



NPC1 mouse mAb

Catalog No	BYmab-11931
Isotype	IgG
Reactivity	Human; Mouse
Applications	WB
Gene Name	NPC1
Protein Name	NPC1
Immunogen	Synthesized peptide derived from human NPC1 AA range: 1073-1123
Specificity	This antibody detects endogenous levels of NPC1 at Human/Mouse
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	141kD
Cell Pathway	Late endosome membrane ; Multi-pass membrane protein . Lysosome membrane ; Multi-pass membrane protein .
Tissue Specificity	
Function	disease:Defects in NPC1 are the cause of Niemann-Pick disease type C1 (NPC1) [MIM:257220]. NPC1 is an autosomal recessive lipid storage disorder, which affects particularly the brain, liver and spleen, and which is characterized by lysosomal accumulation of low density lipoprotein derived cholesterol. Clinical features include variable hepatosplenomegaly and severe progressive neurological dysfunction such as ataxia, dystonia and dementia. The age of onset can vary from infancy to late adulthood.,disease:Defects in NPC1 are the cause of Niemann-Pick disease type D (NPD) [MIM:257220]; also known as Niemann-Pick disease without sphingomyelinase deficiency, or Nova Scotian type. Because of evidence from biochemical changes, lack of complementation, and linkage mapping to the same chromosome site, NPD and NPC1 are considered to be allelic disorders.,domain:A cysteine-rich N-terminal domain a

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Background

This gene encodes a large protein that resides in the limiting membrane of endosomes and lysosomes and mediates intracellular cholesterol trafficking via binding of cholesterol to its N-terminal domain. It is predicted to have a cytoplasmic C-terminus, 13 transmembrane domains, and 3 large loops in the lumen of the endosome - the last loop being at the N-terminus. This protein transports low-density lipoproteins to late endosomal/lysosomal compartments where they are hydrolyzed and released as free cholesterol. Defects in this gene cause Niemann-Pick type C disease, a rare autosomal recessive neurodegenerative disorder characterized by over accumulation of cholesterol and glycosphingolipids in late endosomal/lysosomal compartments.[provided by RefSeq, Aug 2009],

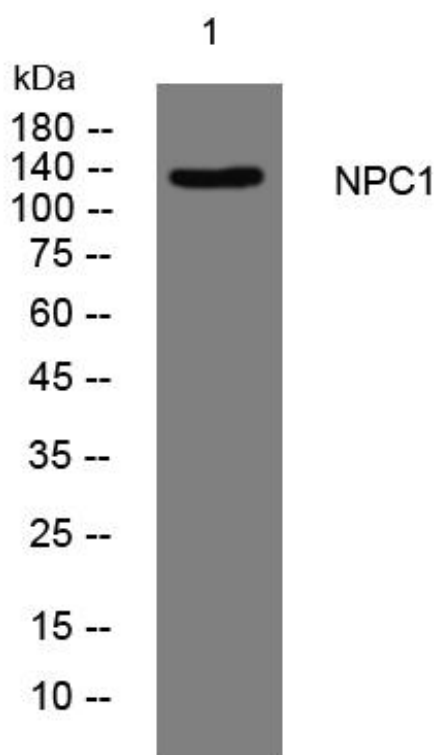
matters needing attention

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 various cells using NPC1 mouse mAb