Product name:	C1INH Rabbit Polyclonal Antibody
Cat number:	ABN07717
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human SERPING1. AA range:342-391
Reactivity:	Human,Rat,Mouse
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:20000-1:40000
Molecular Weight:	55kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

This gene encodes a highly glycosylated plasma protein involved in the regulation of the complement cascade. Its protein inhibits activated C1r and C1s of the first complement component and thus regulates complement activation. Deficiency of this protein is associated with hereditary angioneurotic oedema (HANE). Alternative splicing results in multiple transcript variants encoding the same isoform. [provided by RefSeq, Jul 2008],disease:Defects in SERPING1 are the cause of hereditary angioedema (HAE) [MIM:106100]; also called hereditary angioneurotic edema (HANE). HAE is an autosomal dominant disorder characterized by episodic local subcutaneous edema and submucosal edema involving the upper respiratory and gastrointestinal tracts. HAE due to C1 esterase inhibitor deficiency is comprised of two clinically indistinguishable forms. In HAE type 1, representing 85% of patients, serum levels of C1 esterase inhibitor are less than 35% of normal. In HAE type 2, the levels are normal or elevated, but the protein is non-functional.,function:Activation of the C1 complex is under control of the C1-inhibitor. It forms a proteolytically inactive stoichiometric complex with the C1r or C1s proteases. May play a potentially crucial role in regulating important physiological pathways including complement activation, blood coagulation, fibrinolysis and the generation of kinins. Very efficient inhibitor of FXIIa. Inhibits chymotrypsin and kallikrein.,online information:C1-inhibitor entry,online information:SERPING1 mutation db,polymorphism:Chymotrypsin uses Ala-465 as its reactive site in normal plasma protease C1 inhibitor, and His-466 as its reactive site in the variant His-466.,PTM:Can be proteolytically cleaved by E.coli stcE.,PTM:Highly glycosylated (49%).,similarity:Belongs to the serpin family.,subunit:Binds to E.coli stcE which allows localization of SERPING1 to cell membranes thus protecting the bacteria against complement-mediated lysis. Interacts with MASP1.,