

DESCRIPTION

Source Mouse myeloma cell line, NS0-derived human Complement Factor H-related 1/CFHR1 protein
Glu19-Ala328, with a C-terminal 10-His tag
Accession # Q03591

N-terminal Sequence Analysis Glu19

Predicted Molecular Mass 37 kDa

SPECIFICATIONS

SDS-PAGE 39-48 kDa, reducing conditions

Activity Measured by its binding ability in a functional ELISA.
When Recombinant Human Complement Factor H-related 1/CFHR1 is immobilized at 1 µg/mL (100 µL/well), the concentration of Biotinylated Recombinant Mouse Complement Component C3d that produces 50% of the optimal binding response is 40-240 ng/mL.

Endotoxin Level <0.10 EU per 1 µg of the protein by the LAL method.

Purity >95%, by SDS-PAGE visualized with Silver Staining and quantitative densitometry by Coomassie® Blue Staining.

Formulation Lyophilized from a 0.2 µm filtered solution in Tris and NaCl with Trehalose. See Certificate of Analysis for details.

PREPARATION AND STORAGE

Reconstitution Reconstitute at 500 µg/mL in water.

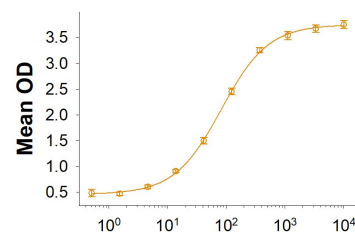
Shipping The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below.

Stability & Storage Use a manual defrost freezer and avoid repeated freeze-thaw cycles.

- 12 months from date of receipt, -20 to -70 °C as supplied.
- 1 month, 2 to 8 °C under sterile conditions after reconstitution.
- 3 months, -20 to -70 °C under sterile conditions after reconstitution.

DATA

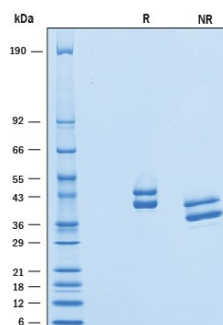
Binding Activity



Biotinylated Recombinant Mouse Complement Component C3d (ng/mL)

When Recombinant Human Complement Factor H-related 1/CFHR1 His-tag (Catalog # 4247-CH) is immobilized at 1 µg/mL, 100 µL/well, Biotinylated Recombinant Mouse Complement Component C3d binds with an ED₅₀ of 40-240 ng/mL.

SDS-PAGE



2 µg/lane of Recombinant Human Complement Factor H-related 1/CFHR1 was resolved with SDS-PAGE under reducing (R) and non-reducing (NR) conditions and visualized by Coomassie® Blue staining, showing bands under reducing conditions at 41 kDa and 45 kDa.

BACKGROUND

Complement factor H-related protein 1 (CFHR1) is a ~40 kDa secreted, primarily homodimeric member of the factor H family of glycoproteins (1). CFHR1 is produced by hepatocytes and circulates as two differentially glycosylated isoforms. The human complement factor H protein family consists of the complement and immune regulators factor H, the factor H-like protein 1 (FHL-1) and five factor H-related proteins (FHR-1 to -5) (2). The genes of this family are located on human chromosome 1q32, which is known as the regulator of complement activation (RCA) gene clusters (3). CFHRs are exclusively composed of individually folded protein domains, termed short consensus repeats (SCRs) or complement control modules. CFHR1 contains 5 SCRs. Although they are considered group 1 CFHRs based on conserved N-termini (2), compared to CFHR1, human CFHR2 and CFHR5 show 67.5% and 34.5% aa identity, respectively. All CFHRs, including CFHR1, are capable of binding complement factor C3b, discriminate between self and non-self cell surfaces, and have been proposed to deregulate complement activation by inhibiting interaction of CFH with C3b (2). CFHR1 inhibits complement C5 convertase activity (4). A common CFHR1/CFHR3 genetic deletion has been implicated as being protective against age-related macular degeneration (5) and nephropathy (6). The same deletion is associated with development of factor H auto-antibodies and susceptibility to atypical hemolytic uremic syndrome due to the deletion of CFHR1 (7-8). Additionally, CFHR1 is recruited to the surface of pathogens which is suggested to result in evasion of host complement attack (9).

References:

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2. Skerka, C. *et al.* (2013) *Mol. Immunol.* **56**:170.
3. Diaz-Guillen, M.A. *et al.* (1999) *Immunogenetics* **49**:549.
4. Heinen, S. *et al.* (2009) *Blood* **114**:2439.
5. Hughes, A.E. *et al.* (2006) *Nat. Genet.* **38**:1173.
6. Gharavi, A.G. *et al.* (2011) *Nat. Genet.* **43**:321.
7. Abarrategui-Garrido, C. (2009) *Blood* **114**:4261.
8. Munch, J. *et al.* (2017) *Clin. Kidney J.* **10**:742.
9. Reuter, M. *et al.* (2010) *J. Biol. Chem.* **285**:38473.