Hermansky-Pudlak 综合征蛋白 4 抗体

产品货号： mlR17382

英文名称： HPS4

中文名称： Hermansky-Pudlak 综合征蛋白 4 抗体

别　　名： Hermansky Pudlak syndrome 4 protein; Light ear protein homolog.

研究领域： 细胞生物　免疫学　信号转导　转运蛋白

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 77kDa

细胞定位： 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human HPS4:531-630/709

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍： 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

Important Note:

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