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Munc 13-4 Rabbit pAb

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Catalog # AP57397

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q70J99
Predicted	Rat, Pig, Dog, Human, Mouse, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	123282
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Munc 13-4
Epitope Specificity	201-300/1090
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Cytoplasm. Membrane. Late endosome. Recycling endosome. Lysosome. Colocalizes with cytotoxic granules at the plasma membrane. Localizes to endosomal exocytic vesicles.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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 cytolytic 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]

Additional Information

Gene ID	201294

Other Names	Protein unc-13 homolog D, Munc13-4, UNC13D
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	UNC13D
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 licences them for exocytosis. Regulates Ca(2+)- dependent secretory lysosome exocytosis in mast cells.
Cellular Location	Cytoplasm. Membrane; Peripheral membrane protein. Late endosome. Recycling endosome. Lysosome. Note=Colocalizes with cytotoxic granules at the plasma membrane. Localizes to endosomal exocytic vesicles
Tissue Location	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.

Background

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