

Primary biliary cholangitis (PBC) is a chronic, or long lasting, progressive liver disorder that mostly affects women and usually appears during middle age. PBC leads to inflammation and scarring of the small bile ducts (the plumbing system of the liver which transports bile, the substance that helps digest fat). When PBC is very severe, it can lead to yellow discoloration of the skin (jaundice). If PBC is untreated, it can lead to cirrhosis, or scarring of the entire liver, which can lead to liver failure. PBC is divided into four stages, with stage 1 being early disease, where there is no significant scarring, to stage 4, or which is defined by cirrhosis. Although the exact cause of PBC is unknown, it is thought that it is likely due to a combination of factors such as autoimmune (when a person's own immune system attacks their body), genetic, and environmental factors. PBC affects mostly women, but more men are now being diagnosed. The disorder usually becomes apparent during middle age, initially affecting most individuals between the ages of 45 to 65 years. However, the disorder has been diagnosed in females as young as 22 years of age and in females in their early 90s. It has been estimated that PBC is one of the most common autoimmune diseases, affecting nearly 1 in 1000 women over the age of 40.