**Autoimmune Liver Disease Testing**

**INDICATIONS FOR TESTING**
Liver disease of unknown etiology (all viral serologies negative, elevated transaminases)

ORDER
- ANA testing by IFA
- F-actin antibody (SMA) testing
- LKM-1 antibody testing
- AMA testing

Positive result(s)
- ANA and/or F-actin/SMA positive

ORDER
- ANCA testing to differentiate between AIH-1 and PSC

Atypical pANCA positive
- Yes
  - Suggests PSC
  - Perform cholangiography
- No
  - Suggests AIH

Inconclusive
- May suggest AIH/PSC overlap

CONSIDER
- Anti-sp100 and anti-gp210 antibody testing by IgG

Positive
- Suggests PBC

Negative result(s)
- Any test positive

ORDER
- ANCA testing
- SLA testing
- Anti-sp100 and anti-gp210 antibody testing
- LC-1 testing

Any test negative
- SLA positive
  - ANCA negative
  - LC-1 positive
  - Anti-sp100 positive
  - Anti-gp210 positive
  - Atypical pANCA positive
  - SLA negative
  - LC-1 negative
  - Suggests AIH

Anti-sp100 positive
Anti-gp210 negative
- Suggests PBC

LC-1 negative
- Suggests AIH-1

LC-1 positive
- Suggests AIH-2

May suggest AIH/PSC overlap

**Test Key**
- AMA: Antimitochondrial antibody
- ANA: Antinuclear antibody
- ANCA: Antineutrophil cytoplasmic antibody
- IFA: Indirect fluorescent antibody
- IgG: Immunoglobulin G
- LC-1: Liver cystolic antigen type 1 antibody
- LKM-1: Liver-kidney microsome type 1 (cytochrome P450 2D6)
- pANCA: Perinuclear antineutrophil cytoplasmic antibody
- SLA: Soluble liver antigen
- SMA: Smooth muscle antibody

**Disease Key**
- AIC: Autoimmune cholangitis
- AIH: Autoimmune hepatitis
- AIH-1: Autoimmune hepatitis type 1
- AIH-2: Autoimmune hepatitis type 2
- CCH: Chronic cryptogenic hepatitis
- PBC: Primary biliary cholangitis
- PSC: Primary sclerosing cholangitis
- UC: Ulcerative colitis

*If ANA and atypical pANCA are both positive, cholangiography and biopsy may be indicated to rule out AIH/PSC overlap syndrome; inconclusive staining patterns may also indicate need for imaging or biopsy.