



Rabbit Anti-HPS5 antibody

SL17383R

Product Name:	HPS5
Chinese Name:	Hermansky-Pudlak综合征蛋白5抗体
Alias:	AIBP63; HPS5_HUMAN; Alpha integrin binding protein 63; Hermansky Pudlak syndrome 5 protein; Ru2; Ruby eye protein 2 homolog.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Horse,Sheep,
Applications:	WB=1:500-2000ELISA=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:	127kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human HPS5:501-600/1129
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 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]

Function:

HPS5 may play a role in organelle biogenesis associated with melanosomes, platelet dense granules, and lysosomes. HPS5 interacts with Hermansky-Pudlak syndrome 6 protein and may interact with the cytoplasmic domain of integrin alpha 3. Mutations in HPS5 gene are associated with Hermansky-Pudlak syndrome type 5. Multiple transcript variants encoding two distinct isoforms have been identified for this gene.

Subunit:

Component of the biogenesis of lysosome-related organelles complex-2 (or BLOC2) composed of HPS3, HPS5 and HPS6. Directly interacts with HPS6. May interact with all alpha-integrin chains that have an aromatic residue before the first lysine of the conserved KXGFFKR motif, including ITGA2, ITGA3, ITGA5 and ITGA6.

Subcellular Location:

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.

DISEASE:

Hermansky-Pudlak syndrome 5 (HPS5) [MIM:614074]: A form of Hermansky-Pudlak syndrome, a genetically heterogeneous 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. Note=The disease is caused by mutations affecting the gene represented in this entry.

Similarity:

Belongs to the HPS5 family.

SWISS:

9UPZ3

Gene ID:

11234

Database links:

[Entrez Gene: 11234](#) Human

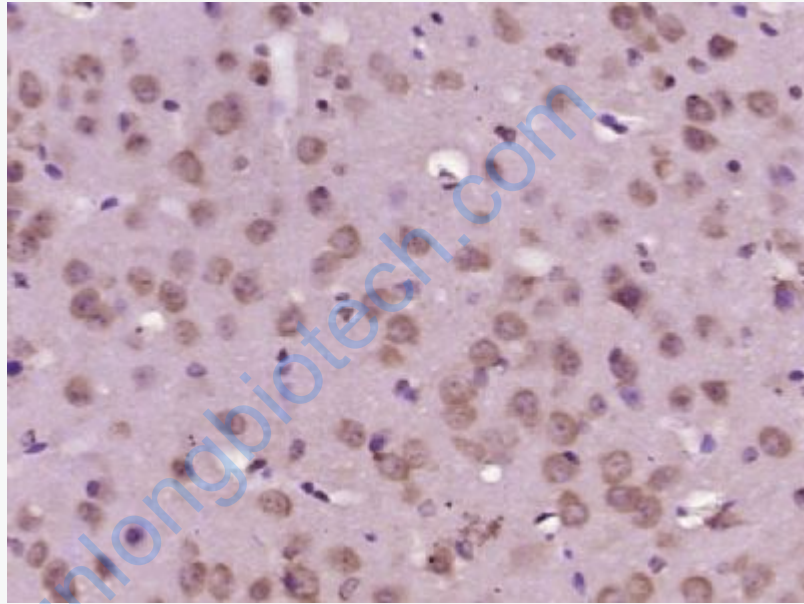
[Omim: 607521](#) Human

[SwissProt: Q9UPZ3](#) Human

[Unigene: 437599](#) Human

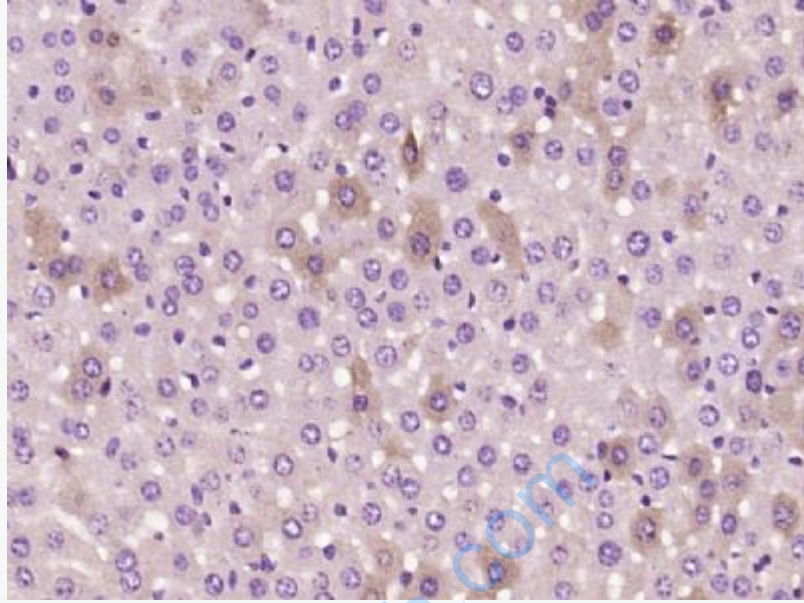
Important Note:

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



Picture:

Paraformaldehyde-fixed, paraffin embedded (mouse brain tissue); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (HPS5) Polyclonal Antibody, Unconjugated (SL17383R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.



Paraformaldehyde-fixed, paraffin embedded (mouse liver tissue); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (HPS5) Polyclonal Antibody, Unconjugated (SL17383R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.