

Overview

Useful For

Assessment of an undetectable total complement (CH50) level

Diagnosing congenital C1 (first component of complement) deficiency

Diagnosing acquired deficiency of C1 inhibitor

Method Name

Nephelometry

NY State Available

Yes

Specimen

Specimen Type

Serum

Specimen Required

Patient Preparation: Fasting

Container/Tube:

Preferred: Red top

Acceptable: Serum gel

Specimen Volume: 1 mL

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	Refrigerated (preferred)	28 days	
	Frozen	28 days	
	Ambient	21 days	

Clinical and Interpretive

Clinical Information

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, C2, C4) results in an inability to clear immune complexes. Inherited deficiency of C1 is rare.

Like the more common C2 deficiency, C1 deficiency is associated with increased incidence of immune complex disease (systemic lupus erythematosus, polymyositis, glomerulonephritis, and Henoch-Schonlein purpura). Low C1 levels have also been reported in patients with abnormal immunoglobulin levels (Bruton and common variable hypogammaglobulinemia and severe combined immunodeficiency). This is most likely due to increased catabolism.

The measurement of C1q is an indicator of the amount of C1 present.

Reference Values

12-22 mg/dL

Interpretation

An undetectable C1q in the presence of an absent total complement (CH50) and normal C2, C3, and C4 suggests a congenital C1 (first component of complement) deficiency.

A low C1q in combination with a low C1 inhibitor and low C4 suggests an acquired C1 inhibitor deficiency.

Cautions

This is a different assay than C1q binding, which is an assay for circulating immune complexes.

Clinical Reference

1. Frank MM: Complement in the pathophysiology of human disease. N Engl J Med 1987 June 11;316(24):1525-1530
2. Frank MM: Complement deficiencies. Pediatr Clin North Am 2000 December;47(6):1339-1354
3. Frigas E: Angioedema with acquired deficiency of the C1 inhibitor: a constellation of syndromes. Mayo Clin Proc 1989 October;64(10):1269-1275

Performance

Method Description

Nephelometry and anti-C1q antiserum are used to quantitate the C1q antigen level.(Instruction manual: Nephelometer II Operations. Siemens, Inc., Newark, DE, 5/2005)

PDF Report

No

Day(s) and Time(s) Test Performed

Monday through Saturday; Continuously until 3 p.m.

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

86160

LOINC® Information

Test ID	Test Order Name	Order LOINC Value
C1Q	Complement C1q, S	4478-4

Result ID	Test Result Name	Result LOINC Value
C1Q	Complement C1q, S	4478-4