

# HPS-6 (H-50): sc-98837

## 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. Hermansky-Pudlak syndrome 6 protein (HPS-6), also designated Ruby-eye protein homolog (Ru), regulates the function and synthesis of lysosomes and other specialized organelles. HPS-6 interacts with HPS-5 to form the BLOC2 complex, also referred to as biogenesis of lysosome-related organelles complex-2. Defects in HPS-6 can cause the Hermansky-Pudlak syndrome 6 (HPS6).

## REFERENCES

- Zhang, Q., Zhao, B., Li, W., Oiso, N., Novak, E.K., Rusiniak, M.E., Gautam, R., Chintala, S., O'Brien, E.P., Zhang, Y., Roe, B.A., Elliott, R.W., Eicher, E.M., Liang, P., Kratz, C., Legius, E., Spritz, R.A., O'Sullivan, T.N., Copeland, N.G., Jenkins, N.A. and Swank, R.T. 2003. Ru2 and Ru encode mouse orthologs of the genes mutated in human Hermansky-Pudlak syndrome types 5 and 6. *Nat. Genet.* 33: 145-153.
- Di Pietro, S.M., Falcón-Pérez, J.M. and Dell'Angelica, E.C. 2004. Characterization of BLOC-2, a complex containing the Hermansky-Pudlak syndrome proteins HPS-3, HPS-5 and HPS-6. *Traffic* 5: 276-283.
- Gautam, R., Chintala, S., Li, W., Zhang, Q., Tan, J., Novak, E.K., Di Pietro, S.M., Dell'Angelica, E.C. and Swank, R.T. 2004. The Hermansky-Pudlak syndrome 3 (cocoa) protein is a component of the biogenesis of lysosome-related organelles complex-2 (BLOC-2). *J. Biol. Chem.* 279: 12935-12942.
- Bossi, G., Booth, S., Clark, R., Davis, E.G., Liesner, R., Richards, K., Starcevic, M., Stinchcombe, J., Trambas, C., Dell'Angelica, E.C. and Griffiths, G.M. 2005. Normal lytic granule secretion by cytotoxic T lymphocytes deficient in BLOC-1, -2 and -3 and Myosins Va, VIIa and XV. *Traffic* 6: 243-251.
- SWISS-PROT/TrEMBL (Q86YV9). World Wide Web URL: <http://www.expasy.ch/sprot/sprot-top.html>

## CHROMOSOMAL LOCATION

Genetic locus: HPS6 (human) mapping to 10q24.32; Hps6 (mouse) mapping to 19 C3.

## SOURCE

HPS-6 (H-50) is a rabbit polyclonal antibody raised against amino acids 400-446 mapping within an internal region of HPS-6 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.

## STORAGE

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

## APPLICATIONS

HPS-6 (H-50) is recommended for detection of Hermansky-Pudlak Syndrome Protein 6 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).

HPS-6 (H-50) is also recommended for detection of Hermansky-Pudlak Syndrome Protein 6 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for HPS-6 siRNA (h): sc-44422, HPS-6 siRNA (m): sc-44999, HPS-6 shRNA Plasmid (h): sc-44422-SH, HPS-6 shRNA Plasmid (m): sc-44999-SH, HPS-6 shRNA (h) Lentiviral Particles: sc-44422-V and HPS-6 shRNA (m) Lentiviral Particles: sc-44999-V.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## RESEARCH USE

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

## PROTOCOLS

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



Try **HPS-6 (C-12): sc-373786**, our highly recommended monoclonal alternative to HPS-6 (H-50).