

Anti-PDHB Antibody [JM66-33]

ET1705-57



Product Type:	Recombinant Rabbit monoclonal IgG, primary antibodies
Species reactivity:	Human, Mouse, Rat
Applications:	WB, IHC-P, FC, IP
Molecular Wt:	Predicted band size: 39 kDa
Clone number:	JM66-33

Description: The pyruvate dehydrogenase complex catalyzes the overall conversion of pyruvate to acetyl-CoA and CO. It contains multiple copies of three enzymatic components: pyruvate dehydrogenase (E1), dihydrolipoamide acetyltransferase (E2) and lipoamide dehydrogenase (E3). Defects in PDHB are a cause of pyruvate dehydrogenase E1 component deficiency (PDHE1 deficiency). 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).

Immunogen: Synthetic peptide within Human PDHB aa 310-359 / 359.

Positive control: HeLa cell lysate, HepG2 cell lysate, C2C12 cell lysate, L6 cell lysate, Mouse heart tissue lysate, Rat heart tissue lysate, human brain tissue, human heart tissue, mouse brain tissue, mouse heart tissue, rat brain tissue, rat heart tissue, Hela.

Subcellular location: Mitochondrion matrix.

Database links: SwissProt: P11177 Human | Q9D051 Mouse | P49432 Rat

Recommended Dilutions:

WB	1:2,000
IHC-P	1:200-1:1,000
FC	1:50-1:100
IP	Use at an assay dependent concentration.

Storage Buffer: 1*TBS (pH7.4), 0.05% BSA, 40% Glycerol. Preservative: 0.05% Sodium Azide.

Storage Instruction: Shipped at 4°C. Store at +4°C short term (1-2 weeks). It is recommended to aliquot into single-use upon delivery. Store at -20°C long term.

Purity: Protein A affinity purified.

Hangzhou Huaan Biotechnology Co., Ltd.

Orders:0086-571-88062880

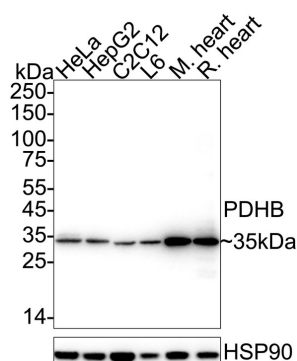
Technical:0086-571-89986345

Service mail:support@huabio.cn

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Images

Fig1: Western blot analysis of PDHB on different lysates with Rabbit anti-PDHB antibody (ET1705-57) at 1/2,000 dilution.

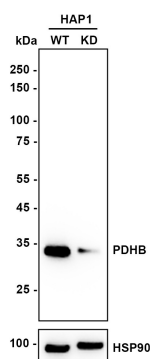


Lane 1: HeLa cell lysate (20 µg/Lane)
 Lane 2: HepG2 cell lysate (20 µg/Lane)
 Lane 3: C2C12 cell lysate (20 µg/Lane)
 Lane 4: L6 cell lysate (20 µg/Lane)
 Lane 5: Mouse heart tissue lysate (40 µg/Lane)
 Lane 6: Rat heart tissue lysate (40 µg/Lane)

Predicted band size: 39 kDa
 Observed band size: 34 kDa
 Exposure time: 8 seconds; ECL: K1801;
 4-20% SDS-PAGE gel.

Proteins were transferred to a PVDF membrane and blocked with 5% NFDM/TBST for 1 hour at room temperature. The primary antibody (ET1705-57) at 1/2,000 dilution was used in 5% NFDM/TBST at 4°C overnight. Goat Anti-Rabbit IgG - HRP Secondary Antibody (HA1001) at 1/50,000 dilution was used for 1 hour at room temperature.

Fig2: Western blot analysis of PDHB on different lysates with Rabbit anti-PDHB antibody (ET1705-57) at 1/2,000 dilution.



Lane 1: HAP1-parental cell lysate
 Lane 2: HAP1-PDHB KD cell lysate

Lysates/proteins at 10 µg/Lane.

Predicted band size: 39 kDa
 Observed band size: 34 kDa

Exposure time: 6 seconds; ECL: K1801;

4-20% SDS-PAGE gel.

Proteins were transferred to a PVDF membrane and blocked with 5% NFDM/TBST for 1 hour at room temperature. The primary antibody (ET1705-57) at 1/2,000 dilution was used in K1803 at 4°C overnight. Goat Anti-Rabbit IgG - HRP Secondary Antibody (HA1001) at 1/50,000 dilution was used for 1 hour at room temperature.

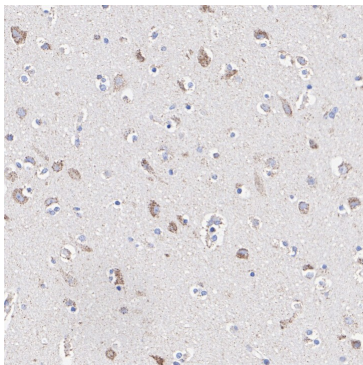


Fig3: Immunohistochemical analysis of paraffin-embedded human brain tissue with Rabbit anti-PDHB antibody (ET1705-57) at 1/200 dilution.

The section was pre-treated using heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0) for 20 minutes. The tissues were blocked in 1% BSA for 20 minutes at room temperature, washed with ddH₂O and PBS, and then probed with the primary antibody (ET1705-57) at 1/200 dilution for 1 hour at room temperature. The detection was performed using an HRP conjugated compact polymer system. DAB was used as the chromogen. Tissues were counterstained with hematoxylin and mounted with DPX.

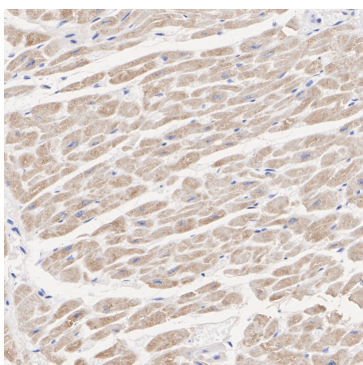


Fig4: Immunohistochemical analysis of paraffin-embedded human heart tissue with Rabbit anti-PDHB antibody (ET1705-57) at 1/1,000 dilution.

The section was pre-treated using heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0) for 20 minutes. The tissues were blocked in 1% BSA for 20 minutes at room temperature, washed with ddH₂O and PBS, and then probed with the primary antibody (ET1705-57) at 1/1,000 dilution for 1 hour at room temperature. The detection was performed using an HRP conjugated compact polymer system. DAB was used as the chromogen. Tissues were counterstained with hematoxylin and mounted with DPX.

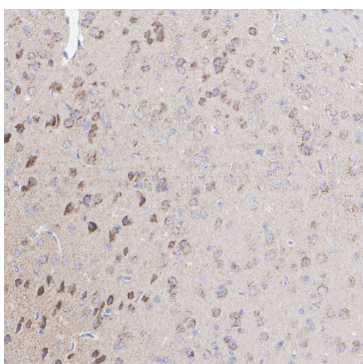


Fig5: Immunohistochemical analysis of paraffin-embedded mouse brain tissue with Rabbit anti-PDHB antibody (ET1705-57) at 1/200 dilution.

The section was pre-treated using heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0) for 20 minutes. The tissues were blocked in 1% BSA for 20 minutes at room temperature, washed with ddH₂O and PBS, and then probed with the primary antibody (ET1705-57) at 1/200 dilution for 1 hour at room temperature. The detection was performed using an HRP conjugated compact polymer system. DAB was used as the chromogen. Tissues were counterstained with hematoxylin and mounted with DPX.

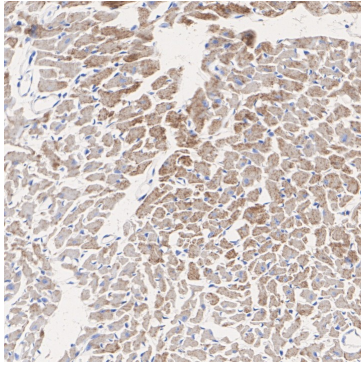


Fig6: Immunohistochemical analysis of paraffin-embedded mouse heart tissue with Rabbit anti-PDHB antibody (ET1705-57) at 1/200 dilution.

The section was pre-treated using heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0) for 20 minutes. The tissues were blocked in 1% BSA for 20 minutes at room temperature, washed with ddH₂O and PBS, and then probed with the primary antibody (ET1705-57) at 1/200 dilution for 1 hour at room temperature. The detection was performed using an HRP conjugated compact polymer system. DAB was used as the chromogen. Tissues were counterstained with hematoxylin and mounted with DPX.

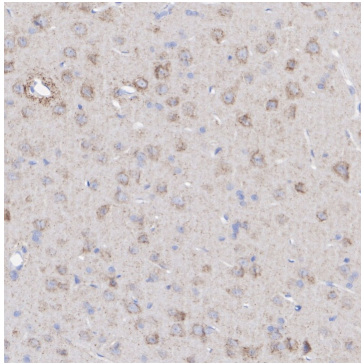


Fig7: Immunohistochemical analysis of paraffin-embedded rat brain tissue with Rabbit anti-PDHB antibody (ET1705-57) at 1/200 dilution.

The section was pre-treated using heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0) for 20 minutes. The tissues were blocked in 1% BSA for 20 minutes at room temperature, washed with ddH₂O and PBS, and then probed with the primary antibody (ET1705-57) at 1/200 dilution for 1 hour at room temperature. The detection was performed using an HRP conjugated compact polymer system. DAB was used as the chromogen. Tissues were counterstained with hematoxylin and mounted with DPX.

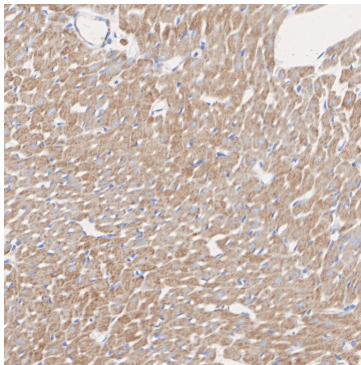


Fig8: Immunohistochemical analysis of paraffin-embedded rat heart tissue with Rabbit anti-PDHB antibody (ET1705-57) at 1/200 dilution.

The section was pre-treated using heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0) for 20 minutes. The tissues were blocked in 1% BSA for 20 minutes at room temperature, washed with ddH₂O and PBS, and then probed with the primary antibody (ET1705-57) at 1/200 dilution for 1 hour at room temperature. The detection was performed using an HRP conjugated compact polymer system. DAB was used as the chromogen. Tissues were counterstained with hematoxylin and mounted with DPX.

Note: All products are "FOR RESEARCH USE ONLY AND ARE NOT INTENDED FOR DIAGNOSTIC OR THERAPEUTIC USE".

Background References

1. Halim ND et al. Phosphorylation status of pyruvate dehydrogenase distinguishes metabolic phenotypes of cultured rat brain astrocytes and neurons. *Glia* 58:1168-76 (2010).
2. Han Z et al. A combined therapeutic approach for pyruvate dehydrogenase deficiency using self-complementary adeno-associated virus serotype-specific vectors and dichloroacetate. *Mol Genet Metab* 93:381-7 (2008).

