

ATP13A2 Rabbit pAb

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Product Information

Application IHC-P, IHC-F, IF

Primary Accession Q9NQ11
Reactivity Rat

Predicted Human, Mouse, Pig, Horse, Rabbit

Host Rabbit
Clonality Polyclonal
Calculated MW 128794
Physical State Liquid

Immunogen KLH conjugated synthetic peptide derived from human ATP13A2

Epitope Specificity 1001-1080/1180

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

SIMILARITY

Membrane; Multi-pass membrane protein (By similarity). Lysosome. Belongs to the cation transport ATPase (P-type) (TC 3.A.3) family. Type V

subfamily.

DISEASE Defects in ATP13A2 are the cause of Kufor-Rakeb syndrome (KRS)

[MIM:606693]; also known as Parkinson disease type 9 (PARK9). KRS is a rare

hereditary disease with juvenile onset. In addition to typical signs of

Parkinson disease, affected individuals show symptoms of more widespread

neurodegeneration, including dementia.

Important Note This product as supplied is intended for research use only, not for use in

human, therapeutic or diagnostic applications.

Background Descriptions ATP13A2 is a 1,180 amino acid multi-pass membrane protein that belongs to

the P5 subfamily of ATPases which play an important role in the

transportation of inorganic cations. Expressed as multiple alternative spliced isoforms, ATP13A2 functions to catalyze the conversion of ATP to ADP and a free phosphate, thereby participating in the active transport of ions across cellular membranes. Defects in the gene encoding ATP13A2 are the cause of Kufor-Rakeb syndrome (KRS), a rare hereditary type of Parkinson's disease that exhibits juvenile onset and is characterized by neurodegeneration and dementia. The ATP13A2 gene maps to human chromosome 1, which spans 260 million base pairs, contains over 3,000 genes and comprises nearly 8% of

the human genome.

Additional Information

Gene ID 23400

Other Names Polyamine-transporting ATPase 13A2, 7.6.2.-, ATP13A2 (HGNC:30213)

Target/Specificity Expressed in brain; protein levels are markedly increased in brain from

subjects with Parkinson disease and subjects with dementia with Lewy bodies. Detected in pyramidal neurons located throughout the cingulate cortex (at protein level). In the substantia nigra, it is found in neuromelanin-positive

dopaminergic neurons (at protein level).

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 ATP13A2 (<u>HGNC:30213</u>)

Function ATPase which acts as a lysosomal polyamine exporter with high affinity for

spermine (PubMed:<u>31996848</u>). Also stimulates cellular uptake of polyamines and protects against polyamine toxicity (PubMed:<u>31996848</u>). Plays a role in intracellular cation homeostasis and the maintenance of neuronal integrity

(PubMed:22186024). Contributes to cellular zinc homeostasis

(PubMed:<u>24603074</u>). Confers cellular protection against Mn(2+) and Zn(2+) toxicity and mitochondrial stress (PubMed:<u>26134396</u>). Required for proper

lysosomal and mitochondrial maintenance (PubMed: 22296644,

PubMed:<u>28137957</u>). Regulates the autophagy-lysosome pathway through the control of SYT11 expression at both transcriptional and post-translational levels (PubMed:<u>27278822</u>). Facilitates recruitment of deacetylase HDAC6 to lysosomes to deacetylate CTTN, leading to actin polymerization, promotion of

autophagosome-lysosome fusion and completion of autophagy

(PubMed:30538141). Promotes secretion of exosomes as well as secretion of SCNA via exosomes (PubMed:24603074, PubMed:25392495). Plays a role in

lipid homeostasis (PubMed:31132336).

Cellular Location Lysosome membrane; Multi-pass membrane protein. Late endosome

membrane; Multi-pass membrane protein. Endosome, multivesicular body

membrane; Multi-pass membrane protein. Cytoplasmic vesicle, autophagosome membrane; Multi-pass membrane protein

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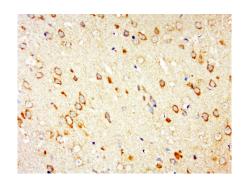
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Images



Paraformaldehyde-fixed, paraffin embedded (rat brain); 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 (ATP13A2) Polyclonal Antibody, Unconjugated (AP54602) at 1:400 overnight at 4°C, followed by a conjugated secondary (sp-0023) for 20 minutes and DAB staining.

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