

Product datasheet for TA379759S

PDHX Rabbit Polyclonal Antibody

Product data:

Product Type: Primary Antibodies

Applications: ELISA, IP, WB

Recommended Dilution: WB,1:500 - 1:2000

IP,0.5μg-4μg antibody for 400μg-600μg extracts of whole cells

ELISA, Recommended starting concentration is 1 µg/mL. Please optimize the concentration

based on your specific assay requirements.

Reactivity: Human, Mouse, Rat

Modifications: Unmodified

Host: Rabbit Isotype: IgG

Clonality: Polyclonal

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

Concentration: lot specific

Purification: Affinity purification

Conjugation: Unconjugated

Storage: Store at -20°C. Avoid freeze / thaw cycles.

Stability: Shelf life: one year from despatch.

Predicted Protein Size: 54kDa

Gene Name: pyruvate dehydrogenase complex component X

Database Link: Entrez Gene 8050 Human

<u>O00330</u>



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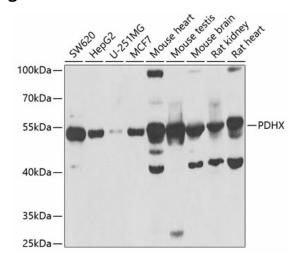


Background:

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.

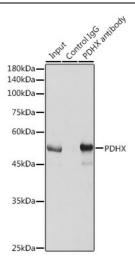
Synonyms: DLDBP; E3BP; OPDX; PDX1; proX

Product images:



Western blot analysis of various lysates using PDHX Rabbit pAb ([TA379759]) at 1:1000 dilution. Secondary antibody: HRP-conjugated Goat anti-Rabbit lgG (H+L) (AS014) at 1:10000 dilution. Lysates/proteins: 25µg per lane. Blocking buffer: 3% nonfat dry milk in TBST. Detection: ECL Basic Kit (RM00020). Exposure time: 5s.





Immunoprecipitation analysis of 600 μ g extracts of Mouse brain cells using 3 μ g PDHX antibody ([TA379759]). Western blot was performed from the immunoprecipitate using PDHX antibody ([TA379759]) at a dilution of 1:1000.