SANTA CRUZ BIOTECHNOLOGY, INC.

PDH-E1α (E-23): sc-133898

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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.
5. Brown, G.K., et al. 1995. Pyruvate dehydrogenase deficiency. J. Med. Genet. 31: 875-879.
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.

CHROMOSOMAL LOCATION

Genetic locus: PDHA1 (human) mapping to Xp22.12; Pdha1 (mouse) mapping to X F4.

SOURCE

PDH-E1α (E-23) is a Protein A purified rabbit polyclonal antibody raised against synthetic PDH-E1α peptide of human origin.

PRODUCT

Each vial contains 100 µg IgG in 1.0 ml PBS with < 0.1% sodium azide, 0.1% gelatin and < 0.02% sucrose.

STORAGE

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

APPLICATIONS

PDH-E1α (E-23) is recommended for detection of PDH-E1α of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for PDH-E1α siRNA (h): sc-91064, PDH-E1α siRNA (m): sc-77407, PDH-E1α shRNA Plasmid (h): sc-91064-SH, PDH-E1α shRNA Plasmid (m): sc-77407-SH, PDH-E1α shRNA (h) Lentiviral Particles: sc-91064-V and PDH-E1α shRNA (m) Lentiviral Particles: sc-77407-V.

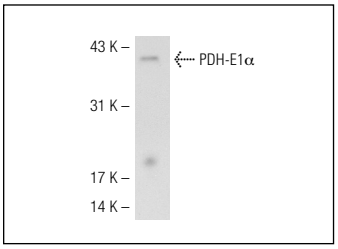
Molecular Weight of PDH-E1α: 43 kDa.

Positive Controls: rat brain extract: sc-2392, Hep G2 cell lysate: sc-2227 or HeLa whole cell lysate: sc-2200.

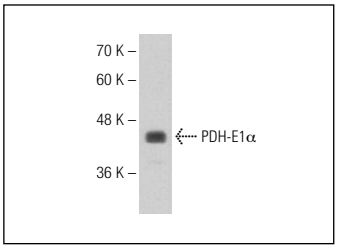
RECOMMENDED SECONDARY REAGENTS

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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

DATA



PDH-E1α (E-23): sc-133898. Western blot analysis of PDH-E1α expression in rat brain tissue extract.



PDH-E1α (E-23): sc-133898. Western blot analysis of PDH-E1α expression in Hep G2 whole cell lysate.

RESEARCH USE

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

PROTOCOLS

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

MONOS Satisfaction Guaranteed

Try **PDH-E1α (D-6): sc-377092**, our highly recommended monoclonal alternative to PDH-E1α (E-23). Also, for AC, HRP, FITC, PE, Alexa Fluor® 488 and Alexa Fluor® 647 conjugates, see **PDH-E1α (D-6): sc-377092**.