

# Antifibrillar Antibody

## Indications for Ordering

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- 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

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Qualitative immunoblot

## Tests to Consider

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### 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

[Fibrillar \(U3 RNP\) Antibody, IgG 2012173](#)

### Related test

[Criteria Systemic Sclerosis Panel 3000479](#)

## Disease Overview

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**Incidence** – 3-20/million

**Age of onset** – peak onset 20-30 years

**Sex** – M<F, 1:3-8

### Ethnicity

Antifibrillar (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

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### 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

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van den Hoogen F, Khanna D, et al. 2013 classification criteria for systemic sclerosis: an American college of rheumatology/European league against rheumatism collaborative initiative. *Ann Rheum Dis.* 2013;72:1747-1755