

Factor VIII light chain (RFFVIII C/5): sc-59512

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.

CHROMOSOMAL LOCATION

Genetic locus: F8 (human) mapping to Xq28.

SOURCE

Factor VIII light chain (RFFVIII C/5) is a mouse monoclonal antibody raised against Factor VIII light chain of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Factor VIII light chain (RFFVIII C/5) is available conjugated to agarose (sc-59512 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-59512 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-59512 PE), fluorescein (sc-59512 FITC), Alexa Fluor[®] 488 (sc-59512 AF488), Alexa Fluor[®] 546 (sc-59512 AF546), Alexa Fluor[®] 594 (sc-59512 AF594) or Alexa Fluor[®] 647 (sc-59512 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-59512 AF680) or Alexa Fluor[®] 790 (sc-59512 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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RESEARCH USE

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

APPLICATIONS

Factor VIII light chain (RFFVIII C/5) is recommended for detection of the 360 kDa band of Factor VIII as well as the 80 kDa doublet bands of human and porcine origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500); 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.

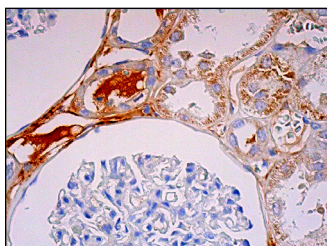
Molecular Weight of Factor VIII light chain: 200/80/73/50/43 kDa.

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

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker[™] Molecular Weight Standards: sc-2035, UltraCruz[®] Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz[®] Mounting Medium: sc-24941 or UltraCruz[®] Hard-set Mounting Medium: sc-359850. 3) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



Factor VIII light chain (RFFVIII C/5): sc-59512. Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic staining of cells in tubules and staining of plasma in blood vessels.

SELECT PRODUCT CITATIONS

1. Serrano, L.J., et al. 2018. Searching for a cell-based therapeutic tool for haemophilia A within the embryonic/foetal liver and the aorta-gonadomesonephros region. *Thromb. Haemost.* 118: 1370-1381.

STORAGE

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