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CFI

Reactivity: Human

Tested applications:WB IHC IF

Recommended Dilution:WB 1:500 - 1:2000 IHC 1:50 - 1:200 IF 1:50 - 1:100

Calculated MW:66kDa

Observed MW: Refer to figures

Immunogen:

Recombinant protein of human CFI

Storage Buffer:

Store at -20. Avoid freeze / thaw cycles. Buffer: PBS with 0.02% sodium azide, 50% glycerol, pH7.3.

Synonym:

FI; IF; KAF; AHUS3; ARMD13; C3BINA; C3b-INA;

Catalog #:A5623

Antibody Type:

Polyclonal Antibody

Species: Rabbit

Gene ID:3426

Isotype: IgG

Swiss Prot:P05156

Purity: Affinity purification

For research use only.

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 uraemic syndrome, a disease characterized by acute renal failure, microangiopathic hemolytic anemia and thrombocytopenia. Primary glomerulonephritis with immmune deposits is another condition associated with mutation of this gene.

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