



BL1S3 rabbit pAb

Catalog No	BYab-11764
Isotype	IgG
Reactivity	Human; Mouse
Applications	WB
Gene Name	BLOC1S3 BLOS3
Protein Name	BL1S3
Immunogen	Synthesized peptide derived from human BL1S3 AA range: 142-192
Specificity	This antibody detects endogenous levels of BL1S3 at Human/Mouse
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Dilution	WB 1: 500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	
Cell Pathway	Cytoplasm .
Tissue Specificity	
Function	disease:Defects in BLOC1S3 are the cause of Hermansky-Pudlak syndrome type 8 (HPS8) [MIM:203300]. 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.,function:May play a role in the biogenesis of melanosomes and other specialized organelles of the endosomal-lysosomal system.,PTM:Phosphorylated.,similarity:Belongs to the BLOC1S3 family.,subunit:Component of the biogenesis of lysosome-related organelles (BLOC-1) complex which is composed of BLOC1S1, BLOC1S2,

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DTNBP1, MUTE

Background

This gene encodes a protein that is a component of the BLOC1 multi-subunit protein complex. This complex is necessary for the biogenesis of specialized organelles of the endosomal-lysosomal system, including platelet dense granules and melanosomes. Mutations in this gene cause Hermansky-Pudlak syndrome 8, a disease characterized by lysosomal storage defects, bleeding due to platelet storage pool deficiency, and oculocutaneous albinism. [provided by RefSeq, Jul 2008],

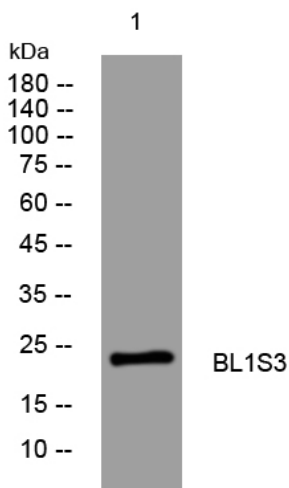
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western blot analysis of lysates from HCT116 cells, primary antibody was diluted at 1:1000, 4° over night