



HPS5 Polyclonal Antibody

Catalog No	BYab-05635
Isotype	IgG
Reactivity	Human;Mouse
Applications	WB;ELISA
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 Polyclonal Antibody detects endogenous levels of protein.
Formulation	Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000 ELISA 1:5000-20000
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