Test Definition: C1ES

C1 Esterase Inhibitor Antigen, Serum

Overview

Useful For

Diagnosis of hereditary angioedema

Monitoring levels of C1 esterase inhibitor in response to therapy

Method Name

Nephelometry

NY State Available

Yes

Specimen

Specimen Type

Serum

Specimen Required

Patient Preparations:

Fasting: 12 hours, preferred but not required.

Supplies: Sarstedt Aliquot Tube, 5 mL (T914)

Collection Container/Tube:

Preferred: Red top

Acceptable: Serum gel

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. Freeze specimen within 30 minutes.

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	Frozen (preferred)	28 days	
	Ambient	72 hours	
	Refrigerated	28 days	

Clinical & Interpretive

Clinical Information

C1 esterase inhibitor blocks the activation of C1 (first component of the complement cascade) to its active form. The deficiency of C1 esterase inhibitor results in the inappropriate activation of C1 and the subsequent release of an activation peptide from C2 with kinin-like activity. This kinin-like peptide enhances vascular permeability. C1 esterase inhibitor deficiency results in hereditary or acquired angioedema. This disease is an autosomal dominant inherited condition, in which exhaustion of the abnormally low levels of C1 esterase inhibitor results in C1 activation, breakdown of C2 and C4, and subsequent acute edema of subcutaneous tissue, the gastrointestinal tract, or the upper respiratory tract. The disease responds to attenuated androgens.

Because 15% of C1 inhibitor deficiencies have nonfunctional protein, some patients will have abnormal functional results (C1INF / C1 Esterase Inhibitor, Functional, Serum) in the presence of normal (or elevated) antigen levels.

Reference Values

19-37 mg/dL

Interpretation

Abnormally low results are consistent with a heterozygous C1 esterase inhibitor deficiency and hereditary angioedema.

Fifteen percent of hereditary angioedema patients have a normal or elevated level but nonfunctional C1 esterase inhibitor protein. Detection of these patients requires a functional measurement of C1 esterase inhibitor; C1INF / C1 Esterase Inhibitor, Functional, Serum.

Measurement of C1q antigen levels (C1Q / Complement C1q, Serum) is key to the differential diagnoses of acquired or hereditary angioedema. Those patients with the hereditary form of the disease will have normal levels of C1q, while those with the acquired form of the disease will have low levels.

Studies in children show that adult levels of C1 inhibitor are reached by 6 months of age.

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. Willrich MAV, Braun KMP, Moyer AM, Jeffrey DH, Frazer-Abel A. Complement testing in the clinical laboratory. Crit Rev Clin Lab Sci. 2021;58(7):447-478. doi:10.1080/10408363.2021.19072972

2. Drouet C, Lopez-Lera A, Ghannam A, et al. SERPING1 variants and C1-INH biological function: A close relationship with C1-INH-HAE. Front Allergy. 2022;3:835503. doi:10.3389/falgy.2022.835503
3. 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. doi:10.1007/s10875-022-01289-3
4. Brodszki N, Frazer-Abel A, Grumach AS, et al. European Society for Immunodeficiencies (ESID) and European Reference Network on Rare Primary Immunodeficiency, Autoinflammatory and Autoimmune Diseases (ERN RITA) Complement Guideline: Deficiencies, Diagnosis, and Management. J Clin Immunol. 2020;40(4):576-591. doi:10.1007/s10875-020-00754-1
5. Patel G, Pongracic JA. Hereditary and acquired angioedema. Allergy Asthma Proc. 2019;40(6):441-445. doi:10.2500/aap.2019.40.4267
6. Longhurst HJ, Tarzi MD, Ashworth F, et al. C1 inhibitor deficiency: 2014 United Kingdom consensus document [published correction appears in Clin Exp Immunol. 2015;182(3):346]. Clin Exp Immunol. 2015;180(3):475-483. doi:10.1111/cei.12584

Performance

Method Description

In this Siemens Nephelometer II method, the light scattered onto the antigen-antibody complexes is measured. The intensity of the measured scattered light is proportional to the amount of antigen-antibody complexes in the sample under certain conditions. If the antibody volume is kept constant, the signal behaves proportionally to the antigen volume.

A reference curve is generated by a standard with a known antigen content on which the scattered light signals of the samples can be evaluated and calculated as an antigen concentration. Antigen-antibody complexes are formed when a sample containing antigen and the corresponding antiserum are put into a cuvette. A light beam is generated with a light-emitting diode, which is transmitted through the cuvette. The light is scattered onto the immuno-complexes that are present. Antigen and antibody are mixed in the initial measurement, but no complex is formed yet. An antigen-antibody complex is formed in the final measurement.

The result is calculated by subtracting value of the final measurement from the initial measurement. The distribution of intensity of the scattered light depends on the ratio of the particle size of the antigen-antibody complexes to the radiated wavelength.(Instruction manual: Siemens Nephelometer II Operations. Siemens, Inc; Version 2.4, 07/2019)

PDF Report

No

Day(s) Performed

Tuesday through Friday

Report Available

2 to 5 days

Specimen Retention Time

14 days

Performing Laboratory Location

Mayo Clinic Laboratories - Rochester Superior Drive

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 has been cleared, approved, or is exempt by the US Food and Drug Administration and is used per manufacturer's instructions. Performance characteristics were verified by Mayo Clinic in a manner consistent with CLIA requirements.

CPT Code Information

83883

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
C1ES	C1 Esterase Inhibitor Antigen, S	4477-6

Result ID	Test Result Name	Result LOINC® Value
C1ES	C1 Esterase Inhibitor Antigen, S	4477-6