Anti-Perforin antibody ab7203

Overview

Product name: Anti-Perforin antibody
Description: Rabbit polyclonal to Perforin
Host species: Rabbit
Tested applications: Suitable for: ICC/IF, WB
Species reactivity: Reacts with: Mouse, Rat
Immunogen: Recombinant full length protein (Rat).
Positive control: ICC/IF: SW480 cells.

Properties

Form: Liquid
Storage instructions: Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.
Storage buffer: Preservative: 0.1% Sodium azide
Constituent: PBS
Purity: Protein A purified
Clonality: Polyclonal
Isotype: IgG

Applications

Our Abpromise guarantee covers the use of ab7203 in the following tested applications.
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

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<th>Application</th>
<th>Abreviews</th>
<th>Notes</th>
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<tr>
<td>ICC/IF</td>
<td></td>
<td>Use a concentration of 5 µg/ml.</td>
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<tr>
<td>WB</td>
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<td>1/1000. Predicted molecular weight: 70 kDa.</td>
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**Function**
Plays a key role in secretory granule-dependent cell death, and in defense against virus-infected or neoplastic cells. 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. Can insert into the membrane of target cells in its calcium-bound form, oligomerize and form large pores. Promotes cytolysis and apoptosis of target cells by facilitating the uptake of cytotoxic granzymes.

**Involvement in 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.

**Sequence similarities**
Belongs to the complement C6/C7/C8/C9 family.
Contains 1 C2 domain.
Contains 1 EGF-like domain.
Contains 1 MACPF domain.

**Domain**
The C2 domain mediates calcium-dependent binding to lipid membranes. A subsequent conformation change leads to membrane insertion of beta-hairpin structures and pore formation. The pore is formed by transmembrane beta-strands.

**Post-translational modifications**
N-glycosylated.

**Cellular localization**
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.

**Images**
ICC/IF image of ab7203 stained SW480 cells. The cells were 4% formaldehyde fixed (10 min) and then incubated in 1%BSA / 10% normal goat serum / 0.3M glycine in 0.1% PBS-Tween for 1h to permeabilise the cells and block non-specific protein-protein interactions. The cells were then incubated with the antibody (ab7203, 5µg/ml) overnight at +4°C. The secondary antibody (green) was ab96899, DyLight® 488 goat anti-rabbit IgG (H+L) used at a 1/250 dilution for 1h. Alexa Fluor® 594 WGA was used to label plasma membranes (red) at a 1/200 dilution for 1h. DAPI was used to stain the cell nuclei (blue) at a concentration of 1.43µM.
Western blot detection of rat Perforin from kidney on Day 3 in a glomerular nephritis model.

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