


HRP Anti-PDHA1 antibody [9H9AF5] ab197956

1 图像

概述

产品名称	HRP Anti-PDHA1抗体[9H9AF5]
描述	HRP小鼠单克隆抗体[9H9AF5] 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 
免疫原	Recombinant full length protein corresponding to Human PDHA1.
阳性对照	WB: Human heart mitochondrial lysate and HepG2 whole cell lysate.
常规说明	<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&As</p>

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot. Store at +4°C. Avoid freeze / thaw cycle. 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	Affinity purified
纯化说明	ab110330 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	9H9AF5
同种型	IgG1

轻链类型

kappa

应用

The Abpromise guarantee

Abpromise™承诺保证使用ab197956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		1/3000. 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 CO₂. 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

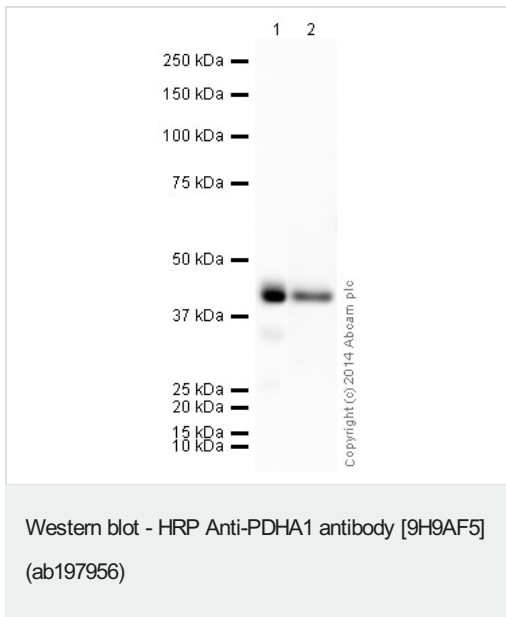
Defects in PDHA1 are a cause of pyruvate decarboxylase E1 component deficiency (PDHE1 deficiency) [MIM:312170]. PDHE1 deficiency is the most common enzyme defect in patients with primary lactic acidosis. It is associated with variable clinical phenotypes ranging from neonatal death to prolonged survival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



All lanes : HRP Anti-PDHA1 antibody [9H9AF5] (ab197956) at 1/3000 dilution

Lane 1 : Human heart tissue lysate - mitochondrial extract (**ab110337**) at 10 µg

Lane 2 : HepG2 (Human hepatocellular liver carcinoma cell line) Whole Cell Lysate at 20 µg

Developed using the ECL technique.

Performed under reducing conditions.

Predicted band size: 43 kDa

Observed band size: 43 kDa

Exposure time: 8 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 ab197956 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"

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