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SLC25A13 Rabbit pAb

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Catalog # AP58119

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q9UJS0
Predicted	Rat, Pig, Dog, Human, Mouse, Chicken, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	74176
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human SLC25A13
Epitope Specificity	351-450/675
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 SLC25A13 are the cause of citrullinemia type 2 (CTLN2) [MIM:603471]. Citrullinemia belongs to the urea cycle disorders. It is an autosomal recessive disease characterized primarily by elevated serum and urine citrulline levels. Ammonia intoxication is another manifestation. CTLN2 is characterized by neuropsychiatric symptoms including abnormal behaviors, loss of memory, seizures and coma. Death can result from brain edema. Onset is sudden and usually between the ages of 20 and 50 years. Defects in SLC25A13 are the cause of neonatal intrahepatic cholestasis due to citrin deficiency (NICCD) [MIM:605814]. NICCD is a form of citrullinemia type 2 with neonatal onset. NICCD is characterized by suppression of the bile flow, hepatic fibrosis, low birth weight, growth retardation, hypoproteinemia, variable liver dysfunction. NICCD is generally not severe and symptoms disappear by one year of age with an appropriate diet. Years or even decades later, however, some individuals develop the characteristic features of citrullinemia type 2 with neuropsychiatric symptoms.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	SLC25A13 is a member of the mitochondrial carrier family. It contains four EF-hand Ca(2+) binding motifs in the N-terminal domain, and localizes to mitochondria. It catalyzes the exchange of aspartate for glutamate and a proton across the inner mitochondrial membrane, and is stimulated by calcium on the external side of the inner mitochondrial membrane. Mutations in the SLC25A13 gene result in citrullinemia, type II. Multiple transcript variants encoding different isoforms have been found for this gene.

Additional Information

Gene ID	10165

Other Names	Electrogenic aspartate/glutamate antiporter SLC25A13, mitochondrial, Calcium-binding mitochondrial carrier protein Aralar2, ARALAR-related gene 2, ARALAR2, Citrin, Mitochondrial aspartate glutamate carrier 2, Solute carrier family 25 member 13 {ECO:0000312|HGNC:HGNC:10983}, SLC25A13 (HGNC:10983)
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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	SLC25A13 (HGNC:10983)
Function	Mitochondrial electrogenic aspartate/glutamate antiporter that favors efflux of aspartate and entry of glutamate and proton within the mitochondria as part of the malate-aspartate shuttle (PubMed:11566871, PubMed:38937634, PubMed:38945283, PubMed:39419476). 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:38937634). Also mediates the uptake of L- cysteinesulfinate (3-sulfino-L-alanine) by mitochondria in exchange for L-glutamate and proton (PubMed:11566871). Can also exchange L- cysteinesulfinate with aspartate in their anionic form without any proton translocation (PubMed:11566871). Lacks transport activity towards gamma-aminobutyric acid (GABA) (PubMed:38945283).
Cellular Location	Mitochondrion inner membrane; Multi-pass membrane protein
Tissue Location	High levels in liver and low levels in kidney, pancreas, placenta, heart and brain.

Background

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