SANTA CRUZ BIOTECHNOLOGY, INC.

EPM2AIP1 (YS-62): sc-100651

BACKGROUND

EPM2AIP1 (EPM2A interacting protein 1), also known as laforin-interacting protein, is the first recognized Laforin binding partner that may play a critical role in discovering the underlying pathogenesis of progressive myoclonic epilepsy type 2 (EPM2), also called Lafora disease (LD). EPM2 is an autosomal recessive disease characterized by grand mal seizures and/or myoclonus at about 15 years of age. Rapid and severe mental deterioration follows, often with psychotic features. Survival is less than ten years after onset. Laforin is the only glycogen phosphatase in mammals that contains a carbohydrate-binding module. Mutations in the glycogen binding domain eliminate the ability of Laforin to dephosphorylate glycogen leading to EPM2 in humans. EPM2AIP1 co-localizes with Laforin to the endoplasmic reticulum. EPM2AIP1 contains two coiled-coil domains and is expressed in heart, brain, placenta, liver, pancreas, kidney and skeletal muscle.

REFERENCES

1. Ianzano, L., Zhao, X.C., Minassian, B.A. and Scherer, S.W. 2003. Identification of a novel protein interacting with laforin, the EPM2A progressive myoclonus epilepsy gene product. Genomics 81: 579-587.
2. Wang, W. and Roach, P.J. 2004. Glycogen and related polysaccharides inhibit the Laforin dual-specificity protein phosphatase. Biochem. Biophys. Res. Commun. 325: 726-730.
3. Chan, E.M., Ackerley, C.A., Lohi, H., Ianzano, L., Cortez, M.A., Shannon, P., Scherer, S.W. and Minassian, B.A. 2004. Laforin preferentially binds the neurotoxic starch-like polyglucosans, which form in its absence in progressive myoclonus epilepsy. Hum. Mol. Genet. 13: 1117-1129.
4. Hitchins, M., Williams, R., Cheong, K., Halani, N., Lin, V.A., Packham, D., Ku, S., Buckle, A., Hawkins, N., Burn, J., Gallinger, S., Goldblatt, J., Kirk, J., Tomlinson, I., Scott, R., Spigelman, A., Suter, C., Martin, D., Suthers, G. and Ward, R. 2005. MLH1 germline epimutations as a factor in hereditary nonpolyposis colorectal cancer. Gastroenterology 129: 1392-1399.
5. Worby, C.A., Gentry, M.S. and Dixon, J.E. 2006. Laforin, a dual specificity phosphatase that dephosphorylates complex carbohydrates. J. Biol. Chem. 281: 30412-30418.
6. Madhavan, D. and Kuzniecky, R.I. 2006. Lafora disease. Rev. Neurol. Dis. 3: 131-135.
7. Tagliabracci, V.S., Turnbull, J., Wang, W., Girard, J.M., Zhao, X., Skurat, A.V., Delgado-Escueta, A.V., Minassian, B.A., Depaoli-Roach, A.A. and Roach, P.J. 2007. Laforin is a glycogen phosphatase, deficiency of which leads to elevated phosphorylation of glycogen *in vivo*. Proc. Natl. Acad. Sci. USA 104: 19262-19266.

CHROMOSOMAL LOCATION

Genetic locus: EPM2AIP1 (human) mapping to 3p22.2; Epm2aip1 (mouse) mapping to 9 F3.

SOURCE

EPM2AIP1 (YS-62) is a mouse monoclonal antibody raised against recombinant EPM2AIP1 of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

EPM2AIP1 (YS-62) is recommended for detection of EPM2AIP1 of mouse, rat and 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 EPM2AIP1 siRNA (h): sc-77976, EPM2AIP1 siRNA (m): sc-144911, EPM2AIP1 shRNA Plasmid (h): sc-77976-SH, EPM2AIP1 shRNA Plasmid (m): sc-144911-SH, EPM2AIP1 shRNA (h) Lentiviral Particles: sc-77976-V and EPM2AIP1 shRNA (m) Lentiviral Particles: sc-144911-V.

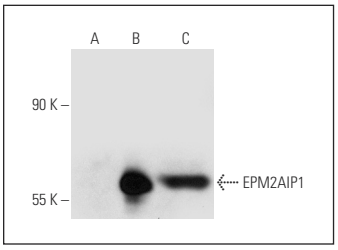
Molecular Weight of EPM2AIP1: 70 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200 or EPM2AIP1 (m): 293T Lysate: sc-120079.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

DATA



EPM2AIP1 (YS-62): sc-100651. Western blot analysis of EPM2AIP1 expression in non-transfected 293T: sc-117752 (**A**), mouse EPM2AIP1 transfected 293T: sc-120079 (**B**) and HeLa (**C**) 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.