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Factor I light chain Rabbit pAb

Catalog # AP99072

Product Information

Application	IHC-P, IHC-F, IF
Reactivity	Rat
Predicted	Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	22 KDa
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from mouse Factor I light chain
Epitope Specificity	451-550/582
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

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	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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.
Name	Factor I light chain Rabbit pAb
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.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.