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Introduction

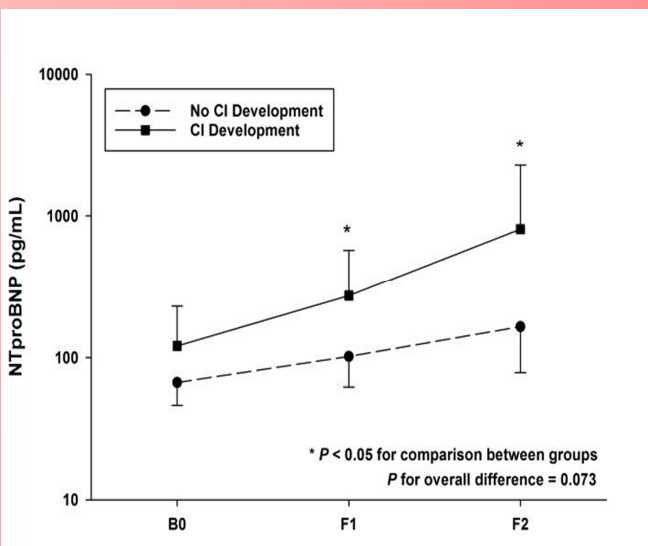
In hereditary transthyretin-derived amyloidosis (ATTRm) multiple organ systems are affected, including the heart. Cardiac involvement is a significant determinant of mortality in ATTRm amyloidosis.

The aim of this study was to describe cardiac disease progression in ATTRm patients from the Groningen Amyloid Cohort (GAC).

Methods

All patients with histologically and genetically confirmed hereditary ATTR were followed prospectively as part of the GAC. Baseline (B0) was defined as the date of the first positive biopsy, first follow-up (F1) period was after 2 years, the second follow-up (F2) was after 4 years. Cardiac involvement as assessed by echocardiography was defined as structural or functional alterations of the myocardial wall (LV >11 mm, RV >6 mm) and/or conduction disturbances. Statistics were performed using the chi-square test, the Kruskal-Wallis equality-of-populations rank test, or the One-way ANOVA where appropriate. To analyse the increase of NT-proBNP a repeated measures mixed model analysis was performed.

Results



In total 77 patients were analysed, mean age was 61 years and 43 (56%) was male. In 39 patients (51%) cardiac involvement was present at baseline. In the remaining 38 patients (49%) who did not have cardiac involvement at baseline, 9 (24%) patients developed cardiac involvement after a median follow up of 2.5 year. In the figure the increase of NT-proBNP (ng/L) depicted by the mean values for the 9 patients who developed cardiac involvement and also for the 29 patients in whom cardiac involvement did not occur during follow-up. In both follow-up time point the NT-proBNP levels differ between the two sub-groups, in both time periods $p < 0.05$, indicated with an *. Overall, no difference between both trends was present, $p = 0.073$.

Conclusion

A substantial part of patients with ATTRm amyloidosis without initial cardiac involvement developed cardiac involvement during follow-up in the first four years after diagnosis.

The biomarker NT-proBNP seems to be an appropriate parameter to monitor the development of cardiac involvement in these patients.