Overview

Useful For
Investigation of a patient with a low (absent) hemolytic complement (CH50), with reflex testing to C3 and C4, if appropriate

Reflex Tests

<table>
<thead>
<tr>
<th>Test ID</th>
<th>Reporting Name</th>
<th>Available Separately</th>
<th>Always Performed</th>
</tr>
</thead>
<tbody>
<tr>
<td>C4</td>
<td>Complement C4, S</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>C3</td>
<td>Complement C3, S</td>
<td>Yes</td>
<td>No</td>
</tr>
</tbody>
</table>

Testing Algorithm
If the C2 result is less than 15 U/mL, then complement C3 and C4 will be performed at an additional charge.

Method Name
Automated Liposome Lysis Assay

NY State Available
Yes

Specimen

Specimen Type
Serum Red

Advisory Information
This test is for assessment of complement C2 and includes assessment of C3 and C4 as reflex testing. Unless a deficiency has already been identified, initial assessment should begin with the total complement assay (COM / Complement, Total, Serum), which is a screen for suspected complement deficiencies and should be performed before ordering individual complement component assays. A deficiency of an individual component of the complement cascade will result in an undetectable total complement level.

Specimen Required

Patient Preparation: Fasting preferred but not required

Supplies: Aliquot Tube, 5 mL (T465)

Collection Container/Tube: Red top

Submission Container/Tube: Plastic vial

Specimen Volume: 1 mL

Collection Instructions:
1. Immediately after specimen collection, place the tube on wet ice.

2. Centrifuge and aliquot serum into plastic vial.

3. Immediately freeze specimen.

**Specimen Minimum Volume**

0.5 mL

**Reject Due To**

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

**Specimen Stability Information**

<table>
<thead>
<tr>
<th>Specimen Type</th>
<th>Temperature</th>
<th>Time</th>
<th>Special Container</th>
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</thead>
<tbody>
<tr>
<td>Serum Red</td>
<td>Frozen</td>
<td>21 days</td>
<td></td>
</tr>
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</table>

**Clinical and Interpretive**

**Clinical Information**

The classic pathway of the complement system is composed of a series of proteins that are activated in response to the presence of immune complexes. This activation process results in the formation of the lytic membrane attack complex, as well as the generation of activation peptides that are chemotactic for neutrophils and that bind to immune complexes and complement receptors. The absence of early components (C1, C2, C4) of the complement cascade results in the inability of immune complexes to activate the cascade. Patients with deficiencies of the early complement proteins are unable to generate lytic activity or to clear immune complexes. These patients have increased susceptibility to infections with encapsulated microorganisms. They may also have symptoms that suggest autoimmune disease, and complement deficiency may be an etiologic factor in the development of autoimmune disease.

Although rare, C2 deficiency is the most common inherited complement deficiency. Homozygous C2 deficiency has an estimated prevalence ranging from 1 in 10,000 to 1 in 40,000 (the prevalence of heterozygotes is 1 in 100 to 1 in 50). Half of the homozygous patients are clinically normal.

However, discoid lupus erythematosus or systemic lupus erythematosus (SLE) occurs in approximately one-third of patients with homozygous C2 deficiency. Patients with SLE and a C2 deficiency frequently have a normal anti-ds DNA titer. Clinically, many have lupus-like skin lesions and photosensitivity, but immunofluorescence studies may fail to demonstrate immunoglobulin or complement along the epidermal-dermal junction.

Other diseases reported to be associated with C2 deficiency include dermatomyositis, glomerulonephritis, vasculitis, atrophoderma, cold urticaria, inflammatory bowel disease, and recurrent infections.

The laboratory findings that suggest C2 deficiency include a hemolytic complement (CH50) of nearly zero, with normal values for C3 and C4.
Reference Values
25-47 U/mL

Interpretation
Low levels of complement may be due to inherited deficiencies, acquired deficiencies, or due to complement consumption (eg, as a consequence of infectious or autoimmune processes).

Absent (or low) C2 levels in the presence of normal C3 and C4 values are consistent with a C2 deficiency.

Low C2 levels in the presence of low C3 and C4 values are consistent with a complement-consumptive process.

Low C2 and C4 values, in the presence of normal values for C3 is suggestive of C1 esterase inhibitor deficiency.

Cautions
As with all complement assays, proper sample handling is of utmost importance to ensure that the complement system is not activated before clinical testing.

Absent (or low) C2 functional levels in the presence of normal C2 antigen levels should be replicated with a new serum specimen to confirm that C2 inactivation has not occurred during shipping.

Clinical Reference

Performance

Method Description
C2 complement activity is measured by mixing patient serum with a C2-deficient serum. The lytic activity of the serum mixture is tested against sensitized, labeled liposomes. If lysis occurs, the patient serum must be the source of the C2. The target liposomes are a commercial reagent (WAKO total complement CH50) and the assay is performed on a Siemens Advia XPT.(Unpublished Mayo method)

PDF Report
No

Day(s) and Time(s) Test Performed
Monday through Friday; 3 p.m.

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 was developed and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the U.S. Food and Drug Administration.

CPT Code Information
86161

86160 x 2 (if appropriate)

LOINC® Information

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<thead>
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<th>Test ID</th>
<th>Test Order Name</th>
<th>Order LOINC Value</th>
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<tbody>
<tr>
<td>C2</td>
<td>C2 Complement, Functional, w/ Reflex, S</td>
<td>93977-7</td>
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<table>
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<th>Test Result Name</th>
<th>Result LOINC Value</th>
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<tbody>
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<td>C2FX</td>
<td>C2 Complement, Functional, S</td>
<td>93977-7</td>
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<tr>
<td>INT53</td>
<td>Interpretation</td>
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