SANTA CRUZ BIOTECHNOLOGY, INC.

# Factor VIII (RFFVIIIC/8): sc-59514

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 occuring in those binding domains as well as at other active sites of Factor VIII likely underly 90-95% of disease cases.

## REFERENCES


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## STORAGE

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

## CHROMOSOMAL LOCATION

Genetic locus: F8 (human) mapping to Xq28.

## SOURCE

Factor VIII (RFFVIIIC/8) is a mouse monoclonal antibody raised against full length purified native protein of human origin.

## PRODUCT

Each vial contains 100 µg $IgG_1$ in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

Factor VIII (RFFVIIIC/8) is recommended for detection of the 360 kDa band of Factor VIII as well as the 210 kDa and 92 kDa band of human and porcine origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000); non cross-reactive with von Willebrand factor.

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.

## RESEARCH USE

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

## PROTOCOLS

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