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STEP BY STEP MAGNETIC RESONANCE IMAGING IN A CASE OF CARDIAC AMYLOIDOSIS – IT IS NOT ALL ABOUT AMYLOID DEPOSITION!

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It is often said that cardiac magnetic resonance (CMR) has the diagnostic power of a noninvasive myocardial biopsy. We illustrate this concept by stepwise analysis of a CMR exam in 65-year-old patient referred to our department for tissue characterization of a left ventricular (LV) hypertrophy of unknown etiology. The subsequent complete laboratory work-up has diagnosed cardiac amyloidosis (CA) of AL type (light chain CA). The reader will become acquainted with the common sequences of the clinical CMR protocol in CA.

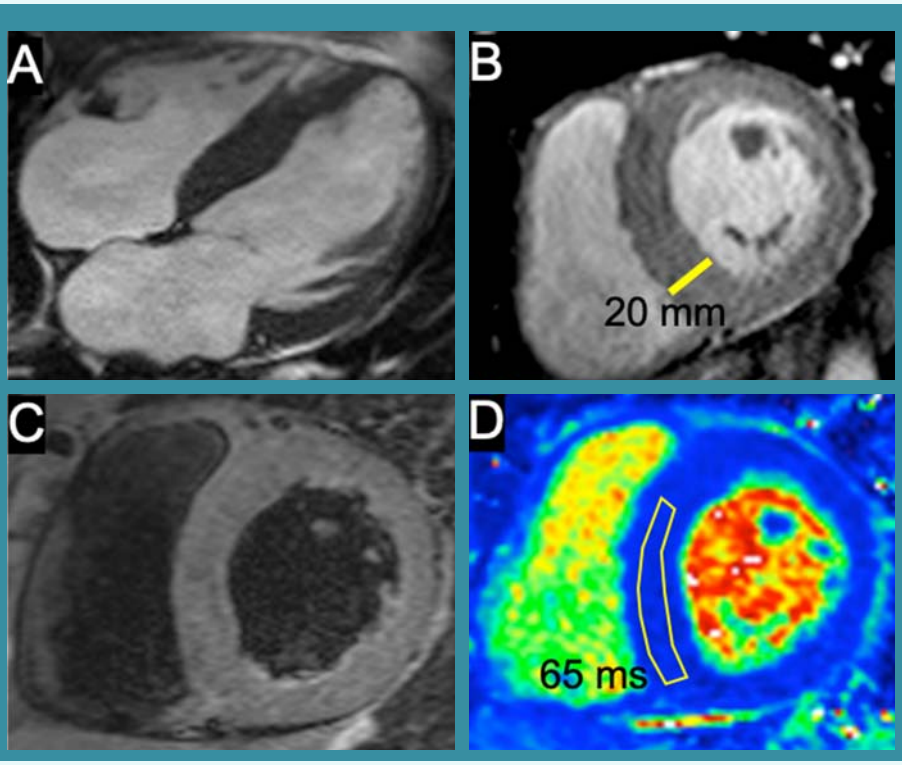
The conventional T2-weighted imaging is a *qualitative* method which can detect only focal oedema as an area of hyperintensity (oedema) surrounded by lower intensity (normal) myocardium. In image **C.** homogenous intensity of the myocardium is seen, excluding focal oedema, however, diffuse myocardial oedema (such is the case of CA) cannot be excluded. T2 mapping **(D)** is a *quantitative* technique allowing the measurement of T2 value in each voxel. A T2 value higher than 50 ms is diagnostic for myocardial oedema (at 1.5 Tesla). In our case, a T2 value of 65 ms was highly suggestive of *myocardial oedema* accompanying the amyloid deposition(1).

Short axis quantitative T1 maps before **(E)** and after **(F)** Gadolinium-based contrast agent injection. Amyloidosis is characterized by the highest native T1 times among all heart conditions, with values >1164 ms being highly suggestive of CA [(2)]. Introducing native and post-contrast T1 values in a formula, the extracellular volume fraction (ECV) can be calculated. In CA the interstitium is expanded due to amyloid deposition and thus ECV can accurately and non-invasively estimate the myocardial amyloid burden. In our patient the calculated ECV was 76% (normal <30%) [(3)].

Two chambers views acquired *early* **(G)** and *10 minutes after* **(H)** Gadolinium injection. The *early* Gadolinium enhancement (EGE) images are acquired in the first minute after contrast injection, allowing for detection of poorly perfused tissues such as areas of microvascular obstruction (MVO). In our case, MVO was identified as a dark subendocardial rim (yellow arrows) surrounded by normally enhanced myocardium.

On the other hand, *late* Gadolinium enhancement (LGE) images are usually pathognomonic for CA: the myocardium appears bright with a dark blood pool appearance (Gadolinium is an extracellular agent which concentrates in tissues with enlarged extracellular spaces such as scars or amyloid infiltration). In the case of our patient, it can be seen that Gadolinium is retained in areas with the highest amyloid burden, mainly the basal LV segments with relative apical sparing (**H.**, yellow arrows).

By gathering information from all the sequences of the CMR protocol we were able to demonstrate that CA is not characterized solely by amyloid deposition in the interstitium. On the contrary, several other



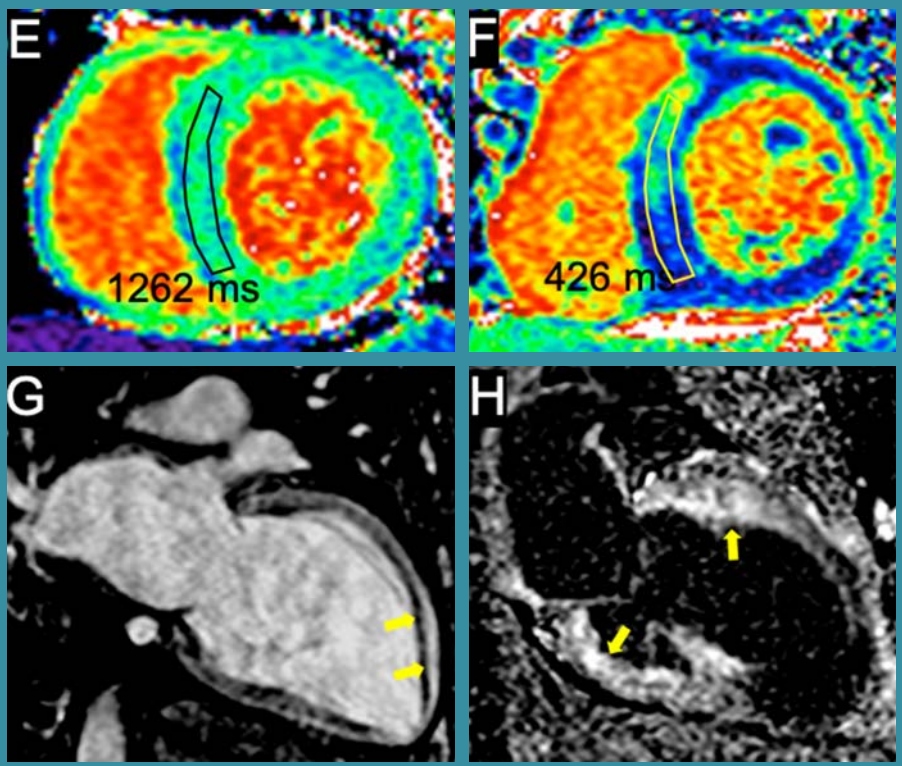


Figure 1. *Non-contrast cine images (balanced steady-state free precession) in 4 chambers* ***(A)*** *and short axis* ***(B)*** *views respectively allow for morphological and functional assessment. The left ventricle (LV) was dilated and the interventricular septum was severely and asymmetrically hypertrophied. The LV ejection fraction was 35%. The right ventricular (RV) free wall was not thickened, however RV was severely dysfunctional, with an ejection fraction of 25%. The atria were only mildly dilated.*
Imaging of myocardial oedema in short axis view using the conventional T2-weighted sequences ***(C)*** *and the more recently developed T2 mapping technique* ***(D).***

pathophysiological processes occur in AL CA, such as: myocardial oedema, myocardiocyte necrosis and microvascular obstruction. These phenomena express the central role of direct light chain toxicity on myocardiocytes resulting in a toxic myocarditis. In AL CA the direct myocardial toxicity of the light chains plays a more important role in the pathophysiology of the disease than strictly interstitial deposition of amyloid.

CMR is the only non-invasive imaging modality able to simultaneously quantify the amyloid burden as well as to identify myocardial oedema, necrosis and microvascular obstruction, providing important prognostic information. This is why CMR undoubtedly deserves the designation of noninvasive myocardial biopsy and should be offered to all CA patients.

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