

Native Human C1 Esterase inhibitor

Cat. No. SERPING1-73H **Lot. No.** (See product label)

SPECIFICATION

Product Overview

Species Human

Source Human Plasma

Description

A single chain glycoprotein which inhibits C1, C1r, C1s, plasma kallikrein, Factors XIa, XIIa, and plasmin. Present in plasma at 16-33 mg per 100 ml. C1 esterase inhibitor deficiency is a rare condition resulting in facial swelling and abdominal cramping. Usually the condition is hereditary, though it may also occur when the C1EI is non-functional. The C1 esterase inhibitor protein is a normal constituent of serum which functions as a serine proteinase inhibitor of the serpin family. The C1 esterase inhibitor inhibits the complement proteases C1r and C1s, as well as the proteases kallikrein, factor XIa, XIIa and plasmin of the blood clotting system. The concentration of C1 esterase inhibitor protein is reduced to 10-30% of normal in patients with angioedema secondary to C1 esterase inhibitor deficiency (85% of patients with Hereditary Angioedema (HAE)); in 15% of patients with HAE, the concentrations of the inhibitor protein is normal but function is markedly reduced.

Form Frozen

Molecular Mass 100 kDa

Purity Purity by SDS-PAGE: $\geq 95\%$

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Usage	For Research Use Only! Not For Use in Humans.
Notes	Centrifuge the vial prior to opening
Storage	-80°C
Storage Buffer	Frozen in 20 mM potassium phosphate, pH 7.0, with 250 mM KCl.
Shipping	Dry Ice

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
Protein Refseq	NP_000053
MIM	606860
UniProt ID	P05155

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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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