10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

ANT-1 Rabbit pAb

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Catalog # AP58511

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P12235
Reactivity	Human
Predicted	Rat, Pig, Dog, Mouse, Rabbit, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	33064
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ANT-1
Epitope Specificity	31-130/298
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 inner membrane; Multi-pass membrane protein.
DISEASE	Defects in SLC25A4 are a cause of progressive external ophthalmoplegia with mitochondrial DNA deletions autosomal dominant type 2 (PEOA2) [MIM:609283]. Progressive external ophthalmoplegia is characterized by progressive weakness of ocular muscles and levator muscle of the upper eyelid. In a minority of cases, it is associated with skeletal myopathy, which predominantly involves axial or proximal muscles and which causes abnormal fatigability and even permanent muscle weakness. Ragged-red fibers and atrophy are found on muscle biopsy. A large proportion of chronic ophthalmoplegias are associated with other symptoms, leading to a multisystemic pattern of this disease. Additional symptoms are variable, and may include cataracts, hearing loss, sensory axonal neuropathy, ataxia, depression, hypogonadism, and parkinsonism.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Defects in SLC25A4 are a cause of progressive external ophthalmoplegia with mitochondrial DNA deletions autosomal dominant type 2 (PEOA2) [MIM:609283]. Progressive external ophthalmoplegia is characterized by progressive weakness of ocular muscles and levator muscle of the upper eyelid. In a minority of cases, it is associated with skeletal myopathy, which predominantly involves axial or proximal muscles and which causes abnormal fatigability and even permanent muscle weakness. Ragged-red fibers and atrophy are found on muscle biopsy. A large proportion of chronic ophthalmoplegias are associated with other symptoms, leading to a multisystemic pattern of this disease. Additional symptoms are variable, and may include cataracts, hearing loss, sensory axonal neuropathy, ataxia, depression, hypogonadism, and parkinsonism.

Additional Information

Gene ID	291
Other Names	ADP/ATP translocase 1, ADP, ATP carrier protein 1, SLC25A4 {ECO:0000303|PubMed:25732997, ECO:0000312|HGNC:HGNC:10990}
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.

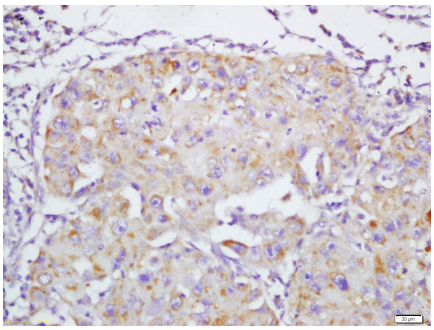
Protein Information

Name	SLC25A4 {ECO:0000303|PubMed:25732997, ECO:0000312|HGNC:HGNC:10990}
Function	ADP:ATP antiporter that mediates import of ADP into the mitochondrial matrix for ATP synthesis, and export of ATP out to fuel the cell (PubMed:21586654, PubMed:27693233, PubMed:23173940, PubMed:30046662). Cycles between the cytoplasmic-open state (c-state) and the matrix-open state (m-state): operates by the alternating access mechanism with a single substrate-binding site intermittently exposed to either the cytosolic (c-state) or matrix (m-state) side of the inner mitochondrial membrane (By similarity). Substrate exchange across the membrane occurs consecutively with one substrate being transported first, then dissociating from the substrate binding site before the second substrate binds for transport in the opposite direction (PubMed:37278158). In addition to its ADP:ATP antiporter activity, also involved in mitochondrial uncoupling and mitochondrial permeability transition pore (mPTP) activity (PubMed:31883789). Plays a role in mitochondrial uncoupling by acting as a proton transporter: proton transport uncouples the proton flows via the electron transport chain and ATP synthase to reduce the efficiency of ATP production and cause mitochondrial thermogenesis (By similarity). Proton transporter activity is inhibited by ADP:ATP antiporter activity, suggesting that SLC25A4/ANT1 acts as a master regulator of mitochondrial energy output by maintaining a delicate balance between ATP production (ADP:ATP antiporter activity) and thermogenesis (proton transporter activity) (By similarity). Proton transporter activity requires free fatty acids as cofactor, but does not transport it (By similarity). Also plays a key role in mPTP opening, a non-specific pore that enables free passage of the mitochondrial membranes to solutes of up to 1.5 kDa, and which contributes to cell death (PubMed:31883789). It is however unclear if SLC25A4/ANT1 constitutes a pore-forming component of mPTP or regulates it (By similarity). Acts as a regulator of mitophagy independently of ADP:ATP antiporter activity: promotes mitophagy via interaction with TIMM44, leading to inhibit the presequence translocase TIMM23, thereby promoting stabilization of PINK1 (By similarity).
Cellular Location	Mitochondrion inner membrane; Multi-pass membrane protein. Membrane; Multi-pass membrane protein. Note=The complex formed with ARL2BP, ARL2 and SLC25A4/ANT1 is expressed in mitochondria (By similarity). May localize to non-mitochondrial membranes (PubMed:27641616) {ECO:0000250|UniProtKB:P48962, ECO:0000269|PubMed:27641616}
Tissue Location	Expressed in erythrocytes (at protein level).

Background

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Images



Tissue/cell: human lung carcinoma; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min; Incubation: Anti-ANT-1 Polyclonal Antibody, Unconjugated(AP58511) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.