

Gigaxonin (F-3): sc-376173

BACKGROUND

Gigaxonin, also referred 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 degradation. 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.

CHROMOSOMAL LOCATION

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

SOURCE

Gigaxonin (F-3) is a mouse monoclonal antibody raised against amino acids 298-597 mapping at the C-terminus of Gigaxonin of human origin.

PRODUCT

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

Gigaxonin (F-3) is available conjugated to agarose (sc-376173 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-376173 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-376173 PE), fluorescein (sc-376173 FITC), Alexa Fluor® 488 (sc-376173 AF488), Alexa Fluor® 546 (sc-376173 AF546), Alexa Fluor® 594 (sc-376173 AF594) or Alexa Fluor® 647 (sc-376173 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-376173 AF680) or Alexa Fluor® 790 (sc-376173 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Alexa Fluor® is a trademark of Molecular Probes, Inc., Oregon, USA

APPLICATIONS

Gigaxonin (F-3) 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

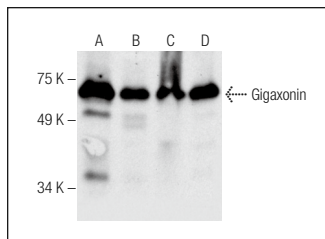
Gigaxonin (F-3) is also recommended for detection of Gigaxonin in additional species, including equine and canine.

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.

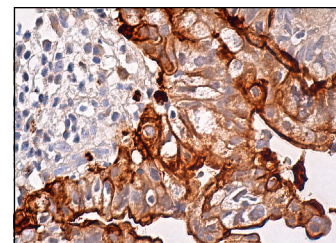
Molecular Weight of Gigaxonin: 68 kDa.

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

DATA



Gigaxonin (F-3): sc-376173. 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-3): sc-376173. Immunoperoxidase staining of formalin fixed, paraffin-embedded human gall bladder tissue showing cytoskeletal staining of glandular cells.

SELECT PRODUCT CITATIONS

1. Tan, H., et al. 2017. HILI destabilizes microtubules by suppressing phosphorylation and Gigaxonin-mediated degradation of TBCB. *Sci. Rep.* 7: 46376.

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.