Imaging: Myocardial Disease

Natural history and disease progression of early cardiac amyloidosis evaluated by echocardiography

Itzhaki Ben Zadok O.; Eisen A.; Shapira Y.; Monakier D.; Iakobishvili Z.; Schwarzenberg S.; Abelow A.; Ofek H.; Kazum S.; Hamdan A.; Bental T.; Sagie A.; Kornowski R.; Vaturi M.

Rabin Medical Center, Department of Cardiology, Petah Tikva, Israel

Funding Acknowledgements: Type of funding sources: None.

Background: Since the diagnosis of cardiac amyloidosis (CA) is often delayed, echocardiographic findings are frequently indicative of already advanced cardiomyopathy.

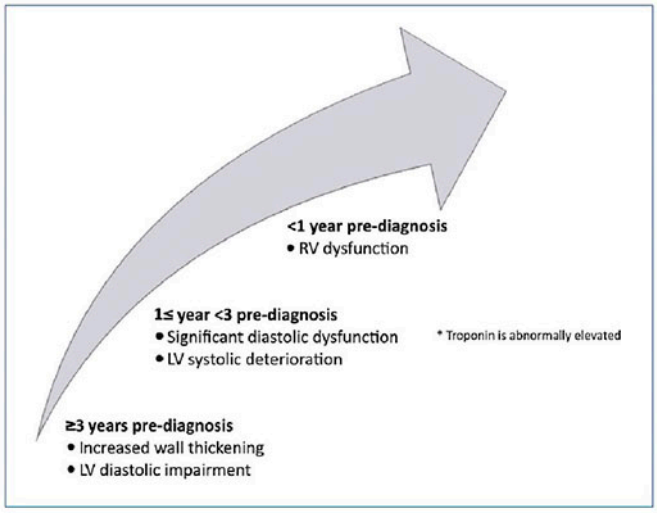
Aims: to describe early echocardiographic features in patients subsequently diagnosed with CA and to delineate disease progression.

Methods: Pre-amyloid diagnosis echocardiographic studies were screened for structural and functional parameters and stratified according to the pathogenetic amyloid subtype (immunoglobulin light-chain (AL) or amyloid transthyretin (ATTR)). Abnormalities were defined based on published guidelines.

Results: Our cohort included 75 CA patients of whom 42 (56%) were diagnosed with AL and 33 (44%) with ATTR. Forty-two patients had an earlier echocardiography exam available for review. Patients presented with increased wall thickness (1.3 (IQR 1.0, 1.5)cm) ≥3 years before the diagnosis of CA and relative wall thickness (RWT) was increased (0.47 (IQR 0.41, 0.50)) ≥7 years pre-diagnosis. Between 1 to 3 years before CA diagnosis restrictive left ventricular (LV) filling pattern was present in 19% of patients and LV ejection fraction (LVEF)≤50% was present in 21% of patients. Right ventricular dysfunction was detected concomitantly with disease diagnosis. The echocardiographic phenotype of ATTR versus AL-CA showed increased RWT (0.74 (IQR 0.62, 0.92) vs. 0.62 (IQR 0.54, 0.76), p = 0.004) and LV mass index (144 (IQR 129, 191) vs. 115 (IQR 105, 146)g/m2,p = 0.020) and reduced LVEF (50 (IQR 44, 58) vs. (60 (IQR 53, 60)%, p = 0.009) throughout the time course of CA progression, albeit survival time was similar.

Conclusions: Increased wall thickness and diastolic dysfunction in CA develop over a time course of several years and can be diagnosed in their earlier stages by standard echocardiography

Abstract Figure. Schematic proposed timeline of CA



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