

HPS-4 (D-10): sc-271425

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-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 (HPS4).

REFERENCES

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4. Anderson, P.D., Huizing, M., Claassen, D.A., White, J. and Gahl, W.A. 2003. Hermansky-Pudlak syndrome type 4 (HPS-4): clinical and molecular characteristics. *Hum. Genet.* 113: 10-17.
5. Nazarian, R., Falcón-Pérez, J.M. and Dell'Angelica, E.C. 2003. Biogenesis of lysosome-related organelles complex 3 (BLOC-3): a complex containing the Hermansky-Pudlak syndrome (HPS) proteins HPS-1 and HPS-4. *Proc. Natl. Acad. Sci. USA* 100: 8770-8775.
6. Bachli, E.B., Brack, T., Eppler, E., Stallmach, T., Trüeb, R.M., Huizing, M. and Gahl, W.A. 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.

SOURCE

HPS-4 (D-10) is a mouse monoclonal antibody raised against amino acids 471-620 mapping near the C-terminus of HPS-4 of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain 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-4 (D-10) is recommended for detection of HPS-4 of 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 shRNA Plasmid (h): sc-44420-SH and HPS-4 shRNA (h) Lentiviral Particles: sc-44420-V.

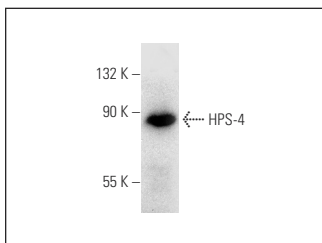
Molecular Weight of HPS-4: 77 kDa.

Positive Controls: HL-60 whole cell lysate: sc-2209, C32 whole cell lysate: sc-2205 or SK-MEL-28 cell lysate: sc-2236.

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 (D-10): sc-271425. Western blot analysis of HPS-4 expression in SK-MEL-28 whole cell lysate.

SELECT PRODUCT CITATIONS

1. Balci, A., Solano-Collado, V., Baldassarre, M. and Spanò, S. 2020. VARP and Rab9 are dispensable for the Rab32/BLOC-3 dependent *Salmonella* killing. *Front. Cell. Infect. Microbiol.* 10: 581024.

RESEARCH USE

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

PROTOCOLS

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