SANTA CRUZ BIOTECHNOLOGY, INC.

SUCLA2 (H-296): sc-68912

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The Power to Question

BACKGROUND

SUCLA2 (succinate-CoA ligase, ADP-forming, β subunit), also known as A-β, SCS-βA or renal carcinoma antigen NY-REN-39, is a 463 amino acid mitochondrial matrix enzyme that belongs to the succinate/malate CoA ligase β subunit family. Widely expressed, SUCLA2 dimerizes with the SCS α subunit to form SCS-A, an essential component of the tricarboxylic acid cycle. Defects in SUCLA2 may be involved in a group of autosomal recessive disorders known as mitochondrial DNA depletion syndromes (MDSs) that are characterized by a decrease in mitochondrial DNA copy numbers in affected tissues. Progressive external ophthalmoplegia (PEO), ataxia-neuropathy and mitochondrial neurogastrointestinal encephalomyopathy (MNGIE) may also be associated with mutations in SUCLA2. Two isoforms of SUCLA2 exists due to alternative splicing events.

REFERENCES

1. Furuyama, K., et al. 2000. Interaction between succinyl CoA synthetase and the heme-biosynthetic enzyme ALAS-E is disrupted in sideroblastic anemia. J. Clin. Invest. 105: 757-764.
2. Elpeleg, O., et al. 2005. Deficiency of the ADP-forming succinyl-CoA synthase activity is associated with encephalomyopathy and mitochondrial DNA depletion. Am. J. Hum. Genet. 76: 1081-1086.
3. Ostergaard, E., et al. 2007. Mitochondrial encephalomyopathy with elevated methylmalonic acid is caused by SUCLA2 mutations. Brain 130: 853-861.
4. Carrozzo, R., et al. 2007. SUCLA2 mutations are associated with mild methylmalonic aciduria, Leigh-like encephalomyopathy, dystonia and deafness. Brain 130: 862-874.
5. Bourdon, A., et al. 2007. Mutation of RRM2B, encoding p53-controlled ribonucleotide reductase (p53R2), causes severe mitochondrial DNA depletion. Nat. Genet. 39: 776-780.
6. Copeland, W.C. 2008. Inherited mitochondrial diseases of DNA replication. Annu. Rev. Med. 59: 131-146.
7. Ostergaard, E. 2008. Disorders caused by deficiency of succinate-CoA ligase. J. Inherit. Metab. Dis. 31: 226-229.
8. Spinazzola, A., et al. 2008. Clinical and molecular features of mitochondrial DNA depletion syndromes. J. Inherit. Metab. Dis. 32: 143-158.
9. Bornstein, B., et al. 2008. Mitochondrial DNA depletion syndrome due to mutations in the RRM2B gene. Neuromuscul. Disord. 18: 453-459.

CHROMOSOMAL LOCATION

Genetic locus: SUCLA2 (human) mapping to 13q14.2; Sucla2 (mouse) mapping to 14 D3.

SOURCE

SUCLA2 (H-296) is a rabbit polyclonal antibody raised against amino acids 53-348 mapping near the N-terminus of SUCLA2 of human origin.

RESEARCH USE

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

PRODUCT

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

APPLICATIONS

SUCLA2 (H-296) is recommended for detection of SUCLA2 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

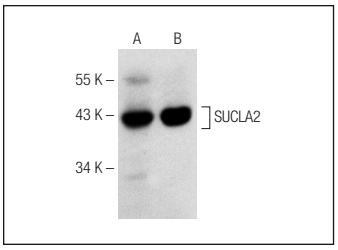
SUCLA2 (H-296) is also recommended for detection of SUCLA2 in additional species, including canine, bovine and porcine.

Suitable for use as control antibody for SUCLA2 siRNA (h): sc-76598, SUCLA2 siRNA (m): sc-76599, SUCLA2 shRNA Plasmid (h): sc-76598-SH, SUCLA2 shRNA Plasmid (m): sc-76599-SH, SUCLA2 shRNA (h) Lentiviral Particles: sc-76598-V and SUCLA2 shRNA (m) Lentiviral Particles: sc-76599-V.

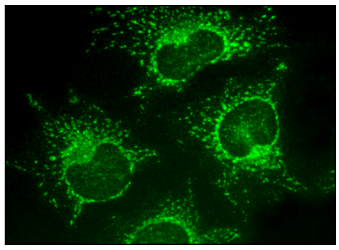
Molecular Weight of SUCLA2: 50 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, MOLT-4 cell lysate: sc-2233 or ES-2 cell lysate: sc-24674.

DATA



SUCLA2 (H-296): sc-68912. Western blot analysis of SUCLA2 expression in Hep G2 (**A**) and MOLT-4 (**B**) whole cell lysates.



SUCLA2 (H-296): sc-68912. Immunofluorescence staining of methanol-fixed HeLa cells showing mitochondrial localization.

STORAGE

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

PROTOCOLS

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

MONOS Satisfaction Guaranteed

Try **SUCLA2 (A-9): sc-374107** or **SUCLA2 (F-2): sc-373959**, our highly recommended monoclonal alternatives to SUCLA2 (H-296).