

Product name:	PCB Rabbit Polyclonal Antibody
Cat number:	ABN15816
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human PC. AA range:357-406
Reactivity:	Human,Mouse,Rat
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:100-1:300,ELISA 1:10000-1:20000
Molecular Weight:	120kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

This gene encodes pyruvate carboxylase, which requires biotin and ATP to catalyse the carboxylation of pyruvate to oxaloacetate. The active enzyme is a homotetramer arranged in a tetrahedron which is located exclusively in the mitochondrial matrix. Pyruvate carboxylase is involved in gluconeogenesis, lipogenesis, insulin secretion and synthesis of the neurotransmitter glutamate. Mutations in this gene have been associated with pyruvate carboxylase deficiency. Alternatively spliced transcript variants with different 5' UTRs, but encoding the same protein, have been found for this gene. [provided by RefSeq, Jul 2008], catalytic activity: $\text{ATP} + \text{pyruvate} + \text{HCO}_3^- = \text{ADP} + \text{phosphate} + \text{oxaloacetate}$, cofactor: Binds 1 manganese ion per subunit, cofactor: Biotin, disease: Defects in PC are the cause of pyruvate carboxylase deficiency (PC deficiency) [MIM:266150]. PC deficiency leads to lactic acidosis, mental retardation and death. It occurs in three forms: mild or type A, severe neonatal or type B, and a very mild lacticacidemia, function: Pyruvate carboxylase catalyzes a 2-step reaction, involving the ATP-dependent carboxylation of the covalently attached biotin in the first step and the transfer of the carboxyl group to pyruvate in the second. Catalyzes in a tissue specific manner, the initial reactions of glucose (liver, kidney) and lipid (adipose tissue, liver, brain) synthesis from pyruvate, online information: Pyruvate carboxylase entry, pathway: Carbohydrate biosynthesis; gluconeogenesis, similarity: Contains 1 ATP-grasp domain, similarity: Contains 1 biotin carboxylation domain, similarity: Contains 1 biotinyl-binding domain, similarity: Contains 1 carboxyltransferase domain, subunit: Homotetramer,