



SZABO SCANDIC

Part of Europa Biosite

Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

Weitere Information auf den folgenden Seiten!
See the following pages for more information!



Lieferung & Zahlungsart

siehe unsere [Liefer- und Versandbedingungen](#)

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

[linkedin.com/company/szaboscandic](https://www.linkedin.com/company/szaboscandic) 

EPM2AIP1 (m): 293T Lysate: sc-120079

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* (mouse) mapping to 9 F3.

PRODUCT

EPM2AIP1 (m): 293T Lysate represents a lysate of mouse EPM2AIP1 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

EPM2AIP1 (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive EPM2AIP1 antibodies. Recommended use: 10-20 µl per lane.

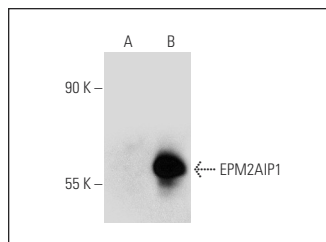
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

EPM2AIP1 (YS-62): sc-100651 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse EPM2AIP1 expression in EPM2AIP1 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

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.

DATA



EPM2AIP1 (YS-62): sc-100651. Western blot analysis of EPM2AIP1 expression in non-transfected: sc-117752 (A) and mouse EPM2AIP1 transfected: sc-120079 (B) 293T whole cell lysates.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. 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.