abcam

Product datasheet

HRP Anti-PDHA1 antibody [8D10E6] ab197962

1 图像

概述

产品名称 HRP Anti-PDHA1抗体[8D10E6]

描述 HRP小鼠单克隆抗体[8D10E6] to PDHA1

宿主 Mouse 偶联物 HRP

 经测试应用
 适用于: WB

 种属反应性
 与反应: Human

预测可用于: Mouse, Rat, Cow, Caenorhabditis elegans, Drosophila melanogaster

A

免疫原 Full length native protein (purified) corresponding to Human PDHA1.

阳性对照 WB: Human heart mitochondrial lysate.

常规说明

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

性能

形式 Liquid

存放说明 Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at +4°C.

Stable for 12 months at -20°C. Store In the Dark.

存储溶液 pH: 7.40

Preservative: 0.1% Proclin 300 Solution

Constituents: 30% Glycerol (glycerin, glycerine), 1% BSA, PBS

纯**度** lgG fraction

纯化说明 <u>ab110334</u> was produced in vitro using hybridomas grown in serum-free medium, and then

purified by biochemical fractionation.

克隆 单克隆

克隆编号 8D10E6

1

同种型 IgG1

轻链类型 kappa

应用

The Abpromise guarantee

Abpromise™承诺保证使用ab197962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		1/5000. Detects a band of approximately 43 kDa (predicted molecular weight: 43 kDa).

靶标

功能

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).

组织特异性

疾病相关

Ubiquitous.

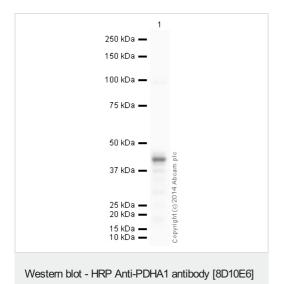
Defects in PDHA1 are a cause of pyruvate decarboxylase E1 component deficiency (PDHE1 deficiency) [MIM:312170]. PDHE1 deficiency is the most common enzyme defect in patients with primary lactic acidosis. It is associated with variable clinical phenotypes ranging from neonatal death to prolonged survival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



(ab197962)

HRP Anti-PDHA1 antibody [8D10E6] (ab197962) at 1/5000 dilution + Human heart tissue lysate - mitochondrial extract (ab110337) at 5 μg

Developed using the ECL technique.

Performed under reducing conditions.

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

Exposure time: 4 minutes

This blot was produced using a 4-12% Bis-tris gel under the MOPS buffer system. The gel was run at 200V for 50 minutes before being transferred onto a Nitrocellulose membrane at 30V for 70 minutes. The membrane was then blocked for an hour using 3% milk before being incubated with ab197962 overnight at 4°C. Antibody binding was visualised using ECL development solution ab133406.

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

Our Abpromise to you: Quality guaranteed and expert technical support

- · Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit https://www.abcam.cn/abpromise or contact our technical team.

Terms and conditions

• Guarantee only valid for products bought direct from Abcam or one of our authorized distributors