# BLOS3 (T-13): sc-167213



The Power to Question

#### **BACKGROUND**

BLOS3, also known as BLOC1S3 (biogenesis of lysosome-related organelles complex 1 subunit 3), is a 202 amino acid protein that belongs to the BLOC1S3 family. BLOS3, along with BLOS1 and BLOS2, are subunits of biogenesis of lysosome-related organelles complex—1 (BLOC1). Localizing to cytoplasm, BLOS3 plays a role in intracellular vesicle trafficking and is required for normal biogenesis of specialized organelles of the endosomal-lysosomal system, such as melanosomes and platelet dense granules. Defects in BLOS3 are the cause of Hermansky-Pudlak syndrome type 8 (HPS8). Hermansky-Pudlak syndrome (HPS) is a genetically heterogeneous and rare autosomal recessive disorder characterized by oculocutaneous albinism, bleeding due to platelet storage pool deficiency and lysosomal storage defects. This syndrome results from defects of diverse cytoplasmic organelles, including melanosomes, platelet dense granules and lysosomes. The BLOS3 gene maps to chromosome 19q13.32.

## REFERENCES

- Starcevic, M., et al. 2004. Identification of snapin and three novel proteins (BLOS1, BLOS2, and BLOS3/reduced pigmentation) as subunits of biogenesis of lysosome-related organelles complex-1 (BLOC-1). J. Biol. Chem. 279: 28393-28401.
- 2. Online Mendelian Inheritance in Man, OMIM™. 2005. Johns Hopkins University, Baltimore, MD. MIM Number: 609762. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
- Morgan, N.V., et al. 2006. A germline mutation in BLOC1S3/reduced pigmentation causes a novel variant of Hermansky-Pudlak syndrome (HPS8). Am. J. Hum. Genet. 78: 160-166.
- 4. Nazarian, R., et al. 2006. Reinvestigation of the dysbindin subunit of BLOC-1 (biogenesis of lysosome-related organelles complex-1) as a dystrobrevin-binding protein. Biochem. J. 395: 587-598.
- Di Pietro, S.M., et al. 2006. BLOC-1 interacts with BLOC-2 and the AP-3 complex to facilitate protein trafficking on endosomes. Mol. Biol. Cell 17: 4027-4038.
- Setty, S.R., et al. 2007. BLOC-1 is required for cargo-specific sorting from vacuolar early endosomes toward lysosome-related organelles. Mol. Biol. Cell 18: 768-780.
- Setty, S.R., et al. 2008. Cell-specific ATP7A transport sustains copper-dependent tyrosinase activity in melanosomes. Nature 454: 1142-1146.
- 8. Cheli, V.T., et al. 2010. Early origin of genes encoding subunits of biogenesis of lysosome-related organelles complex-1, -2 and -3. Traffic 11: 579-586.

## **CHROMOSOMAL LOCATION**

Genetic locus: BLOC1S3 (human) mapping to 19q13.32; Bloc1s3 (mouse) mapping to 7 A3.

## **SOURCE**

BLOS3 (T-13) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of BLOS3 of human origin.

#### **PRODUCT**

Each vial contains 200  $\mu g$  lgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-167213 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## **APPLICATIONS**

BLOS3 (T-13) is recommended for detection of BLOS3 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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); non cross-reactive with BLOS1 or BLOS2.

Suitable for use as control antibody for BLOS3 siRNA (h): sc-97284, BLOS3 siRNA (m): sc-141713, BLOS3 shRNA Plasmid (h): sc-97284-SH, BLOS3 shRNA Plasmid (m): sc-141713-SH, BLOS3 shRNA (h) Lentiviral Particles: sc-97284-V and BLOS3 shRNA (m) Lentiviral Particles: sc-141713-V.

Molecular Weight of BLOS3: 21 kDa.

## **RECOMMENDED SECONDARY REAGENTS**

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

#### **STORAGE**

Store at 4° C, \*\*D0 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 or our catalog for detailed protocols and support products.

Santa Cruz Biotechnology, Inc. 1.800.457.3801 831.457.3801 fax 831.457.3801 Europe +00800 4573 8000 49 6221 4503 0 www.scht.com