

## IGG4-RELATED DISEASE: RESULTS FROM A MULTICENTER PANLAR (PANAMERICAN LEAGUE OF ASSOCIATIONS FOR RHEUMATOLOGY) STUDY GROUP REGISTRY

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**Objective:** Immunoglobulin G4-related disease (IgG4-RD) is a systemic emerging disease. The aim of this study was to investigate, the frequency of different phenotypes, and to describe demographic, analytical, pathological and therapeutic features of this condition.

**Methods:** A multicenter study was performed utilizing the database of PANLAR-IgG4RD study group.

We included patients meeting the 2012 consensus criteria on IgG4-RD. The phenotypes were defined according to Wallace et al 2019 classification. Statistical analyses were performed by SPSS.

**Results:** 174 patients were included in the study, 94 of whom (54%) were male. Median age at diagnosis was 50 ± 14 years, but the average time of onset of

symptoms was  $48 \pm 14$ . The average diagnostic delay was 29 months (median 12). The average follow-up was 30 months.

In total, 211 manifestations were identified in 174 patients at the time of diagnosis. The most common manifestation was pancreatitis, diagnosed in 56 patients (32%), followed by dacryoadenitis in 50 (29%), sialadenitis in 33 (19%). 135 patients (77.6%) had multiple organ involvement. In contrast, 39 patients (22.4%) presented with an isolated lesion.

The most frequent phenotype was Group 1 "Pancreato-Hepatobiliary" in 53 patients (30.5%), followed by Group 3 "Head and neck limited" in 50 patients (28.7%), Group 4 "Mikulicz and systemic" in 29 (16.7%) patients and Group 2 "retroperitoneum and aorta" in 20 patients (11.5%). The remaining 12.6% could not be classified in these phenotypes. Phenotype 1 was significantly more common in males than in females (37.2% vs. 22.5%,  $p = 0.03$ ) while the opposite occurred in relation to phenotype 3 which was significantly more frequent in women (45% vs. 14.9%,  $p < 0.001$ ).

Serum IgG4 was elevated in 94/147 patients (64%). The median concentration was 740 mg / dL (range 4–7020). There were no statistically significant differences between serum IgG4 values among patients with isolated versus multiorgan involvement. Serum IgG levels were elevated in 66/123 patients (53.7%) and serum IgE levels in 38/59 patients (64.5%). 156/174 biopsy samples (89.7%) were obtained. The most frequently biopsied sites were: pancreas in 18%, submaxillary gland in 11.5%, lymph node in 9.6% and tear gland in 9%.

156 patients received medical treatment, 100% with corticosteroids, 37.8% with azathioprine, 27.6% methotrexate, 12.2% with rituximab and 9% with mycophenolate mofetil.

No deaths were attributed directly to IgG4-RD.

**Conclusions:** In our registry, the most frequent initial manifestation was autoimmune pancreatitis. Phenotype 1 was more frequent in men and phenotype 3 in women.