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Response to 'Is the left atrium the key in the amyloid imaging lock?'

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We thank Dr Bart for her commentary on our paper.[1] The echocardiogram is the first-line imaging technique in patients with suspected cardiac amyloidosis (CA).[2] Speckle-tracking analysis of the left ventricle should be systematically performed, as the apical sparing pattern is strongly suggestive of CA, particularly in individuals with unexplained hypertrophy.[2] In patients in sinus rhythm, the left atrial (LA) function may be studies by calculating the peak LA longitudinal strain (PALS), which reflects the reservoir function, and peak LA contraction strain (PACS), which explores the contractile function of the LA. In our study, we reported that both PALS and PACS are severely depressed in patients with amyloid transthyretin (ATTR) CA.[3] The mechanisms are (i) a progressive dilation and remodelling of the LA, due to diastolic dysfunction and leading to a depressed LA contractility, and (ii) amyloid deposition in the atrial walls, with an expansion of extracellular spaces and cardiomyocyte dysfunction. Structural and functional remodelling of the LA is more prominent in ATTR-CA, which is a slowly progressive disease, than in amyloid light-chain (AL)-CA, which has a more rapid progression.[3] We agree with Dr Bart that this difference between ATTR- and AL-CA may be useful for the purposes of differential diagnosis. The exact role of cardiovascular magnetic resonance (CMR) in the diagnostic workup of CA has not been standardized yet, but CMR may implement baseline examination in patients with suspected CA. Several techniques (feature tracking, tagging, and tissue-phase contrast) allow to track the motion of LA throughout the cardiac cycle and represent the CMR equivalents of speckle-tracking imaging. The study cited by Dr Bart investigated the value of CMR-based LA strain in 54 patients with CA (30 AL and 24 ATTR), reporting a lower LA function in patients with ATTR vs. AL despite no significant differences in LA geometry.[4] These findings are in agreement with our study.[1] Echo- or CMR-based LA strain might then help differentiate AL- from ATTR-CA among patients starting the diagnostic workup for suspected CA. On the other hand, further investigations (cardiac scintigraphy with bone tracers, search for a monoclonal protein, and often histological analyses) are still needed to confirm the diagnosis of CA and its subtype, in agreement with the current algorithm.[1] Beyond its possible diagnostic value, we think that LA strain might prove helpful for risk stratification and to track disease evolution. A more depressed LA strain is reasonably associated with more blood stasis and a greater risk of atrial thrombosis, and could then suggest the potential need for an anticoagulant therapy even when there is no history of atrial fibrillation.[1,2] As for the second point, LA strain has a close association with disease severity and is highly reproducible, meaning that a deterioration of LA strain over time reflects disease progression, while unchanged or improved values over times may mirror the stabilizing or positive effects of disease-modifying therapies.[5] In summary, LA strain could really become a 'key in the amyloid imaging lock'.

Conflict of interest: None declared.

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