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**