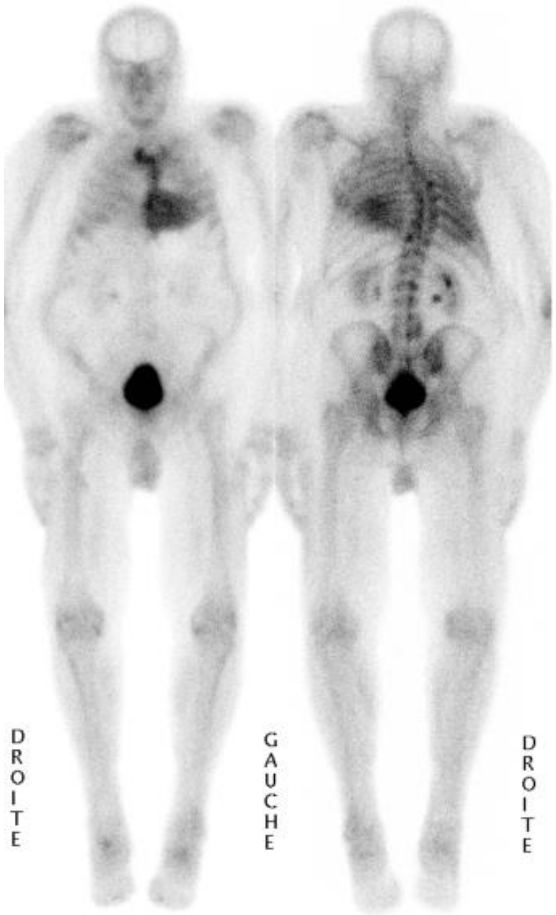
Amylose cardiaque

Cas clinique



- Homme
- 85 ans
- Bilan d'extension d'un cancer de prostate
- PSA 15ng/ml Gleason 10

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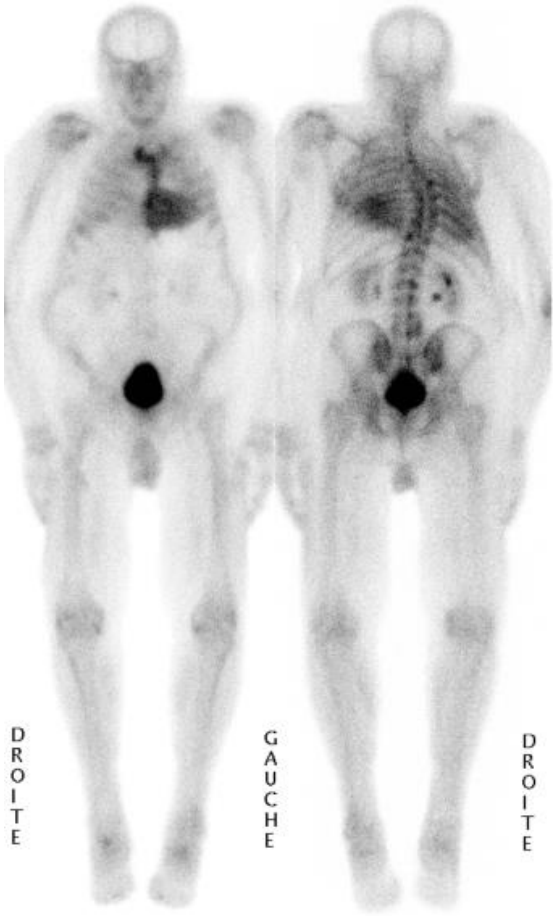
Mai 2017

H 85 ans, ADK prostate (PSA 15ng/ml Gleason 10)



- Hyperfixation myocardique intense à confronter aux antécédents (amylose? cardiopathie ischémique sévère?)
- Pas d'argument scintigraphique pour des localisations osseuses secondaires

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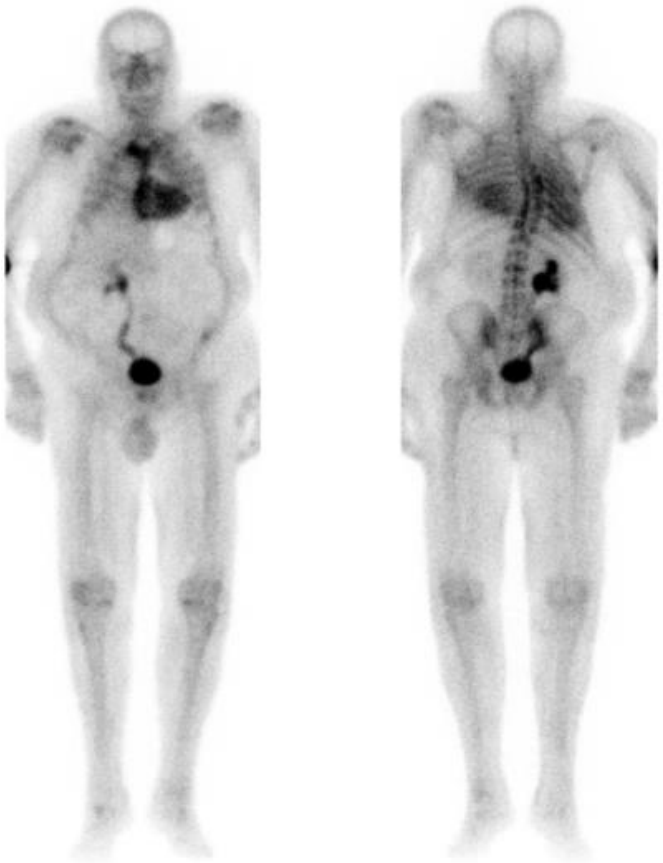
Juin 2019
Suivi évolutif chez ce même patient

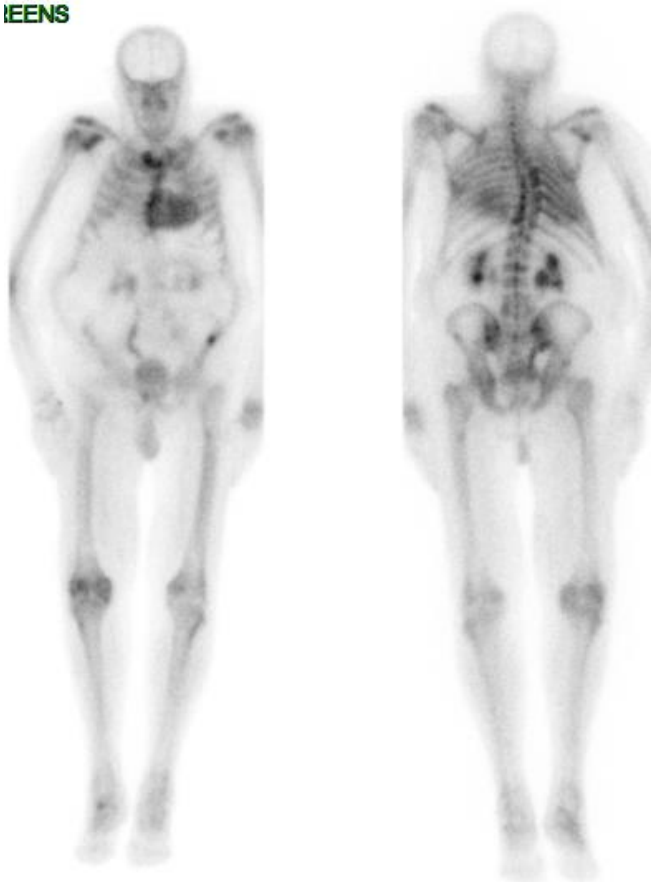


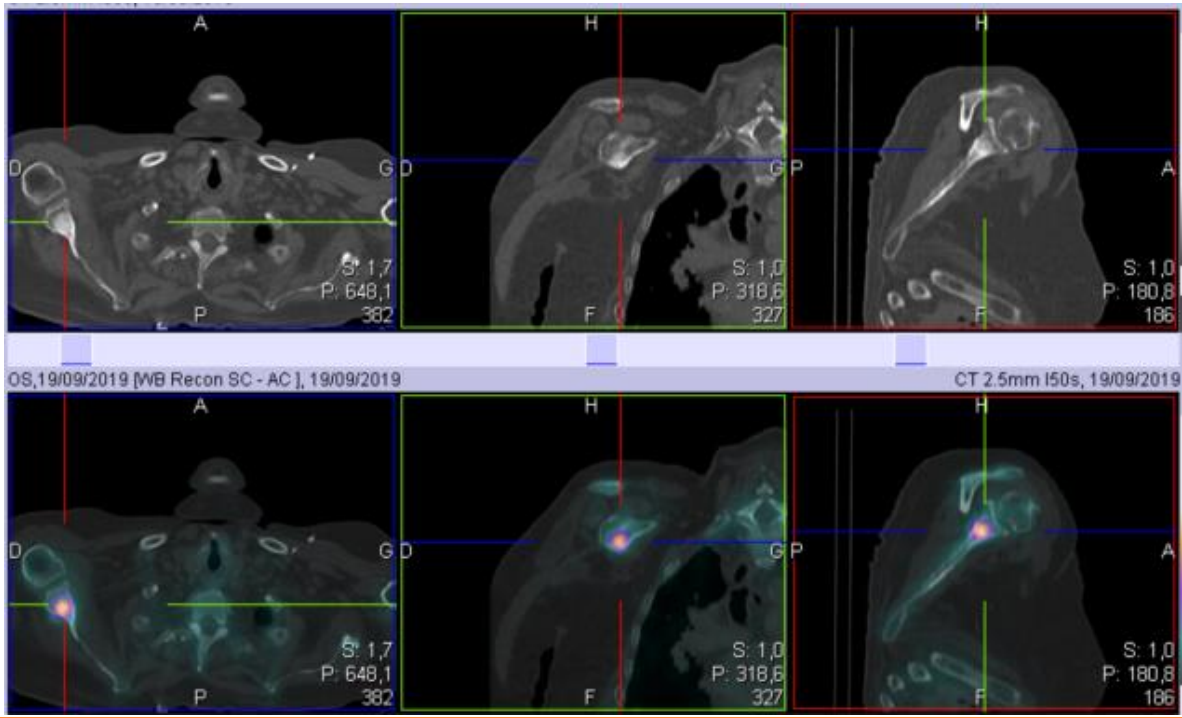
- Stabilité de l'hyperfixation myocardique.
- Hyperfixation suspecte de l'épine iliaque antéro-supérieure gauche

Septembre 2019

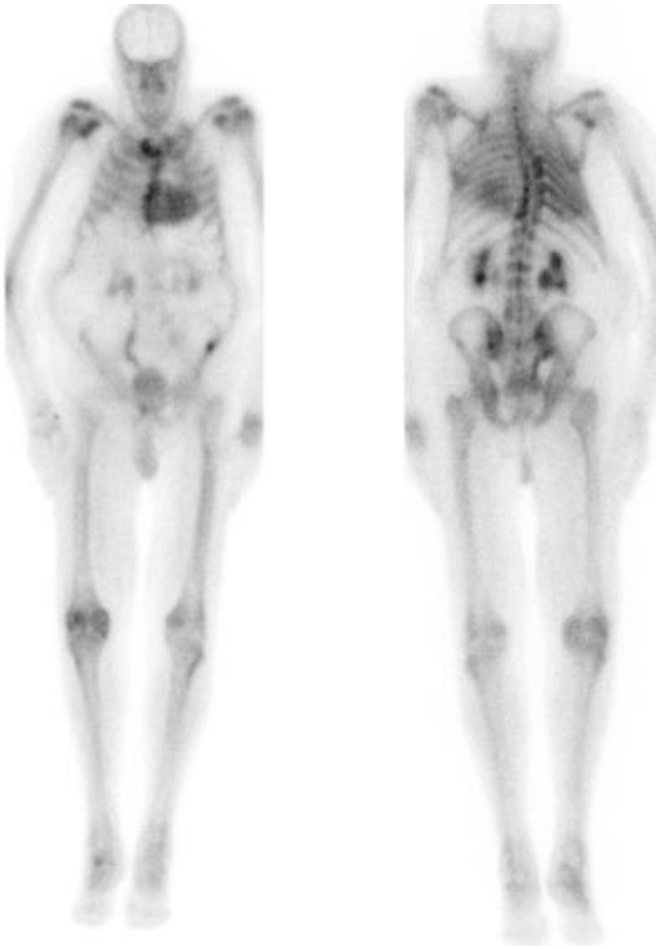
CMH homogène d'allure amyloïde

Recherche d'amylose TTR. Pas de chaine légère ou d'anomalie en immunofixation





Septembre 2019



- Score 2 de Perugini
- Aspect très évocateur d'amylose TTR dans le contexte

- Plusieurs foyers de fixation en regard de lésions osseuses condensées en faveur d'une progression de la pathologie néoplasique prostatique

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Un peu d'histoire

- 1974 : ^{99m}Tc-phosphonates
  - Imagerie du squelette / de l'IDM
- 1975-1977: fixation des parties molles (foie, épaules) correspondant en autoradiographie à des dépôts amyloïdes chez patients suivis pour amylose
- 1976 : fixation myocardique sur cardiomyopathies sans coronaropathie
- 1982 : fixation myocardique suspecte d'AC sur données échographiques chez patients suivis pour amylose
- 1980 : mécanisme :
  - Augmentation de la concentration calcique dans les infiltrations amyloïdes
  - Relation ++ fixation / concentration calcique

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Un peu d'histoire

- 1984 : bonne sensibilité SO (90%)
  - insuffisant pour détection précoce
  - ETT gold-standard
- 1984-1985 : F- amylose AA
- Enthousiasme puis Confusion….
- TTR F+ AL F-
- SO plus précoce que ETT pour TTR
- 2012 : intégration de la SO dans la stratégie >> survie
- 2016 : Gillmore et al
  - Circulation
  - Non biopsy Diagnosis of Cardiac Transthyretin Amyloidosis
  - 1217 patients

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Généralités

- Agrégation de protéines extracellulaires insolubles
- > 30 protéines précurseurs d'amylose
- Peut atteindre tous les organes

Type d'amylose	Amylose AL	Amylose à transthyrétine		Amylose AA
		Héréditaire	Sénile	
Dépots	Chaînes légères d'Ig(Kappa ou Lambda)	Transthyrétine mutée	Transthyrétine sauvage	Protéine inflammatoire
Source	Moelle osseuse	Foie	Foie	Protéine inflammatoire
Organes atteints	Coeur, rein, foie, système nerveux	Système nerveux périphérique, coeur	Coeur	Rein, Foie

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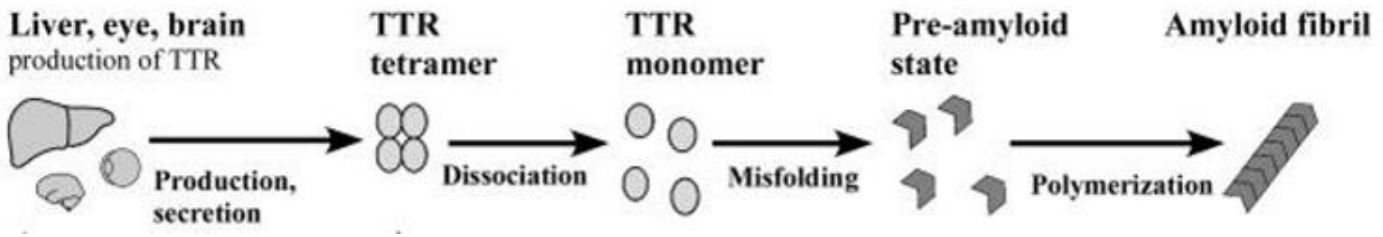
Amylose de la transthyrétine

- TTR : transport T4 et RBP



  - Forme héréditaire
    - 120 mutations différentes avec phénotypes différents
    - Premiers signes vers 30-40 ans, avant 50 ans
    - Rare (5% des amyloses cardiaques)
    - V30m

  - Forme sénile
    - Homme (80% des cas)
    - Dépôts amyloïdes retrouvés à l'autopsie chez 25% des personnes de plus de 85 ans
    - Médiane de survie 75 mois vs 11 mois si AL

- Absence de chaines légères dans le sang
- Traitement ?

Recent advances in transthyretin amyloidosis therapy; Transl Neurodegener. 2014

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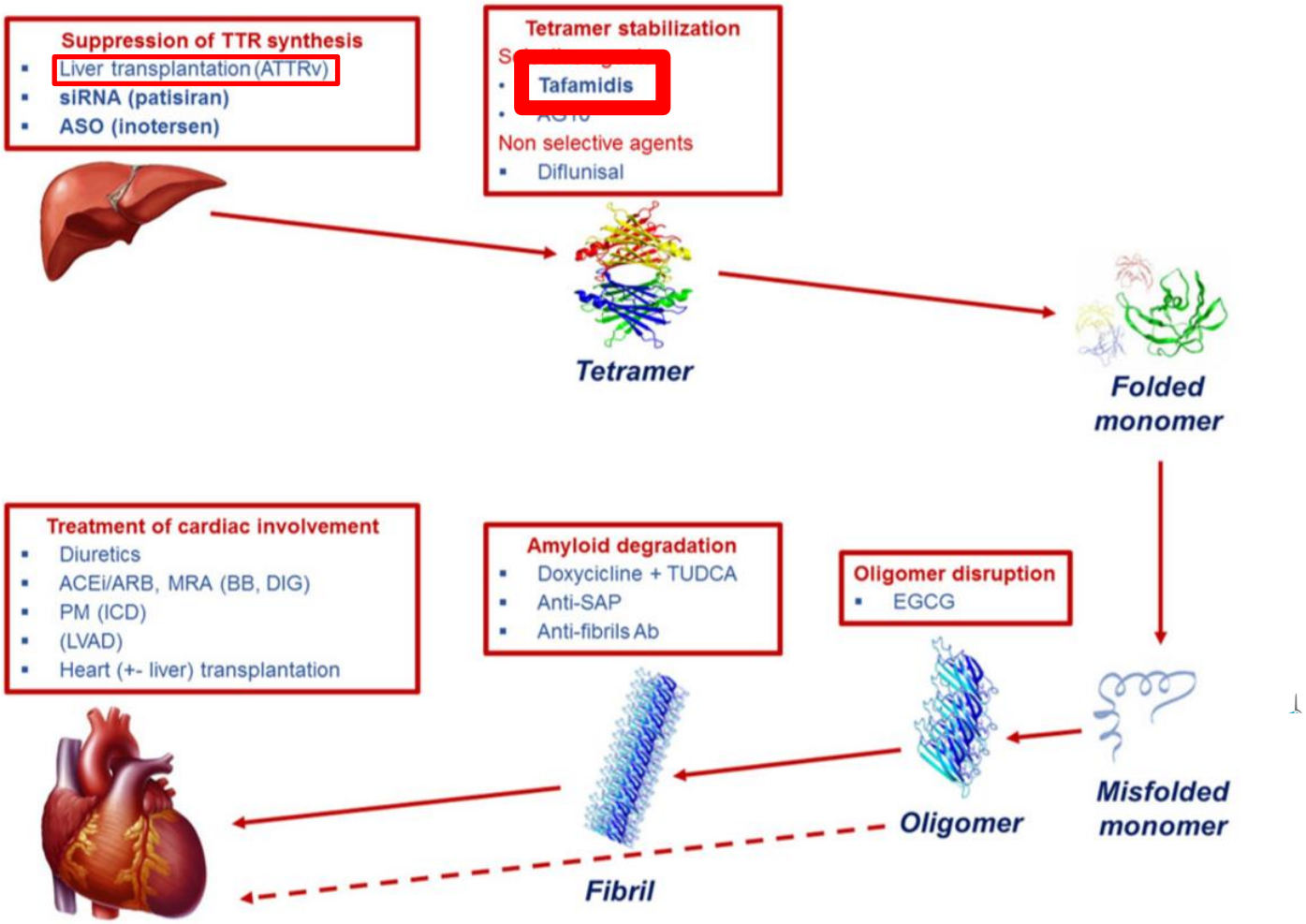
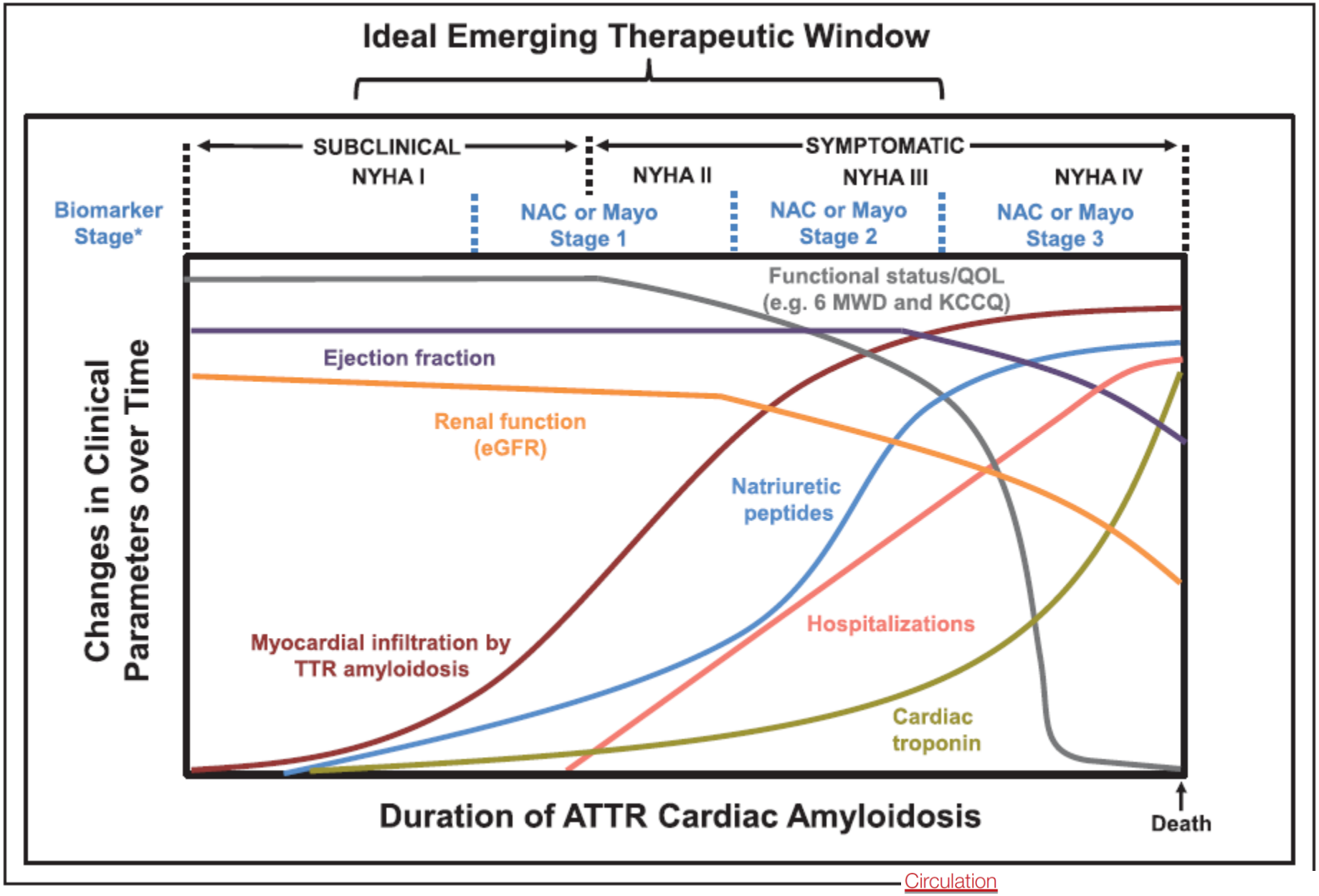
Treatment of cardiac transthyretin amyloidosis: an update

Michele Emdin[1,2*†], Alberto Aimo[3†], Claudio Rapezzi[4], Marianna Fontana[5,6], Federico Perfetto[7,8], Petar M. Seferović[9,10], Andrea Barison[1,2], Vincenzo Castiglione[1,3], Giuseppe Vergaro[1,2], Alberto Giannoni[1,2], Claudio Passino[1,2], and Giampaolo Merlini[11,12]

ESC European Society of Cardiology

European Heart Journal (2019) **0**, 1–10
doi:10.1093/eurheartj/ehz298





Circulation

EDITORIAL

The Truth Is Unfolding About Transthyretin Cardiac Amyloidosis

Evolution

JACC: CARDIOVASCULAR IMAGING
VOL. 7, NO. 5, 2014

Early Identification of Transthyretin-Related Hereditary Cardiac Amyloidosis*

Eloisa Arbustini, MD,† Giampaolo Merlini, MD‡

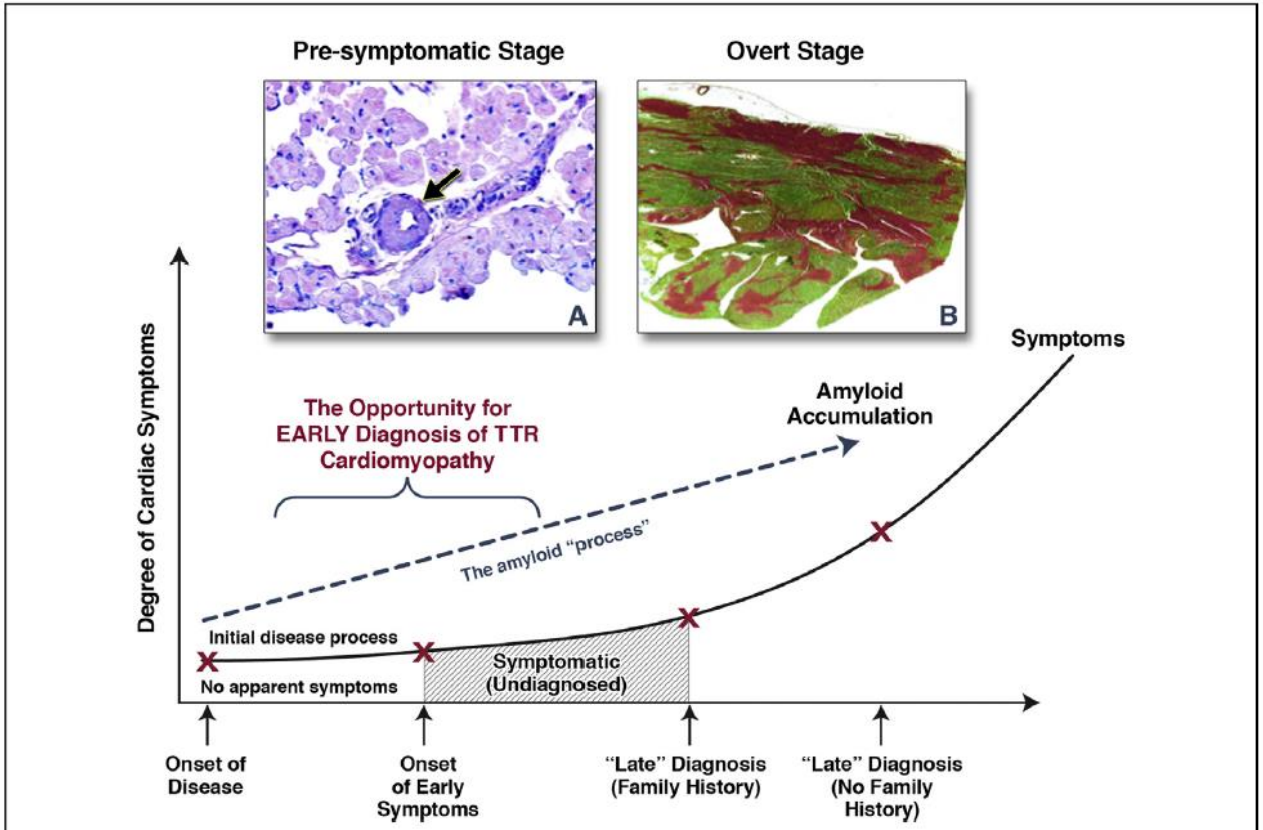


Figure 1. Diagnostic Process in ATTR Amyloidosis

The availability of effective therapies demands early diagnosis to anticipate severe and frequently irreversible end-organ damage (**B,** massive amyloid infiltration, in **green**). Early symptoms are frequently overlooked, and the onset of overt congestive heart failure is associated with end-stage cardiac damage, refractory to therapy. The combination of genetic testing, high-sensitivity troponins, and cardiac imaging techniques allows early diagnosis (**A,** amyloid infiltration of a small intramural vessel), widening the diagnostic windows and, consequently, therapeutic opportunities.

Possible discordance ETT / scinti

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RECOMMANDATION TEMPORAIRE D'UTILISATION (RTU)

PROTOCOLE DE SUIVI DES PATIENTS traités par

VYNDAQEL 20 mg, capsule molle (tafamidis)

dans le

TRAITEMENT DE L'AMYLOSE CARDIAQUE A TRANSTHYRÉTINE

Novembre 2018

Indication de la RTU :

Traitement de l'amylose cardiaque à transthyrétine de forme héréditaire ou sénile, chez les patients adultes présentant une insuffisance cardiaque restrictive de classe NYHA I, II ou III.

Modalités de diagnostic : Selon les données récentes de la littérature[1,2], le **diagnostic** de l'amylose cardiaque à TTR repose sur la combinaison de plusieurs éléments :

- **des signes d'insuffisance cardiaque restrictive** à l'échocardiographie ou l'IRM cardiaque,

Les patients sont généralement âgés de plus de 60 ans, atteints d'hypertrophie ventriculaire gauche concentrique et symptomatiques de classe NYHA II ou III.

- une **scintigraphie osseuse** au Tc99 positive confirmant l'atteinte cardiaque, couplée au dosage des chaines légères libres, à l'électrophorèse des protéines sériques et à la protéinurie de Bence Jones négatifs (afin d'écarter une amylose AL),
- un **test génétique** permettant de caractériser une amylose héréditaire ou sénile.

En dernier lieu, une biopsie extra-cardiaque (salivaire, tissus adipeux,…) ou cardiaque peut confirmer la suspicion d'amylose TTR préétablie par scintigraphie osseuse.

ASNC

EXPERT CONSENSUS RECOMMENDATIONS

ASNC/AHA/ASE/EANM/HFSA/ISA/SCMR/SNMMI EXPERT CONSENSUS RECOMMENDATIONS FOR MULTIMODALITY IMAGING IN CARDIAC AMYLOIDOSIS: PART 2 OF 2—DIAGNOSTIC CRITERIA AND APPROPRIATE UTILIZATION

Table 1. Expert consensus recommendations for diagnosis of cardiac amyloidosis

Criteria for Diagnosis	Subtype
Histological Diagnosis of Cardiac Amyloidosis: Endomyocardial Biopsy*	
1. Endomyocardial biopsy positive for cardiac amyloidosis with Congo red staining with apple-green birefringence under polarized light; typing by immunohistochemistry and/or mass spectrometry at specialized centers	AL, ATTR, Other subtypes
Histological Diagnosis of Cardiac Amyloidosis: Extracardiac Biopsy	
1. ATTR cardiac amyloidosis is diagnosed when below criteria are met: a. Extracardiac biopsy proven ATTR amyloidosis **AND** b. Typical cardiac imaging features (as defined below)	ATTR
2. AL cardiac amyloidosis is diagnosed when below criteria are met: a. Extracardiac biopsy proven AL amyloidosis **AND** b. Typical cardiac imaging features (as defined below) **OR** c. Abnormal cardiac biomarkers: abnormal age-adjusted NT-pro BNP or abnormal Troponin T/I/Hs-Troponin with all other causes for these changes excluded	AL
Clinical Diagnosis of ATTR Cardiac Amyloidosis: ^{99m}Tc-PYP, DPD, HMDP	
3. ATTR cardiac amyloidosis is diagnosed when below criteria are met: a. ^{99m}Tc-PYP, DPD, HMDP Grade 2 or 3 myocardial uptake of radiotracer **AND** b. Absence of a clonal plasma cell process as assessed by serum FLCs and serum and urine immunofixation **AND** c. Typical cardiac imaging features (as defined below)	ATTR
Typical Imaging Features of Cardiac Amyloidosis	
Typical cardiac echo or CMR or PET features: **ANY** of the below imaging features with all other causes for these cardiac manifestations, including hypertension, reasonably excluded.	
1. Echo a. LV wall thickness >12 mm b. Relative apical sparing of global LS ratio (average of apical LS/average of combined mid+basal LS >1) c. ≥ Grade 2 diastolic dysfunction†	ATTR/AL
2. CMR a. LV wall thickness >ULN for sex on SSFP cine CMR b. Global ECV >0.40 c. Diffuse LGE† d. Abnormal gadolinium kinetics typical for amyloidosis, myocardial nulling prior to blood pool nulling	ATTR/AL
3. PET: ^{18}F-florbetapir† or ^{18}F-florbetaben PET† ‡ a. Target to background (LV myocardium to blood pool) ratio >1.5 b. Retention index >0.030 min^{-1}	ATTR/AL

Médecine nucléaire

- Scintigraphie
  - aux phosphonates : PYP, DPD, HMDP
  - ^{123}I-MIBG

- TEP
  - ^{18}FNa
  - ^{18}F-Florbetapir, traceur des plaques amyloides

Scintigraphie aux phosphonates

- Indication sur la charge amyloïde cardiaque

- Quel protocole ?
  - Traceur ?
  - Temps précoce à 5min ? À 10 min
  - Statique ? Corps entier (À 2h, à 3h?) +/- spect-ct?

- Critères d'interprétation ?
  - lecture visuelle?
  - quantification?

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Quel traceur ?

- Pas le MDP
- PYP et DPD les plus répandus et les plus documentés
- Etude sur 1200 pts
  - HMDP = autres traceurs
- HMDP / DPD en phase précoce :
  - Fixation myocardique comparable

Gillmore et al **Diagnosis of Cardiac ATTR Amyloidosis**

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Quel délai ?

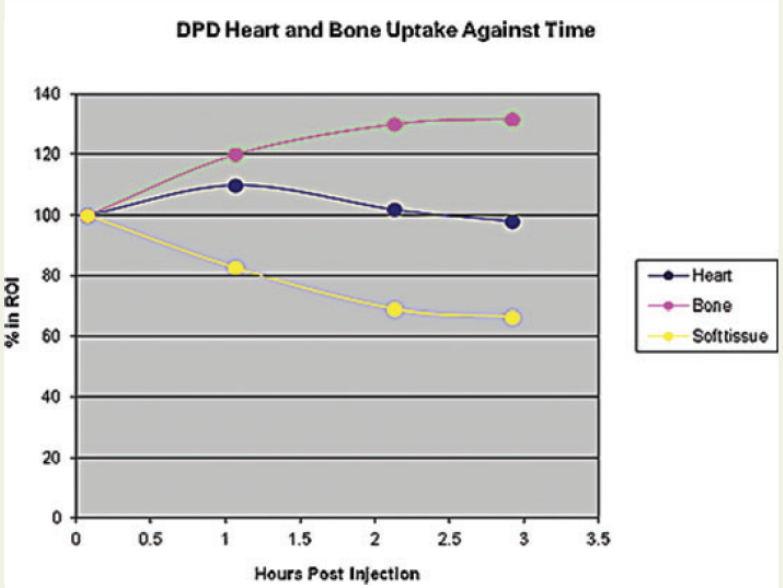

Figure 5 Imaging at multiple time points after ^{99m}Tc-DPD injection showing relative uptake and changing distribution of tracer in a patient with ATTR cardiac amyloidosis. Immediately after injection, tracer is rapidly taken up by heart, skeletal muscle, and bone. Over time, there is a relative gradual decrease in skeletal muscle uptake and progressive increase in bone uptake; cardiac uptake peaks at 1 h post-injection and decreases below baseline at 3 h.

European Heart Journal – Cardiovascular Imaging (2014) **15**, 1289–1298
doi:10.1093/ehjci/jeu107

Utility and limitations of 3,3-diphosphono-1, 2-propanodicarboxylic acid scintigraphy in systemic amyloidosis

David F. Hutt[1], Anne-Marie Quigley[2], Joanne Page[2], Margaret L. Hall[2], Maria Burniston[2], Dorothea Gopaul[1], Thirusha Lane[1], Carol J. Whelan[1], Helen J. Lachmann[1], Julian D. Gillmore[1], Philip N. Hawkins[1], and Ashutosh D. Wechalekar[1]*

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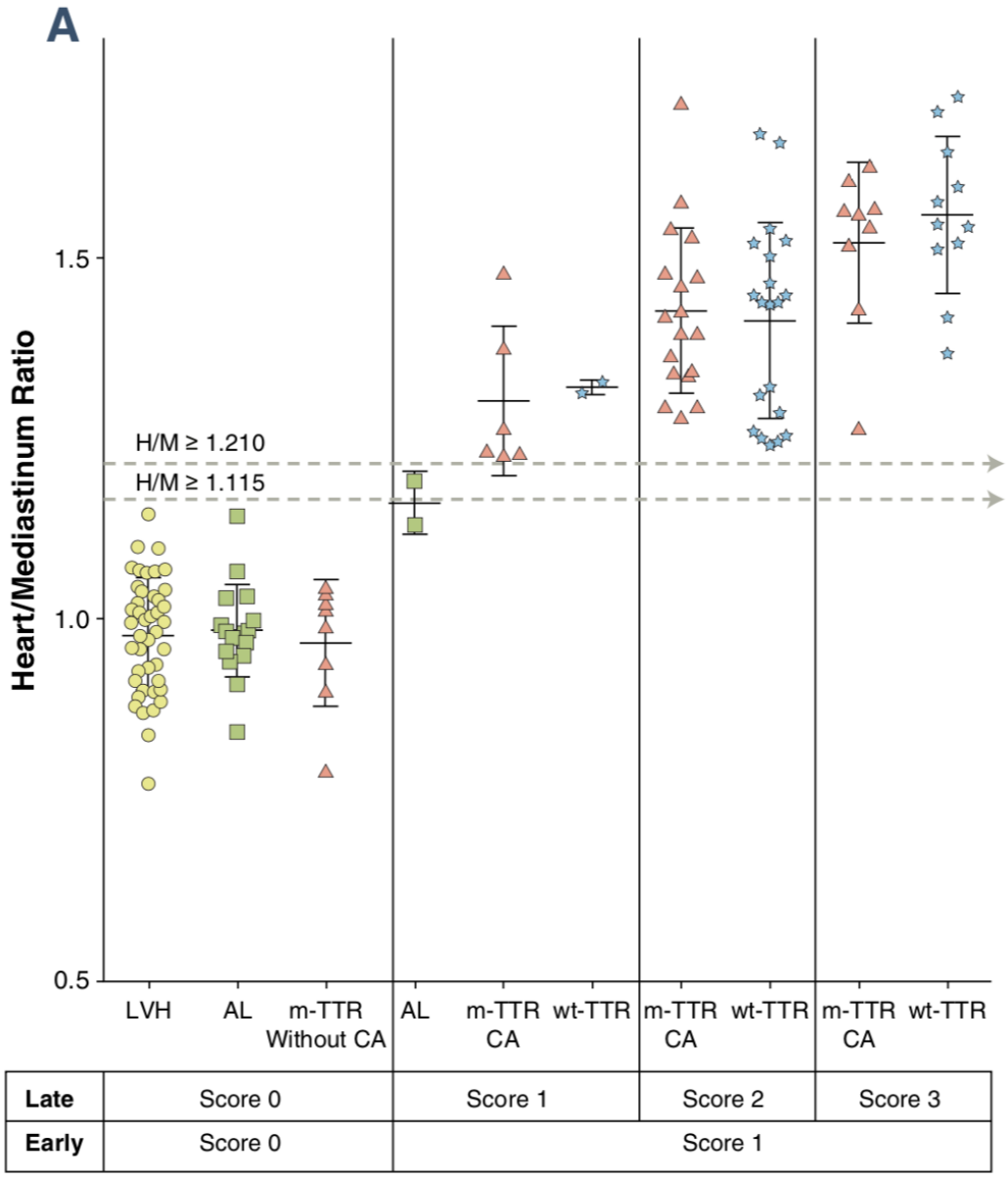
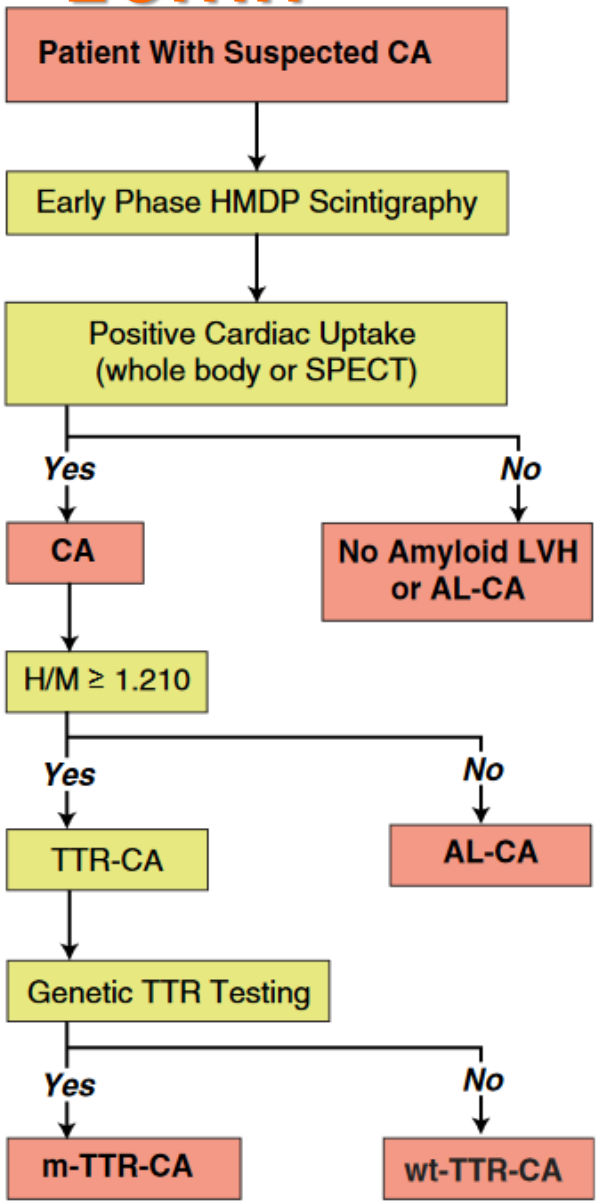
Temps précoce à 10mn



Late	Score 0	Score 1	Score 2	Score 3
Early	Score 0	Score 1		



Galat, A., Van der Gucht, A., Guellich, A., Bodez, D., Cottereau, A.-S., Guendouz, S., … Rosso, J. (2017). Early Phase 99 Tc-HMDP Scintigraphy for the Diagnosis and Typing of Cardiac Amyloidosis. JACC: Cardiovascular Imaging, 10(5), 601–603. doi:10.1016/j.jcmg.2016.05.007

Quelles images ?

- Fixation
  - Myocardique
    - globale/segmentaire
  - Parties molles
  - Squelette
- Acquisition
  - Cliché thoracique planaire
  - Balayage corps entier
  - Spect/ct

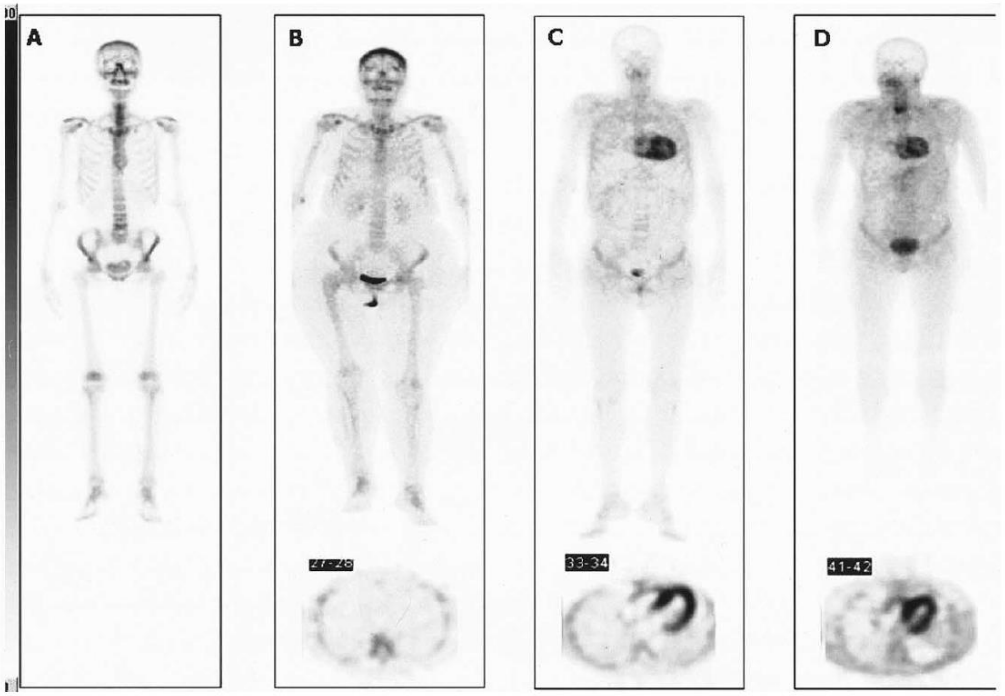


Figure 1. Representative examples illustrating the spectrum of ^{99m}Tc-3,3-diphosphono-1,2-propanodicarboxylic acid (^{99m}Tc-DPD) uptake among patients with transthyretin (TTR)-related or monoclonal immunoglobulin light-chain (AL) cardiac amyloidosis and unaffected controls (**top row** = whole-body scans, anterior view; **bottom row** = cross sectional views of cardiac single-photon emission computed tomography in the same patients). **(A)** Unaffected control subject without visually detectable uptake. **(B)** Patient with AL amyloidosis and echocardiographic documentation of cardiac involvement without any visually detectable sign of myocardial ^{99m}Tc-DPD uptake; mild uptake of the tracer is visible only at the soft tissue level. **(C and D)** Two patients with TTR-related amyloidosis and echocardiographic documentation of cardiac involvement, both showing strong myocardial ^{99m}Tc-DPD uptake (with absent bone uptake); in one of the patients **(D)**, splanchnic uptake is also visible.

Noninvasive Etiologic Diagnosis of Cardiac Amyloidosis Using ^{99m}Tc-3,3-Diphosphono-1,2-Propanodicarboxylic Acid Scintigraphy

Enrica Perugini, MD,* Pier Luigi Guidalotti, MD,† Fabrizio Salvi, MD,‡ Robin M. T. Cooke, MA,* Cinzia Pettinato, MD,† Letizia Riva, MD,* Ornella Leone, MD,§ Mohsen Farsad, MD,† Paolo Ciliberti, MD,* Letizia Bacchi-Reggiani, MSc, MBIOSTAT,* Francesco Fallani, MD,* Angelo Branzi, MD,* Claudio Rapezzi, MD*

Journal of the American College of Cardiology

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SPECT/CT

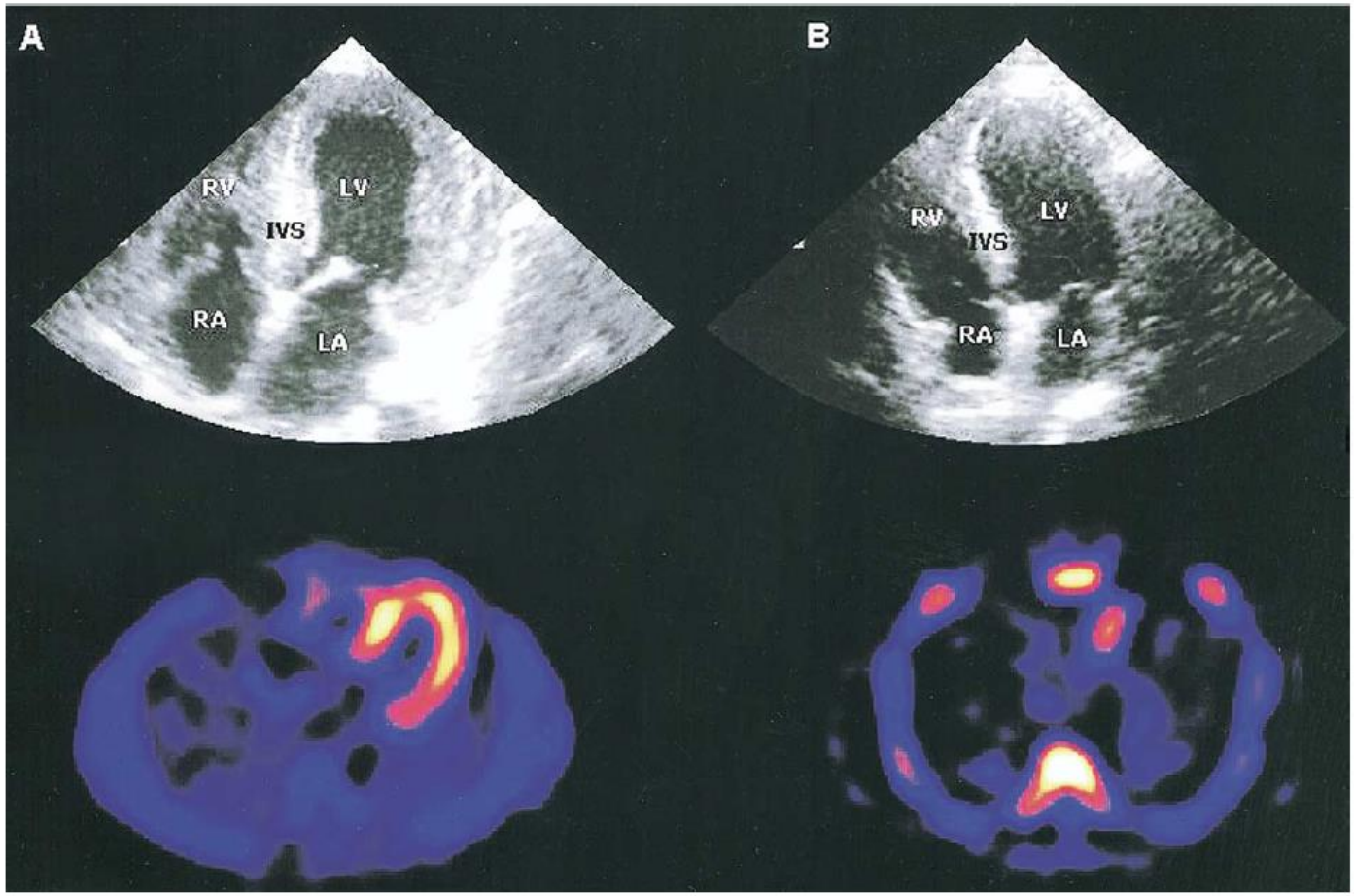
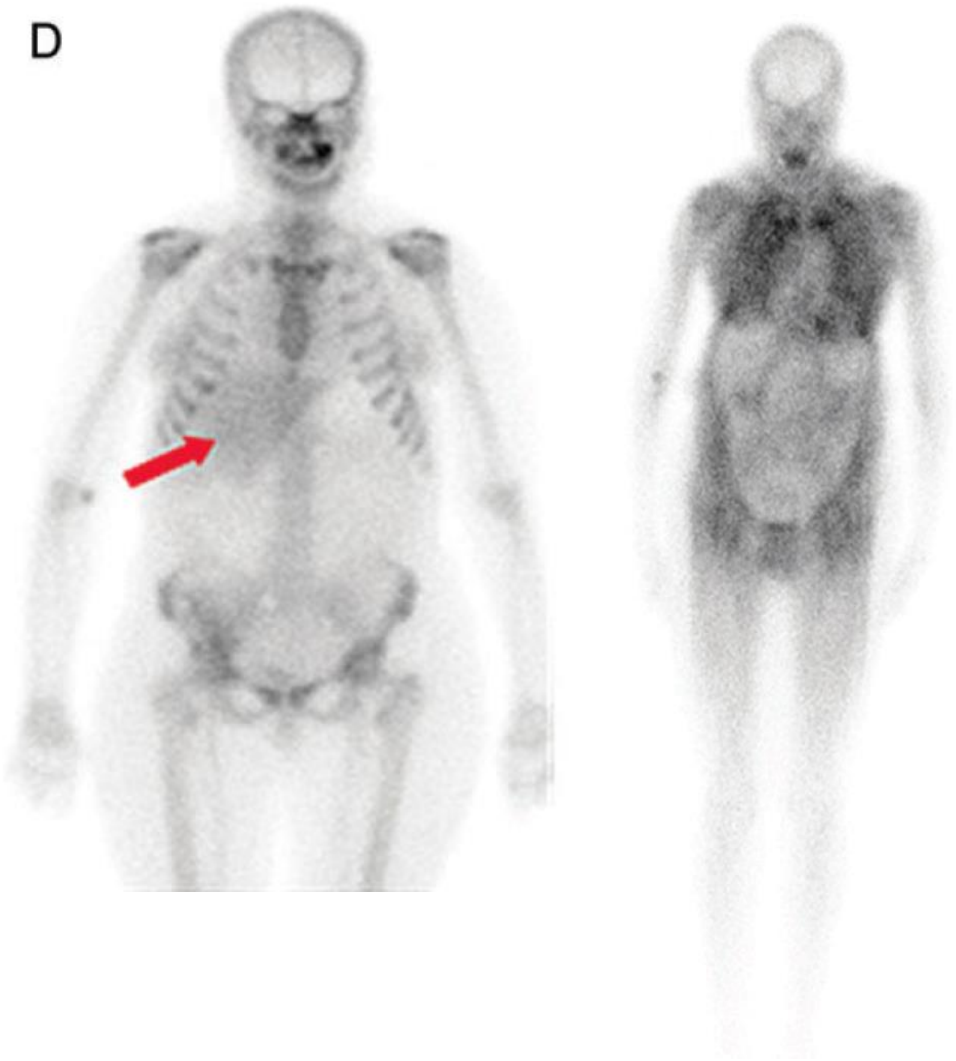


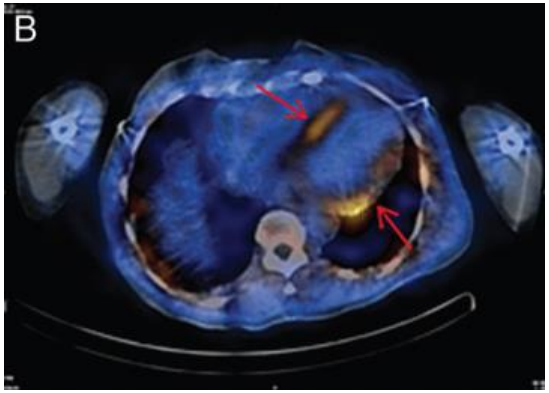
Figure 4. Apical four-chamber-view echocardiograms **(top row)** and cross-sectional views of cardiac single-photon emission computed tomography (SPECT) **(bottom row)** in two patients with transthyretin (TTR)-related cardiac amyloidosis. Topographic correspondences between increased left ventricular parietal thickness and ^{99m}Tc-3,3-diphosphono-1,2-propanodicarboxylic acid (^{99m}Tc-DPD) uptake is evident: **(A)** increased thickness of both the interventricular septum and the lateral wall of the left ventricle at echocardiography in correspondence with diffuse myocardial ^{99m}Tc-DPD uptake at SPECT; **(B)** increased thickness of the medial portion of the interventricular septum (with normal lateral wall) at echocardiography in correspondence with localized myocardial ^{99m}Tc-DPD uptake at the same level. IVS = intraventricular septum; LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

Cas particuliers

- Faux-positifs
  - Infarctus récent

- Faux-négatifs TTR
  - Certaines formes rares mutation non V30M
  - Certaines mutations V30M à début précoce et à fibrilles TTR full-length (type B)





Possible aspect photopénique du foie en rapport avec la congestion HSM liée à IC chronique

Grade 1 possible même si planaire < 0 spect-ct ++

European Heart Journal – Cardiovascular Imaging (2014) **15**, 1289–1298
doi:10.1093/ehjci/jeu107

Utility and limitations of 3,3-diphosphono-1, 2-propanodicarboxylic acid scintigraphy in systemic amyloidosis

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Score visuel de Perugini

- *Score 0 :* absence de fixation cardiaque et fixation osseuse normale
- *Score 1 :* fixation cardiaque légère, inférieure à la fixation osseuse
- *Score 2 :* fixation cardiaque modérée accompagnée d'une diminution de la fixation osseuse
- *Score 3 :* fixation cardiaque intense avec fixation osseuse légère ou absente

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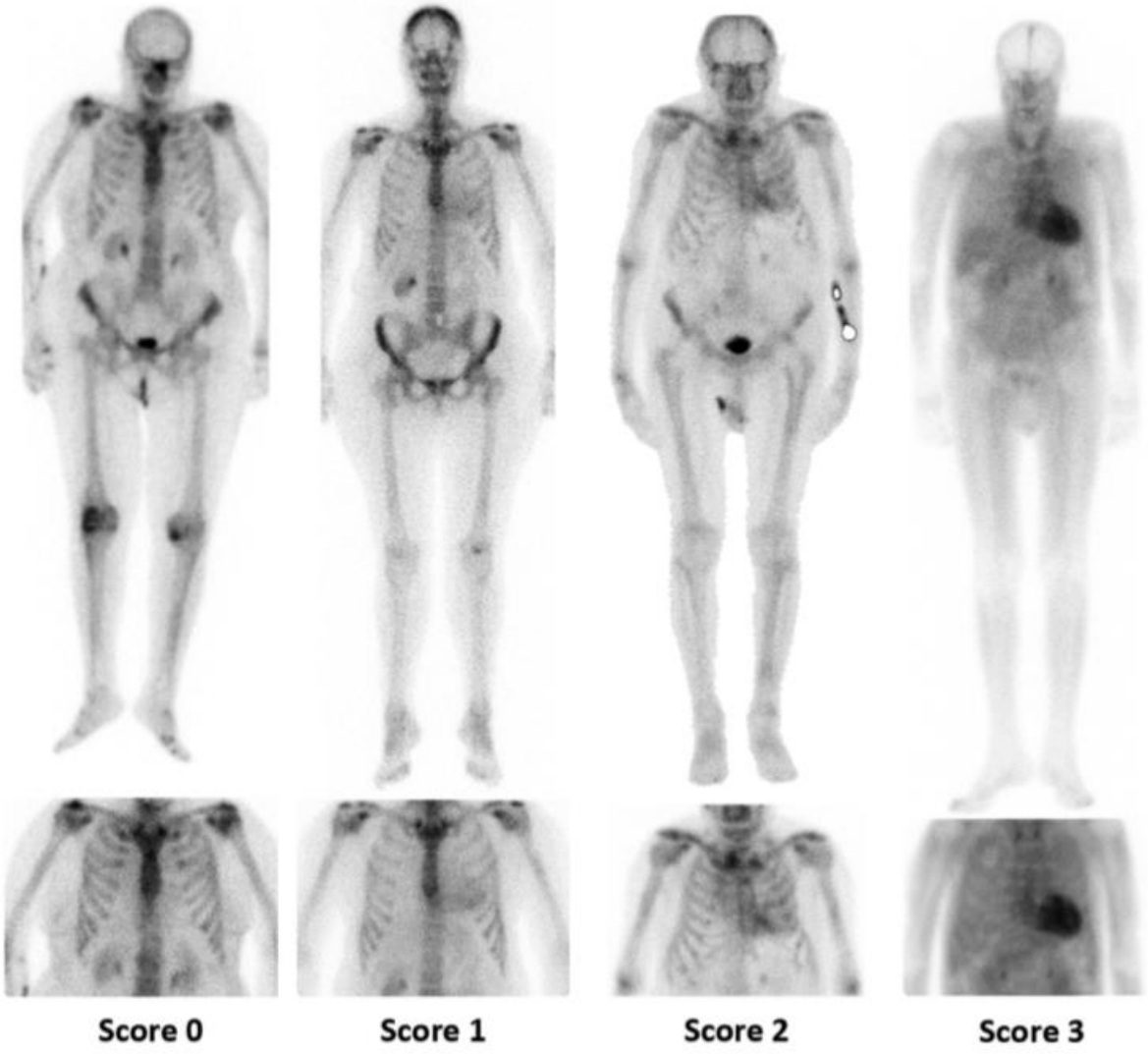
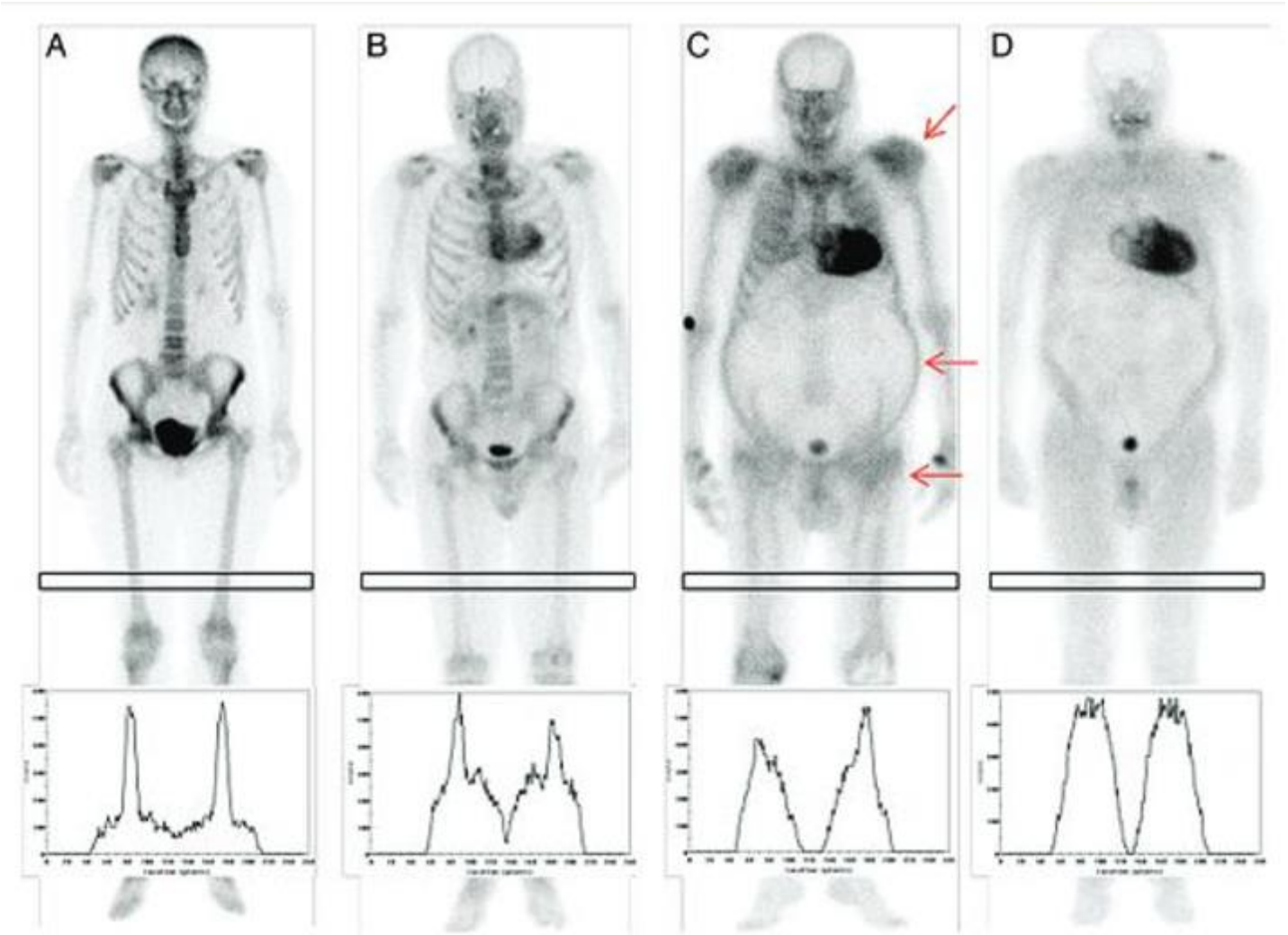
Bone scintigraphy for cardiac amyloidosis imaging: Past, present and future

La scintigraphie osseuse dans l'amylose cardiaque : passé, présent, futur

O. Lairez [a,b,c,d,*], P. Pascal [a,b], G. Victor [a,d], D. Bastié [a,c], Y. Lavie-Badie [a,b,c], A. Pierre [a,e], E. Cassol [a,d], I. Berry [a,c,d]





David F. Hutt : Utility and limitations of DPD scintigraphy in systemic amyloidosis
Eur Heart J Cardiovasc Imaging 2014

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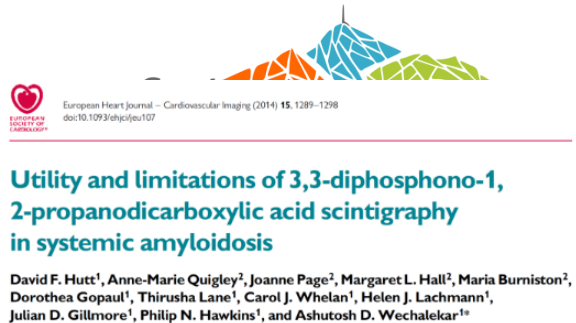
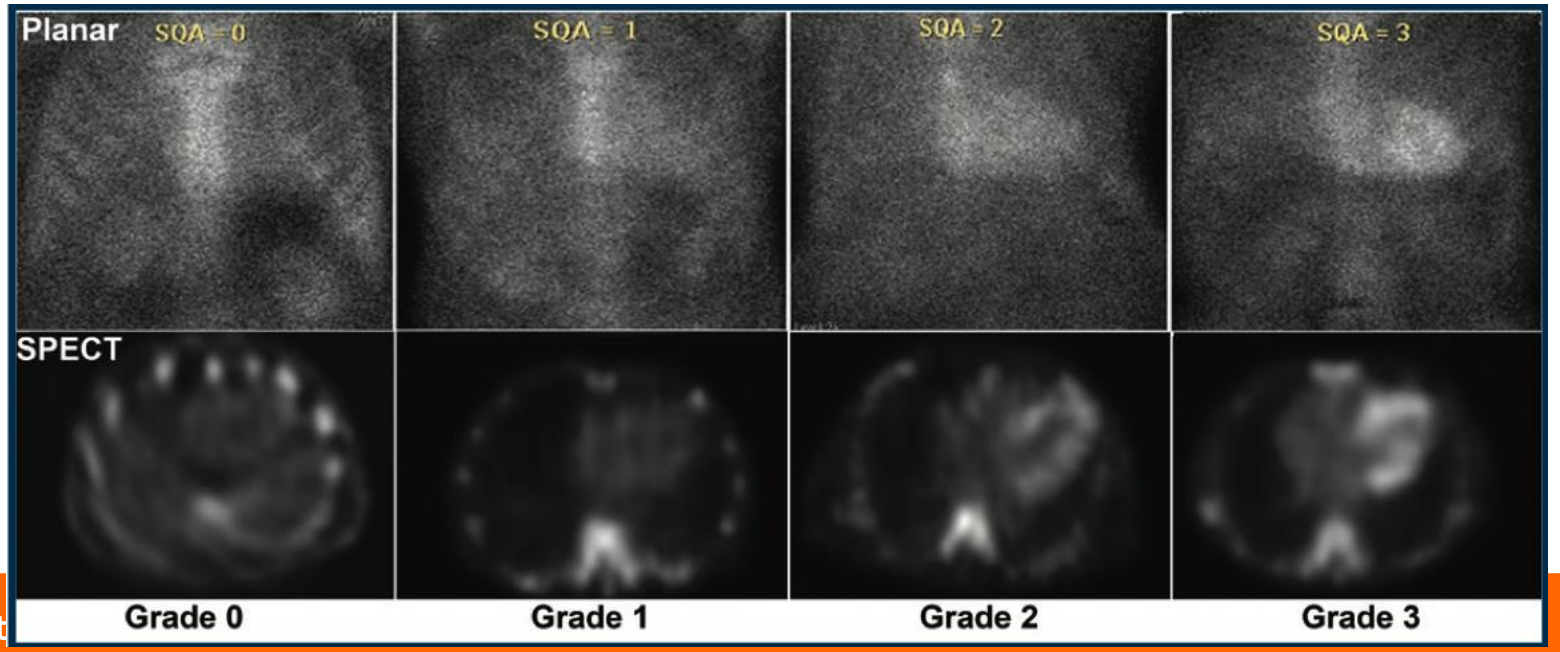
Revised grading system

- Grade 0 — no visible myocardial uptake in either the planar or cardiac SPECT-CT scan;
- Grade 1 — cardiac uptake seen only by SPECT-CT, or minimal cardiac uptake (less intense than bones) evident on the planar scan and with no apparent reduction in intensity of the normal bone images;
- Grade 2 — moderate cardiac uptake, greater in intensity than the bone uptake with apparent reduction of the latter on planar imaging;
- Grade 3 — intense cardiac uptake with little or no bone uptake visualised on planar imaging;
- Grade 4 — intense soft-tissue uptake partly or completely obscuring cardiac uptake on planar imaging



Grade	Myocardial ^{99m}Tc-PYP Uptake
Grade 0	no uptake and normal bone uptake
Grade 1	uptake less than rib uptake
Grade 2	uptake equal to rib uptake
Grade 3	uptake greater than rib uptake with mild/ absent rib uptake



Eur J Nucl Med Mol Imaging (2011) 38:470–478
DOI 10.1007/s00259-010-1642-7

ORIGINAL ARTICLE

Usefulness and limitations of ^{99m}Tc-3,3-diphosphono-1, 2-propanodicarboxylic acid scintigraphy in the aetiological diagnosis of amyloidotic cardiomyopathy

Claudio Rapezzi · Candida Cristina Quarta · Pier Luigi Guidalotti · Simone Longhi · Cinzia Pettinato · Ornella Leone · Alessandra Ferlini · Fabrizio Salvi · Pamela Gallo · Christian Gagliardi · Angelo Branzi

Méta analyse pour un Score ≥ 2 :

-> Pas de différence significative entre les traceurs

99mTc-DPD : **Se : 94.6%/**Sp : 88.4%

99mTc-HMDP: Se : 85.7% **Sp : 97.5%**

Globale : Se : 92.2% / Sp : 95,4%

6 études, 529 patients avec biopsie myocardique

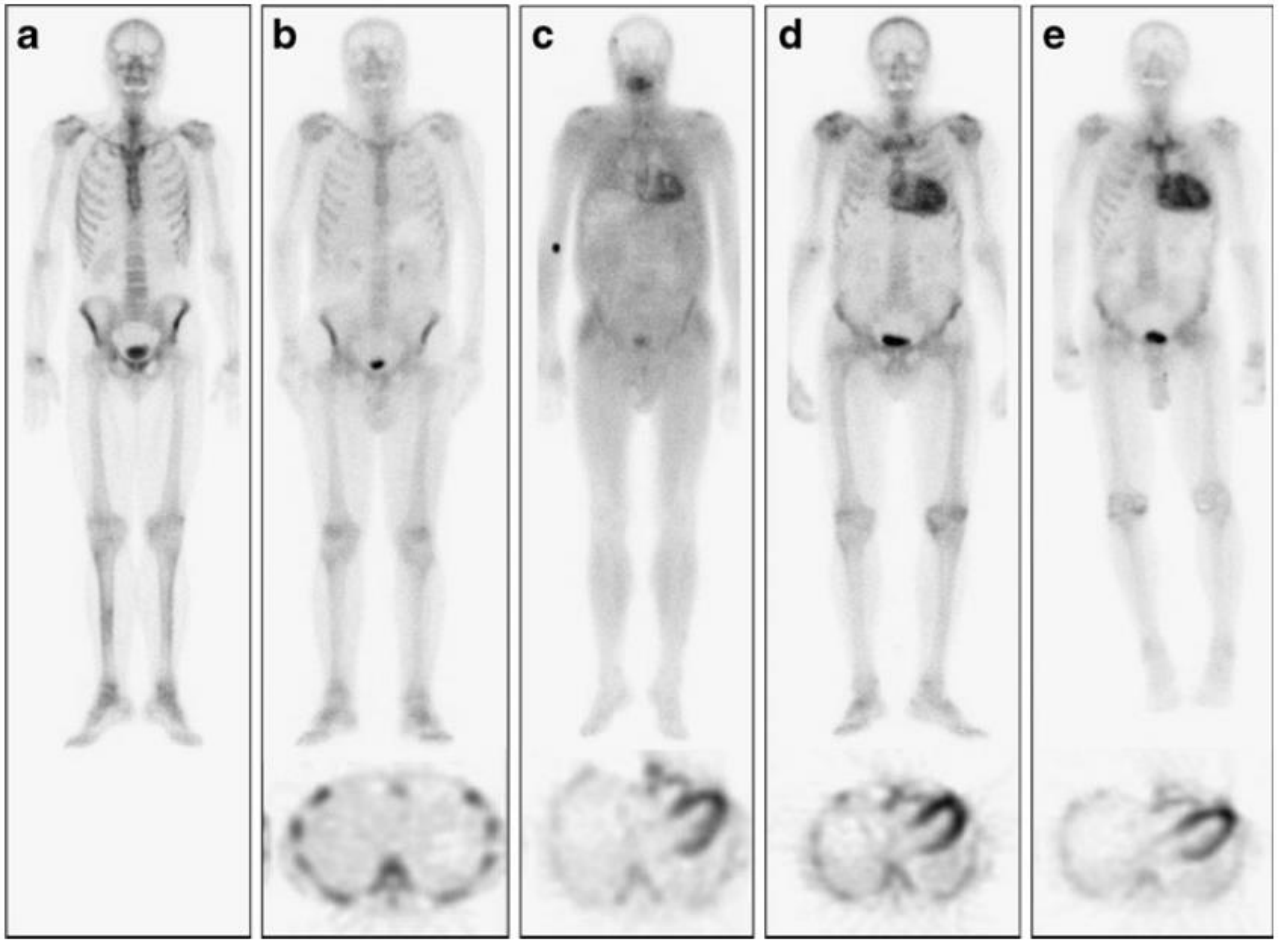


Fig. 1 Examples illustrating the spectrum of ^{99m}Tc-DPD uptake among patients with TTR-related or AL AC and unaffected controls (*top row* whole-body scans, anterior view; *bottom row* cross-sectional views of cardiac SPECT in the same patients). **a** Unaffected control subject without visually detectable uptake. **b** Patient with AL amyloidosis and echocardiographic documentation of cardiac involvement without any visually detectable sign of myocardial tracer uptake; mild uptake of ^{99m}Tc-DPD is visible only at the soft tissue level. **c** Patient with AL amyloidosis and echocardiographic documentation of cardiac involvement, who shows a moderate myocardial tracer uptake along with mild uptake at the soft tissue level. **d**, **e** Two patients with TTR-related amyloidosis (patient in **d** affected by ATTR, patient in **e** affected by SSA) and echocardiographic documentation of AC, both showing strong myocardial ^{99m}Tc-DPD uptake (and attenuated bone uptake)

Treglia, G., Glaudemans, A. W. J. M., Bertagna, F., Hazenberg, B. P. C., Erba, P. A., Giubbini, R., … Slart, R. H. J. A. (2018). Diagnostic accuracy of bone scintigraphy in the assessment of cardiac transthyretin-related amyloidosis: a bivariate meta-analysis. European Journal of Nuclear Medicine and Molecular Imaging. doi:10.1007/s00259-018-4013-4

Quantification

- Heart / WB (Whole body scan retention)
- H / M (Médiastin)
- H / CL (Controlatéral)

Quantification

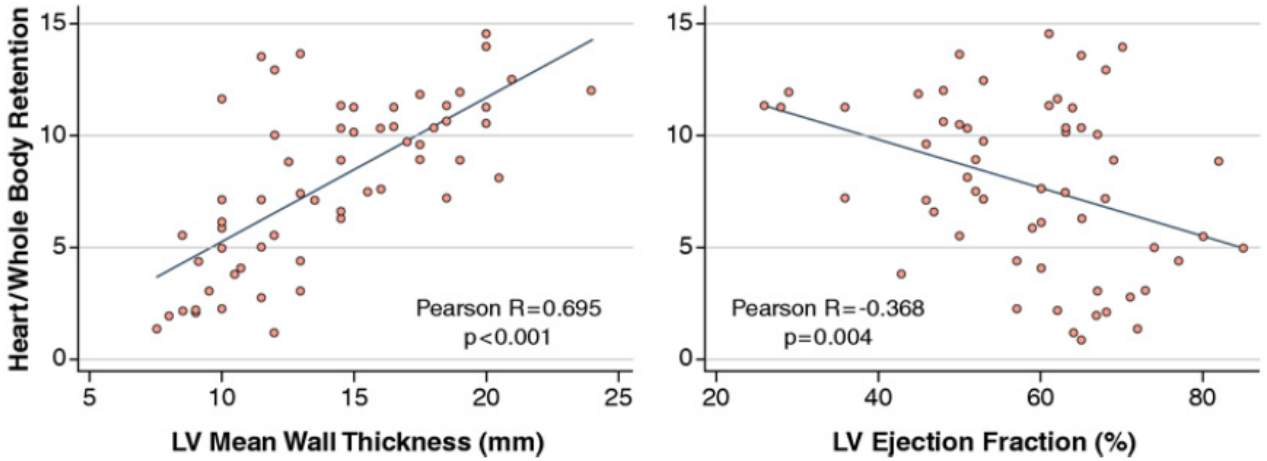


Figure 3. Correlation Between H/WB and Mean LV Wall Thickness and LV Ejection at Pearson's Correlation Test

Mean left ventricular (LV) wall thickness (**left**), and LV ejection (**right**). The heart/whole body retention (H/WB) was positively correlated with LV mean wall thickness and inversely correlated with LV ejection fraction.

Fixation corrélée à l'importance de l'infiltration amyloïde ++

JACC: CARDIOVASCULAR IMAGING
© 2011 BY THE AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION
PUBLISHED BY ELSEVIER INC.
VOL. 4, NO. 6, 2011
ISSN 1936-878X/$36.00
DOI:10.1016/j.jcmg.2011.03.016

Role of ^{99m}Tc-DPD Scintigraphy in Diagnosis and Prognosis of Hereditary Transthyretin-Related Cardiac Amyloidosis

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Bologna and Ferrara, Italy

Quantification

1. An overall interpretation of the findings into categories of 1) not suggestive of TTR amyloidosis; 2) strongly suggestive of TTR amyloidosis or 3) equivocal for TTR amyloidosis

a. Not suggestive: A semi-quantitative visual score of 0 or H/CL ratio < 1.

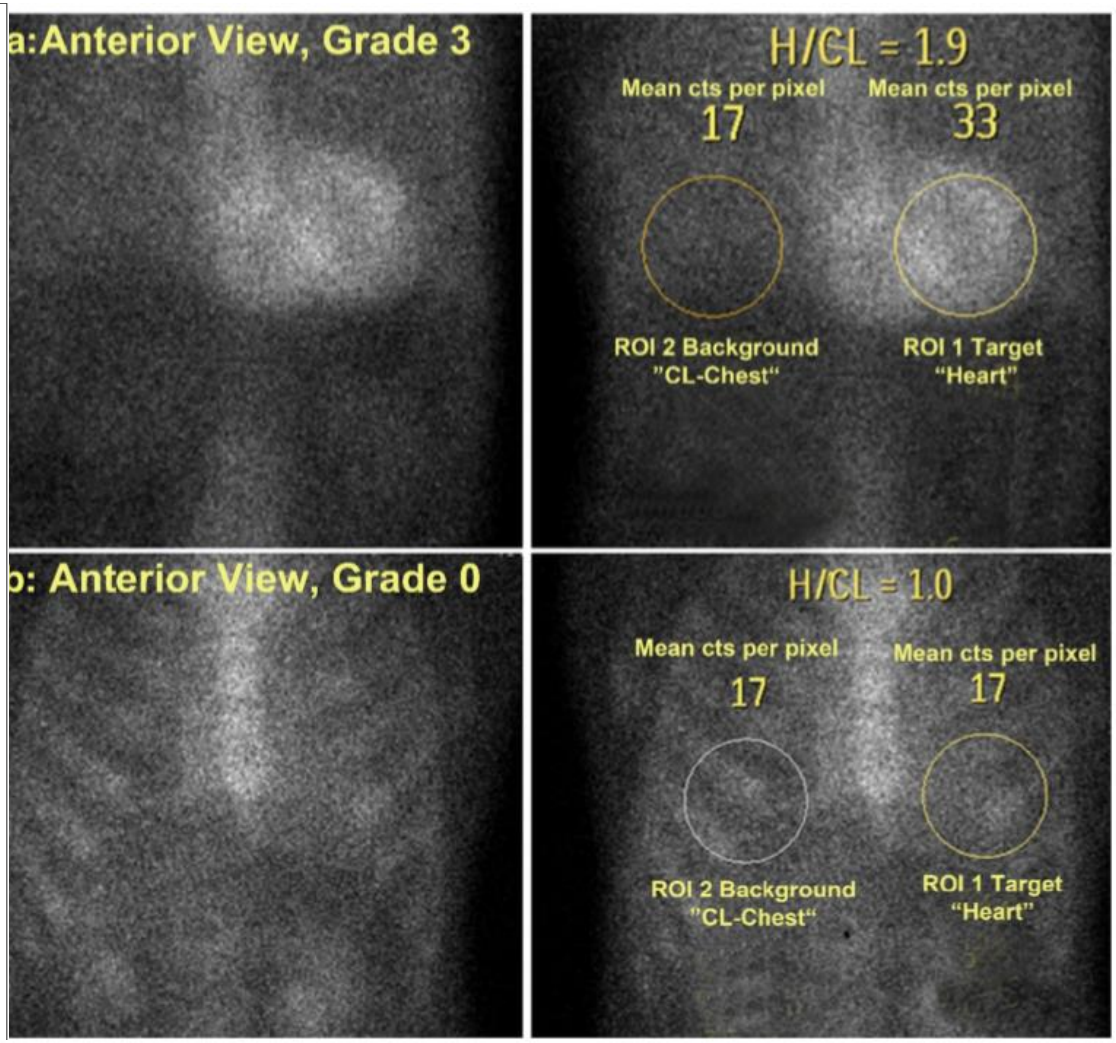
b. Strongly suggestive: A semi-quantitative visual score of 2 or 3 or H/CL ratio >1.5

c. Equivocal: A semi-quantitative visual score of 1 or H/CL ratio 1-1.5

2. Interpret the results in the context of prior evaluation

a. If echo/CMR are strongly positive, and ^{99m}Tc PYP negative, consider further evaluation including endomyocardial biopsy

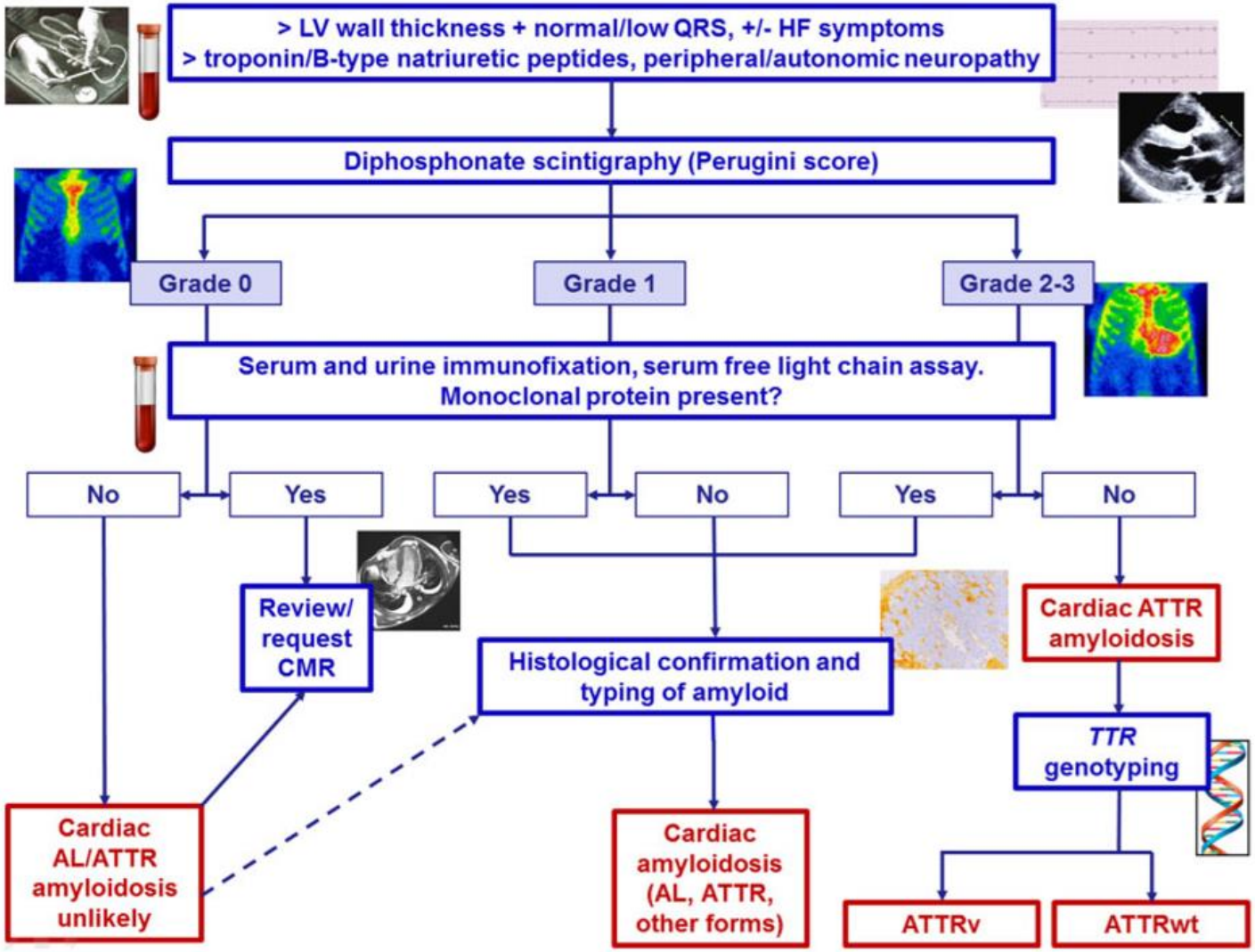
Of note: A negative or mildly positive PYP does not exclude AL amyloid. In addition, equivocal results could represent AL amyloid or early TTR amyloid



Treatment of cardiac transthyretin amyloidosis: an update

ESC European Society of Cardiology
European Heart Journal (2019) **0**, 1–10
doi:10.1093/eurheartj/ehz298

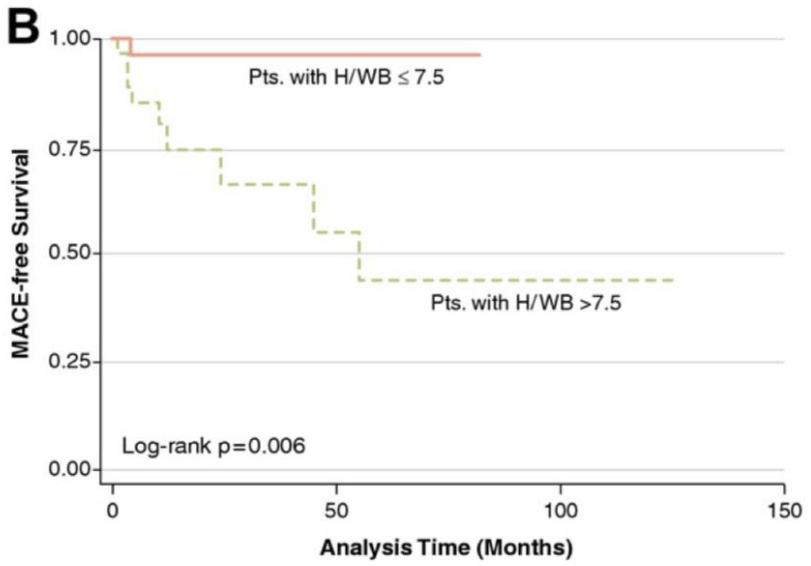
Michele Emdin[1,2*†], Alberto Aimo[3†], Claudio Rapezzi[4], Marianna Fontana[5,6], Federico Perfetto[7,8], Petar M. Seferović[9,10], Andrea Barison[1,2], Vincenzo Castiglione[1,3], Giuseppe Vergaro[1,2], Alberto Giannoni[1,2], Claudio Passino[1,2], and Giampaolo Merlini[11,12]

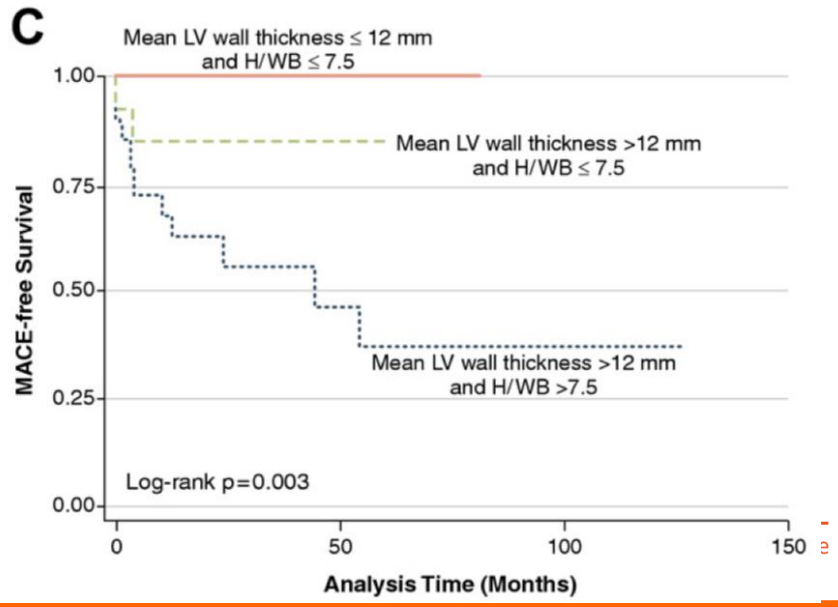


Pronostic

- Heart/whole body > 7.5 risque majoré de MACE

- MACE : Major Adverse Cardiac Events:
  - Mort de causes cardiovasculaires
  - Hospitalisation pour IC
  - BAV 3
  - FA/Flutter
  - SCA





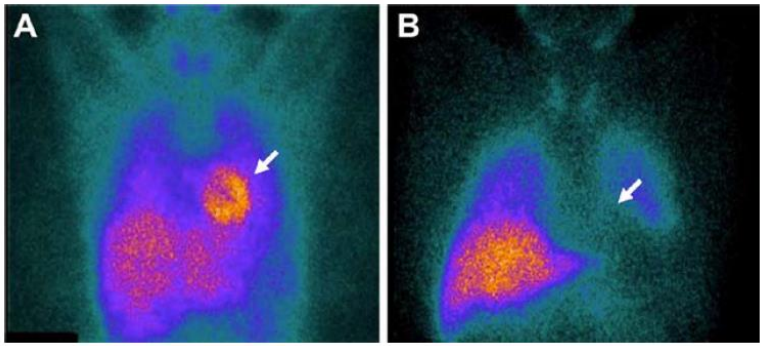
Rapezzi, C., Quarta, C. C., Guidalotti, P. L., Pettinato, C., Fanti, S., Leone, O., … Salvi, F. (2011). Role of 99mTc-DPD Scintigraphy in Diagnosis and Prognosis of Hereditary Transthyretin-Related Cardiac Amyloidosis. JACC: Cardiovascular Imaging, 4(6), 659–670. doi:10.1016/j.jcmg.2011.03.016

MIBG

- Analogue norépinéphrine
- Activité adrénergique
- Evaluation du rapport H/M tardif
- Quantification visuelle de la dénervation (AHA 17)

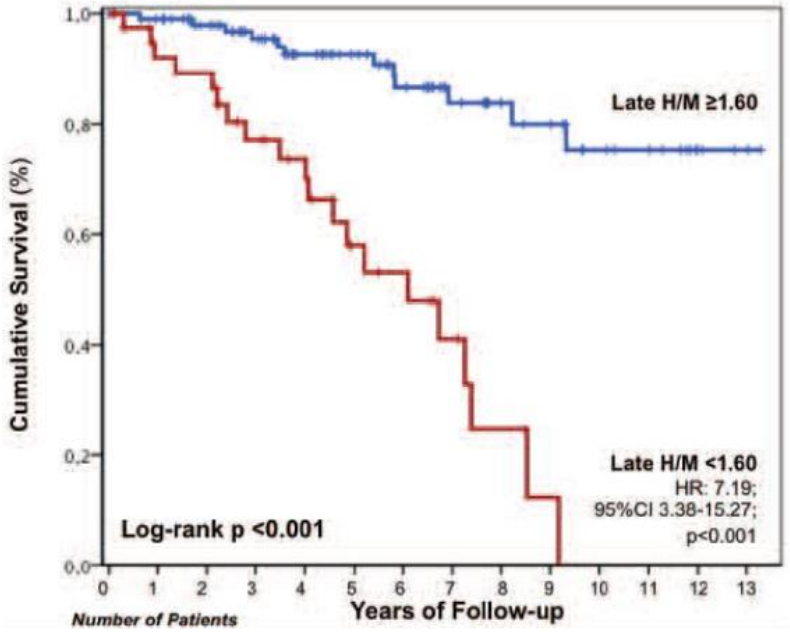
MIBG

- V30M TTR mutation
- Dénervation potentiellement plus précoce que HVG ou signes cliniques
- H/M corrélé à sévérité de polyneuropathie
- Valeur pronostique de la MIBG



H/M 2,5

H/M 1,2



Coutinho et al, Circ CV Imaging 2013

Autres traceurs TEP

- Traceurs de plaques amyloïdes
- ^{18}FNa

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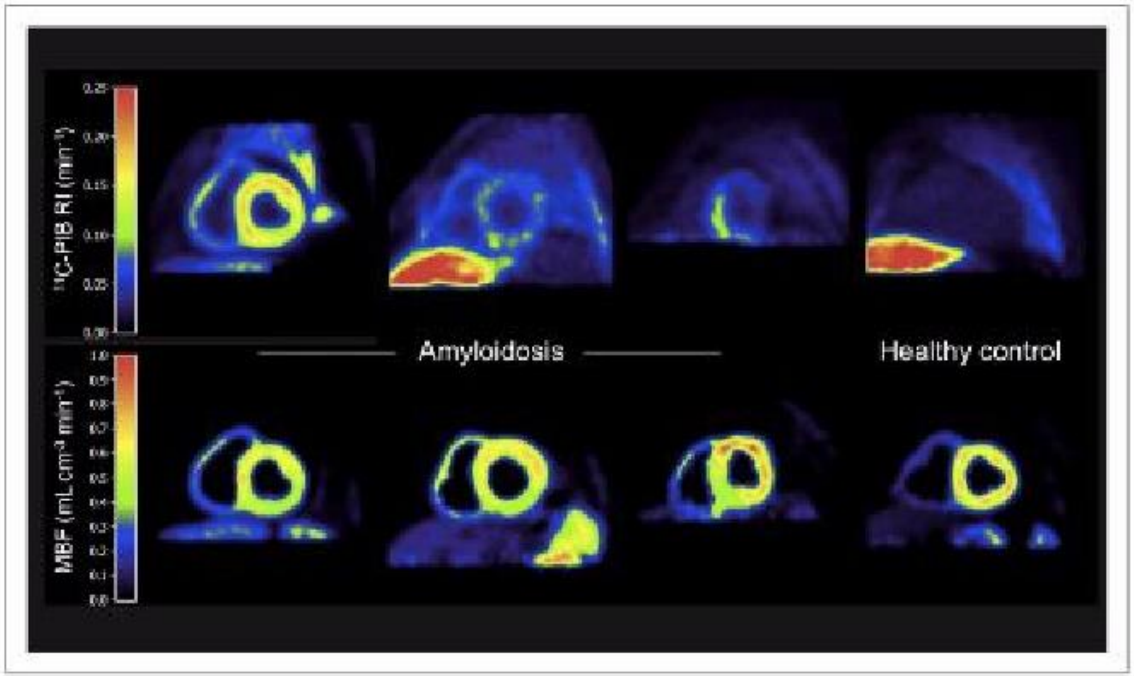
TEP 11C-PIB



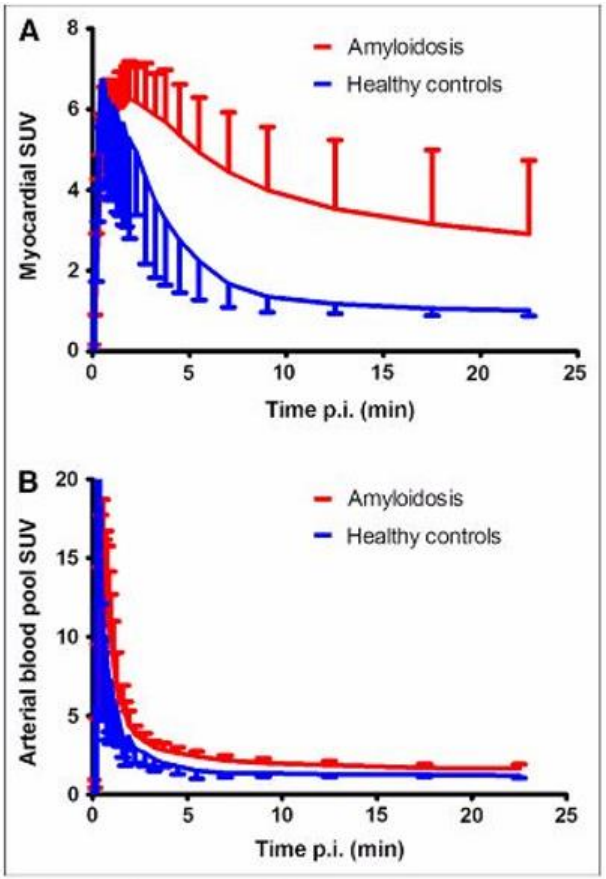


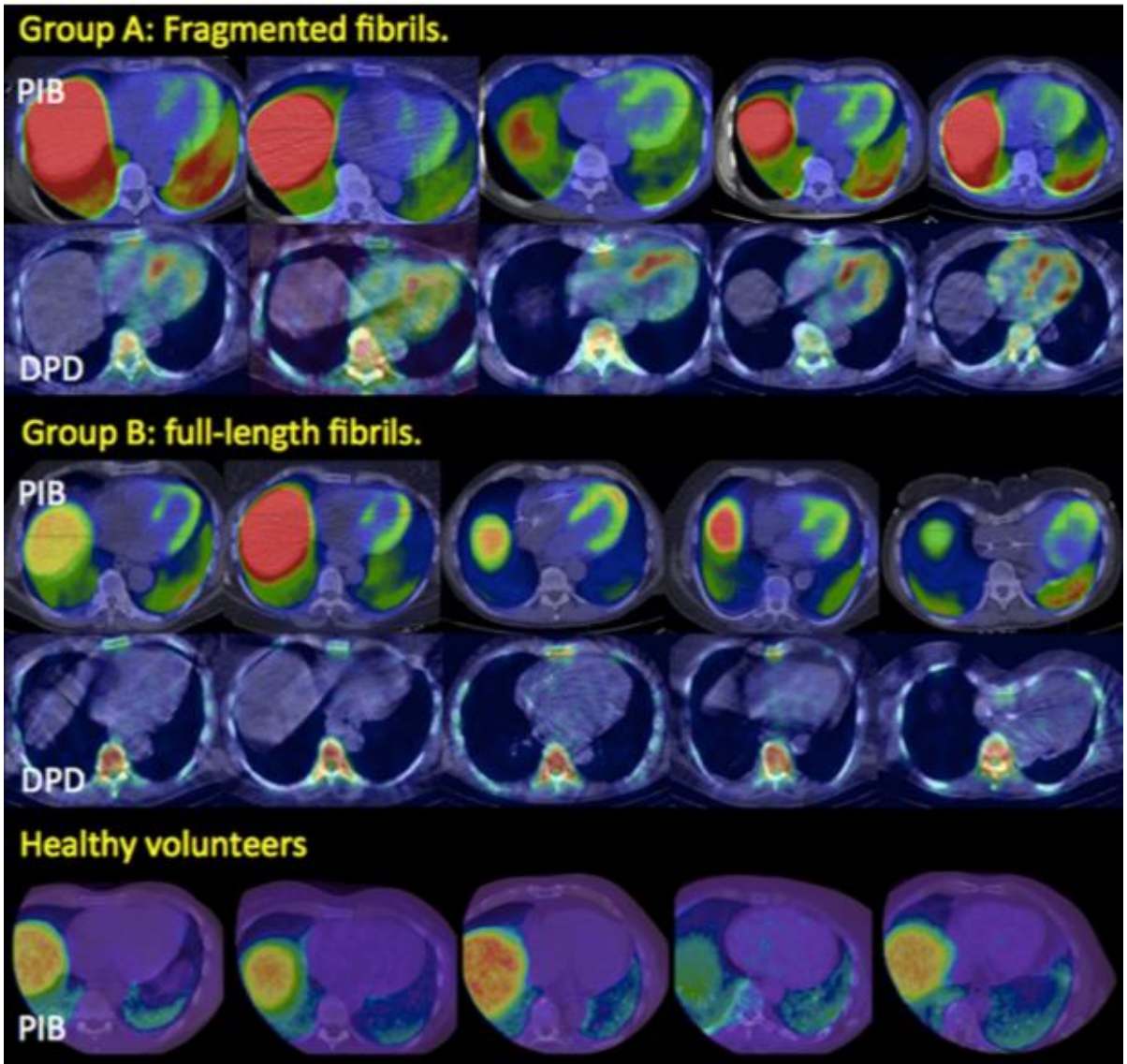
FIGURE 1. Mean standardized uptake value vs. time curves of ^{11}C-PIB in whole myocardium (A) and arterial blood (B) for amyloidosis patients (red) and healthy controls (blue). Error bars represent 95% confidence intervals. p.i. = after injection; SUV = standardized uptake value.

Antoni G et al J Nucl Med 2013
In Vivo visualization of amyloid deposits in the heart with 11c-PIB and pet

Positron emission tomography (PET) utilizing Pittsburgh compound B (PIB) for detection of amyloid heart deposits in hereditary transthyretin amyloidosis (ATTR)

Journal of Nuclear Cardiology®
Volume 25, Number 1;240–8

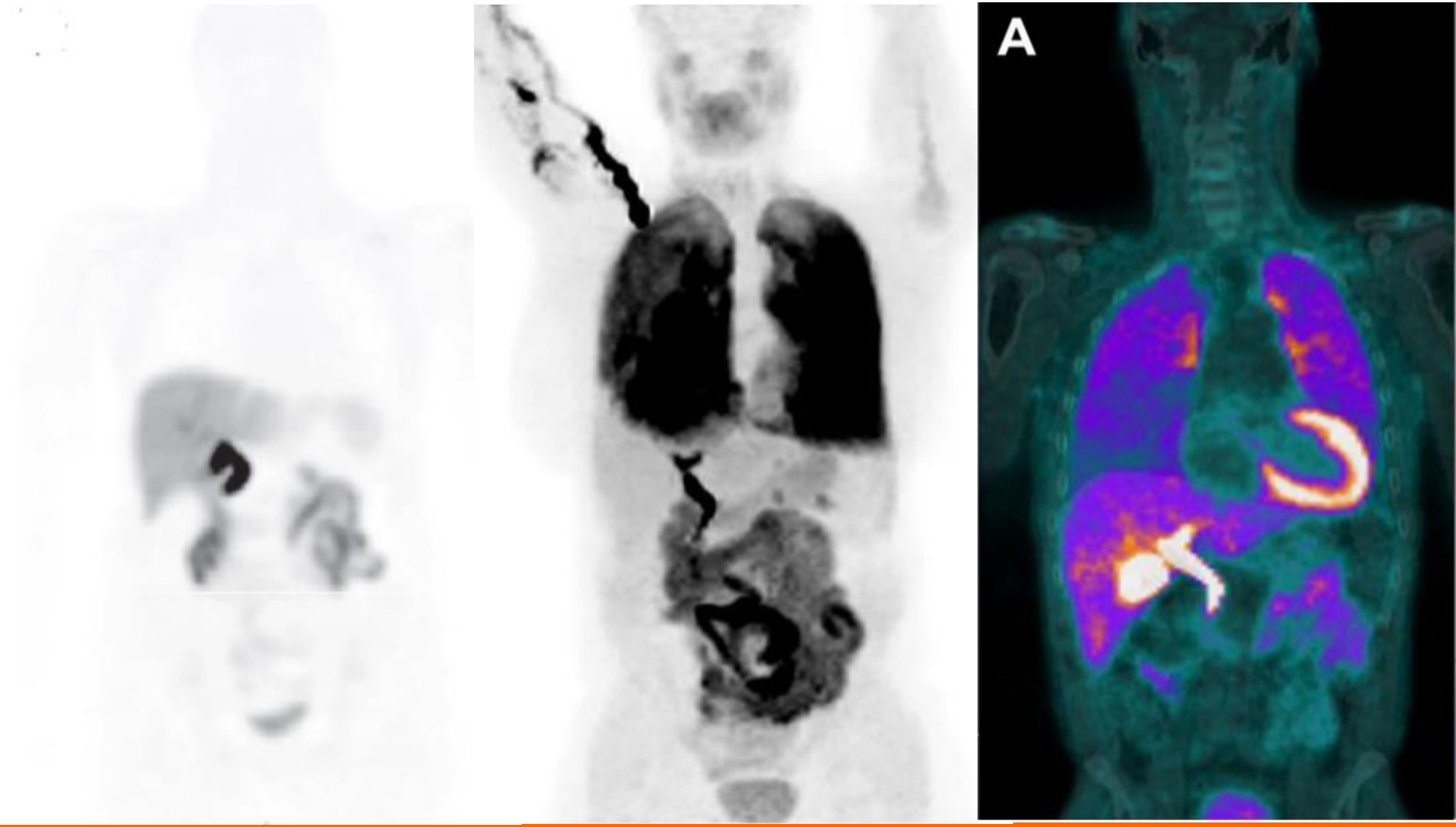
Björn Pilebro,[a] Sandra Arvidsson,[b] Per Lindqvist,[b] Torbjörn Sundström,[c] Per Westermark,[d] Gunnar Antoni,[e] Ole Suhr,[f] and Jens Sörensen[g]



Early Detection of Multiorgan Light Chain (AL) Amyloidosis by Whole Body ^{18}F-florbetapir PET/CT

Eric Christopher Ehman, Mohamed Samir El-Sady, Marie Foley Kijewski, Yiu Ming Khor, Sophia Jacob, Frederick L Ruberg, Vaishali Sanchorawala, Heather Landau, Andrew Jenho Yee, Giada Bianchi, Marcelo F. Di Carli, Rodney H Falk, Hyewon Hyun and Sharmila Dorbala

J Nucl Med.
Published online: April 6, 2019.



TEP 18F-Florbetapir

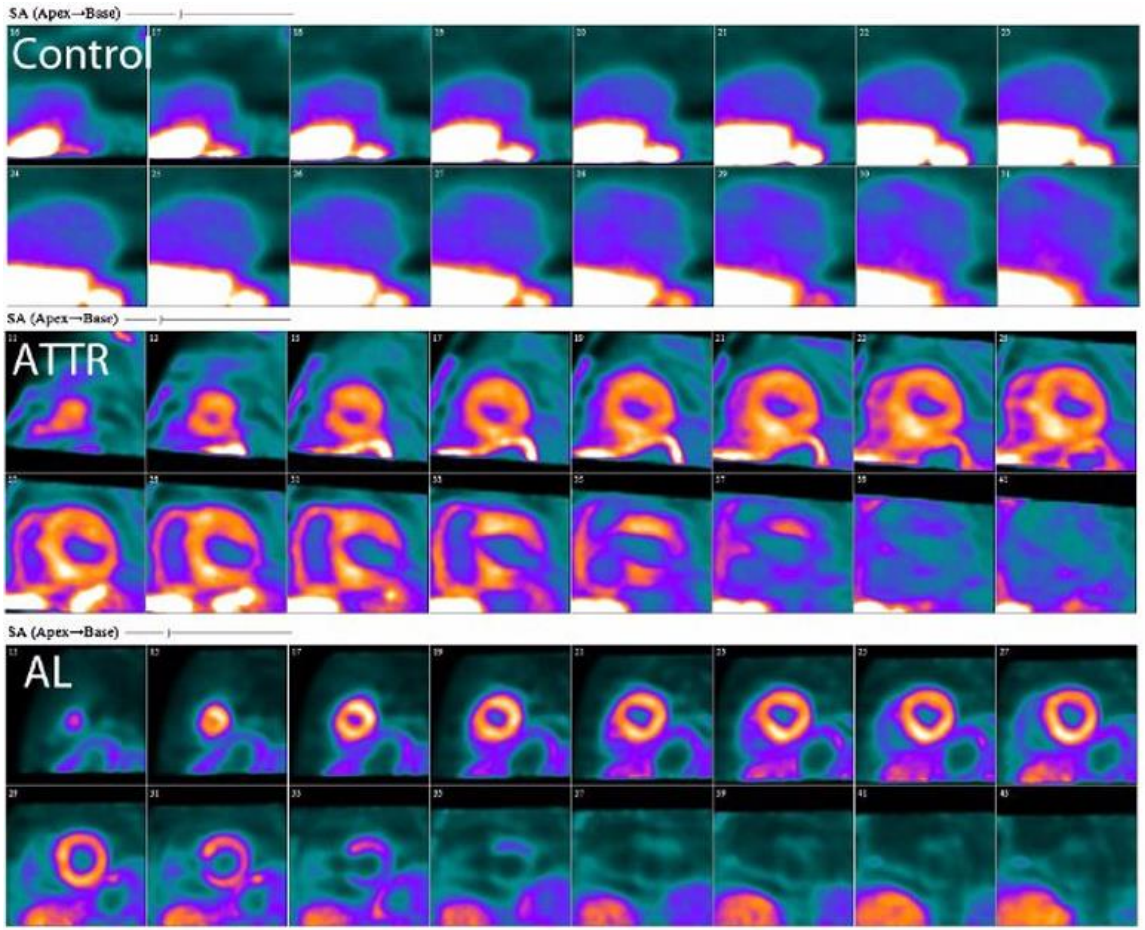


Fig. 1 LV myocardial ^{18}F-florbetapir uptake in representative control, TTR and AL amyloid subjects. The images show ^{18}F-florbetapir uptake in the heart in standard cardiac short axis projections from apex to base in two rows per subject. Note diffuse left > right ventricular uptake of the tracer in the ATTR subjects and in the AL subjects. The images in the control subject are scaled up to show the myocardial boundaries. *AL* light chain amyloidosis, *ATTR* transthyretin amyloidosis

Eur J Nucl Med Mol Imaging. 2014

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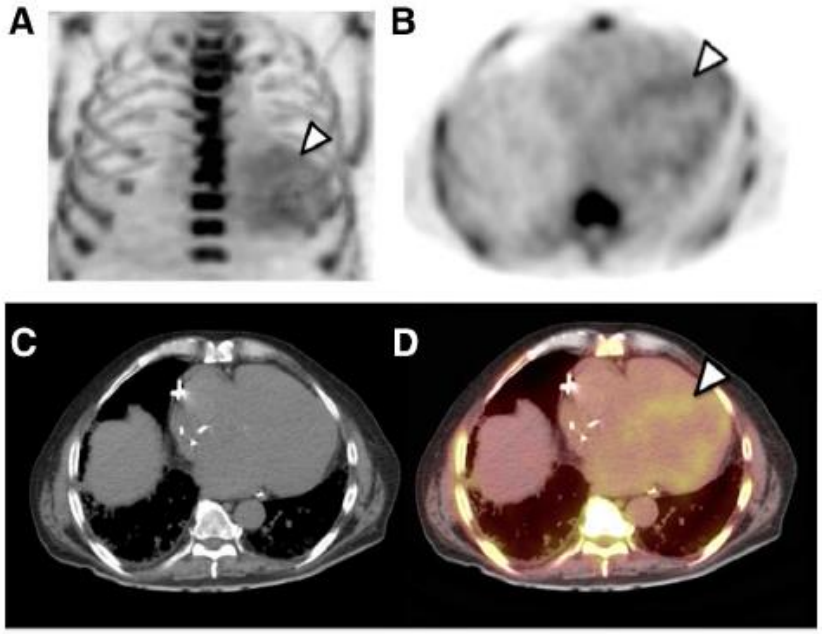
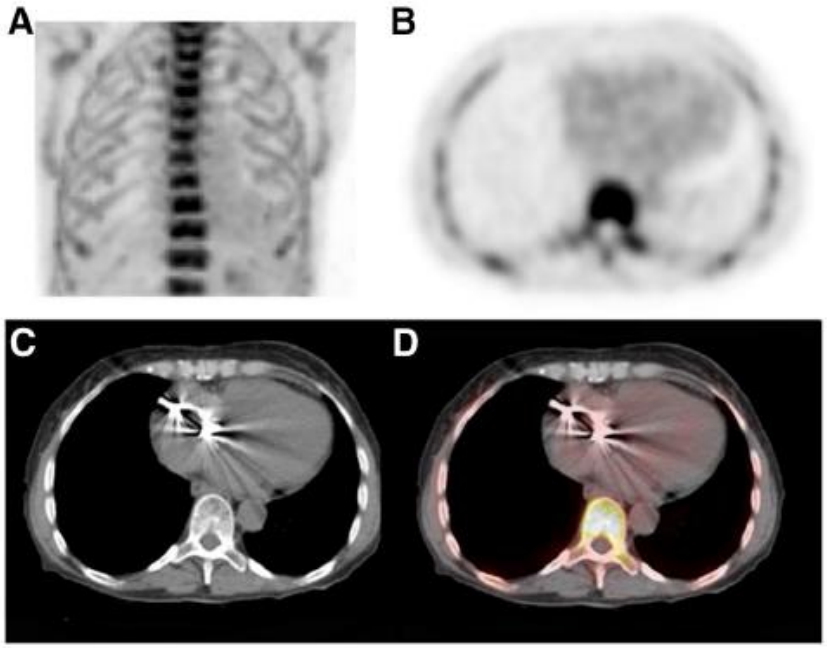
Dorbala S : Imaging cardiac amyloidosis: a pilot study using ^{18}F-florbetapir positron emission tomography

TEP 18F-FNa





Van Der Gucht et al
[18F]-NaF PET/CT in cardiac amyloidosis

Journal of Nuclear Cardiology®

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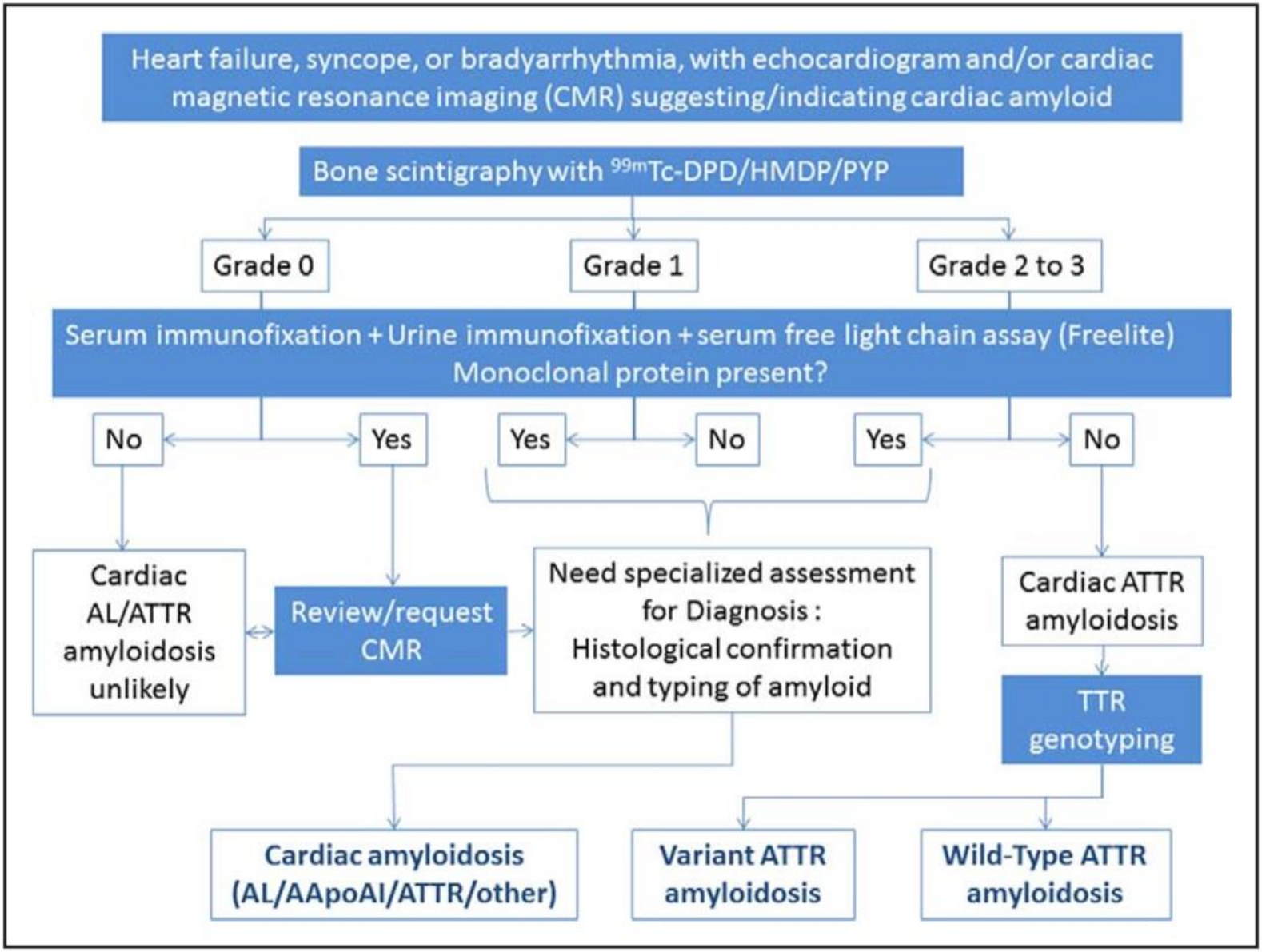
Take home message

- Scintigraphie osseuse aux biphosphonates
  - Identification spécifique des amyloses à TTR
  - Décision thérapeutique
- Homogénéisation progressive des pratiques
- Score de Perugini

Grade 0	no uptake and normal bone uptake
Grade 1	uptake less than rib uptake
Grade 2	uptake equal to rib uptake
Grade 3	uptake greater than rib uptake with mild/ absent rib uptake

- Score 1 : AL/ATTR -> biopsie
- ATTR : Myocarde+ Tissus mous + os –
- TTR : SO Se 99% et Sp 86%
- TTR : SO Score 2/3 et absence de pic monoclonal sérique ou urinaire
  - VPP 100%
  - Sp 98 %

Gillmore et al Circulation 2016



Merci de votre attention

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