

HPS-4 (A-6): sc-398070

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 HSP-4 protein, HPS4, can cause Hermansky-Pudlak syndrome 4 (HPS-4).

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* 127A: 201-207.

CHROMOSOMAL LOCATION

Genetic locus: HPS4 (human) mapping to 22q12.1; Hps4 (mouse) mapping to 5 F.

SOURCE

HPS-4 (A-6) is a mouse monoclonal antibody raised against amino acids 526-671 mapping at the C-terminus of HPS-4 of mouse origin.

PRODUCT

Each vial contains 200 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

HPS-4 (A-6) is available conjugated to agarose (sc-398070 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-398070 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-398070 PE), fluorescein (sc-398070 FITC), Alexa Fluor® 488 (sc-398070 AF488), Alexa Fluor® 546 (sc-398070 AF546), Alexa Fluor® 594 (sc-398070 AF594) or Alexa Fluor® 647 (sc-398070 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-398070 AF680) or Alexa Fluor® 790 (sc-398070 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Alexa Fluor® is a trademark of Molecular Probes, Inc., Oregon, USA

APPLICATIONS

HPS-4 (A-6) is recommended for detection of HPS-4 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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.

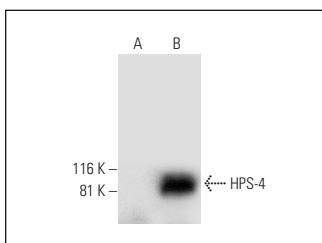
Molecular Weight of HPS-4: 77 kDa.

Positive Controls: HPS-4 (h): 293T Lysate: sc-117112.

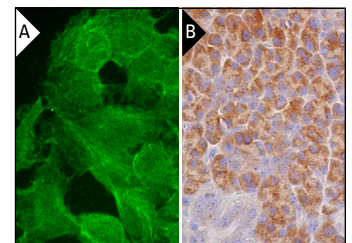
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



HPS-4 (A-6): sc-398070. Western blot analysis of HPS-4 expression in non-transfected: sc-117752 (A) and human HPS-4 transfected: sc-117112 (B) 293T whole cell lysates.



HPS-4 (A-6): sc-398070. Immunofluorescence staining of formalin-fixed Hep G2 cells showing membrane localization (A). Immunoperoxidase staining of formalin fixed, paraffin-embedded mouse pancreas tissue showing cytoplasmic staining of exocrine glandular cells. Blocked with 0.25X UltraCruz® Blocking Reagent: sc-516214. Detection reagents used: m-IgGκ BP-B: sc-516142 and ImmunoCruz® ABC Kit: sc-516216 (B).

STORAGE

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

RESEARCH USE

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