

Overview**Useful For**

Diagnosis of first component of complement (C1) deficiency

Investigation of a patient with an absent total complement (CH50) level

Method Name

Automated Liposome Lysis Assay

NY State Available

Yes

Specimen**Specimen Type**

Serum Red

Advisory Information

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 CH50.

Specimen Required

Patient Preparation: Fasting preferred

Supplies: Aliquot Tube, 5 mL (T465)

Collection Container/Tube: Red top

Submission Container/Tube: Plastic, 5 mL tube

Specimen Volume: 1 mL

Collection Instructions:

1. Immediately after specimen collection, place the tube on wet ice.
2. Centrifuge and separate serum from clot.
3. Immediately freeze specimen.

Specimen Minimum Volume

0.5 mL

Reject Due To

Gross hemolysis	OK
Gross lipemia	OK

Gross icterus	OK
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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 (or 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.

The first component of complement (C1) is composed of 3 subunits designated as C1q, C1r, and C1s. C1q recognizes and binds to immunoglobulin complexed to antigen and initiates the complement cascade. Congenital deficiencies of any of the early complement components (C1-C4) result in an inability to generate the peptides that are necessary to clear immune complexes and to attract neutrophils or generate lytic activity. 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.

Inherited deficiency of C1 is rare. C1 deficiency is associated with increased incidence of immune complex disease (systemic lupus erythematosus [SLE], polymyositis, glomerulonephritis, and Henoch-Schonlein purpura), and SLE is the most common manifestation of C1 deficiency. The SLE associated with C1 deficiency is similar to SLE without complement deficiency, but the age of onset is often prior to puberty.

Low C1 levels have also been reported in patients with abnormal immunoglobulin levels (Bruton and common variable hypogammaglobulinemia and severe combined immunodeficiency), and this is most likely due to increased catabolism.

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

34-63 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).

The measurement of C1q activity is an indicator of the amount of first component of complement (C1) present.

Absent C1q levels in the presence of normal C3 and C4 values are consistent with a C1 deficiency. Low C1q levels in the presence of low C4 but normal C3 may indicate the presence of an acquired inhibitor (autoantibody) to C1 esterase inhibitor.

Cautions

Absent (or low) C1q functional levels in the presence of normal C1q antigen levels should be replicated with a new serum specimen to confirm that C1q 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, Philadelphia, WB Saunders Company. 1984, pp 897-892
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**

C1q complement activity is measured by mixing patient serum with a C1q-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 C1q. The target liposomes are a commercial reagent (WAKO total complement CH50), and the assay is performed on aÂ Advia XPT.(Unpublished Mayo information; Yamamoto S, Kubotsu K, Masaaki K, et al: Automated homogeneous liposome-based assay system for total complement activity. *Clin Chem* 1995;41:586-590)

PDF Report

No

Day(s) and Time(s) Test Performed

Monday through Saturday; Continuous with a 3 p.m. cutoff

Analytic Time

Same day/1 day

Maximum Laboratory Time

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
C1QFX	C1Q Complement, Functional, S	87722-5

Result ID	Test Result Name	Result LOINC Value
C1QFX	C1Q Complement, Functional, S	87722-5