

# HPS-1 (5G12G2): sc-101435

## 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-1 is a component of multiple cytoplasmic organelles and is important for their normal development and function. It plays a role in intracellular protein sorting.

## REFERENCES

- Oh, J., et al. 1996. Positional cloning of a gene for Hermansky-Pudlak syndrome, a disorder of cytoplasmic organelles. *Nat. Genet.* 14: 300-306.
- Bailin, T., et al. 1997. Organization and nucleotide sequence of the human Hermansky-Pudlak syndrome (HPS) gene. *J. Invest. Dermatol.* 108: 923-927.
- Wildenberg, S.C., et al. 1998. Identification of a novel transcript produced by the gene responsible for the Hermansky-Pudlak syndrome in Puerto Rico. *J. Invest. Dermatol.* 110: 777-781.
- Oh, J., et al. 1998. Mutation analysis of patients with Hermansky-Pudlak syndrome: a frameshift hot spot in the HPS gene and apparent locus heterogeneity. *Am. J. Hum. Genet.* 62: 593-598.
- Oetting, W.S., et al. 1999. Molecular basis of albinism: mutations and polymorphisms of pigmentation genes associated with albinism. *Hum. Mutat.* 13: 99-115.
- 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.

## CHROMOSOMAL LOCATION

Genetic locus: HPS1 (human) mapping to 10q24.2; Hps1 (mouse) mapping to 19 C3.

## SOURCE

HPS-1 (5G12G2) is a mouse monoclonal antibody raised against recombinant HPS-1 of human origin.

## PRODUCT

Each vial contains 200 µg IgG<sub>2a</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

HPS-1 (5G12G2) is available conjugated to agarose (sc-101435 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-101435 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-101435 PE), fluorescein (sc-101435 FITC), Alexa Fluor<sup>®</sup> 488 (sc-101435 AF488), Alexa Fluor<sup>®</sup> 546 (sc-101435 AF546), Alexa Fluor<sup>®</sup> 594 (sc-101435 AF594) or Alexa Fluor<sup>®</sup> 647 (sc-101435 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor<sup>®</sup> 680 (sc-101435 AF680) or Alexa Fluor<sup>®</sup> 790 (sc-101435 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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## APPLICATIONS

HPS-1 (5G12G2) is recommended for detection of HPS-1 of mouse, rat and 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)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for HPS-1 siRNA (h): sc-44418, HPS-1 siRNA (m): sc-44993, HPS-1 shRNA Plasmid (h): sc-44418-SH, HPS-1 shRNA Plasmid (m): sc-44993-SH, HPS-1 shRNA (h) Lentiviral Particles: sc-44418-V and HPS-1 shRNA (m) Lentiviral Particles: sc-44993-V.

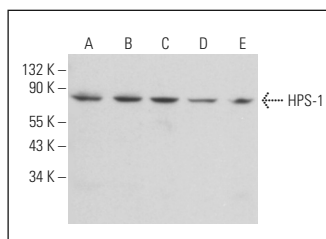
Molecular Weight of HPS-1: 79 kDa.

Positive Controls: A-375 cell lysate: sc-3811, K-562 whole cell lysate: sc-2203 or HEL 92.1.7 cell lysate: sc-2270.

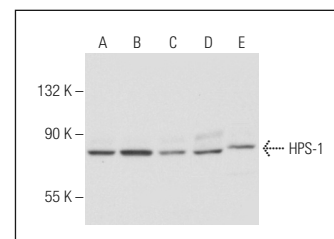
## RECOMMENDED SUPPORT REAGENTS

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<sup>™</sup> Molecular Weight Standards: sc-2035, UltraCruz<sup>®</sup> 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).

## DATA



HPS-1 (5G12G2): sc-101435. Western blot analysis of HPS-1 expression in HEL 92.1.7 (A), TF-1 (B), c4 (C), NIH/3T3 (D) and KNRK (E) whole cell lysates.



HPS-1 (5G12G2): sc-101435. Western blot analysis of HPS-1 expression in K-562 (A), HEL 92.1.7 (B), NCI-H929 (C), SK-MEL-24 (D) and A-375 (E) whole cell lysates.

## 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.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.