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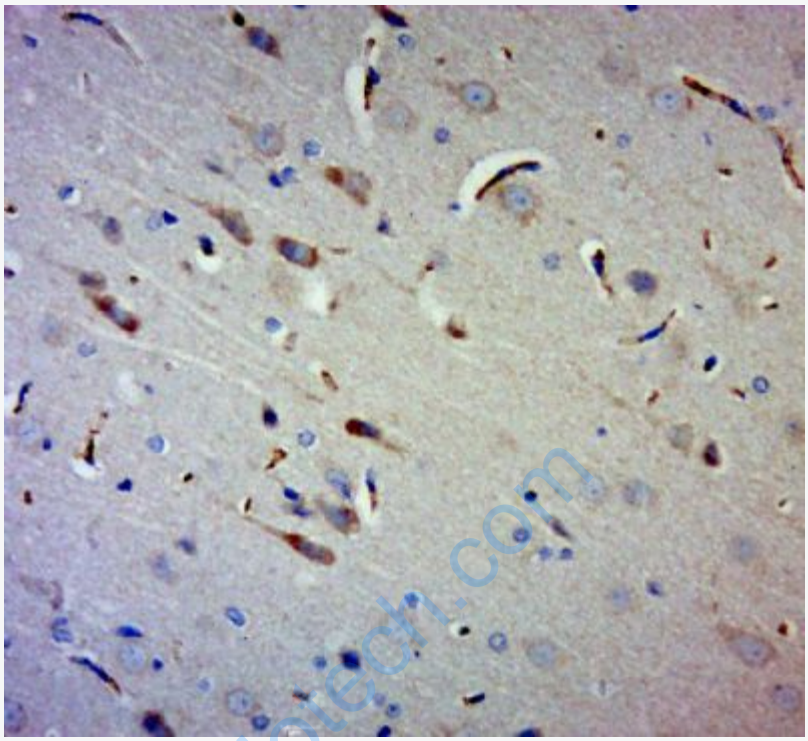
Rabbit Anti-Gigaxonin antibody

SL11025R

Product Name:	Gigaxonin
Chinese Name:	巨轴索神经病蛋白GAN抗体
Alias:	FLJ38059; GAN; GAN1; Kelch-like protein 16; giant axonal neuropathy; KLHL16; GAN_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Pig,Cow,Horse,Rabbit,Sheep,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	68kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Gigaxonin:351-450/597
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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. **Function:** Mutations in gigaxonin result in a sensory and motor neuropathy called Giant Axonal Neuropathy (GAN). Giant axonal neuropathy, a severe autosomal recessive sensorineural neuropathy affecting both the peripheral nerves and the central nervous system, is characterized by neurofilament accumulation, leading to segmental distention of axons. Gigaxonin is a member of the cytoskeletal BTB/kelch (Broad-Complex, Tramtrack and Bric a brac) repeat family. Gigaxonin contains an N-terminal BTB domain followed by 6 kelch repeats, which were predicted to adopt a beta-propeller shape. Gigaxonin controls protein degradation and is essential for neuronal function and survival. 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 **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). **Subcellular Location:** Cytoplasmic; Cytoskeleton. **Tissue Specificity:** Expressed in brain, heart and muscle. **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. **Similarity:** Contains 1 BACK (BTB/Kelch associated) domain. Contains 1 BTB (POZ) domain. Contains 6 Kelch repeats.

	SWISS: Q9H2C0 **Gene ID:** 8139 **Database links:** Entrez Gene: 8139Human Entrez Gene: 209239Mouse Omim: 605379Human SwissProt: Q9H2C0Human SwissProt: Q8CA72Mouse Unigene: 112569Human Unigene: 132992Mouse **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Picture:	 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 (SL11025R) at 1:400 overnight at 4°C, followed by a conjugated secondary (sp-0023) for 20 minutes and DAB staining.