

Total Factor VIII Cell-Based Colorimetric ELISA Kit

Catalog No :	KA3254C
Reactivity :	Human;Mouse
Applications :	ELISA
Gene Name :	F8
Human Gene Id :	2157
Human Swiss Prot No :	P00451
Mouse Swiss Prot No :	Q06194
Storage Stability :	2-8 °C/6 months
Detection Method :	Colorimetric

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.,

Function :

acute inflammatory response, defense response, acute-phase response, inflammatory response, cell adhesion, blood coagulation, hemostasis, response to wounding, biological adhesion, wound healing, coagulation, regulation of body fluid levels, oxidation reduction,

Subcellular Location :

Secreted, extracellular space.

Sort :

22620

No4 :

1

Modifications :

Unmodified

Products Images