Liver ultrasonography

**Bile ducts?**

- **Dilated**
  - Cholangiography and/or surgery

- **Extrahepatic causes**

  - Not dilated
    - Serum gamma GT activity?

      - **Normal**
        - Pruritus?
          - **No**
            - Defect in primary bile acid synthesis (mainly 3β-HSD deficiency)

          - **Yes**
            - PFIC1 or PFIC2
              - BRIC
              - Drug toxicity
              - Hepatitis A

      - **Elevated**
        - Alagille syndrome
          - α1-AT deficiency
          - Cystic fibrosis
          - Drug toxicity

        - Liver biopsy
          - Ductular proliferation?
          - If yes: cholangiography

            - **Bile ducts?**
              - **Abnormal**
                - Sclerosing cholangitis

              - **Normal**
                - PFIC3
                - Autoimmune cholangitis