Physician Algorithm

PBC Algorithm

Algorithm-

The diagnosis of PBC can be established when two of the following three criteria are met:

1. Biochemical evidence of cholestasis based on ALP elevation.
2. Presence of AMA
3. Histologic evidence of nonsuppurative destructive cholangitis and destruction of interlobular bile ducts.

Treatment:

1. UDCA in a dose of 13 to 15 mg/kg/day orally is recommended for patients with PBC who have abnormal liver enzyme values regardless of histologic stage.
2. Biochemical response to UDCA should be evaluated at 6 to 12 months after treatment initiation to determine whether patients should be considered for second-line therapy.
3. If intolerance to UCDA then start Ocaliva or Iqirvo monotherapy. If incomplete biochemical response to UCDA, then add Ocaliva or Iqirvo.
    A. Ocaliva Dosing — 5mg once daily x 3 months. If incomplete response, then increase to 10mg daily
    B. Ocaliva contraindicated with decompensated cirrhosis or compensated cirrhosis with portal hypertension
    C. Iqirvo dosing – 80 mg once daily
    D. Iqirvo – avoid use in Child Pugh class B and C
4. If concern for advanced fibrosis or cirrhosis by imaging, or lab markers (FIB 4 or APRI), then consideration of elastography to determine fibrosis.
5. EGD for variceal screening and HCC screening if cirrhosis

References:

- Lindor KD et al. Primary Biliary Cholangitis: 2018 Practice Guidance From the American Association for the Study of Liver Diseases 2018.
- Louie JS et al. Primary Biliary Cholangitis: A Brief Overview. Clinical Liver Disease, VOL 15, NO 3, MARCH 2020.