

SUCLA2 (C-19): sc-79112

BACKGROUND

SUCLA2 (succinate-CoA ligase, ADP-forming, β subunit), also known as A- β , SCS- β A 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 exist due to alternative splicing events.

REFERENCES

1. Furuyama, K. and Sassa, S. 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 synthetase 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. 2009. 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 (C-19) is an affinity purified rabbit polyclonal antibody raised against a peptide mapping at the C-terminus of SUCLA2 of human origin.

RESEARCH USE

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

PRODUCT

Each vial contains 100 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-79112 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

SUCLA2 (C-19) 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), 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 (C-19) is also recommended for detection of SUCLA2 in additional species, including equine, 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.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

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.