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Gigaxonin Polyclonal Antibody
Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP54346

Specification

Gigaxonin Polyclonal Antibody - Product Information

Application	**IHC-P, IHC-F, IF, ICC, E**
Primary Accession	Q9H2C0
Reactivity	**Rat, Pig, Bovine**
Host	**Rabbit**
Clonality	**Polyclonal**
Calculated MW	**68 KDa**
Physical State	**Liquid**
Immunogen	**KLH conjugated synthetic peptide derived from human Gigaxonin**
Epitope Specificity	**351-450/597**
Isotype	**IgG**
Purity	affinity purified by Protein A
Buffer	**0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.**
SUBCELLULAR LOCATION	**Cytoplasmic; Cytoskeleton.**
SIMILARITY	**Contains 1 BACK (BTB/Kelch associated) domain.Contains 1 BTB (POZ) domain. Contains 6 Kelch repeats.**
SUBUNIT	**Interacts with TBCB. Interacts with CUL3. Part of a complex that contains CUL3, RBX1 and GAN. Interacts (via BTB domain) with UBA1. Interacts (via Kelch domains) with MAP1B (via C-terminus) and MAP1S (via C-terminus).**
Post-translational modifications	**Ubiquitinated by E3 ubiquitin ligase complex formed by CUL3 and RBX1 and probably targeted for proteasome-independent degradation.**
DISEASE	**Defects in GAN are the cause of giant axonal neuropathy (GAN) [MIM:256850]. GAN is a severe autosomal recessive sensorimotor neuropathy affecting both the peripheral nerves and the central nervous system. It is characterized by neurofilament accumulation, leading to segmental distention of axons.**
Important Note	**This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.**

Background Descriptions
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.

Gigaxonin Polyclonal Antibody - Additional Information

Gene ID 8139

Other Names
Gigaxonin, Kelch-like protein 16, GAN, GAN1, KLHL16

Target/Specificity
Expressed in brain, heart and muscle.

Dilution
IHC-P~~N/A<br \>IHC-F~~N/A<br \>IF~~1:50~200<br \>ICC~~N/A<br \>E~~N/A

Storage
Store at -20 ℃ for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ℃.

Gigaxonin Polyclonal Antibody - Protein Information

Name GAN

Synonyms GAN1, KLHL16

Function
Probable cytoskeletal component that directly or indirectly plays an important role in neurofilament architecture. May act as a substrate-specific adapter of an E3 ubiquitin-protein ligase complex which mediates the ubiquitination and subsequent proteasomal degradation of target proteins. Controls degradation of TBCB. Controls degradation of MAP1B and MAP1S, and is critical for neuronal maintenance and survival.

Cellular Location
Cytoplasm. Cytoplasm, cytoskeleton.

Tissue Location
Expressed in brain, heart and muscle.

Gigaxonin Polyclonal Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

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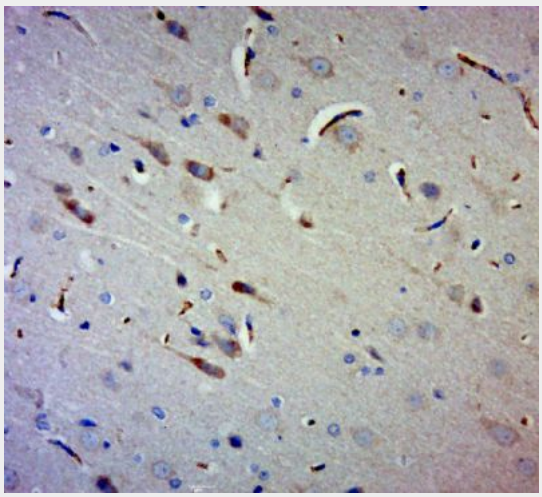
- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

Gigaxonin Polyclonal Antibody - Images



Paraformaldehyde-fixed, paraffin embedded (rat brain tissue); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (GAN) Polyclonal Antibody, Unconjugated (bs-11025R) at 1:400 overnight at 4°C, followed by a conjugated secondary (sp-0023) for 20 minutes and DAB staining.