Product datasheet

Anti-Factor H antibody ab36134

Overview

Product name: Anti-Factor H antibody
Description: Goat polyclonal to Factor H
Host species: Goat
Specificity: This antibody is expected to recognize isoform a (NP_000177.2) only.
Tested applications: Suitable for: WB
Species reactivity: Reacts with: Human
Immunogen: Synthetic peptide: HLVPDRKDKQYK, corresponding to Internal sequence amino acids 577-588 of Human Factor H

Properties

Form: Liquid
Storage instructions: Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
Storage buffer: pH: 7.30
Preservative: 0.02% Sodium azide
Constituents: 0.5% BSA, 0.05% Tris buffered saline
Purity: Immunogen affinity purified
Purification notes: Purified from goat serum by ammonium sulphate precipitation followed by antigen affinity chromatography using the immunizing peptide.
Clonality: Polyclonal
Isotype: IgG

Applications

Our Abpromise guarantee covers the use of ab36134 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.
Function

Factor H functions as a cofactor in the inactivation of C3b by factor I and also increases the rate of dissociation of the C3bBb complex (C3 convertase) and the (C3b)NBB complex (C5 convertase) in the alternative complement pathway.

Tissue specificity

Expressed by the liver and secreted in plasma.

Involvement in disease

Genetic variations in CFH are associated with basal laminar drusen (BLD) [MIM:126700]; also known as drusen of Bruch membrane or cuticular drusen or grouped early adult-onset drusen. Drusen are extracellular deposits that accumulate below the retinal pigment epithelium on Bruch membrane. Basal laminar drusen refers to an early adult-onset drusen phenotype that shows a pattern of uniform small, slightly raised yellow subretinal nodules randomly scattered in the macula. In later stages, these drusen often become more numerous, with clustered groups of drusen scattered throughout the retina. In time these small basal laminar drusen may expand and ultimately lead to a serious pigment epithelial detachment of the macula that may result in vision loss.

Defects in CFH are the cause of complement factor H deficiency (CFH deficiency) [MIM:609814]. CFH deficiency determines uncontrolled activation of the alternative complement pathway with consumption of C3 and often other terminal complement components. It is associated with a number of renal diseases with variable clinical presentation and progression, including membranoproliferative glomerulonephritis and atypical hemolytic uremic syndrome. CFH deficiency patients may show increased susceptibility to meningococcal infections.

Defects in CFH are a cause of susceptibility to hemolytic uremic syndrome atypical type 1 (AHUS1) [MIM:235400]. An atypical form of hemolytic uremic syndrome. It is a complex genetic disease characterized by microangiopathic hemolytic anemia, thrombocytopenia, renal failure and absence of episodes of enterocolitis and diarrhea. In contrast to typical hemolytic uremic syndrome, atypical forms have a poorer prognosis, with higher death rates and frequent progression to end-stage renal disease. Note=Susceptibility to the development of atypical hemolytic uremic syndrome can be conferred by mutations in various components of or regulatory factors in the complement cascade system. Other genes may play a role in modifying the phenotype.

Genetic variation in CFH is associated with age-related macular degeneration type 4 (ARMD4) [MIM:610698]. ARMD is a multifactorial eye disease and the most common cause of irreversible vision loss in the developed world. In most patients, the disease is manifest as ophthalmoscopically visible yellowish accumulations of protein and lipid (known as drusen) that lie beneath the retinal pigment epithelium and within an elastin-containing structure known as Bruch membrane.

Sequence similarities

Contains 20 Sushi (CCP/SCR) domains.

Cellular localization

Secreted.

Images
Anti-Factor H antibody (ab36134) at 0.03 µg/ml + A431 lysate (RIPA buffer, 30µg total protein per lane)

**Predicted band size:** 139 kDa
**Observed band size:** 170 kDa

*why is the actual band size different from the predicted?*

ab36134 staining (0.03µg/ml) of A431 lysate (RIPA buffer, 30µg total protein per lane). Primary incubated for 1 hour. Detected by western blot using chemiluminescence. Primary incubated for 1 hour. Detected by western blot using chemiluminescence.

Anti-Factor H antibody (ab36134) at 1/2000 dilution + Human HepG2 whole cell lysate at 5 µg

**Secondary**

An HRP-conjugated donkey anti-goat IgG polyclonal at 1/2500 dilution

Developed using the ECL technique.

Performed under reducing conditions.

**Predicted band size:** 139 kDa
**Observed band size:** 150 kDa

*why is the actual band size different from the predicted?*

**Blocking Step:** 5% BSA for 1 hour at 24°C.

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