Factor I Monoclonal Antibody

Catalog No	BYmab-13925
Isotype	IgG
Reactivity	Human;Rat;Mouse;
Applications	WB
Gene Name	CFI
Protein Name	Complement factor I
Immunogen	The antiserum was produced against synthesized peptide derived from human CFI. AA range:441-490
Specificity	Factor I Monoclonal Antibody detects endogenous levels of Factor I protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	CFI; IF; Complement factor I; C3B/C4B inactivator
Observed Band	Full lenth:66kD heavy chain: 50-58kD
Cell Pathway	Secreted, extracellular space. Secreted .
Tissue Specificity	Expressed in the liver by hepatocytes (PubMed:6327681). Also present in other cells such as monocytes, fibroblasts or keratinocytes (PubMed:6444659, PubMed:17320177).
Function	catalytic activity:Inactivates complement subcomponents C3b, iC3b and C4b by proteolytic cleavage.,disease: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.,disease:Defects in CFI are the cause of component I deficiency (CFI deficiency) [MIM:217030]. CFI deficiency is an autosomal recessive condition associated with a propensity to pyogenic infections.,disease:Defects in CFI may be associated with or predispose to hemolytic uraemic syndrome (HUS) [MIM:235400]. HUS, the most frequent cause of acute renal failure in childhood, is characterized by the association of acute renal failure, microangiopathic hemolytic

	anemia, and thrombocytopenia. The majority of HUS cases occur after an episode of infectious diarrhea, and are associated with E.coli
Background	This gene encodes a serine proteinase that is essential for regulating the complement cascade. The encoded preproprotein is cleaved to produce both heavy and light chains, which are linked by disulfide bonds to form a heterodimeric glycoprotein. This heterodimer can cleave and inactivate the complement components C4b and C3b, and it prevents the assembly of the C3 and C5 convertase enzymes. Defects in this gene cause complement factor I deficiency, an autosomal recessive disease associated with a susceptibility to pyogenic infections. Mutations in this gene have been associated with a predisposition to atypical hemolytic uremic syndrome, a disease characterized by acute renal failure, microangiopathic hemolytic anemia and thrombocytopenia. Primary glomerulonephritis with immune deposits and age-related macular degeneration are other conditions associated with mutations of this gene. [provided by Ref
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

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