

# How Confident Are You in Diagnosing Primary Biliary Cholangitis (PBC)?

- PBC is an **autoimmune cholestatic condition** that is a significant cause of chronic liver disease<sup>1</sup>
- In PBC, **immune-mediated biliary epithelial cell damage, cholestasis and progressive fibrosis** can lead to **end-stage liver disease**<sup>1</sup>
- It is most prevalent in **women and individuals over 50 years of age**<sup>1-3</sup>

## Early diagnosis of PBC is essential to improve patient outcomes<sup>1</sup>

- **Earlier diagnosis** and initiation of disease management has led to an **improvement in PBC prognosis** in recent years, although it remains a significant cause of liver-related morbidity
- While improved detection of serologic markers facilitates earlier diagnosis,<sup>5</sup> **varying patient presentation** and **non-specific symptoms** mean that diagnosis remains difficult<sup>6</sup>

## What are the signs of PBC?

**~40%** of people with PBC are **symptomatic** at the time of diagnosis<sup>7</sup>

Those with **asymptomatic PBC** are usually diagnosed by chance with **laboratory findings** from a routine health check or during the course of an unrelated illness<sup>8</sup>

### Clinical symptoms may include:

- |  |   |                                       |
|--|---|---------------------------------------|
| <ul style="list-style-type: none"> <li>• <b>Fatigue</b><sup>2</sup></li> <li>• <b>Pruritus</b><sup>2</sup></li> <li>• <b>Autoimmune disease</b><sup>9</sup></li> </ul>   | } | <p><b>Common symptoms</b></p>         |
| <ul style="list-style-type: none"> <li>• <b>Jaundice</b><sup>2</sup></li> <li>• <b>Cognitive impairment</b><sup>10</sup></li> <li>• <b>Abdominal pain</b><sup>11</sup></li> <li>• <b>Nausea, bloating or diarrhea</b><sup>6</sup></li> </ul> | } | <p><b>Other reported symptoms</b></p> |

### Abnormal liver function tests may include:

- **Elevated ALP**<sup>12</sup>
- **Elevated GGT**<sup>1</sup>
- **Elevated ALT and/or AST**<sup>12</sup>
- **Elevated serum total bilirubin**<sup>13</sup>
- **Increased levels of IgM and/or IgG**<sup>12</sup>
- **AMA positive**<sup>12</sup>
- **Anti-gp210 and/or anti-sp100 positive**<sup>12</sup>

ALP, alkaline phosphatase; ALT, alanine aminotransferase; AMA, antimitochondrial autoantibody; AST, aspartate aminotransferase; gp210, 210 kDA glycoprotein; IgG, immunoglobulin G; IgM, immunoglobulin M; sp100, speckled 100 kDA protein.

1. Hirschfield GM et al. *Gut* 2018;67:1568–1594; 2. Dahlqvist G et al. *Hepatology* 2017;65:152–163; 3. Kim WR et al. *Gastroenterol* 2000;119:1631–1636; 4. Prince MI and James OFW. *Clin Liver Dis* 2003;7:795–819; 5. Beuers U et al. *Am J Gastroenterol* 2015;110:1536–1538; 6. PBCers Organization. Available at: [www.pbcers.org/symptoms/](http://www.pbcers.org/symptoms/). Accessed June 2024; 7. Prince MI et al. *Gut* 2004;53:865–870; 8. Kurtovic J et al. *Q J Med* 2005; 98:331–336; 9. Liu Y et al. *Can J Gastroenterol Hepatol* 2021;55:57814; 10. Newton JL et al. *Hepatology* 2008;48:541–549; 11. Laurin JM et al. *J Gastroenterol* 1994;89:1840–1843; 12. Lindor KD et al. *Hepatology* 2019;69:394–420; ; 13. Lammers WJ et al. *Gastroenterology* 2014;147(6):1338–1349

Diagnosis of PBC is confirmed with tests for autoantibodies:<sup>1</sup>

## AMA

AMA is the **hallmark serologic signature** of PBC,<sup>2</sup> present in **~95%** of patients<sup>3,4</sup>

## ANA

**PBC-specific ANA** may be present in **>50%** of patients with PBC,<sup>5</sup> including those who are AMA negative<sup>6</sup>

A diagnosis of PBC can be established when **two out of the three** criteria below are met:<sup>1</sup>

1

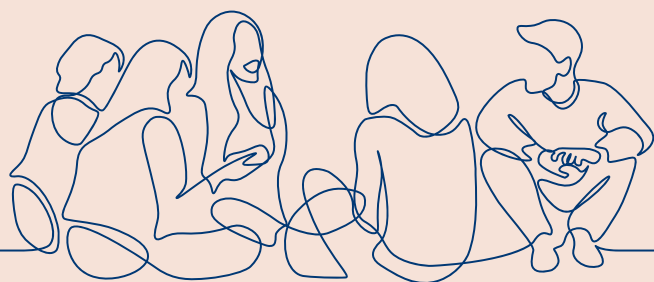
Biochemical evidence of cholestasis based on ALP elevation

2

Presence of AMA or other PBC-specific autoantibodies, including anti-sp100 and/or anti-gp210

3

Histologic evidence of nonsuppurative destructive cholangitis and destruction of interlobular bile ducts



Usually, PBC meets the first two criteria and can be diagnosed without the need for a liver biopsy. **A liver biopsy is only required if laboratory tests are inconclusive<sup>1</sup>**

ALP, alkaline phosphatase; **AMA**, antimitochondrial autoantibody; **ANA**, antinuclear antibodies; **gp210**, 210 kDA glycoprotein; **PBC**, primary biliary cholangitis; **sp100**, speckled 100 kDA protein.

1. Lindor KD et al. *Hepatology* 2019;69:394–420; 2. Hirschfield GM et al. *Gut* 2018;67:1568–1594; 3. Walker JG et al. *Lancet* 1965;1:827–831; 4. Kaplan MM and Gershwin ME. *New Eng J Med* 2005;353:1261–1273; 5. Rigopoulou EI et al. *Gut* 2005;54:528–532; 6. Granito A et al. *Aliment Pharm Ther* 2006;24:1575–1583.

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