



# **GDAP1** Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab) **Catalog # AP54610** 

### **Specification**

# **GDAP1 Polyclonal Antibody - Product Information**

Application **Primary Accession** Reactivity Host Clonality Calculated MW **Physical State** Immunogen

**Epitope Specificity** Isotype **Purity** affinity purified by Protein A

Buffer

SUBCELLULAR LOCATION

**SIMILARITY** 

**SUBUNIT** DISEASE

WB, IHC-P, IHC-F, IF, ICC, E **08TB36** Rat, Pig, Dog, Bovine **Rabbit Polyclonal 41 KDa** Liquid

KLH conjugated synthetic peptide derived from human GDAP1 151-230/358

laG

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

Mitochondrion outer membrane; Multi-pass membrane protein. Cytoplasm (By

Belongs to the GST superfamily, Contains 1 GST C-terminal domain. Contains 1 GST

N-terminal domain.

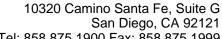
Homodimer.

Defects in GDAP1 are the cause of Charcot-Marie-Tooth disease type 4A (CMT4A) [MIM:214400]. CMT4A is a form of Charcot-Marie-Tooth disease. the most common inherited disorder of the

peripheral nervous system.

Charcot-Marie-Tooth disease is classified in two main groups on the basis of electrophysiologic properties and histopathology: primary peripheral demyelinating neuropathy or CMT1, and primary peripheral axonal neuropathy or CMT2. Demyelinating CMT neuropathies are characterized by severely reduced nerve conduction velocities (less than 38 m/sec), segmental demyelination and remyelination with onion bulb formations on nerve biopsy, slowly progressive distal muscle atrophy and weakness, absent deep tendon reflexes, and hollow feet.

Autosomal recessive forms of demyelinating Charcot-Marie-Tooth





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disease are by convention designated CMT4. CMT4A is a severe form characterized by early age of onset and rapid progression leading to inability to walk in late childhood or adolescence. Defects in GDAP1 are the cause of Charcot-Marie-Tooth disease axonal recessive with vocal cord paresis (CMT2RV) [MIM:607706]. CMT2RV is a form of **Charcot-Marie-Tooth disease characterized** by the association of axonal neuropathy with vocal cord paresis. Defects in GDAP1 are the cause of Charcot-Marie-Tooth disease type 2K (CMT2K) [MIM:607831]. CMT2K is an axonal form of **Charcot-Marie-Tooth disease. Axonal CMT** neuropathies are characterized by signs of axonal regeneration in the absence of obvious myelin alterations, normal or slightly reduced nerve conduction velocities, and progressive distal muscle weakness and atrophy. CMT2K onset is in early childhood (younger than 3 years). This phenotype is characterized by foot deformities, kyphoscoliosis, distal limb muscle weakness and atrophy, areflexia, and diminished sensation in the lower limbs. Weakness in the upper limbs is observed in the first decade, with clawing of the fingers. Inheritance can be autosomal dominant or recessive. Defects in GDAP1 are the cause of Charcot-Marie-Tooth disease recessive intermediate type A (CMTRIA) [MIM:608340]. CMTRIA is a form of Charcot-Marie-Tooth disease characterized by clinical and pathologic features intermediate between demyelinating and axonal peripheral neuropathies, and motor median nerve conduction velocities ranging from 25 to 45 m/sec. This product as supplied is intended for research use only, not for use in human. therapeutic or diagnostic applications.

Important Note

# **Background Descriptions**

Glutathione S-transferases (GSTs) function to conjugate reduced glutathione to many exogenous and endogenous hydrophobic electrophiles. Although it shares the carboxy and amino-terminal glutathione S-transferase domains, GDAP1 is characterized as a GST-like protein because it contains an extended GST domain II and a predicted transmembrane domain, two characteristics which are unusual for GST family members. GDAP1 may function in a signal transduction pathway that is responsible for ganglioside-induced neurite differentiation and also may play a role in protecting myelin membranes from free-radical damage. Mutations in the gene encoding GDAP1 is the cause of many forms of Charcot-Marie-Tooth disease, a common inherited disorder of the peripheral nervous system that is characterized by reduced nerve conduction velocities, slow progressive distal muscle atrophy and absent deep tendon reflexes.



# **GDAP1** Polyclonal Antibody - Additional Information

#### Gene ID 54332

### **Other Names**

Ganglioside-induced differentiation-associated protein 1, GDAP1, GDAP1

### **Target/Specificity**

Highly expressed in whole brain and spinal cord. Predominant expression in central tissues of the nervous system not only in neurons but also in Schwann cells.

#### Dilution

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<span class ="dilution_WB">WB~~1:1000</span><br \><span class
="dilution_IHC-P">IHC-P~~N/A</span><br \><span class
="dilution_IHC-F">IHC-F~~N/A</span><br \><span class
="dilution_IF">IF~~1:50~200</span><br \><span class ="dilution_ICC">ICC~~N/A</span><br \><span class ="dilution_E">E~~N/A</span>
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#### 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.

# **GDAP1 Polyclonal Antibody - Protein Information**

#### Name GDAP1

### **Function**

Regulates the mitochondrial network by promoting mitochondrial fission.

### **Cellular Location**

Mitochondrion outer membrane; Multi-pass membrane protein. Cytoplasm {ECO:0000250|UniProtKB:088741}

### **Tissue Location**

Highly expressed in whole brain and spinal cord. Predominant expression in central tissues of the nervous system not only in neurons but also in Schwann cells

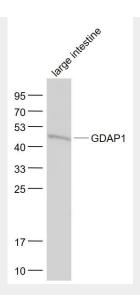
### **GDAP1 Polyclonal Antibody - Protocols**

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

- Western Blot
- Blocking Peptides
- Dot Blot
- <u>Immunohistochemistry</u>
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

# GDAP1 Polyclonal Antibody - Images





# Sample:

Large intestine (Mouse) Lysate at 40 ug

Primary: Anti- GDAP1 (bs-11725R) at 1/1000 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 41 kD Observed band size: 42 kD