





Factor I Monoclonal Antibody

| Catalog No | YP-mAb-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 1:500-1:2000 |
| 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 |



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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.

Products Images