SANTA CRUZ BIOTECHNOLOGY, INC.

PDH-E1α (9H9): sc-65242

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-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.
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; Pdha1 (mouse) mapping to X F4.

SOURCE

PDH-E1α (9H9) is a mouse monoclonal antibody raised against recombinant PDH-E1α of human origin.

PRODUCT

Each vial contains 100 µg IgG_1 in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

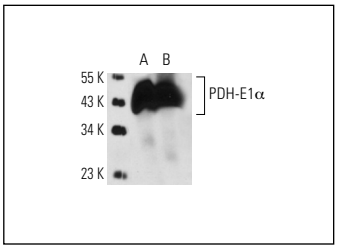
PDH-E1α (9H9) is recommended for detection of PDH-E1α of mouse, rat, human and bovine origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500).

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.

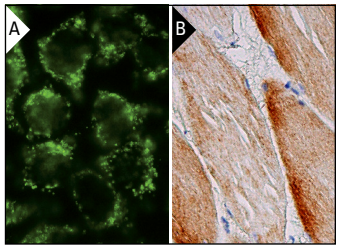
Molecular Weight of PDH-E1α: 43 kDa.

Positive Controls: Mouse brain extract: sc-2253 or mouse liver extract: sc-2256.

DATA



PDH-E1α (9H9): sc-65242. Western blot analysis of PDH-E1α expression in mouse brain (**A**) and mouse liver (**B**) tissue extracts.



PDH-E1α (9H9): sc-65242. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic localization (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human skeletal muscle tissue showing cytoplasmic staining of myocytes.

SELECT PRODUCT CITATIONS

1. Chen, Y.F., et al. 2009. Cisd2 deficiency drives premature aging and causes mitochondria-mediated defects in mice. Genes Dev. 23: 1183-1194.

STORAGE

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

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.