

Laforin (k2A3): sc-135810

BACKGROUND

Laforin (Lafora PTPase) is a dual specificity protein phosphatase expressed in heart, skeletal muscle, kidney, pancreas and brain. It belongs to the protein-tyrosine phosphatase family and contains one CBM20 (carbohydrate binding type-20) domain and one tyrosine-protein phosphatase domain. Laforin may be involved in the control of glycogen metabolism, particularly in monitoring for and preventing the formation of poorly branched glycogen molecules (polyglucosans). Laforin isoform 1 is primarily associated with polyribosomes at the endoplasmic reticulum, however, it is also found at the plasma membrane. Isoform 2 can be found in the nucleus. Defects in the EPM2A gene are a cause of progressive myoclonic epilepsy type 2 (EPM2), also known as Lafora disease. EPM2 is an autosomal recessive disease and a severe form of adolescent-onset progressive epilepsy.

REFERENCES

1. Ganesh, S., et al. 2000. Laforin, defective in the progressive myoclonus epilepsy of Lafora type, is a dual-specificity phosphatase associated with polyribosomes. *Hum. Mol. Genet.* 9: 2251-2261.
2. Ganesh, S., et al. 2001. Mutation screening for Japanese Lafora's disease patients: identification of novel sequence variants in the coding and upstream regulatory regions of EPM2A gene. *Mol. Cell. Probes* 15: 281-289.
3. Wang, J., et al. 2002. A unique carbohydrate binding domain targets the Lafora disease phosphatase to glycogen. *J. Biol. Chem.* 277: 2377-2380.
4. Ganesh, S., et al. 2002. Genotype-phenotype correlations for EPM2A mutations in Lafora's progressive myoclonus epilepsy: exon 1 mutations associate with an early-onset cognitive deficit subphenotype. *Hum. Mol. Genet.* 11: 1263-1271.
5. Ganesh, S., et al. 2002. Alternative splicing modulates subcellular localization of Laforin. *Biochem. Biophys. Res. Commun.* 291: 1134-1137.
6. Ki, C.S., et al. 2003. Two novel mutations in the EPM2A gene in a Korean patient with Lafora's progressive myoclonus epilepsy. *J. Hum. Genet.* 48: 51-54.
7. Ganesh, S., et al. 2003. The Lafora disease gene product Laforin interacts with HIRIP5, a phylogenetically conserved protein containing a NifU-like domain. *Hum. Mol. Genet.* 12: 2359-2368.
8. Ianzano, L., et al. 2003. Identification of a novel protein interacting with Laforin, the EPM2a progressive myoclonus epilepsy gene product. *Genomics* 81: 579-587.
9. Girard, J.M., et al. 2006. Molecular characterization of Laforin, a dual-specificity protein phosphatase implicated in Lafora disease. *Biochimie* 88: 1961-1971.

CHROMOSOMAL LOCATION

Genetic locus: EPM2A (human) mapping to 6q24.3.

SOURCE

Laforin (k2A3) is a mouse monoclonal antibody raised against a recombinant protein corresponding to amino acids 243-331 of Laforin of human origin.

PRODUCT

Each vial contains 50 µg IgG₁ in 0.5 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

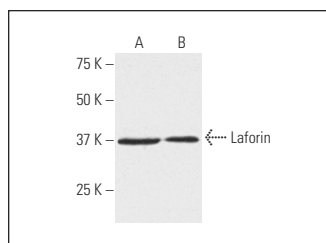
Laforin (k2A3) is recommended for detection of Laforin of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Laforin siRNA (h): sc-75405, Laforin shRNA Plasmid (h): sc-75405-SH and Laforin shRNA (h) Lentiviral Particles: sc-75405-V.

Molecular Weight of Laforin: 38 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200.

DATA



Laforin (k2A3): sc-135810. Western blot analysis of Laforin expression in HeLa (A) and 293T (B) whole cell lysates.

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.