Overview

Useful For
Supporting the diagnosis of IgG4-related disease

Method Name
Nephelometry

NY State Available
Yes

Specimen

Specimen Type
Serum

Advisory Information
This test only quantitates the IgG4 protein. If quantitation of all IgG subclass types is wanted, order IGGS / IgG Subclasses, Serum.

Specimen Required
Patient Preparation: Fasting preferred but not required

Container/Tube:
Preferred: Serum gel
Acceptable: Red top

Specimen Volume: 1 mL

Specimen Minimum Volume
0.5 mL

Reject Due To

<table>
<thead>
<tr>
<th>Condition</th>
<th>Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gross hemolysis</td>
<td>OK</td>
</tr>
<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>14 days</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ambient</td>
<td>14 days</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Frozen</td>
<td>14 days</td>
<td></td>
</tr>
</tbody>
</table>
Clinical and Interpretive

Clinical Information
The most abundant immunoglobulin (Ig) isotype in human serum is immunoglobulin G (IgG). IgG immunoglobulins are comprised of 4 subclasses designated IgG1 through IgG4. Of total IgG, approximately 65% is IgG1, 25% is IgG2, 6% is IgG3, and 4% is IgG4. Each IgG subclass contains structurally unique portions of the constant region of the gamma heavy chain.

IgG subclass 4-related disease is a recently recognized syndrome of unknown etiology most often occurring in middle-aged and older men. Several organ systems can be involved and encompasses many previous and newly described diseases such as type1 autoimmune pancreatitis; Mikulicz disease and sclerosing sialadenitis; inflammatory orbital pseudotumor; chronic sclerosing aortitis; Riedel thyroiditis, a subset of Hashimoto thyroiditis; IgG4-related interstitial pneumonitis; and IgG4-related tubulointerstitial nephritis. Each of these entities is characterized by tumor-like swelling of the involved organs with infiltrative, predominately IgG4-positive, plasma cells with accompanying "storiform" fibrosis. In addition, elevated serum concentrations of IgG4 are found in 60% to 70% of patients diagnosed with IgG4-related disease.

The diagnosis of IgG4-related disease requires a tissue biopsy of the affected organ demonstrating the aforementioned histological features. It is recommended that patients suspected of having an IgG4-related disease have their serum IgG4 level measured.

Reference Values
0-<5 months: < or =19.8 mg/dL
5-<9 months: < or =20.8 mg/dL
9-<15 months: < or =22.0 mg/dL
15-<24 months: < or =23.0 mg/dL
2-<4 years: 0.4-49.1 mg/dL
4-<7 years: 0.8-81.9 mg/dL
7-<10 years: 1.0-108.7 mg/dL
10-<13 years: 1.0-121.9 mg/dL
13-<16 years: 0.7-121.7 mg/dL
16-<18 years: 0.3-111.0 mg/dL
> or =18 years: 2.4-121.0 mg/dL

Interpretation
Elevated levels of IgG4 are consistent with, but not diagnostic of, IgG4-related disease.

Cautions
Elevations in serum IgG4 concentrations are not specific to IgG4-related disease; they are also found in disorders...
such as multicentric Castleman disease, allergic disorders, Churg-Strauss syndrome, sarcoidosis, and a large number of other conditions.

Clinical Reference

Performance

Method Description
In this Siemens Nephelometer II method, the light scattered onto the antigen-antibody complexes is measured. The intensity of the measured scattered light is proportional to the amount of antigen-antibody complexes in the sample under certain conditions. If the antibody volume is kept constant, the signal behaves proportionally to the antigen volume.

A reference curve is generated by a standard with a known antigen content on which the scattered light signals of the samples can be evaluated and calculated as an antigen concentration. Antigen-antibody complexes are formed when a sample containing antigen and the corresponding antiserum are put into a cuvette. A light beam is generated with an LED, which is transmitted through the cuvette. The light is scattered onto the immuno-complexes that are present. Antigen and antibody are mixed in the initial measurement, but no complex is formed yet. An antigen-antibody complex is formed in the final measurement.

The result is calculated by subtracting value of the final measurement from the initial measurement. The distribution of intensity of the scattered light depends on the ratio of the particle size of the antigen-antibody complexes to the radiated wavelength.(Unpublished Mayo method; Instruction manual: Siemens Nephelometer II, Version 3, Siemens, Inc., Newark, DE, 2008)

PDF Report
No

Day(s) and Time(s) Test Performed
Monday through Saturday; Continuously

Analytic Time
Same day/1 day

Maximum Laboratory Time
2 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 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

82787

LOINC® Information

<table>
<thead>
<tr>
<th>Test ID</th>
<th>Test Order Name</th>
<th>Order LOINC Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>IGGS4</td>
<td>Immunoglobulin Subclass IgG4, S</td>
<td>2469-5</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>IGGS4</td>
<td>Immunoglobulin Subclass IgG4, S</td>
<td>2469-5</td>
</tr>
</tbody>
</table>