



Multimodality amyloidosis imaging

EACVI Webinar

Tuesday 9 June 2020 from 18:00 to 19:00 CET

QUESTIONS AND ANSWERS

Answers written by Associate Professor Marianna Fontana

Questions	Answers
Do we need DPD if we have a CMR with high probability of Amyloidosis (very high T1)??	Native T1 needs to be interpreted in the clinical context. For ATTR see algorithm From Gillmore Circulation 2016. For AL characteristics imaging and peripheral biopsy should be considered.
Professor Fontana showed a case of a patient with amyloidosis and an aortic prosthesis, probably due do aortic stenosis. What can you tell about the association amyloidosis / aortic stenosis?	ATTR amyloidosis seems to be highly prevalent in AS. It is not clear, but seems plausible, that increased shear stress and shear forces facilitate TTR misfolding into amyloid and amyloid deposition.
Echo is first line technique when approaching patients with suspected cardiomyopathy. What do you think about echo findings and their reliability and accuracy in patients with late presentation/end stage cardiac amyloid?	Echo is very important to raise the suspicion, but CMR and DPD should be used to confirm diagnosis. Apical sparing on strain is the more specific sign on cardiac amyloidosis.
is any suggestions how to recognise amyloidosis in hypertrophy up to 1.3 cm?	DPD in TTR and CMR in AL are the most sensitive techniques.
Is it possible to have a negative MRI in a patient with histologically determined amyloidosis and why?	LGE is very sensitive, T1 mapping (with ECV) more sensitive. To my knowledge is not possible to have cardiac amyloidosis with a normal LGE AND normal ECV.

which parameter (in CMR or nuclear) renders the best prognostic to describe evolution of patients (survival time of example)	By CMR ECV. DPD is not very good to startify prognosis.
in case of End Stage Renal Disease still not in renal replacement therapy, would you still perform LGE ??? or would you go first for nuclear test?	You can go for nuclear if you suspect TTR, if you suspect AL you should assess risk/benefit ratio and consider CMR with contrast (risk of NSF is very low with some agents - as dotarem).
are there any differences (known or expected) between AL and ATTR?	Many differences are present between AL and ATTR on Imaging. TTR shows typically more increase in mass, more systolic and diastolic dysfunction, higher ECV, lower T1, lower T2, less pericardial and pleural effusion.
are there any fluid dynamic differences (known or expected) between AL and ATTR?	Please see above.
MRI negative with positive cardiac biopsy (post mortem)? possible? why?	See answer number 6.
What about LAB diagnostic?	This is a very wide topic. Probably a review would be more appropriate.
Chemotherapy - mutually in both type?	Only in AL.
Is it possible to have a negative MRI in a patient with histologically determined amyloidosis and why? - very important!!!!	See answer number 6.
Can cardiac MRI differentiate ATTR and AL cardiac amyloidosis? - crucial!	No, it should not be used to differentiate, only to make a diagnosis of cardiac amyloidosis.
if you don't see any LGE in CMRI, detect normal T1 and normal ECV - can you exclude amyloidosis?	Yes
how do we differentiate between amyloidosis and Fabry by echo	Very difficult, you need a CMR with native T1 mapping.
What is the role of BNP to monitor heart failure treatment in amyloid?	It is very established in AL (more than 30% reduction means a cardiac response during chemotherapy). Not established in ATTR.
how reproducible is visual analysis of scintigraphy in amyloidosis (Perumini grade 0-3)?	Very reproducible.
How I important is the apical sparring in amyloidosis?	Very important, it is the most specific sign on echo - but not present when coexistent AS or other conditions that significantly increase afterload
Have you ever seen isolated atrial amyloidosis showing tracer uptake in nuclear imaging?	No
What is the best way to image cardiac AA amyloidosis? And is it important to do so?	Cardiac AA amyloid is not a clinically significant form of cardiac amyloidosis in the vast majority of

	cases. Patients with AA amyloid ca have amyloid deposits, but not significant cardiac amyloidosis.
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