abcam

Product datasheet

Recombinant Human Factor H protein ab131757

画像数 1

製品の詳細

製品名	Recombinant Human Factor H protein
発現系	Wheat germ
アクセッション番号	P08603
タンパク質長	Full length protein
Animal free	No
由来	Recombinant
生物種	Human
配列	DCNELPPRRNTEILTGSWSDQTYPEGTQAIYKCRPGYRSLGN VIMVCRKG EWVALNPLRKCQKRPCGHPGDTPFGTFTLTGGNVFEYGVKAV YTCNEGYQ LLGEINYRECDTDGWTNDIPICEVVKCLPVTAPENGKIVSSA MEPDREYH FGQAVRFVCNSGYKIEGDEEMHCSDDGFWSKEKPKCVEISCK SPDVINGS PISQKIIYKENERFQYKCNMGYEYSERGDAVCTESGWRPLPS CEEKSCDN PYIPNGDYSPLRIKHRTGDEITYQCRNGFYPATRGNTAKCTS TGWIPAPR CTLKPCDYPDIKHGGLYHENMRRPYFPVAVGKYYSYYCDEHF ETPSGSYW DHIHCTQDGWSPAVPCLRKCYFPYLENGYNQNYGRKFVQGKS IDVACHPG YALPKAQTTVTCMENGWSPTPRCIRVSFTL
予測される分子量	73 kDa including tags
領域	20 to 449

特性

Our **Abpromise guarantee** covers the use of **ab131757** in the following tested applications.

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

アプリケーション	Western blot
	ELISA

	SDS-PAGE
製品の状態	Liquid

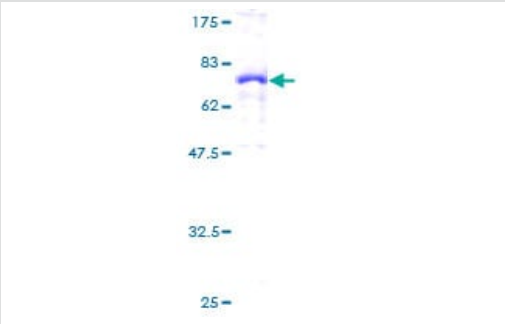
前処理および保存

保存方法および安定性	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl

関連情報

機能	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.
組織特異性	Expressed by the liver and secreted in plasma.
関連疾患	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 serous 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.
配列類似性	Contains 20 Sushi (CCP/SCR) domains.
細胞内局在	Secreted.

画像



SDS-PAGE - Recombinant Human Factor H protein (ab131757)

12.5% SDS-PAGE stained with Coomassie Blue showing ab131757.

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