



PDH-E1 α (h3): 293T Lysate: sc-159233

BACKGROUND

The pyruvate dehydrogenase (PDH) complex is a nuclear-encoded mitochondrial matrix enzyme complex that functions as the primary link between glycolysis and the tricarboxylic acid (TCA) cycle by catalyzing the irreversible conversion of pyruvate into acetyl-CoA. The E1 enzyme of the PDH complex is made up of a heterotetramer of two α and two β subunits. The E1 α subunit (PDH-E1 α) contains the E1 active site and plays a key role in the function of the PDH complex. The PDH complex is regulated by phosphorylation and dephosphorylation of PDH-E1 α . The gene encoding for PDH-E1 α maps to chromosome Xp22.12, and a 20 bp deletion in the last exon of this gene is sufficient to cause PDH deficiency, which causes a broad range of symptoms including the development of seizures, mental retardation and spasticity, as well as intermittent episodes of lactic acidosis associated with cerebellar ataxia.

REFERENCES

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4. Hansen, L.L., et al. 1994. Pyruvate dehydrogenase deficiency caused by a 33 base pair duplication in the PDH-E1 α subunit. *Hum. Mol. Genet.* 3: 1021-1022.
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7. Fitzgerald, J., et al. 1997. Evolution of mammalian X-linked and autosomal PGK and PDH-E1 α subunit genes. *Mol. Biol. Evol.* 13: 1023-1031.
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CHROMOSOMAL LOCATION

Genetic locus: PDHA1 (human) mapping to Xp22.12.

PRODUCT

PDH-E1 α (h3): 293T Lysate represents a lysate of human PDH-E1 α transfected 293T cells and is provided as 100 μ g protein in 200 μ l SDS-PAGE buffer.

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.

APPLICATIONS

PDH-E1 α (h3): 293T Lysate is suitable as a Western Blotting positive control for human reactive PDH-E1 α 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.

RESEARCH USE

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

PROTOCOLS

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