Autoimmune pancreatitis (AIP) is a rare form of chronic pancreatitis characterized by an autoimmune inflammatory process; there is a lymphocytic infiltrate associated with pancreatic fibrosis, causing organic dysfunction. The first descriptions began in the 1990s, and the disease is often underdiagnosed.

The aim of this study is to systematically analyze PAI's characteristics by reviewing the bibliography of the PUBMED and Scielo databases.

Autoimmune mechanisms such as hypergammaglobulinemia, elevated serum IgG levels, especially IgG4, or the presence of autoantibodies are characteristic of AIP. The clinical features are nonspecific and are similar to other pancreatopathies. Studies have shown that 11% of patients with AIP received a diagnosis based on histological findings. Incidence and prevalence values are not known because it is a generally underdiagnosed pathology that has two subtypes: type 1 and type 2. Type 1 AIP is the pancreatic manifestation of a systemic disease associated with a high concentration of IgG4 immunoglobulin.

Type 2 AIP is a specific localized and exclusive disorder of the pancreas. Both show effective responses to corticosteroid therapy. Diagnosis requires a high degree of clinical suspicion and depends on histological study. It is of great importance that physicians carefully manage cases of pancreatitis so that it is possible to diagnose autoimmune pancreatitis, aiming at a better prognosis.

AIP is a recent pathology that represents, in clinical practice, growing relevance. This condition needs to be further studied in order to establish a more accurate diagnosis and treatment with the lowest possible recurrence rate.

Autoimmune pancreatitis is characterized by high concentrations of IgG4 immunoglobulin. The clinical features are nonspecific, and diagnosis is based on histological findings. Type 1 AIP is associated with a systemic disease, while Type 2 is a localized and exclusive disorder of the pancreas. Both respond well to corticosteroids. Early recognition is crucial for better outcomes.