



Rabbit Anti-PCCB antibody

SL19910R

Product Name:	PCCB
Chinese Name:	丙酰辅酶A羧化酶β链抗体
Alias:	DKFZp451E113; PCCase subunit beta; pccB; PCCB_HUMAN; pccBC Complementation group; Propanoyl CoA:carbon dioxide ligase subunit beta; Propanoyl-CoA:carbon dioxide ligase subunit beta; Propionyl CoA carboxylase beta chain, mitochondrial; propionyl Coenzyme A carboxylase, beta polypeptide; Propionyl-CoA carboxylase , beta subunit; Propionyl-CoA carboxylase beta chain, mitochondrial; R74805.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Horse,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	58kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human PCCB:321-420/539
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	The protein encoded by this gene is a subunit of the propionyl-CoA carboxylase (PCC) enzyme, which is involved in the catabolism of propionyl-CoA. PCC is a mitochondrial

enzyme that probably acts as a dodecamer of six alpha subunits and six beta subunits. This gene encodes the beta subunit of PCC. Defects in this gene are a cause of propionic acidemia type II (PA-2). Multiple transcript variants encoding different isoforms have been found for this gene.[provided by RefSeq, May 2010]

Subunit:

Probably a dodecamer composed of six biotin-containing alpha subunits and six beta subunits.

Subcellular Location:

Mitochondrion matrix.

DISEASE:

Defects in PCCB are the cause of propionic acidemia type II (PA-2) [MIM:606054]. PA-2 is a life-threatening disease characterized by episodic vomiting, lethargy and ketosis, neutropenia, periodic thrombocytopenia, hypogammaglobulinemia, developmental retardation, and intolerance to protein.

Similarity:

Belongs to the AccD/PCCB family.
Contains 1 carboxyltransferase domain.

SWISS:

P05166

Gene ID:

5096

Database links:

[Entrez Gene: 5096](#) Human

[Entrez Gene: 66904](#) Mouse

[Entrez Gene: 24624](#) Rat

[Omim: 232050](#) Human

[SwissProt: Q2TBR0](#) Cow

[SwissProt: P05166](#) Human

[SwissProt: Q99MN9](#) Mouse

[SwissProt: P79384](#) Pig

[SwissProt: P07633](#) Rat

[Unigene: 63788](#) Human

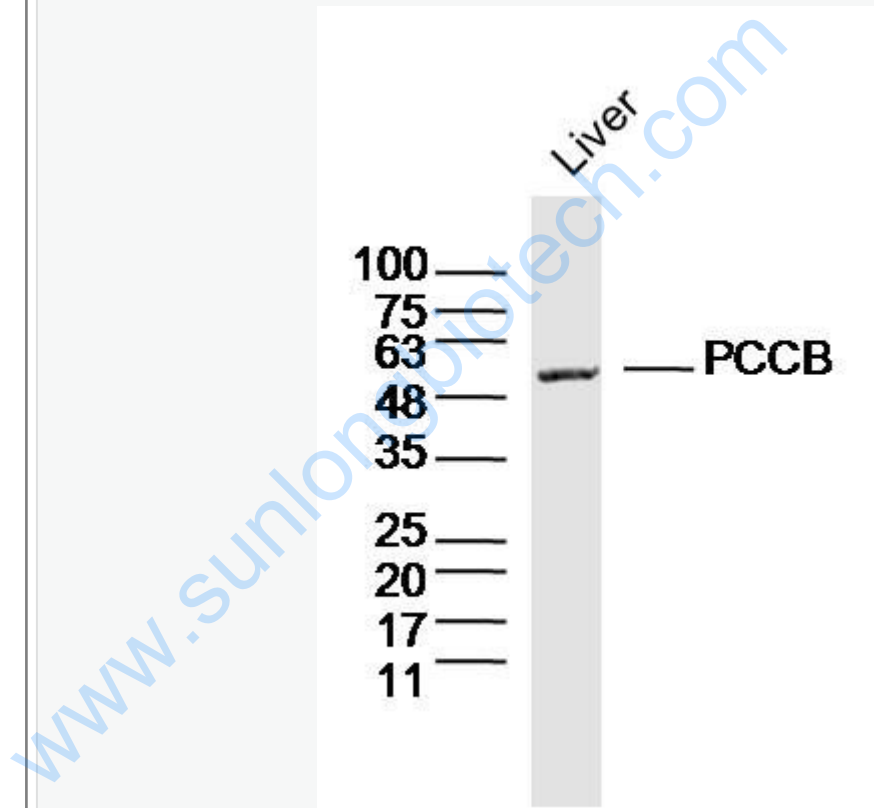
[Unigene: 335385](#) Mouse

[Unigene: 9732](#) Rat

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Picture:



Sample: Liver (Mouse) Lysate at 40 ug

Primary: Anti-PCCB (SL19910R) at 1/300 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 58kD

Observed band size: 58kD

