

PSMB8 Recombinant Mouse mAb

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Catalog # AP94385

Product Information

Application	WB, IHC-P, IHC-F, IF
Reactivity	Rat, Human
Host	Mouse
Clonality	Recombinant
Physical State	Liquid
Isotype	IgG1, Kappa
Purity	affinity purified by Protein G

Buffer 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.

SUBCELLULAR LOCATION Cytoplasm. Nucleus.

DISEASE Defects in PSMB8 are the cause of Nakajo syndrome (NKJO) [MIM:256040]; also called joint contractures muscular atrophy microcytic anemia and panniculitis-induced lipodystrophy. An autosomal recessive autoinflammatory disorder characterized by childhood onset of recurrent fever, joint stiffness and severe contractures of the hands and feet, erythematous skin lesions with subsequent development of lipodystrophy, and laboratory evidence of immune dysregulation. Accompanying features include muscle weakness and atrophy, hepatosplenomegaly, and microcytic anemia. Note=Mutation Met-75 has been found in chronic atypical neutrophilic dermatosis with lipodystrophy and elevated temperature syndrome (CANDLE syndrome). CANDLE patients have some overlapping features with NKJO patients, including a cutaneous eruption and lipodystrophy. They show a characteristic neutrophilic dermatosis with a mononuclear interstitial infiltrate in the dermis that seems pathognomonic for CANDLE syndrome (PubMed:21953331).

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

Background Descriptions The proteasome is a multicatalytic proteinase complex with a highly ordered ring-shaped 20S core structure. The core structure is composed of 4 rings of 28 non-identical subunits; 2 rings are composed of 7 alpha subunits and 2 rings are composed of 7 beta subunits. Proteasomes are distributed throughout eukaryotic cells at a high concentration and cleave peptides in an ATP/ubiquitin-dependent process in a non-lysosomal pathway. An essential function of a modified proteasome, the immunoproteasome, is the processing of class I MHC peptides. This gene encodes a member of the proteasome B-type family, also known as the T1B family, that is a 20S core beta subunit. This gene is located in the class II region of the MHC (major histocompatibility complex). Expression of this gene is induced by gamma interferon and this gene product replaces catalytic subunit 3 (proteasome beta 5 subunit) in the immunoproteasome. Proteolytic processing is required to generate a mature subunit. Two alternative transcripts encoding two isoforms have been identified; both isoforms are processed to yield the same mature subunit. [provided by RefSeq, Jul 2008].

Additional Information

Dilution	WB=1:500-1000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=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.

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