

# Autoimmune Liver Diseases

## Autoantibodies provoking permanent liver damage

### What are autoimmune liver diseases?

Autoimmune liver diseases occur when the body's immune system attacks cells of liver, bile ducts and gallbladder. These diseases affect globally people of all ages and show common characteristics with other liver diseases such as hepatitis, alcohol-induced liver disease and cirrhosis. The three main categories of autoimmune liver disease are autoimmune hepatitis (AIH), primary biliary cirrhosis (PBC), and primary sclerosing cholangitis (PSC); all three diseases are well-defined entities with diagnosis based upon a constellation of clinical, serologic, and liver pathology findings.<sup>1</sup>

### Prevalence of autoimmune hepatitis in different countries of the world

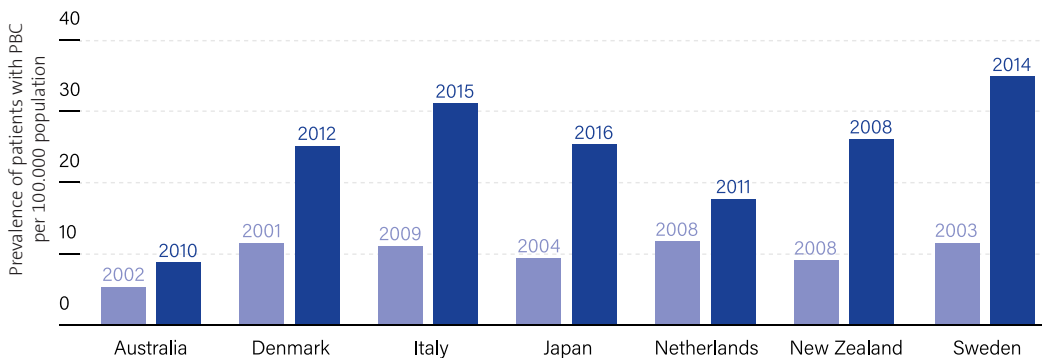
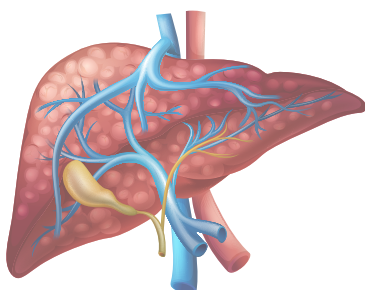


Fig. 1. The point prevalence (per 100,000 population) of autoimmune hepatitis in different countries, reported after 2000. (adapted from Tanaka)<sup>2,3</sup>

### Liver affected by autoimmune diseases

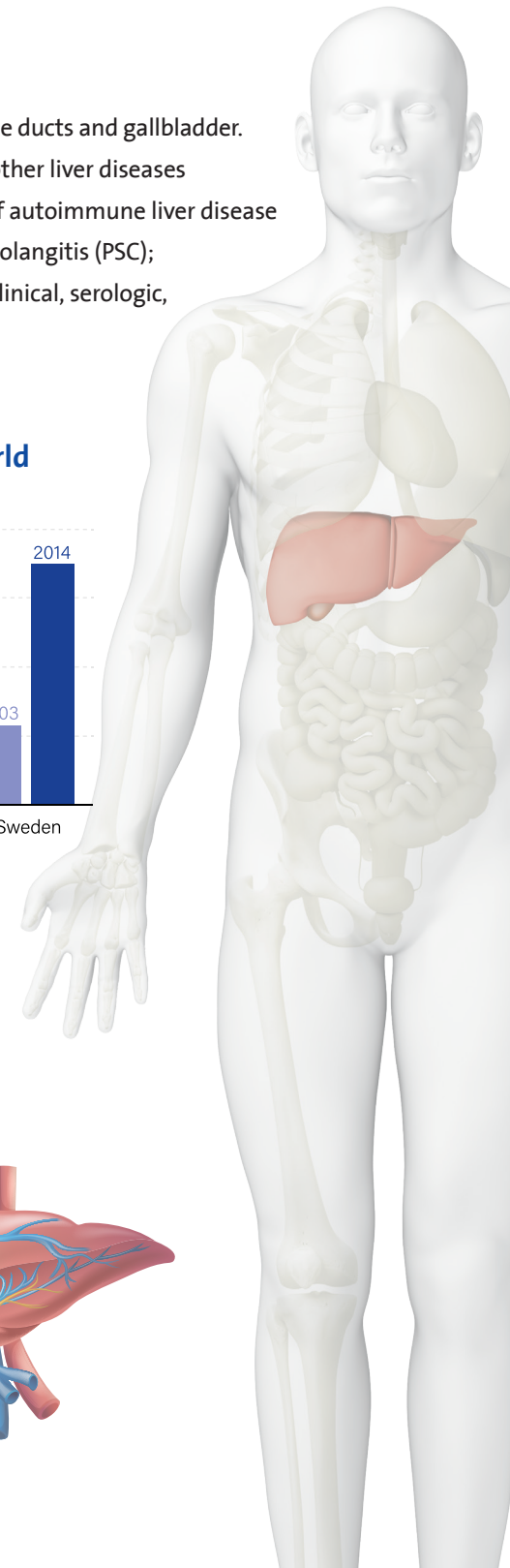
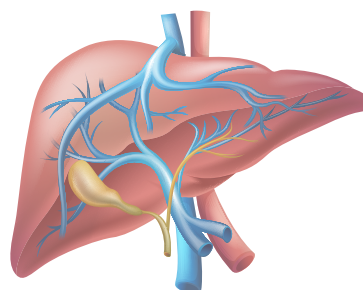
#### Inflamed liver

Symptoms occurring with liver inflammation:



- > Fatigue
- > Abdominal swelling, distention, or bloating
- > Diarrhea
- > Jaundice
- > Low-grade fever
- > Nausea with or without vomiting

#### Healthy liver



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## How are autoantibodies used for diagnostics?

Looking at a characteristic morphologic liver patterns of autoimmune hepatitis, it shows that the pattern is similar to chronic hepatitis. In PBC the destruction of small intrahepatic bile ducts and canals is clearly visible, while in PSC periductal fibrosis and inflammation involving larger bile ducts with variable small duct damage is detectable. A serum testing will offer more insights as antimicrobial antibodies will be found in a patient with PBC, antinuclear, anti-smooth muscle, and anti-LKM antibodies are prominent in AIH, and pANCA in PSC. Although most cases of autoimmune liver disease fit into one of these three categories, overlap syndromes (primarily of AIH with PBC or PSC) can occur in up to 10% of all patients. The diseases can gain additional complexity due to sequential syndromes with transition from one form of autoimmune liver disease to another.<sup>1,4,5</sup>

## Parameter disease correlation

|  | PBC | PSC | AIH Type 1 | AIH Type 2 | AIH Type 3 |
|--|-----|-----|------------|------------|------------|
| <b>ANA</b><br>anti-nuclear antigens            | ●   | ●   | ●          | ●          | ●          |
| <b>LKM-1</b><br>anti-liver-kidney microsomal 1 |     |     |            | ●          |            |
| <b>LC-1</b><br>anti-liver cytosolic 1          |     |     |            | ●          |            |
| <b>SLA/LP</b><br>anti-soluble liver antigen    |     |     | ●          |            | ●          |
| <b>AMA-M2</b><br>anti-mitochondrial M2         | ●   |     |            |            |            |
| <b>Sp100</b><br>anti-nuclear antigen 100 kDa   | ●   |     |            |            |            |
| <b>gp210</b><br>anti-glycoprotein 210 kDa      | ●   |     |            |            |            |
| <b>LBR</b><br>anti-lamin B receptor            | ●   |     |            |            |            |
| <b>p-ANCA</b><br>anti-neutrophil cytoplasmic   |     | ●   |            |            |            |

Fig. 2. Parameter disease correlation. (adapted from Shoenfeld and Meroni)<sup>4</sup>

## Ordering information

| Name                        | Unit/Size | Format      | REF      |
|-----------------------------|-----------|-------------|----------|
| <b>LIVER-LIA</b>            | 24 tests  | IgG         | ITC66205 |
| <b>AMA M2</b>               | 96 Tests  | IgG         | ITC60040 |
| <b>LKM-1--ANTIBODIES</b>    | 96 tests  | IgG         | ITC66050 |
| <b>ANA SCREEN</b>           | 96 tests  | IgG/IgA/IgM | ITC60001 |
| <b>ANA Screen (cut-off)</b> | 96 tests  | IgG/IgA/IgM | ITC70001 |
| <b>ANA LIA</b>              | 24 Tests  | IgG         | ITC92000 |

## References

1. Autoimmune liver disease: overlap and outliers - Washington, Modern Pathology, 2007
2. Autoimmune hepatitis: 2019 Update, Tanaka, Gut and Liver, 2020
3. Current understanding of primary biliary cholangitis, Tanaka, Clinical and Molecular Hepatology, 2021
4. The general practice guide to autoimmune diseases - Shoenfeld and Meroni, 2012
5. Autoantibodies in systemic autoimmune diseases - Conrad et al. 2015

