

# What is Primary Biliary Cholangitis (PBC)?

**PBC is a progressive autoimmune disease of the liver.**<sup>1</sup>

- PBC is estimated to affect **4.3 people per 100,000 per year in the US**<sup>2</sup>
- It is characterized by T-cell mediated **destruction of bile ducts**<sup>3,4</sup>
- **Fatigue and pruritus** are common symptoms of PBC<sup>5</sup>
- PBC can lead to **liver fibrosis, cirrhosis, and end-stage liver disease**<sup>1</sup>

**PBC affects more women than men**<sup>2,5-7</sup>

- Most prevalent in **women aged 40–80 years**<sup>5,7,8</sup>
- Estimated to affect **>1 in 1,000 women** over the age of 40<sup>9</sup>

**The presentation of PBC ranges from asymptomatic to a host of symptoms**

## Common symptoms:



**Fatigue**<sup>5</sup>



**Pruritus**<sup>5</sup>



**Autoimmune diseases**<sup>10</sup>



**Dry eyes and mouth (Sicca syndrome)**<sup>5</sup>

## Other reported symptoms include:



**Jaundice**<sup>11</sup>



**Abdominal discomfort**<sup>12</sup>



**Restless legs**<sup>13</sup>



**Cognitive impairment**<sup>14</sup>

Concurrent rheumatologic or autoimmune disease are common with PBC<sup>10</sup>

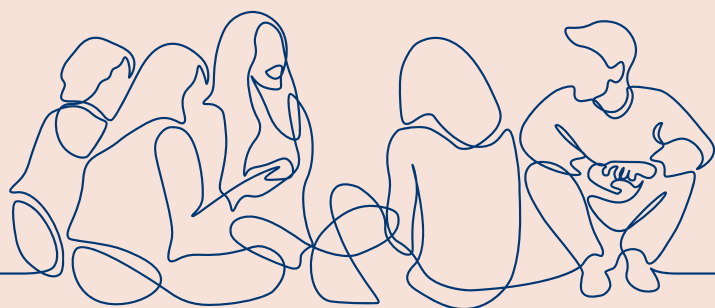
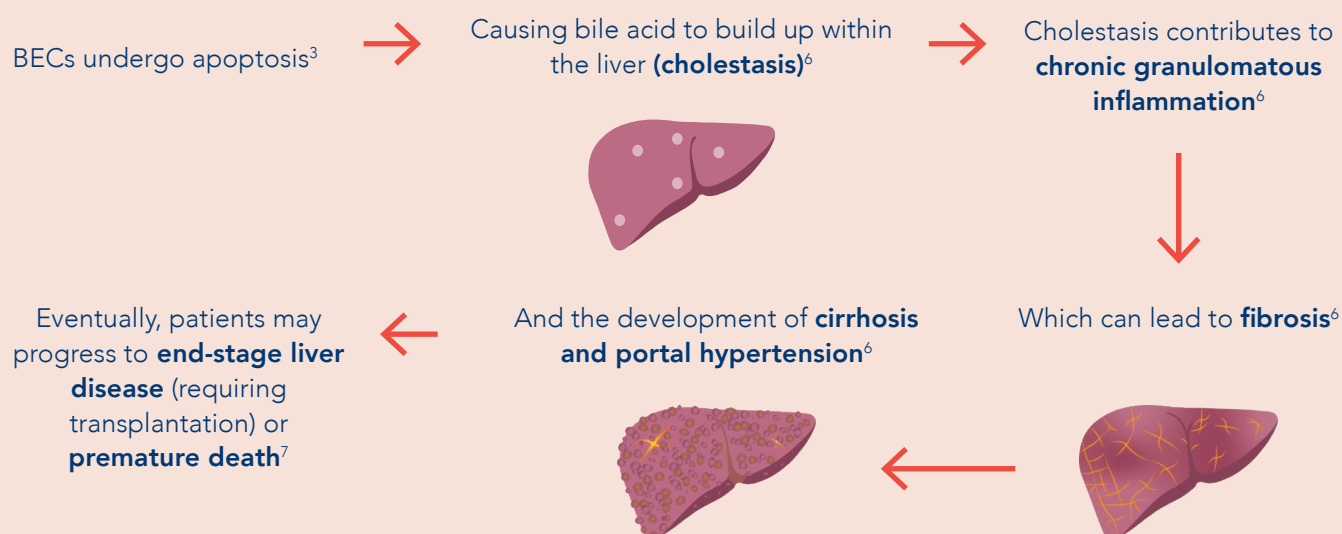
~60% of patients with PBC are asymptomatic at the time of diagnosis<sup>15</sup>

1. Galoosian A et al. *J Clin Transl Hepatol* 2020;8:49–60; 2. Lu M et al. *Clin Gastroenterol Hepatol* 2018;16:1342–1350; 3. Yamada G et al. *Hepatology* 1986;6:385–391; 4. Van Den Oord JJ et al. *Liver* 1984;4: 264–274; 5. Trivella J, et al. *Hepatology Communications* 2023;7:e0179; 6. Gershwin ME et al. *Hepatology* 2005;42:1194–1202; 7. Kim WR et al. *Gastroenterol* 2000;119:1631–1636; 8. Kim KA et al. *Ailment Pharmacol Ther* 2016;43:154–162; 9. Ma D et al. *Heliyon* 10 2024;e25634; 10. Liu Y et al. *Can J Gastroenterol Hepatol* 2021; 55:57814; 11. Reshetnyak V et al. *World J Gastroenterol* 2015;21:7683–7708; 12. Laurin JM et al. *J Gastroenterol* 1994;89:1840–1843; 13. Anderson K et al. *Liver Int* 2013;33:239–243; 14. Newton JL et al. *Hepatology* 2008;48:541–549; 15. Prince MI et al. *Gut* 2004;53:865–870.

## In PBC, T-cell-mediated injury against intralobular biliary epithelial cells (BECs) causes progressive destruction of the bile ducts<sup>1,2</sup>

- Apoptosis of BECs leads to the aberrant expression of autoantigens<sup>3</sup>
- Recognition of autoantigens by **anti-mitochondrial antibody (AMA)** results in the formation of an immune complex<sup>3,4</sup>
  - Further activating the immune system and leading to **persistent and widespread BEC damage**<sup>3,4</sup>

**Bile ducts are integral to the liver's regulation of bile acid metabolism.<sup>5</sup> Their destruction leads to the build-up of bile and other toxins in the liver (cholestasis)<sup>6</sup>**



Progression to fibrosis and cirrhosis among people with PBC is becoming rarer due to increasing awareness and understanding of the natural history, as well as earlier diagnosis and prompt treatment.<sup>6</sup>

1. Jones D et al. *Clin Exp Immunol* 1997;107:562–568; 2. Van Den Oord JJ et al. *Liver* 1984; 4: 264–274; 3. Lleo A et al. *Hepatology* 2009;49:871–879; 4. Lleo A et al. *Hepatology* 2010;52:987–998; 5. Mattner J. *Int J Mol Sci* 2016;17:1864; 6. Galoosian A et al. *J Clin Transl Hepatol* 2020;8:49–60; 7. Lammers W et al. *Ann Hepatol* 2014;13:316–326.