Test Definition: C1ES

C1 Esterase Inhibitor Antigen, S

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 preferred but not required.

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 separate serum from clot.

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

Clinical and 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 (FC1EQ / C1 Esterase Inhibitor, Functional Assay, 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; FC1EQ / C1 Esterase Inhibitor, Functional Assay, 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. Frank MM: Complement deficiencies. Pediatr Clin North Am 2000;47(6):1339-1354

2. Gelfand JA, Boss GR, Conley CL, et al: Acquired C1 esterase inhibitor deficiency and angioedema: a review. Medicine 1979;58(4):321-328

3. Rosen FS, Alper CA, Pensky J, et al: Genetically determined heterogeneity of the C1 esterase inhibitor in patients with hereditary angioneurotic edema. J Clin Invest 1971;50(10):2143-2149

4. Frigas E: Angioedema with acquired deficiency of the C1 inhibitor: a constellation of syndromes. Mayo Clin Proc 1989;64:1269-1275

5. Soldin SJ, Hicks JM, Bailey J, et al: Pediatric reference ranges for estradiol and C1 esterase inhibitor. Clin Chem 1998;44(6s):A17

Performance

Method Description

C1 esterase inhibitor complement antigen (C1ES) is measured by immunonephelometry. Antiserum to C1ES is mixed with patient serum, the light scatter resulting from the antibody interaction with C1ES is measured, and the signal is compared to standard concentrations of C1ES.(Instruction Manual: Siemens 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

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 has been cleared or approved by the U.S. 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