Product Datasheet

PDHX antibody

Catalog No: #38904

Package Size: #38904-1 50ul #38904-2 100ul

Orders: order@signalwayantibody.com
Support: tech@signalwayantibody.com

Description

Product Name	PDHX antibody
Host Species	Rabbit
Clonality	Polyclonal
Isotype	IgG
Purification	Affinity purification
Applications	WB
Species Reactivity	Human,Mouse,Rat
Specificity	The antibody detects endogenous level of total PDHX protein.
Immunogen Type	Recombinant Protein
Immunogen Description	Recombinant fusion protein of human PDHX (NP_003468.2).
Target Name	PDHX
Other Names	PDHX;DLDBP;E3BP;OPDX;PDX1;proX
Accession No.	Uniprot:O00330GeneID:8050
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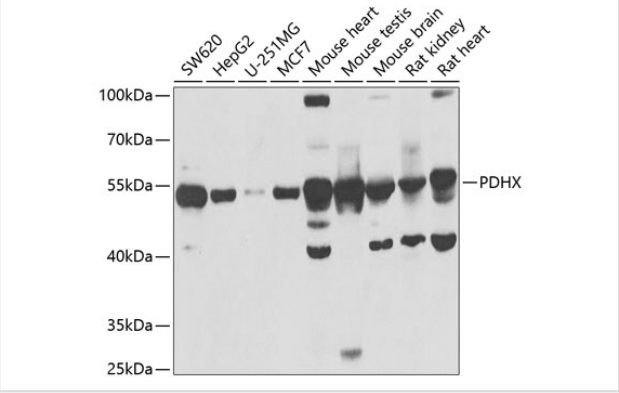
GeneID	8050
SDS-PAGE MW	54kDa
Concentration	1.0mg/ml
Formulation	PBS with 0.02% sodium azide,50% glycerol,pH7.3.
Storage	Store at -20°C. Avoid freeze / thaw cycles.

Application Details

WB□1:500 - 1:2000

Images



Western blot analysis of extracts of various cell lines, using 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.

Note: This product is for in vitro research use only