



ODPB Monoclonal Antibody

Catalog No	BYmab-06002
Isotype	IgG
Reactivity	Human;Rat;Mouse
Applications	WB
Gene Name	PDHB PHE1B
Protein Name	Pyruvate dehydrogenase E1 component subunit beta, mitochondrial (PDHE1-B) (EC 1.2.4.1)
Immunogen	Synthesized peptide derived from human protein . at AA range: 160-240
Specificity	ODPB Monoclonal Antibody detects endogenous levels of protein.
Formulation	Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	39kD
Cell Pathway	Mitochondrion matrix.
Tissue Specificity	Brain,Heart,Lung,
Function	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],

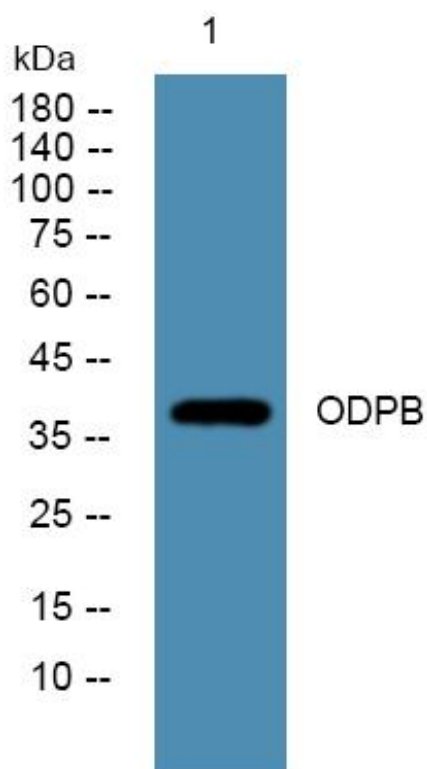
matters needing attention

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 various cells using ODPB Monoclonal Antibody