Relapse-free survival was significantly longer for patients who were completely discordant for about 25% of them.

Our results can lead to test optimisation and recommendations for testing in other immune diseases.

Conclusion.— ANCA alone cannot be considered a reliable marker of GPA remission.

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Are anti-proteinase-3 ANCA a useful marker of granulomatosis with polyangiitis relapses? Results of a longitudinal study on 126 patients

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Introduction.— Predicting granulomatosis with polyangiitis (GPA) relapses by measuring of ANCA titers remains a source of debate. Our objective was to evaluate the relevance of monitoring PR3-ANCA titers for GPA management.

Methods.— This retrospective study included 126 patients recruited from Cochin Hospital, all fulfilling the 1990 ACR criteria for GPA and PR3-ANCA-positive at the time of diagnosis. GPA activity was assessed with the Birmingham Vasculitis Activity Score for Wegener granulomatosis (BWAS/WG). ANCA were detected in an immuno-fluorescence assay and their PR3-ANCA specificity was determined in an ELISA. Risk factors of relapse were assessed using a conditional Andersen–Gill model. Hazard ratios (HR) [95% confidence interval (CI)] are given.

Results.— For the 126 patients (51.6% male, mean age 49 yr), the vasculitis organ-involvement distribution at inclusion was ENT (82.5%), lung (69%), kidney (45.2%), with median BWAS/WG of 7 at diagnosis and 3 at relapse. Remission was obtained in 112 (88%) patients. ANCA became undetectable by IF for 70/115 (60.9%) patients and ELISA for 90/115 (78.3%). After a median follow-up of 70 months, 85/126 (67.5%) patients suffered 154 clinical relapses associated with cANCA-positivity for 122 (79.2%) and PR3-ANCA for 102 (66.2%) of them. Relapse-free survival was significantly longer for patients who remained cANCA-negative (HR0.67 [95% CI 0.47–0.98], p = 0.037) and PR3-ANCA-negative (HR 0.60 [95% CI 0.39–0.92], p = 0.02). When we studied evolution in ANCA titers course and clinical outcome for each patient, a tight parallelism was observed for 60% of them, ie each relapse was associated with ANCA-positivity and relapse-free survival with persistent ANCA-negativity.

Discussion.— We have evidenced that most relapses were associated with IF-positive ANCA and for 60% of the GPA patients, clinical outcome and ANCA-titer changes were tightly correlated. However, they were completely discordant for about 25% of them.

Conclusion.— ANCA alone cannot be considered a reliable marker of GPA remission.