

Overview**Useful For**

Diagnosis of C9 deficiency

Investigation of a patient with a low total (hemolytic) complement (CH50) level

Method Name

Automated Liposome Lysis Assay

NY State Available

Yes

Specimen**Specimen Type**

Serum Red

Ordering Guidance

The total complement (CH50) assay (COM / Complement, Total, Serum) should be used as a screen for suspected complement deficiencies 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

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

| | |
|-----------------|--------|
| Gross hemolysis | OK |
| Gross lipemia | Reject |

| | |
|---------------|----|
| Gross icterus | OK |
|---------------|----|

Specimen Stability Information

| Specimen Type | Temperature | Time | Special Container |
|---------------|-------------|---------|-------------------|
| Serum Red | Frozen | 14 days | |

Clinical and Interpretive

Clinical Information

Complement proteins are components of the innate immune system. There are 3 pathways to complement activation: 1) the classic pathway, 2) the alternative (or properdin) pathway, and 3) the lectin activation (mannan-binding protein: MBP) pathway. 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. The activation process results in the generation of peptides that are chemotactic for neutrophils and that bind to immune complexes and complement receptors. The end result of the complement activation cascade is the formation of the lytic membrane attack complex (MAC).

Patients with deficiencies of the late complement proteins (C5, C6, C7, C8, and C9) are unable to form the MAC, and may have increased susceptibility to neisserial infections.

C9 deficiency is common in the Japanese population and has been reported to occur in almost 1% of the population. The lytic activity of C9-deficient serum is decreased. However, the assembly of C5b-C8 complexes will result in a transmembrane channel with lytic activity, although the lytic activity is reduced. Many C9-deficient patients are therefore asymptomatic. C9-deficient patients may, however, present with invasive neisserial infections.

Complement levels can be detected by antigen assays that quantitate the amount of the protein. For most of the complement proteins, a small number of cases have been described in which the protein is present but is nonfunctional. These rare cases require a functional assay to detect the deficiency.

Reference Values

37-61 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 component 9 (C9) levels in the presence of normal C3 and C4 values are consistent with a C9 deficiency. Absent C9 levels in the presence of low C3 and C4 values suggests complement consumption.

Normal results indicate both normal C9 protein levels and normal functional activity.

Cautions

Absent (or low) complement component 9 (C9) functional levels in the presence of normal C9 antigen levels should be replicated with a new serum specimen to confirm that C9 inactivation did not occur during shipping.

Clinical Reference

1. Sonntag J, Brandenburg U, Polzehl D, et al: Complement systems in healthy term newborns: reference values in umbilical cord blood. *Pediatr Dev Pathol* 1998;1:131-135
2. Prellner K, Sjöholm AG, Truedsson L: Concentrations of C1q, factor B, factor D and properdin in healthy children, and the age-related presence of circulating C1r-C1s complexes. *Acta Paediatr Scand* 1987;76:939-943
3. Davis ML, Austin C, Messmer BL, et al: IFCC-standardization pediatric reference intervals for 10 serum proteins using the Beckman Array 360 system. *Clin Biochem* 1996;29(5):489-492
4. Gaither TA, Frank MM: Complement. In *Clinical Diagnosis and Management by Laboratory Methods*. 17th edition. Edited by JB Henry. WB Saunders Company, 1984, pp 879-892
5. O'Neil KM: Complement deficiency. *Clin Rev Allergy Immunol* 2000;19:83-108
6. Frank MM: Complement deficiencies. *Pediatr Clin North Am* 2000;47(6):1339-1354

Performance

Method Description

Component 9 (C9) complement activity is measured by mixing patient serum with a C9-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 C9. The target liposomes are a commercial reagent (WAKO total complement CH50), and the assay is performed on an Advia XPT. (Unpublished Mayo method)

PDF Report

No

Day(s) Performed

Monday through Friday

Report Available

Same day/1 to 3 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

| Test ID | Test Order Name | Order LOINC Value |
|---------|------------------------------|-------------------|
| C9FX | C9 Complement, Functional, S | 87727-4 |

| Result ID | Test Result Name | Result LOINC Value |
|-----------|------------------------------|--------------------|
| C9FX | C9 Complement, Functional, S | 87727-4 |