

## Overview

### Useful For

Assessment of an undetectable total complement 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 for 12 hours

**Supplies:** Sarstedt Aliquot Tube, 5 mL (T914)

**Collection Container/Tube:**

**Preferred:** Serum gel

**Acceptable:** Red top

**Submission Container/Tube:** Plastic vial

**Specimen Volume:** 1 mL

**Collection Instructions:** Centrifuge and aliquot serum into a 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	Ambient	21 days	
	Refrigerated (preferred)	28 days	
	Frozen	28 days	

## Clinical & 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 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. Stegert M, Bock M, Trendelenburg M. Clinical presentation of human C1q deficiency: How much of a lupus? *Mol Immunol*. 2015;67(1):3-11
2. Tangye SG, Al-Herz W, Bousfiha A, et al. Human inborn errors of immunity: 2022 update on the classification from the International Union of Immunological Societies Expert Committee. *J Clin Immunol*. 2022;42(7):1473-1507
3. Beurskens FJ, van Schaarenburg RA, Trouw LA. C1q, antibodies and anti-C1q autoantibodies. *Mol Immunol*. 2015;68(1):6-13

## Performance

**Method Description**

Nephelometry and anti-C1q antiserum are used to quantitate the C1q antigen level. (Instruction manual: Siemens Nephelometer II. Siemens, Inc; Version 2.3, 2008; Addendum to the Instruction Manual 2.3, 08/2017)

**PDF Report**

No

**Day(s) Performed**

Monday through Friday

**Report Available**

2 to 5 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**

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