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Perforin Recombinant Rabbit mAb

Catalog # AP100320

Product Information

Application	WB, IHC-P, IHC-F, IF, IP, E
Primary Accession	P14222
Other Accession	P14222
Reactivity	Rat, Mouse
Host	Rabbit
Clonality	Recombinant
Calculated MW	61377
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the complement C6/C7/C8/C9 family. Contains 1 C2 domain. Contains 1 EGF-like domain. Contains 1 MACPF domain.
SUBUNIT	Monomer, as sobluble protein. Homooligomer. Oligomerization is required for pore formation.
Post-translational modifications	N-glycosylated.
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.
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	5551
Other Names	HPLH2; P1; PFP; Pfn; Prf-1; Cyta; RATCYTA; PERF_HUMAN; PRF1; Cytolysin; Lymphocyte pore-forming protein (PFP); PERF_MOUSE; Lymphocyte

	pore-forming protein; PERF_RAT; perforin 1; perforin 1 (pore forming protein); Perforin; perforin 1 (preforming protein); Perforin-1; FLH2; pore forming protein; PGFL; PIGF; PIGF-2; PLGF; SHGC-10760
Dilution	IHC-P=1:50-200,IHC-F=1:50-200,WB=1:500-2000,IF=1:50-200,ELISA=1:5000-10000,IP=1:50-200
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:20889983, PubMed:21037563, PubMed:24558045, PubMed:9058810, PubMed:9164947). Plays an important role in killing other cells that are recognized as non-self by the immune system, e.g. in transplant rejection or some forms of autoimmune disease (PubMed:9058810). Can insert into the membrane of target cells in its calcium-bound form, oligomerize and form large pores (PubMed:20889983, PubMed:21037563). Promotes cytolysis and apoptosis of target cells by mediating the passage and uptake of cytotoxic granzymes (PubMed:20038786, PubMed:20225066, PubMed:24558045, PubMed:32299851). Facilitates the delivery of cationic cargo protein, while anionic or neural proteins are not delivered efficiently (PubMed:24558045). Perforin pores allow the release of mature caspase-7 (CASP7) into the extracellular milieu (By similarity).
Cellular Location	Cytolytic granule. Secreted. Cell membrane; Multi-pass membrane protein. Endosome lumen. Note=Stored in cytolytic granules of cytolytic T-lymphocytes and secreted into the cleft between T- lymphocyte and target cell (PubMed:20038786). Inserts into the cell membrane of target cells and forms pores (PubMed:20889983). Membrane insertion and pore formation requires a major conformation change (PubMed:20889983). May be taken up via endocytosis involving clathrin- coated vesicles and accumulate in a first time in large early endosomes (PubMed:20038786).

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