

Note: Centrifuge before opening to ensure complete recovery of vial contents.

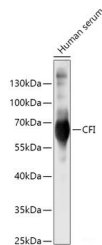
Description

Reactivity	Human,Mouse
Immunogen	Recombinant fusion protein of human CFI
Host	Rabbit
Isotype	IgG
Purification	Affinity purification
Conjugation	Unconjugated
Formulation	PBS with 0.02% sodium azide,50% glycerol,pH7.3.

Applications Recommended Dilution

WB	1:500-1:2000
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Data



Western blot analysis of extracts of Human serum using CFI Polyclonal Antibody at 1:500 dilution.

Observed Mw:60kDa
Calculated Mw:65kDa

Preparation & Storage

Storage Store at -20°C. Avoid freeze / thaw cycles.

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.

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