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OTC/Ornithine Carbamoyltransferase Rabbit pAb

Catalog # AP99664

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P00480
Other Accession	P00480
Reactivity	Rat, Human, Mouse
Predicted	Pig, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	39935
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human OTC/Ornithine Carbamoyltransferase
Epitope Specificity	31-130/354
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 ATCase/OTCase family.
SUBUNIT	Homotrimer.
Post-translational modifications	Acetylation at Lys-88 negatively regulates ornithine carbamoyltransferase activity in response to nutrient signals.
DISEASE	Defects in OTC are the cause of ornithine carbamoyltransferase deficiency (OTCD) [MIM:311250]. OTCD is an X-linked disorder of the urea cycle which causes a form of hyperammonemia. Mutations with no residual enzyme activity are always expressed in hemizygote males by a very severe neonatal hyperammonemic coma that generally proves to be fatal. Heterozygous females are either asymptomatic or express orotic aciduria spontaneously or after protein intake. The disorder is treatable with supplemental dietary arginine and low protein diet. The arbitrary classification of patients into the 'neonatal' group (clinical hyperammonemia in the first few days of life) and 'late' onset (clinical presentation after the neonatal period) has been used to differentiate severe from mild forms.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This nuclear gene encodes a mitochondrial matrix enzyme. Missense, nonsense, and frameshift mutations in this enzyme lead to ornithine transcarbamylase deficiency, which causes hyperammonemia. Since the gene for this enzyme maps close to that for Duchenne muscular dystrophy, it may play a role in that disease also. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	5009

Other Names	OCTD; OTC1; OTCD; OTCase; Sf; spf; OTC_BOVIN; OTC; Ornithine carbamoyltransferase, mitochondrial; 2.1.3.3; OTC_HUMAN; OTC_MOUSE; OTC_PIG; OTC_RAT; OTC_SHEEP; ornithine transcarbamylase; ornithine carbamoyltransferase; ornithine transcarbamylase deficiency
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,Flow-Cyt=1ug /Test
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	OTC (HGNC:8512)
Function	Catalyzes the second step of the urea cycle, the condensation of carbamoyl phosphate with L-ornithine to form L-citrulline (PubMed:2556444, PubMed:6372096, PubMed:8112735). The urea cycle ensures the detoxification of ammonia by converting it to urea for excretion (PubMed:2556444).
Cellular Location	Mitochondrion matrix
Tissue Location	Mainly expressed in liver and intestinal mucosa.

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