

factor VIII Rabbit pAb

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Catalog # AP52057

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P00451
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Calculated MW	267009
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human factor VIII
Epitope Specificity	1451-1550/2351
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Secreted, extracellular space.
DISEASE	Hemophilia A (HEMA) [MIM:306700]: A disorder of blood coagulation characterized by a permanent tendency to hemorrhage. About 50% of patients have severe hemophilia resulting in frequent spontaneous bleeding into joints, muscles and internal organs. Less severe forms are characterized by bleeding after trauma or surgery. Note=The disease is caused by mutations affecting the gene represented in this entry. Of particular interest for the understanding of the function of F8 is the category of CRM (cross-reacting material) positive patients (approximately 5%) that have considerable amount of F8 in their plasma (at least 30% of normal), but the protein is non-functional; i.e. the F8 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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes coagulation factor VIII, which participates in the intrinsic pathway of blood coagulation; factor VIII is a cofactor for factor IXa which, in the presence of Ca ²⁺ and phospholipids, converts factor X to the activated form Xa. This gene produces two alternatively spliced transcripts. Transcript variant 1 encodes a large glycoprotein, isoform a, which circulates in plasma and associates with von Willebrand factor in a noncovalent complex. This protein undergoes multiple cleavage events. Transcript variant 2 encodes a putative small protein, isoform b, which consists primarily of the phospholipid binding domain of factor VIIIc. This binding domain is essential for coagulant activity. Defects in this gene results in hemophilia A, a common recessive X-linked coagulation disorder. [provided by RefSeq, Jul 2008].

Additional Information

Gene ID	2157
Other Names	Coagulation factor VIII, Antihemophilic factor, AHF, Procoagulant component, Factor VIIIa heavy chain, 200 kDa isoform, Factor VIIIa heavy chain, 92 kDa isoform, Factor VIII B chain, Factor VIIIa light chain, F8, F8C
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	F8
Synonyms	F8C
Function	Factor VIII, along with calcium and phospholipid, acts as a cofactor for F9/factor IXa when it converts F10/factor X to the activated form, factor Xa.
Cellular Location	Secreted, extracellular space.

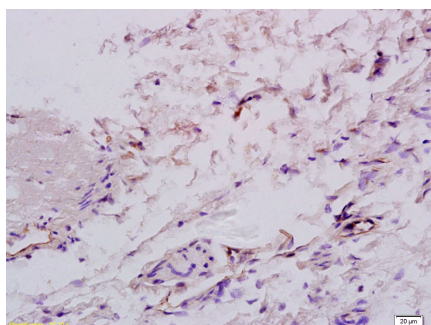
Background

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References

Truett M.A.,et al.DNA 4:333-349(1985).
Wood W.I.,et al.Nature 312:330-337(1984).
Levinson B.,et al.Genomics 14:585-589(1992).
Toole J.J.,et al.Nature 312:342-347(1984).
Gitschier J.,et al.Hum. Mol. Genet. 1:199-200(1992).

Images



Tissue/cell: human colon carcinoma; 4%
Paraformaldehyde-fixed and paraffin-embedded;
Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling
bathing for 15min; Block endogenous peroxidase by 3%
Hydrogen peroxide for 30min; Blocking buffer (normal
goat serum,C-0005) at 37??for 20 min;
Incubation: Anti-factor VIII(FVIII)(human) Polyclonal
Antibody, Unconjugated(AP52057) 1:200, overnight at
4°C, followed by conjugation to the secondary
antibody(SP-0023) and DAB(C-0010) staining

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.