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 (h3): 293T Lysate: sc-128830

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

1. Hirosawa, M., Nagase, T., Murahashi, Y., Kikuno, R. and Ohara, O. 2001. Identification of novel transcribed sequences on human chromosome 22 by expressed sequence tag mapping. DNA Res. 8: 1-9.
2. Suzuki, T., Li, W., Zhang, Q., Karim, A., Novak, E.K., Sviderskaya, E.V., Hill, S.P., Bennett, D.C., Levin, A.V., Nieuwenhuis, H.K., Fong, C.T., Castellan, C., Miterski, B., Swank, R.T. and Spritz, R.A. 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., Boissy, R.E. and Gahl, W.A. 2002. Hermansky-Pudlak syndrome: vesicle formation from yeast to man. Pigment Cell Res. 15: 405-419.
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.

PRODUCT

HPS-4 (h3): 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.

APPLICATIONS

HPS-4 (h3): 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.

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.

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.