Antifibrillarin 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
Criteria Systemic Sclerosis Panel 3000479

Disease Overview
Incidence: 3-20/million
Age of onset: peak onset 20-30 years
Sex: M<F, 1:3-8

Ethnicity
Antifibrillarin (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
- Sicca syndrome

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 three 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