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NAGA Rabbit pAb

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Catalog # AP57346

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P17050
Reactivity	Rat
Predicted	Pig, Dog, Human, Mouse, Rabbit, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	46565
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human NAGA
Epitope Specificity	101-200/411
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Lysosome.
DISEASE	Schindler disease (SCHIND) [MIM:609241]: Form of NAGA deficiency characterized by early-onset neuroaxonal dystrophy and neurological signs (convulsion during fever, epilepsy, psychomotor retardation and hypotonia). NAGA deficiency is typically classified in three main phenotypes: NAGA deficiency type I (Schindler disease or Schindler disease type I) with severe manifestations; NAGA deficiency type II (Kanzazi disease or Schindler disease type II) which is mild; NAGA deficiency type III (Schindler disease type III) characterized by mild-to-moderate neurologic manifestations. NAGA deficiency results in the increased urinary excretion of glycopeptides and oligosaccharides containing alpha-N-acetylgalactosaminyl moieties. Inheritance is autosomal recessive. Note: The disease is caused by mutations affecting the gene represented in this entry. Ref.13 Ref.15 Kanzaki disease (KANZD) [MIM:609242]: Autosomal recessive disorder characterized by late-onset, angiokeratoma corporis diffusum and mild intellectual impairment. Note: The disease is caused by mutations affecting the gene represented in this entry.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	NAGA encodes the lysosomal enzyme alpha-N-acetylgalactosaminidase, which cleaves alpha-N-acetylgalactosaminyl moieties from glycoconjugates. Mutations in NAGA have been identified as the cause of Schindler disease types I and II (type II also known as Kanzaki disease). [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	4668

Other Names	Alpha-N-acetylgalactosaminidase, 3.2.1.49, Alpha-galactosidase B, NAGA (HGNC:7631)
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	NAGA (HGNC:7631)
Function	Removes terminal alpha-N-acetylgalactosamine residues from glycolipids and glycopeptides. Required for the breakdown of glycolipids.
Cellular Location	Lysosome.

Background

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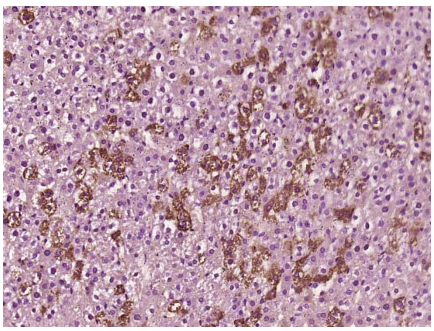
Images



Paraformaldehyde-fixed, paraffin embedded (Rat liver); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (NAGA) Polyclonal Antibody, Unconjugated (AP57346) at 1:500 overnight at 4°C, followed by a conjugated secondary (sp-0023) for 20 minutes and DAB staining.