Human/Mouse/Rat Niemann-Pick Type C1/NPC1 Alexa Fluor® 594-conjugated Antibody

Monoclonal Rabbit IgG_1 Clone # 1318A

Catalog Number: FAB10105T

100 µg

DESCRIPTION

Species Reactivity	Human/Mouse/Rat
Specificity	Detects human Niemann-Pick Type C1/NPC1 in direct ELISAs. Detects human, mouse, and rat Niemann-Pick Type C1/NPC1 in Western blots.
Source	Monoclonal Rabbit IgG_1 Clone # 1318A
Purification	Protein A or G purified from cell culture supernatant
Immunogen	Synthetic peptide containing Human Niemann-Pick Type C1/NPC1
Conjugate	Alexa Fluor 594 Excitation Wavelength: 590 nm Emission Wavelength: 617 nm
Formulation	Supplied 0.2mg/ml in 1X PBS with RDF1 and 0.09% Sodium Azide *Contains <0.1% Sodium Azide, which is not hazardous at this concentration according to GHS classifications. Refer to the Safety Data Sheet (SDS) for additional information and handling instructions.

APPLICATIONS

Please Note: *Optimal dilutions should be determined by each laboratory for each application. General Protocols are available in the Technical Information section on our website.*

Knockout Validated	Optimal dilution of this antibody should be experimentally determined.
Western Blot	Optimal dilution of this antibody should be experimentally determined.
Immunohistochemistry	Optimal dilution of this antibody should be experimentally determined.

PREPARATION AND STORAGE

Shipping	The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below.
Stability & Storage	Protect from light. Do not freeze. 12 months from date of receipt, 2 to 8 °C as supplied

BACKGROUND

NPC intracellular cholesterol transporter 1 or Niemann-Pick C1 protein (NPC1) is a 1,278 aminoacids (aa) intracellular cholesterol transporter which plays an important role in cholesterol transport from the endosomal/lysosomal compartment. NPC1 was identified as the gene that when mutated, results in Niemann-Pick disease, type C, a rare neurovisceral lipid storage disorder resulting from autosomal recessively inherited loss-of-function mutations in either NPC1 or NPC2. This disrupts intracellular lipid transport, leading to the accumulation of lipid products in the late endosomes and lysosomes. Approximately 95% of NPC patients are found to have mutations in the NPC1 gene. In humans, at least one other isoform, missing aa 519-586 is known. Human NPC1 protein sequence is 87% identical to both, mouse and rat NPC1.

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Bio-Techne®
Global | bio-techne.com info@bio-techne.com techsupport@bio-techne.com TEL: 1.612.379.2956
USA | TEL: 800.343.7475 Canada | TEL: 855.668.8722 Europe | Middle East | Africa TEL: +44.0.1235.529449
China | info.cn@bio-techne.com TEL: 400.821.3475