



Munc 13-4 Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP57397

Specification

Munc 13-4 Polyclonal Antibody - Product Information

Application
Primary Accession
Reactivity
Host
Clonality
Calculated MW
Physical State
Immunogen

Epitope Specificity Isotype **Purity** affinity purified by Protein A

Buffer

SUBCELLULAR LOCATION

SIMILARITY

DISEASE

IHC-P, IHC-F, IF, ICC, E

Q70199

Rat, Pig, Dog, Bovine

Rabbit Polyclonal 123 KDa Liquid

KLH conjugated synthetic peptide derived

from human Munc 13-4

201-300/1090

laG

0.01M TBS (pH7.4) with 1% BSA, 0.02%

Proclin300 and 50% Glycerol.

Cytoplasm. Membrane. Late endosome.

Recycling endosome. Lysosome.

Colocalizes with cytotoxic granules at the plasma membrane. Localizes to endosomal

exocytic vesicles.

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.

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



Important Note

other organs is also found. 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]

Munc 13-4 Polyclonal Antibody - Additional Information

Gene ID 201294

Other Names

Protein unc-13 homolog D, Munc13-4, UNC13D

Target/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.

Dilution

IHC-P~~N/A<br \><span class
="dilution_IHC-F">IHC-F~~N/A<br \><span class
="dilution_IF">IF~~1:50~200<br \>ICC~~N/A<br \>E~~N/A

Format

0.01M TBS(pH7.4), 0.09% (W/V) sodium azide and 50% Glyce

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.

Munc 13-4 Polyclonal Antibody - 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.

Munc 13-4 Polyclonal Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- <u>Immunofluorescence</u>
- <u>Immunoprecipitation</u>
- Flow Cytomety
- Cell Culture

Munc 13-4 Polyclonal Antibody - Images