SANTA CRUZ BIOTECHNOLOGY, INC.

HPS-3 (N-15): sc-33375

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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-3 is a cytoplasmic protein that is important in early stages of melanosome maturation and biogenesis. HPS-3 is a widely expressed protein, but higher levels can be detected in placenta, liver and kidney.

REFERENCES

1. Huizing, M., et al. 2001. Hermansky-Pudlak syndrome type 3 in Ashkenazi Jews and other non-Puerto Rican patients with hypopigmentation and platelet storage-pool deficiency. Am. J. Hum. Gen. 69: 1022-1032.
2. Anikster, Y., et al. 2001. Mutation of a new gene causes a unique form of Hermansky-Pudlak syndrome in a genetic isolate of central Puerto Rico. Nat. Genet. 28: 376-380.
3. Di Pietro, S.M., et al. 2004. Characterization of BLOC-2, a complex containing the Hermansky-Pudlak syndrome proteins HPS3, HPS5 and HPS6. Traffic 5: 276-283.
4. Gautam, R., et al. 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.
5. Boissy, R.E., et al. 2005. Melanocyte-specific proteins are aberrantly trafficked in melanocytes of Hermansky-Pudlak syndrome-type 3. Am. J. Pathol. 166: 231-240.

CHROMOSOMAL LOCATION

Genetic locus: HPS3 (human) mapping to 3q24; Hps3 (mouse) mapping to 3 A2.

SOURCE

HPS-3 (N-15) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of HPS-3 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-33375 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 or our catalog for detailed protocols and support products.

APPLICATIONS

HPS-3 (N-15) is recommended for detection of Hermansky-Pudlak Syndrome Protein 3 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

HPS-3 (N-15) is also recommended for detection of Hermansky-Pudlak Syndrome Protein 3 in additional species, including canine, bovine and porcine.

Suitable for use as control antibody for HPS-3 siRNA (h): sc-44419, HPS-3 siRNA (m): sc-44867, HPS-3 shRNA Plasmid (h): sc-44419-SH, HPS-3 shRNA Plasmid (m): sc-44867-SH, HPS-3 shRNA (h) Lentiviral Particles: sc-44419-V and HPS-3 shRNA (m) Lentiviral Particles: sc-44867-V.

Molecular Weight of HPS-3: 114 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227 or A-431 whole cell lysate: sc-2201.

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.

RESEARCH USE

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