

PDH-E1 α (H-131): sc-292543

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

1. Sermon, K., et al. 1990. Characterisation of a cDNA for porcine PDH-E1 α 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) E1 α gene. *Am. J. Hum. Genet.* 49: 414-420.
3. Chun, K., et al. 1993. Mutations in the X-linked E1 α 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-E1 α subunit. *Hum. Mol. Genet.* 3: 1021-1022.
5. Brown, G.K., et al. 1995. Pyruvate dehydrogenase deficiency. *J. Med. Genet.* 31: 875-879.

CHROMOSOMAL LOCATION

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

SOURCE

PDH-E1 α (H-131) is a rabbit polyclonal antibody raised against amino acids 31-161 mapping near the N-terminus of PDH-E1 α of human origin.

PRODUCT

Each vial contains 200 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

Store at 4 $^{\circ}$ 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.

APPLICATIONS

PDH-E1 α (H-131) is recommended for detection of PDH-E1 α of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

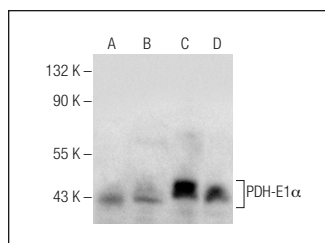
PDH-E1 α (H-131) is also recommended for detection of PDH-E1 α in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for PDH-E1 α siRNA (h): sc-91064, PDH-E1 α siRNA (m): sc-77407, PDH-E1 α shRNA Plasmid (h): sc-91064-SH, PDH-E1 α shRNA Plasmid (m): sc-77407-SH, PDH-E1 α shRNA (h) Lentiviral Particles: sc-91064-V and PDH-E1 α shRNA (m) Lentiviral Particles: sc-77407-V.

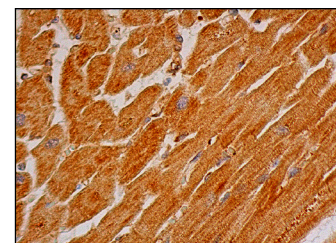
Molecular Weight of PDH-E1 α : 43 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, K-562 whole cell lysate: sc-2203 or rat brain extract: sc-2392.

DATA



PDH-E1 α (H-131): sc-292543. Western blot analysis of PDH-E1 α expression in Hep G2 (A) and K-562 (B) whole cell lysates and rat brain (C) and mouse brain (D) tissue extracts.



PDH-E1 α (H-131): sc-292543. Immunoperoxidase staining of formalin fixed, paraffin-embedded human heart muscle tissue showing cytoplasmic staining of myocytes.

SELECT PRODUCT CITATIONS

1. Pomierny, B., et al. 2015. Ethylene glycol ethers induce apoptosis and disturb glucose metabolism in the rat brain. *Sci. Dir.* E-published.

PROTOCOLS

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



Try **PDH-E1 α (D-6): sc-377092**, our highly recommended monoclonal alternative to PDH-E1 α (H-131). Also, for AC, HRP, FITC, PE, Alexa Fluor[®] 488 and Alexa Fluor[®] 647 conjugates, see **PDH-E1 α (D-6): sc-377092**.