

Gigaxonin Rabbit pAb

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Catalog # AP54346

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q9H2C0
Reactivity	Rat
Predicted	Pig, Human, Mouse, Rabbit, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	67638
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.
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 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.

Additional Information

Gene ID	8139
Other Names	Gigaxonin, Kelch-like protein 16, GAN, GAN1, KLHL16

Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C 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 °C.

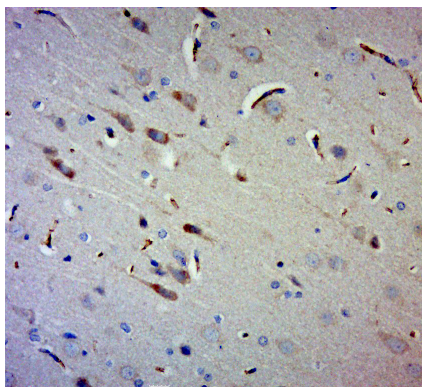
Protein Information

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.

Background

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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 (AP54346) at 1:400 overnight at 4°C, followed by a conjugated secondary (sp-0023) for 20 minutes and DAB staining.

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