

FREM2 (N-19): sc-82599



The Power to Question

BACKGROUND

FREM2 (FRAS1 related extracellular matrix protein 2) is a 3,169 amino acid single-pass type I membrane protein that localizes to the extracellular side of the cell membrane and contains five Calx- β domains, as well as 12 CSPG repeats. Functioning as an extracellular matrix protein, FREM2 is required for the maintenance of skin and renal epithelia and is also thought to be involved in epidermal adhesion events. Defects or mutations in the gene encoding FREM2, which maps to human chromosome 13, are associated with Fraser syndrome, a multisystem malformation that is characterized by ear abnormalities, congenital heart defects and cutaneous syndactyly. FREM2 exists as multiple alternatively spliced isoforms.

REFERENCES

1. 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.
2. Jadeja, S., Smyth, I., Pitera, J.E., Taylor, M.S., van Haelst, M., Bentley, E., McGregor, L., Hopkins, J., Chalepakis, G., Philip, N., Perez Aytes, A., Watt, F.M., Darling, S.M., Jackson, I., Woolf, A.S. and Scambler, P.J. 2005. Identification of a new gene mutated in Fraser syndrome and mouse myelencephalic blebs. *Nat. Genet.* 37: 520-525.
3. Timmer, J.R., Mak, T.W., Manova, K., Anderson, K.V. and Niswander, L. 2005. Tissue morphogenesis and vascular stability require the *Frem2* protein, product of the mouse myelencephalic blebs gene. *Proc. Natl. Acad. Sci. USA* 102: 11746-11750.
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. Shafeghati, Y., Kniepert, A., Vakili, G. and Zenker, M. 2008. Fraser syndrome due to homozygosity for a splice site mutation of FREM2. *Am. J. Med. Genet. A* 146A: 529-531.
6. van Haelst, M.M., Maiburg, M., Baujat, G., Jadeja, S., Monti, E., Bland, E., Pearce, K., Hennekam, R.C. and Scambler, P.J. 2008. Molecular study of 33 families with Fraser syndrome new data and mutation review. *Am. J. Med. Genet. A* 146A: 2252-2257.
7. Online Mendelian Inheritance in Man, OMIM™. 2009. Johns Hopkins University, Baltimore, MD. MIM Number: 608945. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>

CHROMOSOMAL LOCATION

Genetic locus: FREM2 (human) mapping to 13q13.3; *Frem2* (mouse) mapping to 3 C.

SOURCE

FREM2 (N-19) is an affinity purified rabbit polyclonal antibody raised against a peptide mapping within an N-terminal extracellular domain of FREM2 of human origin.

PRODUCT

Each vial contains 100 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-82599 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

FREM2 (N-19) is recommended for detection of FREM2 of mouse, rat and human 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 solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000); non cross-reactive with family members FREM1 or FREM3.

FREM2 (N-19) is also recommended for detection of FREM2 in additional species, including equine, canine and bovine.

Suitable for use as control antibody for FREM2 siRNA (h): sc-75061, FREM2 siRNA (m): sc-75062, FREM2 shRNA Plasmid (h): sc-75061-SH, FREM2 shRNA Plasmid (m): sc-75062-SH, FREM2 shRNA (h) Lentiviral Particles: sc-75061-V and FREM2 shRNA (m) Lentiviral Particles: sc-75062-V.

Molecular Weight of FREM2: 220 kDa.

Positive Controls: C2C12 whole cell lysate: sc-364188 or NIH/3T3 whole cell lysate: sc-2210.

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) 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 or our catalog for detailed protocols and support products.