

Factor VIII (GMA-8002): sc-65949

BACKGROUND

Factor VIII is a glycoprotein cofactor that serves as a critical component in the blood coagulation pathway. Insufficient expression levels or expression of nonfunctional Factor VIII results in hemophilia A, a common severe hereditary bleeding disorder. In the liver, the main site of Factor VIII synthesis, the mature polypeptide chain of 2,332 amino acids is secreted into the lumen of the endoplasmic reticulum, where it interacts with various chaperone proteins, including Calreticulin, Calnexin and IgG-binding protein. From the lumen, a portion of Factor VIII translocates to the Golgi and undergoes activation via proteolysis of both the heavy and light chain portions of the protein into three fragments. Finally, proteolysis of activated Factor VIII by Factor XIa, Protein C or Thrombin results in inactivation of Factor VIII. Survival of Factor VIII in the bloodstream requires binding to von Willebrand factor (VWF) at both the amino- and carboxy-termini of the light chain. Point mutations occurring in those binding domains as well as at other active sites of Factor VIII likely underly 90-95% of disease cases.

REFERENCES

1. Fulcher, C.A., Roberts, J.R. and Zimmerman, T.S. 1983. Thrombin proteolysis of purified Factor VIII: correlation of activation with generation of a specific polypeptide. *Blood* 61: 807-811.
2. Eaton, D., Rodriguez, H. and Vehar, G.A. 1986. Proteolytic processing of human Factor VIII: correlation of specific cleavages by Thrombin, Factor XIa, and activated Protein C with activation and inactivation of Factor VIII coagulant activity. *Biochemistry* 25: 505-512.
3. Foster, P.A. and Zimmerman, T.S. 1989. Factor VIII structure and function. *Blood Rev.* 3: 180-191.
4. Kaufman, R.J. 1992. Biological regulation of Factor VIII activity. *Annu. Rev. Med.* 43: 325-339.
5. Saenko, E.L. and Scandella, D. 1997. The acidic region of the Factor VIII light chain and the C2 domain together form the high affinity binding site for von Willebrand factor. *J. Biol. Chem.* 272: 18007-18014.
6. Bhopale, G.M. and Nanda, R.K. 2003. Blood coagulation Factor VIII: An overview. *J. Biosci.* 28: 783-789.

CHROMOSOMAL LOCATION

Genetic locus: F8 (human) mapping to Xq28.

SOURCE

Factor VIII (GMA-8002) is a mouse monoclonal antibody raised against B domain deleted recombinant Factor VIII of human origin, with epitope mapping to the A1 domain.

PRODUCT

Each vial contains 100 µg IgG_{2a} in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

APPLICATIONS

Factor VIII (GMA-8002) is recommended for detection of Factor VIII of human origin by solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Factor VIII siRNA (h): sc-43756, Factor VIII shRNA Plasmid (h): sc-43756-SH and Factor VIII shRNA (h) Lentiviral Particles: sc-43756-V.

Molecular Weight of Factor VIII heavy chain: 200 kDa.

Molecular Weight of Factor VIII light chain: 80 kDa.

Molecular Weight of Factor VIII cleaved fragments: 50/43/73 kDa.

Positive Controls: Caki-1 cell lysate: sc-2224 or human liver extract: sc-363766.

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.



See **Factor VIII light chain (RFFVIII C/5): sc-59512** for Factor VIII light chain antibody conjugates, including AC, HRP, FITC, PE, and Alexa Fluor[®] 488, 546, 594, 647, 680 and 790.