

# PCCA (I-17): sc-68997

## BACKGROUND

PCCA (propionyl coenzyme A carboxylase,  $\alpha$  polypeptide) is a 703 amino acid protein that localizes to the mitochondrion matrix. PCCA is a mitochondrial, biotin-dependent enzyme involved in the catabolism of branched chain amino acids, odd chain fatty acids and other metabolites. PCCA is responsible for the formation of carboxybiotin upon hydrolysis of ATP and contains a C-terminal biotin-binding domain and a biotin carboxylase domain. Inherited mutations in the gene that encodes PCCA result in a deficiency of the protein, which leads to propionic acidemia (PA), an autosomal recessive inborn error in the catabolism of methionine, isoleucine, threonine, valine, odd-numbered chain length fatty acids and cholesterol. Two isoforms exist due to alternative splicing events.

## REFERENCES

1. Kelson, T.L., et al. 1996. Chaperonin-mediated assembly of wild-type and mutant subunits of human propionyl-CoA carboxylase expressed in *Escherichia coli*. Hum. Mol. Genet. 5: 331-337.
2. Richard, E., et al. 1997. Three novel splice mutations in the PCCA gene causing identical exon skipping in propionic acidemia patients. Hum. Genet. 101: 93-96.
3. Ugarte, M., et al. 1999. Overview of mutations in the PCCA and PCCB genes causing propionic acidemia. Hum. Mutat. 14: 275-282.
4. Clavero, S., et al. 2002. Functional characterization of PCCA mutations causing propionic acidemia. Biochim. Biophys. Acta 1588: 119-125.
5. Kim, S.N., et al. 2002. Molecular analysis of PCCB gene in Korean patients with propionic acidemia. Mol. Genet. Metab. 77: 209-216.
6. Yang, X., et al. 2004. Mutation spectrum of the PCCA and PCCB genes in Japanese patients with propionic acidemia. Mol. Genet. Metab. 81: 335-342.
7. Desviat, L.R., et al. 2004. Propionic acidemia: mutation update and functional and structural effects of the variant alleles. Mol. Genet. Metab. 83: 28-37.
8. Desviat, L.R., et al. 2006. New splicing mutations in propionic acidemia. J. Hum. Genet. 51: 992-997.
9. Desviat, L.R., et al. 2009. High frequency of large genomic deletions in the PCCA gene causing propionic acidemia. Mol. Genet. Metab. 96: 171-176.

## CHROMOSOMAL LOCATION

Genetic locus: PCCA (human) mapping to 13q32.3; Pcca (mouse) mapping to 14 E5.

## SOURCE

PCCA (I-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of PCCA of human origin.

## STORAGE

Store at 4° C, \*\*DO NOT FREEZE\*\*. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

## PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-68997 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

PCCA (I-17) is recommended for detection of PCCA of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

PCCA (I-17) is also recommended for detection of PCCA in additional species, including equine, canine and bovine.

Suitable for use as control antibody for PCCA siRNA (h): sc-76077, PCCA siRNA (m): sc-76078, PCCA shRNA Plasmid (h): sc-76077-SH, PCCA shRNA Plasmid (m): sc-76078-SH, PCCA shRNA (h) Lentiviral Particles: sc-76077-V and PCCA shRNA (m) Lentiviral Particles: sc-76078-V.

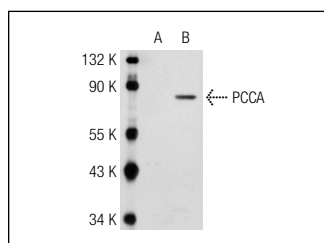
Molecular Weight of PCCA: 70 kDa.

Positive Controls: PCCA (m): 293T Lysate: sc-122423.

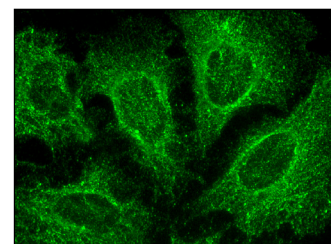
## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## DATA



PCCA (I-17): sc-68997. Western blot analysis of PCCA expression in non-transfected: sc-117752 (A) and mouse PCCA transfected: sc-122423 (B) 293T whole cell lysates.



PCCA (I-17): sc-68997. Immunofluorescence staining of methanol-fixed HeLa cells showing mitochondrial localization.

## RESEARCH USE

For research use only, not for use in diagnostic procedures.