SANTA CRUZ BIOTECHNOLOGY, INC.

Factor VIII light chain (H-100): sc-33584

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 polypetide 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.

CHROMOSOMAL LOCATION

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

SOURCE

Factor VIII light chain (H-100) is a rabbit polyclonal antibody raised against amino acids 2252-2351 mapping at the C-terminus 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.

STORAGE

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

RESEARCH USE

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

APPLICATIONS

Factor VIII light chain (H-100) is recommended for detection of Factor VIII precursor and light chain 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

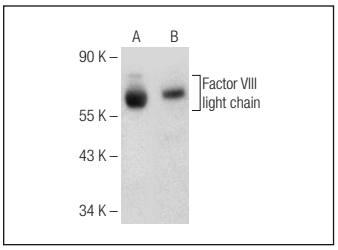
Factor VIII light chain (H-100) is also recommended for detection of Factor VIII precursor and light chain in additional species, including equine and bovine.

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.

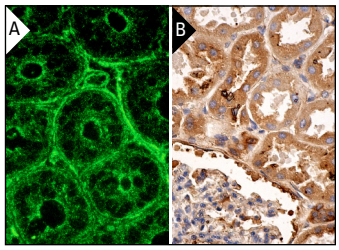
Molecular Weight of Factor VIII: 200/80/73/50/43 kDa.

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

DATA



Factor VIII light chain (H-100): sc-33584. Western blot analysis of Factor VIII light chain expression in human platelet whole cell lysate (**A**) and human kidney tissue extract (**B**).



Factor VIII light chain (H-100): sc-33584. Immunofluorescence staining of normal mouse intestine frozen section showing cytoplasmic and extracellular staining (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic staining of cells in glomeruli and tubules (**B**).

SELECT PRODUCT CITATIONS

1. Yadav, N., et al. 2009. The therapeutic effect of bone marrow-derived liver cells in the phenotypic correction of murine hemophilia A. Blood 114: 4552-4561.
2. Yadav, N., et al. 2011. Factor VIII can be synthesized in hemophilia A mice liver by bone marrow progenitor cells-derived hepatocytes and sinusoidal endothelial cells. Stem Cells Dev. 21: 110-120.