



Rabbit Anti-Munc 13-4 antibody

SL19103R

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| Product Name: | Munc 13-4 |
| Chinese Name: | UNC13D蛋白抗体 |
| Alias: | FHL 3; FHL3; FLJ00067; HLH 3; HLH3; HPLH 3; HPLH3; Jinx; Munc13 4; Munc13-4; Protein unc 13 homolog D; Protein unc-13 homolog D; UN13D_HUMAN; Unc 13 homolog D; UNC 13D; Unc-13 homolog D (C. elegans); Unc13 homolog D (C elegans); Unc13 homolog D; UNC13, C. elegans, homolog of, D; UNC13D; Unc13h4 |
| Organism Species: | Rabbit |
| Clonality: | Polyclonal |
| React Species: | Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rhesus monkey |
| Applications: | ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user. |
| Molecular weight: | 123kDa |
| Cellular localization: | cytoplasmicThe cell membrane |
| Form: | Lyophilized or Liquid |
| Concentration: | 1mg/ml |
| immunogen: | KLH conjugated synthetic peptide derived from human Munc 13-4:201-300/1090 |
| Lsotype: | IgG |
| Purification: | affinity purified by Protein A |
| Storage Buffer: | 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol. |
| Storage: | Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C. |
| PubMed: | PubMed |
| Product Detail: | This gene encodes a protein that is a member of the UNC13 family, containing similar domain structure as other family members but lacking an N-terminal phorbol ester-binding C1 domain present in other Munc13 proteins. The protein appears to play a role in vesicle maturation during exocytosis and is involved in regulation of cytolitic |

granules secretion. Mutations in this gene are associated with familial hemophagocytic lymphohistiocytosis type 3, a genetically heterogeneous, rare autosomal recessive disorder. [provided by RefSeq, Jul 2008]

Function:

Plays a role in cytotoxic granule exocytosis in lymphocytes. Required for both granule maturation and granule docking and priming at the immunologic synapse. Regulates assembly of recycling and late endosomal structures, leading to the formation of an endosomal exocytic compartment that fuses with perforin-containing granules at the immunologic synapse and licenses them for exocytosis. Regulates Ca(2+)-dependent secretory lysosome exocytosis in mast cells.

Subcellular Location:

Cytoplasm. Membrane. Late endosome. Recycling endosome. Lysosome. Colocalizes with cytotoxic granules at the plasma membrane. Localizes to endosomal exocytic vesicles.

Tissue Specificity:

Expressed at high levels in spleen, thymus and leukocytes. Also expressed in lung and placenta, and at very low levels in brain, heart, skeletal muscle and kidney. Expressed in cytotoxic T-lymphocytes (CTL) and mast cells.

DISEASE:

Defects in UNC13D are the cause of hemophagocytic lymphohistiocytosis familial type 3 (FHL3) [MIM:608898]; also known as HPLH3. Familial hemophagocytic lymphohistiocytosis (FHL) is a genetically heterogeneous, rare autosomal recessive disorder. It is characterized by immune dysregulation with hypercytokinemia and defective natural killer cell function. The clinical features of the disease include fever, hepatosplenomegaly, cytopenia, hypertriglyceridemia, hypofibrinogenemia, and neurological abnormalities ranging from irritability and hypotonia to seizures, cranial nerve deficits, and ataxia. Hemophagocytosis is a prominent feature of the disease, and a non-malignant infiltration of macrophages and activated T lymphocytes in lymph nodes, spleen, and other organs is also found.

Similarity:

Belongs to the unc-13 family.

Contains 2 C2 domains.

Contains 1 MHD1 (MUNC13 homology domain 1) domain.

Contains 1 MHD2 (MUNC13 homology domain 2) domain.

SWISS:

Q70J99

Gene ID:

201294

Database links:

[Entrez Gene: 201294](#) Human

[Entrez Gene: 70450](#) Mouse

[Entrez Gene: 192177](#) Rat

[Entrez Gene: 704431](#) Rhesus monkey

[Omin: 608897](#) Human

[SwissProt: Q70J99](#) Human

[SwissProt: B2RUP2](#) Mouse

[SwissProt: Q9R189](#) Rat

[Unigene: 41045](#) Human

[Unigene: 259460](#) Mouse

[Unigene: 198919](#) Rat

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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