Active Native Human C1r Enzyme

Cat. No. C1R-96H **Lot. No.** (See product label)

SPECIFICATION

Product Overview	
Species	Human
Source	Human Serum
Description	C1r enzyme is the activated form of C1r proenzyme. C1r is a subunit of the C1 complex which is the first complement component in the classical pathway of complement. C1r proenzyme is an inactive zymogen until C1 is activated. C1r is activated when C1 binds to and is activated by antibodies bound to antigens (immune complexes) yielding C1r enzyme, the first protease that initiates the cascade. C1 is a non-covalent calcium-dependent complex of one C1q, two C1r and two C1s molecules. Each C1q binds through two or more of its six arms to the Fc domains of IgG or IgM. The binding of multiple arms to immune complexes causes the two C1r proteins in the complex (protease zymogens) to activate producing two proteases that cleave and activate the two C1s protease zymogens in the complex. The activation of C1r results from cleavage of C1r into two fragments of 57,000 and 35,000 daltons. Activation of the bound C1s molecule is the only known function of C1r enzyme. Activated C1s cleaves complement component C4 releasing C4a and initiating covalent attachment of C4b to the activating surface. Activated C1s also cleaves C2 and the larger fragment of C2 binds to the surface-attached C4b forming C4b,C2a the C3/C5 convertase of the classical pathway.
Form	Frozen liquid

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Bio-activity	>90 % C1r will bind with excess C1-INH
Molecular Mass	92,000 Da (2 chains)
Purity	>90 % by SDS PAGE (Note: C1r enzyme is 92,000 unreduced, but upon reduction runs as 57,000 and 35,000 chains on SDS PAGE)
Characteristic	C1r enzyme is a 92,000 dalton, two chain, trypsin-like enzyme. C1r is present in plasma at 31 ug/mL. C1r proenzyme is an unstable zymogen and it spontaneously activates (Dodds, A.W. and Sim, R.B. editors (1997); Morikis, D. and Lambris, J.D. editors. (2005)) by cleaving a peptide bond in C1r producing a 57,000 dalton heavy chain and a 35,000 dalton light chain. This is the form sold as C1r enzyme. This self-activation occurs rapidly in the C1 complex upon binding to an immune complex and it occurs slowly with pure C1r. Two C1r form a C1r-C1r complex in the presence of calcium which in turn forms a stable complex with two C1s molecules in the presence of calcium. This tetramer can exist in solution, but in the presence of C1q it binds to C1q forming the C1 complex, which is stable in the presence of calcium. C1r self-activation is controlled in part by a weak association with C1esterase inhibitor (C1-INH) when it is in the C1 complex and similar stabilization occurs with purified C1r (Ziccardi, R.J. (1982)). C1r enzyme, however, is irreversibly inactivated by binding to C1-INH.
Applications	C1r enzyme can be used to activate C1s proenzyme. In the presence of calcium it is still capable of binding another C1r enzyme and two C1s molecules and this complex is capable of binding to C1q. The activated C1r enzyme will rapidly activate the two C1s proenzymes to form C1s enzymes and the resulting C1q-C1r2-C1s2 complex is a fully active C1 molecule which will activate C4 and C2 in the fluid phase or on a cell bearing antibodies, such as EA (Dodds, A.W. and Sim, R.B. editors (1997); Morgan, B.P. ed. (2000)). EA are sheep erythrocytes with rabbit IgM anti-sheep erythrocytes antibodies bound to their surface(Morgan, B.P. ed. (2000)).

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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 Imidazole, 140 mM NaCl, 2 mM EDTA, pH 7.4
Preservative	None, 0.22 μm filtered.
Warning	Use normal precautions for handling human blood products.

GENE INFORMATION

Gene Name	C1R complement component 1, r subcomponent [Homo sapiens]
Official Symbol	C1R
Synonyms	C1R; complement component 1, r subcomponent; complement C1r subcomponent;
Gene ID	715
mRNA Refseq	NM_001733
Protein Refseq	NP_001724
MIM	613785
UniProt ID	P00736
Chromosome Location	12p13.31

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Pathway	Classical antibody-mediated complement activation, organism-specific biosystem; Complement Activation, Classical Pathway, organism-specific biosystem; Complement and Coagulation Cascades, organism-specific biosystem; Complement and coagulation cascades, organism-specific biosystem; Complement and coagulation cascades, conserved biosystem; Complement cascade, organism-specific biosystem; Creation of C4 and C2 activators, organism-specific biosystem;
Function	calcium ion binding; peptidase activity; serine-type endopeptidase activity; serine-type peptidase activity;

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