Dysbindin (B-5): sc-398872



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. 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 (human) mapping to 6p22.3; Dtnbp1 (mouse) mapping to 13 A5.

SOURCE

Dysbindin (B-5) is a mouse monoclonal antibody raised against amino acids 1-90 mapping at the N-terminus of Dysbindin of human origin.

PRODUCT

Each vial contains 200 μ g IgG $_{2b}$ lambda light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

Dysbindin (B-5) is recommended for detection of Dysbindin isoforms 1 and 2 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], 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).

Suitable for use as control antibody for Dysbindin siRNA (h): sc-60560, Dysbindin siRNA (m): sc-60561, Dysbindin siRNA (r): sc-106988, Dysbindin shRNA Plasmid (h): sc-60560-SH, Dysbindin shRNA Plasmid (m): sc-60561-SH, Dysbindin shRNA Plasmid (r): sc-106988-SH, Dysbindin shRNA (h) Lentiviral Particles: sc-60560-V, Dysbindin shRNA (m) Lentiviral Particles: sc-60561-V and Dysbindin shRNA (r) Lentiviral Particles: sc-106988-V.

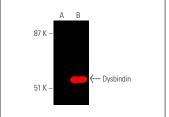
Molecular Weight of Dysbindin: 40-50 kDa.

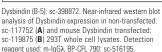
Positive Controls: Dysbindin (m): 293T Lysate: sc-119875.

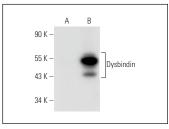
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-lgG λ BP-HRP: sc-516132 or m-lgG λ BP-HRP (Cruz Marker): sc-516132-CM (dilution range: 1:1000-1:10000), Cruz MarkerTM 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-lgG λ BP-FITC: sc-516185 or m-lgG λ BP-PE: sc-516186 (dilution range: 1:50-1:200) with UltraCruz* Mounting Medium: sc-24941 or UltraCruz* Hard-set Mounting Medium: sc-359850.

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.

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.