补体因子 H 抗体

产品货号： mlR9525

英文名称： Factor H

中文名称： 补体因子 H 抗体

别　　名： adrenomedullin binding protein; age related maculopathy susceptibility 1; AHUS 1; AHUS1; AMBP 1; AMBP1; ARMD 4; ARMD4; ARMS 1; ARMS1; beta 1 H globulin; beta 1H; beta1H; CFAH_HUMAN; CFH; CFHL 3; CFHL3; complement factor H; complement factor H, isoform b; Factor H; factor H like 1; FH; FHL 1; FHL1; H factor 1 (complement); H factor 1; H factor 2 (complement); HF 1; HF 2; HF; HF1; HF2; HUS; MGC88246.

研究领域： 细胞生物　免疫学

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 IF=1:50-200 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 137kDa

细胞定位： 分泌型蛋白

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human Factor H:621-710/1231

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍： 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]

Function:

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.

Subcellular Location:

Secreted.

Tissue Specificity:

Expressed by the liver and secreted in plasma.

DISEASE:

Genetic variations in CFH are associated with basal laminar drusen (BLD) [MIM:126700]; 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) [MIM:609814]. 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) [MIM:235400]. 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.

Genetic variation in CFH is associated with age-related macular degeneration type 4 (ARMD4) [MIM:610698]. 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.

Similarity:

Contains 20 Sushi (CCP/SCR) domains.

SWISS:

P08603

Gene ID:

3075

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

产品图片