

## Rabbit Anti-Factor I heavy chain antibody

SL10339R

<b>Product Name</b>	Factor I heavy chain
<b>Chinese Name</b>	补体因子 I 重链抗体
<b>Alias</b>	Complement factor I heavy chain; Factor I heavy chain; AHUS3; C3b INA; C3b inactivator; C3B/C4B inactivator; C3BINA; CFAI_HUMAN; CFI; Complement component I; Complement control protein factor I; Complement factor I; F1; factor I; FactorI; FI; I factor; IF; KAF; Konglutinogen activating factor; Heavy chain of factor I; OTTHUMP00000219728.
<b>Research Area</b>	Cardiovascular immunology
<b>Immunogen Species</b>	Rabbit
<b>Clonality</b>	Polyclonal
<b>React Species</b>	Human, (predicted: Mouse, Rat, ) WB=1:500-2000 (Paraffin sections need antigen repair)
<b>Applications</b>	not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
<b>Theoretical molecular weight</b>	35/63kDa
<b>Cellular localization</b>	Extracellular matrix Secretory protein
<b>Form</b>	Liquid
<b>Concentration</b>	1mg/ml
<b>immunogen</b>	KLH conjugated synthetic peptide derived from human Complement factor I heavy chain: 261-360/582
<b>Lsotype</b>	IgG
<b>Purification</b>	affinity purified by Protein A
<b>Buffer Solution</b>	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol.
<b>Storage</b>	Shipped at 4°C. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
<b>Attention</b>	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
<b>PubMed</b>	<a href="#">PubMed</a>

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.

**Tissue Specificity:**

Plasma.

**Product Detail**

**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.

**SWISS:**  
P05156

**Gene ID:**  
3426

**Database links:**

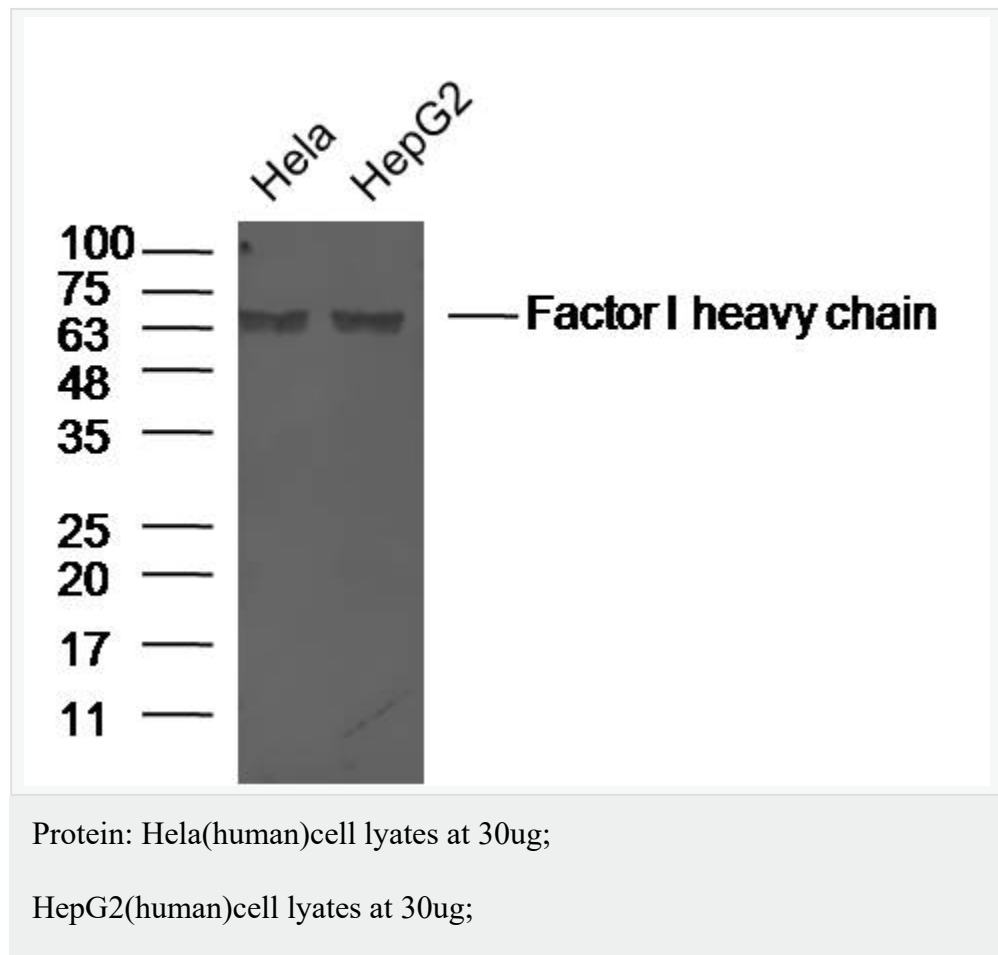
[Entrez Gene: 3426](#) Human

[Olim: 217030](#) Human

[SwissProt: P05156](#) Human

[Unigene: 312485](#) Human

**Product Picture**





Primary: Rabbit Anti-Factor I heavy chain(SL10339R) at 1:300;

Secondary: 800CW Conjugated Goat(polyclonal) Anti-Rabbit IgG(H+L)

at 1: 10000;

Predicted band size:39/63 kD

Observed band size:63 kD