



## FREM1 (M-300): sc-98447

### BACKGROUND

FREM1 (FRAS1 related extracellular matrix 1), also known as QBRICK or C9orf154, is a 2,179 amino acid protein that contains one C-type lectin domain, one Calx- $\beta$  domain and twelve CSPG repeats. Localized to the basement membrane of embryonic epidermal cells and secreted into extracellular space, FREM1 functions as an extracellular matrix protein that is essential for epidermal adhesion during embryogenesis and may also participate in epidermal differentiation. FREM1 exists as multiple alternatively spliced isoforms and is encoded by a gene which maps to human chromosome 9. Chromosome 9 contains 145 million base pairs and comprises 4% of the human genome, encoding nearly 900 genes. Hereditary hemorrhagic telangiectasia, which is characterized by harmful vascular defects, and familial dysautonomia, are both associated with chromosome 9. Notably, chromosome 9 encompasses the largest interferon family gene cluster.

### REFERENCES

1. Puente, X.S., Sánchez, L.M., Overall, C.M. and López-Otín, C. 2003. Human and mouse proteases: a comparative genomic approach. *Nat. Rev. Genet.* 4: 544-558.
2. Smyth, I., Du, X., Taylor, M.S., Justice, M.J., Beutler, B. and Jackson, I.J. 2004. The extracellular matrix gene FREM1 is essential for the normal adhesion of the embryonic epidermis. *Proc. Natl. Acad. Sci. USA* 101: 13560-13565.
3. Kiyozumi, D., Osada, A., Sugimoto, N., Weber, C.N., Ono, Y., Imai, T., Okada, A. and Sekiguchi, K. 2005. Identification of a novel cell-adhesive protein spatiotemporally expressed in the basement membrane of mouse developing hair follicle. *Exp. Cell Res.* 306: 9-23.
4. Kiyozumi, D., Sugimoto, N. and Sekiguchi, K. 2006. Breakdown of the reciprocal stabilization of QBRICK/FREM1, FRAS1, and FREM2 at the basement membrane provokes Fraser syndrome-like defects. *Proc. Natl. Acad. Sci. USA* 103: 11981-11986.
5. Online Mendelian Inheritance in Man, OMIM™. 2006. Johns Hopkins University, Baltimore, MD. MIM Number: 608944. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
6. Chiotaki, R., Petrou, P., Giakoumaki, E., Pavlakis, E., Sitaru, C. and Chalepakis, G. 2007. Spatiotemporal distribution of FRAS1/FREM proteins during mouse embryonic development. *Gene Expr. Patterns* 7: 381-388.
7. Petrou, P., Pavlakis, E., Dalezios, Y. and Chalepakis, G. 2007. Basement membrane localization of FREM3 is independent of the FRAS1/FREM1/FREM2 protein complex within the sublamina densa. *Matrix Biol.* 26: 652-658.
8. Petrou, P., Makrygiannis, A.K. and Chalepakis, G. 2008. The FRAS1/FREM family of extracellular matrix proteins: structure, function, and association with Fraser syndrome and the mouse bleb phenotype. *Connect. Tissue Res.* 49: 277-282.
9. Pavlakis, E., Makrygiannis, A.K., Chiotaki, R. and Chalepakis, G. 2008. Differential localization profile of FRAS1/FREM proteins in epithelial basement membranes of newborn and adult mice. *Histochem. Cell Biol.* 130: 785-793.

### CHROMOSOMAL LOCATION

Genetic locus: FREM1 (human) mapping to 9p22.3; Frem1 (mouse) mapping to 4 C3.

### SOURCE

FREM1 (M-300) is a rabbit polyclonal antibody raised against amino acids 1825-2124 mapping near the C-terminus of FREM1 of mouse origin.

### PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

### APPLICATIONS

FREM1 (M-300) is recommended for detection of FREM1 of mouse, rat and, to a lesser extent, human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g 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).

Suitable for use as control antibody for FREM1 siRNA (m): sc-75060, FREM1 siRNA (h): sc-75059, FREM1 shRNA Plasmid (m): sc-75060-SH, FREM1 shRNA Plasmid (h): sc-75059-SH, FREM1 shRNA (m) Lentiviral Particles: sc-75060-V and FREM1 shRNA (h) Lentiviral Particles: sc-75059-V.

Molecular Weight of FREM1: 244 kDa.

### RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

### 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.

### PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.