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GDAP1 Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP54610

Product Information

Application WB, IHC-P, IHC-F, IF, ICC, E

Primary Accession Q8TB36

Reactivity Rat, Pig, Dog, Bovine

Host Rabbit
Clonality Polyclonal
Calculated MW 41346
Physical State Liquid

Immunogen KLH conjugated synthetic peptide derived from human GDAP1

Epitope Specificity 151-230/358

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 Mitochondrion outer membrane; Multi-pass membrane protein. Cytoplasm

(By similarity).

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

1 GST N-terminal domain.

SUBUNIT Homodimer.

DISEASE 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 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.

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.

Additional Information

Important Note

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 WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC=1:100-500,IF=1:100-50

0,ELISA=1:5000-10000

Format 0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce

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 GDAP1

Function Regulates the mitochondrial network by promoting mitochondrial fission.

Cellular Location Mitochondrion outer membrane; Multi-pass membrane protein. Cytoplasm

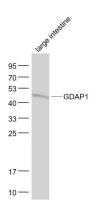
{ECO:0000250|UniProtKB:088741}

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

Images



Sample:

Large intestine (Mouse) Lysate at 40 ug Primary: Anti- GDAP1 (AP54610) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

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

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