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PCCA Antibody (Center)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP12328c

Product Information

Application	WB, IHC-P, E
Primary Accession	P05165
Other Accession	NP_001121164.1, NP_000273.2
Reactivity	Human, Rat, Mouse
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB31944
Calculated MW	80059
Antigen Region	362-390

Additional Information

Gene ID	5095
Other Names	Propionyl-CoA carboxylase alpha chain, mitochondrial, PCCase subunit alpha, Propanoyl-CoA:carbon dioxide ligase subunit alpha, PCCA
Target/Specificity	This PCCA antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 362-390 amino acids from the Central region of human PCCA.
Dilution	WB~~1:2000 IHC-P~~1:100~500 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	PCCA Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

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, PubMed:8434582). Propionyl-CoA carboxylase catalyzes the carboxylation of propionyl-CoA/propanoyl-CoA to D-methylmalonyl-CoA/(S)-methylmalonyl-CoA (PubMed:10101253, PubMed:6765947, PubMed: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). Other alternative minor substrates include (2E)-butenoyl-CoA/crotonoyl-CoA (By similarity).

Cellular Location Mitochondrion matrix

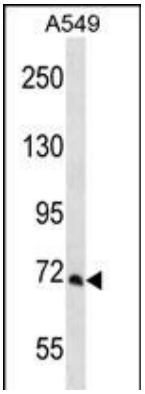
Background

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.

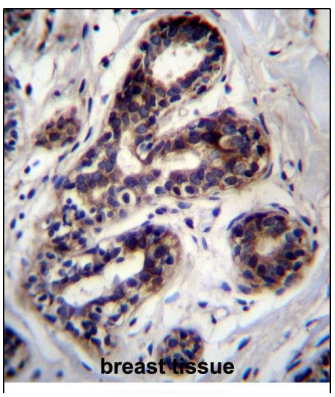
References

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)

Images



PCCA Antibody (Center) (Cat. #AP12328c) western blot analysis in A549 cell line lysates (35ug/lane).This demonstrates the PCCA antibody detected the PCCA protein (arrow).



PCCA Antibody (Center) (Cat. #AP12328c)immunohistochemistry analysis in formalin fixed and paraffin embedded human breast tissue followed by peroxidase conjugation of the secondary antibody and DAB staining.This data demonstrates the use of PCCA Antibody (Center) for immunohistochemistry. Clinical relevance has not been evaluated.

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