

HPS-1 Mouse mAb

CatalogNo: YM0336

| Key Features

Host Species

- Mouse

Reactivity

- Human

Applications

- WB,ELISA

MW

- 79kD (Calculated)

| Recommended Dilution Ratios

WB 1:500-1:2000

ELISA 1:10000

Not yet tested in other applications.

| Storage

Storage* -15°C to -25°C/1 year(Do not lower than -25°C)

Formulation Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

| Basic Information

Clonality Monoclonal

| Immunogen Information

Immunogen Purified recombinant fragment of HPS-1 expressed in E. Coli.

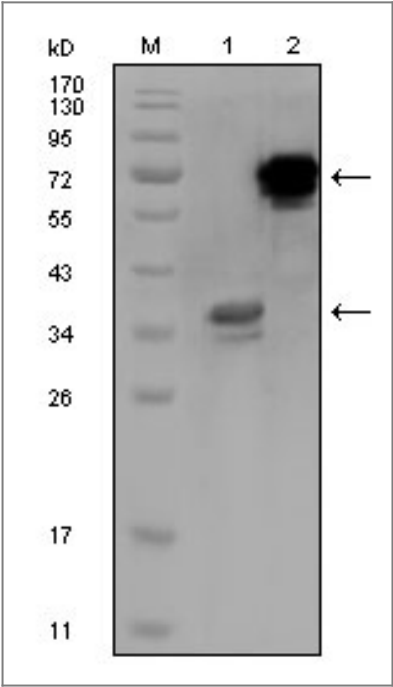
Specificity HPS-1 Monoclonal Antibody detects endogenous levels of HPS-1 protein.

| Target Information

Gene name HPS1

Protein Name	Hermansky-Pudlak syndrome 1 protein		
	Organism	Gene ID	UniProt ID
	Human	3257;	Q92902;
	Mouse		O08983;
Cellular Localization	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 specificity:Ubiquitous.,		

Validation Data



Western Blot analysis using HPS-1 Monoclonal Antibody against truncated HPS1 recombinant protein (1) and HPS1-hlgGfc transfected CHO-K1 cell lysate (2).

Contact information

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HPS-1 Mouse mAb

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