

# Liver disease

The liver is an important organ with responsibility for many critical functions (for example, metabolism of protein and fats, storage, bile acid production and detoxification (toxic drugs & chemicals). Inability to perform these functions leads to liver disease. In this section, autoimmune liver disease (such as autoimmune hepatitis (AIH), primary biliary cirrhosis (PBC), primary sclerosing cholangitis (PSC), and autoimmune cholangiopathy and overlap syndromes) and its immune markers are reviewed.

[Open all sections](#)

## Autoimmune liver diseases

A variety of clinical signs and symptoms may be associated with the liver disease and the evidence of autoimmune involvement is characterised by chronic progressive inflammation and hypergammaglobulinaemia and the presence of specific high titre auto-antibodies is often evident.

**Primary Biliary Cirrhosis (PBC):** Anti-mitochondrial antibodies (AMA) are associated with this disease and affects older females with an incident of 90%. The cause is unknown but it is thought that infection might be a trigger. The most common target for these antibodies is M2 antigen, the pyruvate dehydrogenase complex. A number of the subunits of the pyruvate dehydrogenase may be targeted.

**Autoimmune hepatitis (AIH):** This affects about 90% of younger female patients and is categorised by seropositivity for antinuclear antibody (ANA) and/or smooth muscle or antibodies to liver kidney microsomes (LKM). **Type 1 AIH** is associated with high titre of ANA and /or AMA, whereas **type 2 AIH** is associated with LKM. There are several antigens identified associated with AIH, for example, liver kidney microsome, liver cytosolic (LC-1), liver cytokeratine, glutathione-S-transferase, soluble liver antigen, actin in smooth muscle and ANA.

Table: Summary of antibodies in liver disease.

Antibody	Antigen	AIH1	AIH2	PBC	Overlap
<a href="#">ANA (/facilities/clinical-immunology-services/autoimmunity/hep-2-image-library/nuclearplus.aspx)</a>	Sp 100 (nuclear matrix), Gp 210 (Rim ANA)	+		+	+
ASM	Actin, Vimentin, Tubulin	+			+/-
<a href="#">LKM1 (/facilities/clinical-immunology-services/autoimmunity/liver-associated-antibodies/Liver-antibodies.aspx)</a>	Cytochrome P450		+		
LC1	Formiminotransferase Cyclodeaminase		+		
SLA/LP	UGA suppressor tRNA associated protein	+	+		+
M2	pyruvate dehydrogenase complex			+	+

The decision tree below shows diagnostic outcome based on the immunological profile of a serum from a patient with liver disease.

## Decision tree



