

Goat Anti-Laforin (isoform a) Antibody

Peptide-affinity purified goat antibody Catalog # AF1610a

Specification

Goat Anti-Laforin (isoform a) Antibody - Product Information

Application WB, IHC, E
Primary Accession O95278

Other Accession <u>NP_005661</u>, <u>7957</u>, <u>13853 (mouse)</u>, <u>680683 (rat)</u>

Reactivity Human

Predicted Mouse, Rat, Dog

Host Goat
Clonality Polyclonal
Concentration 100ug/200ul

Isotype IgG
Calculated MW 37158

Goat Anti-Laforin (isoform a) Antibody - Additional Information

Gene ID 7957

Other Names

Laforin, 3.1.3.-, 3.1.3.16, 3.1.3.48, Glucan phosphatase, Lafora PTPase, LAFPTPase, EPM2A

Dilution

WB~~1:1000 IHC~~1:100~500

E~~N/A

Format

0.5 mg lgG/ml in Tris saline (20mM Tris pH7.3, 150mM NaCl), 0.02% sodium azide, with 0.5% bovine serum albumin

Storage

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions

Goat Anti-Laforin (isoform a) Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Goat Anti-Laforin (isoform a) Antibody - Protein Information

Name EPM2A

Function

Plays an important role in preventing glycogen hyperphosphorylation and the formation of



insoluble aggregates, via its activity as glycogen phosphatase, and by promoting the ubiquitination of proteins involved in glycogen metabolism via its interaction with the E3 ubiquitin ligase NHLRC1/malin. Shows strong phosphatase activity towards complex carbohydrates in vitro, avoiding glycogen hyperphosphorylation which is associated with reduced branching and formation of insoluble aggregates (PubMed:169019012392272925538239255445602523121026231210<a href="http://www.uniprot.org/citat

href="http://www.uniprot.org/citations/11001928" target="_blank">11001928, PubMed:11220751, PubMed:11739371, PubMed:14532330, PubMed:14722920, PubMed:16971387, PubMed:18617530, PubMed:22036712, PubMed:23922729). Has been shown to dephosphorylate MAPT (By similarity). Forms a complex with NHLRC1/malin and HSP70, which suppresses the cellular toxicity of misfolded proteins by promoting their degradation through the ubiquitin-proteasome system (UPS). Acts as a scaffold protein to facilitate PPP1R3C/PTG ubiquitination by NHLRC1/malin (PubMed:23922729, Also

Cellular Location

Cytoplasm. Note=Under glycogenolytic conditions localizes to the nucleus [Isoform 2]: Cytoplasm. Endoplasmic reticulum membrane; Peripheral membrane protein; Cytoplasmic side. Cell membrane. Nucleus. Note=Also found in the nucleus. [Isoform 5]: Cytoplasm. Nucleus

promotes proteasome-independent protein degradation through the macroautophagy pathway (PubMed:20453062).

Tissue Location

Expressed in heart, skeletal muscle, kidney, pancreas and brain. Isoform 4 is also expressed in the placenta

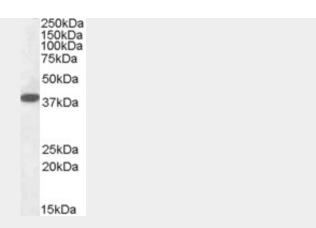
Goat Anti-Laforin (isoform a) Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

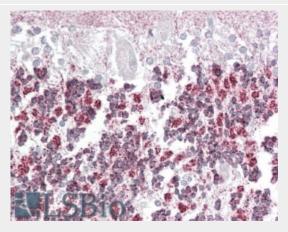
- Western Blot
- Blocking Peptides
- Dot Blot
- <u>Immunohistochemistry</u>
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

Goat Anti-Laforin (isoform a) Antibody - Images

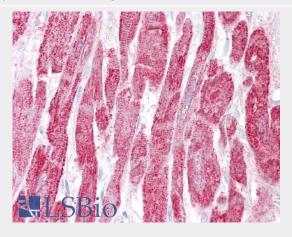




AF1610a (0.1 μ g/ml) staining of Human Cerebellum lysate (35 μ g protein in RIPA buffer). Detected by chemiluminescence.



AF1610a (2.5 μ g/ml) staining of paraffin embedded Human Cerebellum. Steamed antigen retrieval with citrate buffer pH 6, AP-staining.



AF1610a (2.5 μ g/ml) staining of paraffin embedded Human Heart. Steamed antigen retrieval with citrate buffer pH 6, AP-staining.

Goat Anti-Laforin (isoform a) Antibody - Background

This gene encodes a dual-specificity phosphatase that associates with polyribosomes. The encoded protein may be involved in the regulation of glycogen metabolism. Mutations in this gene have been associated with myoclonic epilepsy of Lafora. Alternative splicing results in multiple transcript variants.

Goat Anti-Laforin (isoform a) Antibody - References





Tel: 858.875.1900 Fax: 858.875.1999

Increased endoplasmic reticulum stress and decreased proteasomal function in lafora disease models lacking the phosphatase laforin. Vernia S, et al. PLoS One, 2009 Jun 16. PMID 19529779. Deletions and missense mutations of EPM2A exacerbate unfolded protein response and apoptosis of neuronal cells induced by endoplasm reticulum stress. Liu Y, et al. Hum Mol Genet, 2009 Jul 15. PMID 19403557.

Lafora progressive myoclonus epilepsy: a meta-analysis of reported mutations in the first decade following the discovery of the EPM2A and NHLRC1 genes. Singh S, et al. Hum Mutat, 2009 May. PMID 19267391.

AMP-activated protein kinase phosphorylates R5/PTG, the glycogen targeting subunit of the R5/PTG-protein phosphatase 1 holoenzyme, and accelerates its down-regulation by the laforin-malin complex. Vernia S, et al. J Biol Chem, 2009 Mar 27. PMID 19171932.

The malin-laforin complex suppresses the cellular toxicity of misfolded proteins by promoting their degradation through the ubiquitin-proteasome system. Garyali P, et al. Hum Mol Genet, 2009 Feb 15. PMID 19036738.