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CFHL5 Rabbit pAb

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Catalog # AP55328

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q9BXR6
Predicted	Human
Host	Rabbit
Clonality	Polyclonal
Calculated MW	64419
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CFHL5
Epitope Specificity	21-120/569
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Secreted.
DISEASE	Note=Defects in CFHR5 have been found in patients with atypical hemolytic uremic syndrome and may contribute to the disease. Atypical hemolytic uremic syndrome 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. 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	This gene is a member of a small complement factor H (CFH) gene cluster on chromosome 1. Each member of this gene family contains multiple short consensus repeats (SCRs) typical of regulators of complement activation. The protein encoded by this gene has nine SCRs with the first two repeats having heparin binding properties, a region within repeats 5-7 having heparin binding and C reactive protein binding properties, and the C-terminal repeats being similar to a complement component 3 b (C3b) binding domain. This protein co-localizes with C3, binds C3b in a dose-dependent manner, and is recruited to tissues damaged by C-reactive protein. Allelic variations in this gene have been associated, but not causally linked, with two different forms of kidney disease: membranoproliferative glomerulonephritis type II (MPGNII) and hemolytic uraemic syndrome (HUS). [provided by RefSeq, Jan 2010]

Additional Information

Gene ID	81494
Other Names	Complement factor H-related protein 5, FHR-5, CFHR5, CFHL5, FHR5
Dilution	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	CFHR5
Synonyms	CFHL5, FHR5
Function	Involved in complement regulation. The dimerized forms have avidity for tissue-bound complement fragments and efficiently compete with the physiological complement inhibitor CFH.
Cellular Location	Secreted.
Tissue Location	Expressed by the liver and secreted in plasma.

Background

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