Niemann Pick C1 Conjugated Antibody

Catalog No: #C49838

Package Size: #C49838-AF350 100ul #C49838-AF405 100ul #C49838-AF488 100ul #C49838-AF555 100ul #C49838-AF594 100ul #C49838-AF647 100ul #C49838-AF680 100ul #C49838-AF750 100ul #C49838-Biotin 100ul

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Description

Product Name	Niemann Pick C1 Conjugated Antibody
Host Species	Rabbit
Clonality	Monoclonal
Species Reactivity	Hu, Ms, Rt
Immunogen Description	Recombinant protein
Conjugates	Biotin AF350 AF405 AF488 AF555 AF594 AF647 AF680 AF750
Other Names	Niemann Pick C1 protein precursor antibody Niemann Pick disease, type C1 antibody Niemann-Pick C1 protein antibody NPC antibody NPC1 antibody NPC1_HUMAN antibody
Accession No.	Swiss-Prot#:O15118
Uniprot	O15118
GeneID	4864;
Excitation Emission	AF350: 346nm/442nm AF405: 401nm/421nm AF488: 493nm/519nm AF555: 555nm/565nm AF594: 591nm/614nm AF647: 651nm/667nm AF680: 679nm/702nm AF750: 749nm/775nm
Calculated MW	142 kDa
Formulation	0.01M Sodium Phosphate, 0.25M NaCl, pH 7.6, 5mg/ml Bovine Serum Albumin, 0.02% Sodium Azide
Storage	Store at 4°C in dark for 6 months

Application Details

Suggested Dilution:

AF350 conjugated: most applications: 1: 50 - 1: 250

AF405 conjugated: most applications: 1: 50 - 1: 250

AF488 conjugated: most applications: 1: 50 - 1: 250

AF555 conjugated: most applications: 1: 50 - 1: 250

AF594 conjugated: most applications: 1: 50 - 1: 250

AF647 conjugated: most applications: 1: 50 - 1: 250

AF680 conjugated: most applications: 1: 50 - 1: 250

AF750 conjugated: most applications: 1: 50 - 1: 250

Biotin conjugated: working with enzyme-conjugated streptavidin, most applications: 1: 50 - 1: 1,000

Background

Cells obtain cholesterol via two distinct pathways, endogenous synthesis in the endoplasmic reticulum and exogenous uptake through the low-density lipoprotein (LDL) receptor pathway. NPC1 is a protein that resides in late endosomes and lysosomes and is involved in the intracellular trafficking of cholesterol. The human NPC1 gene maps to chromosome 18q11.2 and produces proteins which undergo N-glycosylation and are expressed in brain and liver. NPC1 contains a cysteine-rich domain, which is critical for proper protein function, but is highly mutated. Mutations in NPC1 result in Niemann-Pick disease type C (NPC), an autosomal recessive disease characterized by the accumulation of unesterified cholesterol in the endosomal/lysosomal system. The accumulation of cholesterol results in progressive neurodegeneration and death. More than 90% of cases of NPC are due to mutations in NPC1 and patients with NPC display multiple neurological symptoms, such as hepatosplenomegaly, ataxia, dystonia and dementia.

Note: This product is for in vitro research use only