

Factor VIII heavy chain (H-140): sc-33583

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 Xa, 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., et al. 1983. Thrombin proteolysis of purified Factor VIII: Correlation of activation with generation of a specific polypeptide. *Blood* 61: 807-811.
2. Eaton, D., et al. 1986. Proteolytic processing of human Factor VIII: Correlation of specific cleavages by Thrombin, Factor Xa, and activated Protein C with activation and inactivation of Factor VIII coagulant activity. *Biochemistry* 25: 505-512.
3. Foster, P.A., et al. 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. et al. 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., et al. 2003. Blood coagulation Factor VIII: an overview. *J. Biosci.* 28: 783-789.

CHROMOSOMAL LOCATION

Genetic locus: F8 (human) mapping to Xq28; F8 (mouse) mapping to X A7.3.

SOURCE

Factor VIII heavy chain (H-140) is a rabbit polyclonal antibody raised against amino acids 261-400 mapping within an internal region of Factor VIII of human origin.

PRODUCT

Each vial contains 200 µg IgG 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 heavy chain (H-140) is recommended for detection of Factor VIII precursor and 200 kDa and 92 kDa heavy chain isoforms of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

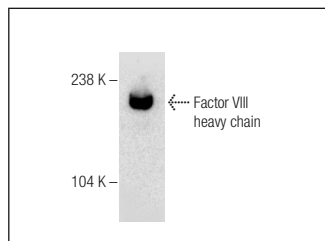
Factor VIII heavy chain (H-140) is also recommended for detection of Factor VIII precursor and 200 kDa and 92 kDa heavy chain isoforms in additional species, including equine, canine and bovine.

Suitable for use as control antibody for Factor VIII siRNA (h): sc-43756, Factor VIII siRNA (m): sc-44757, Factor VIII shRNA Plasmid (h): sc-43756-SH, Factor VIII shRNA Plasmid (m): sc-44757-SH, Factor VIII shRNA (h) Lentiviral Particles: sc-43756-V and Factor VIII shRNA (m) Lentiviral Particles: sc-44757-V.

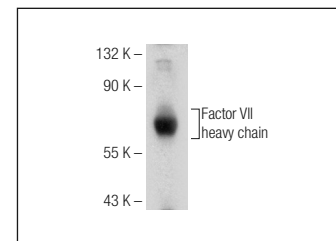
Molecular Weight of Factor VIII: 200/80/73/50/43 kDa.

Positive Controls: mouse kidney extract: sc-2255, human kidney extract: sc-363764 or human platelet extract: sc-363773.

DATA



Factor VIII heavy chain (H-140): sc-33583. Western blot analysis of Factor VIII heavy chain expression in human kidney tissue extract.



Factor VIII heavy chain (H-140): sc-33583. Western blot analysis of Factor VIII heavy chain expression in human platelet extract.

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 or our catalog for detailed protocols and support products.