Overview

Useful For
Evaluating patients suspected of having systemic sclerosis, when used in conjunction with centromere and Scl70 antibodies

Providing diagnostic and prognostic information in patients with systemic sclerosis

Method Name
Enzyme-LinkedImmunosorbentAssay(ELISA)

NY State Available
Yes

Specimen

Specimen Type
Serum

Specimen Required
Container/Tube:

Preferred: Serum gel

Acceptable: Red top

Specimen Volume: 0.5 mL

Specimen Minimum Volume
0.4 mL

Reject Due To

<table>
<thead>
<tr>
<th>Gross hemolysis</th>
<th>Reject</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gross lipemia</td>
<td>Reject</td>
</tr>
<tr>
<td>Gross icterus</td>
<td>OK</td>
</tr>
</tbody>
</table>

Specimen Stability Information

<table>
<thead>
<tr>
<th>Specimen Type</th>
<th>Temperature</th>
<th>Time</th>
<th>Special Container</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum</td>
<td>Refrigerated (preferred)</td>
<td>21 days</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Frozen</td>
<td>21 days</td>
<td></td>
</tr>
</tbody>
</table>

Clinical and Interpretive
Clinical Information

Systemic sclerosis is a multisystem connective tissue (systemic rheumatic) disease characterized by fibroblast dysfunction leading to fibrosis of the skin and internal organs, microvascular injury leading to tissue hypoxia, and an autoimmune response manifested by production of autoantibodies. The severity of the disease is highly variable among individual patients. Limited cutaneous systemic sclerosis and diffuse cutaneous systemic sclerosis have been recognized as distinct subsets, with worse survival for those with the diffuse form. Clinical features of CREST syndrome (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasias) can be seen in both limited cutaneous and diffuse cutaneous forms but, overall, are associated with a better prognosis. Several disease-specific and mutually exclusive autoantibodies have been identified that are helpful in both diagnosis and disease classification. Centromere and topoisomerase I (Scl 70) autoantibodies are associated with limited cutaneous systemic sclerosis and diffuse cutaneous systemic sclerosis, respectively.

RNA polymerase III is a complex, 16-subunit enzyme directing transcription of small, stable nontranslated RNA genes: tRNA, 5S rRNA, Alu RNA, U6 snRNA, and 7SK snRNA genes. The immunodominant epitope for autoantibodies with anti-RNA polymerase I/III specificity has been identified on the RNA polymerase-specific subunit RPC155.

Autoantibodies to RNA polymerase III antigen are found in 11% to 23% of patients with systemic sclerosis. Systemic sclerosis patients who are positive for RNA polymerase III antibodies form a distinct serologic subgroup and usually do not have any of the other antibodies typically found in systemic sclerosis patients such as anticentromere or anti-Scl70. Numerous studies have shown that systemic sclerosis patients with anti-RNA polymerase III have an increased risk of the diffuse cutaneous form of scleroderma, with a high likelihood of skin involvement and hypertensive renal disease.

Reference Values

- <20.0 U (negative)
- 20.0-39.9 U (weak positive)
- 40.0-80.0 U (moderate positive)
- >80.0 U (strong positive)

Interpretation

A positive result supports a possible diagnosis of systemic sclerosis (see Cautions). This autoantibody is strongly associated with diffuse cutaneous scleroderma and with an increased risk of acute renal crisis.

A negative result indicates no detectable IgG antibodies to RNA polymerase III, but does not rule out the possibility of systemic sclerosis (11%-23% sensitivity).

Cautions

A positive result indicates the presence of measurable IgG antibodies to RNA polymerase III, but does not unequivocally establish the diagnosis of systemic sclerosis or other autoimmune disease.

The level of RNA polymerase III autoantibodies does not indicate the severity of disease in patients with systemic sclerosis.

The presence of immune complexes or other immunoglobulin aggregates in the patient specimen may cause an increased level of nonspecific binding and produce false-positive results with this assay.

Clinical Reference
Test Definition: RNAP

RNA Polymerase III Ab, IgG, S


Performance

Method Description

The immunodominant fragment of RNA polymerase III antigen is derived from recombinant DNA technology. Purified RNA polymerase III antigen is adsorbed to the wells of a polystyrene microtiter plate under conditions that preserve the antigen in its antigenic state. Prediluted controls and diluted patient sera are added to separate wells. Unbound sample is washed away and an enzyme-labeled antihuman IgG conjugate is added to each well. After incubation and washing away of unbound enzyme-labeled antihuman IgG, the bound conjugate is measured by adding a chromogenic substrate. The intensity of the absorbance produced is measured with an automated microwell plate reader. Results are calculated by comparison to a single-point calibrator. (Package insert: QUANTA Lite RNA Pol III, INOVA Diagnostics, San Diego, CA. 1/2015)

PDF Report

No

Day(s) and Time(s) Test Performed

Wednesday

Analytic Time

1 day

Maximum Laboratory Time

7 days

Specimen Retention Time

14 days

Performing Laboratory Location

Rochester

Fees and Codes

Fees

- Authorized users can sign in to Test Prices for detailed fee information.
- Clients without access to Test Prices can contact Customer Service 24 hours a day, seven days a week.
- Prospective clients should contact their Regional Manager. For assistance, contact Customer Service.
**Test Definition: RNAP**
RNA Polymerase III Ab, IgG, S

**Test Classification**
This test has been cleared, approved or is exempt by the U.S. Food and Drug Administration and is used per manufacturer's instructions. Performance characteristics were verified by Mayo Clinic in a manner consistent with CLIA requirements.

**CPT Code Information**
83516

<table>
<thead>
<tr>
<th>LOINC® Information</th>
</tr>
</thead>
<tbody>
<tr>
<td>Test ID</td>
</tr>
<tr>
<td>RNAP</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Result ID</th>
<th>Test Result Name</th>
<th>Result LOINC Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>RNAP</td>
<td>RNA Polymerase III Ab, IgG, S</td>
<td>79182-2</td>
</tr>
</tbody>
</table>