ALPHA DIAGNOSTIC INTERNATIONAL

Product Specification Sheet

C1 Esterase Inhibitor, Human Plasma

Cat # C1E15-N | C1 Esterase Inhibitor, Human Plasma | **SIZE**: 1 mg

C1-inhibitor (C1-inh, C1 esterase inhibitor) is a serine protease inhibitor (serpin) protein, the main function of which is the inhibition of the complement system to prevent spontaneous activation. C1-inh is an acute phase protein, it circulates in blood at levels of around 0.25 g/L, which levels rise ~2-fold during inflammation. C1-inh irreversibly binds to and inactivates C1r and C1s proteinases in the C1 complex of classical pathway of complement. MASP-1 and MASP-2 proteinases in MBL complexes of the lectin pathway are also inactivated. This way, C1-inh prevents the proteolytic cleavage of later complement components C4 and C2 by C1 and MBL. Although named after its complement inhibitory activity, C1-inh also inhibits proteinases of the fibrinolytic, clotting, and kinin pathways. Most notably, C1-inh is the most important physiological inhibitor of plasma kallikrein, fXIa and fXIIa.

C1-inh is the largest member among the serpin class of proteins. Remarkably, C1-inh has a 2-domain structure, unlike most family members. The C-terminal serpin domain is similar to other serpins, and this part of C1-inh provides the inhibitory activity of C1-inh. The N-terminal domain (also some times referred to as the N-terminal tail) is not essential for C1-inh to inhibit proteinases. This domain has no similarity to other proteins. C1-inh is highly glycosylated glycosylated, bearing both N- and O-glycans. N-terminal domain is especially heavily glycosylated. Deficiency of this protein is associated with hereditary angioedema ("hereditary angioneurotic edema"), or swelling due to leakage of fluid from blood vessels into connective tissue.

With 35% carbohydrate by weight, C1 inhibitor is the most highly glycosylated serum protein. In the region sequenced, C1NH showed about 22% identity with antithrombin III, 26% with alpha-1-antitrypsin and alpha-1-antichymotrypsin, and 18% with angiotensinogen. Human C1 inhibitor is a single chain moiety, which has 478 amino acid residues, accounts for little more than half the molecular weight of the circulating protein, which is 104 kD.

COMPLEMENT COMPONENT 1 INHIBITOR; C1NH
Alternative titles; symbols: C1I; C1INH, SERPING1
Gene map locus 11q11-q13.1

Sources of antigen and antibodies

Human C1 esterase inhibitor (**cat # C1E15-N**) is purified (>95%, ~100 kda) from human plasma that has been shown by certified tests to be negative for HbsAg and HIV and HCV. However, all precautions must be taken to avoid contamination.

Form & Storage

It is supplied in 20 mM potassium phosphate, pH 7.0, with 250 mM NaCl in liquid or powder form. Store at –70oC. Reconstitute in PBS at no <100 ug/100 ul. Lot specific concentrations are provided on the vial.

Stability: 6-12 months at –70oC or below.

General References: (1) Bock SC (1986) Biochem. 25, 4292-4301; Davis AE (2008) Ann. Allergy Asthma Immunol. 100 (1 Suppl 2): S7–12; Caliezi C (2000) Pharmacol. Rev. 52 (1): 91–112; Bernsterin JA (2008) Ann. Allergy Asthma Immunol. 100 (1 Suppl 2): S41–6.

This product is for In vitro research use only.

All human derived material has been tested negative for HIV, HCV, and HbSAg. Nevertheless, all precautions should be taken and samples be treated as potentially hazardous.

C1E15-N 100330A