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Product Information

Anti-Gigaxonin (N-terminal)
produced in rabbit, affinity isolated antibody

Product Number **SAB4200104**

Product Description

Anti-Gigaxonin (N-terminal) is produced in rabbit using as the immunogen a synthetic peptide corresponding to a sequence at the N-terminal of human gigaxonin (GeneID 8139), conjugated to KLH. The corresponding sequence is identical in rat and mouse gigaxonin. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-Gigaxonin (N-terminal) specifically recognizes human, rat, and mouse gigaxonin. The antibody can be used in several immunochemical techniques including immunoblotting (~70 kDa), immunoprecipitation, and immunofluorescence. Detection of the gigaxonin band by immunoblotting is specifically inhibited by the gigaxonin immunizing peptide.

Giant axonal neuropathy (GAN) is a severe, autosomal recessive sensorimotor neuropathy affecting both the peripheral and central nervous system. GAN is characterized by aberrant neurofilament accumulation, leading to segmental distension of the axons and generalized disorganization of the cytoskeletal intermediate filaments (IFs).[1,2] GAN is caused by mutations in gigaxonin (also known as GAN, GAN1, Kelch-like protein 16, KLHL16), a distant member of the BTB/kelch superfamily.[1]

Gigaxonin is an ubiquitously expressed protein, composed of an N-terminal BTB domain followed by six kelch repeats. It plays an important role in cytoskeletal functions and dynamics by directing ubiquitin-mediated degradation of cytoskeletal proteins, MAP1B, MAP8, and the tubulin-cofactor B (TBCB), via the Cul3-E3-ubiquitin-ligase family.[3-5] Gigaxonin co-localizes with MAP1B in neurons. It binds directly with the light-chain (LC) of MAP1B to enhance microtubule stability in neurons that is required for axonal transport. Gigaxonin controlled degradation of MAP1B is essential for neuronal function and survival.[5] Gene knockout of GAN in mice has been shown to cause toxic accumulation of MAP8, alterations of microtubule network, and impaired retrograde axonal transport, leading to neuronal death.[6]

Reagent

Supplied as a solution in 0.01 M phosphate buffered saline, pH 7.4, containing 15 mM sodium azide as a preservative.

Antibody concentration: ~1.5 mg/mL

Precautions and Disclaimer

For R&D use only. Not for drug, household, or other uses. Please consult the Safety Data Sheet for information regarding hazards and safe handling practices.

Storage/Stability

Store at –20 °C. For continuous use, the product may be stored at 2–8 °C for up to one month. For extended storage, freeze in working aliquots. Repeated freezing and thawing, or storage in “frost-free” freezers, is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilutions should be discarded if not used within 12 hours.

Product Profile

Immunoblotting: a working antibody concentration of 1-2 μg/mL is recommended using mouse brain extracts (S1 fraction), and 1.5-3 μg/mL using SH-SY5Y cell lysates.

Immunoprecipitation: a working antibody amount of 15-30 μg is recommended using rat brain extracts (S1 fraction).

Immunofluorescence: a working antibody concentration of 10-20 μg/mL is recommended using HeLa cells expressing human gigaxonin.

Note: In order to obtain best results in various techniques and preparations, it is recommended to determine optimal working dilutions by titration.

References

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5. Allen, E., et al., *Nature,* **438**, 224-228 (2005).
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