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Factor I light chain Rabbit pAb

Catalog # AP99648

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	P05156
Other Accession	P05156
Predicted	Rat, Human, Mouse, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	65750
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Factor I Complement factor I light chain
Epitope Specificity	281-380/582
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the peptidase S1 family. Contains 1 Kazal-like domain. Contains 2 LDL-receptor class A domains. Contains 1 peptidase S1 domain. Contains 1 SRCR domain.
DISEASE	Defects in CFI are a cause of susceptibility to hemolytic uremic syndrome atypical type 3 (AHUS3) [MIM:612923]. An atypical form of hemolytic uremic syndrome. It is a complex genetic disease characterized by microangiopathic hemolytic anemia, thrombocytopenia, renal failure and absence of episodes of enterocolitis and diarrhea. In contrast to typical hemolytic uremic syndrome, atypical forms have a poorer prognosis, with higher death rates and frequent progression to end-stage renal disease. Note=Susceptibility to the development of atypical hemolytic uremic syndrome can be conferred by mutations in various components of or regulatory factors in the complement cascade system. Other genes may play a role in modifying the phenotype. Defects in CFI are the cause of complement factor I deficiency (CFI deficiency) [MIM:610984]. CFI deficiency is an autosomal recessive condition associated with a propensity to pyogenic infections.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The complement pathway is an important host defense system that contributes to both innate and acquired immunity. There are three pathways of complement activation: the classical pathway, lectin pathway and alternative pathway. Complement protein Factor I is a key serine protease that modulates the complement cascade by regulating the levels of C3 convertases. It circulates in plasma as a heavily N-glycosylated heterodimer made up of two disulfide linked chains, each carrying three N-linked oligosaccharide chains that may have both structural and functional roles in the interactions with the natural substrate and the cofactor during catalysis. Factor I is a serine protease with a high degree of specificity for C3b and C4b. It requires protein cofactors for cleavage of these complement proteins; Factor H, CR1 or MCP are required for C3b cleavage, and C4bp or CR1 are required for C4b cleavage.

Additional Information

Gene ID	3426
Other Names	AHUS3; ARMD13; C3BINA; C3b-INA; FI; IF; KAF; CFAI_HUMAN; CFI; C3B/C4B inactivator; 3.4.21.45; complement factor I; I factor (complement); Konglutinogen-activating factor; C3b-inactivator
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	CFI
Synonyms	IF
Function	Trypsin-like serine protease that plays an essential role in regulating the immune response by controlling all complement pathways. Inhibits these pathways by cleaving three peptide bonds in the alpha- chain of C3b and two bonds in the alpha-chain of C4b thereby inactivating these proteins (PubMed:17320177, PubMed:7360115). Essential cofactors for these reactions include factor H and C4BP in the fluid phase and membrane cofactor protein/CD46 and CR1 on cell surfaces (PubMed:12055245, PubMed:2141838, PubMed:9605165). The presence of these cofactors on healthy cells allows degradation of deposited C3b by CFI in order to prevent undesired complement activation, while in apoptotic cells or microbes, the absence of such cofactors leads to C3b-mediated complement activation and subsequent opsonization (PubMed:28671664).
Cellular Location	Secreted, extracellular space. Secreted
Tissue Location	Expressed in the liver by hepatocytes (PubMed:6327681). Also present in other cells such as monocytes, fibroblasts or keratinocytes (PubMed:17320177, PubMed:6444659)

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