

LA PRISE EN CHARGE CARDIOLOGIQUE DES AMYLOSES NON SPÉCIFIQUES

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CHU de Toulouse

2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure

McDonagh, et al. *Eur Heart J* 2021;42:3599-3726

Diagnosis and treatment of cardiac amyloidosis: a position statement of the ESC Working Group on Myocardial and Pericardial Diseases

Garcia-Pavia P, et al. *Eur Heart J*. 2021;42:1554-1568

2023 Focused Update of the 2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure

McDonagh, et al. *Eur Heart J*. 2023;44:3627-3639

2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure

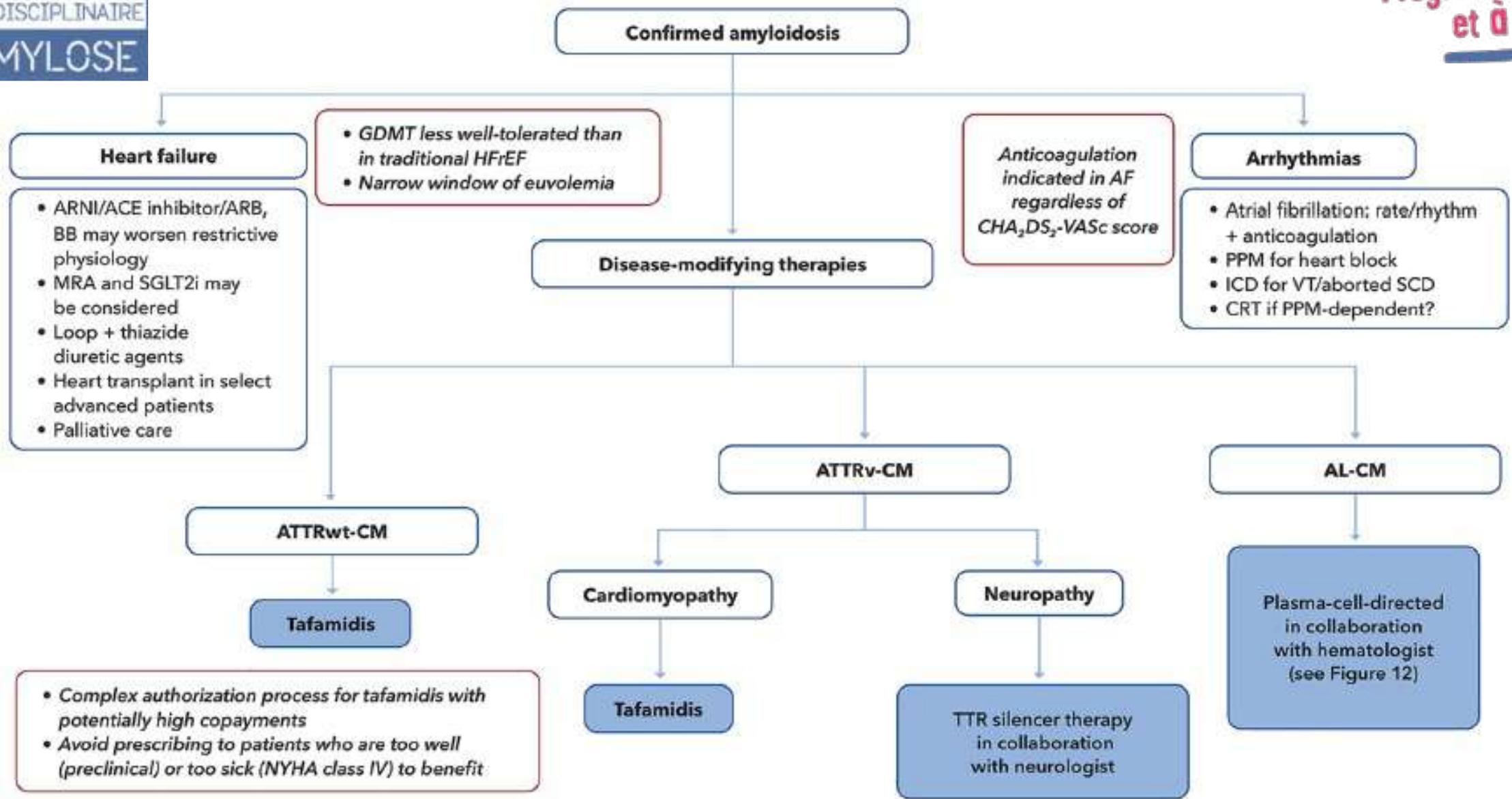
Progrès réalisés
et à venir !

Type of HF		HFrEF	HFmrEF	HFpEF
CRITERIA	1	Symptoms ± Signs ^a	Symptoms ± Signs ^a	Symptoms ± Signs ^a
	2	LVEF ≤40%	LVEF 41 – 49% ^b	LVEF ≥50%
	3	–	–	Objective evidence of cardiac structural and/or functional abnormalities consistent with the presence of LV diastolic dysfunction/raised LV filling pressures, including raised natriuretic peptides ^c

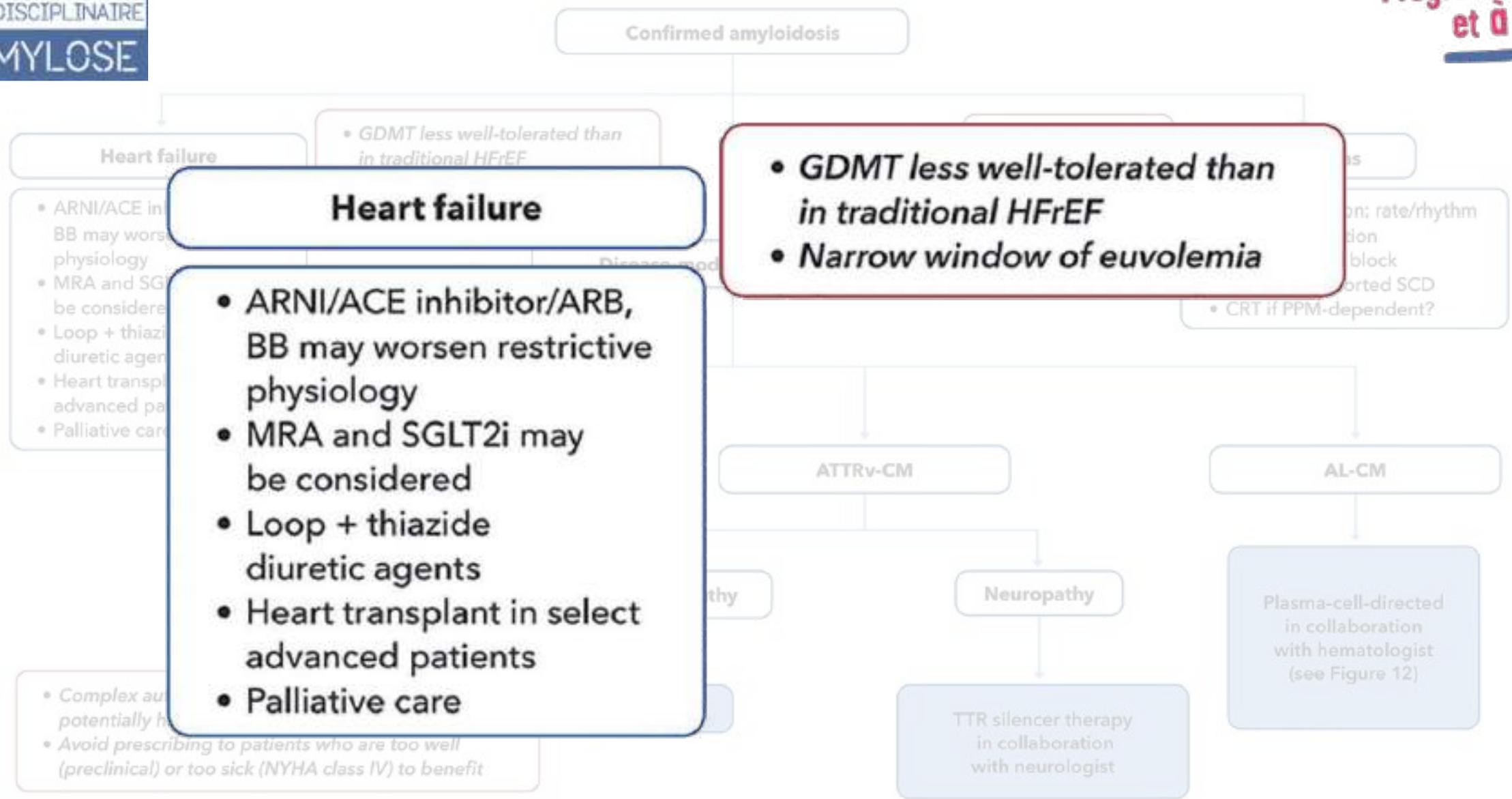
2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure

**Progrès réalisés
et à venir !**

Type of HF		HFpEF
CRITERIA	1	Symptoms ± Signs ^a
	2	LVEF \geq 50%
	3	Objective evidence of cardiac structural and/or functional abnormalities consistent with the presence of LV diastolic dysfunction/raised LV filling pressures, including raised natriuretic peptides ^c

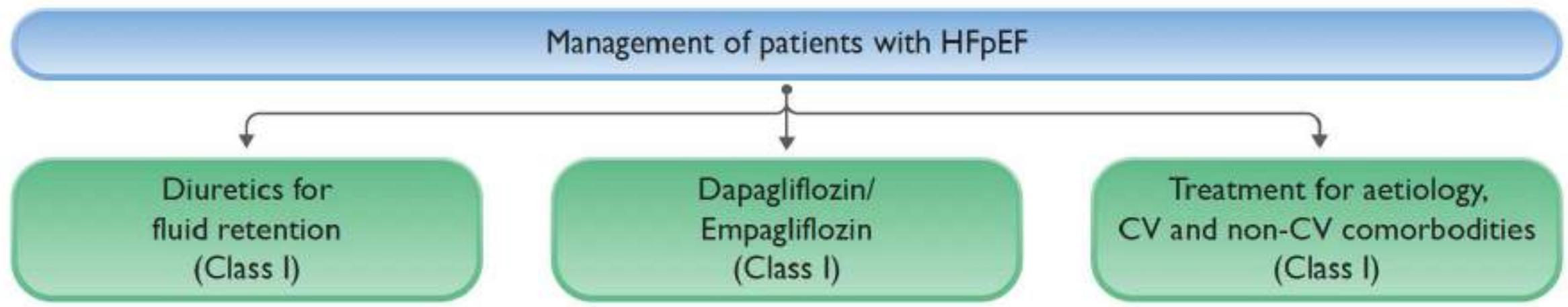


• *Complex authorization process for tafamidis with potentially high copayments*
 • *Avoid prescribing to patients who are too well (preclinical) or too sick (NYHA class IV) to benefit*



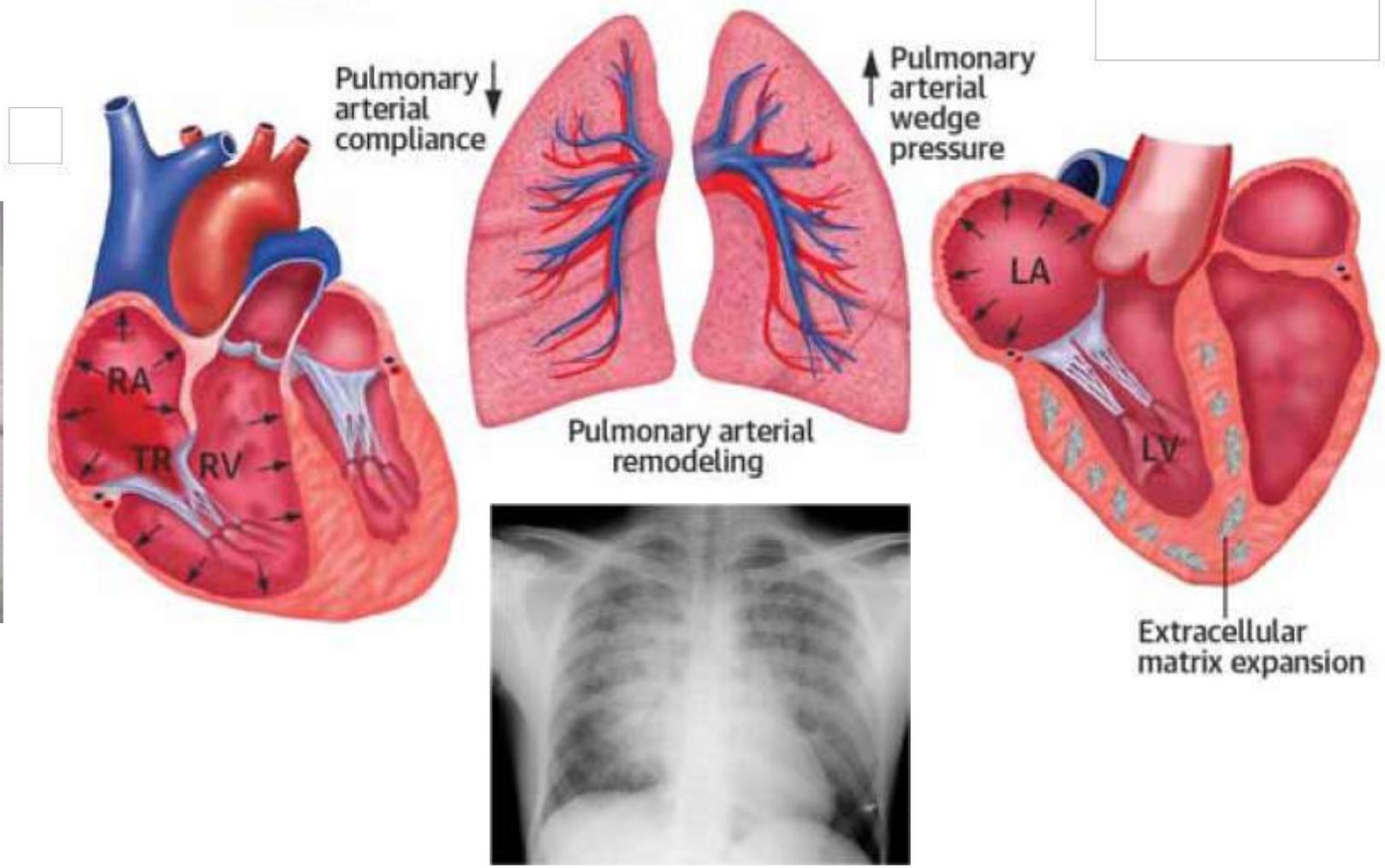
2023 Focused Update of the 2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure

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PRISE EN CHARGE DES SYMPTÔMES : GESTION DE LA VOLÉMIE

Progrès réalisés
et à venir !



BÉTABLOQUANTS & AMYLOSE CARDIAQUE

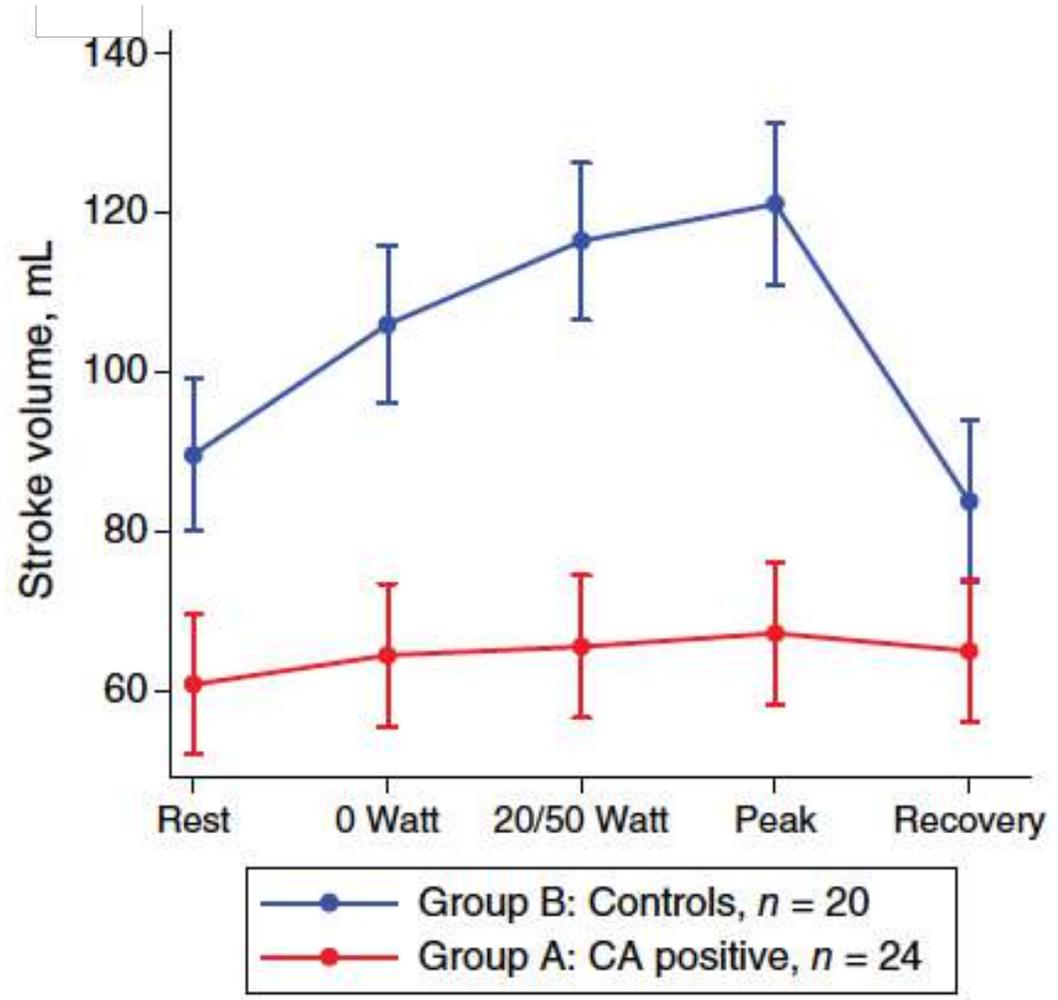
Progrès réalisés
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$$DC = VES \times FC$$

DC : débit cardiaque
VES : volume d'éjection systolique
FC : fréquence cardiaque

Amylose cardiaque

$$DC = VES \times FC$$



IEC, ARA2, ARNI & AMYLOSE CARDIAQUE

Progrès réalisés
et à venir !

$$PA = DC \times Ra = VES \times FC \times Ra$$

PA : pression artérielle

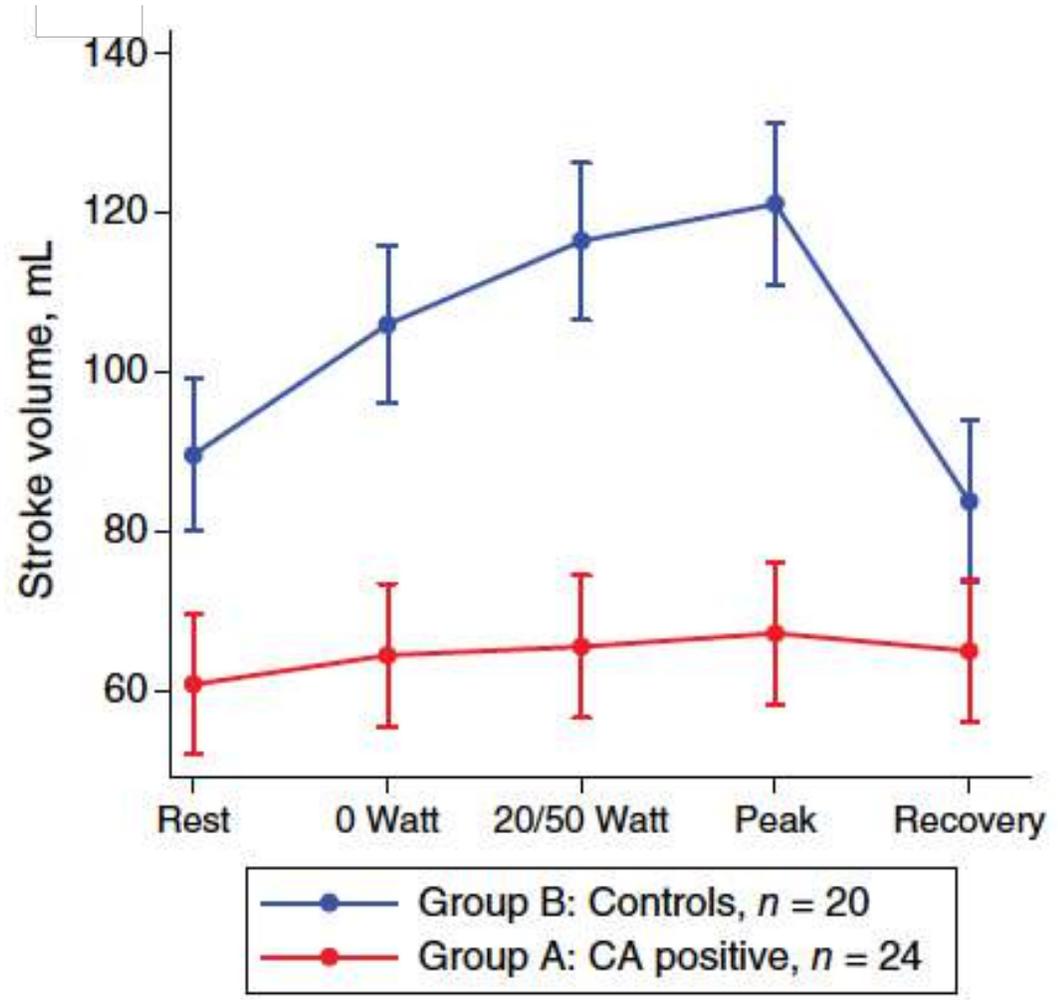
DC : débit cardiaque

VES : volume d'éjection systolique

FC : fréquence cardiaque

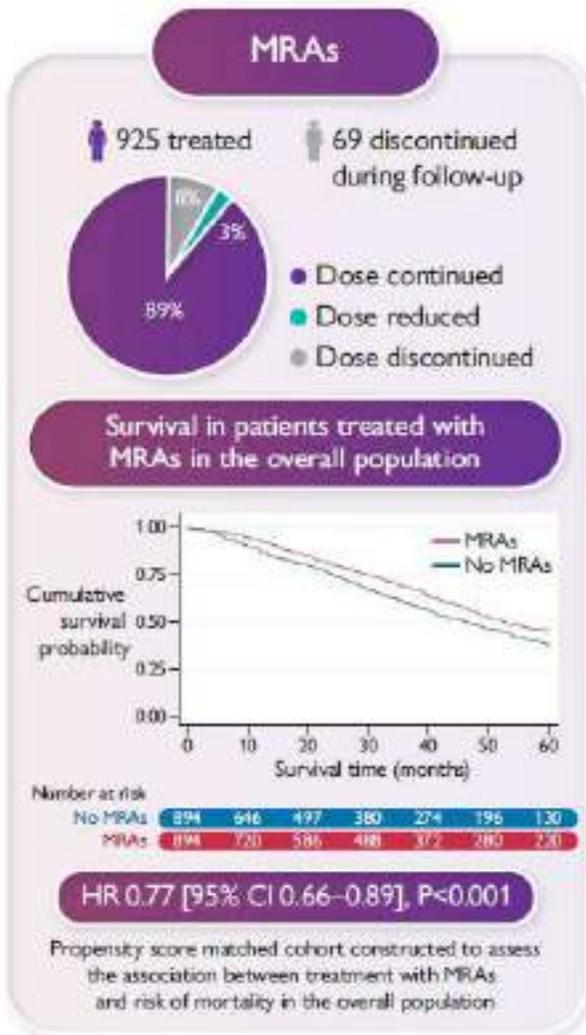
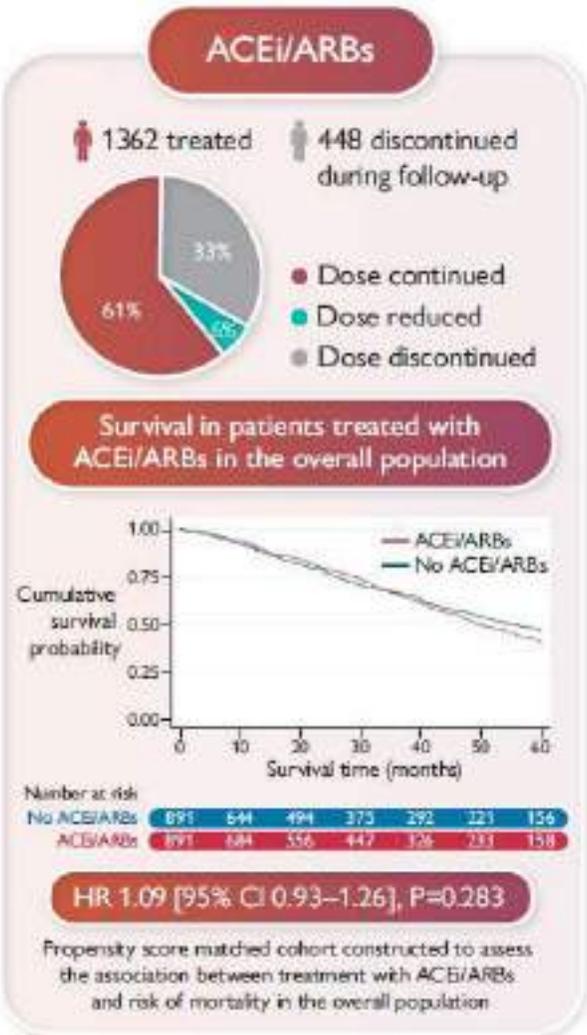
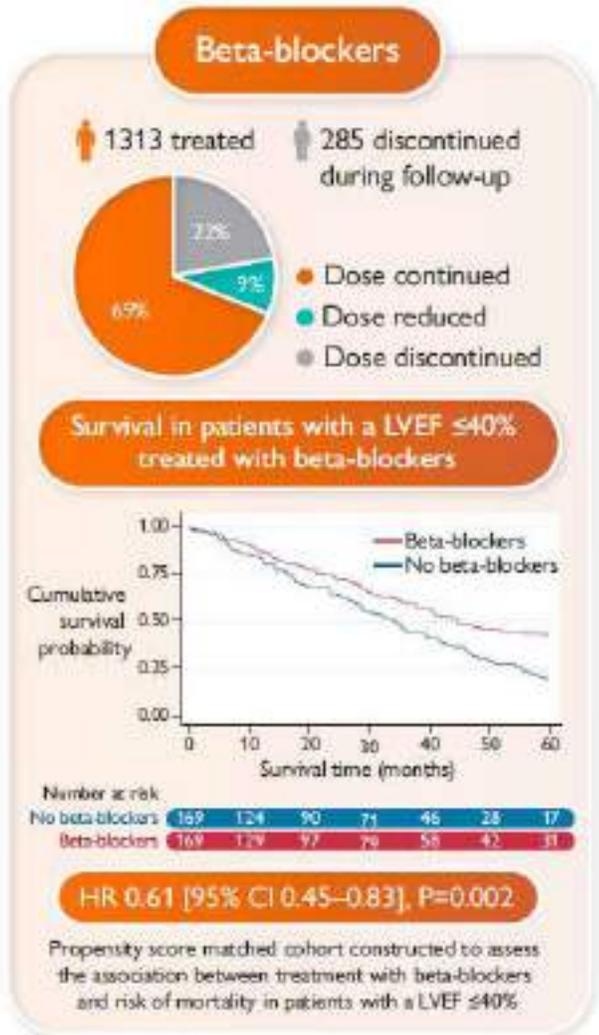
Amylose cardiaque

$$PA = VES \times FC \times Ra$$



Conventional heart failure therapy in cardiac ATTR amyloidosis

Progrès réalisés
et à venir !



BÉTABLOQUANTS & AMYLOSE CARDIAQUE

**Progrès réalisés
et à venir !**

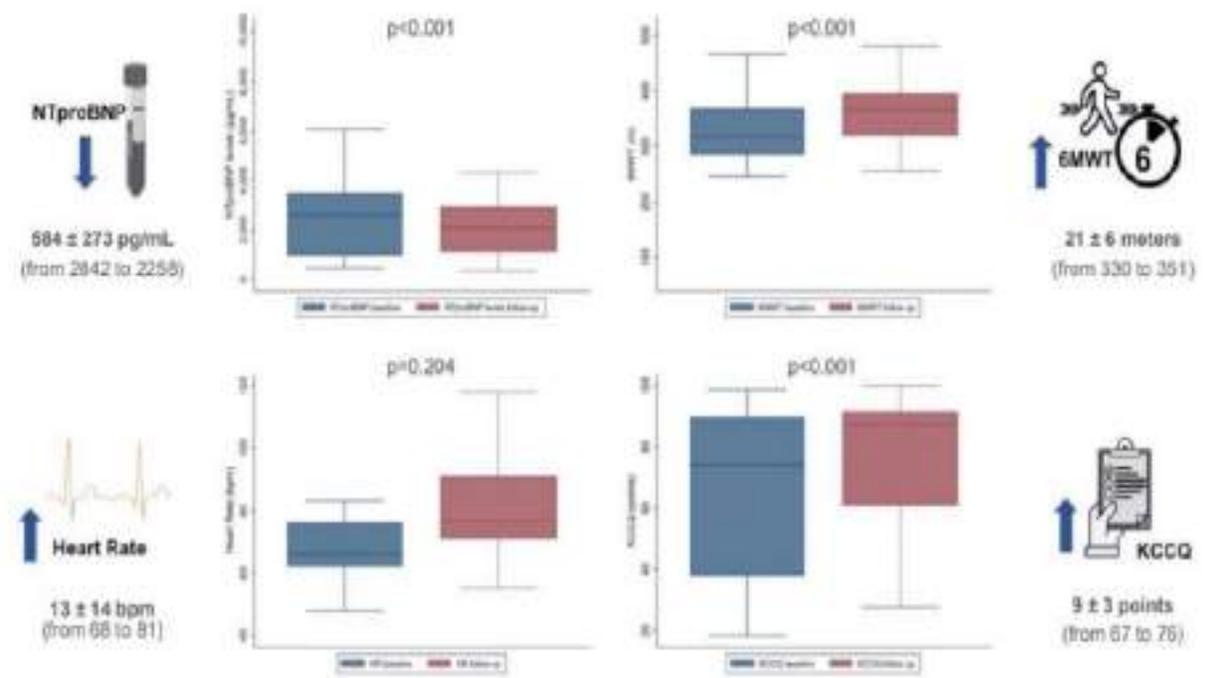
METHODS: WITHDRAWAL PROTOCOL



Discontinuation of beta-blockers in patients with ATTR-CM leads to clinical improvement at short term with increase in quality of life and functional capacity.

There is an urgent need for randomized controlled clinical trials to assess the use of HF medications in ATTR-CM.

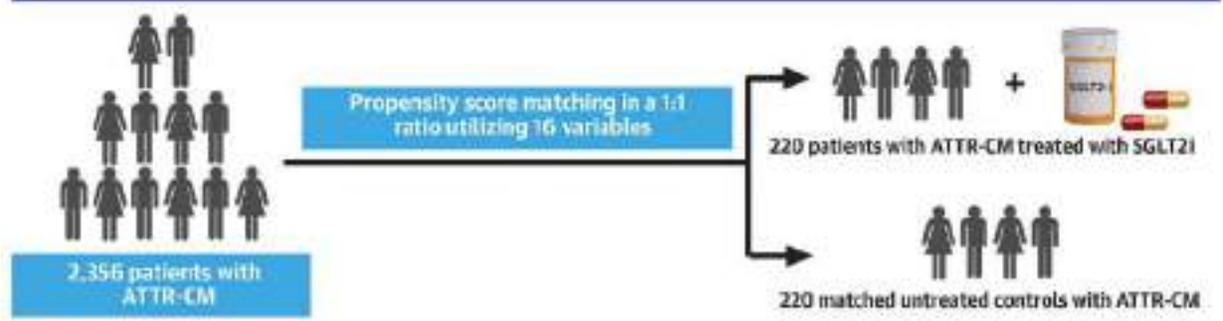
RESULTS: BETA-BLOCKERS WITHDRAWAL-PROTOCOL



SGLT2 Inhibitor Therapy in Patients With Transthyretin Amyloid Cardiomyopathy

Progrès réalisés
et à venir!

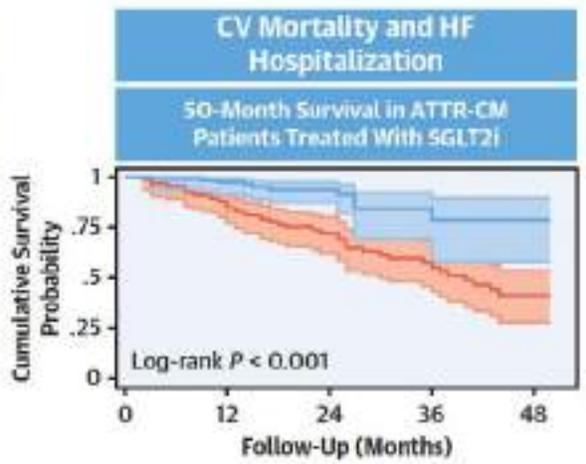
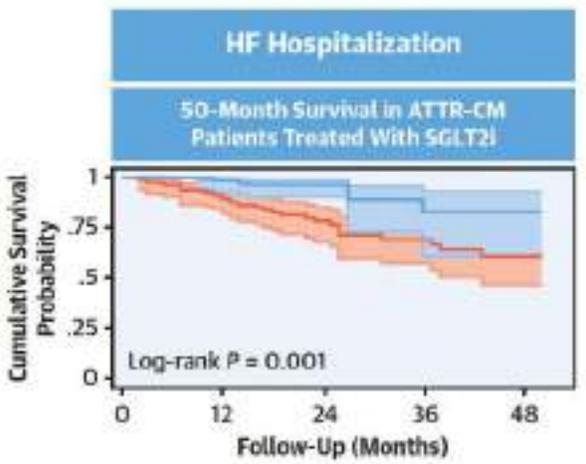
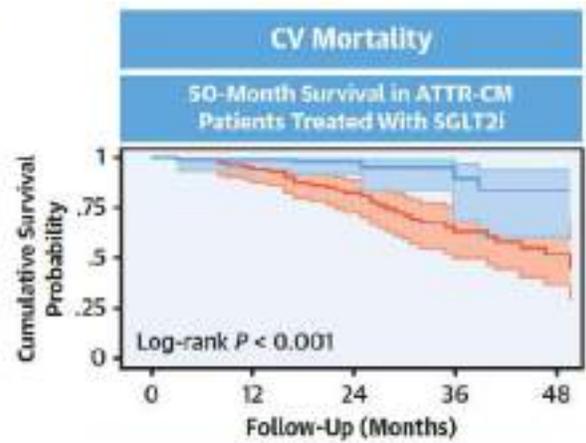
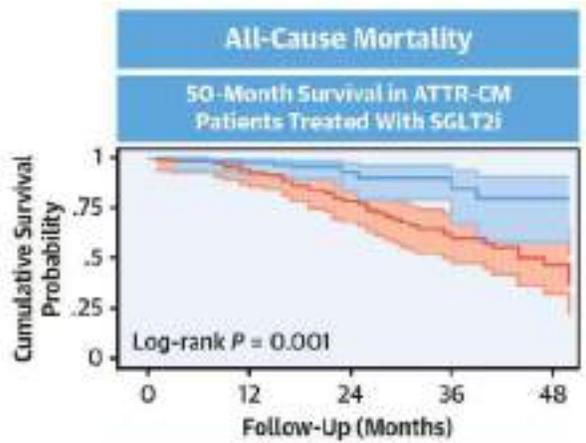
Study Design and Population



Main Findings

Safe treatment: 4.5% discontinuation rate over 28 months

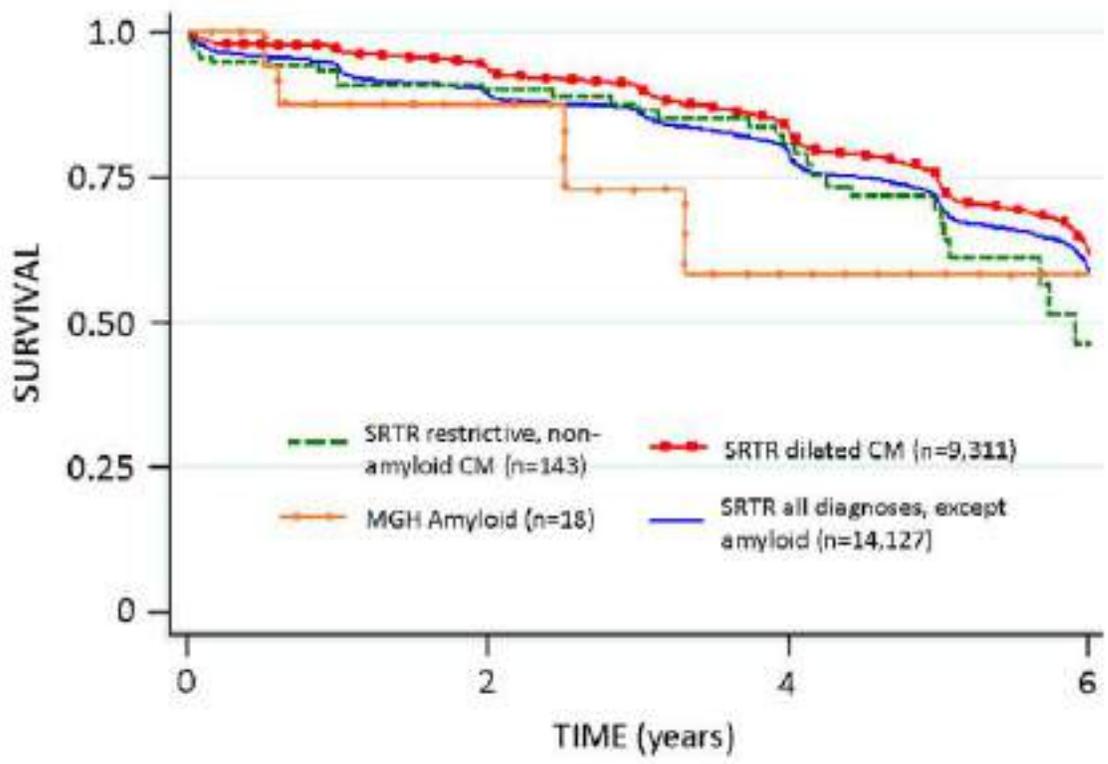
- All-cause mortality
- Cardiovascular mortality
- HF hospitalization
- Cardiovascular mortality and HF hospitalization
- Slower decline in eGFR
- Attenuated rise in NT-proBNP
- Lower loop diuretic requirement
- Stable blood pressure profile



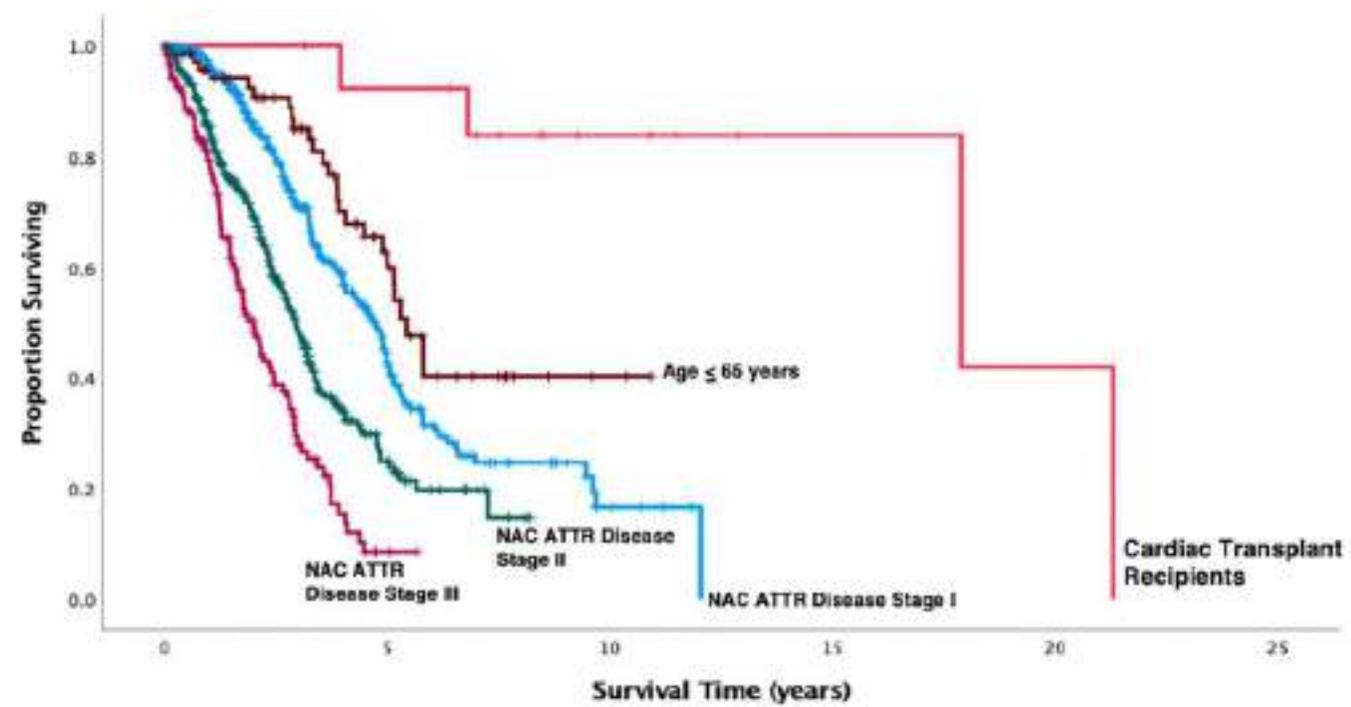
TRANSPLANTATION & AMYLOSE CARDIAQUE

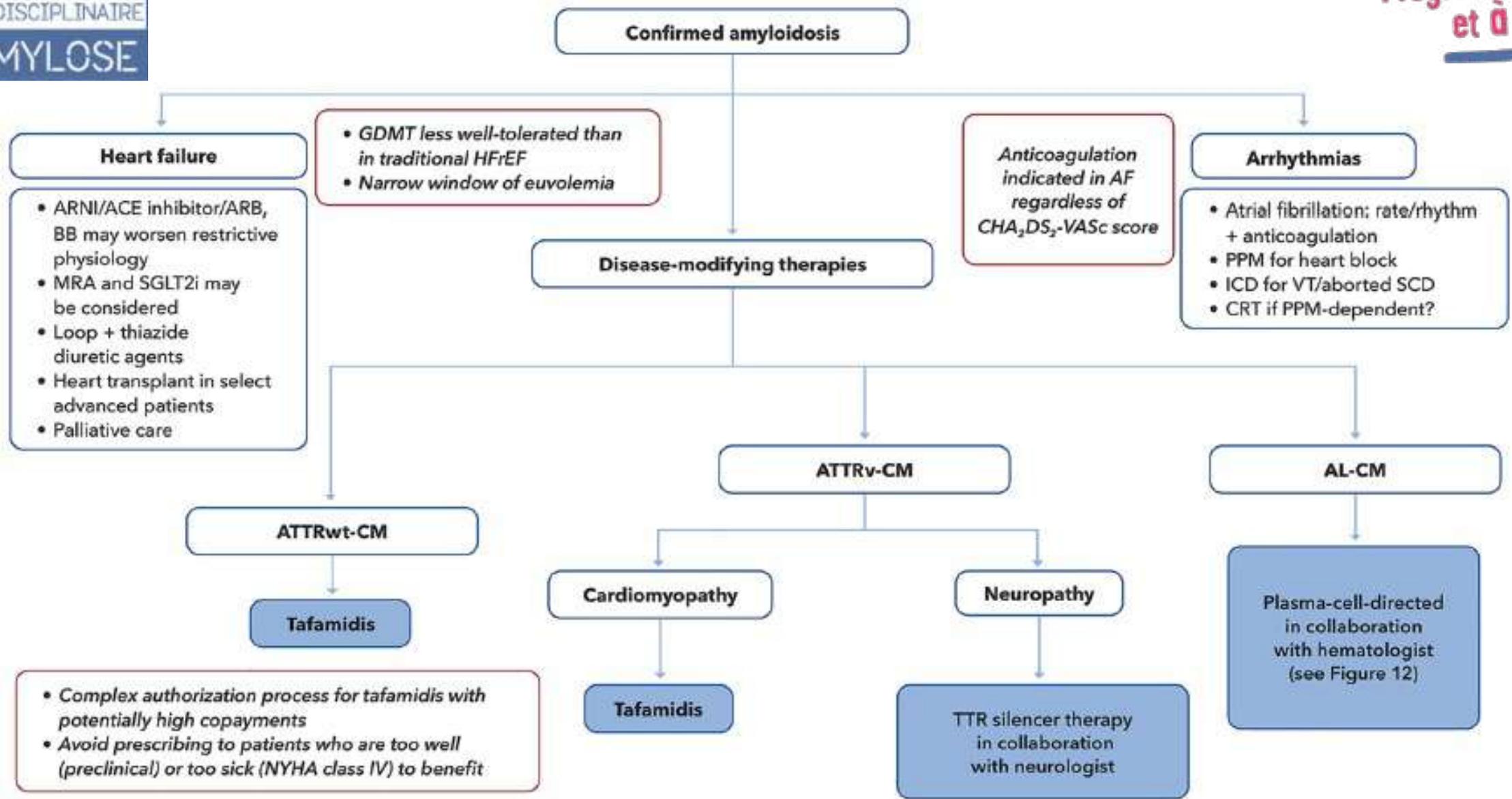
Progrès réalisés
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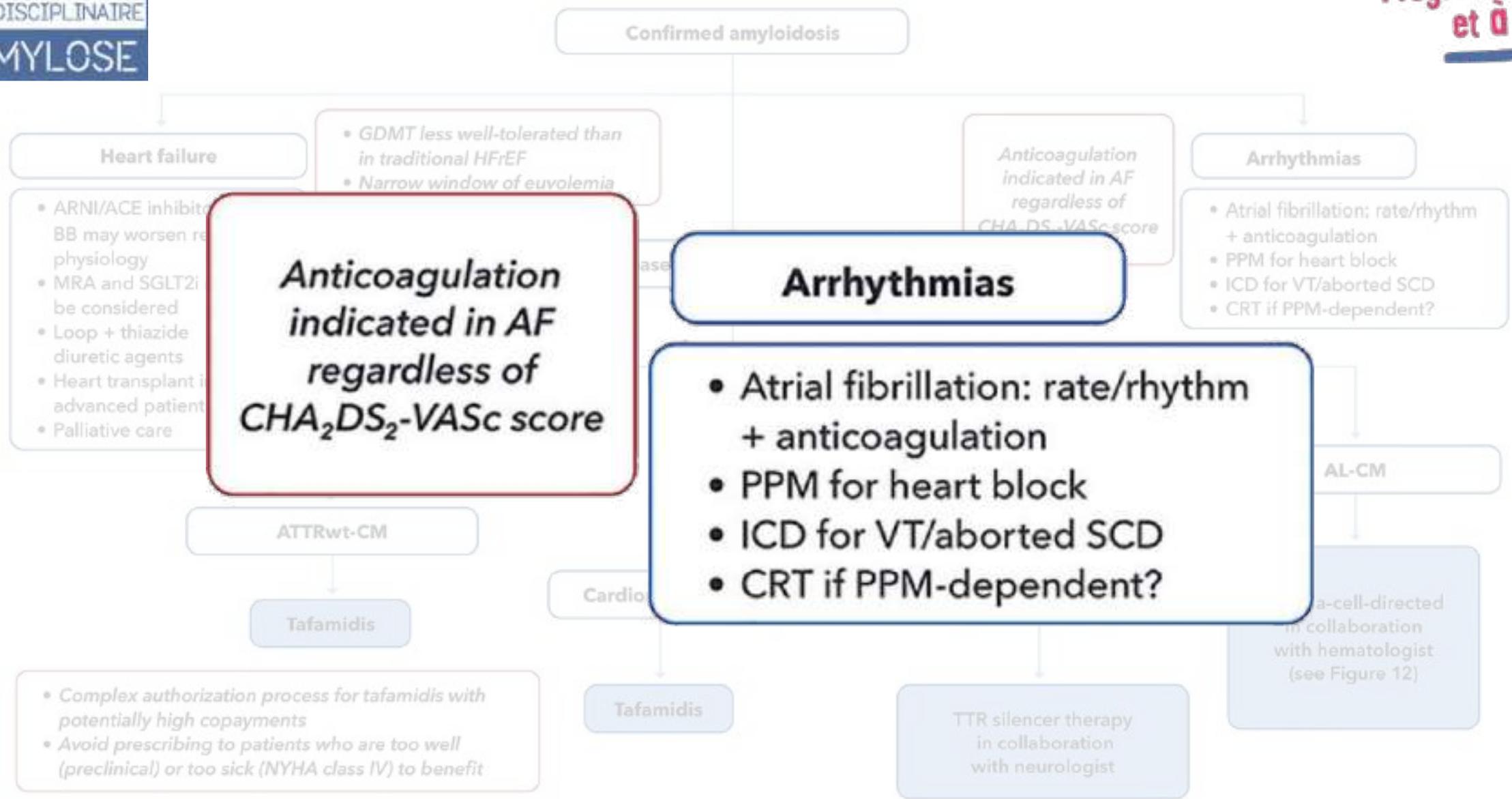
Amylose cardiaque AL



Amylose cardiaque ATTR







2020 ESC Guidelines for the diagnosis and management of atrial fibrillation

**Progrès réalisés
et à venir !**

INTEGRATED AF MANAGEMENT



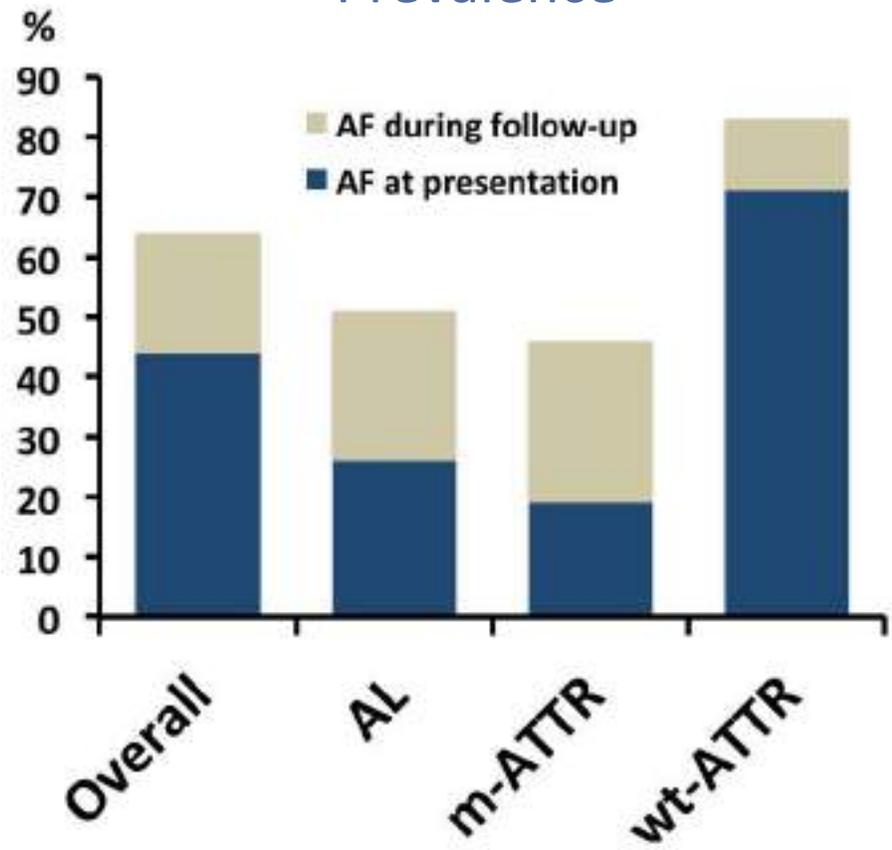
Patient-centred

Optimised stroke prevention

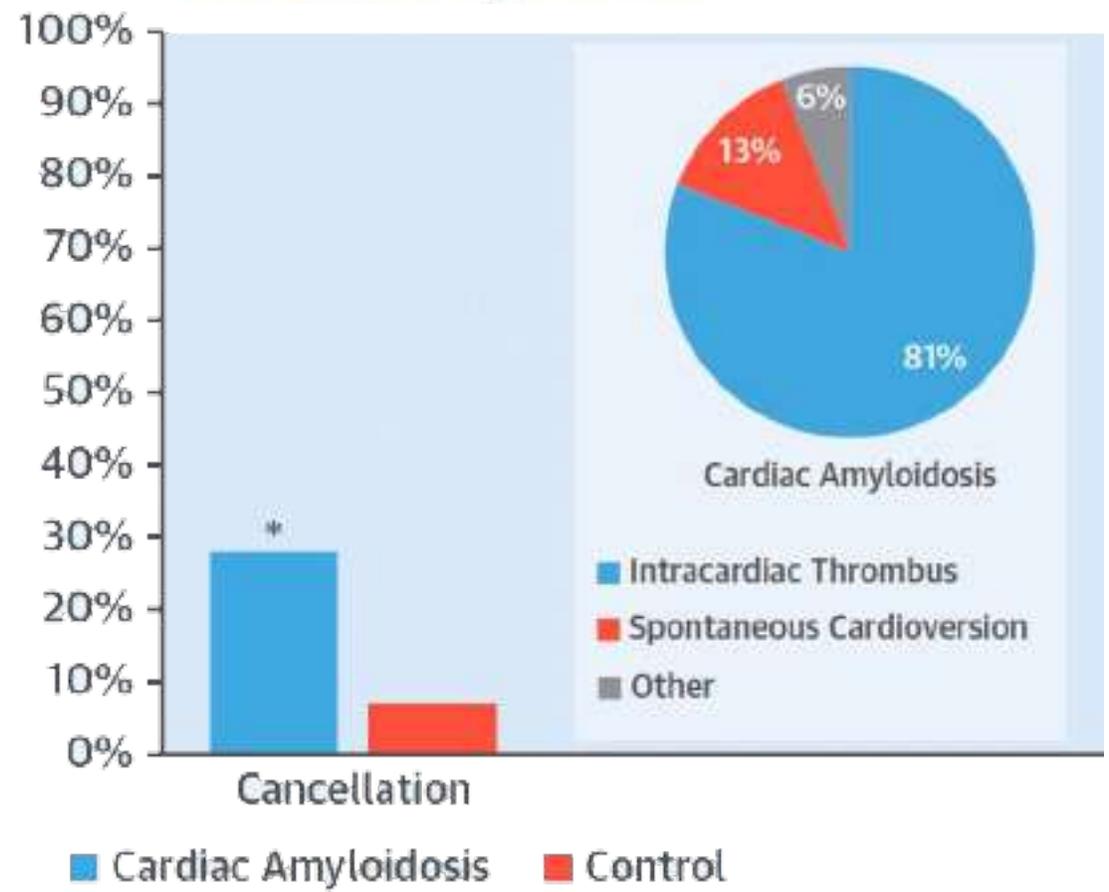
FA & AMYLOSE CARDIAQUE

Progrès réalisés
et à venir !

Prévalence

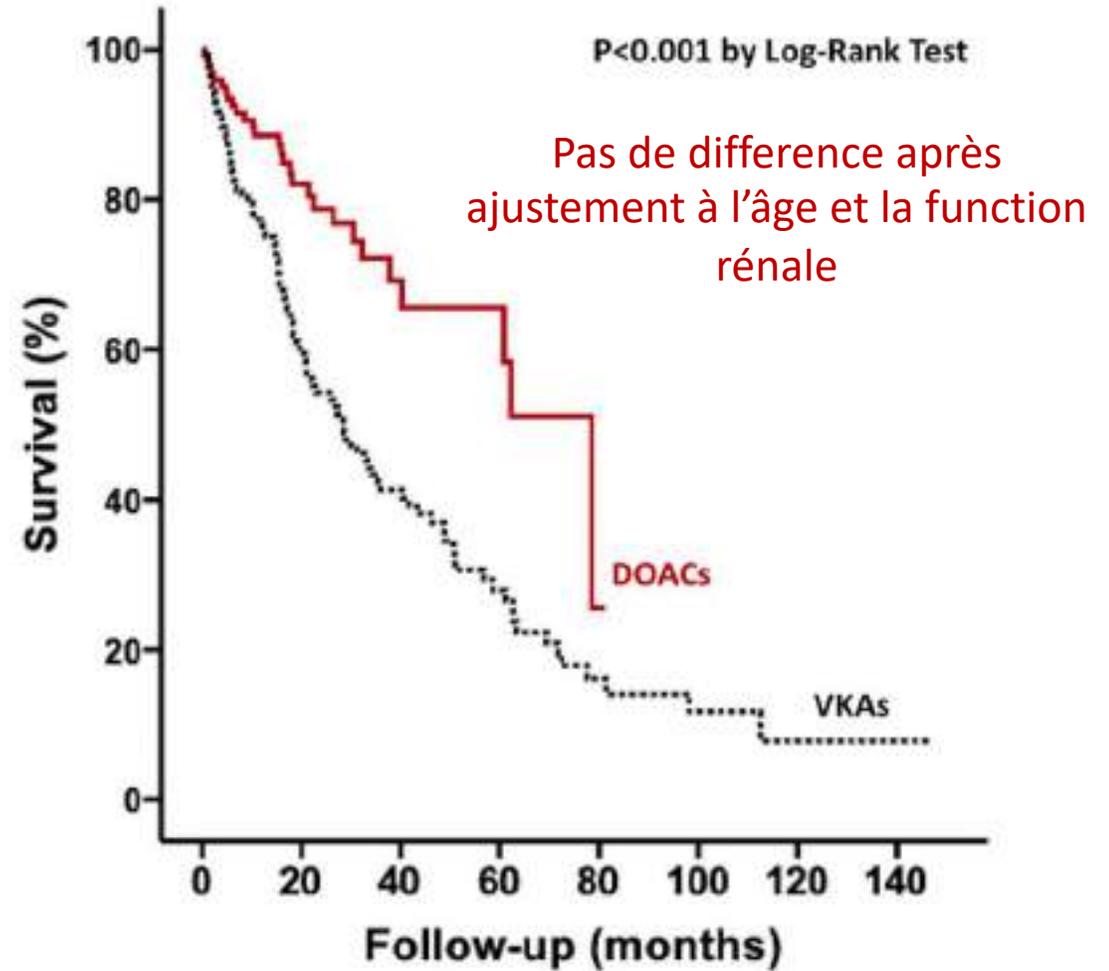
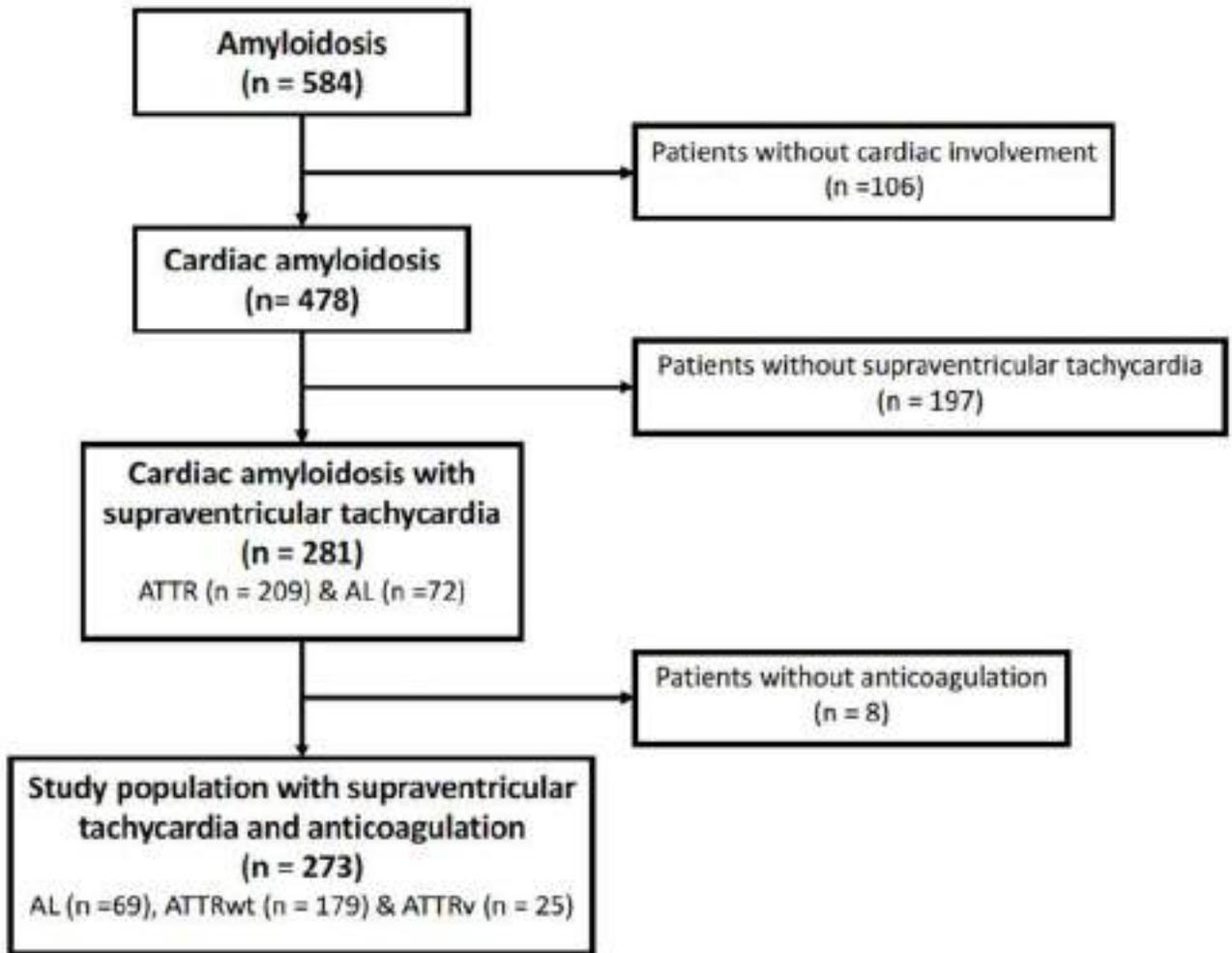


Direct Current Cardioversion of Atrial Arrhythmias in Adults With Cardiac Amyloidosis



ANTICOAGULATION & AMYLOSE CARDIAQUE

Progrès réalisés
et à venir !



2020 ESC Guidelines for the diagnosis and management of atrial fibrillation

Progrès réalisés
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INTEGRATED AF MANAGEMENT



Patient-centred

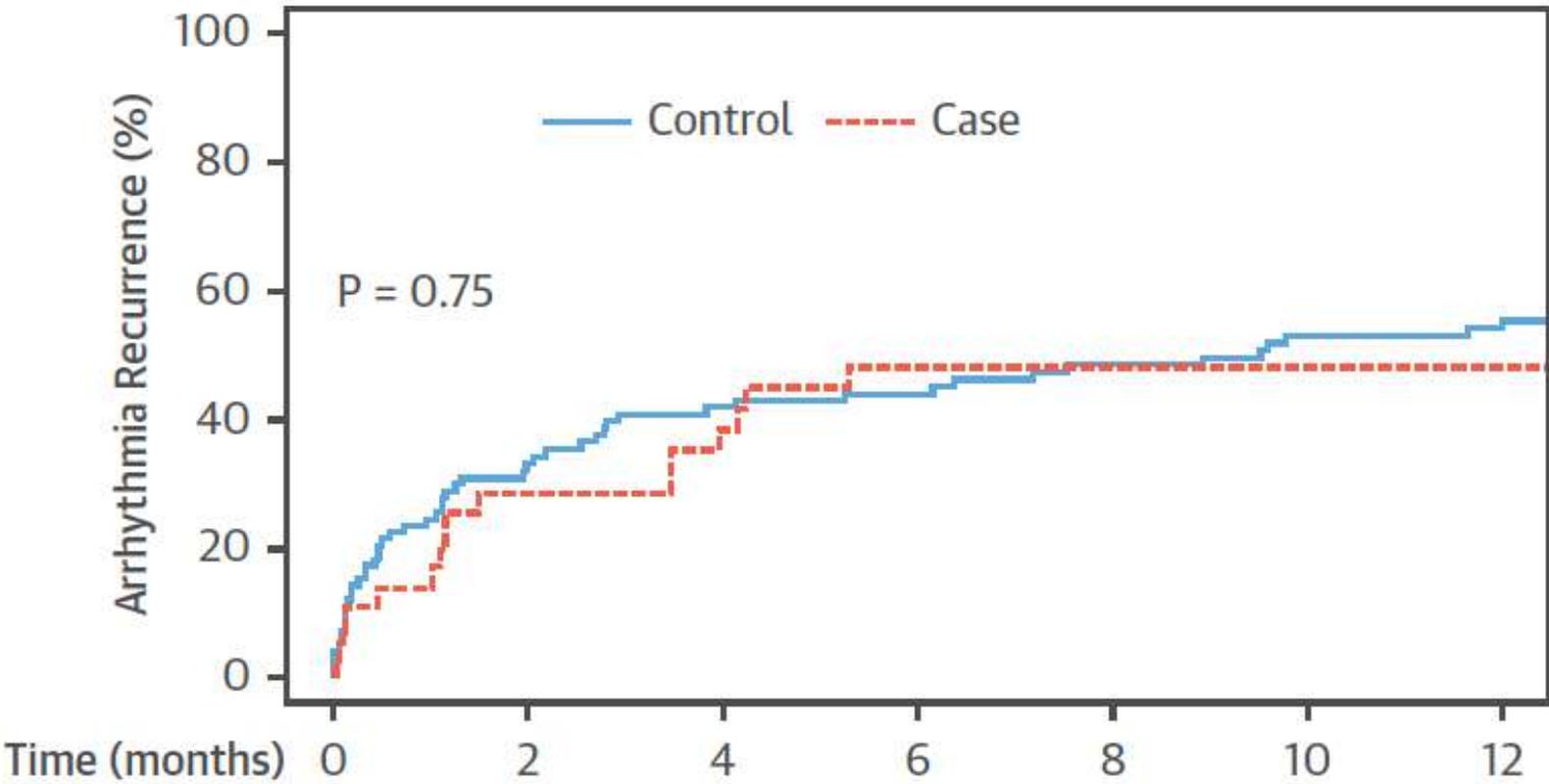
Optimised stroke prevention

Symptom control with rate or rhythm control

CARDIOVERSION & AMYLOSE CARDIAQUE

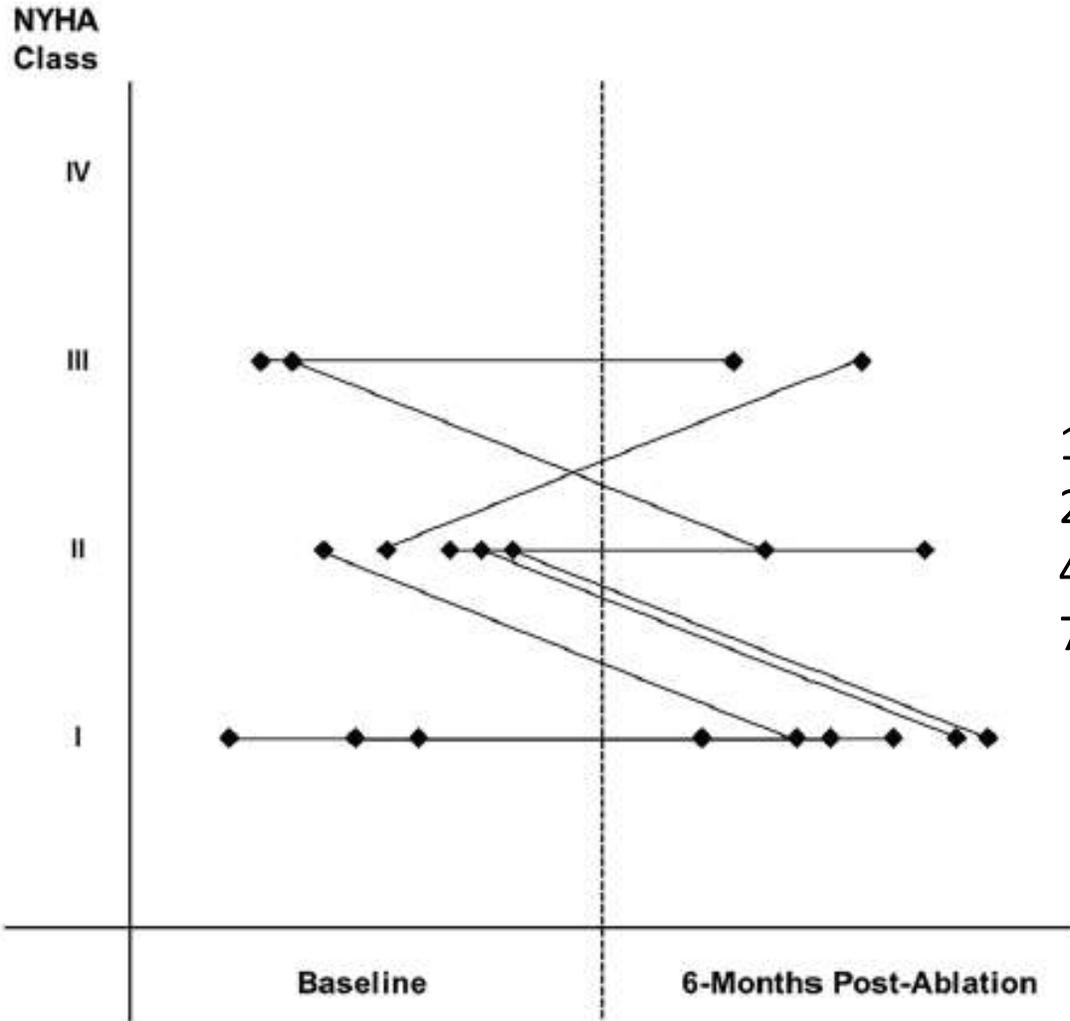
Progrès réalisés
et à venir !

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



ABLATION & AMYLOSE CARDIAQUE

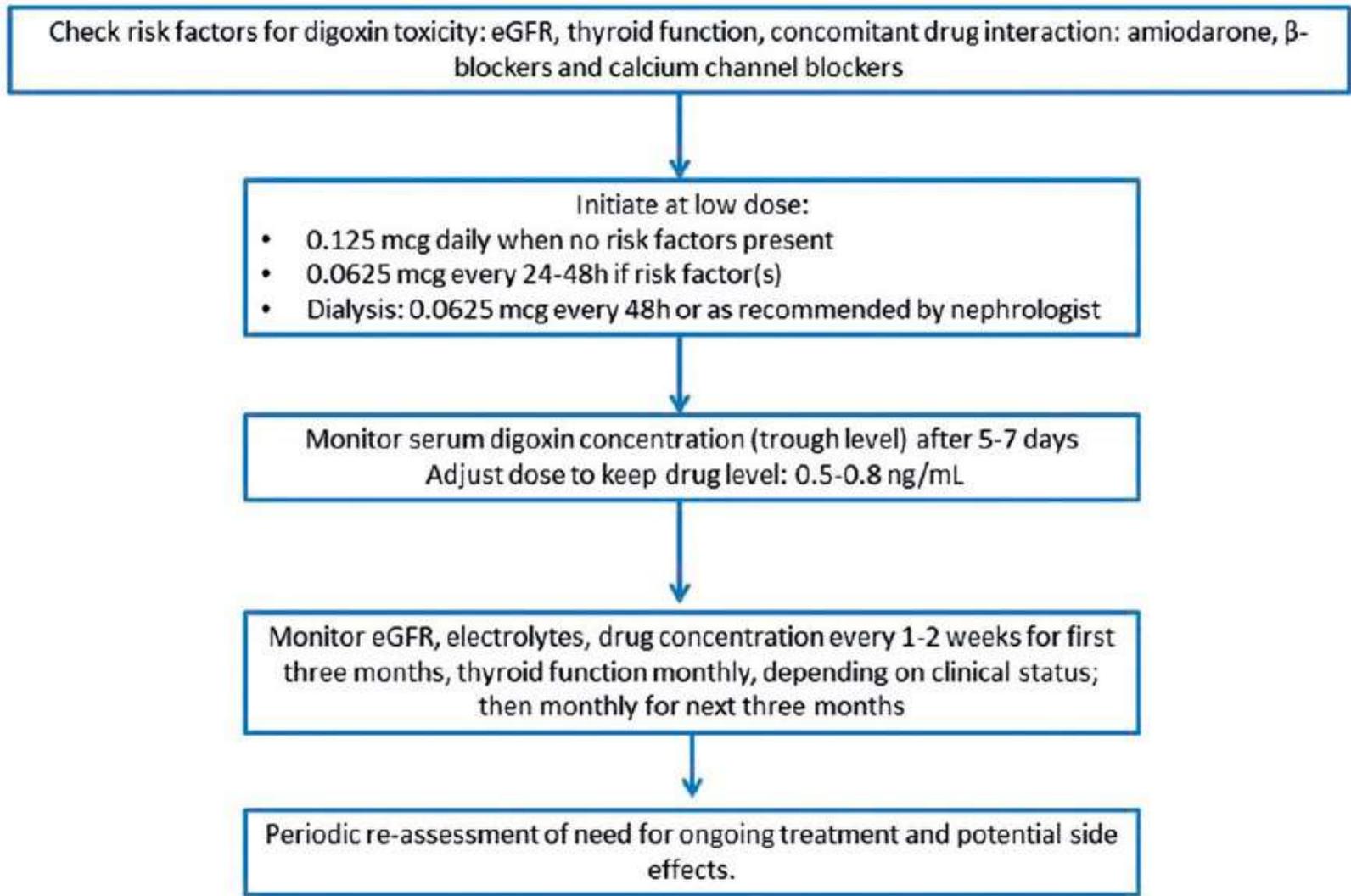
Progrès réalisés
et à venir !



13 ablations de FA
25% récidives à 1 an
40% récidives à 3 ans
70% amélioration des symptômes à 6 mois

Digoxin use in systemic light-chain (AL) amyloidosis: contra-indicated or cautious use?

Progrès réalisés
et à venir !



Diagnosis and treatment of cardiac amyloidosis: a position statement of the ESC Working Group on Myocardial and Pericardial Diseases

Progrès réalisés
et à venir !

Treatment of Cardiac Complications and Comorbidities in Cardiac Amyloidosis

Aortic Stenosis

- Severe AS confers worse prognosis.
- Concomitant ATTRwt risk factor for periprocedural AV block.
- TAVR improves outcome in amyloid-AS.

Ventricular arrhythmias

- ICD for secondary prevention.
- ICD in primary prevention usually not recommended.
- Transvenous ICD preferred over subcutaneous ICD.

Conduction disorders

- PPM according to standard indications.
- Consider CRT if high paced burden expected.

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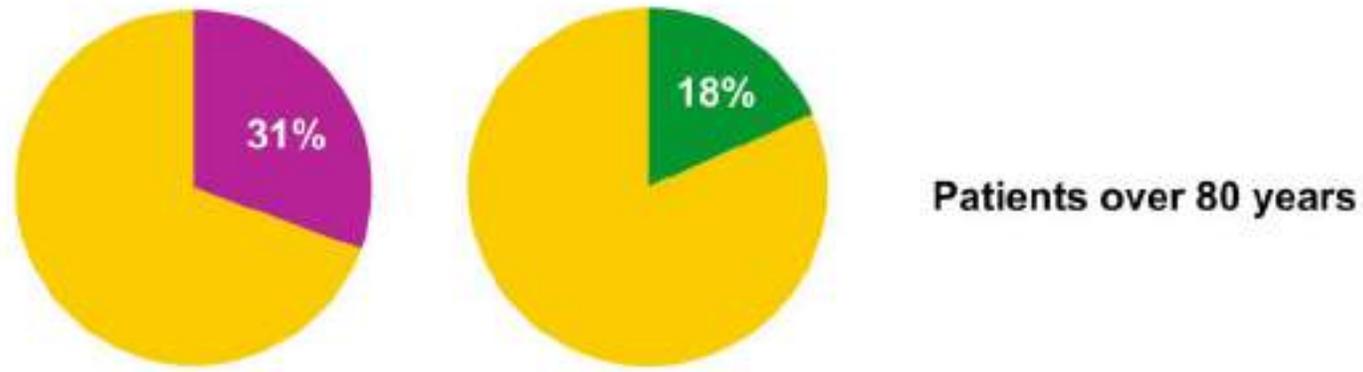
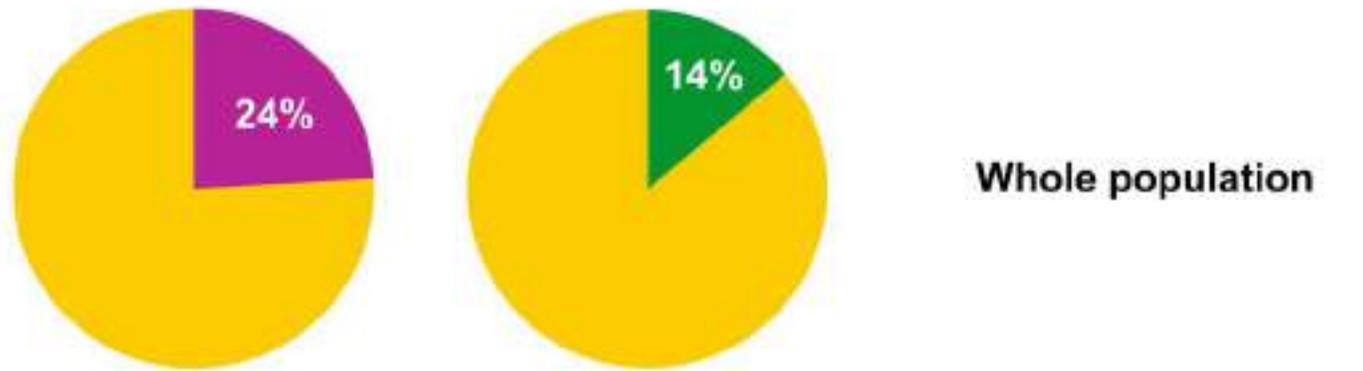
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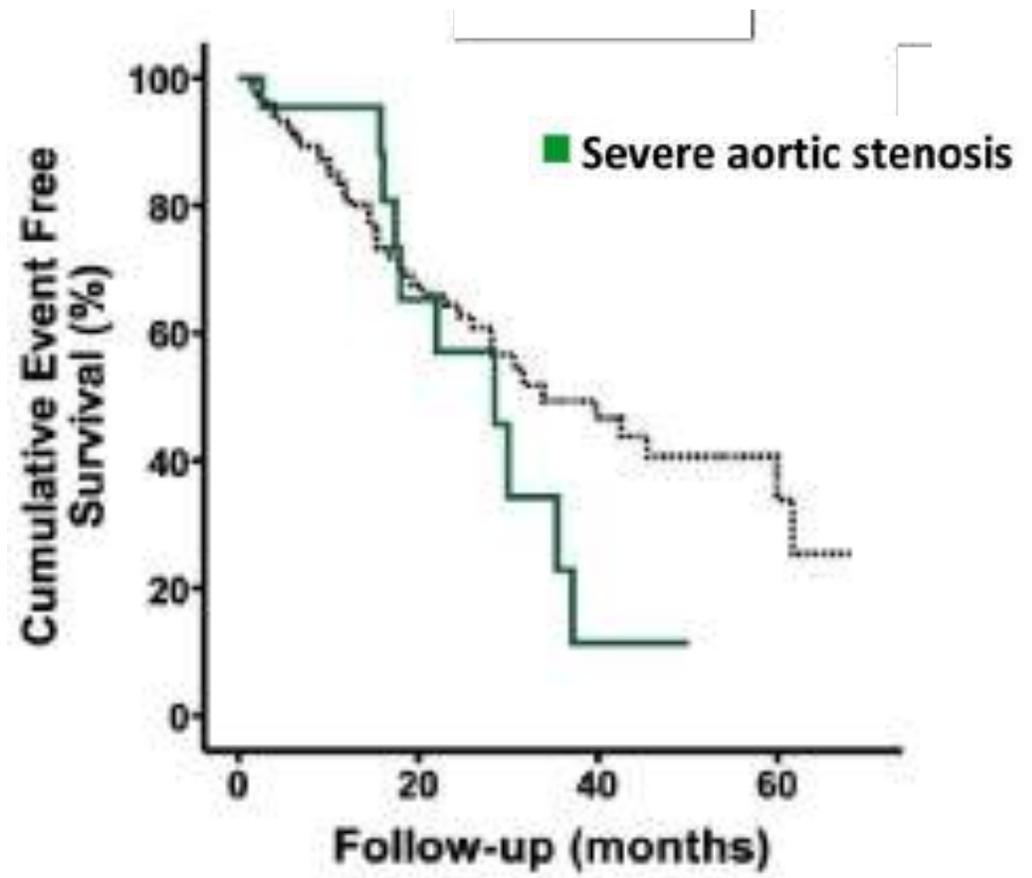
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RAO & AMYLOSE CARDIAQUE (ATTRWT)

Progrès réalisés
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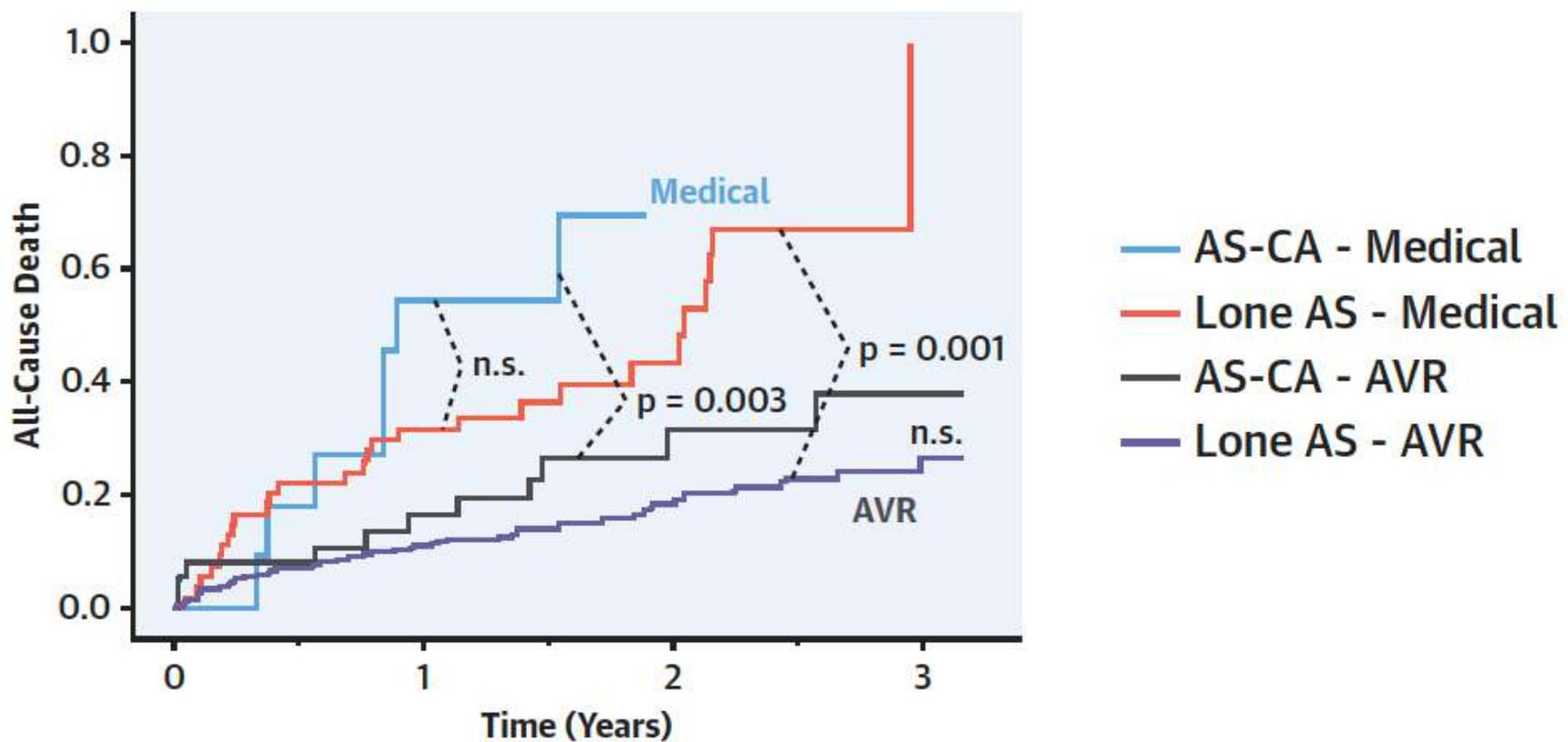
- Aortic stenosis
- Severe aortic stenosis
- No aortic stenosis



Prevalence and Outcomes of Concomitant Aortic Stenosis and Cardiac Amyloidosis

Progrès réalisés
et à venir !

All-Cause Mortality in Lone AS Versus AS-CA Following Aortic Valve Replacement or With Medical Therapy



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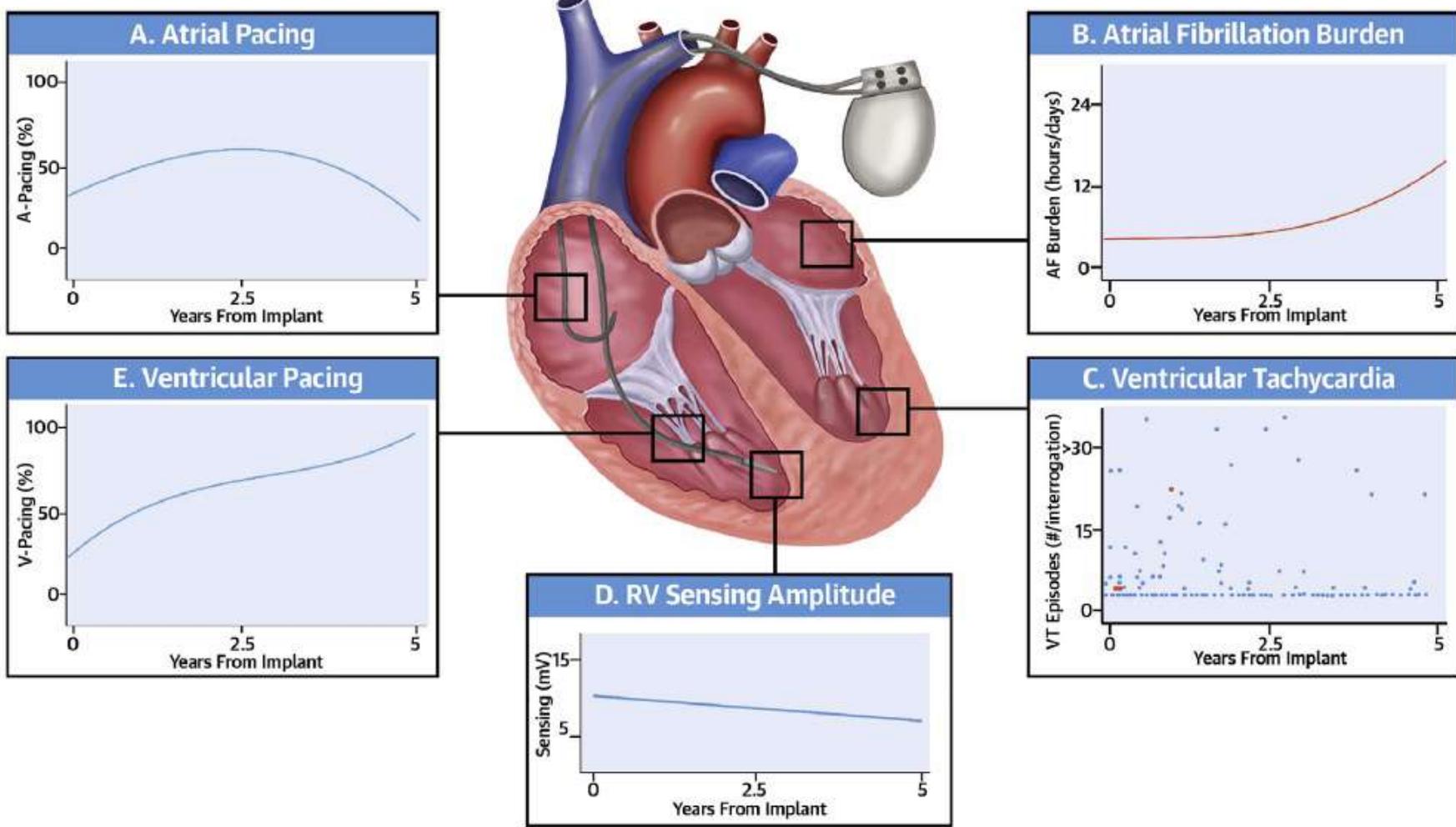
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Cardiac Implantable Electronic Devices

A Window Into the Evolution of Conduction Disease in Cardiac Amyloidosis

Progrès réalisés
et à venir !



HYPOTENSION ORTHOSTATIQUE & AMYLOSE CARDIAQUE

Progrès réalisés
et à venir !



Traitement de l'hypotension orthostatique
Mesures relatives au mode de vie - Repas légers et plus fréquents - Réduire la consommation d'alcool
Réadaptation à l'orthostatisme
Bas de contention
Midodrine
Fludrocortisone - Rétention hydro-sodée

LA PRISE EN CHARGE CARDIOLOGIQUE DES AMYLOSES NON SPÉCIFIQUES

Progrès réalisés
et à venir !



Equilibrer la **volémie**

Traiter l'**insuffisance cardiaque**

- Antagonistes des récepteurs aux minéralocorticoïdes
- Inhibiteurs de SGLT2
- Transplantation le cas échéant

Dépister et prendre en charge les **arythmies supraventriculaires**

- Anticoagulation
- Contrôle du rythme ou de la fréquence

Anticiper les **troubles du rythme ventriculaire ou de la conduction**

Prendre en charge les rétrécissements aortiques serrés

Traiter les **hypotensions orthostatiques**