ELK Biotechnology

FA8A (light chain, Cleaved-Glu1668) rabbit pAb

Cat No.:ES19999

For research use only

Overview

Product Name	FA8A (light chain, Cleaved-Glu1668) rabbit pAb
Host species	Rabbit
Applications	WB; ELISA
Species Cross-Reactivity	Human;Mouse
Recommended dilutions	WB 1:1000-2000 ELISA 1:5000-20000
Immunogen	Synthesized peptide derived from human FA8A (light chain, Cleaved-Glu1668)
Specificity	This antibody detects endogenous levels of Human,Mouse FA8A (light chain, Cleaved-Glu1668, protein was cleaved amino acid sequence between 1667-1668)
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Storage	Store at -20℃. Avoid repeated freeze-thaw cycles.
Protein Name	FA8A (light chain, Cleaved-Glu1668)
Gene Name	F8 F8C
Cellular localization	Secreted, extracellular space.
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Clonality	Polyclonal
Concentration	1 mg/ml
Observed band	75 260kD
Human Gene ID	2157
Human Swiss-Prot Number	P00451
Alternative Names	Coagulation factor VIII (Antihemophilic factor;AHF;Procoagulant component) [Cleaved into: Factor VIIIa heavy chain, 200 kDa isoform; Factor VIIIa heavy chain, 92 kDa isoform; Factor VIII B chain; Factor VIIIa light chain]
Background	disease:Defects in F8 are the cause of hemophilia A (HEMA) [MIM:306700]. HEMA is a common

recessive X-linked coagulation disorder. The frequency of hemophilia A is 1-2 in 10,000 male births in all ethnic groups. About 50% of patients have severe hemophilia A with F8C activity less than 1% of normal; they have frequent spontaneous bleeding into joints, muscles and internal organs. Moderately severe hemophilia A occurs in about 10% of patients; F8C activity is 2-5% of normal, and there is bleeding after minor trauma. Mild hemophilia A, which occurs in 30-40% of patients, is associated with F8C activity of 5-30% and bleeding occurs only after significant trauma or surgery. Of particular interest for the understanding of the function of F8C is the category of CRM (cross-reacting material) positive patients (approximately 5%) that have considerable amount of F8C in their plasma (at least 30% of normal), but the protein is non-functional; i.e., the F8C activity is much less than the plasma protein level. CRM-reduced is another category of patients in which the F8C antigen and activity are reduced to approximately the same level. Most mutations are CRM negative, and probably affect the folding and stability of the protein.,domain:Domain F5/8 type C 2 is responsible for phospholipid-binding and essential for factor VIII activity.,function:Factor VIII, along with calcium and phospholipid, acts as a cofactor for factor IXa when it converts factor X to the activated form, factor Xa.,mass spectrometry:Disulfated PubMed:10368977,mass spectrometry:Monosulfated PubMed:10368977,mass spectrometry:Nonsulfated PubMed:10368977,mass spectrometry:Sulfated PubMed:10368977,mass spectrometry:Trisulfated PubMed:10368977,online information:Factor VIII entry,online information:Factor VIII mutation db,pharmaceutical:Available under the names Kogenate (Bayer) and Recombinate (Baxter and American Home Products). Used to treat hemophilia A.,PTM:Sulfation on Tyr-1699 is essential for binding vWF.,similarity:Belongs to the multicopper oxidase family.,similarity:Contains 1 F5/8 type C

domain.,similarity:Contains 2 F5/8 type C domains.,similarity:Contains 3 F5/8 type A domains.,similarity:Contains 6 plastocyanin-like domains.,subunit:Interacts with vWF. vWF binding is essential for the stabilization of F8 in circulation.,