Liver Cytosolic Antigen Type 1 (LC-1) Antibody

Indications for Ordering

- Differential evaluation of autoimmune liver disease (ALD) of unknown etiology, especially autoimmune hepatitis (AIH) of childhood onset
- Consider:
  - If other serologic tests in ALD panel are negative, **or**
  - In combination with specific tests:
    - Anti-liver-kidney microsome 1 (LKM1)
    - ANA
    - F-actin
    - SMA

Test Description

Liver-Kidney Microsome-1 Antibody, IgG
- Semiquantitative enzyme-linked immunosorbent assay (ELISA)

Liver Cytosolic Antigen Type 1 (LC-1) Antibody, IgG
- Qualitative immunoblot

Tests to Consider

Primary Tests

Liver-Kidney Microsome - 1 Antibody, IgG 0055241
- Use in combination with Liver Cytosolic Antigen Type 1 (LC-1) Antibody, IgG
- More likely to be positive than LC-1

Liver Cytosolic Antigen Type 1 (LC-1) Antibody, IgG 2010711
- Use in combination with Liver-Kidney Microsome - 1 Antibody, IgG

Related Tests

Autoimmune Liver Disease Reflexive Panel 3002479
- Recommended initial screening panel when ALD strongly suspected
- Negative results do not rule out disease
- Consider further evaluation with less frequently occurring autoantibody tests based on clinical presentation and history

ANCA-Associated Vasculitis Profile (ANCA/MPO/PR3) with Reflex to ANCA Titer 2006480
- Initial test in conjunction with Autoimmune Liver Disease Reflexive Panel for evaluation of ALD

Disease Overview

Incidence

AIH: 0.85-1.9/100,000 per year for white adults of northern European ancestry

Prevalence

- ALD
  - 5% of all liver diseases
- AIH type 1
  - Most common type of AIH
- AIH type 2
  - Rare: 4% of AIH individuals in the U.S.

Age

- AIH type 1: bimodal peaks (10-30 years, 40-50 years)
- AIH type 2: childhood

Physiology

AIH
- Etiology: antibodies directed against the liver
- Chronic course with slow progression: may resemble other chronic liver diseases (eg, alcoholic cirrhosis, chronic viral hepatitis)

Clinical Presentation

- Clinical features vary widely, ~25% asymptomatic
  - Arthralgias
  - Anorexia, fatigue, lethargy, malaise
  - Hepatomegaly, nausea, upper abdominal pain, jaundice
    - Progression to cirrhosis and liver failure possible
- Antibody-negative disease
  - Same presentation and histology as antibody-positive AIH

Diagnostic/Prognostic Issues

- May be difficult to clinically distinguish between AIH types 1 and 2
- Important to distinguish AIH type 1 from type 2 (prognostically)
  - Higher risk of fulminant course and progression to cirrhosis in type 2
  - Cirrhosis often present in children at time of diagnosis

- Antibody testing may be helpful for diagnosis
  - Antibodies to LKM1 and/or LC-1 are diagnostic for AIH type 2
    - LKM1 antibodies should be analyzed by measuring antibodies to cytochrome P4502D6
- Diagnosis of exclusion
  - No other etiology found for liver disease/cirrhosis – key to this diagnosis
- Liver biopsy may be appropriate in certain individuals
  - Should be performed when diagnosis is still unclear
  - Considered "gold standard"

Test Interpretation

Typical antibody pattern in AIH-1

- pANCA: atypical staining
- SMA, F-actin: positive
- ANA: homogeneous pattern most common
- LC-1: negative
- SLA: variably positive, more common in children

Results

Typical antibody pattern in AIH-2

- pANCA: positive (rare) or negative
- LKM-1: positive
- LC-1: positive
- ANA, SMA, F-actin, M2, SLA: negative

Limitations

- Negative antibody testing does not rule out ALD
- All interpretation of antibody patterns must be done in conjunction with clinical presentation
  - Overlap may occur between diseases and antibodies
- Neither LKM-1 nor LC-1 has absolute diagnostic sensitivity for AIH type 2