

HPS-1 (C-18): sc-33374

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 10 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-1 is a component of multiple cytoplasmic organelles and is important for their normal development and function. It plays a role in intracellular protein sorting.

REFERENCES

1. Oh, J., et al. 1996. Positional cloning of a gene for Hermansky-Pudlak syndrome, a disorder of cytoplasmic organelles. *Nat. Genet.* 14: 300-306.
2. Bailin, T., et al. 1997. Organization and nucleotide sequence of the human Hermansky-Pudlak syndrome (HPS) gene. *J. Invest. Dermatol.* 108: 923-927.

CHROMOSOMAL LOCATION

Genetic locus: HPS1 (human) mapping to 10q24.2; Hps1 (mouse) mapping to 19 C3.

SOURCE

HPS-1 (C-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of HPS-1 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-33374 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

RESEARCH USE

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

APPLICATIONS

HPS-1 (C-18) is recommended for detection of Hermansky-Pudlak Syndrome Protein 1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (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-1 siRNA (h): sc-44418, HPS-1 siRNA (m): sc-44993, HPS-1 shRNA Plasmid (h): sc-44418-SH, HPS-1 shRNA Plasmid (m): sc-44993-SH, HPS-1 shRNA (h) Lentiviral Particles: sc-44418-V and HPS-1 shRNA (m) Lentiviral Particles: sc-44993-V.

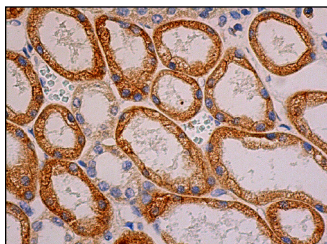
Molecular Weight of HPS-1: 79 kDa.

Positive Controls: K-562 whole cell lysate: sc-2203, HEL 92.1.7 cell lysate: sc-2270 or A-375 cell lysate: sc-3811.

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) 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. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



HPS-1 (C-18): sc-33374. Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic staining of cells in tubules.

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 or our catalog for detailed protocols and support products.

MONOS
Satisfaction
Guaranteed

Try **HPS-1 (5G12G2): sc-101435**, our highly recommended monoclonal alternative to HPS-1 (C-18).