Active Native Human C1 Esterase Inhibitor (C1-INH)

Cat. No. SERPING1-97H **Lot. No.** (See product label)

SPECIFICATION

Species	Human
Source	Human Serum
Description	The protease inhibitor C1-INH prevents the spontaneous activation of complement and limits consumption of C2 and C4 by rapidly inactivating C1r, C1s and MASP2. It is the only plasma serine protease inhibitor (Serpin) capable of interacting with and inhibiting activated C1. C1-INH interacts with the catalytic sites of both C1r and C1s. The interaction with activated C1r and C1s is covalent resulting in complexes which are stable to SDS. The binding of C1-INH to activated C1 releases both C1r and C1s from the complex leaving C1q bound to the immune complex. The released complexes contain four molecules: C1-INH-C1r-C1s-C1-INH. The reaction of C1 esterase inhibitor with activated C1 is very fast with the estimated half-life of C1r and C1s being approximately 15 seconds in serum. In fact, at serum concentrations of C1-INH little or no additional C4 or C2 activation occurs 3 min after immune complexes are added because all the C1r and C1s molecules have been inactivated and removed from the C1q which remains bound to the immune complex (Ross, G.D. (1986); Morley, B.J. and Walport, M.J. (2000); Rother, K., et al. (1998); Ziccardi, R.J. (1982a and 1982b); Morgan, B.P. (1990)). C1-INH is thought to bind to and stabilize unactivated C1r and C1s in the C1 complex thus retarding their spontaneous activation (Ziccardi, RJ. (1982b)). C1-INH plays an important role in suppression of inflammation and control of vascular permeability. Through its ability to inhibit complement proteases (C1r, C1s and MASP2) and to express a variety of other biological functions (Davis III, A.E. et al. (2008)) C1-INH is able to regulate

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	inflammatory reactions in sepsis, endotoxic shock, ischemia-reperfusion injury, transplantation, and other bacterial and parasitic infections. Through its ability to control contact system activation it inhibits bradykinin generation and this controls vascular permeability. This function is most apparent in patients with a physical or functional deficiency of C1-INH. Hereditary angioedema (HAE) patients suffer from enhanced blood vessel permeability and tissue swelling or edema. Most patients are heterozygous and have 15% to 30 % of the normal level of functional C1-INH in blood.
Form	Frozen liquid
Bio-activity	>90 % active protein
Molecular Mass	110,000 Da (single chain)
Purity	>95 % by SDS PAGE
Characteristic	C1-INH has an apparent molecular weight of 110,000 Da. It is a single chain protein that is highly glycosylated with approximately 30 to 40% carbohydrate. It is synthesized as a 500 amino acid protein. Removal if the signal peptide results in a plasma protein that contains 478 amino acids. The calculated molecular weight based on amino acids is 53,000 g/mole, however due to extensive glycosylation its mass is closer to 75,000. It runs abnormally on many gel systems giving apparent molecular weights from 90,000 to 115,000 MW. The glycosylation sites include six N-linked and six or seven O-linked sites. C1-INH is an extremely acidic protein with a pI of less than 3.0.
Applications	The genetic disorder HAE is caused by a partial deficiency of C1-INH. Replacement therapy with a C1-INH concentrate produced by a number of drug companies has been approved for use in both Europe and the USA. These concentrates are

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	administered intravenously and increase blood levels of C1-INH 2- to 3-fold. A typical treatment includes administration of 1000 Units (approximately 200 mg) and the blood level may remain elevated for 2-4 days.
Usage	For research use only. Not for human or drug use.
Storage	-70°C or below. Avoid freeze/thaw.
Concentration	1.0 mg/mL (see Certificate of Analysis for actual concentration)
Storage Buffer	10 mM sodium phosphate, 145 mM NaCl, pH 7.3
Preservative	None, 0.22 μm filtered.
Warning	Use normal precautions for handling human blood products.

GENE INFORMATION

Gene Name	SERPING1 serpin peptidase inhibitor, clade G (C1 inhibitor), member 1 [Homo sapiens]
Official Symbol	SERPING1
Synonyms	SERPING1; serpin peptidase inhibitor, clade G (C1 inhibitor), member 1; C1NH, serine (or cysteine) proteinase inhibitor, clade G (C1 inhibitor), member 1, (angioedema, hereditary); plasma protease C1 inhibitor; angioedema; hereditary; C1 INH; C1IN; HAE1;
Gene ID	710
mRNA Refseq	NM_000062

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Protein Refseq	NP_000053
MIM	606860
UniProt ID	P05155
Chromosome Location	11q12-q13.1
Pathway	Complement and Coagulation Cascades, organism-specific biosystem; Complement and coagulation cascades, organism-specific biosystem; Complement and coagulation cascades, conserved biosystem; Formation of Fibrin Clot (Clotting Cascade), organism-specific biosystem; Hemostasis, organism-specific biosystem; Intrinsic Pathway, organism-specific biosystem; Pertussis, organism-specific biosystem;
Function	complement binding; peptidase inhibitor activity; protein binding; serine-type endopeptidase inhibitor activity;

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