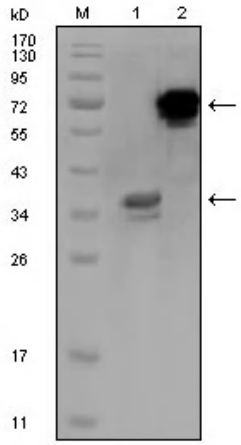
HPS-1 Monoclonal Antibody

Catalog No	YP-Ab-03403
Isotype	IgG
Reactivity	Human
Applications	WB;ELISA
Gene Name	HPS1
Protein Name	Hermansky-Pudlak syndrome 1 protein
Immunogen	Purified recombinant fragment of HPS-1 expressed in E. Coli.
Specificity	HPS-1 Monoclonal Antibody detects endogenous levels of HPS-1 protein.
Formulation	Ascitic fluid containing 0.03% sodium azide,0.5% BSA, 50%glycerol.
Source	Monoclonal, Mouse
Purification	Affinity purification
Dilution	Western Blot: 1/500 - 1/2000. ELISA: 1/10000. Not yet tested in other applications.
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	HPS1; HPS; Hermansky-Pudlak syndrome 1 protein
Observed Band	
Cell Pathway	cytoplasm,lysosome,integral component of plasma membrane,cytoplasmic, membrane-bounded vesicle,BLOC-3 complex,cytoplasmic vesicle,
Tissue Specificity	Ubiquitous.
Function	alternative products:Additional isoforms seem to exist,disease:Defects in HPS1 are the cause of Hermansky-Pudlak syndrome type 1 (HPS1) [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:Component of multiple cytoplasmic organelles. Apparently crucial for their normal development and function. May be involved in intracellular protein sorting.,online information:HPS1 mutations,online information:Retina International's Scientific Newsletter,tissue
Background	This gene encodes a protein that may play a role in organelle biogenesis associated with melanosomes, platelet dense granules, and lysosomes. The encoded protein is a component of three different protein complexes termed biogenesis of lysosome-related organelles complex (BLOC)-3, BLOC4, and

	BLOC5. Mutations in this gene are associated with Hermansky-Pudlak syndrome type 1. Alternative splicing results in multiple transcript variants. A pseudogene related to this gene is located on chromosome 22. [provided by RefSeq, Aug 2015],
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 using HPS-1 Monoclonal Antibody against truncated HPS1 recombinant protein (1) and HPS1-hIgGFc transfected CHO-K1 cell lysate (2).