

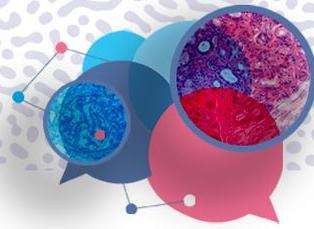
Controverses de la prise en charge cardiologique

Défibrillateur dans les amyloses AL

CONTRE

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Défibrillateur dans les amyloses

Esperance de vie > 1 an

DAI en prévention secondaire



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

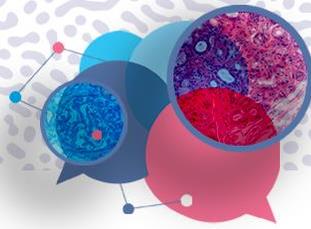
Recommendations	Class ^a	Level ^b
An ICD should be considered in patients with light-chain amyloidosis or transthyretin-associated cardiac amyloidosis and haemodynamically not-tolerated VT.	IIa	C

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DAI en prévention primaire



?

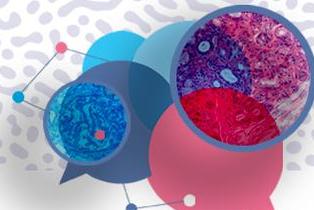


Que disent les recommandations?

- **ESC guidelines 2015 et 2022:** DAI uniquement en prévention secondaire IIa/C
- **American guidelines 2013** DAI **prévention primaire** si amylose avec LVEF < 50% + syncope inexpliquée
- **Update American guidelines 2017** DAI : **prévention primaire est retiré**
- **Réccomandation Heart Ryhtme Association** **classe II b/C**

stratification du risque / les paramètres biologiques (troponine et NT-proBNP)+ imagerie (IRM) + survie >1 an

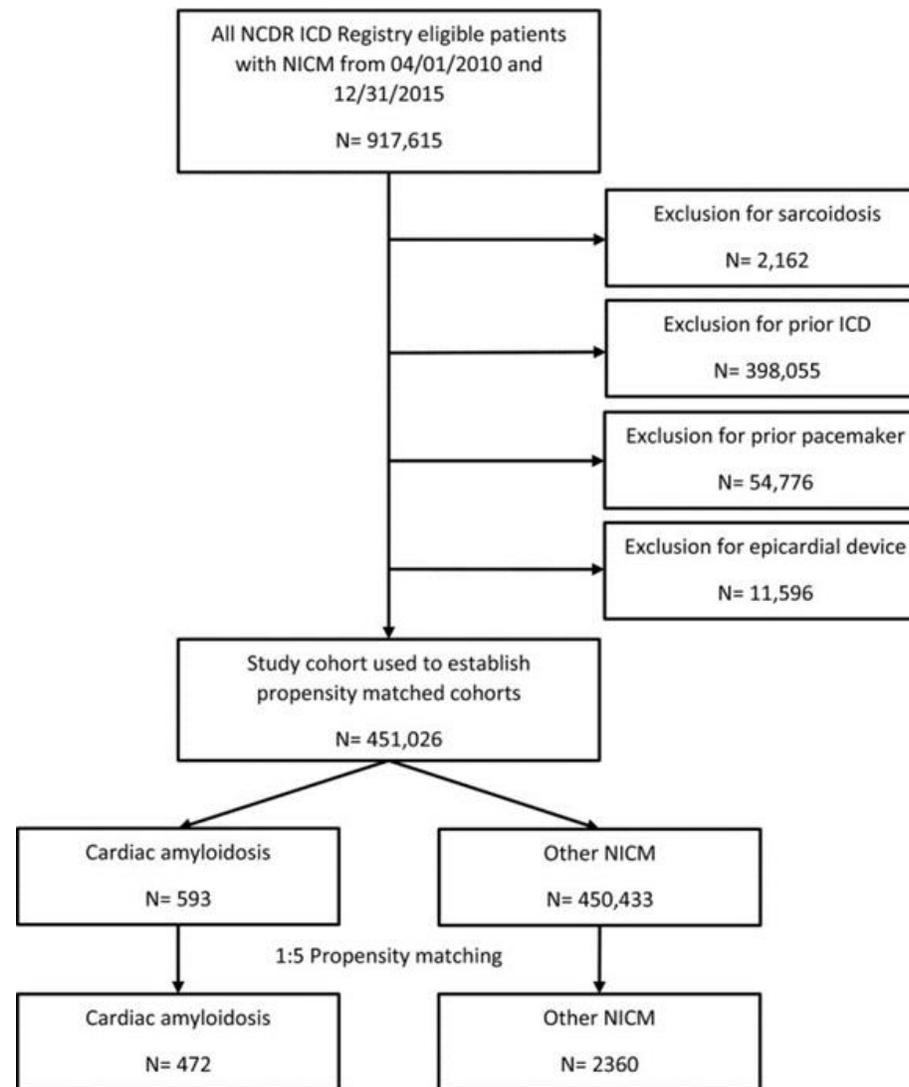
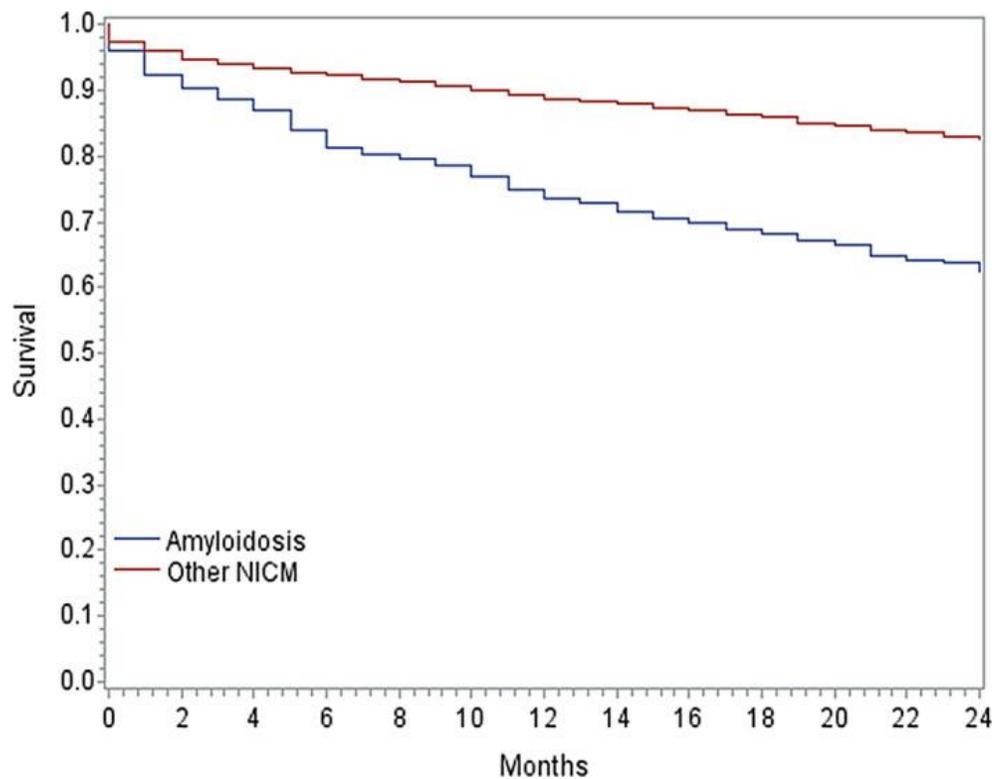
- **German consensus :**
 - (a) decision cas par cas
 - b) plutot pour en prevention primaire si risque de mortalité élevée (paramètres biologiques, imagerie + VT + si espérance vie >1 an).

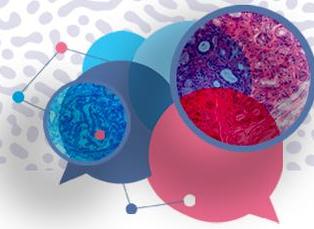


► J Am Heart Assoc. 2020 Sep 1;9(18):e016038. doi: [10.1161/JAHA.120.016038](https://doi.org/10.1161/JAHA.120.016038)

Survival Following Implantable Cardioverter-Defibrillator Implantation in Patients With Amyloid Cardiomyopathy

Angela Y Higgins¹, Amarnath R Annapureddy^{1,2}, Yongfei Wang², Karl E Minges², Rachel Lampert¹, Lynda E Rosenfeld¹, Daniel L Jacoby¹, Jephtha P Curtis^{1,2}, Edward J Miller¹, James V Freeman^{1,2}✉





- **Mortalité** après l'implantation de DAI de **25% à 1 ans** chez les patient ayant une amylose cardiaque
- **2 fois plus** par rapport aux patients NICMs
- 6 variables pour mortalité a 1 an pour les patients avec **amylose cardiaque + DAI:**

⇒ *syncope, TV*

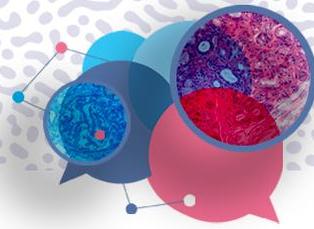
⇒ *Diabète, l'insuffisance rénale , maladie cerebro-vasculaire sévère*

Table 3.

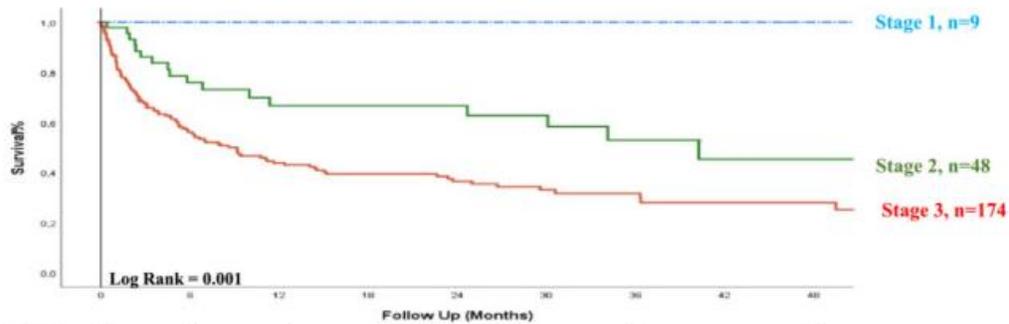
Patient Risk Factors Significantly Associated With 1-Year Survival in Multivariable Analysis Among Those With Cardiac Amyloidosis

Parameter	Hazard Ratio	95% CI	P Value
Syncope	1.78	1.22–2.59	0.003
Ventricular tachycardia	1.65	1.15–2.38	0.01
Prior cerebrovascular disease	2.03	1.28–3.23	0.003
Diabetes mellitus	1.55	1.05–2.27	0.03
Creatinine = 1.6–2.5 g/dL	1.99	1.32–3.02	0.001
Creatinine >2.5 g/dL	4.34	2.72–6.93	<0.001

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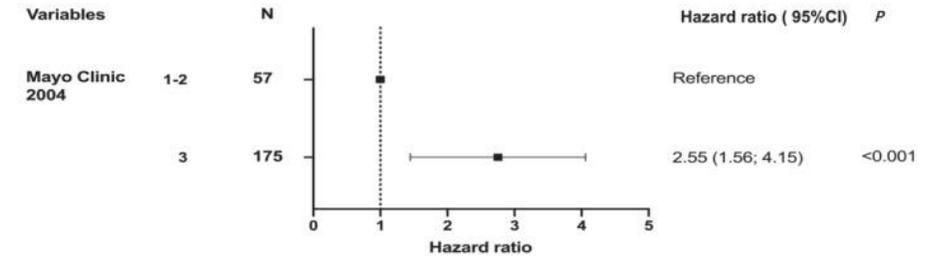


(B) Overall survival and cardiac transplant according to Mayo Clinic 2004 staging

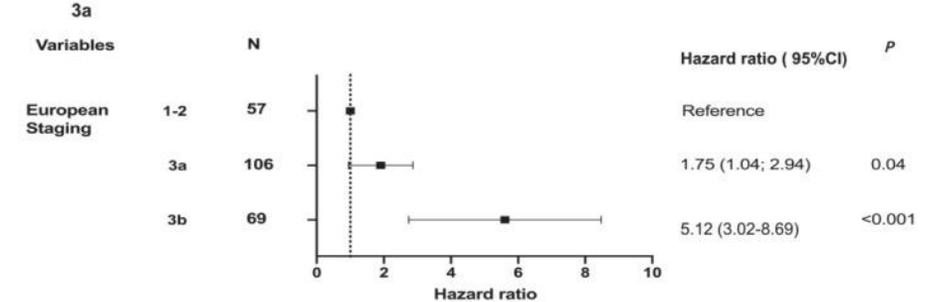


	0	6	12	18	24	30	36	42	48
Stage1	9	9	9	9	9	9	9	9	9
Stage2	48	37	35	34	32	31			
Stage3	175	98	84	76	70	69			

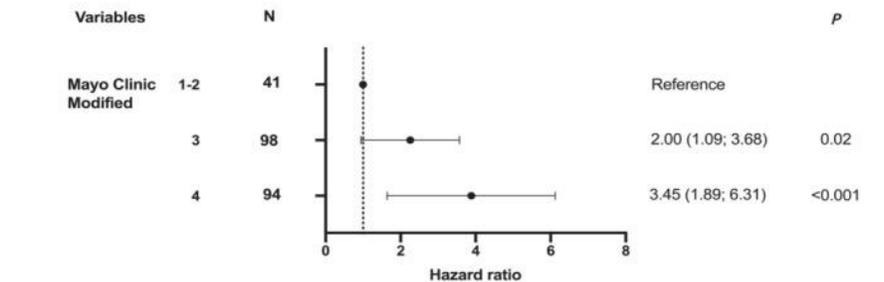
(A) Mortality and heart transplant risk is higher among Mayo clinic 2004 stage 3 than stage 2



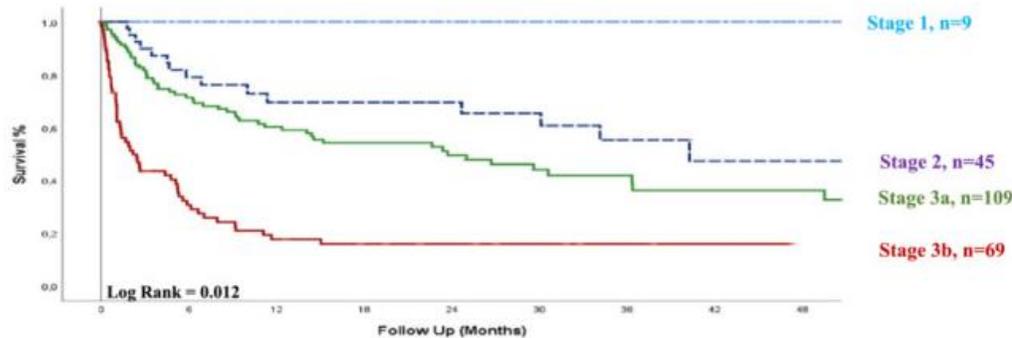
(B) Mortality and heart transplant risk is three times higher among European stage 3b than stage 3a



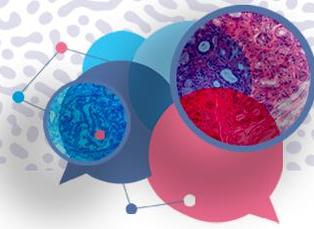
(C) Mortality and cardiac transplant risk is one and half times higher among modified 2012 Mayo stage 3 than stage 4



(C) Overall survival and cardiac transplant according to European Staging



	0	6	12	18	24	30	36	42	48
Stage1	9	9	9	9	9	9	9	9	9
Stage2	45	36	34	33	31	30			
Stage 3A	109	77	69	62	56	55			
Stage 3b	69	22	16	15	15	15			



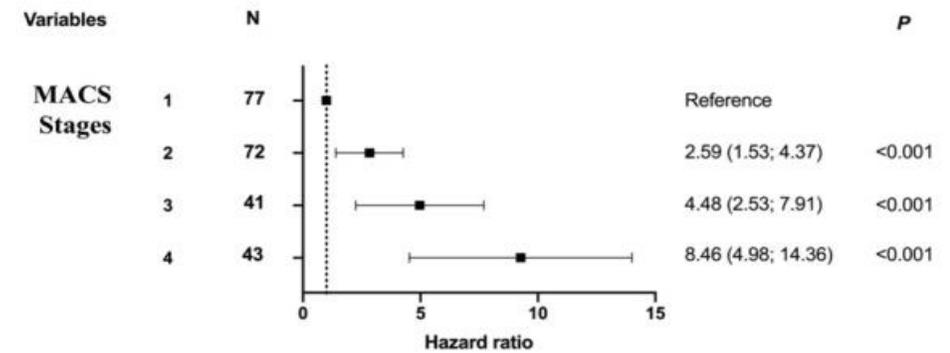
Prognostic mortality factors in advanced light chain cardiac amyloidosis: A prospective cohort study

A.Zaroui et al, ESC Heart Fail. 2024 Jun;11(3):1707-1719. doi: 10.1002/ehf2.14671.

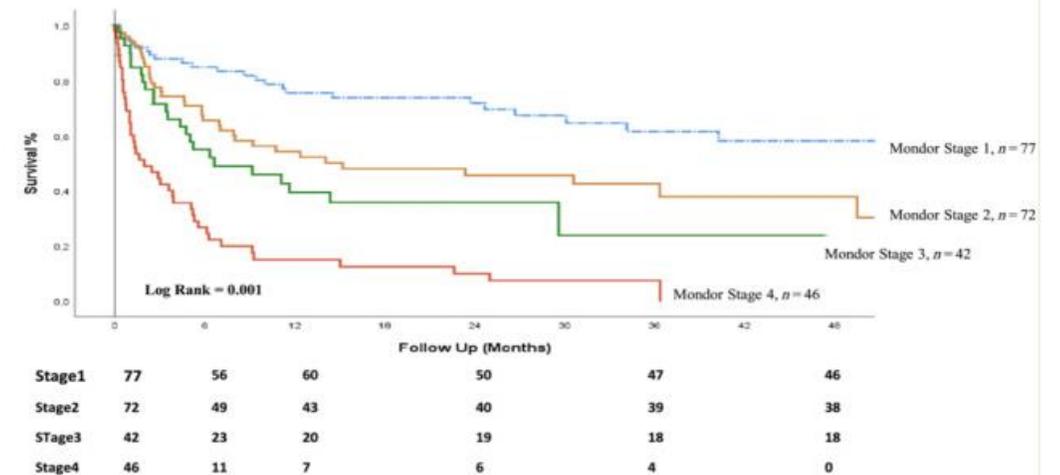
Survie a 1 an:

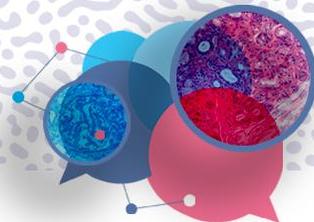
- 75% for stage 1,
- 52%, for stage 2,
- 39% for stage 3 and
- 12% for stage 4 ($P = 0.002$)

(A) Mortality and cardiac transplant risk doubles between MACS stage 2, 3 and 4.



(B) All cause mortality and cardiac transplant according to Mondor Amyloidosis Cardiac Stage





Review

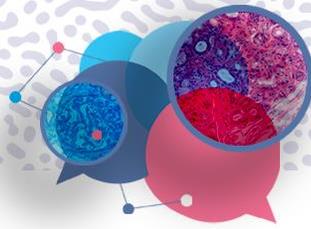
Arrhythmic Manifestations of Cardiac Amyloidosis: Challenges in Risk Stratification and Clinical Management

Natallia Laptseva ^{1,2,4}, Valentina A. Rossi ¹, Isabella Sudano ¹, Rahel Schwotzer ^{2,3}, Frank Ruschitzka ^{1,2},
Andreas J. Flammer ^{1,2} and Firat Duru ^{4,5}

J. Clin. Med. 2023, 12, 2581. <https://doi.org/10.3390/jcm12072581>

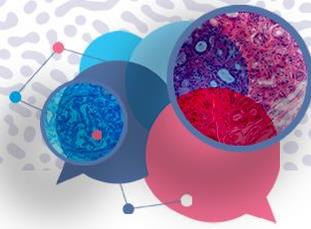
Table 2. Arrhythmia risk and indication for device therapy in various forms of cardiac amyloidosis.

	AL	vATTR	wtATTR
Arrhythmia risk			
Sinus node disease	+ / ++	++	++
AV conduction disease	++	+++	+++
Atrial fibrillation	+++ / ++++	+++	+++
nsVT / VT	+++	++ / +++	++ / +++
SCD risk	+++	++	++
Device indication			
PM	+	+++	++ / +++
CRT	+ / ++	++ / +++	++
ICD (primary prevention)	+ / ?	+ / ?	+ / ?



Conclusions : DAI dans l'amylose

- 1) **Taux élevé d'arythmies ventriculaires** : 20 à 40% des patients implantés avec une amylose ont présenté des arythmies ventriculaires déclenchant des thérapies.
- 2) Dans l'amylose cardiaque, être porteur d'un DAI **n'était pas associé à une amélioration du pronostic vital**
 - ***la survenue d'une arythmie ventriculaire pourrait être plus un marqueur de l'évolution terminale*** de la maladie qu'une complication
 - **les recommandations actuelles réservent l'implantation d'un DAI en prévention secondaire**
amylose +TV mal tolérée cliniquement



Conclusions : DAI dans l'amylose

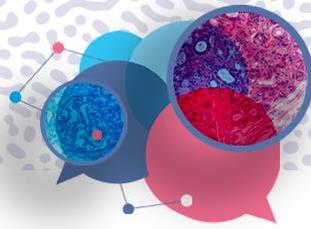
- Manque de données dans la littérature:

études peu robuste / cohortes limitées / résultats contradictoires / sans diminution de la mortalité ni du risque de MS

- **Les MS observées dans l'amylose cardiaque ne sont pas toujours rythmiques!**

- Incidence élevé des dissociations électromécaniques

désamorçage ventriculaire (BAV haut degré, découplage electro-mecanique)



Autres solutions /thérapies ?

- ? **MÉDICAMENTEUSE**: Amiodarone
- ? **L'ABLATION** (endocavitaire/ epicardique) des arythmies ventriculaires
- ? **LIFEVEST** ... bridge to decision /bridge to recovery