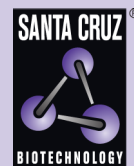


PCCB (E-12): sc-393929



The Power to Question

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

- 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.
- 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.
- Tahara, T., et al. 1993. Three independent mutations in the same exon of the PCCB gene: differences between Caucasian and Japanese propionic acidemia. *J. Inherit. Metab. Dis.* 16: 353-360.
- Lamhonwah, A.M., et al. 1994. Correction of the metabolic defect in propionic acidemia fibroblasts by microinjection of a full-length cDNA or RNA transcript encoding the propionyl-CoA carboxylase β subunit. *Genomics* 19: 500-505.

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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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.

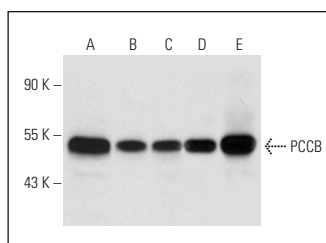
Molecular Weight of PCCB: 58 kDa.

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

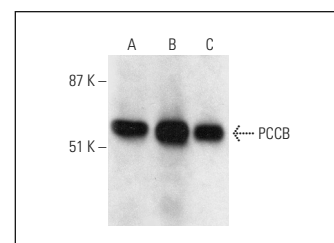
RECOMMENDED SUPPORT REAGENTS

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.

STORAGE

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

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.