## Type 2 AIP-Korean experience and tips on diagnosis/management

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Autoimmune pancreatitis (AIP) has two distinct subtypes; type 1 & type 2. Whereas type 1 AIP is recognized as the pancreatic manifestation of a systemic fibroinflammatory disorder named IgG4-related disease, type 2 AIP is a pancreas-specific disorder not associated with IgG4. In Korea, type 1 appears to be the predominant form of AIP while in Western countries, a mix of type 1 & 2 is encountered.

The similarities between both types are the pancreatic imaging findings and the response to the steroid therapy. However, patients with type 2 tend to be younger and are more likely to present with abdominal pain and clinical pancreatitis than those with type 1. There is an absence of elevated serum IgG4 and extrapancreatic manifestations, which provide clues in diagnosing type 1 AIP. Inflammatory bowel disease is more common in type 2 than type 1. Type 2 has no specific biomarker and shows minimal/absent IgG4 immunostaining from pancreatic tissues. The identifying granulocytic epithelial lesion (GEL) in the core biopsy/resection specimen is needed for a definitive diagnosis of type 2 AIP is likely to be underrecognized. Type 2 AIP patients experienced a lower relapse after steroid therapy. Accurate subtyping of AIP may have an impact on predicting a maintenance therapy. Our understanding of the clinical spectrum of type 2 AIP continues to evolve.