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NHLRC1 Antibody (Center)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP13383c

Product Information

Application	WB, E
Primary Accession	Q6VVB1
Other Accession	NP_940988.2
Reactivity	Human, Rat, Mouse
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB32737
Calculated MW	42293
Antigen Region	149-179

Additional Information

Gene ID	378884
Other Names	E3 ubiquitin-protein ligase NHLRC1, 632-, Malin, NHL repeat-containing protein 1, NHLRC1, EPM2B
Target/Specificity	This NHLRC1 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 149-179 amino acids from the Central region of human NHLRC1.
Dilution	WB~~1:1000 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.05% (V/V) Proclin 300. This antibody is prepared by Saturated Ammonium Sulfate (SAS) precipitation followed by dialysis against PBS.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	NHLRC1 Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	NHLRC1
Synonyms	EPM2B
Function	E3 ubiquitin-protein ligase. Together with the phosphatase EPM2A/laforin,

	appears to be involved in the clearance of toxic polyglucosan and protein aggregates via multiple pathways. In complex with EPM2A/laforin and HSP70, suppresses the cellular toxicity of misfolded proteins by promoting their degradation through the ubiquitin-proteasome system (UPS). Ubiquitinates the glycogen-targeting protein phosphatase subunits PPP1R3C/PTG and PPP1R3D in a laforin- dependent manner and targets them for proteasome-dependent degradation, thus decreasing glycogen accumulation. Polyubiquitinates EPM2A/laforin and ubiquitinates AGL and targets them for proteasome-dependent degradation. Also promotes proteasome-independent protein degradation through the macroautophagy pathway.
Cellular Location	Endoplasmic reticulum. Nucleus. Note=Localizes at the endoplasmic reticulum and, to a lesser extent, in the nucleus
Tissue Location	Expressed in brain, cerebellum, spinal cord, medulla, heart, liver, skeletal muscle and pancreas

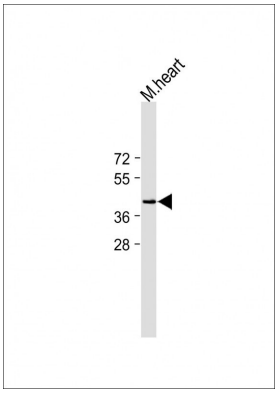
Background

The protein encoded by this gene is a single subunit E3 ubiquitin ligase. Laforin is polyubiquitinated by the encoded protein. Defects in this intronless gene lead to an accumulation of laforin and onset of Lafora disease, also known as progressive myoclonic epilepsy type 2 (EPM2).

References

Moreno, D., et al. Mol. Biol. Cell 21(15):2578-2588(2010)
Rao, S.N., et al. J. Biol. Chem. 285(2):1404-1413(2010)
Traore, M., et al. Neurogenetics 10(4):319-323(2009)
Singh, S., et al. Hum. Mutat. 30(5):715-723(2009)
Vernia, S., et al. PLoS ONE 4 (6), E5907 (2009) :

Images



All lanes : Anti-NHLRC1 Antibody (Center) at 1:500 dilution Lane 1: mouse heart lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated (ASP1615) at 1/15000 dilution. Observed band size : 42kDa Blocking/Dilution buffer: 5% NFDM/TBST.