THE RECOMMENDED DIAGNOSTIC PROCESS

**Patient with suspected primary biliary cholangitis (PBC)**

- Persistent cholestatic abnormalities in serum biochemistry
  - Elevated liver enzymes including ALP, GGT, AST, ALT
  - Elevated conjugated bilirubin
- Symptoms of PBC, including:
  - Pruritus
  - Sicca
  - Arthralgias
  - Fatigue

**Initial assessment**

- Personal/family history, physical examination, abdominal ultrasound
- Serum (blood) biochemistry test for ALP
- Serum antibody (serology) test for AMA and/or PBC-specific ANA
  - AMA is present in 90% of PBC cases
  - ANA is present in 30% of PBC cases

**Establish a secure diagnosis of PBC**

- Elevated ALP
- AMA-positive (>1/140) or anti-gp210/anti-sp100-positive

**Confirmed PBC**

---

RECOMMENDATIONS FOR MANAGING PBC

**Initiate 1st-line treatment**

- Ursodeoxycholic acid (UDCA) at 13-15 mg/kg/day

**Assess risk of progression based on response to treatment**

<table>
<thead>
<tr>
<th>Low risk</th>
<th>Intermediate-to-high risk</th>
<th>Consider referral for further assessment</th>
</tr>
</thead>
</table>
| ■ Adequate response to UDCA, e.g.  
  □ ALP <1.5 x ULN AND  
  □ Normal bilirubin AND  
  □ Early fibrosis  
| ■ Intolerance to UDCA OR  
  □ ALP >1.5 x ULN OR  
  □ Bilirubin >ULN OR  
  □ Albumin <LLN OR  
  □ Increasing fibrosis/cirrhosis  
| ■ Presence of any one of:  
  □ Decompensated cirrhosis (Child-Pugh B or C)  
  □ Bilirubin >2 x ULN  
  □ AST or ALT >5 x ULN  
  □ Severe pruritus  

**Continue UDCA and assess response every 12 months**

**Further assessment to evaluate risk-benefit of 2nd-line treatment**

**Is risk-benefit positive in favour of treatment?**

- NO Tertiary referral
- YES Continue UDCA and assess response every 12 months

**Initiate 2nd-line treatment**

- Obeticholic acid

**On-treatment assessment and regular follow-up**

- Monitor blood tests every 3-6 months (depending on patient risk profile): ALP, bilirubin, AST, ALT, GGT, albumin and platelets
- Monitor evolution of fibrosis yearly using elastography (imaging test that measures liver stiffness)

**Continue therapy if treatment response is adequate**

**Evidence of disease progression?**

- NO
- YES Further assessment

ALP: Alkaline phosphatase.  
AMA: Antimitochondrial antibodies.  
ANA: Antinuclear antibodies.  
AST: Aspartate aminotransferase.  
GGT: Gamma-glutamyltranspeptidase.  
LLN: Lower limit of normal.  
ULN: Upper limit of normal.

---

THE RECOMMENDED DIAGNOSTIC PROCESS

**Patient with suspected primary biliary cholangitis (PBC)**

- Persistent cholestatic abnormalities in serum biochemistry
  - Elevated liver enzymes including ALP, GGT, AST, ALT
  - Elevated conjugated bilirubin
- Symptoms of PBC, including:
  - Pruritus
  - Sicca
  - Arthralgias
  - Fatigue

**Initial assessment**

- Personal/family history, physical examination, abdominal ultrasound
- Serum (blood) biochemistry test for ALP
- Serum antibody (serology) test for AMA and/or PBC-specific ANA
  - AMA is present in 90% of PBC cases
  - ANA is present in 30% of PBC cases

**Establish a secure diagnosis of PBC**

- Elevated ALP
- AMA-positive (>1/140) or anti-gp210/anti-sp100-positive

**Confirmed PBC**

---

RECOMMENDATIONS FOR MANAGING PBC

**Initiate 1st-line treatment**

- Ursodeoxycholic acid (UDCA) at 13-15 mg/kg/day

**Assess risk of progression based on response to treatment**

<table>
<thead>
<tr>
<th>Low risk</th>
<th>Intermediate-to-high risk</th>
<th>Consider referral for further assessment</th>
</tr>
</thead>
</table>
| ■ Adequate response to UDCA, e.g.  
  □ ALP <1.5 x ULN AND  
  □ Normal bilirubin AND  
  □ Early fibrosis  
| ■ Intolerance to UDCA OR  
  □ ALP >1.5 x ULN OR  
  □ Bilirubin >ULN OR  
  □ Albumin <LLN OR  
  □ Increasing fibrosis/cirrhosis  
| ■ Presence of any one of:  
  □ Decompensated cirrhosis (Child-Pugh B or C)  
  □ Bilirubin >2 x ULN  
  □ AST or ALT >5 x ULN  
  □ Severe pruritus  

**Continue UDCA and assess response every 12 months**

**Further assessment to evaluate risk-benefit of 2nd-line treatment**

**Is risk-benefit positive in favour of treatment?**

- NO Tertiary referral
- YES Continue UDCA and assess response every 12 months

**Initiate 2nd-line treatment**

- Obeticholic acid

**On-treatment assessment and regular follow-up**

- Monitor blood tests every 3-6 months (depending on patient risk profile): ALP, bilirubin, AST, ALT, GGT, albumin and platelets
- Monitor evolution of fibrosis yearly using elastography (imaging test that measures liver stiffness)

**Continue therapy if treatment response is adequate**

**Evidence of disease progression?**

- NO
- YES Further assessment

ALP: Alkaline phosphatase.  
AMA: Antimitochondrial antibodies.  
ANA: Antinuclear antibodies.  
AST: Aspartate aminotransferase.  
GGT: Gamma-glutamyltranspeptidase.  
LLN: Lower limit of normal.  
ULN: Upper limit of normal.