melphalan/dexamethasone (NA) | NA | Died, 81 |
| 17 | SR, 1st degree HB | None detected | Bisoprolol 1.25 mg od | CVD (12) | NA | Died, 65 |
| 18 | SR, 1st degree HB Paroxysmal AF | AF | None | CVD (NA) | Amiodarone | Alive, 186 |
| 19 | SR, 1st degree HB | AF at 140 bpm | None | CVD (1) | Bisoprolol | Alive, 193 |
| 20 | AF | None detected | None | CVD (NA) | NA | Died, 49 |

SR, sinus rhythm; CVD, cyclophophosphamide, velcade, and dexamethasone; HB, heart block; CAVB, complete atrioventricular block; TP, temporary pacing; PPM, permanent pacemaker; CPR, cardiopulmonary resuscitation; AF, atrial fibrillation; NK, not known; ChemoRx, chemotherapy treatment; od, once daily.
[a]Medication at time of censor among those without a cardiac rhythm change.

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*Sayed, RH. et al. Eur Heart J. 2015;36(18):1098-105.*

-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 |

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Reisinger et al. JACC 1997

# 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

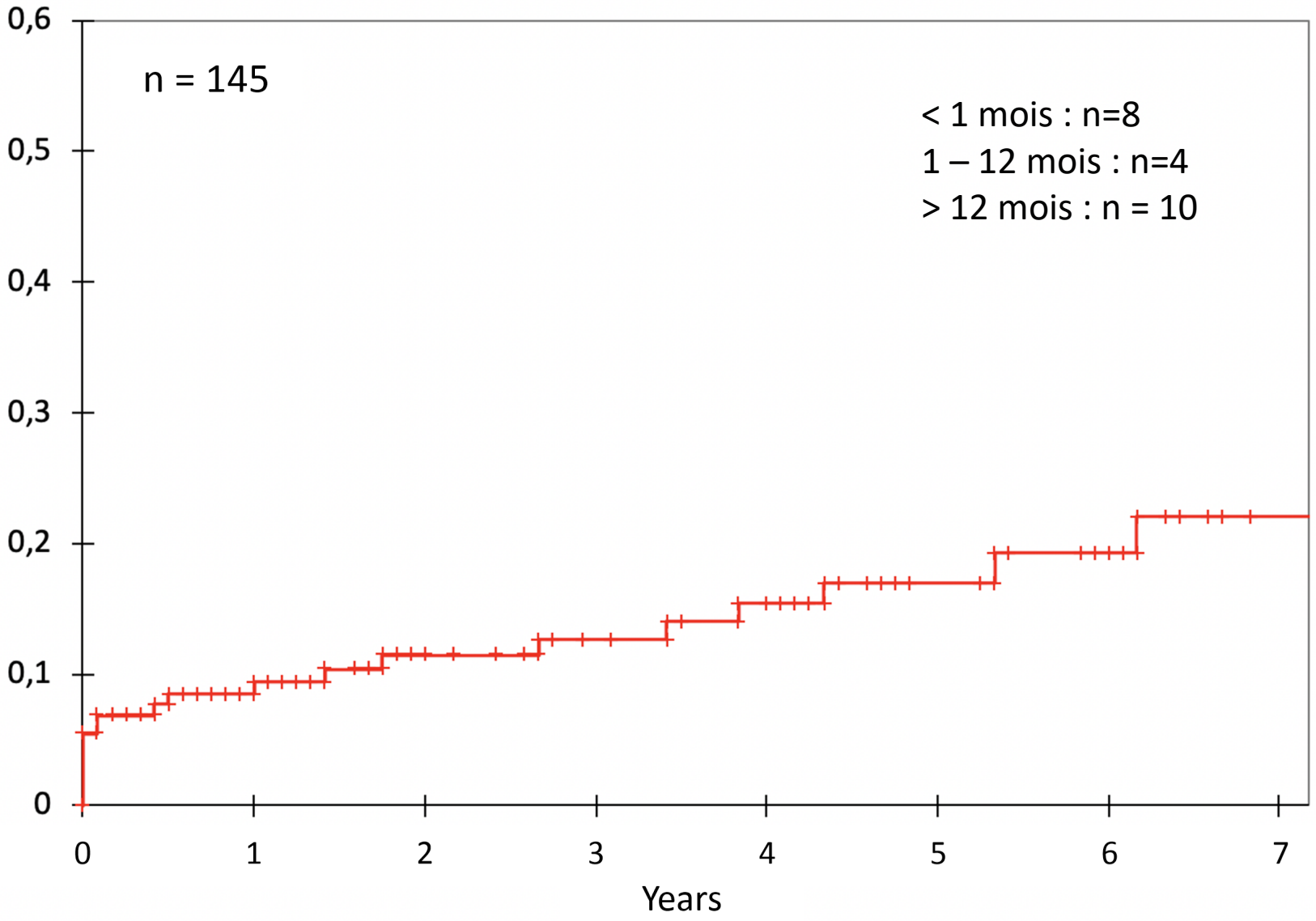
# Critère primaire

- **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)

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

## Cumulative incidence of first appropriate ICD therapy



# Thérapies

- 37 thérapies chez 22 patients

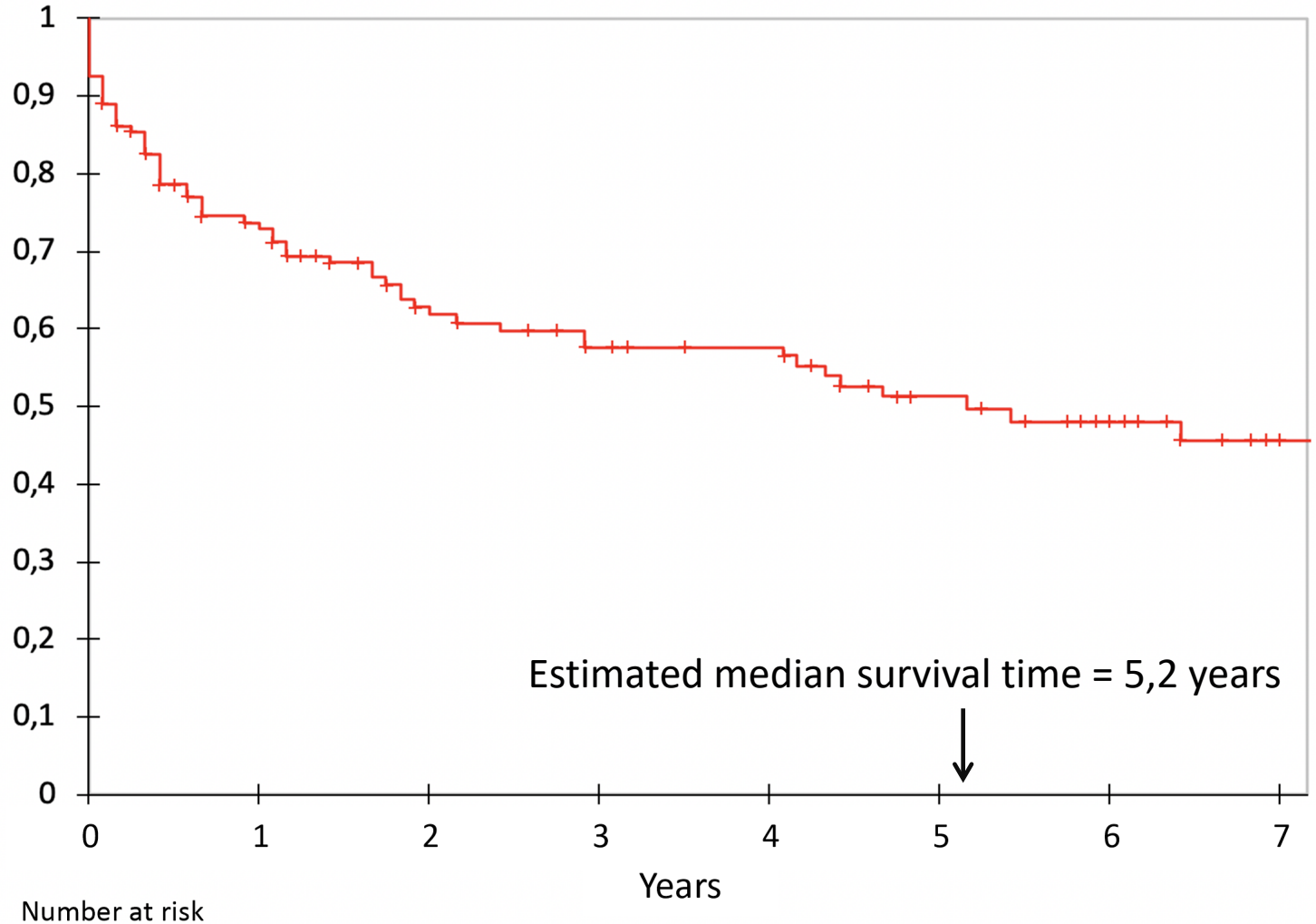
- 5 patients (3,5%) avec > 1 thérapie

- 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

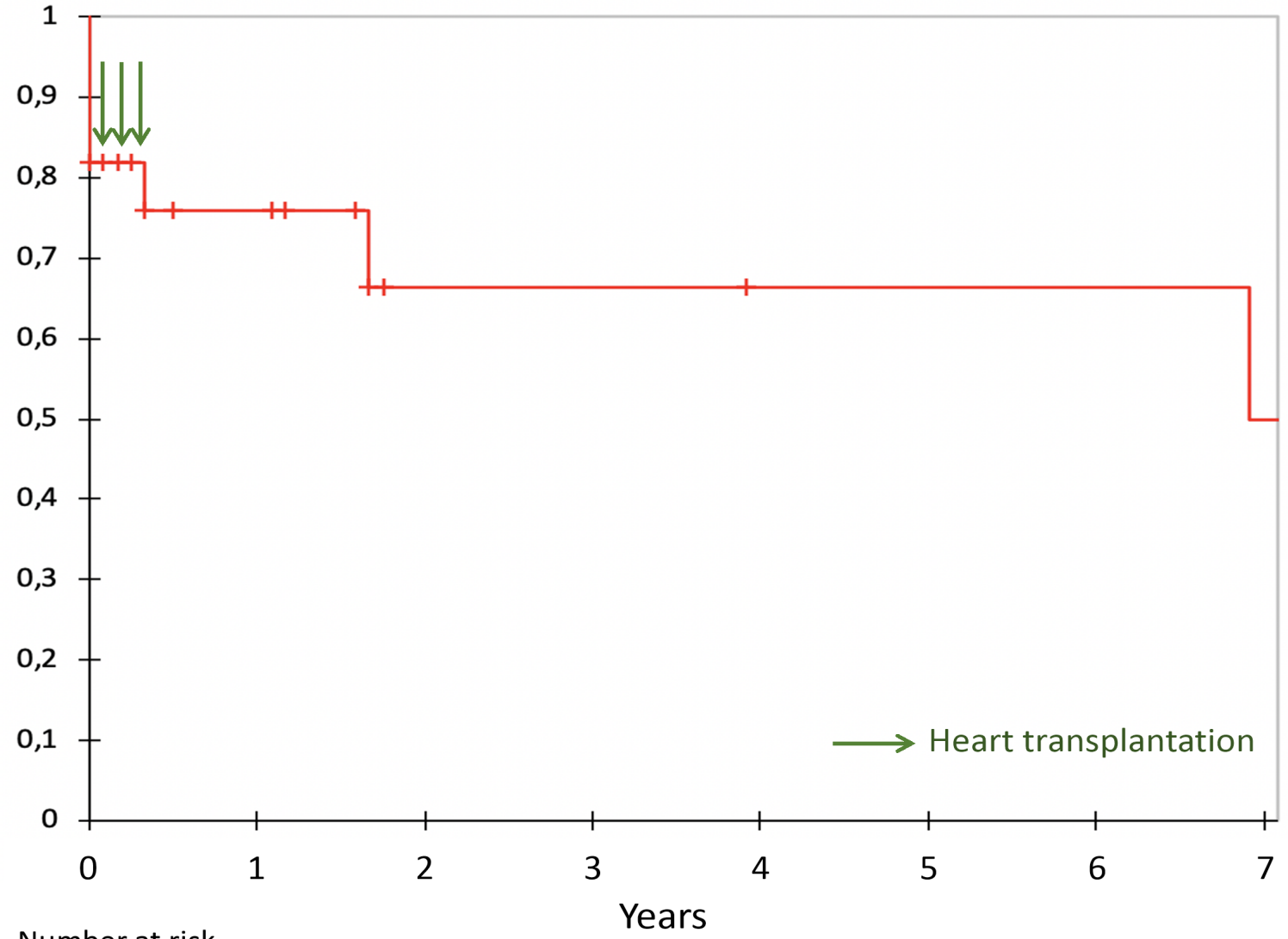
## Kaplan-Meier curve of survival in AL-CA patients with ICD




Number at risk

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

## 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

## 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)

## LIFE vest

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

# Take Home Messages

-La mort subite n'est pas le mode de DC principal au cours de l'amylose mais représente environ 20% des DC

-Ce taux semble plus élevé au cours des amyloses AL vs. TTR

-La FEVG semble moins stratifiante de ce risque/ autres cardiopathies
(ne pas implanter trop tardivement++, espérance de vie > 1 an)

-Taux de thérapies appropriées assez élevés environ 15-30% /an surtout chez les AL

-Facteurs de mort subite à définir: strain, VG, TVNS, syncope, fibrose IRM?

-Effet des thérapies de la maladie elle-même?

# MERCI DE VOTRE ATTENTION!!