Overview

Useful For
Investigation of suspected alternative pathway complement deficiency, atypical hemolytic uremic syndrome, C3 glomerulonephritis, dense-deposit disease

Method Name
Enzyme-Linked Immunosorbent Assay (ELISA)

NY State Available
Yes

Specimen

Specimen Type
Serum Red

Advisory Information
To screen for most complement abnormalities, order COM / Complement, Total, Serum.

Specimen Required
Patient Preparation: Patient should be fasting.

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 at 4 degrees C and separate serum from clot.
3. Freeze specimen within 30 minutes.

Forms
If not ordering electronically, complete, print, and send a Renal Diagnostics Test Request (T830) with the specimen.

Specimen Minimum Volume
0.2 mL

Reject Due To
<table>
<thead>
<tr>
<th>Gross hemolysis</th>
<th>OK</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gross lipemia</td>
<td>OK</td>
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<tr>
<td>Gross icterus</td>
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**Test Definition: AH50**
Complement, Alternate Path, Func, S

### Specimen Stability Information

<table>
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<tr>
<th>Specimen Type</th>
<th>Temperature</th>
<th>Time</th>
<th>Special Container</th>
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<tbody>
<tr>
<td>Serum Red</td>
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<td>14 days</td>
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### Clinical and Interpretive

#### Clinical Information

Complement proteins are components of the innate immune system. There are 3 pathways to complement activation: (1) the classical pathway, (2) the alternative (or properdin) pathway, and (3) the lectin (or mannose-binding lectin, MBL) pathway.

The total complement (CH50) assay (COM / Complement, Total, Serum) is the best screening assay for most complement abnormalities. It assesses the classical complement pathway including early components that activate the pathway in response to immune complexes, as well as the late components involved in the membrane attack complex. The CH50 assay will be abnormal if there are specific hereditary or acquired C1-C9 complement component deficiencies or if there is consumption of complement due to immune (or autoimmune) complexes.

The AH50 assay is a screening test for complement abnormalities in the alternative pathway. The alternate pathway shares C3 and C5-C9 components, but has unique early complement components designated factors D, B, and P, as well as regulatory factors H and I. This pathway can be activated by hydrolysis of C3 or by microbial polysaccharides and does not require immune complex formation. Patients with disseminated infections with pyogenic bacteria in the presence of a normal CH50 may have a decreased AH50 due to hereditary or acquired deficiencies of the alternate pathway. Patients with deficiencies in the alternate pathway factors (D, B, P, H, and I) or late complement components (C3, C5-C9) are unusually susceptible to recurrent Neisserial meningitis. The use of the CH50 and AH50 assays allow identification of the specific pathway abnormality.

Unregulated alternative pathway can also result in disease. The majority of these diseases present with renal function impairment such as atypical hemolytic uremic syndrome (a-HUS), dense deposit disease (DDD), and C3 glomeulonephritis (C3GN).

#### Reference Values

> or =46% normal

#### Interpretation

Absent complement alternate pathway (AH50) in the presence of a normal total hemolytic complement (CH50) suggests an alternate pathway component deficiency.

Normal AH50 with absent CH50 suggests an early (C1, C2, C4) classic pathway deficiency.

Absent AH50 and CH50 suggests a late (C3, C5, C6, C7, C8, C9) component deficiency or complement consumption.

Absent AH50 and CH50 in the presence of a normal C3 and C4 suggests a late (C5, C6, C7, C8, C9) component deficiency.

#### Cautions

This assay is a functional test and is dependent on correct sampling, storage, and shipping conditions.
An absent complement alternate pathway (AH50) should be confirmed with a repeat test on a different specimen.

**Clinical Reference**


**Performance**

**Method Description**

The Weislab complement assay for the alternative pathway combines principles of the hemolytic assay for complement activation with the use of labeled antibodies specific for neoantigens produced as a result of complement activation. The micro titer plate strips are coated with lipopolysaccharide (LPS). Patient serum is diluted in diluent containing specific blocker to ensure that only the alternative pathway is activated. During the first incubation, the diluted patient serum in the wells is activated by the coating. The wells are then washed and C5b-9 (MAC) is detected with a specific alkaline phosphatase labeled antibody to the neoantigen expressed during MAC formation. After a final wash, an alkaline phosphatase substrate is added. The amount of alternative pathway complement activity correlates with the color intensity of the solution and is measured in terms of absorbance (optical density: OD). (Nordin JG, Truedsson L, Sjoholm A: New procedure for detection of complement deficiency by ELISA, J Imm Methods 1993;166:655-668)

**PDF Report**

No

**Day(s) and Time(s) Test Performed**

Varies

**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 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

LOINC® Information

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