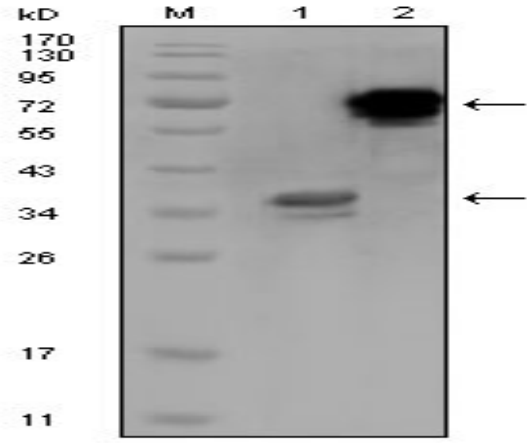
Best Tools for immunology Research

HPS-1 Monoclonal Antibody

Catalog No :	YM0336
Reactivity :	Human
Applications :	WB;ELISA
Target :	HPS-1
Gene Name :	HPS1
Protein Name :	Hermansky-Pudlak syndrome 1 protein
Human Gene Id :	3257
Human Swiss Prot No :	Q92902
Mouse Swiss Prot No :	O08983
Immunogen :	Purified recombinant fragment of HPS-1 expressed in E. Coli.
Specificity :	HPS-1 Monoclonal Antibody detects endogenous levels of HPS-1 protein.
Formulation :	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source :	Monoclonal, Mouse
Dilution :	WB 1:500 - 1:2000. ELISA: 1:10000. Not yet tested in other applications.
Purification :	Affinity purification
Storage Stability :	-15°C to -25°C/1 year(Do not lower than -25°C)
Molecularweight :	79kD
P References :	1. Platelets. 2007 Mar;18(2):150-7. 2. Intern Med. 2005 Jun;44(6):616-21. 3. Proc Natl Acad Sci U S A. 2003 Jul 22;100(15):8770-5. Epub 2003 Jul 7.

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],
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
Subcellular Location :	cytoplasm,lysosome,integral component of plasma membrane,cytoplasmic, membrane-bounded vesicle,BLOC-3 complex,cytoplasmic vesicle,
Expression :	Ubiquitous.
Sort :	7775
No4 :	1
Host :	Mouse
Modifications :	Unmodified

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).