


# Anti-PDHA1 antibody ab97352

[1 References](#) [3 图像](#)

### 概述

<b>产品名称</b>	Anti-PDHA1抗体
<b>描述</b>	兔多克隆抗体to PDHA1
<b>宿主</b>	Rabbit
<b>经测试应用</b>	<b>适用于:</b> WB, ICC/IF
<b>种属反应性</b>	<b>与反应:</b> Human <b>预测可用于:</b> Mouse, Cow, Zebrafish 
<b>免疫原</b>	Recombinant fragment within Human PDHA1 aa 39-295. The exact sequence is proprietary. (NP_000275).
<b>阳性对照</b>	WB: 293T, A431, H1299, HeLa S3, HepG2, MOLT4 and Raji ICC/IF: HeLa cells
<b>常规说明</b>	<p>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.</p> <p>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&amp;As</p>

### 性能

<b>形式</b>	Liquid
<b>存放说明</b>	Shipped at 4°C. Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.
<b>存储溶液</b>	pH: 7.00 Preservative: 0.025% Proclin 300 Constituents: 79% PBS, 20% Glycerol (glycerin, glycerine)
<b>纯度</b>	Immunogen affinity purified
<b>克隆</b>	多克隆
<b>同种型</b>	IgG

### 应用

## The Abpromise guarantee

**Abpromise™** 承诺保证使用 ab97352 于以下的经测试应用

“应用说明”部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
WB		1/500 - 1/3000. Predicted molecular weight: 43 kDa.
ICC/IF		1/100 - 1/200.

## 靶标

### 功能

The pyruvate dehydrogenase complex catalyzes the overall conversion of pyruvate to acetyl-CoA and CO<sub>2</sub>. It contains multiple copies of three enzymatic components: pyruvate dehydrogenase (E1), dihydrolipoamide acetyltransferase (E2) and lipoamide dehydrogenase (E3).

### 组织特异性

Ubiquitous.

### 疾病相关

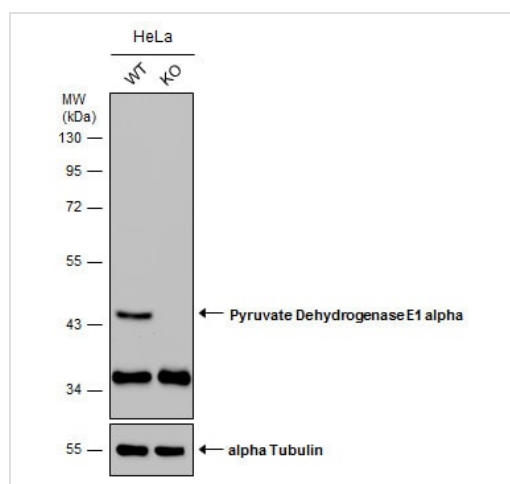
Defects in PDHA1 are a cause of pyruvate decarboxylase E1 component deficiency (PDHE1 deficiency) [MIM:312170]. PDHE1 deficiency is the most common enzyme defect in patients with primary lactic acidosis. It is associated with variable clinical phenotypes ranging from neonatal death to prolonged survival 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.

### 细胞定位

Mitochondrion matrix.

## 图片



Western blot - Anti-PDHA1 antibody (ab97352)

**All lanes** : Anti-PDHA1 antibody (ab97352) at 1/1000 dilution

**Lane 1** : WT HeLa cell extract

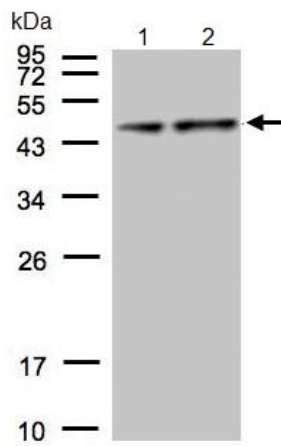
**Lane 2** : Pyruvate Dehydrogenase E1-alpha subunit Knockout HeLa cell extracts

Lysates/proteins at 30 µg per lane.

### Secondary

**All lanes** : HRP-conjugated anti-rabbit IgG

**Predicted band size:** 43 kDa



Western blot - Anti-PDHA1 antibody (ab97352)

**All lanes :** Anti-PDHA1 antibody (ab97352) at 1/1000 dilution

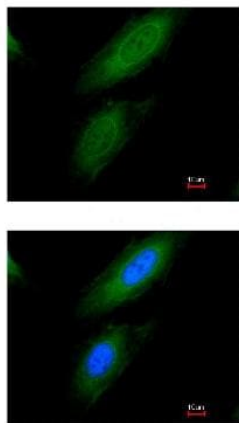
**Lane 1 :** HepG2 whole cell lysate

**Lane 2 :** MOLT4 whole cell lysate

Lysates/proteins at 30 µg per lane.

**Predicted band size:** 43 kDa

12% SDS PAGE



Immunocytochemistry/ Immunofluorescence - Anti-PDHA1 antibody (ab97352)

ab97352, at a 1/200 dilution, staining PDHA1 in paraformaldehyde fixed HeLa cells by Immunofluorescence. Lower image was merged with DNA probe.

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

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