

# 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

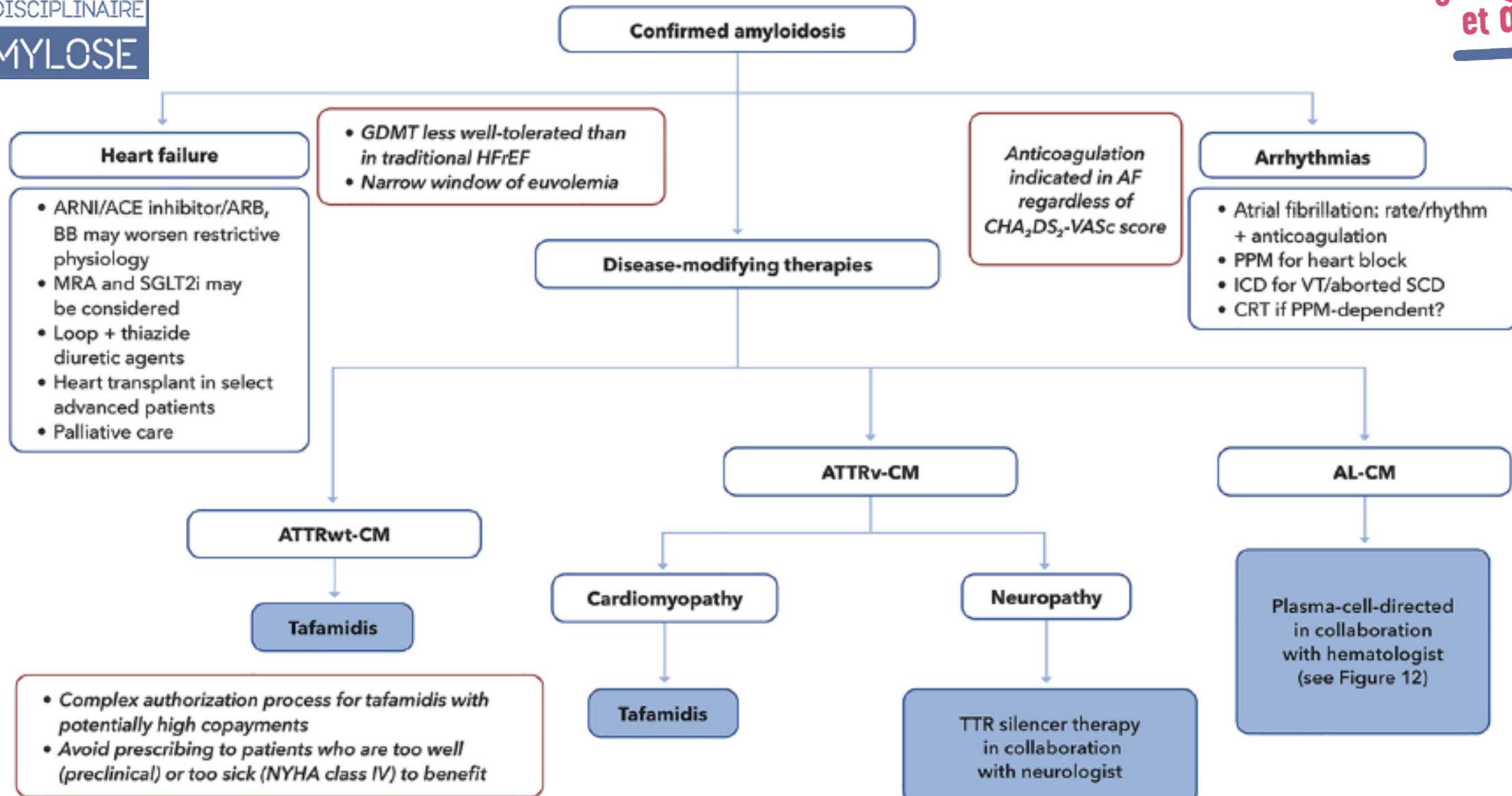
## 2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure

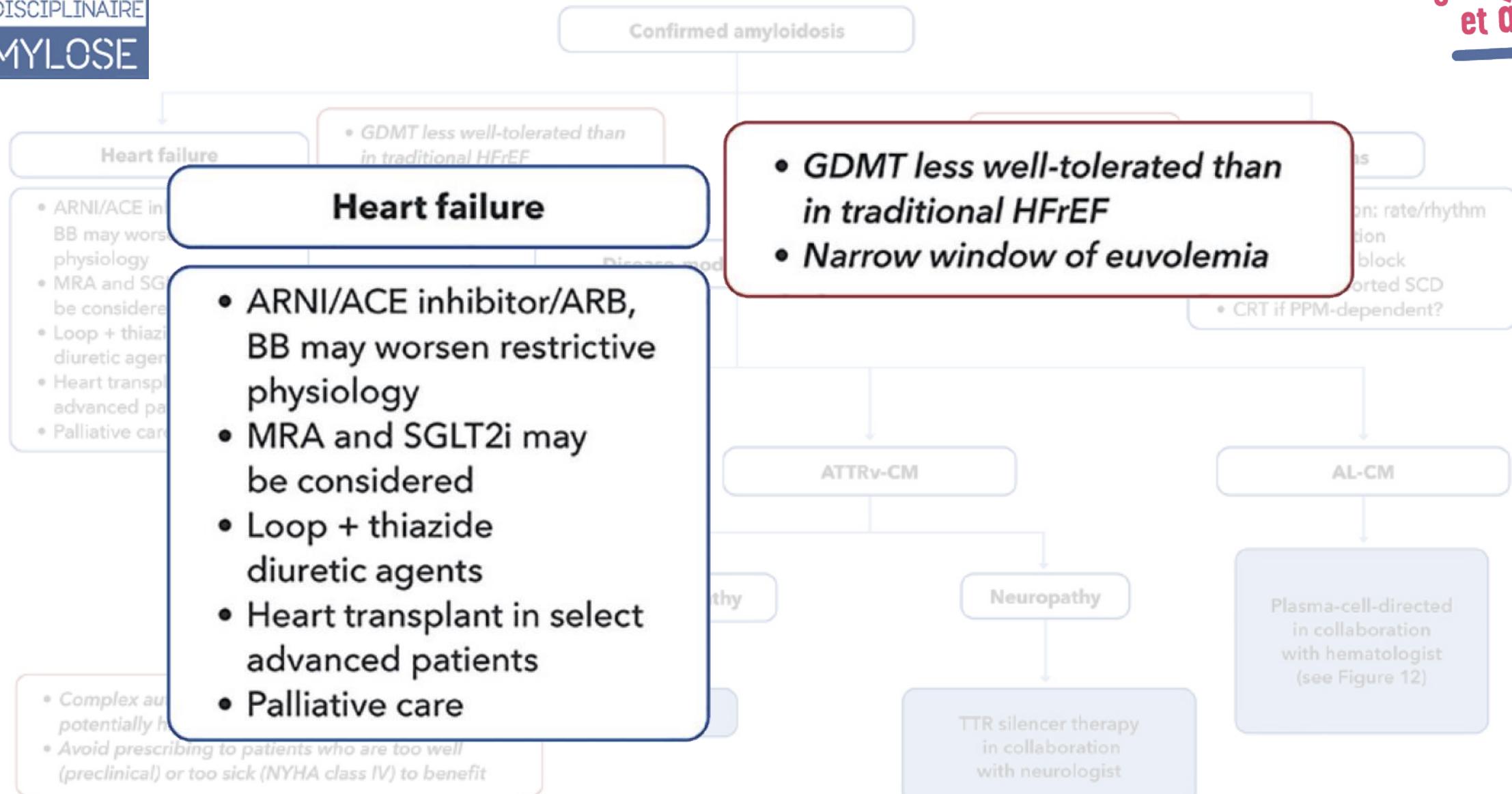
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et à venir !

Type of HF	HFrEF	HFmrEF	HFpEF
CRITERIA			
1	Symptoms ± Signs <sup>a</sup>	Symptoms ± Signs <sup>a</sup>	Symptoms ± Signs <sup>a</sup>
2	LVEF $\leq$ 40%	LVEF 41–49% <sup>b</sup>	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 <sup>c</sup>

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

Type	Type of HF	
CRITERIA		
1	HFpEF	Symptoms ± Signs <sup>a</sup>
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 <sup>c</sup>





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

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## Management of patients with HFpEF

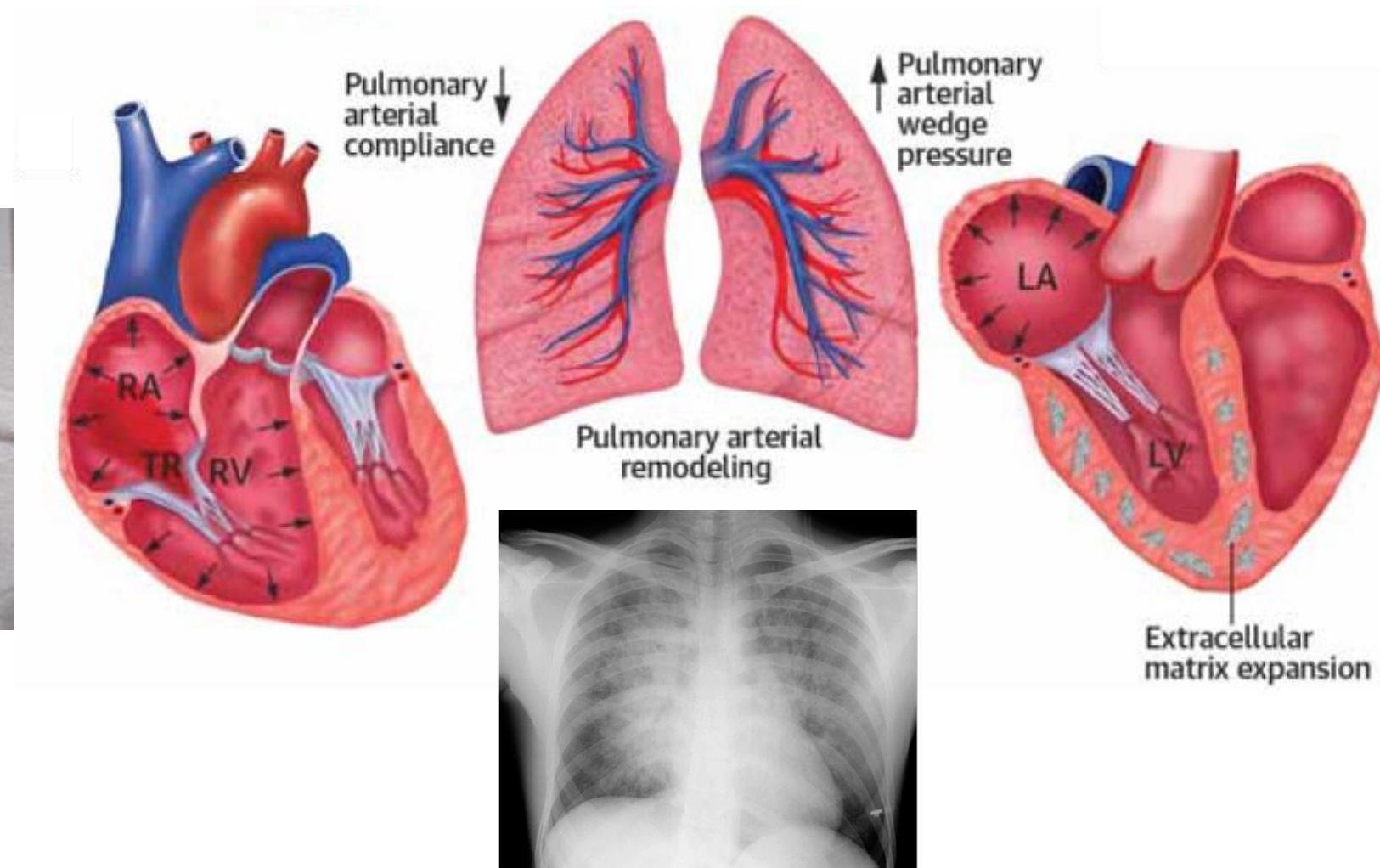
Diuretics for  
fluid retention  
(Class I)

Dapagliflozin/  
Empagliflozin  
(Class I)

Treatment for aetiology,  
CV and non-CV comorbidities  
(Class I)

# PRISE EN CHARGE DES SYMPTÔMES : GESTION DE LA VOLÉMIE

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# BÉTABLOQUANTS & AMYLOSE CARDIAQUE

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$$DC = VES \times FC$$

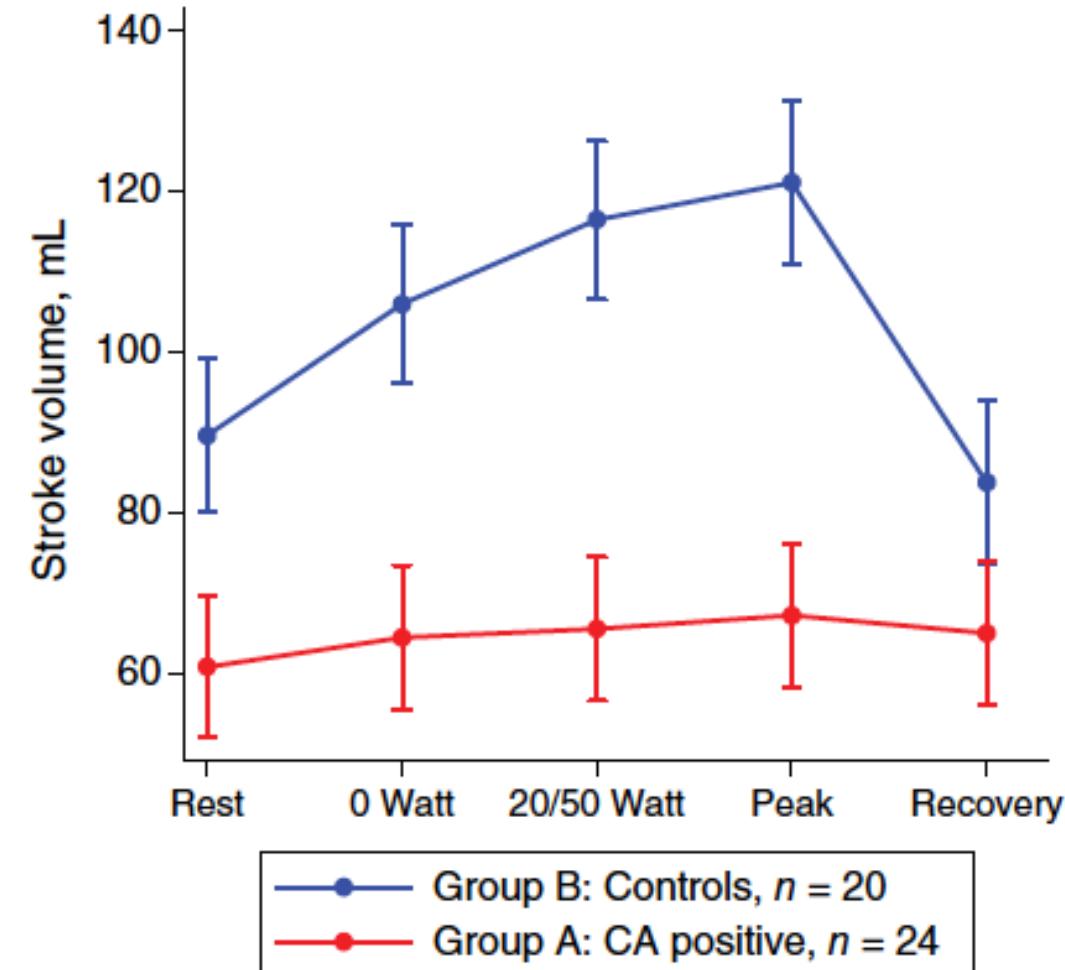
DC : débit cardiaque

VES : volume d'éjection systolique

FC : fréquence cardiaque

Amylose cardiaque

$$DC = VES \times FC$$



# IEC, ARA2, ARNI & AMYLOSE CARDIAQUE

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

PA : pression artérielle

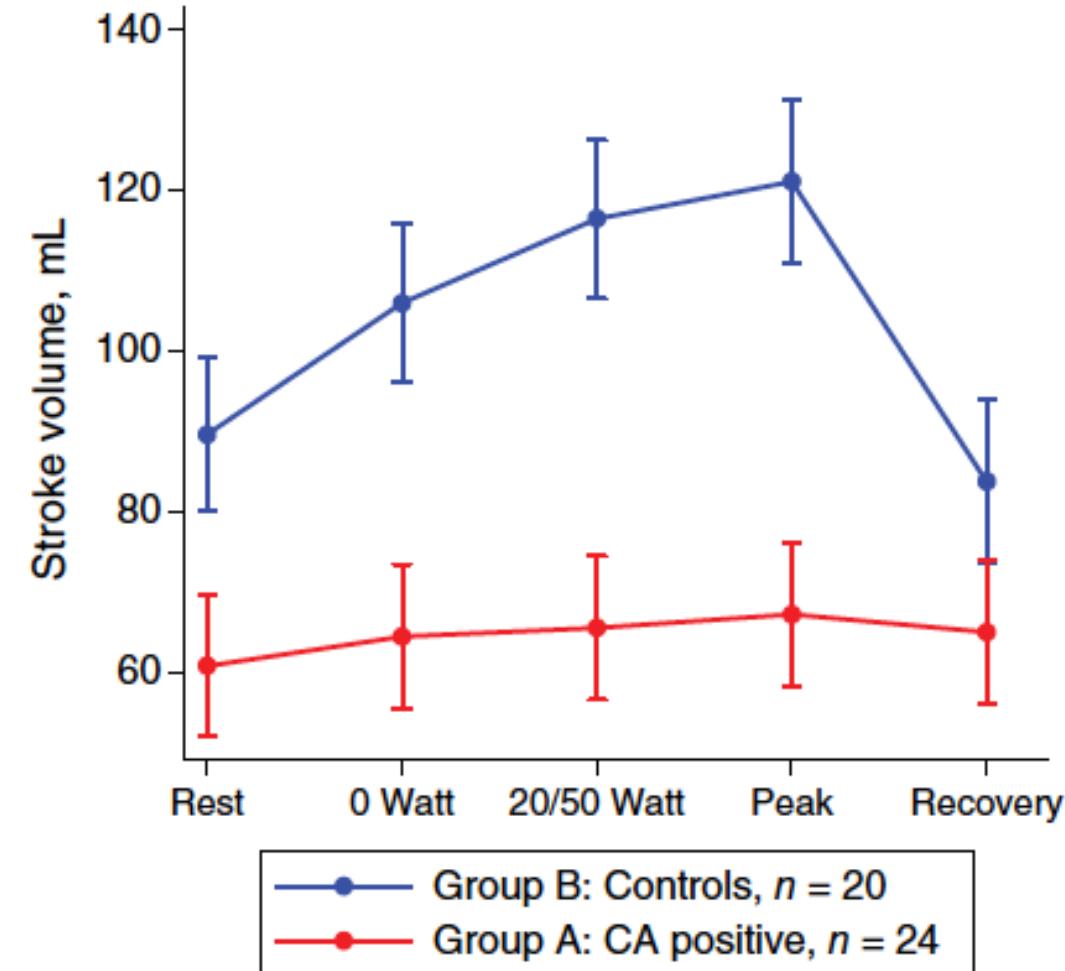
DC : débit cardiaque

VES : volume d'éjection systolique

FC : fréquence cardiaque

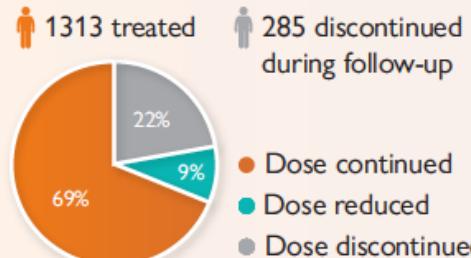
Amylose cardiaque

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

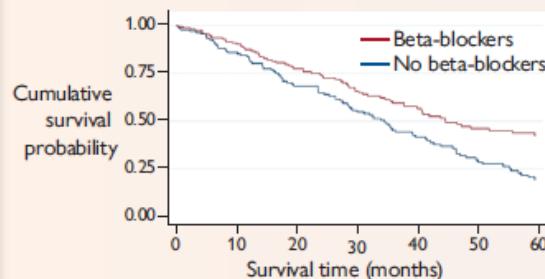


# Conventional heart failure therapy in cardiac ATTR amyloidosis

## Beta-blockers



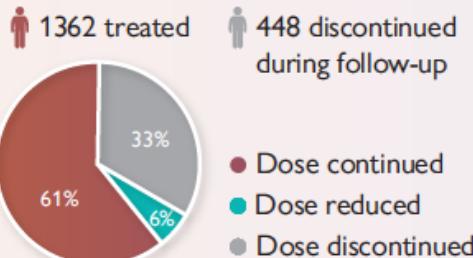
Survival in patients with a LVEF ≤40% treated with beta-blockers



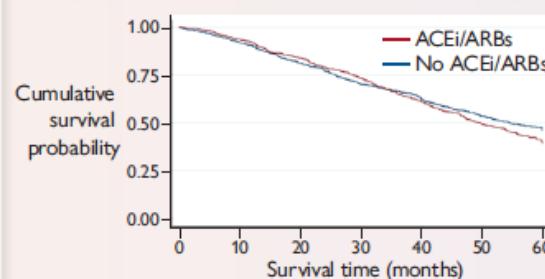
HR 0.61 [95% CI 0.45–0.83], P=0.002

Propensity score matched cohort constructed to assess the association between treatment with beta-blockers and risk of mortality in patients with a LVEF ≤40%

## ACEi/ARBs



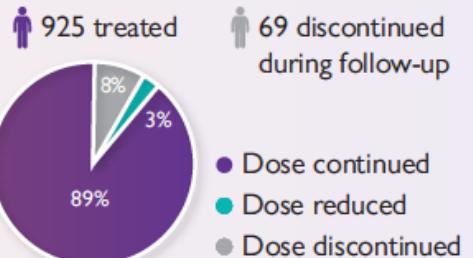
Survival in patients treated with ACEi/ARBs in the overall population



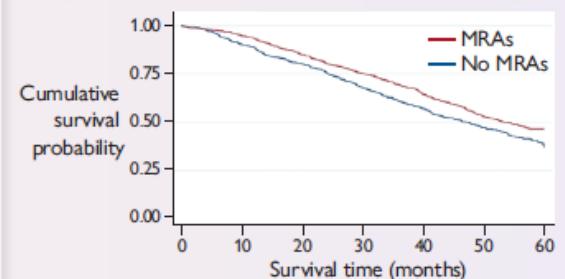
HR 1.09 [95% CI 0.93–1.26], P=0.283

Propensity score matched cohort constructed to assess the association between treatment with ACEi/ARBs and risk of mortality in the overall population

## MRAs



Survival in patients treated with MRAs in the overall population

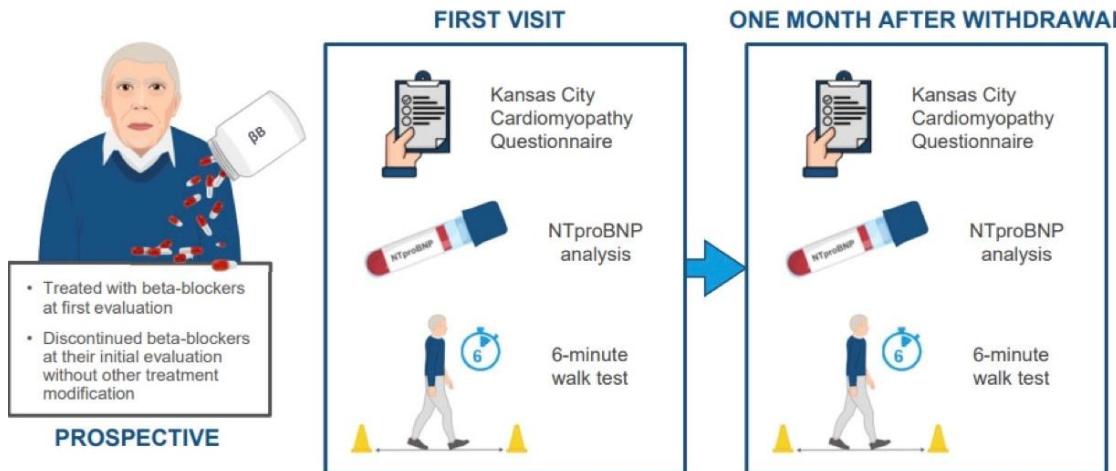


HR 0.77 [95% CI 0.66–0.89], P<0.001

Propensity score matched cohort constructed to assess the association between treatment with MRAs and risk of mortality in the overall population

# BÉTABLOQUANTS & AMYLOSE CARDIAQUE

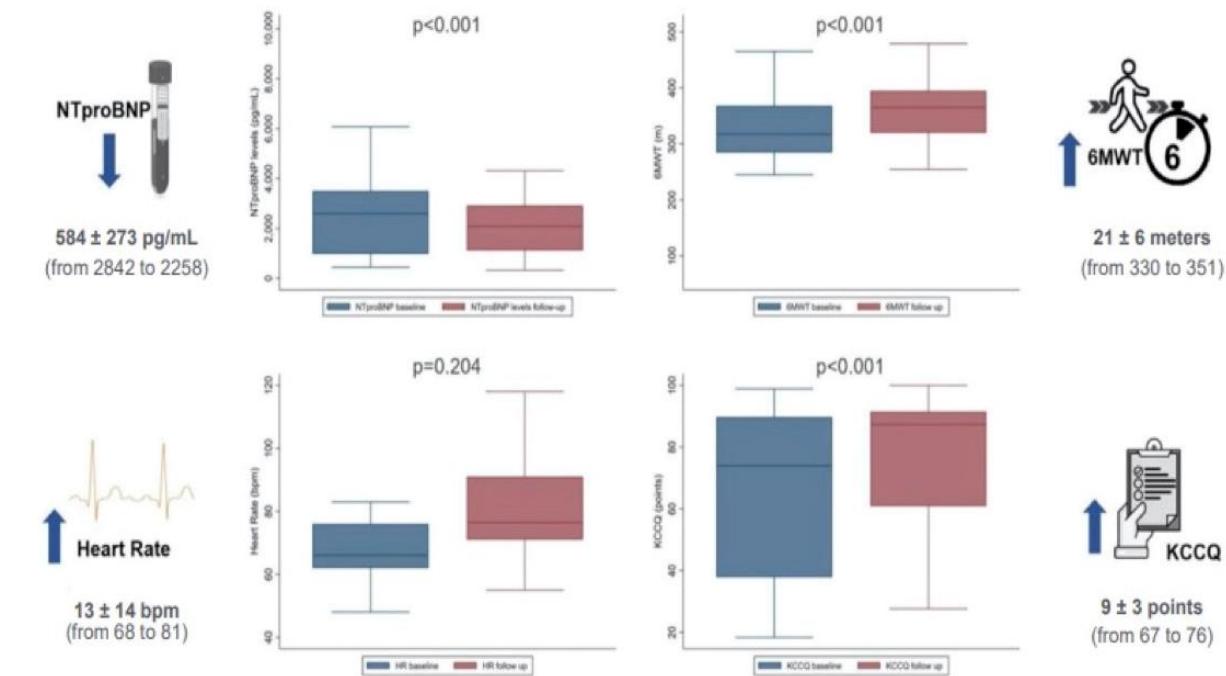
## METHODS: WITHDRAWAL PROTOCOL



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.

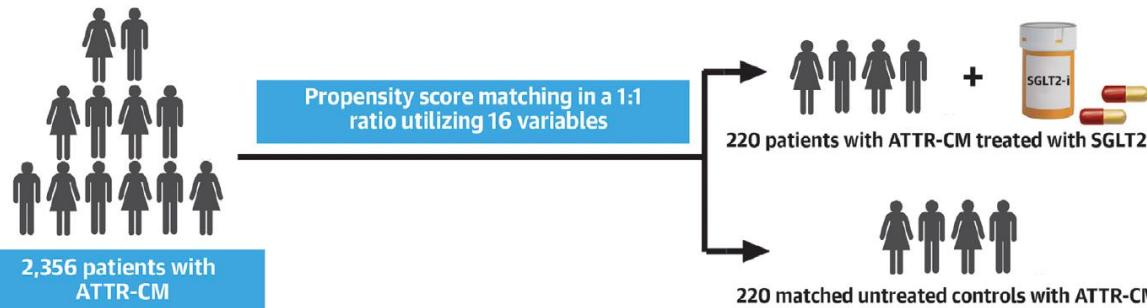
## RESULTS: BETA-BLOCKERS WITHDRAWAL-PROTOCOL



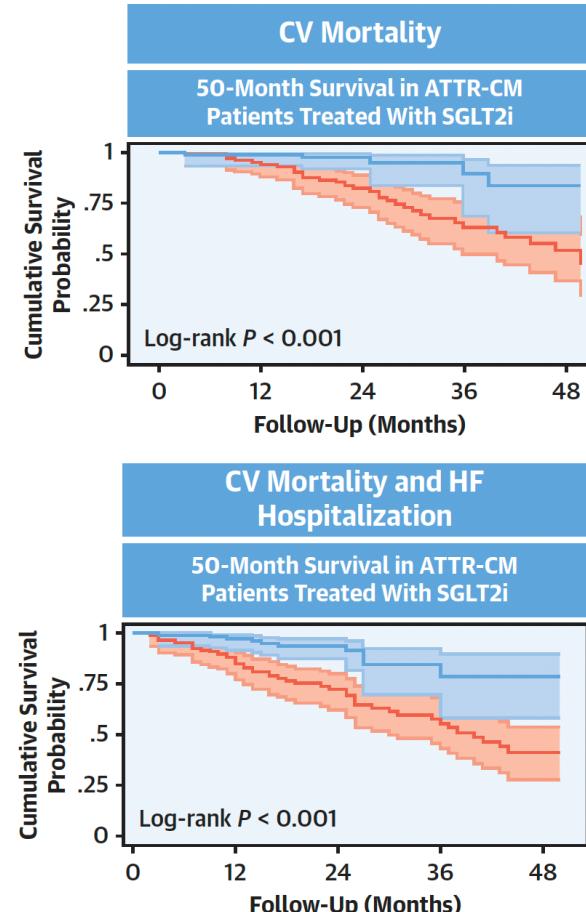
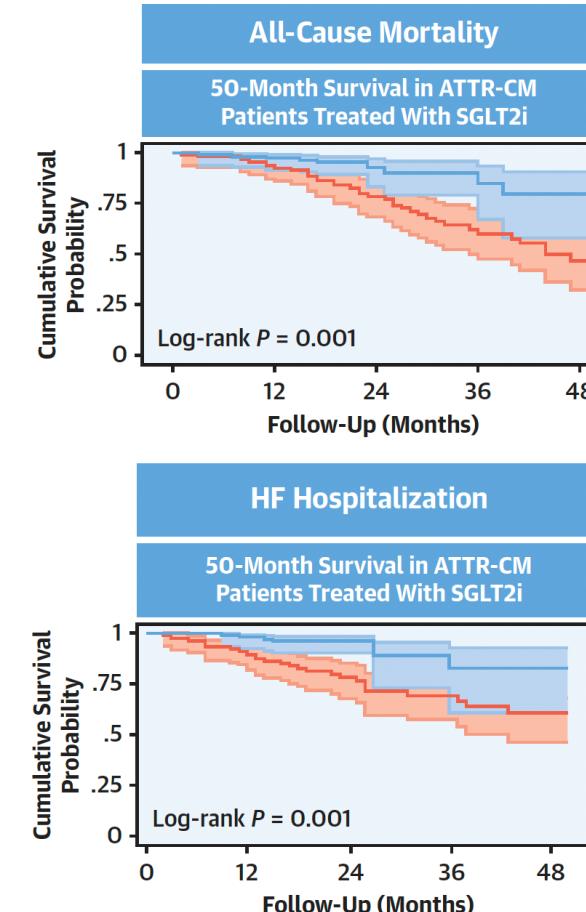
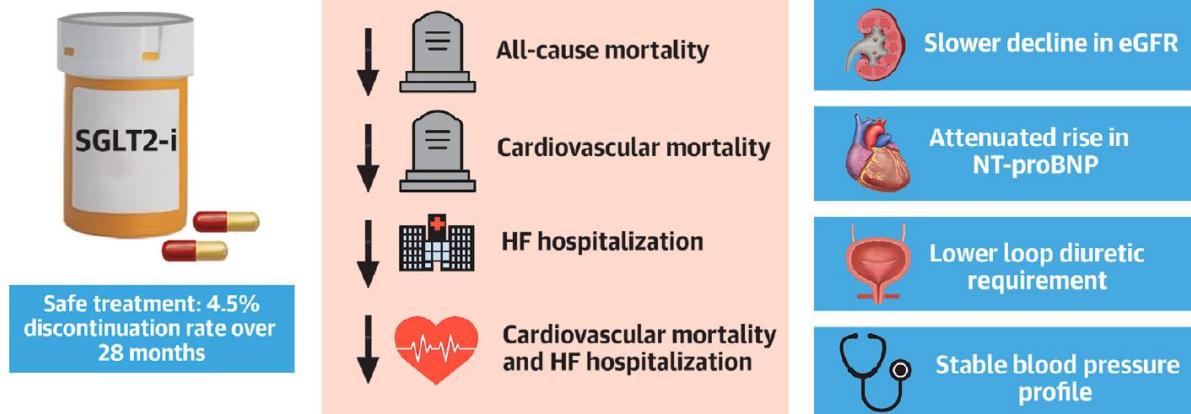
# SGLT2 Inhibitor Therapy in Patients With Transthyretin Amyloid Cardiomyopathy

Progrès réalisés  
et à venir !

## Study Design and Population



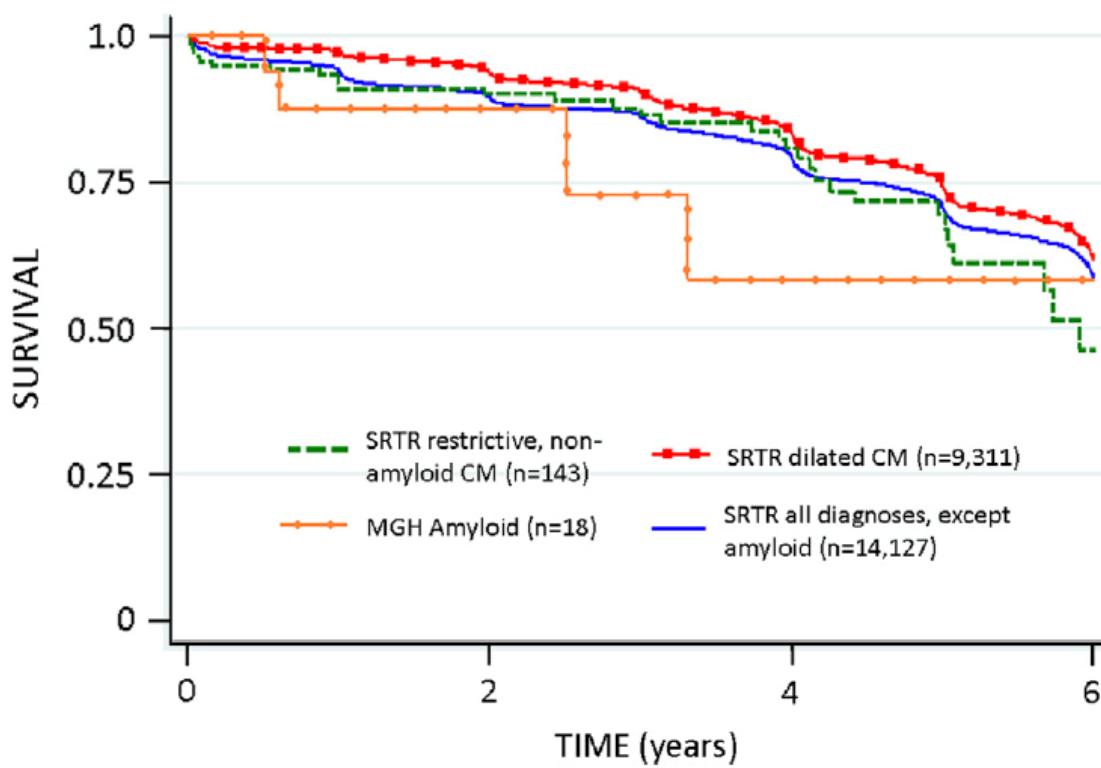
## Main Findings



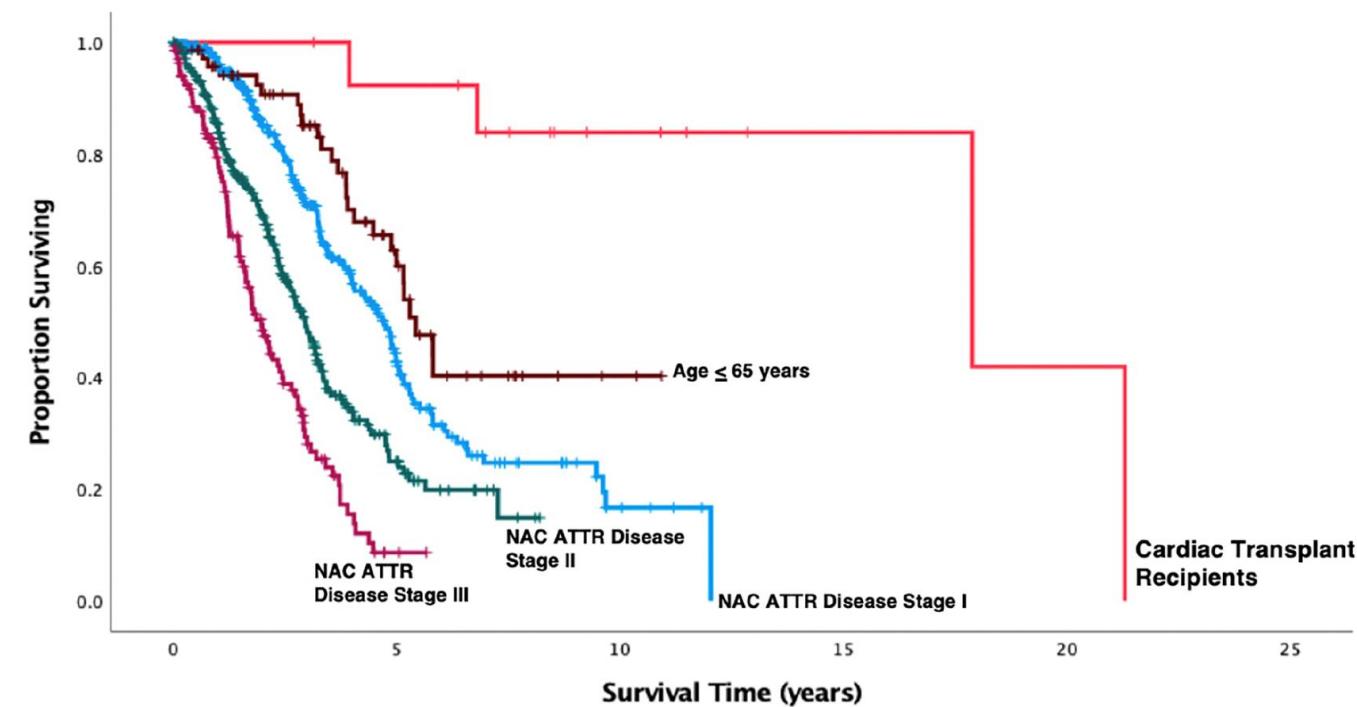
# TRANSPLANTATION & AMYLOSE CARDIAQUE

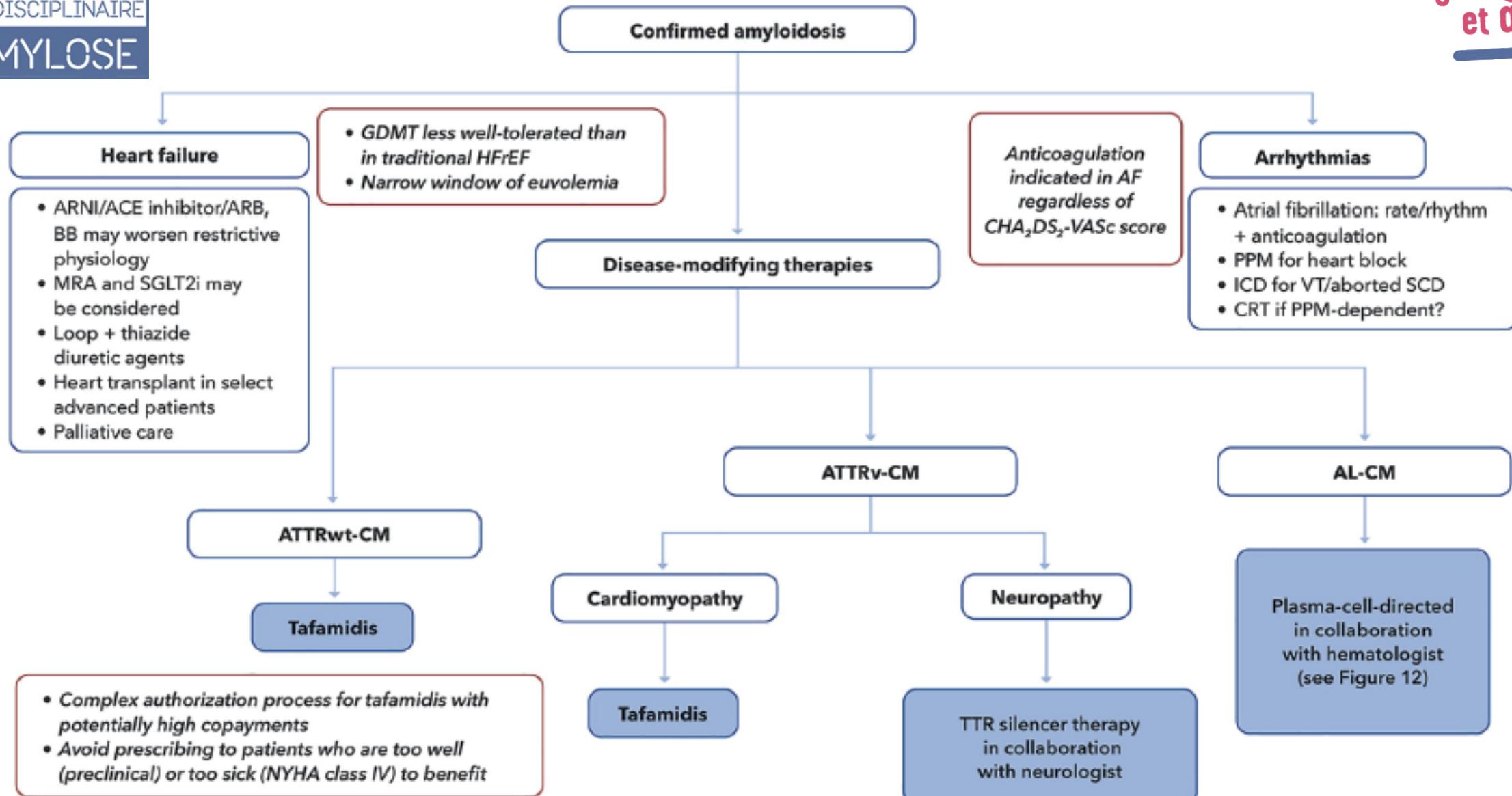
Progrès réalisés  
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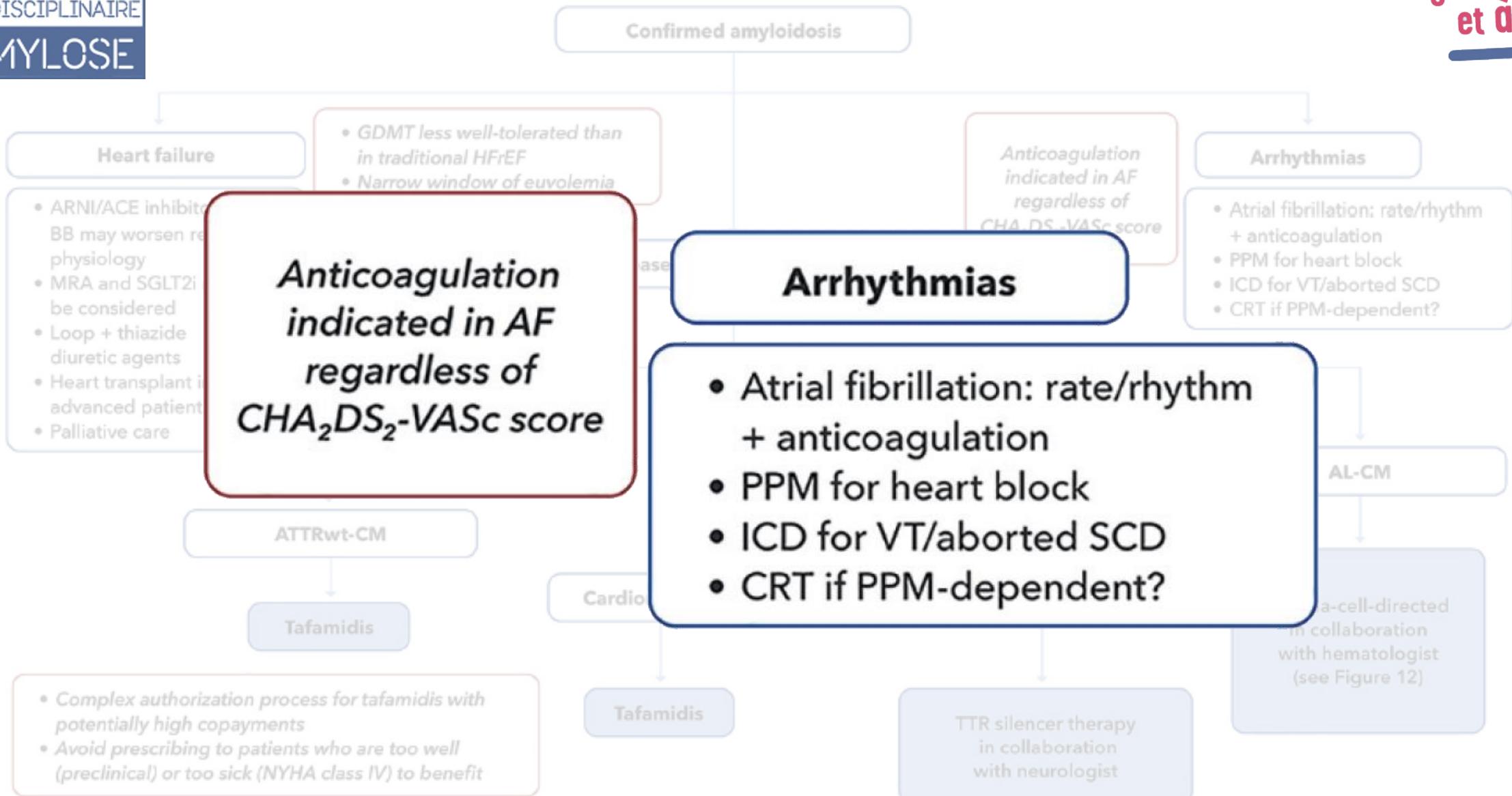
Amylose cardiaque AL



Amylose cardiaque ATTR







# **2020 ESC Guidelines for the diagnosis and management of atrial fibrillation**

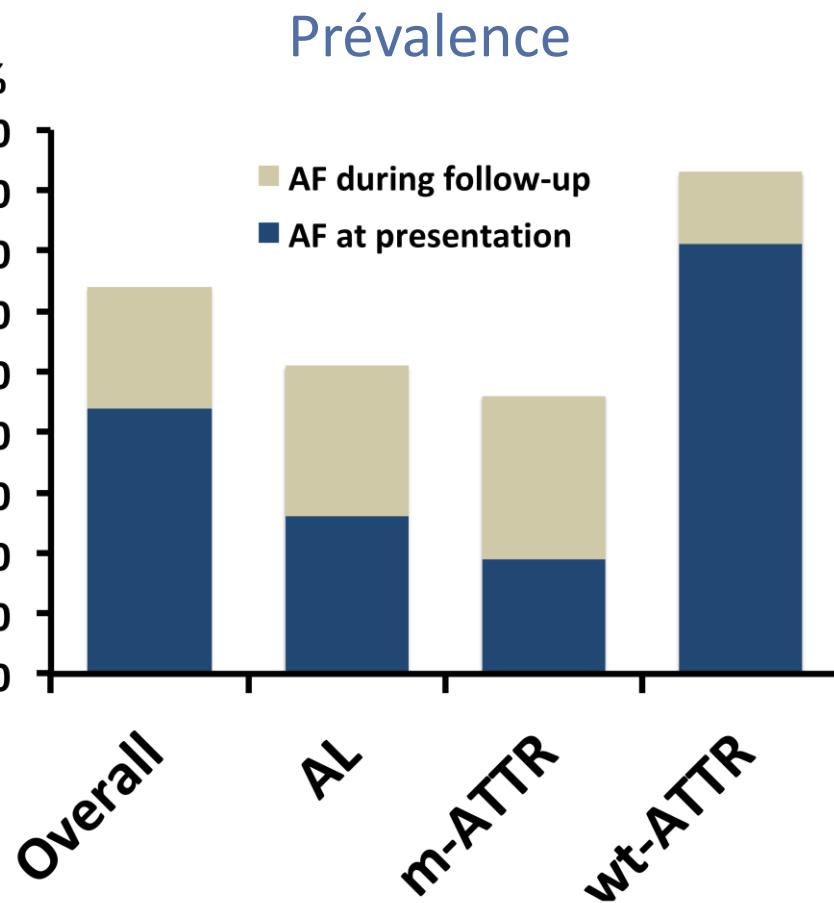
## **INTEGRATED AF MANAGEMENT**



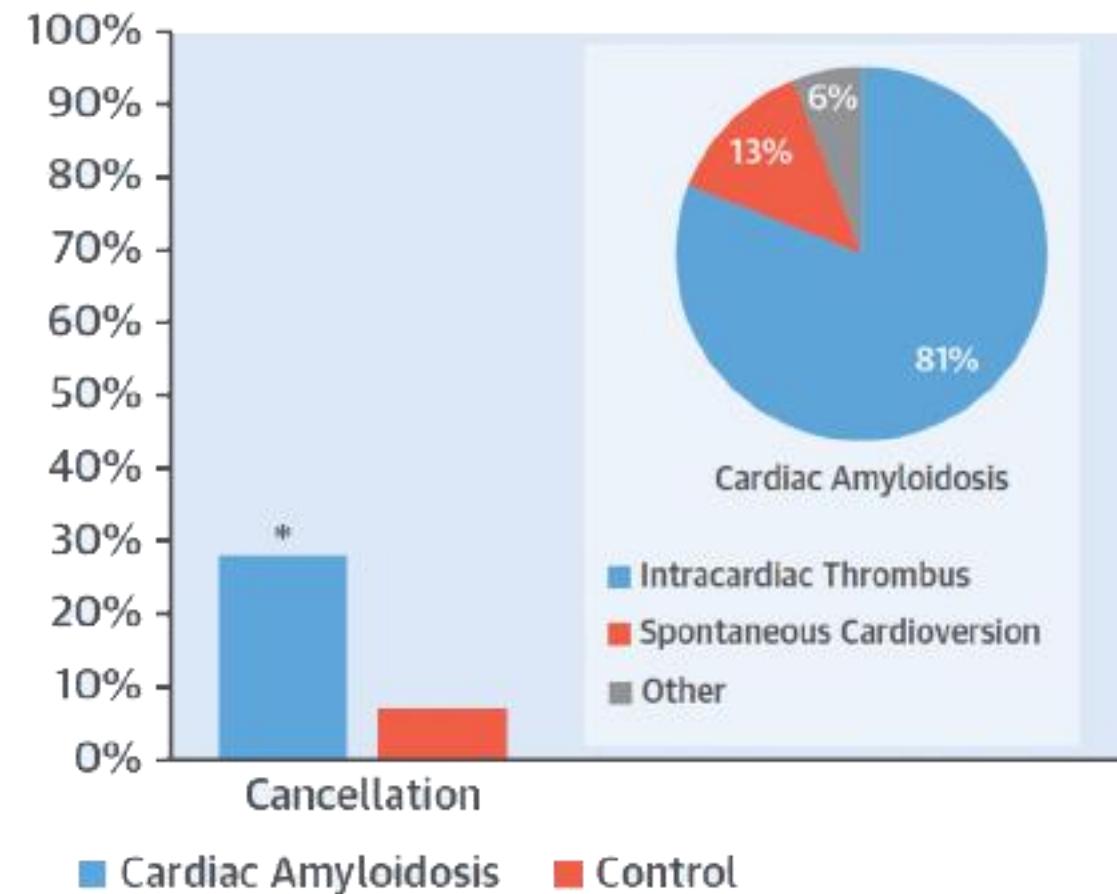
**Patient-centred**

**Optimised stroke prevention**

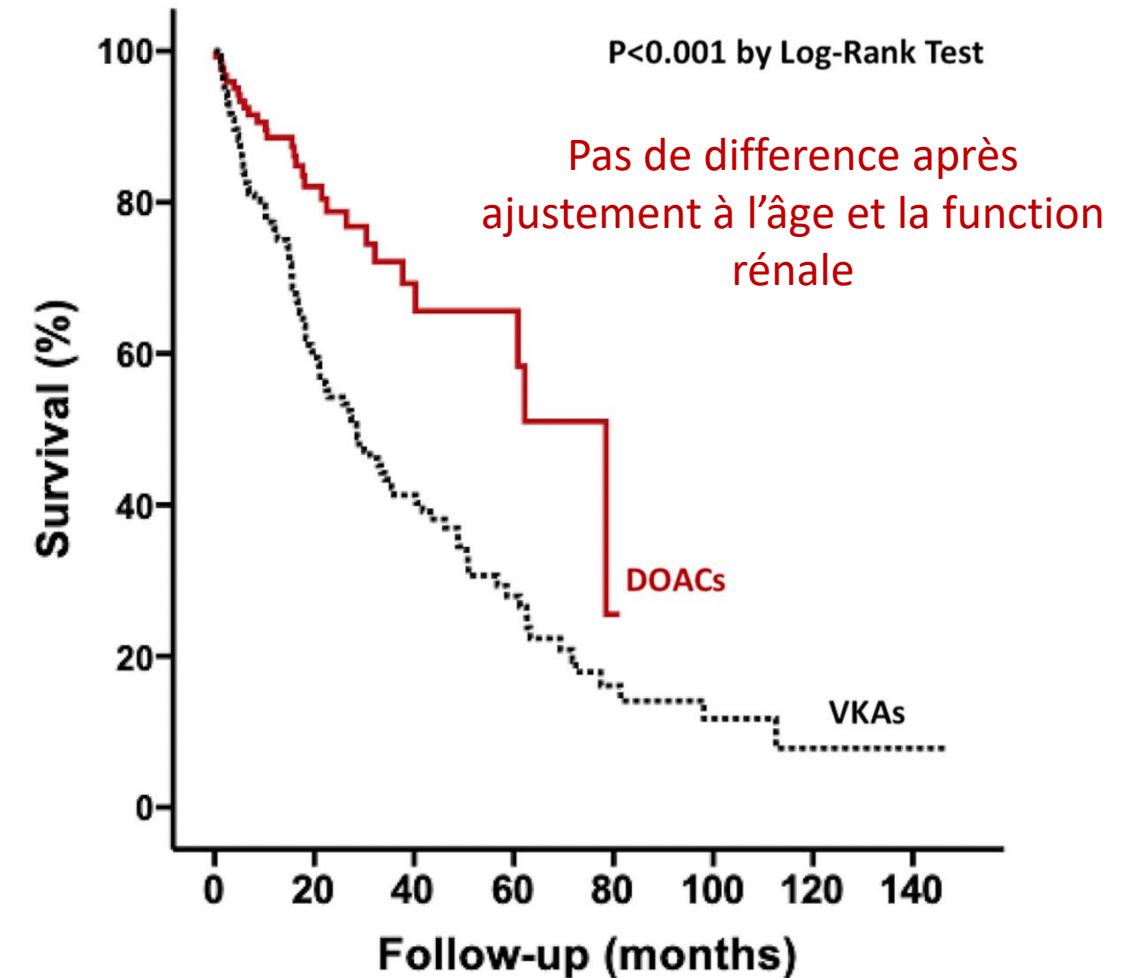
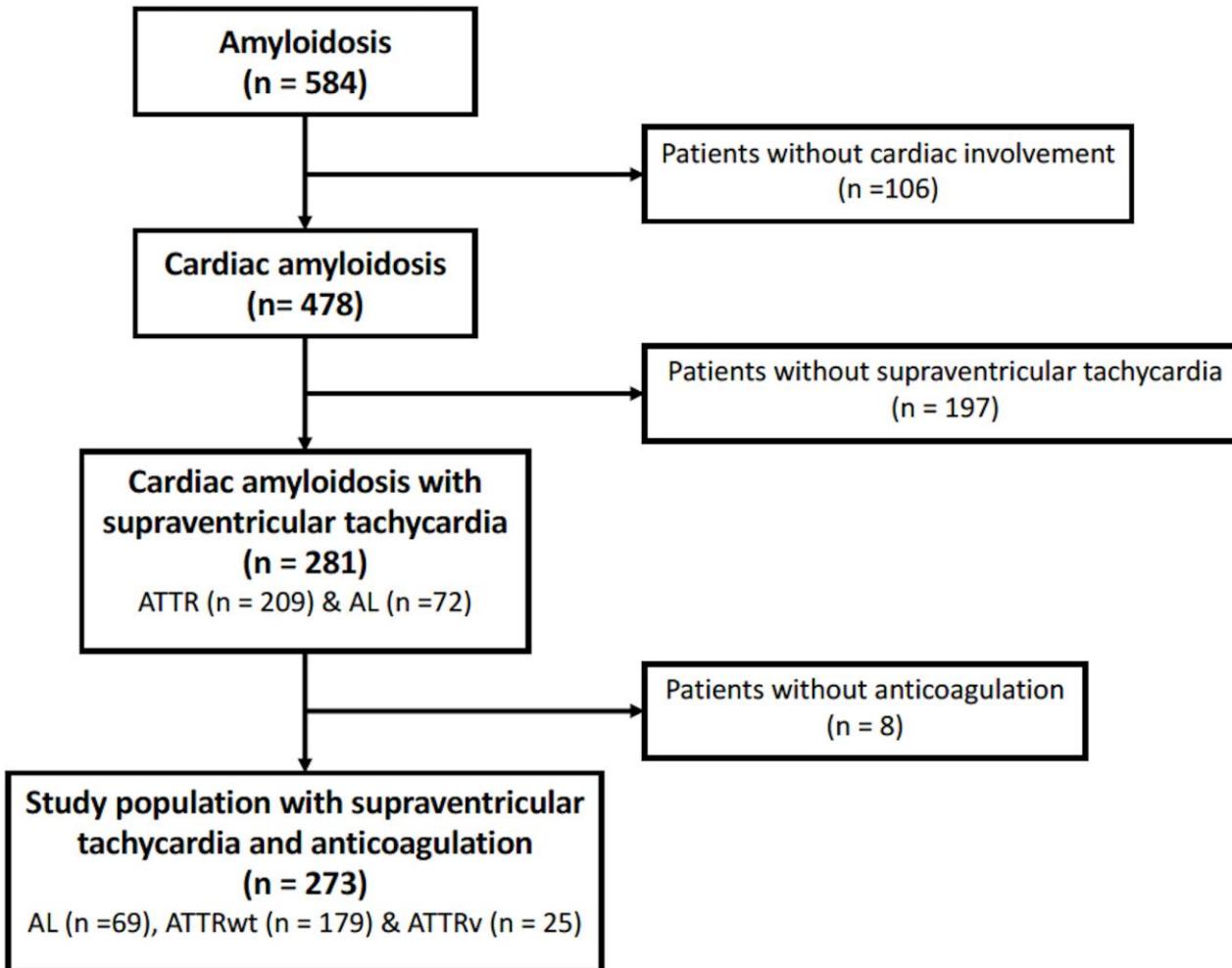
## FA & AMYLOSE CARDIAQUE



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



# ANTICOAGULATION & AMYLOSE CARDIAQUE



# 2020 ESC Guidelines for the diagnosis and management of atrial fibrillation

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## INTEGRATED AF MANAGEMENT



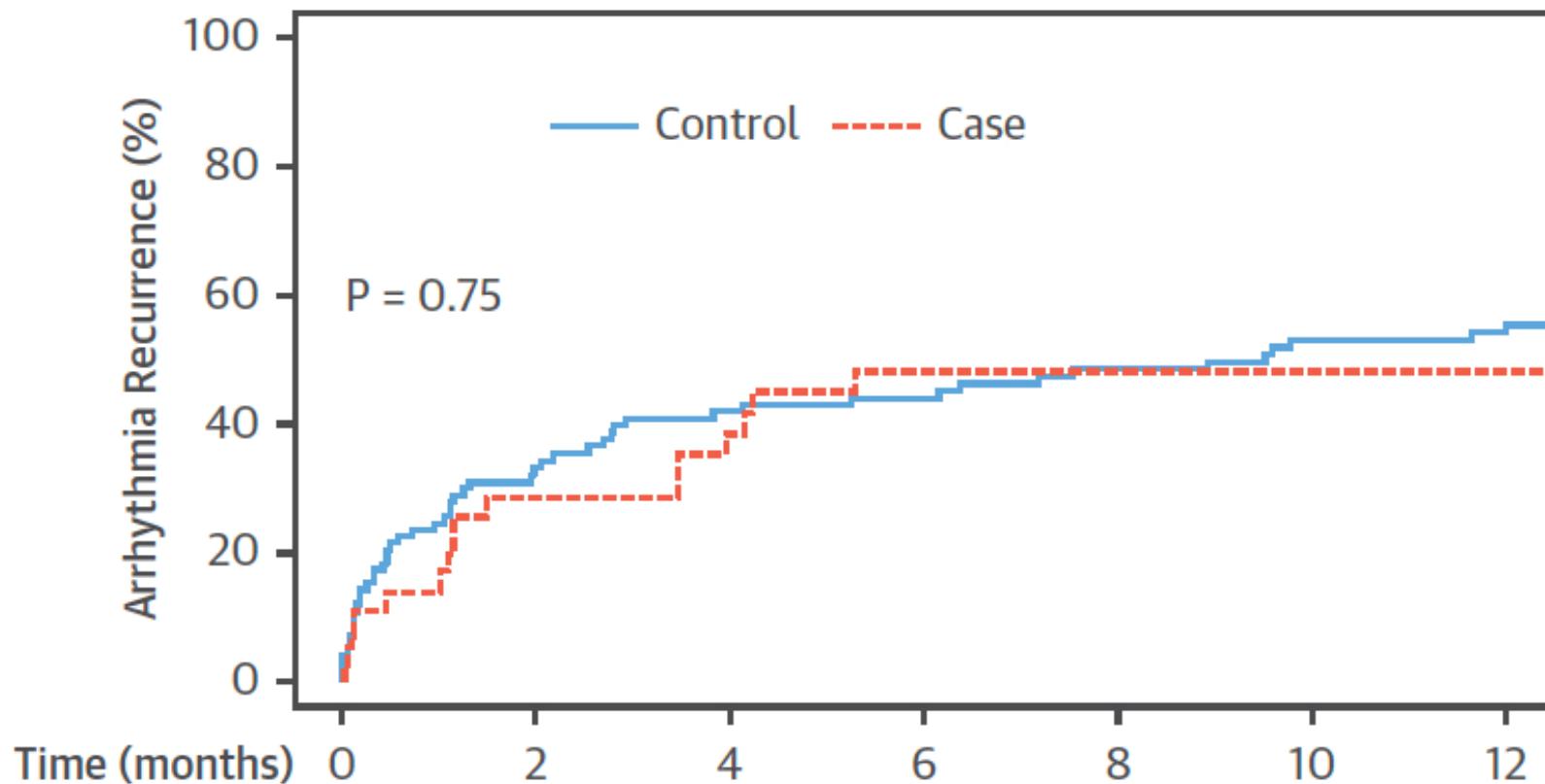
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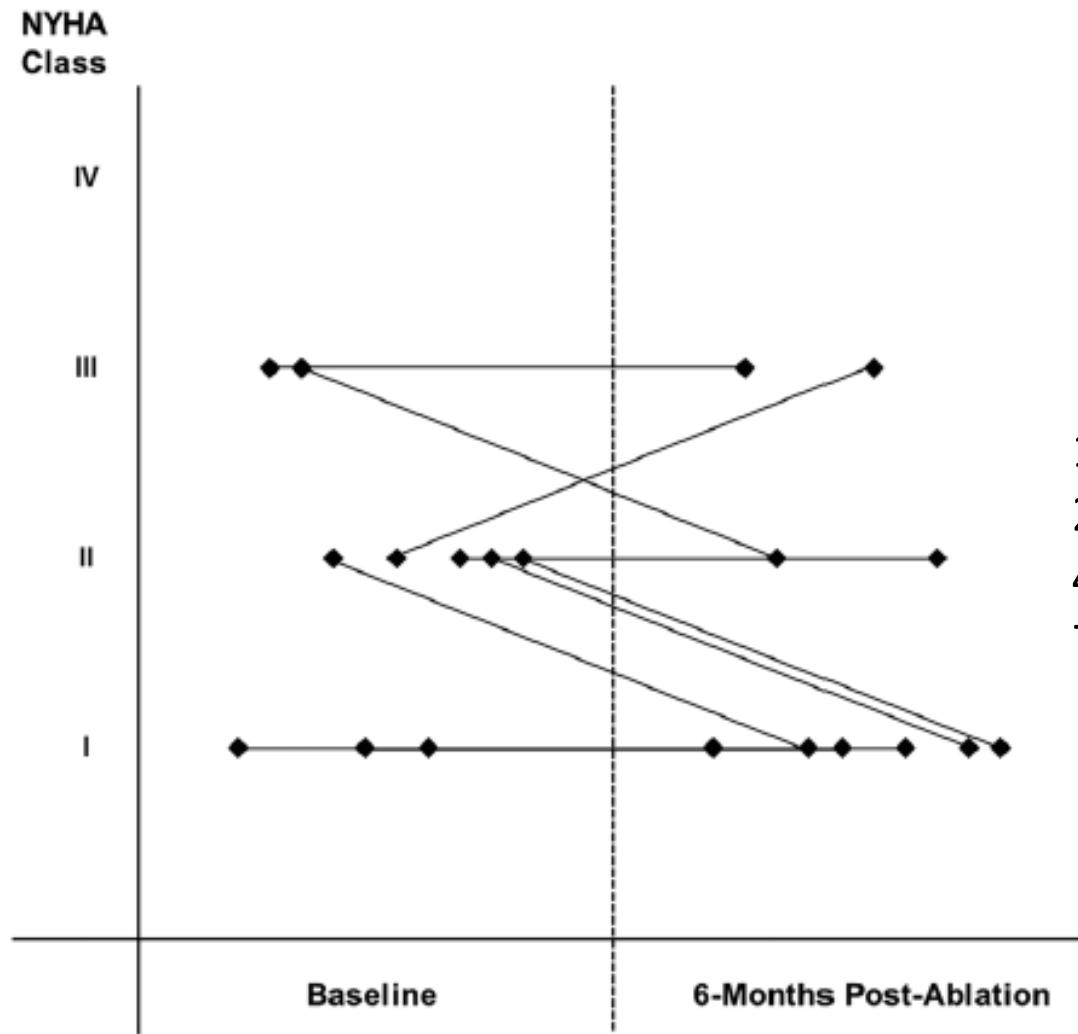
Symptom control with rate or rhythm control

# CARDIOVERSION & AMYLOSE CARDIAQUE

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



## ABLATION & AMYLOSE CARDIAQUE



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?

Check risk factors for digoxin toxicity: eGFR, thyroid function, concomitant drug interaction: amiodarone,  $\beta$ -blockers and calcium channel blockers

Initiate at low dose:

- 0.125 mcg daily when no risk factors present
- 0.0625 mcg every 24-48h if risk factor(s)
- Dialysis: 0.0625 mcg every 48h or as recommended by nephrologist

Monitor serum digoxin concentration (trough level) after 5-7 days  
Adjust dose to keep drug level: 0.5-0.8 ng/mL

Monitor eGFR, electrolytes, drug concentration every 1-2 weeks for first three months, thyroid function monthly, depending on clinical status; then monthly for next three months

Periodic re-assessment of need for ongoing treatment and potential side effects.

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

Progrès réalisés  
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## 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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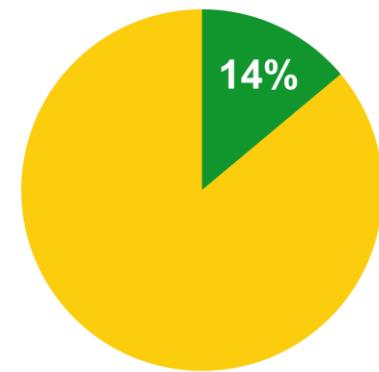
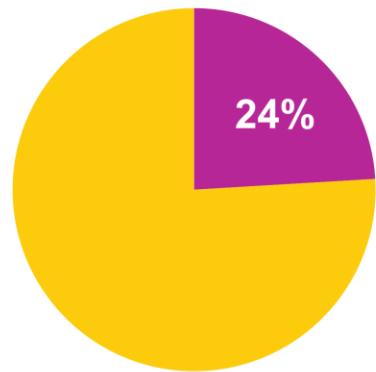
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# RAo & AMYLOSE CARDIAQUE (ATTRWT)

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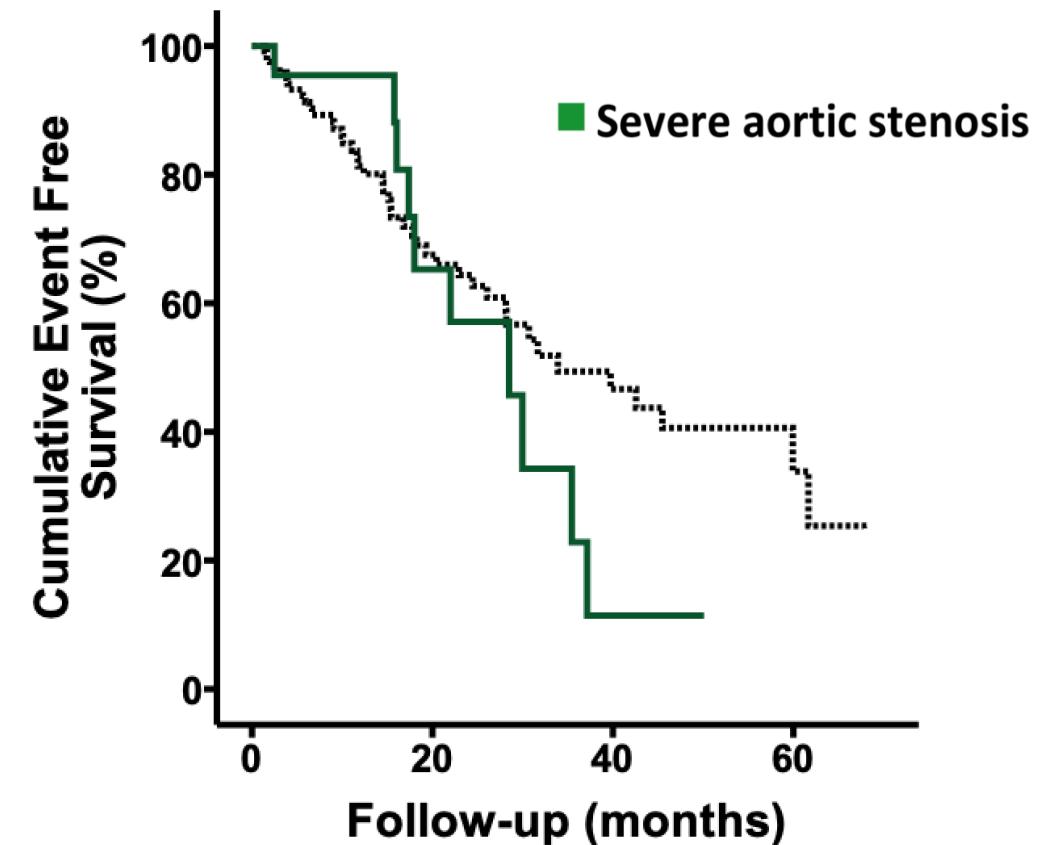
Whole population

Patients over 80 years

■ Aortic stenosis

■ Severe aortic stenosis

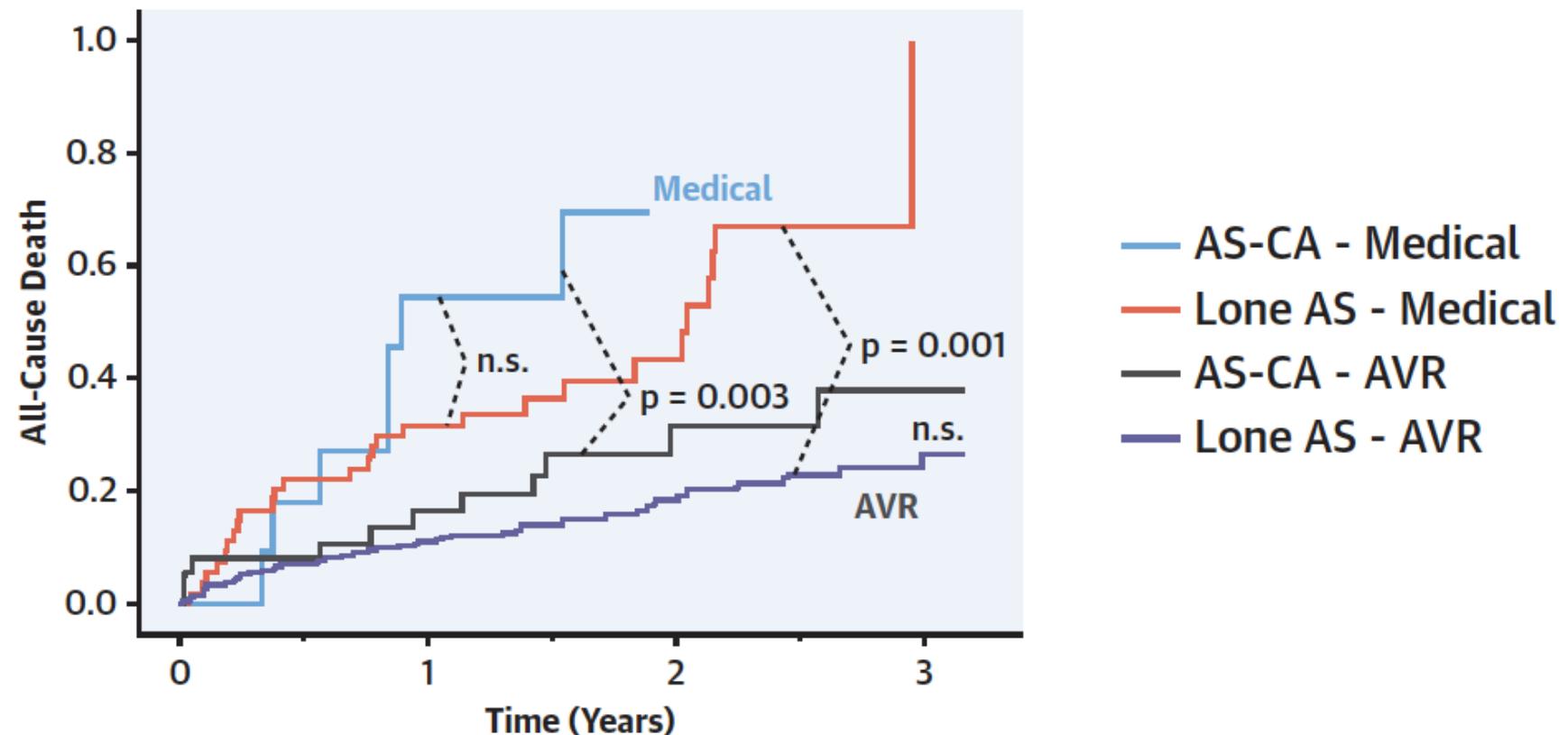
■ No aortic stenosis



# Prevalence and Outcomes of Concomitant Aortic Stenosis and Cardiac Amyloidosis

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



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

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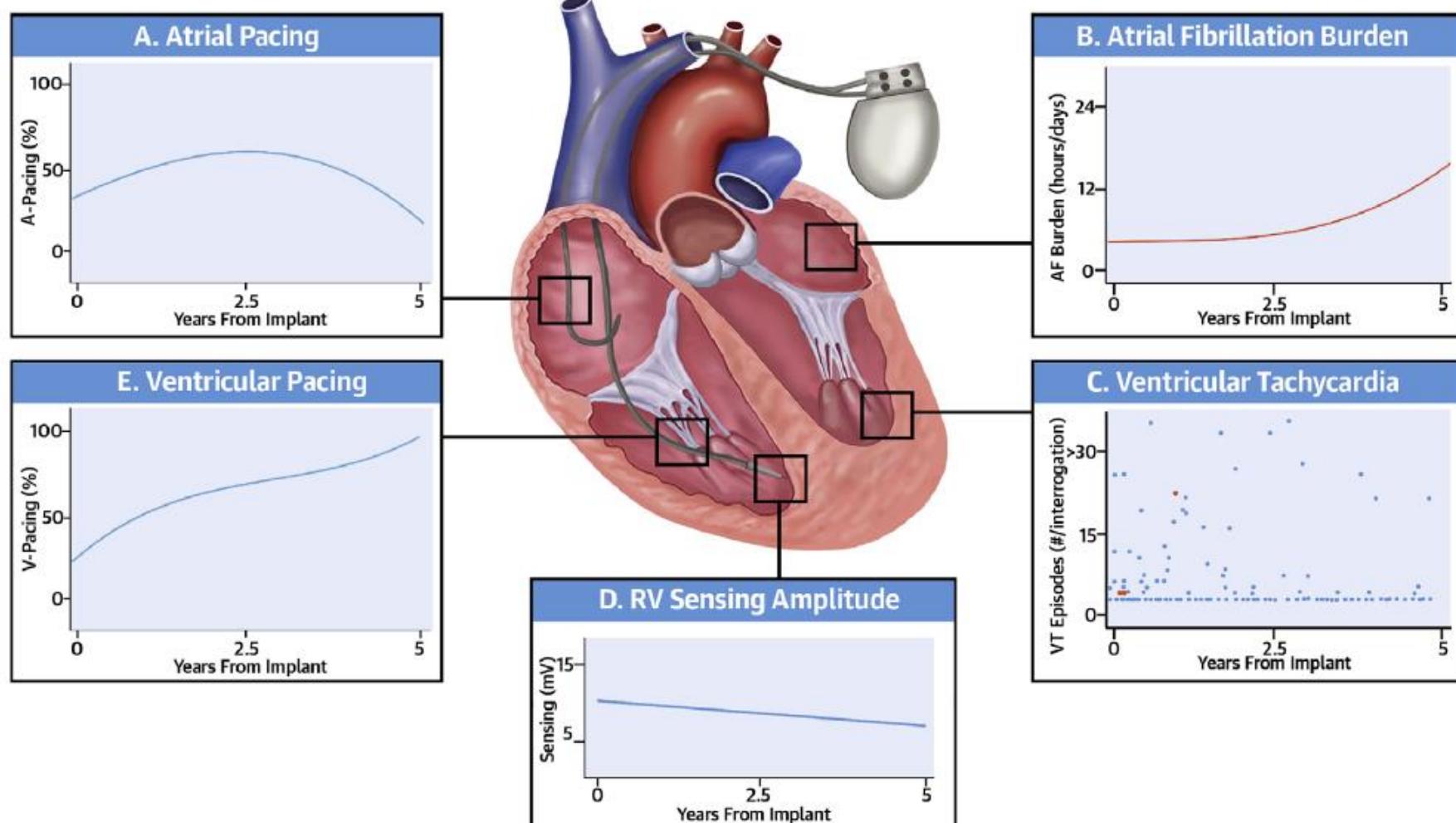
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# Cardiac Implantable Electronic Devices

A Window Into the Evolution of Conduction Disease in Cardiac Amyloidosis

Progrès réalisés  
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# HYPOTENSION ORTHOSTATIQUE & AMYLOSE CARDIAQUE

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## 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

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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**