补体因子 I 轻链抗体

产品货号： mlR24314

英文名称： Factor I light chain

中文名称： 补体因子 I 轻链抗体

别　　名： AHUS3; ARMD13; C3b INA; C3b inactivator; C3B/C4B inactivator; C3BINA; CFAI_HUMAN; Cfi; Complement component I; Complement control protein factor I; Complement factor I; Complement factor I heavy chain; Complement factor I light chain; F1; factor I; FactorI; FI; I factor; IF; KAF; Konglutinogen activating factor; Light chain of factor I; OTTHUMP00000219728; OTTHUMP00000221928

研究领域： 心血管　免疫学

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应　： Mouse, Rat, Cow,

产品应用： IHC-P=1:400-800 （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： 22kDa

细胞定位： 细胞外基质 分泌型蛋白

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from mouse Factor I light chain :451-550/582

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： 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.

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.

Subcellular Location:

Secreted; extracellular space.

Tissue Specificity:

Plasma.

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.

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.

SWISS:

P05156

Gene ID:

3426

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