

# SUCLA2 (K-15): sc-79115

## BACKGROUND

SUCLA2 (succinate-CoA ligase, ADP-forming,  $\beta$  subunit), also known as A- $\beta$ , SCS- $\beta$ A or renal carcinoma antigen NY-REN-39, is a 463 amino acid mitochondrial matrix enzyme that belongs to the succinate/malate CoA ligase  $\beta$  subunit family. Widely expressed, SUCLA2 dimerizes with the SCS  $\alpha$  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.

## CHROMOSOMAL LOCATION

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

## SOURCE

SUCLA2 (K-15) is an affinity purified rabbit polyclonal antibody raised against a peptide mapping within an internal region of SUCLA2 of human origin.

## STORAGE

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

## PRODUCT

Each vial contains 100  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-79115 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

SUCLA2 (K-15) 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 (K-15) 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.

## RESEARCH USE

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

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.