

## Anti-DTNBP1(Dystrobrevin binding protein 1) Polyclonal Antibody

Cat. #: 60B758

### Description:

DTNBP1(Dystrobrevin binding protein 1) plays a role in the biogenesis of lysosome-related organelles such as platelet dense granule and melanosomes. Dystrobrevin binding protein 1 binds to alpha and beta dystrobrevins that are components of the dystrophin-associated protein complex (DPC). It interacts with pallidin and MUTED. Dystrobrevin binding protein 1 is a part of the biogenesis of lysosome-related organelles complex 1 (BLOC-1). Defects in DTNBP1 are the cause of Hermansky-Pudlak syndrome 7 (HPS7). Hermansky-Pudlak syndrome (HPS) is a genetically heterogeneous, 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. Ceroid storage in the lungs is associated with pulmonary fibrosis, a common cause of premature death in individuals with HPS. Dystrobrevin binding protein 1 belongs to the dysbindin family.

### Immunogen/Specificity:

Polyclonal antibody produced in rabbits immunizing with a synthetic peptide corresponding to N-terminal residues of human DTNBP1(Dystrobrevin binding protein 1)

### References

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Funke,B., et al, Am. J. Hum. Genet. 75 (5), 891-898 (2004)  
Hall,D., et al, Genes Brain Behav. 3 (4), 240-248 (2004)  
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Kirov,G., et al, Biol. Psychiatry 55 (10), 971-975 (2004)

Clone Number:

Isotype:

Species: human, mouse

Storage and Stability: at -20oC

Storage buffer:

This antibody is stored in PBS, 0.01% sodium azide and 50% glycerol.

Preparation:

Purified by antigen-specific affinity chromatography.

Applications :

ELISA

Western Blotting (1µg/ml for 2hrs)