



Rabbit Anti-HPS2 antibody

SL17380R

Product Name:	HPS2
Chinese Name:	HPS2蛋白抗体
Alias:	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.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,Sheep,Cat, Xenopus laevis
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:	121kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human HPS2:1-100/1094
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. 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

Database links:

[Entrez Gene: 767602](#) Cow

[Entrez Gene: 403459](#) Dog

[Entrez Gene: 8546](#) Human

[Entrez Gene: 11774](#) Mouse

[Entrez Gene: 100049670](#) Pig

[Entrez Gene: 443724](#) Xenopus laevis

[Omim: 603401](#) Human

[SwissProt: Q32PG1](#) Cow

[SwissProt: Q7YRF1](#) Dog

[SwissProt: O00203](#) Human

[SwissProt: Q9Z1T1](#) Mouse

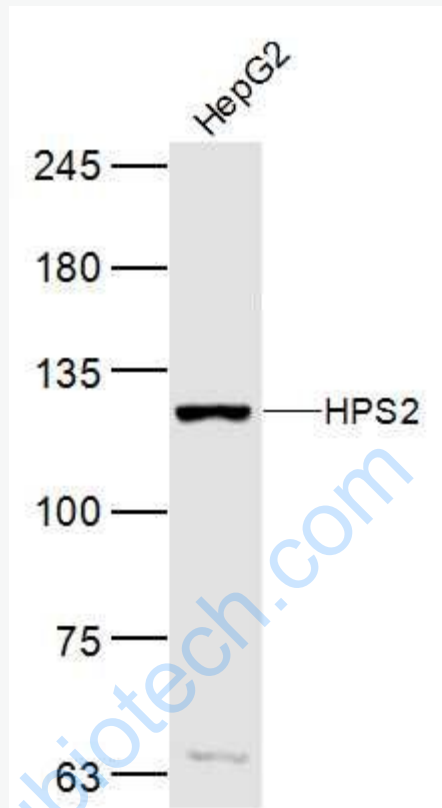
[Unigene: 532091](#) Human

[Unigene: 21185](#) Mouse

Important Note:

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

Picture:



Sample:

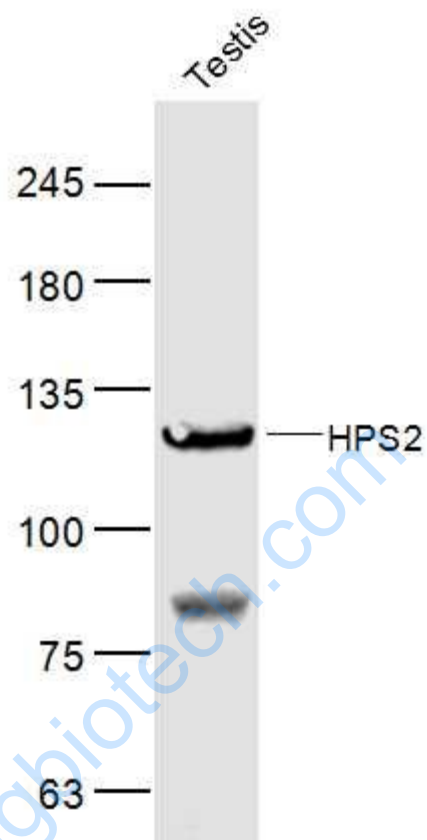
HepG2(Human) Lysate at 40 ug

Primary: Anti- HPS2 (SL17380R) at 1/300 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 121kD

Observed band size: 121 kD



Sample:

Testis(Mouse) Lysate at 40 ug

Primary: Anti- HPS2 (SL17380R) at 1/300 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 121kD

Observed band size: 121 kD