

Overview

Useful For

Diagnosis of C5 deficiency
Investigation of a patient with an absent total complement (CH50) level

Method Name

Nephelometry

NY State Available

Yes

Specimen

Specimen Type

Serum

Ordering Guidance

The total complement 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 but not required

Collection Container/Tube:

Preferred: Red top

Acceptable: Serum gel

Submission Container/Tube: Plastic vial

Specimen Volume: 1 mL

Collection Instructions: Centrifuge and separate serum from clot.

Reject Due To

Gross hemolysis OK

Gross lipemia Reject

Gross icterus OK

Specimen Minimum Volume

0.5 mL

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Serum	Frozen (preferred)	60 days	
	Refrigerated	28 days	

	Ambient	7 days	
--	---------	--------	--

Clinical & 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).

The absence of early components (C1-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 clear immune complexes or to 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.

More than 30 cases of C5 deficiency have been reported. Most of these patients have neisserial infections.

Reference Values

10.6-26.3 mg/dL

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 C5 levels in the presence of normal C3 and C4 values are consistent with a C5 deficiency. Absent C5 levels in the presence of low C3 and C4 values suggests complement consumption.

A small number of cases have been described in which the complement protein is present but is nonfunctional. These rare cases require a functional assay to detect the deficiency (C5FX / C5 Complement, Functional, Serum).

Cautions

Quantitation of specific proteins by nephelometric means may not be possible in lipemic sera due to the extreme light scattering properties of the specimen. Turbidity and particles in the specimen may result in extraneous light scattering signals, resulting in variable specimen analysis.

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 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

C5 complement antigen is measured by immunonephelometry. Antiserum to C5 is mixed with patient serum, the light scatter resulting from the antibody interaction with C5 is measured, and the signal is compared to standard concentrations of C5. (Unpublished Mayo information; Instruction manual: Siemens Nephelometer II I Version 3, Siemens, Inc., Newark, DE, 2008)

PDF Report

No

Specimen Retention Time

14 days

Performing Laboratory Location

Rochester

Fees & Codes

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 US Food and Drug Administration.

CPT Code Information

86160