



Dysbindin (m): 293T Lysate: sc-119875

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. HPS is a result of defects in various cytoplasmic organelles such as melanosomes, platelet dense granules and lysosomes. The HPS proteins, including HPS-1-6 and Dysbindin (also designated HPS-7), all interact within three distinct, ubiquitously expressed protein complexes or biogenesis of lysosome-related organelle complexes. Defects in the genes encoding for these proteins are the cause of HPS. Dysbindin binds to dystrobrevins in the dystrophin-associated protein complex (DPC) complex. Dysbindin is a cytoplasmic protein. Isoforms 1 and 2 are the result of alternative splicing.

REFERENCES

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CHROMOSOMAL LOCATION

Genetic locus: Dtnbp1 (mouse) mapping to 13 A5.

PRODUCT

Dysbindin (m): 293T Lysate represents a lysate of mouse Dysbindin transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

Dysbindin (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive Dysbindin antibodies. Recommended use: 10-20 µl per lane.

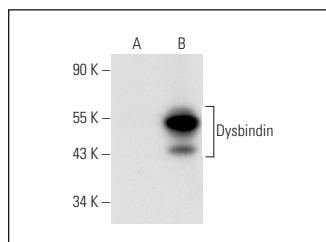
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

Dysbindin (B-5): sc-398872 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse Dysbindin expression in Dysbindin transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

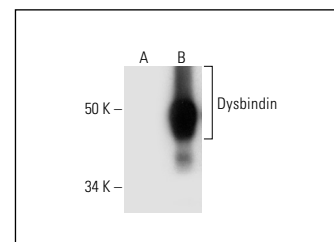
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended:
1) Western Blotting: use m-IgGλ BP-HRP: sc-516132 or m-IgGλ BP-HRP (Cruz Marker): sc-516132-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



Dysbindin (B-5): sc-398872. Western blot analysis of Dysbindin expression in non-transfected: sc-117752 (A) and mouse Dysbindin transfected: sc-119875 (B) 293T whole cell lysates.



Dysbindin (D-8): sc-390626. Western blot analysis of Dysbindin expression in non-transfected: sc-117752 (A) and mouse transfected: sc-119875 (B) 293T whole cell lysates.

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.