SZABO SCANDIC

Part of Europa Biosite

Produktinformation

- Forschungsprodukte & Biochemikalien
- Zellkultur & Verbrauchsmaterial
- Diagnostik & molekulare Diagnostik
- Laborgeräte & Service

Weitere Information auf den folgenden Seiten!
See the following pages for more information!

Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

linkedin.com/company/szaboscandic

SANTA CRUZ BIOTECHNOLOGY, INC.

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

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 3 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

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. Anderson, P.D., et al. 2003. Hermansky-Pudlak syndrome type 4 (HPS-4): clinical and molecular characteristics. Hum. Genet. 113: 10-17.
5. 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.
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.

PRODUCT

HPS-4 (h): 293T Lysate represents a lysate of human HPS-4 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

APPLICATIONS

HPS-4 (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive HPS-4 antibodies. Recommended use: 10-20 µl per lane.

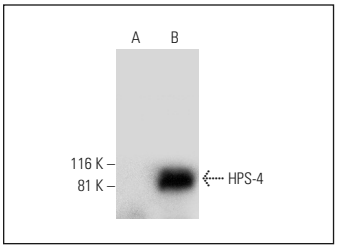
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

HPS-4 (A-6): sc-398070 is recommended as a positive control antibody for Western Blot analysis of enhanced human HPS-4 expression in HPS-4 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

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.

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.

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.