# Is it time to reassess your patients with progressive cholestasis?

Certain patient presentations of progressive cholestatic liver disease (ChLD) may signal the need for further assessment

Actor portrayal.

## Idiopathic Cholestasis

**CONSIDER REASSESSING IF** signs of cholestasis manifest without apparent cause. Presentation is highly variable, but symptoms can include[1-3]:

- Jaundice
- Pruritus
- Abnormal stools
- Abnormal liver parameters for bilirubin or transaminases*

In addition to clinical symptoms, **MEDICAL HISTORY MAY ALSO REFLECT:**

- **Suffers from long-term symptoms and has not found proper relief**
- **Has sought care without receiving a definitive diagnosis**

## Cholestasis With Pruritus or Unusual Presentation

**CONSIDER REASSESSING IF** your patient is receiving care for another liver disease but has unusual symptoms, including:

- **Small duct PSC[1]**
- **AMA negative PBC[4,5]**
- **NAFLD with pruritus[6]**
- **Lean NAFLD without metabolic syndrome[6]**
- **Lean NASH with pruritus and without metabolic syndrome[6]**

## Secondary Cholestasis Triggered by Liver Issue

**CONSIDER REASSESSING IF** symptoms of cholestatic pruritus arise in patients who have recently experienced liver issues, including:

- **All women with ICP[1]**
- **Drug-induced cholestasis[1]**
- **Hormonal-induced cholestasis triggered by birth control, menopause, etc[1,7]**

## History of Complicated Gallstones

**CONSIDER REASSESSING IF** your patient has a complicated history of gallstones, including:

- **Intrahepatic gallstones[1]**
- **Very strong family history of gallstones and incident at a young age[8,9]**
- **LPAC leading to stones in the gallbladder or liver[10]**

*Including GGT, AST, ALT, or ALP.

ALP=alkaline phosphatase; ALT=alanine aminotransferase; AMA=antimitochondrial antibody; AST=aspartate aminotransferase; GGT=gamma-glutamyl transferase; ICP=intrahepatic cholestasis of pregnancy; LPAC=low phospholipid-associated cholelithiasis; NAFLD=nonalcoholic fatty liver disease; NASH=nonalcoholic steatohepatitis; PBC=primary biliary cholangitis; PSC=primary sclerosing cholangitis.

# Could adult progressive familial intrahepatic cholestasis (PFIC) be hiding in your practice?

PFIC is a rare and life-threatening type of cholestatic liver disease with several subtypes. While previously believed to present only in early infancy, PFIC can manifest later in life after a trigger—or patients can experience a long and complicated path to diagnosis.[11]

## **Diagnosing** PFIC can be challenging in adolescent and older patients

**REASONS include[11,12]:**

- Symptoms are difficult to identify and often overlooked
- Presentation is highly variable and often not considered classical

## **Identifying** PFIC is the first step for providing appropriate treatment

**MULTIPLE ASSESSMENTS can be used to help support a clinical diagnosis of PFIC:**

- **Common symptoms**, especially pruritus and jaundice, along with elevated serum bile acids and the presence of gastrointestinal symptoms, like diarrhea, are key indicators of PFIC[11]
- **Lab results** with abnormal transaminase levels and high levels of bilirubin, as well as abnormally high levels of serum bile acids, could be predictors of PFIC[13]
- **Imaging** can be used to help rule out other conditions:
  - Performing a cholangiography can rule out extrahepatic conditions[14,15]
  - An ultrasound can identify liver damage progression, extrahepatic causes of cholestasis, and help distinguish PFIC from other forms of cholestasis[14,16]

Genetic testing can reinforce a suspected diagnosis of PFIC, but in some patients, testing can be inconclusive or indeterminate. Support from a geneticist may be required.[13]

**There may be more adolescent and adult patients with PFIC in your practice than you think. Confirming a diagnosis is vital to addressing their ChLD.**

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