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Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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SANTA CRUZ BIOTECHNOLOGY, INC.

PCCA (m2): 293T Lysate: sc-122423

BACKGROUND

PCCA (propionyl coenzyme A carboxylase, α 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.
8. Desviat, L.R., et al. 2006. New splicing mutations in propionic acidemia. J. Hum. Genet. 51: 992-997.
9. Desviat, L.R., et al. 2009. High frequency of large genomic deletions in the PCCA gene causing propionic acidemia. Mol. Genet. Metab. 96: 171-176.

CHROMOSOMAL LOCATION

Genetic locus: Pcca (mouse) mapping to 14 E5.

PRODUCT

PCCA (m2): 293T Lysate represents a lysate of mouse PCCA transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

APPLICATIONS

PCCA (m2): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive PCCA antibodies.

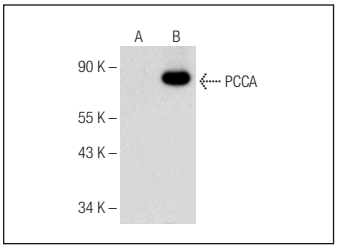
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

PCCA (D-5): sc-374341 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse PCCA expression in PCCA transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

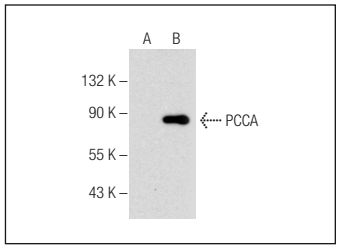
RECOMMENDED SUPPORT REAGENTS

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.

DATA



PCCA (D-5): sc-374341. Western blot analysis of PCCA expression in non-transfected: sc-117752 (**A**) and mouse PCCA transfected: sc-122423 (**B**) 293T whole cell lysates.



PCCA (F-4): sc-376428. Western blot analysis of PCCA expression in non-transfected: sc-117752 (**A**) and mouse PCCA transfected: sc-122423 (**B**) 293T whole cell lysates.

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.