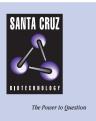
# SANTA CRUZ BIOTECHNOLOGY, INC.

# FREM2 (H-300): sc-98471



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- $\beta$  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 chromsome 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

- 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.
- 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.
- 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.
- 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.
- 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.
- 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.

#### CHROMOSOMAL LOCATION

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

#### SOURCE

FREM2 (H-300) is a rabbit polyclonal antibody raised against amino acids 2281-2580 mapping within an internal region of FREM2 of human origin.

# **STORAGE**

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

#### PRODUCT

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

#### **APPLICATIONS**

FREM2 (H-300) 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), 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 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.

#### **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<sup>™</sup> compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker<sup>™</sup> 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<sup>™</sup> Mounting Medium: sc-24941.

## **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.