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PCK2 Recombinant Rabbit mAb

Catalog # AP96455

Product Information

Application	WB, IHC-P, IHC-F, IF
Reactivity	Rat, Human, Mouse
Host	Rabbit
Clonality	Recombinant
Physical State	Liquid
Isotype	IgG/Kappa
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 phosphoenolpyruvate carboxykinase [GTP] family.
SUBUNIT	Monomer.
Post-translational modifications	Phosphorylated upon DNA damage, probably by ATM or ATR.
DISEASE	Defects in PCK2 are the cause of mitochondrial phosphoenolpyruvate carboxykinase deficiency (M-PEPCKD) [MIM:261650]. A metabolic disorder resulting from impaired gluconeogenesis. It is a rare disease with less than 10 cases reported in the literature. Clinical characteristics include hypotonia, hepatomegaly, failure to thrive, lactic acidosis and hypoglycemia. Autoposy reveals fatty infiltration of both the liver and kidneys. The disorder is transmitted as an autosomal recessive trait.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	PCK2 is a a member of the phosphoenolpyruvate carboxykinase (GTP) family. The protein is a mitochondrial enzyme that catalyzes the conversion of oxaloacetate to phosphoenolpyruvate in the presence of GTP. A cytosolic form encoded by a different gene has also been characterized and is the key enzyme of gluconeogenesis in the liver. The encoded protein may serve a similar function, although it is constitutively expressed and not modulated by hormones such as glucagon and insulin that regulate the cytosolic form. Alternatively spliced transcript variants have been described.

Additional Information

Other Names	PCKGM_HUMAN; PCK2; PEPCK-M; Phosphoenolpyruvate carboxykinase 2, mitochondrial (mtPCK2); 4.1.1.32; PEPCK2; PEPCK; mtPCK2; phosphoenolpyruvate carboxykinase 2, mitochondrial; phosphoenolpyruvate carboxykinase 2 (mitochondrial)
Dilution	WB=1:500-2000,Flow-Cyt=1:50-100,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.

Name	PCK2 Recombinant Rabbit mAb
Function	Catalyzes the conversion of oxaloacetate (OAA) to phosphoenolpyruvate (PEP), the rate-limiting step in the metabolic pathway that produces glucose from lactate and other precursors derived from the citric acid cycle (By similarity).

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