SANTA CRUZ BIOTECHNOLOGY, INC.

α-KGD (m): 293T Lysate: sc-124905

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

The α-ketoglutarate dehydrogenase (α-KGD) complex is a multienzyme complex which localizes to the mitochondrial matrix and consists of three protein subunits: α-ketoglutarate dehydrogenase, also designated α-KGD, E1k or oxoglutarate dehydrogenase (OGDH); dihydrolipoyl succinyltransferase (E2k or DLST); and dihydrolipoyl dehydrogenase (E3). The α-KGD subunit of the α-KGD complex catalyzes the conversion of α-ketoglutarate to succinyl-CoA and CO_2, an essential reaction of the tricarboxylic acid cycle. A definciency in α-KGD results in hypotonia, metabolic acidosis, hyperlactatemia immediately after birth, and neurologic deterioration resulting in death at about 30 months of age. Low molar ratios of ketone bodies in plasma of neonates with congenital lactic acidosis are proposed indicators of tricarboxylic acid cycle dysfunction.

REFERENCES

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2. Bunik, V.I., et al. 2005. Phosphonate analogues of α-ketoglutarate inhibit the activity of the α-KGD complex isolated from brain and in cultured cells. Biochemistry 44: 10552-10561.
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4. Strumilo, S., et al. 2005. Short-term regulation of the α-KGD complex by energy-linked and some other effectors. Biochemistry 70: 726-729.
5. Senthilnathan, P., et al. 2005. Modulation of TCA cycle enzymes and electron transport chain systems in experimental lung cancer. Life Sci. 78: 1010-1014.
6. Tian, J., et al. 2005. Variant tricarboxylic acid cycle in *Mycobacterium tuberculosis*: identification of α-KGD. Proc. Natl. Acad. Sci. USA 102: 10670-10675.
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8. Toyoshima, M., et al. 2005. Thiamine-responsive congenital lactic acidosis: clinical and biochemical studies. Pediatr. Neurol. 33: 98-104.
9. Waagepetersen, H.S., et al. 2006. Cellular mitochondrial heterogeneity in cultured astrocytes as demonstrated by immunogold labeling of α-KGD. Glia 53: 225-231.

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.

CHROMOSOMAL LOCATION

Genetic locus: Ogdh (mouse) mapping to 11 A1.

PRODUCT

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

APPLICATIONS

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

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

RESEARCH USE

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