Anti-Fibrillarin Antibody

Indications for Ordering

- Recommended for the diagnosis of systemic sclerosis in patients negative for centromere, Scl-70, or RNA polymerase III antibodies
- May predict skeletal muscle involvement and pulmonary arterial hypertension

Test Description

Qualitative immunoblot

Tests to Consider

Typical testing strategy

- Initial testing
  - CBC with platelet count and automated differential
  - Antinuclear antibody (ANA) by IFA
- Secondary testing based on IFA pattern
  - Scleroderma (Scl-70)
  - RNA polymerase III

Primary test

Fibrillarin (U3 RNP) Antibody, IgG 2012173

Related test

Systemic Sclerosis Panel 2012057

Disease Overview

Incidence – 3-20/million

Age of onset – peak onset 20-30 years

Sex – M<F, 1:3-8

Ethnicity

Anti-fibrillarin (U3-RNP) antibody has a higher prevalence in individuals of African-American descent

Symptoms

Constellation of symptoms, including

- Cardiovascular – eg, rhythm disturbances
- Dermatologic – eg, digital ulcers
- Gastrointestinal – eg, dysmotility
- Musculoskeletal – eg, myopathy
- Pulmonary – eg, fibrosis
- Renal – glomerulonephritis
- Scleroderma

Diagnostic issues

- Autoimmune connective tissue diseases may present with similar features, particularly early in disease, making diagnosis difficult
- ANA IFA patterns may help define diagnostic pathways
  - Most patients with SSc will have at least one of the following antibodies, and these antibody tests are adequate for initial evaluation (van den Hoogen, 2013)
    - Centromere
    - Scl-70
    - RNA polymerase III
  - Some patients with clinical suspicion of SSc are negative for the 3 antibodies above
    - May have a less common antibody (eg, U3-RNP IgG)
    - U3-RNP IgG
    - Detected more frequently in African-American patients with SSc compared to other ethnic groups
  - Distinct clinical features
    - Younger age at disease onset
    - Organ involvement
      - Myositis
      - Pulmonary hypertension
      - Renal disease

Test Interpretation

Results

Negative

Limitations

- Negative test result does not rule out the diagnosis of SSc
- Test results alone are not diagnostic
  - Results should be used in conjunction with other laboratory tests and clinical findings

Reference