Conclusion: Our results suggest that the coexistence of MEFV exon 2 or exon 3 variants and a MEFV exon 10 mutation has combined effects on inflammasome activation in the Japanese population.

References:

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SAT0528

CLINICAL PHENOTYPES OF IGG4-RELATED DISEASE REFLECT DIFFERENCES IN EPIDEMIOLOGICAL FEATURES, SEROLOGICAL FINDINGS, AND PROGNOSTIC OUTCOMES

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Background: Four clinical phenotypes of IgG4-Related Disease (IgG4-RD) have been recently identified by Latent Class Analysis (LCA) - Pancreato/biliary (Group 1), Retropertitoneum/Aortitis (Group 2); Head-and-neck limited (Group 3); Miekulicz/Systemic (Group 4) - but the relevance of this classification for patient management remains unknown (1,2).

Objectives: We aimed to assess whether clinical judgment can replicate LCA classification and to evaluate potential differences in epidemiological features, serological findings, and disease outcomes between disease phenotypes.

Methods: The study included 179 patients. Four IgG4-RD experts were asked to classify a validation cohort of 40 patients according to published LCA derived phenotypes. The following variables were recorded on additional 139 patients: serum IgG4 and IgE; inflammatory markers; esoinphils; plasmablasts; IgG4-RD Responder Index (RI); history of atopy, diabetes, osteoporosis, relapses, and tumors; cumulative dose of glucocorticoids and use of rituximab.

Results: Clinical judgment recapitulated LCA classification with strong agreement between IgG4-RD experts (κ = 0.841, p < 0.0005). Group 1 showed the highest levels of serum IgG4 and IgE; inflammatory markers; esoinphils; plasmablasts; IgG4-RD Responder Index (RI); history of atopy, diabetes, osteoporosis, relapses, and tumors; cumulative dose of glucocorticoids and use of rituximab.

Conclusion: SAT0528