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Rabbit Anti-HPS3 antibody

SL17381R

Product Name:	HPS3
Chinese Name:	Hermansky-Pudlak综合征蛋白3抗体
Alias:	Cocoa; DKFZp686F0413; FLJ22704; Hermansky Pudlak syndrome 3; Hermansky Pudlak syndrome 3 protein; SUTAL.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,Sheep,
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:	114kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human HPS3:561-660/1004
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 containing a potential clathrin-binding motif, consensus dileucine signals, and tyrosine-based sorting signals for targeting to vesicles of lysosomal lineage. The encoded protein may play a role in organelle biogenesis associated with melanosomes, platelet dense granules, and lysosomes. Mutations in this gene are associated with Hermansky-Pudlak syndrome type 3. Alternate splice variants exist, but their full length sequence has not been determined. [provided by RefSeq, Jul

2008]

Function:
HPS3 is involved in early stages of melanosome biogenesis and maturation. Defects in HPS3 are the cause of the cocoa (coa) mutant, and of Hermansky-Pudlak syndrome type 3 (HPS3). HPS3 is an 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.

Subcellular Location:
Cytoplasmic

SWISS:
Q969F9

Gene ID:
84343

Database links:

Entrez Gene: 84343 Human

Omim: 606118 Human

SwissProt: Q969F9 Human

Unigene: 591311 Human

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