10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

Perforin Rabbit pAb

Perforin Rabbit pAb
Catalog # AP94127

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P10820
Reactivity	Mouse
Predicted	Rat
Host	Rabbit
Clonality	Polyclonal
Calculated MW	62081
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from mouse Perforin
Epitope Specificity	161-250/554
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	Cytoplasmic granule lumen. Secreted. Cell membrane. Endosome lumen. Stored in cytoplasmic granules of cytolytic T-lymphocytes and secreted into the cleft between T-lymphocyte and target cell. Inserts into the cell membrane of target cells and forms pores. Membrane insertion and pore formation requires a major conformation change. May be taken up via endocytosis involving clathrin-coated vesicles and accumulate in a first time in large early endosomes.
DISEASE	Defects in PRF1 are the cause of hemophagocytic lymphohistiocytosis familial type 2 (FHL2) [MIM:603553]; also known as HPLH2. 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	The protein encoded by this gene has structural and functional similarities to complement component 9 (C9). Like C9, this protein creates transmembrane tubules and is capable of lysing non-specifically a variety of target cells. This protein is one of the main cytolytic proteins of cytolytic granules, and it is known to be a key effector molecule for T-cell- and natural killer-cell-mediated cytolysis. Defects in this gene cause familial hemophagocytic lymphohistiocytosis type 2 (HPLH2), a rare and lethal autosomal recessive disorder of early childhood. Alternative splicing results in multiple transcript variants encoding the same protein. [provided by RefSeq, Jul 2008].

Additional Information

Gene ID	18646
Other Names	Perforin-1, P1, Cytolysin, Lymphocyte pore-forming protein, Prf1, Pfp
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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.

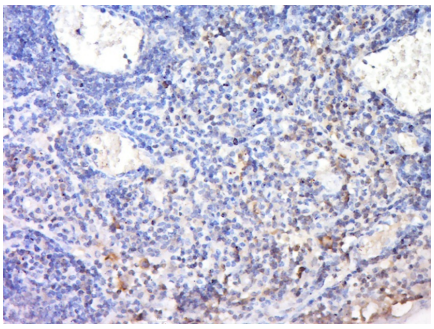
Protein Information

Name	Prf1
Synonyms	Pfp
Function	Pore-forming protein that plays a key role in granzyme- mediated programmed cell death, and in defense against virus-infected or neoplastic cells (PubMed:19446473, PubMed:21037563, PubMed:26306037, PubMed:2783478, PubMed:3261391, PubMed:35148176, PubMed:35705808, PubMed:7520535, PubMed:7526382, PubMed:7972104, PubMed:8164737). Can insert into the membrane of target cells in its calcium-bound form, oligomerize and form large pores (PubMed:19446473, PubMed:21037563, PubMed:26306037, PubMed:3261391, PubMed:35148176, PubMed:35705808, PubMed:7526382, PubMed:8164737). Promotes cytolysis and apoptosis of target cells by mediating the passage and uptake of cytotoxic granzymes (PubMed:19446473, PubMed:21037563, PubMed:26306037, PubMed:3261391, PubMed:35148176, PubMed:7526382, PubMed:8164737). Facilitates the delivery of cationic cargo protein, while anionic or neural proteins are not delivered efficiently (By similarity). Perforin pores allow the release of mature caspase-7 (CASP7) into the extracellular milieu (PubMed:35705808).
Cellular Location	Cytolytic granule. Secreted. Cell membrane; Multi-pass membrane protein. Endosome lumen {ECO:0000250|UniProtKB:P14222}. Note=Released from cytotoxic lymphocytes, together with proapoptotic granzymes: stored in cytolytic granules of cytolytic T-lymphocytes and secreted into the cleft between T-lymphocyte and target cell (PubMed:2040805, PubMed:8164737). May be taken up via endocytosis involving clathrin-coated vesicles and accumulate in a first time in large early endosomes (By similarity) Inserts into the cell membrane of target cells and forms pores (PubMed:19446473, PubMed:21037563). Membrane insertion and pore formation requires a major conformation change (PubMed:19446473, PubMed:21037563). {ECO:0000250|UniProtKB:P14222, ECO:0000269|PubMed:19446473, ECO:0000269|PubMed:2040805, ECO:0000269|PubMed:21037563, ECO:0000269|PubMed:8164737}
Tissue Location	Detected in cytotoxic T-lymphocytes and natural killer cells.

Background

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

Images



Paraformaldehyde-fixed, paraffin embedded (mouse lymph); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Perforin) Polyclonal Antibody, Unconjugated (AP94127) at 1:200 overnight at 4°C, followed by a conjugated secondary (sp-0023) for 20 minutes and DAB staining.