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Goat Anti-Pyruvate dehydrogenase / PDHX Antibody
Peptide-affinity purified goat antibody
Catalog # AF1889a

Specification

Goat Anti-Pyruvate dehydrogenase / PDHX Antibody - Product Information

Application	**WB**
Primary Accession	O00330
Other Accession	NP_001159630, 8050, 27402 (mouse)
Reactivity	**Human**
Predicted	**Mouse, Rat, Dog**
Host	**Goat**
Clonality	**Polyclonal**
Concentration	**100ug/200ul**
Isotype	**IgG**
Calculated MW	**54122**

Goat Anti-Pyruvate dehydrogenase / PDHX Antibody - Additional Information

Gene ID 8050

Other Names
Pyruvate dehydrogenase protein X component, mitochondrial, Dihydrolipoamide dehydrogenase-binding protein of pyruvate dehydrogenase complex, E3-binding protein, E3BP, Lipoyl-containing pyruvate dehydrogenase complex component X, proX, PDHX, PDX1

Format
0.5 mg IgG/ml in Tris saline (20mM Tris pH7.3, 150mM NaCl), 0.02% sodium azide, with 0.5% bovine serum albumin

Storage
Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions
Goat Anti-Pyruvate dehydrogenase / PDHX Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Goat Anti-Pyruvate dehydrogenase / PDHX Antibody - Protein Information

Name PDHX

Synonyms PDX1

Function
Required for anchoring dihydrolipoamide dehydrogenase (E3) to the dihydrolipoamide transacetylase (E2) core of the pyruvate dehydrogenase complexes of eukaryotes. This specific

binding is essential for a functional PDH complex.

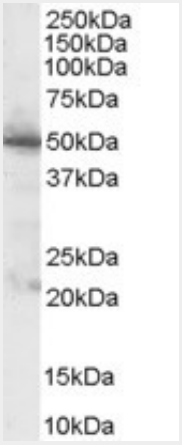
Cellular Location
Mitochondrion matrix.

Goat Anti-Pyruvate dehydrogenase / PDHX Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

Goat Anti-Pyruvate dehydrogenase / PDHX Antibody - Images



EB06371 (1 µg/ml) staining of Human Pancreas lysate (35 µg protein in RIPA buffer). Primary incubation was 1 hour. Detected by chemiluminescence.

Goat Anti-Pyruvate dehydrogenase / PDHX Antibody - Background

The pyruvate dehydrogenase (PDH) complex is located in the mitochondrial matrix and catalyzes the conversion of pyruvate to acetyl coenzyme A. The PDH complex thereby links glycolysis to Krebs cycle. The PDH complex contains three catalytic subunits, E1, E2, and E3, two regulatory subunits, E1 kinase and E1 phosphatase, and a non-catalytic subunit, E3 binding protein (E3BP). This gene encodes the E3 binding protein subunit; also known as component X of the pyruvate dehydrogenase complex. This protein tethers E3 dimers to the E2 core of the PDH complex. Defects in this gene are a cause of pyruvate dehydrogenase deficiency which results in neurological dysfunction and lactic acidosis in infancy and early childhood. This protein is also a minor antigen for antimitochondrial antibodies. These autoantibodies are present in nearly 95% of patients with the autoimmune liver disease primary biliary cirrhosis (PBC). In PBC, activated T lymphocytes attack and destroy epithelial cells in the bile duct where this protein is abnormally distributed and overexpressed. PBC eventually leads to cirrhosis and liver failure. Alternative splicing results in multiple transcript variants encoding distinct isoforms.

Goat Anti-Pyruvate dehydrogenase / PDHX Antibody - References

Enzyme-assisted photosensitization with rose Bengal acetate induces structural and functional alteration of mitochondria in HeLa cells. Bottone MG, et al. Histochem Cell Biol, 2007 Mar. PMID 17024456.
Pyruvate dehydrogenase E3 binding protein (protein X) deficiency. Brown RM, et al. Dev Med Child Neurol, 2006 Sep. PMID 16904023.
A novel gross deletion caused by non-homologous recombination of the PDHX gene in a patient with pyruvate dehydrogenase deficiency. Min M, et al. Mol Genet Metab, 2006 Sep-Oct. PMID 16843025.
PDC-E3BP is not a dominant T-cell autoantigen in primary biliary cirrhosis. McHugh A, et al. Liver Int, 2006 May. PMID 16629643.
Leigh's disease due to a new mutation in the PDHX gene. Schiff M, et al. Ann Neurol, 2006 Apr. PMID 16566017.