Renal amyloidosis: validation of a proposed histological scoring system in an independent cohort

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Background

In systemic amyloidosis, the kidney is frequently affected and renal involvement has major impact on survival. Renal involvement is clinically characterized by decreased estimated Glomerular Filtration Rate (eGFR) and proteinuria. The two most common renal amyloidosis types are light chain related amyloidosis (AL) and serum amyloid A (AA) amyloidosis. Standardized histopathological scoring of amyloid deposits is crucial to assess disease progression. Therefore, we aimed to validate the proposed scoring system from Rubinstein et.al. (1) in an independent patient cohort.

Methods

We attempt to reproduce the scoring system, consisting of an Amyloid Score (AS) and a Composite Scarring Injury Score (CSIS), in a multicenter AL and AA case series. Additionally, we analyzed all renal amyloidosis kidney biopsies performed in the Netherlands between 1993 and 2012.
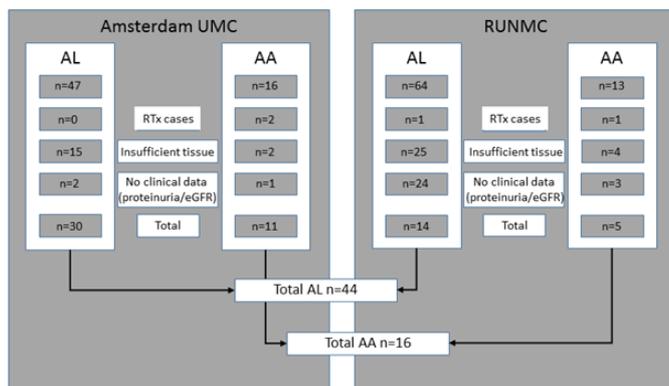
Figure 1 Flow chart showing the number of included and excluded renal amyloidosis cases in our patient cohort.

Results

Similar to the original study, AS and CSIS correlated to eGFR (r:-0.45, p=0.0061 and r:-0.60, p<0.0001, respectively), but not to proteinuria at diagnosis. Furthermore, AS, but not CSIS, was associated with renal outcome. The scoring system was not reproducible in AA patients. The median incidence rate (IR) for renal amyloidosis in the Netherlands was 2.3 per million population per year (pmp/y) and increased during the study period.

Reference
1. Rubinstein S, Cornell RF, Du L, et al.; Novel pathologic scoring tools predict end-stage kidney disease in light chain (AL) amyloidosis. Amyloid 2017; 24(3):205-211.

Figure 1
Incidencerate of renal amyloidosis in the Netherlands between 1993 and 2012, values are presented as per million population (pmp).
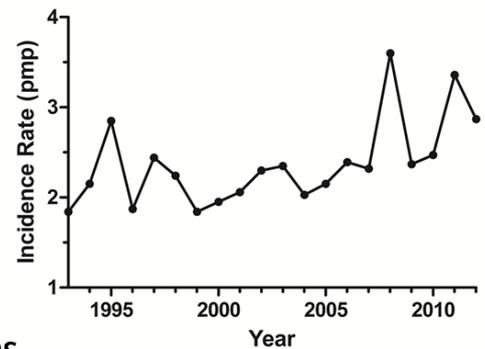
Conclusions

In our AL case series and the original study, AS and CSIS were correlated to eGFR, but not to proteinuria and AS correlated with renal outcome. Overall, we regard this scoring system competent for standardized histopathological assessment of amyloid deposits burden and thereby disease advancement in renal biopsies.
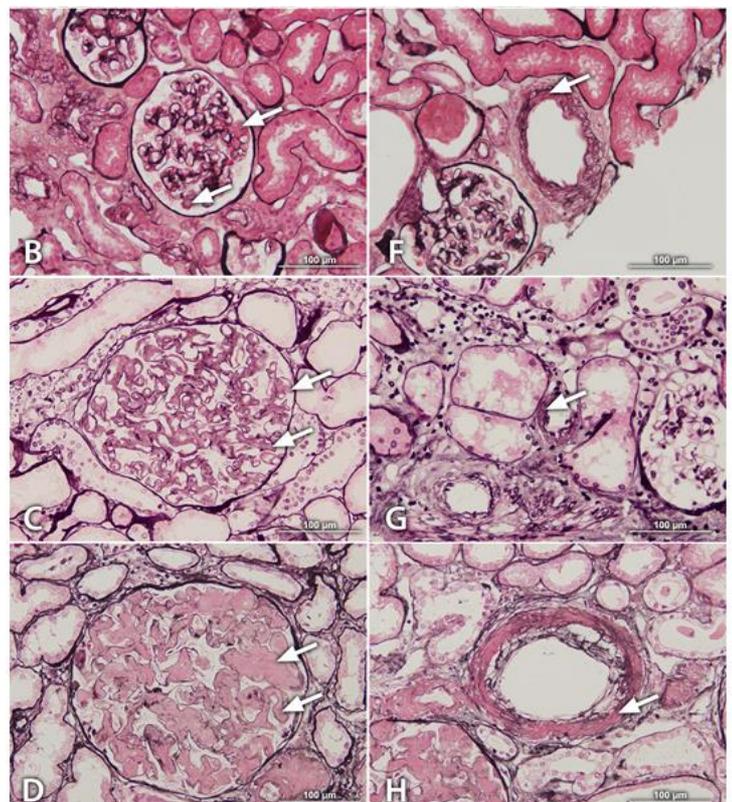
Figure 3 Examples of amyloid deposits score. B-D: Representative slides showing the mesangial and capillary wall amyloid scores. B=1;1, C=2;2, and D=3;3, respectively (arrows showing deposits, silver staining, x20). F-H: Representative slides showing the vessel amyloid score. F=1, G=2, H=3 (arrows showing deposits, silver staining, x20).