



Rabbit Anti-PCCA antibody

SL19909R

Product Name:	PCCA
Chinese Name:	丙酰辅酶A羧化酶 α 链抗体
Alias:	mitochondrial; Pcca; pccA COMPLEMENTATION GROUP; PCCA_HUMAN; PCCase subunit alpha; Propanoyl CoA:carbon dioxide ligase subunit alpha; Propanoyl-CoA:carbon dioxide ligase subunit alpha; Propionyl CoA carboxylase alpha chain; Propionyl CoA carboxylase alpha chain mitochondrial; Propionyl CoA carboxylase alpha polypeptide; Propionyl Coenzyme A Carboxylase Alpha Polypeptide; Propionyl-CoA carboxylase alpha chain.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
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:	74kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human PCCA:451-550/728
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 the alpha subunit of the heterodimeric mitochondrial enzyme Propionyl-CoA carboxylase. PCCA encodes the biotin-binding

region of this enzyme. Mutations in either PCCA or PCCB (encoding the beta subunit) lead to an enzyme deficiency resulting in propionic acidemia. 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 PCCA are the cause of propionic acidemia type I (PA-1) [MIM:606054]. PA-1 is a life-threatening disease characterized by episodic vomiting, lethargy and ketosis, neutropenia, periodic thrombocytopenia, hypogammaglobulinemia, developmental retardation, and intolerance to protein.

Similarity:

Contains 1 ATP-grasp domain.

Contains 1 biotin carboxylation domain.

Contains 1 biotinyl-binding domain.

SWISS:

P05165

Gene ID:

5095

Database links:

[Entrez Gene: 418774](#) Chicken

[Entrez Gene: 738775](#) Chimpanzee

[Entrez Gene: 614302](#) Cow

[Entrez Gene: 476975](#) Dog

[Entrez Gene: 101127469](#) Gorilla

[Entrez Gene: 100062251](#) Horse

[Entrez Gene: 5095](#) Human

[Entrez Gene: 110821](#) Mouse

[Entrez Gene: 100456529](#) Orangutan

[Entrez Gene: 100524103](#) Pig

[Entrez Gene: 687008](#) Rat

[Entrez Gene: 699844](#) Rhesus monkey

[Entrez Gene: 437019](#) Zebrafish

[Omin: 232000](#) Human

[SwissProt: P05165](#) Human

[SwissProt: Q91ZA3](#) Mouse

[SwissProt: P14882](#) Rat

[Unigene: 80741](#) Human

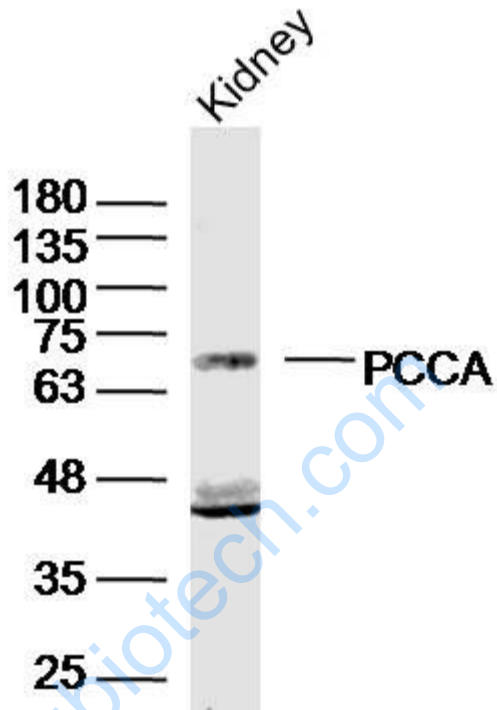
[Unigene: 23876](#) Mouse

[Unigene: 6033](#) Rat

Important Note:

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

Picture:



Sample: Kidney (Mouse) Lysate at 40 ug

Primary: Anti-PCCA (SL19909R) at 1/300 dilution

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

Predicted band size: 74 kD

Observed band size: 74 kD