

# PCCA (H-1): sc-393527



The Power to Question

## 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.

## CHROMOSOMAL LOCATION

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

## SOURCE

PCCA (H-1) is a mouse monoclonal antibody raised against amino acids 447-728 mapping at the C-terminus of PCCA of human origin.

## PRODUCT

Each vial contains 200  $\mu$ g IgG<sub>1</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## STORAGE

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

## APPLICATIONS

PCCA (H-1) is recommended for detection of PCCA of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

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.

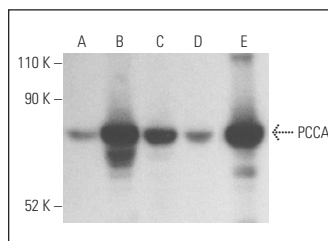
Molecular Weight of PCCA: 70 kDa.

Positive Controls: PCCA (h): 293T Lysate: sc-110766, Hep G2 cell lysate: sc-2227 or Jurkat whole cell lysate: sc-2204.

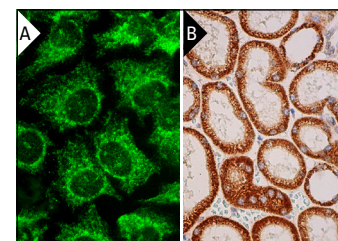
## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgG $\kappa$  BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

## DATA



PCCA (H-1): sc-393527. Western blot analysis of PCCA expression in non-transfected 293T: sc-117752 (A), human PCCA transfected 293T: sc-110766 (B), Hep G2 (C) and Jurkat (D) whole cell lysates and human kidney tissue extract (E). Detection reagent used: m-IgG Fc BP-HRP: sc-525409.



PCCA (H-1): sc-393527. Immunofluorescence staining of methanol-fixed HeLa cells showing mitochondrial localization (A). Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic staining of cells in tubules (B).

## SELECT PRODUCT CITATIONS

1. Jiang, L., et al. 2020. Dual mRNA therapy restores metabolic function in long-term studies in mice with propionic acidemia. *Nat. Commun.* 11: 5339.

## RESEARCH USE

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