Valve disease in cardiac amyloidosis: an echocardiographic score

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Funding Acknowledgement: Type of funding sources: None.

Background: Cardiac amyloidosis (CA) may affect all cardiac structures, including the valves.

Methods: From 423 patients undergoing a diagnostic workup for CA we selected 2 samples of 20 patients with amyloid transthyretin (ATTR-) or light-chain (AL-) CA, and age- and sex-matched controls. We chose 31 echocardiographic items related to the mitral, aortic and tricuspid valves, giving a value of 1 to each abnormal item.

Results: Patients with ATTR-CA displayed more often a shortened/hidden and restricted posterior mitral valve leaflet (PMVL), thickened mitral chordae tendineae and aortic stenosis than those with AL-CA, and less frequent PMVL calcification than matched controls. Score values were 15.8 (13.6–17.4) in ATTR-CA, 11.0 (9.3–14.9) in AL-CA, 12.8 (11.1–14.4) in ATTR-CA controls, and 11.0 (9.1–13.0) in AL-CA controls (p=0.004 for ATTR- vs. AL-CA, 0.009 for ATTR-CA vs. their controls, and 0.461 for AL-CA vs. controls). Area under the curve values to diagnose ATTR-CA were 0.782 in patients with ATTR-CA or matched controls, and 0.773 in patients with LV hypertrophy.

Conclusions: Patients with ATTR-CA have a prominent impairment of mitral valve structure and function, and higher score values. The valve score is quite effective in identifying patients with ATTR-CA among patients with CA or unexplained hypertrophy.