

Gigaxonin (H-219): sc-292621

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

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CHROMOSOMAL LOCATION

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

SOURCE

Gigaxonin (H-219) is a rabbit polyclonal antibody raised against amino acids 201-419 mapping within an internal region of Gigaxonin of human origin.

RESEARCH USE

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

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

Gigaxonin (H-219) is recommended for detection of Gigaxonin 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)], 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 (H-219) is also recommended for detection of Gigaxonin in additional species, including equine, canine, bovine, porcine and avian.

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.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

STORAGE

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

PROTOCOLS

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