Test Definition: C2FXN

C2 Complement, Functional, S, NR

Overview

Useful For

Investigation of a patient with a low (absent) hemolytic complement (CH50)

Method Name

Automated Liposome Lysis Assay

NY State Available

Yes

Specimen

Specimen Type

Serum Red

Advisory Information

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.

To evaluate for complement C2, C3, and C4 in one orderable, consider ordering C2 / C2 Complement, Functional, with Reflex, Serum.

Specimen Required

Patient Preparation: Fasting preferred but not required

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	21 days	

Clinical and Interpretive

Clinical Information

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. This activation process results in the formation of the lytic membrane attack complex, as well as the generation of activation peptides that are chemotactic for neutrophils and that bind to immune complexes and complement receptors. The absence of early components (C1, C2, 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 generate lytic activity or to clear immune complexes.

Although rare, C2 deficiency is the most common inherited complement deficiency. Homozygous C2 deficiency has an estimated prevalence ranging from 1 in 10,000 to 1 in 40,000 (the prevalence of heterozygotes is 1 in 100 to 1 in 50). Half of the homozygous patients are clinically normal.

However, discoid lupus erythematosus or systemic lupus erythematosus (SLE) occurs in approximately one-third of patients with homozygous C2 deficiency. Patients with SLE and a C2 deficiency frequently have a normal anti-ds DNA titer. Clinically, many have lupus-like skin lesions and photosensitivity, but immunofluorescence studies may fail to demonstrate immunoglobulin or complement along the epidermal-dermal junction.

Other diseases reported to be associated with C2 deficiency include dermatomyositis, glomerulonephritis, vasculitis, atrophodema, cold urticaria, inflammatory bowel disease, and recurrent infections.

The laboratory findings that suggest C2 deficiency include a hemolytic complement (CH50) of nearly zero, with normal values for C3 and C4.

Reference Values

25-47 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 (or low) C2 levels in the presence of normal C3 and C4 values are consistent with a C2 deficiency.

Low C2 levels in the presence of low C3 and C4 values are consistent with a complement-consumptive process.

Low C2 and C4 values, in the presence of normal values for C3 is suggestive of C1 esterase inhibitor deficiency.

Cautions

As with all complement assays, proper sample handling is of utmost importance to ensure that the complement

system is not activated before clinical testing.

Clinical Reference

1. 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

2. Agnello V: Complement deficiency states. Medicine 1978;57:1-23

3. Buckley D, Barnes L: Childhood subacute cutaneous lupus erythematosus associated with homozygous complement 2 deficiency. Pediatr Dermatol 1995;12:327-330

Performance

Method Description

C2 complement activity is measured by mixing patient serum with a C2-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 C2. The target liposomes are a commercial reagent (WAKO total complement CH50), and the assay is performed on a Siemens Advia XPT.(Unpublished Mayo method)

PDF Report

No

Day(s) and Time(s) Test Performed

Monday through Friday; 3 p.m.

Analytic Time

Same day/1 day

Maximum Laboratory Time

2 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
C2FXN	C2 Complement, Functional, S, NR	93977-7

Result ID	Test Result Name	Result LOINC Value
C2FX	C2 Complement,Functional,S	93977-7
INT53	Interpretation	69048-7