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Lieferung & Zahlungsart

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- Mindermengenzuschlag
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SANTA CRUZ BIOTECHNOLOGY, INC.

PDH-E1α (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 E1α subunit (PDH-E1α) 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-E1α. The gene encoding for PDH-E1α 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

1. Sermon, K., et al. 1990. Characterisation of a cDNA for porcine PDH-E1α and comparison with the human cDNA. Nucleic Acids Res. 18: 4925.
2. Chun, K., et al. 1991. Pyruvate dehydrogenase deficiency due to a 20-bp deletion in exon II of the pyruvate dehydrogenase (PDH) E1α gene. Am. J. Hum. Genet. 49: 414-420.
3. Chun, K., et al. 1993. Mutations in the X-linked E1α subunit of pyruvate dehydrogenase leading to deficiency of the pyruvate dehydrogenase complex. Hum. Mol. Genet. 2: 449-454.
4. Hansen, L.L., et al. 1994. Pyruvate dehydrogenase deficiency caused by a 33 base pair duplication in the PDH-E1α subunit. Hum. Mol. Genet. 3: 1021-1022.
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6. Otero, L.J., et al. 1996. Association of cerebral dysgenesis and lactic acidemia with X-linked PDH-E1α subunit mutations in females. Pediatr. Neurol. 13: 327-332.
7. Fitzgerald, J., et al. 1997. Evolution of mammalian X-linked and autosomal PGK and PDH-E1α subunit genes. Mol. Biol. Evol. 13: 1023-1031.
8. Seyda, A., et al. 2001. A case of PDH-E1α acidosis. J. Inherit. Metab. Dis. 24: 551-559.

CHROMOSOMAL LOCATION

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

PRODUCT

PDH-E1α (h3): 293T Lysate represents a lysate of human PDH-E1α transfected 293T cells and is provided as 100 µg protein in 200 µl 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-E1α (h3): 293T Lysate is suitable as a Western Blotting positive control for human reactive PDH-E1α 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.

PROTOCOLS

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