8e MASTERCLASS AMYLOSES CARDIAQUES

ÉVÉNEMENT HYBRIDE

Jeudi 18 décembre 2025
Fondation Biermans-Lapôtre ■ PARIS

www.masterclass-amylose.com

Evolution de rétrécissement aortique : POUR la dégénérescence

Dr Stephanie CAZALBOU

CHU de Toulouse

Journée organisée par :

cardiogen filière nationale de santé maladies cardiaques héréditaires

Réseau Amylose

ASSOCIATION FRANÇAISE CONTRE L'AMYLOSE

GRACE GROUP FOR RESEARCH ON AMYLOIDOSIS AND CARDIAC EXCELLENCE F-CRIN NETWORK

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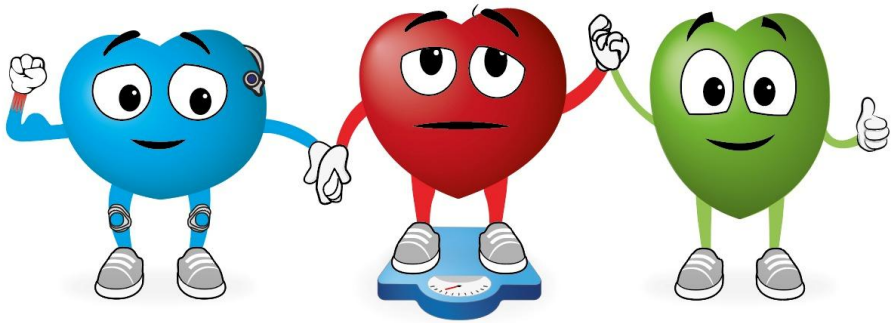
GICC

REPOF

cardiogen filière nationale de santé maladies cardiaques héréditaires ou rares

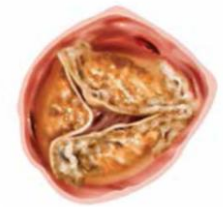
Liens d'intérêts

- Aucun



R' EPOF EPOF EPON

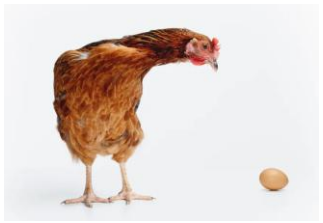
Prévalence de l'amylose et RAC



Prévalence RAO > 75 ans

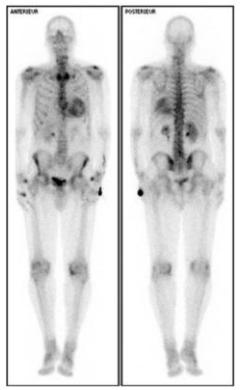
3% Rao serré

12% Rao tout venant



Prévalence ATTR+Rao serré

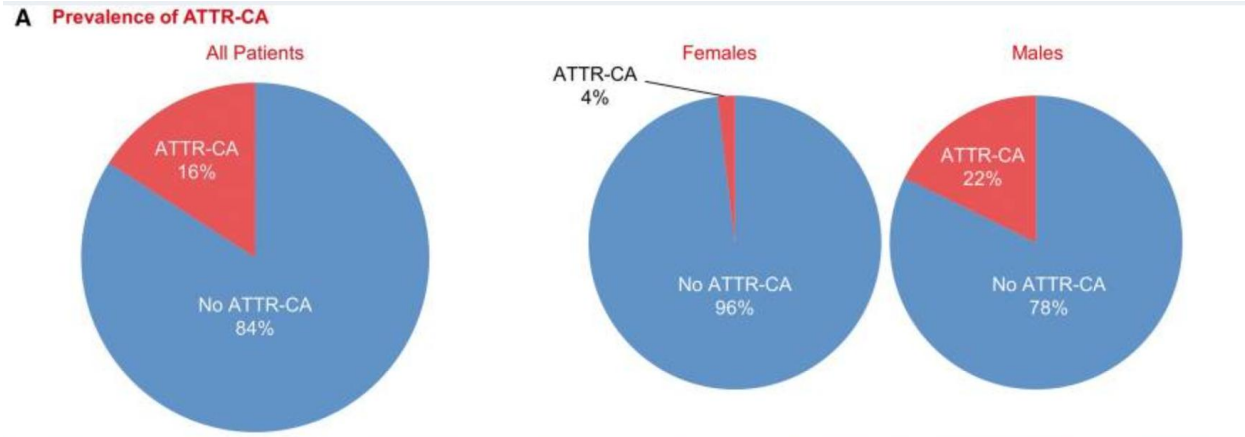
6-16%



Prévalence de l'amylose à TTR

>70 ans: 3,3%

>80 ans: 20%



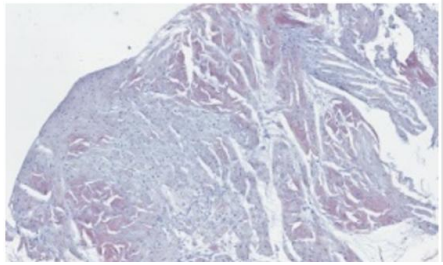
Castaño A, Narotsky DL. Eur Heart J. 2017 Oct 7;38(38):2879-2887

Thaden J.J.,. Prog Cardiovasc Dis. 2014;56:565–571

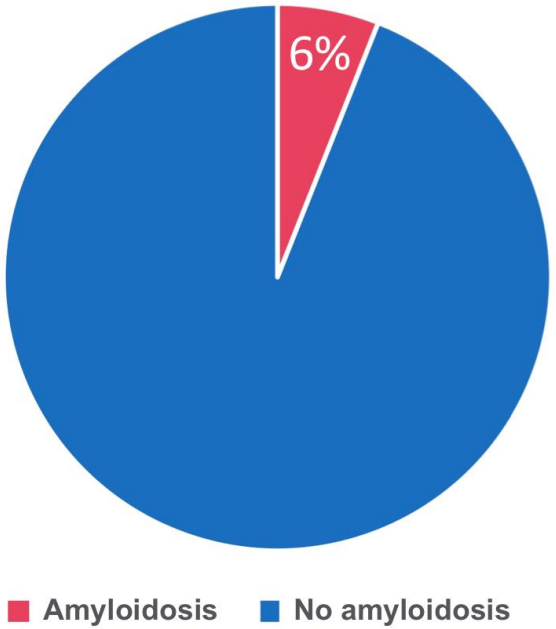
J Lebek, *European Heart Journal*, November 2025

Occult Transthyretin Cardiac Amyloid in Severe Calcific Aortic Stenosis

Prevalence and Prognosis in Patients Undergoing Surgical Aortic Valve Replacement



Prévalence de l'amylose cardiaque à TTR de **6%** parmi les patients > 65 ans référés pour remplacement valvulaire aortique chirurgical avec un ratio homme/femme 2:1



Treibel TA, *Circ Cardiovasc Imaging*. 2016;9:e005066

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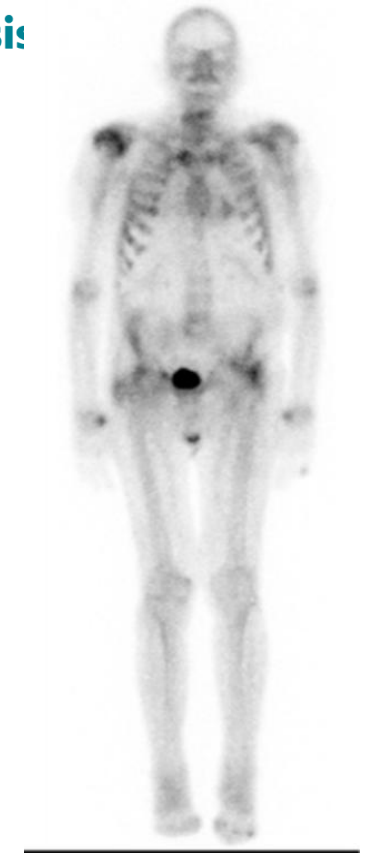
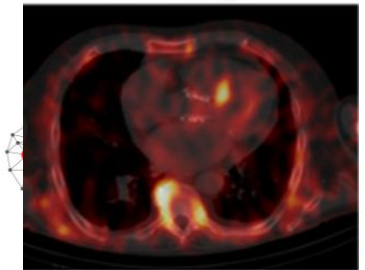
Prevalence and outcome of dual aortic stenosis and cardiac amyloid pathology in patients referred for transcatheter aortic valve implantation

Paul R. Scully[1,2], Kush P. Patel[1,2], Thomas A. Treibel[1,2], George D. Thornton[1], Rebecca K. Hughes[1,2], Sucharitha Chadalavada[1], Michail Katsoulis[3], Neil Hartman[4], Marianna Fontana[5], Francesca Pugliese[1,6], Nikant Sabharwal[7], James D. Newton[7], Andrew Kelion[7], Muhiddin Ozkor[1], Simon Kennon[1], Michael Mullen[1], Guy Lloyd[1,2,6], Leon J. Menezes[1,8,9], Philip N. Hawkins[5], and James C. Moon[1,2*]

- **Prévalence AS-AC 16%**
  - Grade 1: 30%
  - Grade 2: 70%
  - Grade 3: 0





Prevalence and Outcomes of Concomitant Aortic Stenosis and Cardiac Amyloidosis

Christian Nitsche, MD,[a] Paul R. Scully, PhD,[b,c] Kush P. Patel, MBBS,[b,d] Andreas A. Kammerlander, MD, Matthias Koschutnik, MD,[a] Carolina Dona, MD,[a] Tim Wollenweber, MD,[e] Nida Ahmed, MBBS,[b,d] George D. Thornton, MBBS,[b,d] Andrew D. Kelion, MD,[f] Nikant Sabharwal, MD,[f] James D. Newton, MD,[f] Muhiddin Ozkor, MD,[d] Simon Kennon, MD,[d] Michael Mullen, MD,[d] Guy Lloyd, MD,[b,d,g] Marianna Fonta Philip N. Hawkins, FMedSci,[h] Francesca Pugliese, MD,[b,g] Leon J. Menezes, MD,[d,i] James C. Moon, MD,[b] Julia Mascherbauer, MD,[a] Thomas A. Treibel, PhD[b,d]

- **Prévalence AS-AC 12%**
  - Grade 1: 33%
  - Grade: 2/3: 67 %

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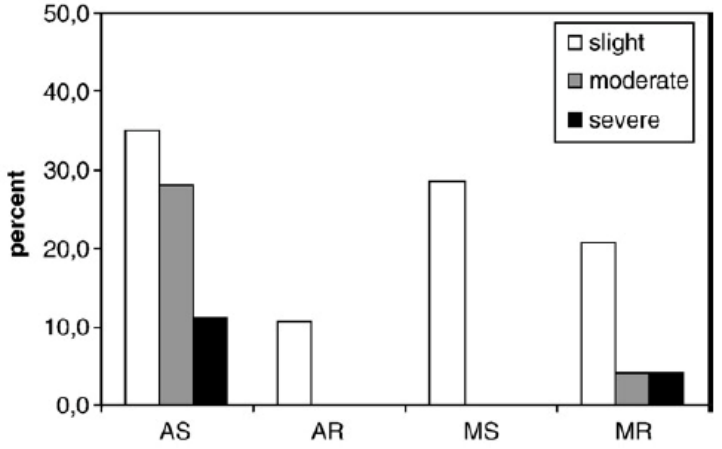
Dépôts amyloids et valvulopathies

High prevalence of amyloid in 150 surgically removed heart valves—a comparison of histological and clinical data reveals a correlation to atheroinflammatory conditions

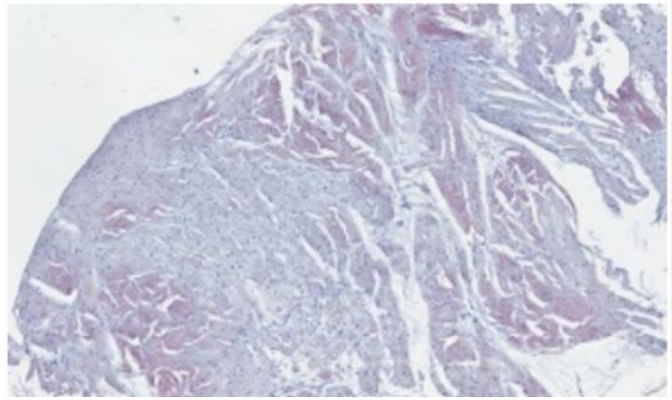


Dépôts amyloïdes retrouvés dans 53% des valvulopathies

74% dans la sténose aortique

AS		Aortic regurgitation		Mitral stenosis		Mitral regurgitation	
Without amyloid (n=26)	With amyloid (n=74)	Without amyloid (n=17)	With amyloid (n=2)	Without amyloid (n=5)	With amyloid (n=2)	Without amyloid (n=19)	With amyloid (n=5)

Kristen AV, *Cardiovasc Pathol*. 2010;19:228-35



Contents lists available at ScienceDirect

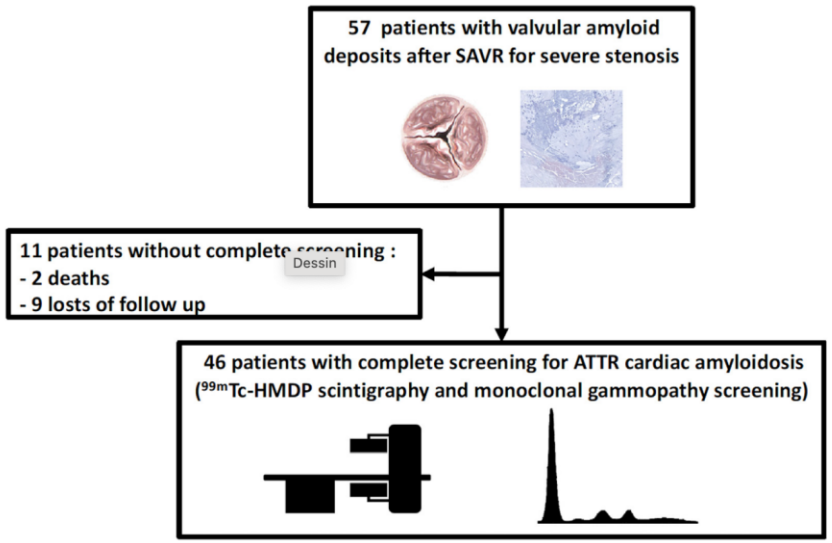
Cardiovascular Pathology

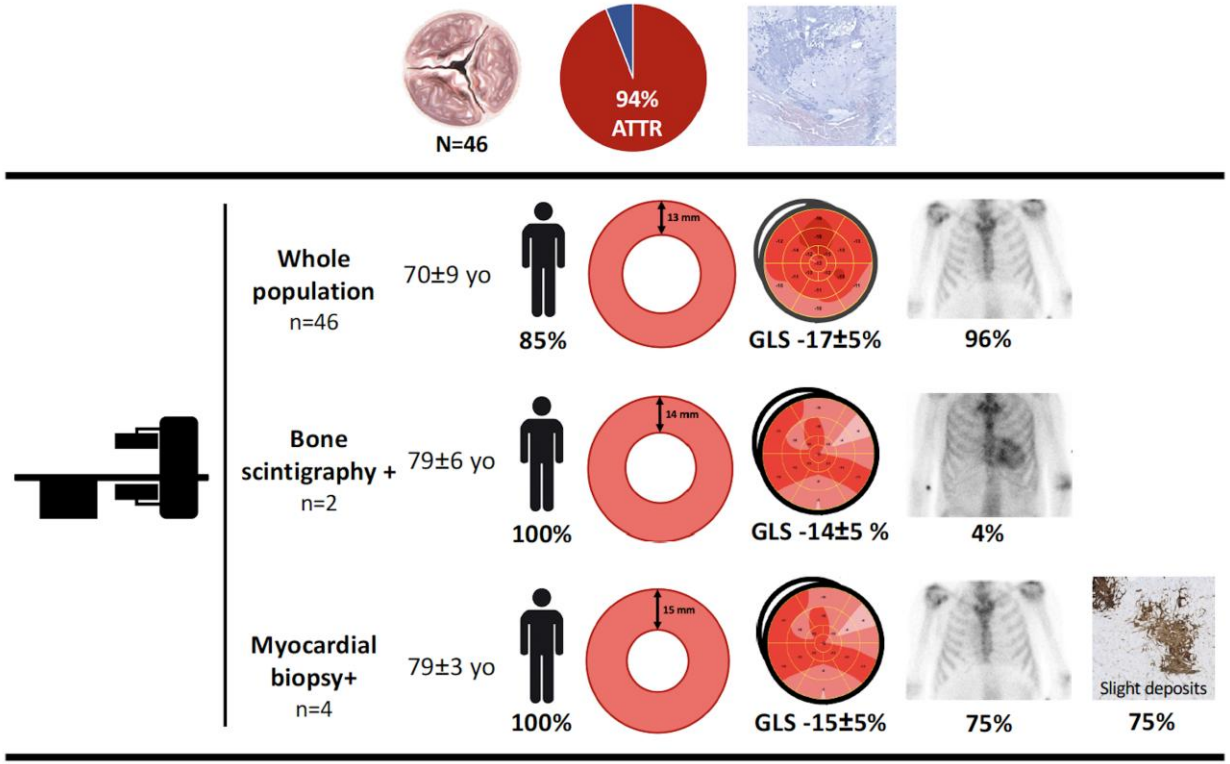
journal homepage: www.elsevier.com/locate/carpath

Original Article

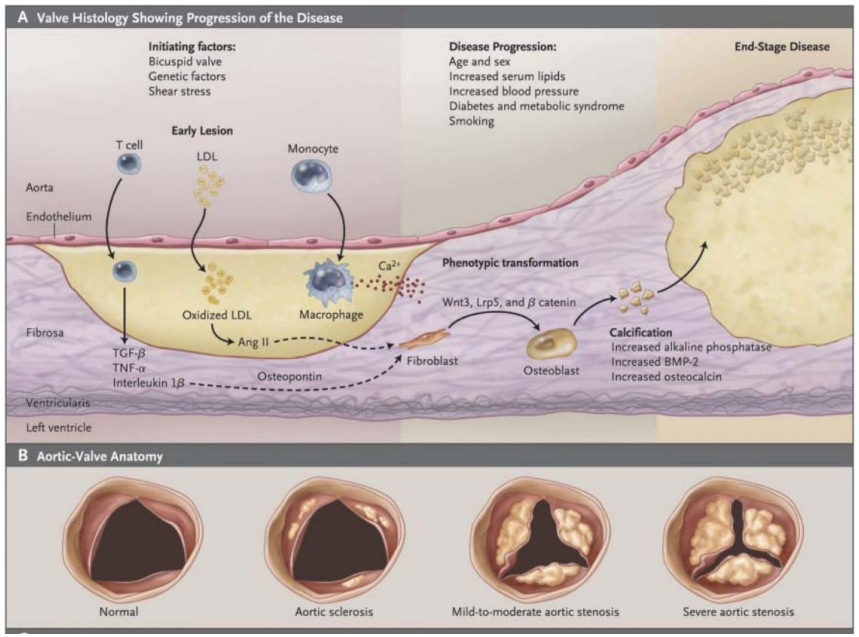
Descriptive study of the clinical and myocardial status of a population with anatomopathological aortic valve amyloidosis ☆

Jean-Baptiste Brette [a,b], Magali Colombat [d,e], Pauline Fournier [a,b], Maxime Moninhas [a,b], Bertrand Marcheix [d,f], Olivier Lairez [a,b,c,d,*], Eve Cariou [a,b], On behalf of the Toulouse Amyloidosis Research Network collaborators [1]





Histoire naturelle du RAC



Iung B, Vahanian A Degenerative calcific aortic stenosis: a natural history *Heart* 2012;98:iv7-iv13.

Progression/an

0,1cm2 et 7 mmHG

Progression en cas d'amylose associée ?

Dweck MR, Boon NA,. J Am Coll Cardiol. 2012 Nov 6;60(19):1854-63.

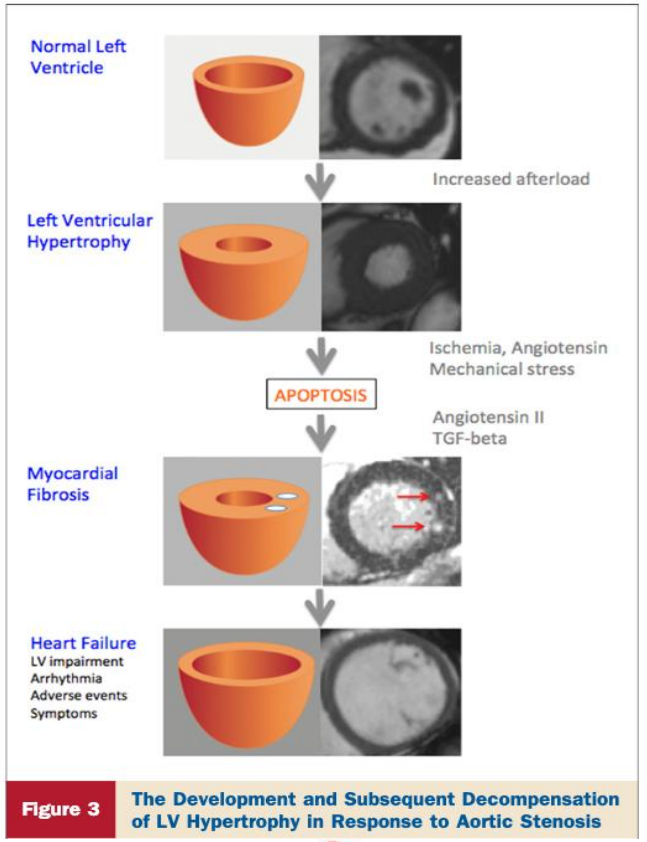
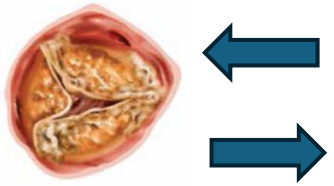
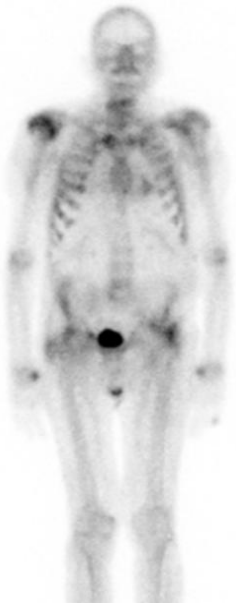


Figure 3 The Development and Subsequent Decompensation of LV Hypertrophy in Response to Aortic Stenosis

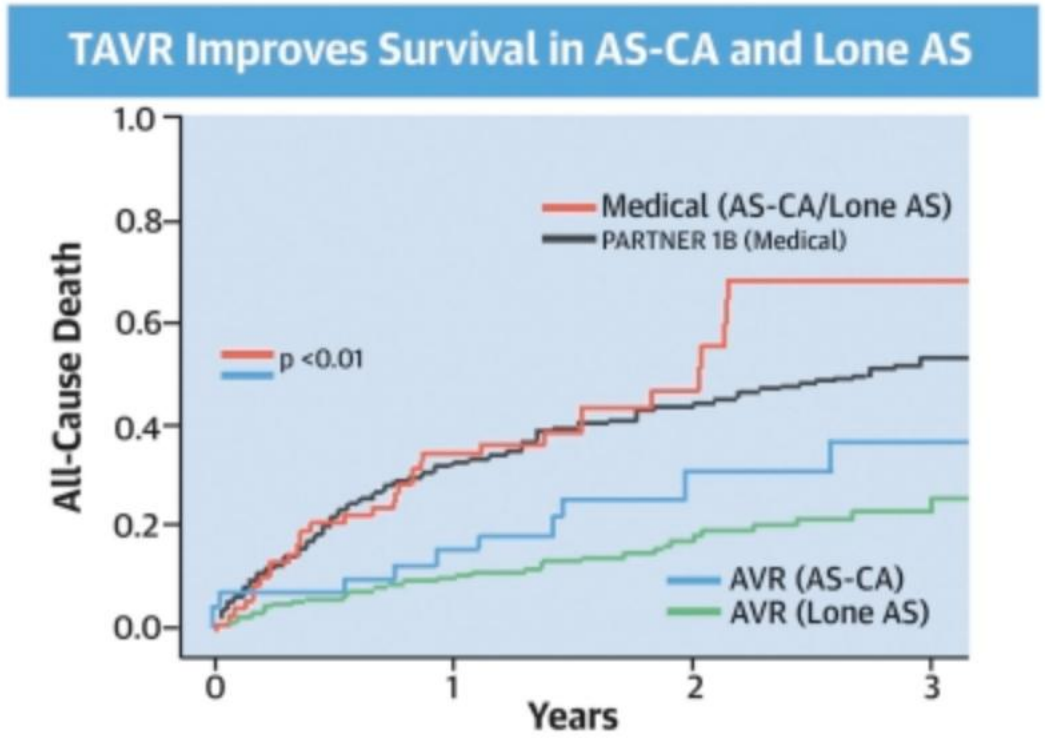
Dégénérescence ?

- RAO > maladie évolutive avec une participation inflammatoire
- Forte prévalence de dépôts amyloïdes valvulaires > atteinte cardiaque
  - *Stade précoce de la maladie ?*
  - *Amylose opportuniste?*
  - *Marqueur de vieillissement ?*
- Manque de données sur le suivi de sténose aortique chez les patients suivis initialement pour une amylose cardiaque





Pronostic/Traitement



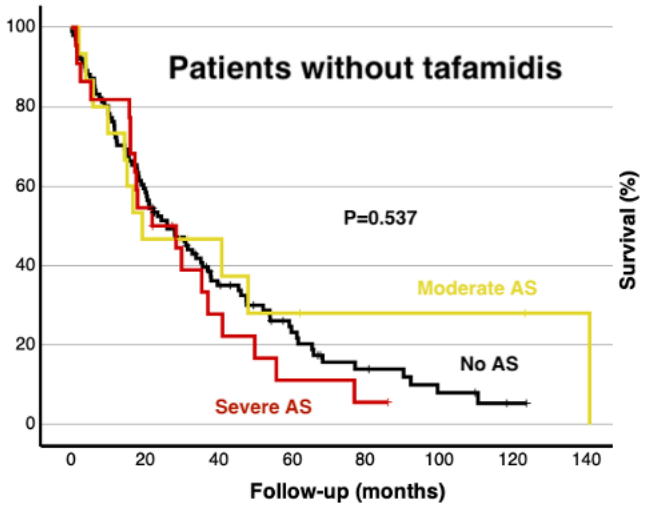
Nitsche C, et al. J Am Coll Cardiol. 2020:S0735-1097(20)37735-4.

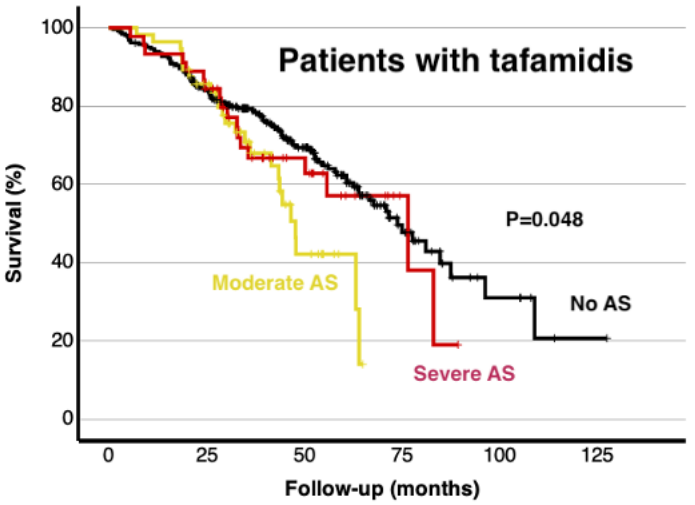
UNIVERSITÉ DE TOULOUSE

Prevalence and Prognostic Impact of Aortic Stenosis in Wild-Type Transthyretin Cardiac Amyloidosis

CHU TOULOUSE

Hugo Malerbi, MD; Hugo Cavalerie; Yassmine Boumzebra; Pauline Fournier; Yoan Lavie-Badie; Stéphanie Cazalbou; Eve Cariou and Olivier Lairez





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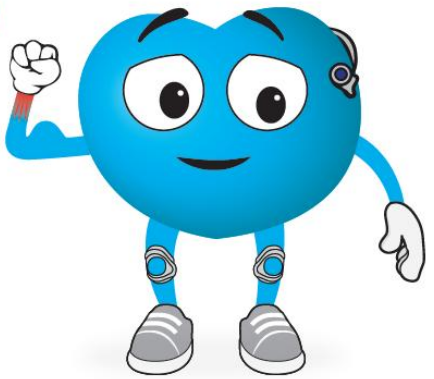
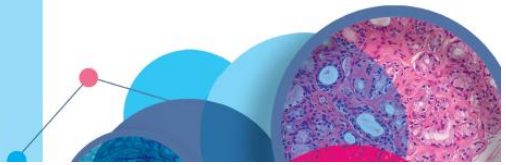
Les atteintes rhumatologiques (synoviales) de l'amylose surviennent plusieurs années avant l'insuffisance cardiaque (EPOF)

R² EPOF

- Surdité
- Canal carpien
- Doigt à ressaut
- Rupture du tendon du long biceps
- Canal lombaire étroit
- Prothèse de hanche/genou
- Essoufflement
- Prise de poids
- Œdèmes
- Fatigue



Et si c'était une Amylose Cardiaque à transthyrétine ?



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GICC SOCIÉTÉ FRANÇAISE DE CARDIOLOGIE
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TIC-TAC,

*Il est temps de changer de **TAC-TIC**,*

*Passons du **TACØTAC** !!!*

R³ EPOF

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Prevalence and Prognostic Impact of Aortic Stenosis in Wild-Type Transthyretin Cardiac Amyloidosis

Background

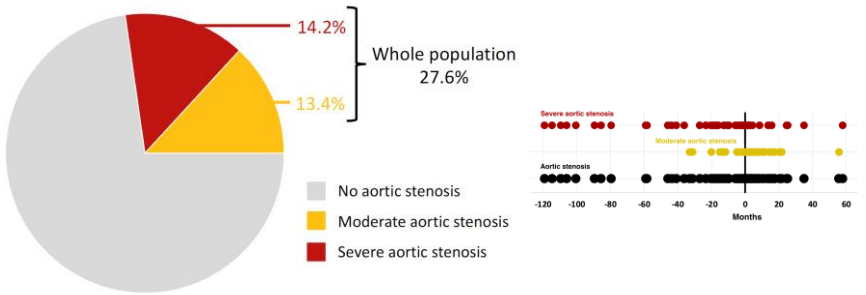
Wild-type transthyretin cardiomyopathy (wtATTR-CM) is an age-related disease frequently associated with aortic stenosis (AS). The aim of this study was to investigate the prevalence and prognostic impact of AS in the natural history of wtATTR-CM.

Methods

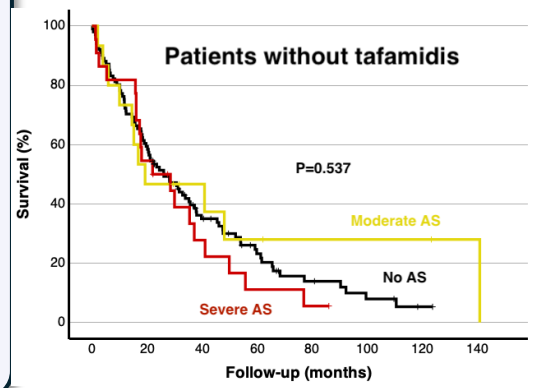
Five hundred patients with wtATTR-CM and full medical records were retrospectively enrolled in the study. Patient's medical records were reviewed to determine the prevalence of AS in ATTRwt-CM, the timing of diagnoses between the two diseases, and their reciprocal prognostic impact. Cox proportional hazards model was used for statistical analysis.

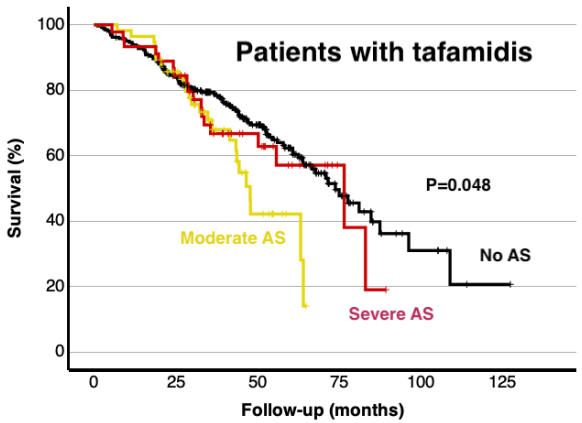
Results

The average age was 81±6 years, with 82% of male. One hundred and thirty-eight (28%) patients had a concomitant diagnosis of AS, including 67 (14%) with severe AS, at the time of diagnosis or during follow-up. Among them, 79% (53/67) underwent an aortic valve replacement. AS was diagnosed before, simultaneously and after wtATTR-CM in 51 (10%), 42 (8%), and 45 (9%) patients, respectively. The presence of AS has no impact on all-cause mortality (hazard ratio 1.22; 95% confidence interval -CI- 0.93 – 1.61, P=0.152). Timing of diagnosis between AS and wtATTR-CM had no impact on all-cause mortality.



Three hundred and sixty-two (72%) patients received tafamidis. Among patients not treated with tafamidis, neither moderate or severe AS had impact on all-cause mortality (hazard ratio 0.75, 95%CI 0.43 – 1.32, P=0.316 and 1.24, 95%CI 0.75 – 2.03, P=0.399, respectively). However, among patients treated with tafamidis, after adjustment on age, moderate AS had a trend to increase all-cause mortality (hazard ratio 1.51; 95%CI 0.97 – 2.35, P=0.07).





Conclusion

AS is found in a quarter of patients with wtATTR-CM. Patients with severe AS mainly undergo aortic valve replacement, and AS had no impact on mortality before the advent of ATTR-targeted therapy. In patients receiving tafamidis, there is a trend towards increased mortality in the presence of moderate AS. These results show that the introduction of ATTR-targeted therapy shifts the prognosis from myocardial to valvular disease.

Unveiling transthyretin cardiac amyloidosis and its predictors among elderly patients with severe aortic stenosis undergoing transcatheter aortic valve replacement

Castano A, *European Heart Journal* 2017;38:2879–2887