

Product name:	NPC1 Mouse Monoclonal Antibody
Cat number:	MABN81253
Conjugate:	Unconjugated
Size:	100µL
Clone:	Monoclonal
Concentration:	1mg/ml
Host:	Mouse
Isotype:	Mouse IgG1
Immunogen:	Purified recombinant fragment of human NPC1 (AA: 34-174) expressed in E. Coli.
Reactivity:	Human
Applications:	IHC 1:200-1:1000,ELISA 1:5000-1:20000
Molecular Weight:	142.2kDa
Purification:	Affinity Purification
Form:	Liquid
Buffer:	Purified antibody in PBS with 0.05% sodium azide
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.
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.