

Product datasheet

Alexa Fluor® 488 Anti-PDHA1 antibody [8D10E6] ab197961

[1 Image](#)

Overview

Product name	Alexa Fluor® 488 Anti-PDHA1 antibody [8D10E6]
Description	Alexa Fluor® 488 Mouse monoclonal [8D10E6] to PDHA1
Host species	Mouse
Conjugation	Alexa Fluor® 488. Ex: 495nm, Em: 519nm
Tested applications	Suitable for: ICC/IF
Species reactivity	Reacts with: Human Predicted to work with: Mouse, Rat, Cow, Caenorhabditis elegans, Drosophila melanogaster



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

Positive control ICC/IF: HeLa cells.

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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

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot. Store at +4°C. Avoid freeze / thaw cycle. Store In the Dark.
Storage buffer	pH: 7.40 Preservative: 0.02% Sodium azide Constituents: 1% BSA, PBS, 30% Glycerol (glycerin, glycerine)
Purity	IgG fraction
Purification notes	ab110334 was produced in vitro using hybridomas grown in serum-free medium, and then purified by biochemical fractionation.
Clonality	Monoclonal
Clone number	8D10E6
Isotype	IgG1
Light chain type	kappa

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab197961 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
ICC/IF		1/100. This product gave a positive signal in HeLa cells fixed with 4% formaldehyde (10 min).

Target

Function 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).

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).

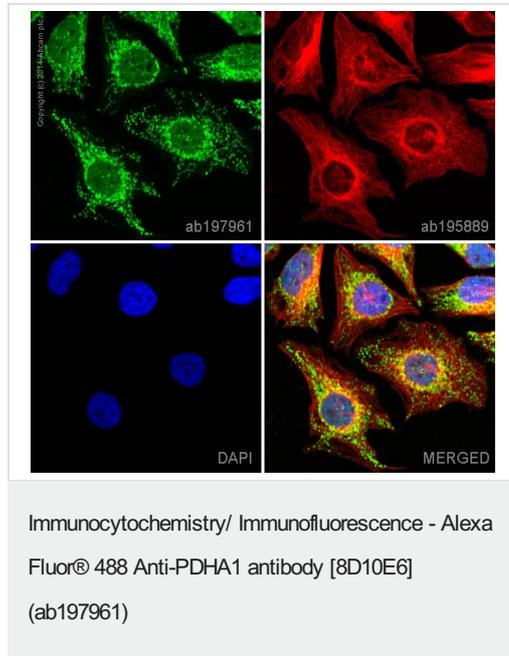
Defects in PDHA1 are the cause of X-linked Leigh syndrome (X-LS) [MIM:308930]. X-LS is an early-onset progressive neurodegenerative disorder with a characteristic neuropathology consisting of focal, bilateral lesions in one or more areas of the central nervous system, including the brainstem, thalamus, 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



ab197961 staining PDHA1 in HeLa cells. The cells were fixed with 4% formaldehyde (10 min), permeabilized with 0.1% Triton X-100 for 5 minutes and then blocked with 1% BSA/10% normal goat serum/0.3M glycine in 0.1%PBS-Tween for 1h. The cells were then incubated overnight at +4°C with ab197961 at a 1/100 dilution (shown in green) and **ab195889**, Mouse monoclonal to alpha Tubulin (Alexa Fluor® 594), at a 1/250 dilution (shown in red). Nuclear DNA was labelled with DAPI (shown in blue).

Image was taken with a confocal microscope (Leica-Microsystems, TCS SP8).

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