

PCCB (E-12): sc-393929

BACKGROUND

Propionyl-CoA is an important intermediate of amino acid metabolism and is also produced by oxidation of odd-numbered fatty acids. Propionyl-CoA carboxylase (PCC) catalyzes the biotin-dependent carboxylation of propionyl-CoA to D-methylmalonyl-CoA. PCCB (propionyl coenzyme A carboxylase, β chain), also known as propanoyl-CoA:carbon dioxide ligase subunit β , is a 539 amino acid subunit of PCC that localizes to the mitochondrion matrix. Inherited mutations in the gene encoding PCCB result in mutations near the amino-terminus, which contains the biotin-binding site of the protein. This mutation leads to propionic acidemia type II (PA-2), an autosomal recessive disease characterized by neutropenia, hypogammaglobulinemia, episodic vomiting, ketosis and lethargy, periodic thrombocytopenia, developmental retardation and general intolerance to dietary protein.

REFERENCES

1. Lamhonwah, A.M., et al. 1986. Isolation of cDNA clones coding for the α and β chains of human propionyl-CoA carboxylase: chromosomal assignments and DNA polymorphisms associated with PCCA and PCCB genes. *Proc. Natl. Acad. Sci. USA* 83: 4864-4868.
2. Ohura, T., et al. 1993. The molecular defect in propionic acidemia: exon skipping caused by an 8-bp deletion from an intron in the PCCB allele. *Hum. Genet.* 92: 397-402.
3. Tahara, T., et al. 1993. Three independent mutations in the same exon of the PCCB gene: differences between Caucasian and Japanese propionic acidemia. *J. Inher. Metab. Dis.* 16: 353-360.

CHROMOSOMAL LOCATION

Genetic locus: PCCB (human) mapping to 3q22.3.

SOURCE

PCCB (E-12) is a mouse monoclonal antibody raised against amino acids 240-539 mapping at the C-terminus of PCCB of human origin.

PRODUCT

Each vial contains 200 μ g IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

PCCB (E-12) is available conjugated to agarose (sc-393929 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-393929 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-393929 PE), fluorescein (sc-393929 FITC), Alexa Fluor® 488 (sc-393929 AF488), Alexa Fluor® 546 (sc-393929 AF546), Alexa Fluor® 594 (sc-393929 AF594) or Alexa Fluor® 647 (sc-393929 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-393929 AF680) or Alexa Fluor® 790 (sc-393929 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

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STORAGE

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

APPLICATIONS

PCCB (E-12) is recommended for detection of PCCB of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ 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).

Suitable for use as control antibody for PCCB siRNA (h): sc-76079, PCCB shRNA Plasmid (h): sc-76079-SH and PCCB shRNA (h) Lentiviral Particles: sc-76079-V.

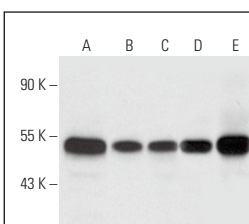
Molecular Weight of PCCB: 58 kDa.

Positive Controls: human liver extract: sc-363766, human kidney extract: sc-363764 or HeLa whole cell lysate: sc-2200.

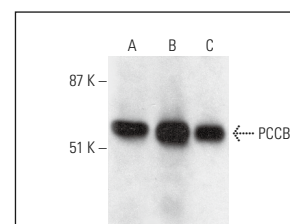
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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 κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



PCCB (E-12): sc-393929. Western blot analysis of PCCB expression in Hep G2 (A), Jurkat (B) and HeLa (C) whole cell lysates and human liver (D) and human kidney (E) tissue extracts.



PCCB (E-12): sc-393929. Western blot analysis of PCCB expression in Hep G2 whole cell lysate (A) and human liver (B) and human brain (C) tissue extracts.

SELECT PRODUCT CITATIONS

1. Martínez-Pizarro, A., et al. 2024. Functional analysis of novel variants identified in *cis* in the PCCB gene in a patient with propionic acidemia. *Gene* 893: 147902.
2. Spangsberg Petersen, U.S., et al. 2024. Regulating PCCA gene expression by modulation of pseudoexon splicing patterns to rescue enzyme activity in propionic acidemia. *Mol. Ther. Nucleic Acids* 35: 102101.

RESEARCH USE

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