

NPC1 Antibody (monoclonal) (M02)

Mouse monoclonal antibody raised against a partial recombinant NPC1. Catalog # AT3083a

Product Information

Application	WB, E
Primary Accession	<u>015118</u>
Other Accession	<u>BC063302</u>
Reactivity	Human
Host	mouse
Clonality	monoclonal
Isotype	IgG2a Kappa
Clone Names	4H2
Calculated MW	142167

Additional Information

Gene ID	4864
Other Names	Niemann-Pick C1 protein, NPC1
Target/Specificity	NPC1 (AAH63302, 151 a.a. ~ 250 a.a) partial recombinant protein with GST tag. MW of the GST tag alone is 26 KDa.
Dilution	WB~~1:500~1000 E~~N/A
Format	Clear, colorless solution in phosphate buffered saline, pH 7.2 .
Storage	Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.
Precautions	NPC1 Antibody (monoclonal) (M02) is for research use only and not for use in diagnostic or therapeutic procedures.

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 hydrolized 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.

References

Variation at the NFATC2 Locus Increases the Risk of Thiazolinedinedione-Induced Edema in the Diabetes REduction Assessment with ramipril and rosiglitazone Medication (DREAM) Study. Bailey SD, et al. Diabetes Care, 2010 Jul 13. PMID 20628086.Evaluating the discriminative power of multi-trait genetic risk scores for type 2 diabetes in a northern Swedish population. Fontaine-Bisson B, et al. Diabetologia, 2010 Oct. PMID 20571754.Epistasis between intracellular cholesterol trafficking-related genes (NPC1 and ABCA1) and Alzheimer's disease risk. Rodr?guez-Rodr?guez E, et al. J Alzheimers Dis, 2010 Jan 1. PMID 20571217.Increased expression of the lysosomal cholesterol transporter NPC1 in Alzheimer's disease. K?gedal K, et al. Biochim Biophys Acta, 2010 Aug. PMID 20497909.Niemann-Pick C1 modulates hepatic triglyceride metabolism and its genetic variation contributes to serum triglyceride levels. Uronen RL, et al. Arterioscler Thromb Vasc Biol, 2010 Aug. PMID 20489167.

Images



Citations

• <u>Genetic and chemical correction of cholesterol accumulation and impaired autophagy in hepatic and neural cells</u> <u>derived from Niemann-Pick Type C patient-specific iPS cells.</u>

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.