

Munc13-4 (C-12): sc-271301

BACKGROUND

Munc13-4 is a member of the Munc13 family and is a homolog to Munc13-1. Munc13-4 lacks the C1 domain and N-terminal extension that are present in other Munc13 family members. It is a peripheral membrane, GTP-Rab 27a-binding protein. Munc13-4 has a ubiquitous tissue distribution; however, unlike related proteins Munc13-1, -2 and -3, Munc13-4 is mainly expressed outside the nervous system. High expression levels of Munc13-4 have been seen in mucous goblet and alveolar type II cells of the lung, as well as in cytotoxic T lymphocytes and mast cells. Munc13-4 localizes to secretory lysosomes. Overexpression of Munc13-4 enhances degranulation of mast cell secretory lysosomes, suggesting that it positively regulates secretory lysosome fusion and exocytosis. Mutations in Munc13-4 cause familial hemophagocytic lymphohistiocytosis subtype 3.

REFERENCES

- Feldmann, J., et al. 2003. Munc13-4 is essential for cytolytic granules fusion and is mutated in a form of familial hemophagocytic lymphohistiocytosis (FHL-3). *Cell* 115: 461-473.
- Shirakawa, R., et al. 2004. Munc13-4 is a GTP-Rab 27-binding protein regulating dense core granule secretion in platelets. *J. Biol. Chem.* 279: 10730-10737.
- Neef, M., et al. 2005. Munc13-4 is an effector of Rab 27a and controls secretion of lysosomes in hematopoietic cells. *Mol. Biol. Cell* 16: 731-741.
- Ishii, E., et al. 2005. Genetic subtypes of familial hemophagocytic lymphohistiocytosis: correlations with clinical features and cytotoxic T lymphocyte/natural killer cell functions. *Blood* 105: 3442-3448.
- Yamamoto, K., et al. 2005. Mutations of Syntaxin 11 and SNAP 23 genes as causes of familial hemophagocytic lymphohistiocytosis were not found in Japanese people. *J. Hum. Genet.* 50: 600-603.
- Hong, W., et al. 2005. Cytotoxic T lymphocyte exocytosis: bring on the SNAREs! *Trends Cell Biol.* 15: 644-650.
- Schneider, E.M., et al. 2006. Mutations of Perforin and Munc13-4 do not mark HLH by NK defects. *Pediatr. Blood Cancer* 46: 409-411.
- Ueda, I., et al. 2006. Correlation between phenotypic heterogeneity and gene mutational characteristics in familial hemophagocytic lymphohistiocytosis (FHL). *Pediatr. Blood Cancer* 46: 482-488.
- Mizumoto, H., et al. 2006. Familial hemophagocytic lymphohistiocytosis with the Munc13-4 mutation: a case report. *Eur. J. Pediatr.* 165: 384-388.

CHROMOSOMAL LOCATION

Genetic locus: UNC13D (human) mapping to 17q25.1.

SOURCE

Munc13-4 (C-12) is a mouse monoclonal antibody raised against amino acids 136-285 mapping near the N-terminus of Munc13-4 of human origin.

RESEARCH USE

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

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

Munc13-4 (C-12) is recommended for detection of Munc13-4 of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg 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 Munc13-4 siRNA (h): sc-62651, Munc13-4 shRNA Plasmid (h): sc-62651-SH and Munc13-4 shRNA (h) Lentiviral Particles: sc-62651-V.

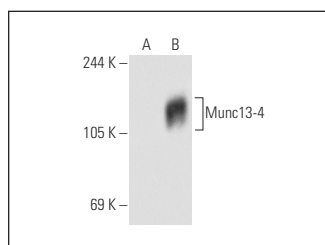
Molecular Weight of Munc13-4: 120 kDa.

Positive Controls: Munc13-4 (h): 293T Lysate: sc-177578, HeLa whole cell lysate: sc-2200 or Jurkat whole cell lysate: sc-2204.

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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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.

DATA



Munc13-4 (C-12): sc-271301. Western blot analysis of Munc13-4 expression in non-transfected: sc-117752 (A) and human Munc13-4 transfected: sc-177578 (B) 293T whole cell lysates.

STORAGE

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

PROTOCOLS

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