

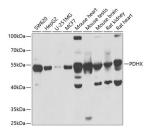
Tel:240-252-7368(USA) Fax: 240-252-7376(USA) techsupport@elabscience.com Website: www.elabscience.com

PDHX Polyclonal Antibody

Catalog No.E-AB-61954ReactivityH,M,RStorageStore at -20°C. Avoid freeze / thaw cycles.HostRabbitApplicationsWBIsotypeIgG

Note: Centrifuge before opening to ensure complete recovery of vial contents.

Images



Western blot analysis of extracts of various cell lines using PDHX Polyclonal Antibody at 1:1000 dilution.

Immunogen Information

Immunogen Recombinant fusion protein of human PDHX

GeneID 8050 **Swissprot** 000330

Synonyms PDHX,DLDBP,E3BP,OPDX,PDX1,proX

Product Information

Calculated MW 29kDa/51kDa/54kDa

Observed MW 54kDa

Buffer PBS with 0.02% sodium azide,50% glycerol,pH7.3.

Purify Affinity purification **Dilution** WB 1:500-1:2000

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.

For Research Use Only

Thank you for your recent purchase

If you would like to learn more about antibodies, please visit www.elabscience.com.

Focus on your research Service for life science