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Rabbit Anti-Factor I light chain antibody

SL13131R

Product Name:	Factor I light chain
Chinese Name:	补体因子I轻链抗体
Alias:	AHUS3; ARMD13; C3b INA; C3b inactivator; C3B/C4B inactivator; C3BINA; CFAI_HUMAN; Cfi; Complement component I; Complement control protein factor I; Complement factor I; Complement factor I heavy chain; Complement factor I light chain; F1; factor I; FactorI; FI; I factor; IF; KAF; Konglutinogen activating factor; Light chain of factor I; OTTHUMP00000219728; OTTHUMP00000221928
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Cow,Horse,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	22kDa
Cellular localization:	Extracellular matrixSecretory protein
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Factor I Complement factor I light chain:281-380/582
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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. **Function:** Responsible for cleaving the alpha-chains of C4b and C3b in the presence of the cofactors C4-binding protein and factor H respectively. **Subcellular Location:** Secreted; extracellular space. **Tissue Specificity:** Plasma. **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. **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. **SWISS:** P05156 **Gene ID:** 3426

	Database links: Entrez Gene: 3426Human Omim: 217030Human SwissProt: P05156Human Unigene: 312485Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.