

PCCB (C-14): sc-68999

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. 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.

CHROMOSOMAL LOCATION

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

SOURCE

PCCB (C-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of PCCB of human origin.

PRODUCT

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

Blocking peptide available for competition studies, sc-68999 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

PCCB (C-14) is recommended for detection of PCCB of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

PCCB (C-14) is also recommended for detection of PCCB in additional species, including canine, porcine and avian.

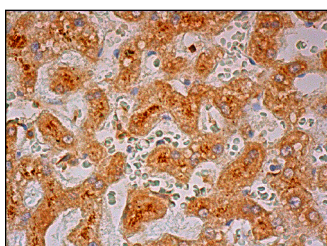
Suitable for use as control antibody for PCCB siRNA (h): sc-76079, PCCB siRNA (m): sc-76080, PCCB shRNA Plasmid (h): sc-76079-SH, PCCB shRNA Plasmid (m): sc-76080-SH, PCCB shRNA (h) Lentiviral Particles: sc-76079-V and PCCB shRNA (m) Lentiviral Particles: sc-76080-V.

Molecular Weight of PCCB: 58 kDa.

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) 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. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



PCCB (C-14): sc-68999. Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic staining of hepatocytes.

RESEARCH USE

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

MONOS
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Try **PCCB (E-12): sc-393929**, our highly recommended monoclonal alternative to PCCB (C-14).