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MUP1 Rabbit pAb

Catalog # AP97665

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q9HC21
Other Accession	Q9HC21
Reactivity	Mouse
Predicted	Rat, Human
Host	Rabbit
Clonality	Polyclonal
Calculated MW	35511
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human MUP1
Epitope Specificity	201-300/320
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the mitochondrial carrier (TC 2.A.29) family. Contains 3 Solcar repeats.
DISEASE	The disease is caused by mutations affecting the gene represented in this entry. Disease description:A disorder characterized by severe congenital microcephaly and severe 2-ketoglutaric aciduria leading to death within the first year. The disease is caused by mutations affecting the gene represented in this entry. Disease description:A disease characterized by recurrent episodes of flaccid paralysis and encephalopathy associated with bilateral striatal necrosis and chronic progressive polyneuropathy.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes a mitochondrial protein that is a member of the solute carrier family. Although this protein was initially thought to be the mitochondrial deoxynucleotide carrier involved in the uptake of deoxynucleotides into the matrix of the mitochondria, further studies have demonstrated that this protein instead functions as the mitochondrial thiamine pyrophosphate carrier, which transports thiamine pyrophosphates into mitochondria. Mutations in this gene cause microcephaly, Amish type, a metabolic disease that results in severe congenital microcephaly, severe 2-ketoglutaric aciduria, and death within the first year. Multiple alternatively spliced variants, encoding the same protein, have been identified for this gene. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	60386
Other Names	DNC; MCPHA; MTPPT; MUP1; THMD3; THMD4; TPC; TPC_HUMAN; SLC25A19; Mitochondrial thiamine pyrophosphate transporter (MTPPT); Mitochondrial

	uncoupling protein 1; Solute carrier family 25 member 19; solute carrier family 25 (mitochondrial deoxynucleotide carrier), member 19; microcephaly, Amish; solute carrier family 25 (mitochondrial thiamine pyrophosphate carrier), member 19; Mitochondrial thiamine pyrophosphate carrier; Deoxynucleotide carrier
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	SLC25A19 (HGNC:14409)
Function	Mitochondrial transporter mediating uptake of thiamine diphosphate into mitochondria. It is not clear if the antiporter activity is affected by the membrane potential or by the proton electrochemical gradient.
Cellular Location	Mitochondrion membrane; Multi-pass membrane protein
Tissue Location	Expressed in all tissues examined except for placenta. Highest levels in colon, kidney, lung, testis, spleen, and brain.

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