Complement FACTOR H (CFH) Antibody – Mouse Monoclonal

PRODUCT OVERVIEW

Product Name	COMPLEMENT FACTOR H (CFH), Monoclonal Antibody
Full Product Name	MOUSE ANTI HUMAN COMPLEMENT FACTOR H
Product Gene Name	anti-CFH antibody
Clonality	Monoclonal
Isotype	IgG1
Clone Number	10-10
Host	Mouse
Form/Format	Purified Purified IgG - liquid
Concentration	IgG concentration 1.0 mg/ml (lot specific)
Specificity	This item recognises human complement factor H which exists in 2 forms. The most common form, of 150 kDa, and the less common form of 43 kDa. Factor H is secreted by the liver into the blood serum. It is important in regulating the complement pathway, preventing unnecessary inflammation which can damage the host tissue. Complement factor H functions as a cofactor in the inactivation of C3b by factor I. It makes C3b susceptible to cleavage by factor I, resulting in iC3b. Factor H also inhibits the formation of the C3bBb complex (C3 convertase) and increases the rate of dissociation of both C3 convertase and the (C3b)NBB complex (C5 convertase). This prevents these components of the classical and of alternative complement pathways from forming a positive feedback loop. Mutations in factor H are associated with hemolytic-uremic syndrome, age-related macular degeneration, membranoproliferative glomerulonephritis (MPGN) type II and chronic hypocomplementemic nephropathy.
Preparation and Storage	Store at 4 degree C or at -20 degree C if preferred. Storage in frost-free freezers is not recommended. This product should be stored undiluted. Avoid repeated freezing and thawing as this may denature the antibody. Should this product contain a precipitate we recommend microcentrifugation before use. Shelf Life: 18 months from date of despatch.
Molecular Weight	51,034 Da
OMIM	126700
3D Structure	ModBase 3D Structure for P08603
Perservative Stabilisers	0.1% Sodium Azide (NaN3) Preparation||Purified IgG prepared by affinity chromatography on Protein A.
Immunogen	Purified human complement factor H from serum. Histology Positive Control Tissue||Kidney from post streptococcal glomerulonephritis patients. Buffer Solution||Borate buffered saline
Target Species	Human
Applications Tested/Suitable for anti-CFH antibody	Immunohistology Frozen, ELISA (EIA), Flow cytometry (FC/FACS), Western Blot (WB)

Related Product Information for anti-CFH antibody	MBS212919 recognises human complement factor H which exists in 2 forms. The most common form, of 150 kDa, and the less common form of 43 kDa. Factor H is secreted by the liver into the blood serum. It is important in regulating the complement pathway, preventing unnecessary inflammation which can damage the host tissue. Complement factor H functions as a cofactor in the inactivation of C3b by factor I. It makes C3b susceptible to cleavage by factor I, resulting in iC3b. Factor H also inhibits the formation of the C3bBb complex (C3 convertase) and increases the rate of dissociation of both C3 convertase and the (C3b)NBB complex (C5 convertase). This prevents these components of the classical and of alternative complement pathways from forming a positive feedback loop. Mutations in factor H are associated with hemolytic-uremic syndrome, age-related macular degeneration, membranoproliferative glomerulonephritis (MPGN) type II and chronic hypocomplementemic nephropathy.
Other Notes	Small volumes of anti-CFH antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.
Searchable Terms for CFH purchase	MBS212919 is an antibody targeting CFH. Also known as MOUSE ANTI HUMAN COMPLEMENT FACTOR H. Host: Mouse.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for CFH. It may not necessarily be applicable to this product.

NCBI GI #	62739186
NCBI GeneID	3075
NCBI Accession #	NP_000177.2
UniProt Primary Accession #	P08603
UniProt Secondary Accession #	P78435; Q14570; Q2TAZ5; Q38G77; Q5TFM3; Q8N708; Q9NU86; A5PL14
UniProt Related Accession #	P08603
GenBank Nucleotide #	NM_000186.3
NCBI Official Full Name	complement factor H isoform a
NCBI Official Synonym Full Names	complement factor H
NCBI Official Symbol	CFH
NCBI Official Synonym Symbols	FH; HF; HF1; HF2; HUS; FHL1; AHUS1; AMBP1; ARMD4; ARMS1; CFHL3
UniProt Protein Name	Complement factor H
UniProt Synonym Protein Names	H factor 1
Protein Family	Complement factor
UniProt Gene Name	CFH
UniProt Synonym Gene Names	HF; HF1; HF2
UniProt Entry Name	CFAH_HUMAN
NCBI Protein Information	complement factor H; H factor 1 (complement); H factor 2 (complement); adrenomedullin binding protein; age-related maculopathy susceptibility 1; beta-1-H-globulin; beta-1H; complement factor H, isoform b; factor H-like 1
NCBI Summary for CFH	This gene is a member of the Regulator of Complement Activation (RCA) gene cluster and encodes a protein with twenty short consensus repeat (SCR) domains. This protein is secreted into the bloodstream and has an essential role in the regulation of complement activation, restricting this innate defense mechanism to microbial infections. Mutations in this gene have been associated with hemolytic-uremic syndrome (HUS) and chronic hypocomplementemic nephropathy. Alternate transcriptional splice variants, encoding different isoforms, have been characterized. [provided by RefSeq, Oct 2011]

UniProt Comments for CFH	CFH: Factor H functions as a cofactor in the inactivation of C3b by factor I and also increases the rate of dissociation of the C3bBb complex (C3 convertase) and the (C3b)NBB complex (C5 convertase) in the alternative complement pathway. Genetic variations in CFH are associated with basal laminar drusen (BLD); also known as drusen of Bruch membrane or cuticular drusen or grouped early adult-onset drusen. Drusen are extracellular deposits that accumulate below the retinal pigment epithelium on Bruch membrane. Basal laminar drusen refers to an early adult-onset drusen phenotype that shows a pattern of uniform small, slightly raised yellow subretinal nodules randomly scattered in the macula. In later stages, these drusen often become more numerous, with clustered groups of drusen scattered throughout the retina. In time these small basal laminar drusen may expand and ultimately lead to a serous pigment epithelial detachment of the macula that may result in vision loss. Defects in CFH are the cause of complement factor H deficiency (CFHD). A disorder that can manifest as several different phenotypes, including asymptomatic, recurrent bacterial infections, and renal failure. Laboratory features usually include decreased serum levels of factor H, complement component C3, and a decrease in other terminal complement components, indicating activation of the alternative complement pathway. It is associated with a number of renal diseases with variable clinical presentation and progression, including membranoproliferative glomerulonephritis and atypical hemolytic uremic syndrome. Defects in CFH are a cause of susceptibility to hemolytic uremic syndrome atypical type 1 (AHUS1). 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. 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. Genetic variation in CFH is associated with age-related macular degeneration type 4 (ARMD4). ARMD is a multifactorial eye disease and the most common cause of irreversible vision loss in the developed world. In most patients, the disease is manifest as ophthalmoscopically visible yellowish accumulations of protein and lipid (known as drusen) that lie beneath the retinal pigment epithelium and within an elastin- containing structure known as Bruch membrane. 2 isoforms of the human protein are produced by alternative splicing. Protein type: Secreted, signal peptide; Secreted Chromosomal Location of Human Ortholog: 1q32 Cellular Component: extracellular space; extracellular region Molecular Function: heparin binding; heparan sulfate proteoglycan binding; protein binding Biological Process: complement activation, alternative pathway; regulation of complement activation; innate immune response; complement activation Disease: Complement Factor H Deficiency; Basal Laminar Drusen; Hemolytic Uremic Syndrome, Atypical, Susceptibility To, 1; Macular Degeneration, Age-related, 4
Research Articles on CFH	**1.** Genetic analysis revealed a novel heterozygous mutation in complement factor H (R83S) in addition to known risk polymorphisms carried by individuals with membranoproliferative glomerulonephritis.
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