

CAB6426

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## Product Information

<b>Product SKU:</b>	CAB6426	<b>Gene ID:</b>	8050	<b>Size:</b>	20uL, 100uL
<b>Clone No:</b>	-	<b>Host Species:</b>	Rabbit	<b>Reactivity:</b>	Human,Mouse,Rat

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## Additional Information

<b>Observed MW:</b>	54kDa	<b>Conjugate:</b>	Unconjugated
<b>Calculated MW:</b>	54kDa	<b>Isotype:</b>	IgG

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## Immunogen Information

<b>Background:</b>	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.
<b>Recommended Dilution:</b>	WB,1:500 - 1:2000 IP,0.5µg-4µg antibody for 400µg-600µg extracts of whole cells
<b>Synonyms:</b>	E3BP; OPDX; PDX1; proX; DLDBP; PDHDX; PDHX
<b>Purification Method:</b>	Affinity purification
<b>Immunogen:</b>	Recombinant fusion protein containing a sequence corresponding to amino acids 1-300 of human PDHX (NP_003468.2).
<b>Storage:</b>	Store at -20°C. Avoid freeze / thaw cycles.Buffer: PBS with 0.02% sodium azide,50% glycerol,pH7.3.