



Rabbit Anti-HPS4 antibody

SL17382R

Product Name:	HPS4
Chinese Name:	Hermansky-Pudlak综合征蛋白4抗体
Alias:	Hermansky Pudlak syndrome 4 protein; Light ear protein homolog.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	77kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human HPS4:531-630/709
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	This gene encodes a protein component of biogenesis of lysosome-related organelles complexes (BLOC). BLOC complexes are important for the formation of endosomal-lysosomal organelles such as melanosomes and platelet dense granules. Mutations in this gene result in subtype 4 of Hermansky-Pudlak syndrome, a form of albinism. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Aug 2012]

Function:

Hermansky-Pudlak syndrome is a disorder of organelle biogenesis in which oculocutaneous albinism, bleeding, and pulmonary fibrosis result from defects of melanosomes, platelet dense granules, and lysosomes. Mutations in HPS4 gene as well as several others can cause this syndrome. HPS4 appears to be important in organelle biogenesis and is similar to the mouse 'light ear' protein. Five transcript variants encoding different isoforms have been found for this gene. In addition, transcript variants utilizing alternative polyadenylation signals exist.

Subcellular Location:

lysosome, melanosome, membrane fraction and platelet dense granule

SWISS:

Q9NQG7

Gene ID:

89781

Database links:

[Entrez Gene: 89781](#) Human

[Omim: 606682](#) Human

[SwissProt: Q6P1K3](#) Human

[SwissProt: Q9NQG7](#) Human

[Unigene: 474436](#) Human

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.