SANTA CRUZ BIOTECHNOLOGY, INC.

HPS-5 (C-12): sc-33384

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. Hermansky-Pudlak syndrome 5 protein (HPS-5), also designated Ruby-eye protein 2 homolog (Ru2), plays a role in Integrin general function regulation. It regulates the function and synthesis of lysosomes and other specialized organelles. HPS-5 interacts with HPS-6 to form the BLOC-2 complex, also referred to as biogenesis of lysosome-related organelles complex 2. Defects in the gene encoding for HPS-5 can cause Hermansky-Pudlak syndrome 5.

REFERENCES

1. Wixler, V., et al. 1999. Identification of novel interaction partners for the conserved membrane proximal region of α Integrin cytoplasmic domains. FEBS Lett. 445: 351-355.
2. Zhang, Q., et al. 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.
3. Di Pietro, S.M., et al. 2004. Characterization of BLOC-2, a complex containing the Hermansky-Pudlak syndrome proteins HPS-3, HPS-5 and HPS-6. Traffic 5: 276-283.
4. Huizing, M., et al. 2004. Cellular, molecular and clinical characterization of patients with Hermansky-Pudlak syndrome type 5. Traffic 5: 711-722.
5. 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.

CHROMOSOMAL LOCATION

Genetic locus: HPS5 (human) mapping to 11p14; Hps5 (mouse) mapping to 7 B4.

SOURCE

HPS-5 (C-12) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus of HPS-5 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-33384 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.

APPLICATIONS

HPS-5 (C-12) is recommended for detection of Hermansky-Pudlak syndrome protein 5 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).

Suitable for use as control antibody for HPS-5 siRNA (h): sc-44421 and HPS-5 siRNA (m): sc-44998.

Molecular Weight of HPS-5: 141 kDa.

Positive Controls: mouse liver extract: sc-2256.

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.

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.