MAYO CLINIC LABORATORIES

Test Definition: C6FX

C6 Complement, Functional, Serum

Overview

Useful For

Diagnosis of C6 deficiency

Investigation of a patient with an undetectable total complement level

Method Name

Automated Liposome Lysis Assay

NY State Available

Yes

Specimen

Specimen Type

Serum Red

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

Supplies: Sarstedt 5 mL Aliquot Tube (T914)

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.

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 & 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 (mannan-binding lectin) pathway. The classical pathway of the complement system is composed of a series of proteins that are activated in response to the presence of immune complexes. A single IgM molecule or 2 IgG molecules are sufficient to trigger activation of the recognition complex initiated by C1q. The activation process triggers a cascade that includes an amplification loop. The amplification loop is mediated by C3, with cleavage of a series of proteins, and results in 3 main end products: 1) anaphylatoxins that promote inflammation (C3a, C5a), 2) opsonization peptides that are chemotactic for neutrophils (C3b) and facilitate phagocytosis, and 3) the membrane attack complex (MAC), which promotes cell lysis.

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.

C6 deficiency is relatively rare, over 50 cases have been described. Most of these patients have systemic meningococcal infection and some have had invasive gonococcal infections. Normal levels of C6 antigen have been reported in patients with dysfunctional C6 lytic activity, hence the recommendation of functional testing.

Reference Values

32-57 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 C6 levels in the presence of normal C3 and C4 values are consistent with a C6 deficiency. Absent C6 levels in the presence of low C3 and C4 values suggests complement consumption.

Normal results indicate both normal C6 protein levels and normal functional activity.

Cautions

Absent (or low) C6 functional levels in the presence of normal C6 antigen levels should be replicated with a new serum specimen to confirm that C6 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 Mar-Apr; 1(2):131-135
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the age-related presence of circulating C1r-C1s complexes. Acta Paediatr Scand. 1987 Nov;76(6):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 Oct;29(5):489-492
4. Gaither TA, Frank MM: Complement. In: Henry JB, ed. Clinical Diagnosis and Management by Laboratory Methods. 17th ed. WB Saunders Company; 1984:879-892
5. O'Neil KM: Complement deficiency. Clin Rev Allergy Immunol. 2000 Oct;19:83-108
6. Frank MM: Complement deficiencies. Pediatr Clin North Am. 2000 Dec;47(6):1339-1354
7. Willrich MAV, Braun KMP, Moyer AM, Jeffrey DH, Frazer-Abel A. Complement testing in the clinical laboratory. Crit Rev Clin Lab Sci. 2021 Nov;58(7):447-478. doi: 10.1080/10408363.2021.1907297

Performance

Method Description

C6 complement activity is measured by mixing patient serum with a C6-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 C6. 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

1 to 3 days

Specimen Retention Time

14 days

Performing Laboratory Location

Rochester

Fees & 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 account representative. 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. It has not been cleared or approved by the US Food and Drug Administration.

CPT Code Information

86161

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
C6FX	C6 Complement, Functional, S	60459-5

Result ID	Test Result Name	Result LOINC® Value
C6FX	C6 Complement, Functional, S	60459-5