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SUCLA2 (m): 293T Lysate: sc-123830

BACKGROUND

SUCLA2 (succinate-CoA ligase, ADP-forming, beta subunit), also known as A-BETA, 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., Miller, C., Hershkovitz, E., Bitner-Glindzicz, M., Rahman, S., Bondi-Rubinstein, G., Pagnamenta, A., Eshhar, S. and Saada, A. 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., Hansen, F.J., Sorensen, N., Duno, M., Vissing, J., Larsen, P.L., Faeroe, O., Thorgrimsson, S., Wibrand, F., Christensen, E. and Schwartz, M. 2007. Mitochondrial encephalomyopathy with elevated methylmalonic acid is caused by SUCLA2 mutations. *Brain* 130: 853-861.
4. Carrozzo, R., Carrozzo, R., Dionisi-Vici, C., Steuerwald, U., Lucioi, S., Deodato, F., Di Giandomenico, S., Bertini, E., Franke, B., Kluijtmans, L.A., Meschini, M.C., Rizzo, C., Piemonte, F., Rodenburg, R., Santer, R., Santorelli, F.M., van Rooij, A., Vermunt-de Koning, D., Morava, E. and Wevers, R.A. 2007. SUCLA2 mutations are associated with mild methylmalonic aciduria, Leigh-like encephalomyopathy, dystonia and deafness. *Brain* 130: 862-874.
5. Bourdon, A., Minai, L., Serre, V., Jais, J.P., Sarzi, E., Aubert, S., Chrétien, D., de Lonlay, P., Paquis-Flucklinger, V., Arakawa, H., Nakamura, Y., Munnich, A. and Rötig, A. 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. Spinazzola, A., Invernizzi, F., Carrara, F., Lamantea, E., Donati, A., Dirocco, M., Giordano, I., Meznaric-Petrusa, M., Baruffini, E., Ferrero, I. and Zeviani, M. 2008. Clinical and molecular features of mitochondrial DNA depletion syndromes. *J. Inherit. Metab. Dis.* 32:143-158.

CHROMOSOMAL LOCATION

Genetic locus: *Sucla2* (mouse) mapping to 14 D3.

RESEARCH USE

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

PRODUCT

SUCLA2 (m): 293T Lysate represents a lysate of mouse SUCLA2 transfected 293T cells and is provided as 100 μ g protein in 200 μ l SDS-PAGE buffer.

APPLICATIONS

SUCLA2 (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive SUCLA2 antibodies. Recommended use: 10-20 μ l per lane.

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

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.

PROTOCOLS

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