

HPS6 Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP56681

Product Information

Application WB, IHC-P, IHC-F, IF, ICC, E

Primary Accession Q86YV9

Reactivity Rat, Pig, Bovine

Host Rabbit Clonality Polyclonal Calculated MW 82975 **Physical State** Liquid

Immunogen KLH conjugated synthetic peptide derived from human HPS6

501-600/775 **Epitope Specificity**

Isotype IgG

affinity purified by Protein A **Purity**

Buffer

SUBCELLULAR LOCATION

DISEASE

0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Microsome membrane. Cytoplasm > cytosol. Early endosome membrane. Defects in HPS6 are the cause of Hermansky-Pudlak syndrome type 6 (HPS6)

[MIM:614075]. 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.

This product as supplied is intended for research use only, not for use in **Important Note**

human, therapeutic or diagnostic applications.

This intronless gene encodes a protein that may play a role in organelle **Background Descriptions**

biogenesis associated with melanosomes, platelet dense granules, and

lysosomes. This protein interacts with Hermansky-Pudlak syndrome 5 protein. Mutations in this gene are associated with Hermansky-Pudlak syndrome type

6. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID 79803

Other Names Hermansky-Pudlak syndrome 6 protein, Ruby-eye protein homolog, Ru, HPS6

Target/Specificity Ubiquitous.

WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC=1:100-500,IF=1:100-50 Dilution

0,ELISA=1:5000-10000

Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When Storage

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.

Protein Information

Name HPS6

Function May regulate the synthesis and function of lysosomes and of highly

specialized organelles, such as melanosomes and platelet dense granules (PubMed:17041891). Acts as a cargo adapter for the dynein- dynactin motor complex to mediate the transport of lysosomes from the cell periphery to the perinuclear region. Facilitates retrograde lysosomal trafficking by linking the motor complex to lysosomes, and perinuclear positioning of lysosomes is crucial for the delivery of endocytic cargos to lysosomes, for lysosome

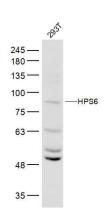
maturation and functioning (PubMed: 25189619).

Cellular Location Microsome membrane. Cytoplasm, cytosol. Early endosome membrane.

Lysosome membrane

Tissue Location Ubiquitous.

Images



Sample:

293T(Human) Cell Lysate at 30 ug

Primary: Anti-HPS6 (AP56681) at 1/300 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at

1/20000 dilution

Predicted band size: 83 kD Observed band size: 83 kD

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