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SENILE SYSTEMIC AMYLOIDOSIS IS A MARKEDLY UNDERDIAGNOSED CARDIOMYOPATHY: EXPERIENCE OF A CARDIAC AMYLOIDOSIS PROGRAM

Poster Contributions
Poster Sessions, Expo North
Saturday, March 09, 2013, 3:45 p.m.-4:30 p.m.

Session Title: What Is Unfolding in Cardiac Amyloidosis Research?
Abstract Category: 23. Pericardial/Myocardial Disease
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Authors: *Rodney H. Falk, Jenna Kruger, Candida Cristina Quarta, Brigham and Women's Hospital, Boston, MA, USA, Harvard Vanguard Medical Associates, Boston, MA, USA*

Background: Senile systemic amyloidosis (SSA) is an infiltrative cardiomyopathy of older men due to amyloid deposits derived from wild-type transthyretin (TTR). Although TTR amyloid in autopsied hearts >70 yr old, is common, infiltration severe enough to cause thick LV and CHF is unusual. Nevertheless, it might be expected to be seen in an older CHF population. In published series of amyloidosis, the proportion of patients with SSA is <2.5%, whereas light-chain (AL) amyloidosis is about 75%. If this is the true prevalence of SSA, it would be very rare, which is discrepant with autopsy data. We therefore sought to determine the prevalence of SSA as a cause of CA in consecutive patients at our institution, to investigate whether it was commoner than previously reported.

Methods: All patients with a diagnosis of CA were evaluated for precise etiology. SSA was defined as TTR amyloid without mutation, familial amyloid as CA with mutant TTR and AL amyloidosis as CA with plasma cell dyscrasia.To eliminate referral bias we also analyzed patients in whom the amyloid type was unknown at the time of referral, but later determined.

Results: See table.

Conclusion: SSA was10 times as common in our referral population than previously reported, accounting for 29-33% of all cases of CA seen. With an aging population and better diagnostic techniques SSA is likely to be recognized as a much commoner cause of cardiomyopathy in elderly men than previously realized.

Breakdown of CA Etiology

	AL	SSA	Familial
Total (%)	125 (53.4%)	77 (32.9%)	32 (13.7%)
Mean age yr.	62.6	75.2	70
M/F	88/37	73/4	21/11
Number previously untyped ((%)	68 (58.6%)	34 (29.8%)	14(12.1%)