## bs-11025R

# [ Primary Antibody ]

# Gigaxonin Rabbit pAb

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DATASHEET -

Host: Rabbit Isotype: IgG

Clonality: Polyclonal

**GenelD:** 8139 SWISS: Q9H2C0

Target: Gigaxonin

Immunogen: KLH conjugated synthetic peptide derived from human Gigaxonin:

351-450/597.

**Purification:** affinity purified by Protein A

Concentration: 1mg/ml

Storage: 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50%

Glycerol.

Shipped at 4°C. Store at -20°C for one year. Avoid repeated

freeze/thaw cycles.

Background: 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.

**Applications: IHC-P** (1:100-500)

IHC-F (1:100-500) **IF** (1:100-500)

Reactivity: Rat (predicted: Human,

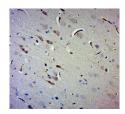
Mouse, Rabbit, Pig, Sheep,

Cow, Horse)

Predicted MW.: 68 kDa

Subcellular Cytoplasm Location:

### VALIDATION 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.