SANTA CRUZ BIOTECHNOLOGY, INC.

PCCB (E-7): sc-515741

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, hypogammagloubinemia, 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 acidaemia. J. Inherit. Metab. Dis. 16: 353-360.
4. 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.
5. Rodríguez-Pombo, P., et al. 1998. Human propionyl-CoA carboxylase β subunit gene: exon-intron definition and mutation spectrum in Spanish and Latin American propionic acidemia patients. Am. J. Hum. Genet. 63: 360-369.
6. Chloupkova, M., et al. 2000. Changes in the carboxyl terminus of the β subunit of human propionyl-CoA carboxylase affect the oligomer assembly and catalysis: expression and characterization of seven patient-derived mutant forms of PCC in *Escherichia coli*. Mol. Genet. Metab. 71: 623-632.
7. Desviat, L.R., et al. 2006. New splicing mutations in propionic acidemia. J. Hum. Genet. 51: 992-997.

CHROMOSOMAL LOCATION

Genetic locus: PCCB (human) mapping to 3q22.3; Pccb (mouse) mapping to 9 E4.

SOURCE

PCCB (E-7) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 281-302 within an internal region of PCCB of human origin.

PRODUCT

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

APPLICATIONS

PCCB (E-7) is recommended for detection of PCCB of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg 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 siRNA (m): sc-76080, PCCB shRNA Plasmid (h): sc-76079-SH, PCCB shRNA Plasmid (m): sc-76080-SH, PCCB shRNA (h) Lentiviral Particles: sc-76079-V and PCCB shRNA (m) Lentiviral Particles: sc-76080-V.

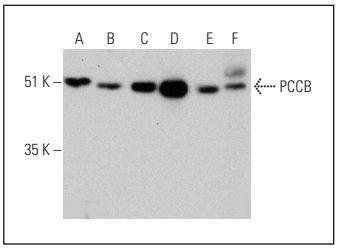
Molecular Weight of PCCB: 58 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, Hep G2 cell lysate: sc-2227 or human kidney extract: sc-363764.

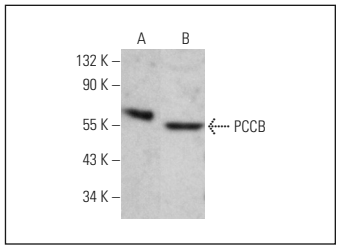
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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-IgGκ BP-FITC: sc-516140 or m-IgGκ 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-7): sc-515741. Western blot analysis of PCCB expression in Hep G2 (**A**) and HeLa (**B**) whole cell lysates and human liver (**C**), human kidney (**D**), human brain (**E**) and mouse brain (**F**) tissue extracts.



PCCB (E-7): sc-515741. Western blot analysis of PCCB expression in Hep G2 (**A**) and Neuro-2A (**B**) whole cell lysates.

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.