

PCCA Antibody (Center) Blocking peptide
Synthetic peptide
Catalog # BP12328c**Specification**

PCCA Antibody (Center) Blocking peptide - Product InformationPrimary Accession [P05165](#)**PCCA Antibody (Center) Blocking peptide - Additional Information****Gene ID** 5095**Other Names**Propionyl-CoA carboxylase alpha chain, mitochondrial, PCCase subunit alpha,
Propanoyl-CoA:carbon dioxide ligase subunit alpha, PCCA**Format**

Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

Storage

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

Precautions

This product is for research use only. Not for use in diagnostic or therapeutic procedures.

PCCA Antibody (Center) Blocking peptide - Protein Information**Name** PCCA ([HGNC:8653](#))**Function**

This is one of the 2 subunits of the biotin-dependent propionyl-CoA carboxylase (PCC), a mitochondrial enzyme involved in the catabolism of odd chain fatty acids, branched-chain amino acids isoleucine, threonine, methionine, and valine and other metabolites (PubMed: [6765947](http://www.uniprot.org/citations/6765947), PubMed: [8434582](http://www.uniprot.org/citations/8434582)). Propionyl-CoA carboxylase catalyzes the carboxylation of propionyl-CoA/propanoyl-CoA to D-methylmalonyl-CoA/(S)-methylmalonyl-CoA (PubMed: [10101253](http://www.uniprot.org/citations/10101253), PubMed: [6765947](http://www.uniprot.org/citations/6765947), PubMed: [8434582](http://www.uniprot.org/citations/8434582)). Within the holoenzyme, the alpha subunit catalyzes the ATP-dependent carboxylation of the biotin carried by the biotin carboxyl carrier (BCC) domain, while the beta subunit then transfers the carboxyl group from carboxylated biotin to propionyl-CoA (By similarity). Propionyl-CoA carboxylase also significantly acts on butyryl-CoA/butanoyl-CoA, which is converted to ethylmalonyl-CoA/(2S)-ethylmalonyl-CoA at a much lower rate (PubMed: [6765947](http://www.uniprot.org/citations/6765947)). Other alternative minor substrates include (2E)-butenoyl-CoA/crotonoyl-CoA (By similarity).

Cellular Location
Mitochondrion matrix

PCCA Antibody (Center) Blocking peptide - Protocols

Provided below are standard protocols that you may find useful for product applications.

- [Blocking Peptides](#)

PCCA Antibody (Center) Blocking peptide - Images

PCCA Antibody (Center) Blocking peptide - Background

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.

PCCA Antibody (Center) Blocking peptide - References

Bailey, S.D., et al. Diabetes Care 33(10):2250-2253(2010) Huang, C.S., et al. Nature 466(7309):1001-1005(2010) Rose, J.E., et al. Mol. Med. 16 (7-8), 247-253 (2010) :Talmud, P.J., et al. Am. J. Hum. Genet. 85(5):628-642(2009) MacDonald, M.J., et al. Diabetologia 52(6):1087-1091(2009)