HPS2 蛋白抗体

产品货号： mlR17380

英文名称： HPS2

中文名称： HPS2 蛋白抗体

别　　名： Adapter related protein complex 3 beta 1 subunit; Adapter-related protein complex 3 subunit beta-1; Adaptor protein complex AP-3 subunit beta-1; Adaptor protein complex AP3 beta1 subunit; ADTB3; ADTB3A; AP-3 complex subunit beta-1; AP3 complex beta1 subunit; AP3B1; AP3B1_HUMAN; Beta-3A-adaptin; Beta3A adaptin; Clathrin assembly protein complex 3 beta 1 large chain; Clathrin assembly protein complex 3 beta-1 large chain; HPS; PE.

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, Dog, Pig, Cow, Horse, Rabbit, Sheep, Cat,　Xenopus laevis

产品应用： WB=1:500-2000 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： 121kDa

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 HPS2:1-100/1094

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 that may play a role in organelle biogenesis associated with melanosomes, platelet dense granules, and lysosomes. The encoded protein is part of the heterotetrameric AP-3 protein complex which interacts with the scaffolding protein clathrin. Mutations in this gene are associated with Hermansky-Pudlak syndrome type 2. Two transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Nov 2012]

Function:

Subunit of non-clathrin- and clathrin-associated adaptor protein complex 3 that plays a role in protein sorting in the late-Golgi/trans-Golgi network (TGN) and/or endosomes. The AP complexes mediate both the recruitment of clathrin to membranes and the recognition of sorting signals within the cytosolic tails of transmembrane cargo molecules. AP-3 appears to be involved in the sorting of a subset of transmembrane proteins targeted to lysosomes and lysosome-related organelles.

Subcellular Location:

Golgi apparatus. Cytoplasmic vesicle > clathrin-coated vesicle membrane. Golgi apparatus. Component of the coat surrounding the cytoplasmic face of coated vesicles located at the Golgi complex.

Tissue Specificity:

Ubiquitously expressed.

Post-translational modifications:

Phosphorylated on serine residues.

DISEASE:

Defects in AP3B1 are the cause of Hermansky-Pudlak syndrome type 2 (HPS2) [MIM:608233]. 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. HPS2 differs from the other forms of HPS in that it includes immunodeficiency in its phenotype and patients with HPS2 have an increased susceptibility to infections.

Similarity:

Belongs to the adaptor complexes large subunit family.

SWISS:

O00203

Gene ID:

8546

Important Note:

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

applications.

产品图片