

# HPS-6 (C-12): sc-373786

## 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. Hermansky-Pudlak syndrome 6 protein (HPS-6), also designated Ruby-eye protein homolog (Ru) regulates the function and synthesis of lysosomes and other specialized organelles. HPS-6 interacts with HPS-5 to form the BLOC2 complex, also referred to as biogenesis of lysosome-related organelles complex-2. Defects in HPS6 can cause the Hermansky-Pudlak syndrome 6 (HPS6).

## REFERENCES

1. 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.
2. Di Pietro, S.M., et al. 2004. Characterization of BLOC-2, a complex containing the Hermansky-Pudlak syndrome proteins HPS3, HPS5 and HPS6. *Traffic* 5: 276-283.
3. 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.
4. Bossi, G., et al. 2005. Normal lytic granule secretion by cytotoxic T lymphocytes deficient in BLOC-1, -2 and -3 and myosins Va, VIIa and XV. *Traffic* 6: 243-251.
5. SWISS-PROT/TrEMBL (Q86YV9). World Wide Web URL: <http://www.expasy.ch/sprot/sprot-top.html>

## CHROMOSOMAL LOCATION

Genetic locus: HPS6 (human) mapping to 10q24.32.

## SOURCE

HPS-6 (C-12) is a mouse monoclonal antibody raised against amino acids 400-446 mapping within an internal region of HPS-6 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-6 (C-12) is available conjugated to agarose (sc-373786 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-373786 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-373786 PE), fluorescein (sc-373786 FITC), Alexa Fluor® 488 (sc-373786 AF488), Alexa Fluor® 546 (sc-373786 AF546), Alexa Fluor® 594 (sc-373786 AF594) or Alexa Fluor® 647 (sc-373786 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-373786 AF680) or Alexa Fluor® 790 (sc-373786 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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## APPLICATIONS

HPS-6 (C-12) is recommended for detection of HPS-6 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

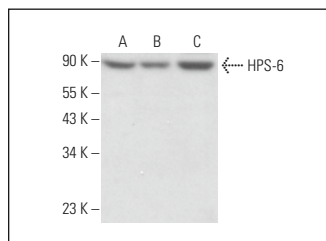
Suitable for use as control antibody for HPS-6 siRNA (h): sc-44422, HPS-6 shRNA Plasmid (h): sc-44422-SH and HPS-6 shRNA (h) Lentiviral Particles: sc-44422-V.

Positive Controls: A-431 whole cell lysate: sc-2201, MCF7 whole cell lysate: sc-2206 or Raji whole cell lysate: sc-364236.

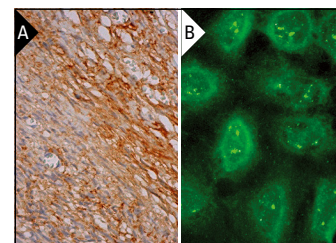
## RECOMMENDED SUPPORT REAGENTS

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. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

## DATA



HPS-6 (C-12): sc-373786. Western blot analysis of HPS-6 expression in Raji (A), A-431 (B) and MCF7 (C) whole cell lysates.



HPS-6 (C-12): sc-373786. Immunoperoxidase staining of formalin fixed, paraffin-embedded human ovary tissue showing cytoplasmic and membrane staining of ovarian stroma cells (A). Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic localization (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.