



# ODPB Polyclonal Antibody

|                           |   |
|---------------------------|---|
| <b>Catalog No</b>         | BYab-06002  |
| <b>Isotype</b>            | IgG   |
| <b>Reactivity</b>         | Human;Rat;Mouse   |
| <b>Applications</b>       | WB;ELISA  |
| <b>Gene Name</b>          | PDHB PHE1B  |
| <b>Protein Name</b>       | Pyruvate dehydrogenase E1 component subunit beta, mitochondrial (PDHE1-B) (EC 1.2.4.1)  |
| <b>Immunogen</b>          | Synthesized peptide derived from human protein . at AA range: 160-240   |
| <b>Specificity</b>        | ODPB Polyclonal Antibody detects endogenous levels of protein.  |
| <b>Formulation</b>        | Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.  |
| <b>Source</b>             | Polyclonal, Rabbit,IgG  |
| <b>Purification</b>       | The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.   |
| <b>Dilution</b>           | WB 1:500-2000 ELISA 1:5000-20000  |
| <b>Concentration</b>      | 1 mg/ml   |
| <b>Purity</b>             | ≥90%  |
| <b>Storage Stability</b>  | -20°C/1 year  |
| <b>Synonyms</b>           |   |
| <b>Observed Band</b>      | 39kD  |
| <b>Cell Pathway</b>       | Mitochondrion matrix.   |
| <b>Tissue Specificity</b> | Brain,Heart,Lung,   |
| <b>Function</b>           | catalytic activity:Pyruvate + [dihydrolipoyllysine-residue acetyltransferase] lipoyllysine = [dihydrolipoyllysine-residue acetyltransferase] S-acetyldihydrolipoyllysine + CO(2).,cofactor:Thiamine pyrophosphate.,disease:Defects in PDHB are a cause of pyruvate dehydrogenase E1 component deficiency (PDHE1 deficiency) [MIM:312170]. PDHE1 deficiency is the most common enzyme defect in patients with primary lactic acidosis. It is associated with variable clinical phenotypes ranging from neonatal death to prolonged survival complicated by developmental delay, seizures, ataxia, apnea, and in some cases to an X-linked form of Leigh syndrome (LS) (Leigh encephalomyelopathy).,function:The pyruvate dehydrogenase complex catalyzes the overall conversion of pyruvate to acetyl-CoA and CO(2). It contains multiple copies of three enzymatic components: pyruvate |

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## dehydrogenase (E1), dihydrolipoamide acetyl

### Background

The pyruvate dehydrogenase (PDH) complex is a nuclear-encoded mitochondrial multienzyme complex that catalyzes the overall conversion of pyruvate to acetyl-CoA and carbon dioxide, and provides the primary link between glycolysis and the tricarboxylic acid (TCA) cycle. The PDH complex is composed of multiple copies of three enzymatic components: pyruvate dehydrogenase (E1), dihydrolipoamide acetyltransferase (E2) and lipoamide dehydrogenase (E3). The E1 enzyme is a heterotetramer of two alpha and two beta subunits. This gene encodes the E1 beta subunit. Mutations in this gene are associated with pyruvate dehydrogenase E1-beta deficiency. Alternatively spliced transcript variants have been found for this gene. [provided by RefSeq, Mar 2012],

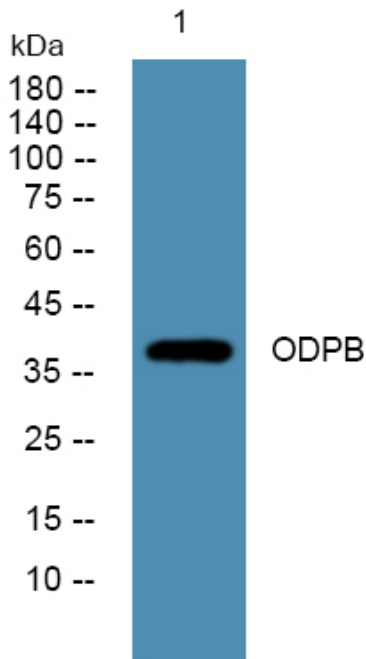
### matters needing attention

Avoid repeated freezing and thawing!

### Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

## Products Images



Western blot analysis of lysates from SW480 cells, primary antibody was diluted at 1:1000, 4° over night