



HPS5 Monoclonal Antibody

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|--------------------|---|
| Catalog No | BYmab-05635 |
| Isotype | IgG |
| Reactivity | Human;Mouse |
| Applications | WB |
| Gene Name | HPS5 AIBP63 KIAA1017 |
| Protein Name | Hermansky-Pudlak syndrome 5 protein (Alpha-integrin-binding protein 63) (Ruby-eye protein 2 homolog) (Ru2) |
| Immunogen | Synthesized peptide derived from part region of human protein |
| Specificity | HPS5 Monoclonal Antibody detects endogenous levels of protein. |
| Formulation | Liquid in PBS containing 50% glycerol, 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 | |
| Observed Band | 124kD |
| Cell Pathway | Cytoplasm, cytosol . |
| Tissue Specificity | Widely expressed. Isoform 1:Highly expressed in lungs and testis. Isoform 2:Highly expressed in placenta, kidney, testis ovary, lung and thymus. |
| Function | disease:Defects in HPS5 are the cause of Hermansky-Pudlak syndrome type 5 (HPS5) [MIM:203300]. Hermansky-Pudlak syndrome (HPS) is a genetically heterogeneous, rare, autosomal recessive disorder characterized by oculocutaneous albinism, bleeding due to platelet storage pool deficiency, and lysosomal storage defects. This syndrome results from defects of diverse cytoplasmic organelles including melanosomes, platelet dense granules and lysosomes. Ceroid storage in the lungs is associated with pulmonary fibrosis, a common cause of premature death in individuals with HPS.,function:May regulate the synthesis and function of lysosomes and of highly specialized organelles, such as melanosomes and platelet dense granules. Might be involved in the regulation of general functions of integrins.,PTM:Phosphorylated upon DNA |

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damage, probably by ATM or ATR.,similarity:Belongs to the HPS5 family.,subunit

Background

This gene encodes a protein that may play a role in organelle biogenesis associated with melanosomes, platelet dense granules, and lysosomes. This protein interacts with Hermansky-Pudlak syndrome 6 protein and may interact with the cytoplasmic domain of integrin, alpha-3. Mutations in this gene are associated with Hermansky-Pudlak syndrome type 5. Multiple transcript variants encoding two distinct isoforms have been identified for this gene. [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

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