SANTA CRUZ BIOTECHNOLOGY, INC.

HPS-4 (H-150): sc-98835

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

REFERENCES

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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 127: 201-207.

CHROMOSOMAL LOCATION

Genetic locus: HPS4 (human) mapping to 22q12.1.

SOURCE

HPS-4 (H-150) is a rabbit polyclonal 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 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 (H-150) is recommended for detection of HPS-4 of 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).

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.

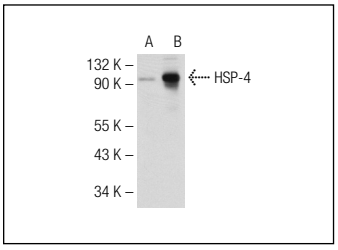
Molecular Weight of HPS-4: 77 kDa.

Positive Controls: HL-60 whole cell lysate: sc-2209 or HPS-4 (h3): 293T Lysate: sc-128830.

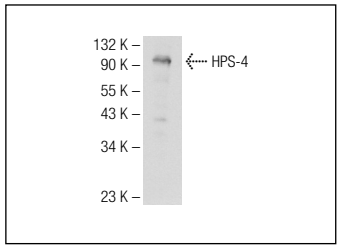
RECOMMENDED SECONDARY REAGENTS

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.

DATA



HPS-4 (H-150): sc-98835. Western blot analysis of HPS-4 expression in non-transfected: sc-117752 (**A**) and human HPS-4 transfected: sc-128830 (**B**) 293T whole cell lysates.



HPS-4 (H-150): sc-98835. Western blot analysis of HPS-4 expression in HL-60 whole cell lysate.

RESEARCH USE

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