

# AUTOIMMUNE PANCREATITIS

*Autoimmune pancreatitis (AIP) is chronic inflammation of the pancreas of autoimmune etiology.*

## ■ CHARACTERISTIC FEATURES

- Enlargement of pancreas and narrowing of the main pancreatic duct.
- Presence of autoantibodies and elevated immunoglobulins, particularly IgG4.
- Histopathology showing lymphoplasmacytic inflammation and fibrosis.

## ■ EPIDEMIOLOGY

The prevalence of AIP is between 5 and 6% of all patients with chronic pancreatitis. AIP affects both sexes, but it is twice as common in men as in women. Most patients are older than 50 years.

## ■ CAUSE

The cause of AIP is unknown, but current evidence strongly suggests an autoimmune basis for the disease. It is frequently associated with rheumatoid arthritis, Sjögren's syndrome, and inflammatory bowel disease.

## ■ CLINICAL FEATURES

Patients with AIP present with a wide variety of symptoms, but most common symptoms are abdominal pain and

jaundice. Occasionally, patients present with symptoms related to extrapancreatic organ involvement. Inflammatory bowel disease (ulcerative colitis or, less commonly, Crohn's disease) is frequently associated with AIP.

## ■ DIAGNOSTIC IMAGING

Endoscopic retrograde cholangiopancreatography is mandatory for the diagnosis of AIP. The enlargement of the pancreas (*Figure 1*) with ductal narrowing often with stenosis. The differential diagnosis of the focal form of AIP rather than pancreatic carcinoma can be very difficult on the basis of CT imaging only. However, the finding of diffuse pancreatic-ductal narrowing is highly diagnostic of AIP.

## ■ PATHOLOGY

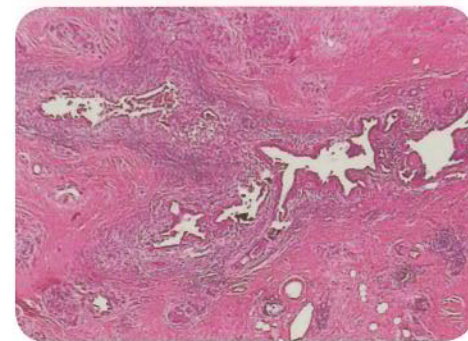
The histologic hallmark of AIP is lymphoplasmacytic infiltration with fibrosis in the pancreas (*Figure 2*). Abundance of IgG4-positive plasma cells has been found both within the pancreas and at the extrapancreatic sites of involvement.

## ■ LAB STUDIES

The elevated serum IgG4 level is nearly diagnostic of AIP. IgG4 shows specificity of 98% and sensitivity of 80-92% when cutoff values of 135mg/dl are used. IgG levels are also elevated and shows specificity of 73% and sensitivity of 75%. Antinuclear antibodies (ANA) and rheumatoid factor (RF) levels are also increased. A combination of IgG4, IgG, ANA and RF increases the sensitivity of detecting AIP to 97%. (*See Table.*)



**Figure 1.** Enlargement of Pancreas in a CT in a patient with AIP.



**Figure 2.** Lymphoplasmacytic infiltration with fibrosis in the pancreas around a duct.

## ■ MANAGEMENT

AIP responds dramatically to oral steroid therapy in contrast to other chronic pancreatitis patients. The usual initial dose of corticosteroids (prednisone) is of 40 mg daily for 1 week, followed by a taper of the daily dose by 5 mg per week.

**TABLE: SENSITIVITY AND SPECIFICITY OF IMMUNOLOGICAL MARKERS FOR AIP**

Analyte	Sensitivity (%)	Specificity (%)
IgG4	80 – 92	98
IgG	72 – 73	75
Antinuclear Antibodies (ANA)	63 – 66	–
Rheumatoid Factor (RF)	20 – 28	–
IgG4, IgG, ANA and RF	97	98

#### DIAGNOSTIC CRITERIA\*

**1. Imaging studies of pancreatic parenchyma:**

- a. Show a diffuse/segmental/focally enlarged gland, occasionally with a mass and/or a hypoattenuation rim.
- b. Show diffuse/segmental/focal pancreatic ductal narrowing, often with stenosis of the bile duct.

Both (a.) and (b.) are required for diagnosis.

**2. Common lymphoplasmacytic infiltration and fibrosis, with abundant IgG4-positive cell infiltration.**

**3. Elevated level of serum IgG or IgG4, and detection of autoantibodies.**

**AIP is diagnosed when criterion 1 and one of the other two criteria are satisfied.**

*\*Otsuki M et al. Asian diagnostic criteria for autoimmune pancreatitis: consensus of the Japan-Korea Symposium on Autoimmune Pancreatitis. J Gastroenterol. 2008;43:403-8.*

#### SUGGESTED READING

Finkelberg DL, Sahani D, Deshpande V, Brugge W. Autoimmune Pancreatitis. N Engl J Med 2006; 355:2670-2676.

Kamisawa T, Okazaki K, Kawa S. Diagnostic criteria for autoimmune pancreatitis in Japan. World J Gastroenterol. 2008; 14:4992-4.

Kawa S, Hamano H. Serological markers for the diagnosis of autoimmune pancreatitis. J Jpn Pancreas Soc 2007; 22:641-645.

#### IMMCO TESTS

**Autoimmune Pancreatitis Panel:**

Test Code: 188 (ANA; Rheumatoid Factor; IgG; IgG4)

Methodology: IFA, ELISA and Nephelometry

Schedule/Turnaround Time: 7 days.

**Specimen Requirements:** Specimen need not be refrigerated or frozen. Collect 5-10ml of blood in a red top or serum separator tube (minimum: 0.5 mL). If possible, separate serum from the clot and place into orange tube provided with the IMMCO collection kits. Do not puncture top of orange tube. If separation facilities are not available, the blood can be sent in the tube used for collection.

**Sample Stability:** Sample is stable at ambient temperature during shipment. If sample is stored prior to shipment, it is stable at refrigerated (2-8°C) up to five days and frozen (-20°C) up to a year.

#### SAMPLE SUBMISSION

Specimen collection kits are available free of charge. **Please call 1-800-537-8378 or e-mail service@immco.com for supplies.**

Specimen can be shipped by courier services, U.S. Postal service and overnight carriers free of charge. Results are reported within two business days of the receipt of the specimen via mail, fax and through IMMCO online, a HIPAA-compliant web tool at [www.immco.com](http://www.immco.com).

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