

Cardiac sympathetic denervation in wild-type transthyretin amyloidosis

A. Aimo¹, A. Gimelli², G. Vergaro², D. Genovesi², A. Kusch², M. Emdin², P. Marzullo²

¹Scuola Superiore Sant'Anna, Pisa, Italy; ²Fondazione Toscana Gabriele Monasterio, Pisa, Italy

Funding Acknowledgement: Type of funding source: None

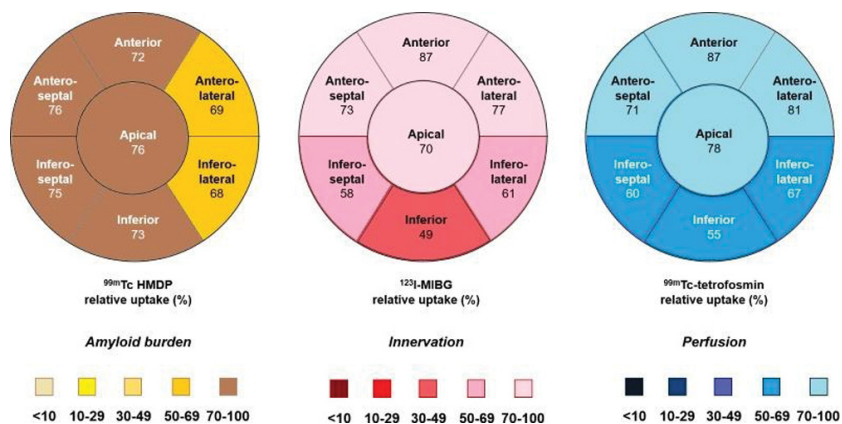
Background: Tissue accumulation of misfolded transthyretin (TTR) may occur because of TTR gene mutations (variant amyloid TTR amyloidosis, ATTRv), or as an age-related phenomenon (wild-type ATTR, ATTRwt). Cardiac sympathetic denervation has been reported in ATTRv, but has never been investigated in ATTRwt.

Methods: Fifteen consecutive patients with ATTRwt cardiomyopathy (81% men, median age 82 years, no one with prior myocardial infarction) underwent Cadmium Zinc Telluride tomographic imaging for amyloid burden (99mTc-hydroxymethylene diphosphonate - 99mTc-HMDP), innervation (123I-metaiodobenzylguanidine - 123I-MIBG), and perfusion (99mTc-tetrofosmin).

Results: Median summed 99mTc-HMDP score was 60 (58–62), denoting a severe and diffuse amyloid burden. Planar 123I-MIBG examination showed decreased early and late H/M ratios (late H/M ratio: 1.5 [1.3–1.6], range 1.2–1.9, reference value ≥ 2.0). Summed 123I-MIBG score was 12

(6–22), with the most prominent denervation in the infero-septal, inferior, and infero-lateral regions; summed rest score was 7 (5–11), with lowest degrees of myocardial perfusion in the inferior and infero-septal regions. The correlation between amyloid burden (as relative 99mTc-HMDP uptake) and innervation (as relative 123I-MIBG uptake) did not achieve statistical significance at both segmental ($p=0.252$) and regional level ($p=0.251$). Nevertheless, denervation tended to worsen in parallel with the amyloid burden, and 123I-MIBG scores increased with 99mTc-HMDP scores. Segments and regions with more prominent hypoperfusion were those showing the more intense denervation ($r=0.500$ and 0.591 , respectively; both $p<0.001$).

Conclusions: Patients with ATTRwt cardiomyopathy display cardiac sympathetic denervation, particularly in the inferior and septal myocardial wall. Myocardial hypoperfusion has a similar regional pattern, while the amyloid burden is more extensive.



Downloaded from https://academic.oup.com/eurheartj/article/41/Supplement_2/ehaa946.2043/6003511 by guest on 25 April 2024