



Perforin 1 Monoclonal Antibody

Catalog No	BYmab-10607
Isotype	IgG
Reactivity	Human;Rat;Mouse;
Applications	WB
Gene Name	PRF1
Protein Name	Perforin 1
Immunogen	The antiserum was produced against synthesized peptide derived from the C-terminal region of human PRF1. AA range:451-500
Specificity	Perforin 1 Monoclonal Antibody detects endogenous levels of Perforin 1
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	Perforin-1 (P1) (Cytolysin) (Lymphocyte pore-forming protein) (PFP)
Observed Band	61kD
Cell Pathway	Cytolytic granule . Secreted. Cell membrane ; Multi-pass membrane protein . Endosome lumen . 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). .
Tissue Specificity	Liver,Natural killer cell,Spleen,
Function	disease:Defects in PRF1 are the cause of familial hemophagocytic lymphohistiocytosis 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

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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. function: In the presence of calcium, perforin polymerizes into transmembrane tubules and is capable of lysing

Background

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 cytotoxicity. 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],

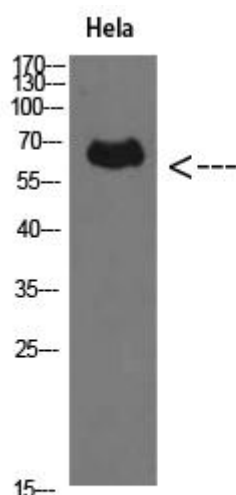
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using Perforin 1 Monoclonal Antibody