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Rabbit Anti-phospho-PDHA1 (Ser293) antibody

SL4036R

Product Name:	phospho-PDHA1 (Ser293)
Chinese Name:	磷酸化丙酮酸脱氢酶α1抗体
Alias:	PDHA1 (phospho Ser293); PDHA1 (phospho S293); mitochondrial; somatic form; ODPA_HUMAN; PDH; PDHA1; PDHCE1A; PDHE1 A type I; PDHE1-A type I; PHE1A; Pyruvate Dehydrogenase (lipoamide) alpha 1; Pyruvate Dehydrogenase E1 alpha; Pyruvate dehydrogenase E1 component subunit alpha; Pyruvate dehydrogenase E1 component subunit alpha, somatic form, mitochondrial.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Horse,Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	40kDa
Cellular localization:	cytoplasmicMitochondrion
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated Synthesised phosphopeptide derived from human PDHA1 around the phosphorylation site of Ser293:GH(p-S)MS
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	The pyruvate dehydrogenase (PDH) complex is a nuclear-encoded mitochondrial multienzyme complex that catalyzes the overall conversion of pyruvate to acetyl-CoA

and CO(2), and provides the primary link between glycolysis and the tricarboxylic acid (TCA) cycle. The PDH complex is composed of multiple copies of three enzymatic components: pyruvate dehydrogenase (E1), dihydrolipoamide acetyltransferase (E2) and lipoamide dehydrogenase(E3). The E1 enzyme is a heterotetramer of two alpha and two beta subunits. This gene encodes the E1 alpha 1 subunit containing the E1 active site, and plays a key role in the function of the PDH complex. Mutations in this gene are associated with pyruvate dehydrogenase E1-alpha deficiency and X-linked Leigh syndrome. Alternatively spliced transcript variants encoding different isoforms have been found for this gene.

Function:
The pyruvate dehydrogenase complex catalyzes the overall conversion of pyruvate to acetyl-CoA and CO(2), and thereby links the glycolytic pathway to the tricarboxylic cycle.

Subunit:
Tetramer of 2 alpha and 2 beta subunits.

Subcellular Location:
Mitochondrion matrix.

Tissue Specificity:
Testis. Expressed in postmeiotic spermatogenic cells.

Post-translational modifications:
Phosphorylation at Ser-232, Ser-293 and Ser-300 by PDK family kinases inactivates the enzyme; for this phosphorylation at a single site is sufficient. Dephosphorylation at all three sites, i.e. at Ser-232, Ser-293 and Ser-300, is required for reactivation.

DISEASE:
Pyruvate dehydrogenase E1-alpha deficiency (PDHAD) [MIM:312170]: An enzymatic defect causing primary lactic acidosis in children. It is associated with a broad clinical spectrum ranging from fatal lactic acidosis in the newborn to chronic neurologic dysfunction with structural abnormalities in the central nervous system without systemic acidosis. Note=The disease is caused by mutations affecting the gene represented in this entry.
X-linked Leigh syndrome (X-LS) [MIM:308930]: Early-onset progressive neurodegenerative disorder with a characteristic neuropathology consisting of focal, bilateral lesions in one or more areas of the central nervous system, including the brainstem, thalamus, basal ganglia, cerebellum, and spinal cord. The lesions are areas of demyelination, gliosis, necrosis, spongiosis, or capillary proliferation. Clinical symptoms depend on which areas of the central nervous system are involved. The most common underlying cause is a defect in oxidative phosphorylation. LS may be a feature of a deficiency of any of the mitochondrial respiratory chain complexes. Note=The disease is caused by mutations affecting the gene represented in this entry.

	SWISS: P08559 **Gene ID:** 5160 **Database links:** Entrez Gene: 5160Human Entrez Gene: 18597Mouse Entrez Gene: 29554Rat Omim: 300502Human SwissProt: A7MB35Cow SwissProt: P08559Human SwissProt: P35486Mouse SwissProt: P26284Rat Unigene: 530331Human Unigene: 34775Mouse Unigene: 3655Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.