

C1- Esterase Inhibitor

General:

C1-esterase inhibitor is an initial complement cascade regulator glycoprotein with a molecular weight of 105 kD, which is formed predominantly in hepatocytes. It is a mediator in coagulation, fibrinolysis and the kinin system by spontaneous activation of factor XIIIa (Hagemann factor). Through initial activation of the complement system, a permanent decrease is observed. Hereditary deficiency leads to episodic disturbances of tissue permeability of the skin, gastrointestinal tract and, rarely, respiratory tract.

Main symptoms are circumscribed edemas and convulsive, recurrent abdominal pain. Deficiency can lead to angioneurotic edema with acute swellings of lips and eyelids, gastrointestinal colics and particularly dangerous swellings of larynx and pharynx. An edema of glottis can lead to asphyxia.

It is important to distinguish this type of autosomal dominant hereditary angioedema from other histamine-mediated angioedemas. The examination of C1-esterase inhibitors reflects their functional activity.

The following tests are available:

- **C1-Esterase Inhibitor Activity in citrate plasma**

Indication: Suspicion of angioneurotic edema, Quincke's disease

Material: 3 ml citrate plasma, **frozen**

TAT: 7-10 days*

Method: photometric

Units: %

Ref.- range: 70 – 130

- **C1-Esterase Inhibitor Protein in serum**

General:

The immunological examination of the inhibitor protein reflects its concentration and recognizes defect deficiencies. 85% of angioneurotic edema cases show decreased C1-esterase inhibitor protein concentration, 15% show normal C1-esterase inhibitor protein concentration with low C1-esterase inhibitor activity.

Material: 1 ml serum

TAT: 7-10 days*

Method: TURB

Units: mg/dl

Ref.- range: 21-38

- **C1-Esterase Inhibitor antibodies**

Material: 1 ml serum

TAT: 7-10 days*

Method: blot

Ref.- range: see report

For complete list of laboratory test offered at Freiburg Medical Laboratory, please visit <http://www.fml-dubai.com/parameter-listings/>