

BL1S3 rabbit pAb

Catalog No :	YT7129
Reactivity :	Human;Mouse
Applications :	WB
Target :	BL1S3
Gene Name :	BLOC1S3 BLOS3
Protein Name :	BL1S3
Human Gene Id :	388552
Human Swiss Prot No :	Q6QNY0
Mouse Gene Id :	232946
Mouse Swiss Prot No :	Q5U5M8
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
Dilution :	WB 1:500-2000
Purification :	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Concentration :	1 mg/ml
Storage Stability :	-15°C to -25°C/1 year(Do not lower than -25°C)

Molecularweight : 22kD

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],

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, DTNBP1, MUTE

Subcellular Location : Cytoplasm .

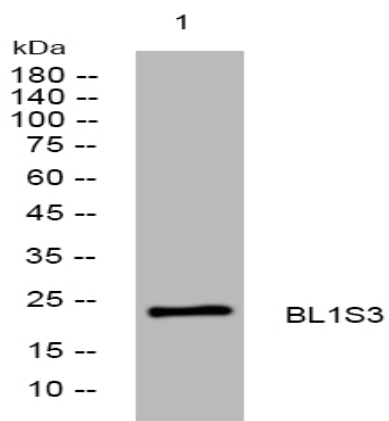
Sort : 2762

No4 : 1

Host : Rabbit

Modifications : Unmodified

Products Images



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