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Factor B Rabbit pAb

Catalog # AP98959

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P00751
Other Accession	P00751
Reactivity	Rat, Mouse
Predicted	Human
Host	Rabbit
Clonality	Polyclonal
Calculated MW	85533
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Complement factor-B Bb fragment
Epitope Specificity	301-400/764
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 peptidase S1 domain. Contains 3 Sushi (CCP/SCR) domains. Contains 1 VWFA domain.
SUBUNIT	Monomer.
DISEASE	Defects in CFB are a cause of susceptibility to hemolytic uremic syndrome atypical type 4 (AHUS4) [MIM:612924]. 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.
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 component proteins, C3, C4 and C5, are potent anaphylatoxins that are released during complement activation. Binding of these proteins to their respective G protein-coupled receptors, C3aR, C1R and C5aR, induces proinflammatory events, such as cellular degranulation, smooth muscle contraction, arachidonic acid metabolism, cytokine release, leukocyte activation and cellular chemotaxis. Complement Factor B, also designated Properdin Factor B or PBF2, is part of the alternate pathway of the complement system and is cleaved by Factor D into two fragments: Ba and Bb. Bb combines with complement Factor 3b to produce the C3 or C5 convertase and plays a role in the differentiation and proliferation of preactivated B lymphocytes, lysis of erythrocytes, stimulation of lymphocyte blastogenesis and rapid spreading of peripheral blood monocytes. Ba is important in inhibiting the proliferation of preactivated B lymphocytes. Adipsin, also designated complement Factor D, is a serine protease that cleaves complement Factor B and may be involved in obesity. Factor H

controls the function of the alternative complement pathway. FHR-1 (complement Factor H related protein 1) may play a role in lipid metabolism.

Additional Information

Gene ID	629
Other Names	AHUS4; ARMD14; BF; BFD; CFAB; CFBD; FB; FBI12; GBG; H2-Bf; PBF2; C2; Da1-24; CFAB_HUMAN; CFB; C3/C5 convertase; Glycine-rich beta glycoprotein (GBG); Properdin factor B; 3.4.21.47; CFAB_MOUSE; complement factor B; B-factor, properdin; properdin B
Dilution	WB=1:500-2000,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.

Protein Information

Name	CFB (HGNC:1037)
Synonyms	BF, BFD
Function	Precursor of the catalytic component of the C3 and C5 convertase complexes of the alternative pathway of the complement system, a cascade of proteins that leads to phagocytosis and breakdown of pathogens and signaling that strengthens the adaptive immune system (PubMed:3638964, PubMed:624565, PubMed:6554279, PubMed:6919543, PubMed:9748277). The alternative complement pathway acts as an amplification loop that enhances other complement pathways (classical, lectin and GZMK) by promoting formation of additional C3 and C5 convertases (PubMed:3638964, PubMed:624565, PubMed:6554279, PubMed:6919543, PubMed:9748277). CFB is cleaved and activated by CFD to generate Ba and Bb chains; Bb chain constituting the catalytic component of the C3 and C5 convertases (PubMed:6769474, PubMed:9748277).
Cellular Location	Secreted.

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