# abcam

### Product datasheet

# Anti-PDHA1 antibody [9H9AF5] ab110330



## \*\*\* \* \* \* 8 Abreviews 78 References 5 Images

#### Overview

Product name Anti-PDHA1 antibody [9H9AF5]

**Description** Mouse monoclonal [9H9AF5] to PDHA1

Host species Mouse

**Tested applications** Suitable for: WB, ICC/IF, Flow Cyt

**Species reactivity** Reacts with: Mouse, Rat, Cow, Human

**Immunogen** Recombinant full length protein corresponding to Human PDHA1.

Positive control Isolated mitochondria from Human, Bovine, Rat and Mouse hearts, HepG2 lysate; cultured, normal

Human embryonic lung fibroblasts (strain MRC5); HL60 cells.

General notes

This antibody clone is manufactured by Abcam. If you require a custom buffer formulation or

conjugation for your experiments, please contact orders@abcam.com.

The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets

your needs before purchasing.

If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be

found below, along with publications, customer reviews and Q&As

Product was previously marketed under the MitoSciences sub-brand.

#### **Properties**

Form Liquid

**Storage instructions** Shipped at 4°C. Store at +4°C. Do Not Freeze.

Storage buffer pH: 7.5

Preservative: 0.02% Sodium azide Constituent: HEPES buffered saline

**Purity** lgG fraction

Purification notes ab110330 was produced in vitro using hybridomas grown in serum-free medium, and then

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purified by biochemical fractionation.

Clonality Monoclonal
Clone number 9H9AF5
Isotype IgG1
Light chain type kappa

#### **Applications**

The Abpromise guarantee

Our Abpromise quarantee covers the use of ab110330 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB	****(6)	Use a concentration of 1 $\mu$ g/ml. Predicted molecular weight: 43 kDa.
ICC/IF	**** <u>*</u> (1)	Use a concentration of 5 µg/ml. (heat-induced antigen-retrieval improves signal)
Flow Cyt		Use a concentration of 1 µg/ml.  ab170190 - Mouse monoclonal lgG1, is suitable for use as an isotype control with this antibody.

#### **Target**

**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 dehydrogenase (E1), dihydrolipoamide acetyltransferase (E2) and lipoamide dehydrogenase (E3).

**Tissue specificity** 

Ubiquitous.

Involvement in disease

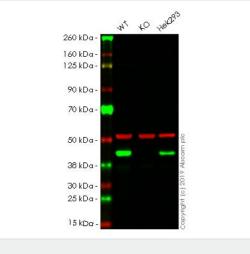
Defects in PDHA1 are a cause of pyruvate decarboxylase E1 component deficiency (PDHE1 deficiency) [MIM:312170]. PDHE1 deficiency is the most common enzyme defect in patients with primary lactic acidosis. It is associated with variable clinical phenotypes ranging from neonatal death to prolonged survival complicated by developmental delay, seizures, ataxia, apnea, and in some cases to an X-linked form of Leigh syndrome (X-LS).

Defects in PDHA1 are the cause of X-linked Leigh syndrome (X-LS) [MIM:308930]. X-LS is an early-onset progressive neurodegenerative disorder with a characteristic neuropathology consisting of focal, bilateral lesions in one or more areas of the central nervous system, including the brainstem, thalamus, basal ganglia, cerebellum, and spinal cord. The lesions are areas of demyelination, gliosis, necrosis, spongiosis, or capillary proliferation. Clinical symptoms depend on which areas of the central nervous system are involved. The most common underlying cause is a defect in oxidative phosphorylation. LS may be a feature of a deficiency of any of the mitochondrial respiratory chain complexes.

**Cellular localization** 

Mitochondrion matrix.

#### **Images**



Western blot - Anti-PDHA1 antibody [9H9AF5] (ab110330)

All lanes: Anti-PDHA1 antibody [9H9AF5] (ab110330) at 1 µg/ml

Lane 1: Wild-type HeLa whole cell lysate

Lane 2: PDHA1 knockout HeLa whole cell lysate

Lane 3: HEK-293 whole cell lysate

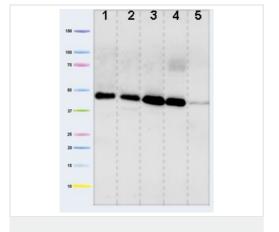
Lysates/proteins at 20 µg per lane.

**Predicted band size:** 43 kDa **Observed band size:** 43 kDa

**Lanes 1 - 3:** Merged signal (red and green). Green - ab110330 observed at 43 kDa. Red - loading control, **ab52866**, observed at 50 kDa.

ab110330 was shown to specifically react with PDHA1 in wild-type HeLa cells as signal was lost in PDHA1 knockout cells. Wild-type and PDHA1 knockout samples were subjected to SDS-PAGE.

Ab110330 and <u>ab52866</u> (Rabbit anti alpha Tubulin loading control) were incubated overnight at 4°C at 1 ug/ml and 1/1000 dilution respectively. Blots were developed with Goat anti-Mouse lgG H&L (IRDye® 800CW) preabsorbed <u>ab216772</u> and Goat anti-Rabbit lgG H&L (IRDye® 680RD) preabsorbed <u>ab216777</u> secondary antibodies at 1/20000 dilution for 1 hour at room temperature before imaging.



Western blot - Anti-PDHA1 antibody [9H9AF5] (ab110330)

All lanes : Anti-PDHA1 antibody [9H9AF5] (ab110330) at 1  $\mu$ g/ml

**Lane 1 :** Isolated mitochondria from Human heart at 10  $\mu g$ 

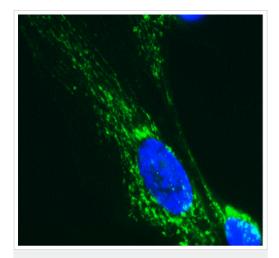
**Lane 2 :** Isolated mitochondria from Bovine heart at 4  $\mu g$ 

Lane 3 : Isolated mitochondria from Rat heart at 10  $\mu g$ 

Lane 4: Isolated mitochondria from Mouse heart at 10  $\mu g$ 

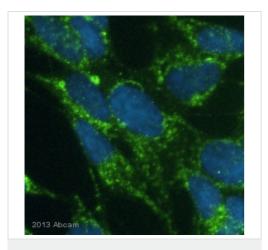
Lane 5: hepG2 cell lysate at 20 µg

Predicted band size: 43 kDa



Immunocytochemistry/ Immunofluorescence - Anti-PDHA1 antibody [9H9AF5] (ab110330)

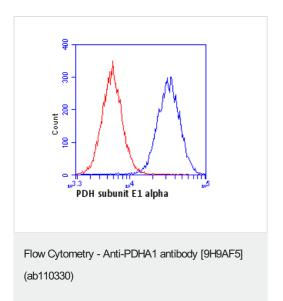
Immunocytochemistry analysis using ab110330 at  $5\mu g/ml$  staining PDHA1 in cultured, normal Human embryonic lung fibroblasts (strain MRC5) followed by AlexaFluor® 488 goat anti-mouse IgG1 secondary antibody (2  $\mu g/ml$ ).



Immunocytochemistry/ Immunofluorescence - Anti-PDHA1 antibody [9H9AF5] (ab110330)

'This image is courtesy of an Abreview submitted by George Allen'

Immunocytochemical analysis of SH-SY5Y human neuroblastoma labeling PDHA1 with ab110330 at 1/500 dilution.



Flow cytometric analysis using ab110330 at  $1\mu g/ml$  staining PDHA1 in HL60 cells (blue). Isotype control antibody (red).

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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