

## HPS-4 (S-16): sc-33380

### BACKGROUND

Hermansky-Pudlak syndrome (HPS) is a rare, genetically heterogeneous, autosomal recessive disorder. It is characterized by oculocutaneous albinism, lysosomal storage defects and prolonged bleeding due to platelet storage pool deficiency. There are ten HPS genes encoding HPS proteins that all interact within three distinct ubiquitously expressed protein complexes or biogenesis of lysosome-related organelle complexes. Defects in these genes cause HPS. HPS-4, also designated light-ear protein homolog, is important in organelle biosynthesis. Defects in the gene encoding for the HPS-4 protein, HPS4, can cause Hermansky-Pudlak syndrome 4 (HPS4).

### REFERENCES

1. Hirosawa, M., et al. 2001. Identification of novel transcribed sequences on human chromosome 22 by expressed sequence tag mapping. DNA Res. 8: 1-9.
2. Suzuki, T., et al. 2002. Hermansky-Pudlak syndrome is caused by mutations in HPS4, the human homolog of the mouse light-ear gene. Nat. Genet. 30: 321-324.
3. Huizing, M., et al. 2002. Hermansky-Pudlak syndrome: vesicle formation from yeast to man. Pigment Cell Res. 15: 405-419.
4. Nazarian, R., et al. 2003. Biogenesis of lysosome-related organelles complex 3 (BLOC-3): a complex containing the Hermansky-Pudlak syndrome (HPS) proteins HPS1 and HPS4. Proc. Natl. Acad. Sci. USA 100: 8770-8775.
5. Anderson, P.D., et al. 2003. Hermansky-Pudlak syndrome type 4 (HPS-4): clinical and molecular characteristics. Hum. Genet. 113: 10-17.
6. Bachli, E.B., et al. 2004. Hermansky-Pudlak syndrome type 4 in a patient from Sri Lanka with pulmonary fibrosis. Am. J. Med. Genet. A. 127: 201-207.

### CHROMOSOMAL LOCATION

Genetic locus: HPS4 (human) mapping to 22cen-q12.3; Hps4 (mouse) mapping to 5 F.

### SOURCE

HPS-4 (S-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of HPS-4 of human origin.

### PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-33380 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

### STORAGE

Store at 4° C, \*\*DO NOT FREEZE\*\*. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

### PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.

### APPLICATIONS

HPS-4 (S-16) is recommended for detection of HPS-4 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for HPS-4 siRNA (h): sc-44420, HPS-4 siRNA (m): sc-44996, HPS-4 shRNA Plasmid (h): sc-44420-SH, HPS-4 shRNA Plasmid (m): sc-44996-SH, HPS-4 shRNA (h) Lentiviral Particles: sc-44420-V and HPS-4 shRNA (m) Lentiviral Particles: sc-44996-V.

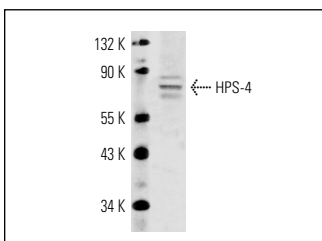
Molecular Weight of HPS-4: 77 kDa.

Positive Controls: HL-60 whole cell lysate: sc-2209.

### RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

### DATA



HPS-4 (S-16): sc-33380. Western blot analysis of HPS-4 expression in HL-60 whole cell lysate.

### RESEARCH USE

For research use only, not for use in diagnostic procedures.

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Try **HPS-4 (A-6): sc-398070** or **HPS-4 (H-3): sc-166638**, our highly recommended monoclonal alternatives to HPS-4 (S-16).