

C1 Esterase Inhibitor

Synonyms

C1INH

Clinical Indication

Diagnosis of hereditary angioedema (HAE). C1 esterase inhibitor deficiency is the commonest of the inherited complement deficiencies (approx. 1:50,000) and is autosomal dominant. Several members of a family are usually affected.

The much more common condition of urticaria/angioedema tends to cause repeated episodes of intensely itchy swellings which last a shorter period of time, often no more than hours. No obvious trigger is detected in the majority of such cases and laboratory investigation is usually not helpful.

Usually C1 esterase inhibitor analysis will only be undertaken if C4 levels are low with normal C3 levels (Basildon Hospital specific criteria).

The request form should show:

- Site, duration, frequency and clinical nature of attacks
- Details of relatives similarly affected

Part of Profile / See Also

C1 esterase inhibitor quantitative and functional activity, C3 and C4 levels.

Request Form

Combined Pathology manual Blood form or ICE request

Availability / Frequency of Analysis

Referred Test: Analysed by Immunology, Royal London Hospital ([8285](#)) if specific criteria met.

Turnaround Time

2 weeks

Patient Preparation

Sample Requirements

URGENT - Samples must be transported to the laboratory immediately and remain at the ESL.

Specimen Type

Serum

Volume

2 ml

Container



Yellow top (SST) tube



Or Paediatric orange top (lithium-heparin)



Or Paediatric green top (lithium-heparin)

Reference Range & Units

Quantitation: 150 – 350 mg/L. Functional activity: 70-130%.

Please note that quantitation may not be undertaken if functional activity is normal.

Interferences

Interpretation & Clinical

Decision Value (if applicable)

C1 esterase inhibitor quantitation and functional activity should be interpreted in conjunction for diagnosis of HAE/AAE. Reduced C1 inhibitor function may be caused by delay in sample transit and may also be caused by the use of exogenous oestrogens, including oral contraceptive agents. C1

inhibitor is an acute-phase protein and can be increased in inflammatory processes as well as the administration of exogenous androgen and related steroids. Interpretation should consider family and clinical/past medical history

References

<https://www.eselpathology.nhs.uk/test-a-z/?testid=575&letter=C>

<https://www.uptodate.com/contents/an-overview-of-angioedema-clinical-features-diagnosis-and-management>

Test code

C1ES

Lab Handling

Sample must be bled and processed at the ESL. Centrifuge SST sample and aliquot at least 0.5ml into 2 separate tubes and freeze at -20C. Ensure sample type is written on all aliquots. Sample MUST be in the freezer within 30 minutes otherwise a rebleed may be required. Sample is sent frozen with the Global courier to the Royal London Hospital.



8285
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ISO 15189:2012