SANTA CRUZ BIOTECHNOLOGY, INC.

Gigaxonin (F-11): sc-390067

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Gigaxonin, also refered to as giant axonal neuropathy, GAN1, or KLHL16, controls protein degradation and is essential for neuronal function and survival. Gigaxonin is a member of the cytoskeletal BTB/kelch repeat family and influences cytoskeletal organization and dynamics, playing a large role in neurofilament architecture. The amino terminal BTB domain of Gigaxonin binds to the ubiquitin-activating enzyme E1, while the carboxy-terminal kelch repeat domain interacts directly with the light chain of microtubule-associated protein 1B (MAP1B), and tags it for degredation. Overexpression of MAP1B may lead to neuronal cell death, whereas a reduction of MAP1B significantly improves the survival rate of neurons. Mutations in the Gigaxonin gene result in human giant axonal neuropathy (GAN), an autosomal recessive neurodegenerative disorder characterized by axonal degeneration caused by cytoskeletal abnormalities, including accumulated intermediate filaments.

REFERENCES

1. Ding, J., et al. 2002. Microtubule-associated protein 1B: a neuronal binding partner for Gigaxonin. J. Cell Biol. 158: 427-433.
2. Bomont, P., et al. 2003. Identification of seven novel mutations in the GAN gene. Hum. Mutat. 21: 446.
3. Bomont, P. and Koenig, M. 2003. Intermediate filament aggregation in fibroblasts patients is aggravated in non dividing cells and by microtubule destabilization. Hum. Mol. Genet. 12: 813-822.
4. Nakagawa, M. and Takashima, H. 2003. Molecular mechanisms of hereditary neuropathy: genotype-phenotype correlation. Rinsho Byori 51: 536-543.
5. Cullen, V.C. et al. 2004. Gigaxonin is associated with the Golgi and dimerises via its BTB domain. Neuroreport 15: 873-876.
6. Bruno, C., et al. 2004. Clinical and molecular findings in patients with giant axonal neuropathy (GAN). Neurology 62: 13-16.
7. Allen, E., et al. 2005. Gigaxonin-controlled degradation of MAP1B light chain is critical to neuronal survival. Nature 438: 224-228.
8. Wang, W., et al. 2005. Gigaxonin interacts with Tubulin folding cofactor B and controls its degradation through the ubiquitin-proteasome pathway. Curr. Biol. 15: 2050-2055.

CHROMOSOMAL LOCATION

Genetic locus: GAN (human) mapping to 16q23.2; Gan (mouse) mapping to 8 E1.

SOURCE

Gigaxonin (F-11) is a mouse monoclonal antibody raised against amino acids 201-419 mapping within an internal region of Gigaxonin of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

Gigaxonin (F-11) is recommended for detection of Gigaxonin of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Gigaxonin (F-11) is also recommended for detection of Gigaxonin in additional species, including equine.

Suitable for use as control antibody for Gigaxonin siRNA (h): sc-60687, Gigaxonin siRNA (m): sc-60688, Gigaxonin siRNA (r): sc-156074, Gigaxonin shRNA Plasmid (h): sc-60687-SH, Gigaxonin shRNA Plasmid (m): sc-60688-SH, Gigaxonin shRNA Plasmid (r): sc-156074-SH, Gigaxonin shRNA (h) Lentiviral Particles: sc-60687-V, Gigaxonin shRNA (m) Lentiviral Particles: sc-60688-V and Gigaxonin shRNA (r) Lentiviral Particles: sc-156074-V.

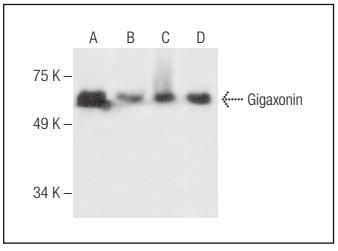
Molecular Weight of Gigaxonin: 68 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, SJRH30 cell lysate: sc-2287 or SH-SY5Y cell lysate: sc-3812.

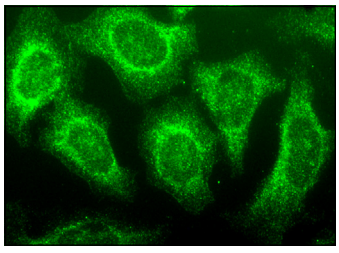
RECOMMENDED SUPPORT REAGENTS

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). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



Gigaxonin (F-11): sc-390067. Western blot analysis of Gigaxonin expression in HeLa (**A**), SJRH30 (**B**) and SH-SY5Y (**C**) whole cell lysates and human hippocampus tissue extract (**D**).



Gigaxonin (F-11): sc-390067. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic localization.

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.