

### Anti-PDHA1 antibody [9H9AF5] ab110330

KO 評価済

★★★★★ [8 Abreviews](#) [78 References](#) [画像数 5](#)

#### 製品の概要

製品名	Anti-PDHA1 antibody [9H9AF5]
製品の詳細	Mouse monoclonal [9H9AF5] to PDHA1
由来種	Mouse
アプリケーション	<b>適用あり:</b> WB, ICC/IF, Flow Cyt
種交差性	<b>交差種:</b> Mouse, Rat, Cow, Human
免疫原	Recombinant full length protein corresponding to Human PDHA1.
ポジティブ・コントロール	Isolated mitochondria from Human, Bovine, Rat and Mouse hearts, HepG2 lysate; cultured, normal Human embryonic lung fibroblasts (strain MRC5); HL60 cells.
特記事項	<p>This antibody clone is manufactured by Abcam. If you require a custom buffer formulation or conjugation for your experiments, please contact <a href="mailto:orders@abcam.com">orders@abcam.com</a>.</p> <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> <p>Product was previously marketed under the MitoSciences sub-brand.</p>

#### 製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at +4°C. Do Not Freeze.
バッファー	pH: 7.5 Preservative: 0.02% Sodium azide Constituent: HEPES buffered saline
精製度	IgG fraction
特記事項 (精製)	ab110330 was produced in vitro using hybridomas grown in serum-free medium, and then

	purified by biochemical fractionation.
ポリ/モノ	モノクローナル
クローン名	9H9AF5
アイソタイプ	IgG1
軽鎖の種類	kappa

## アプリケーション

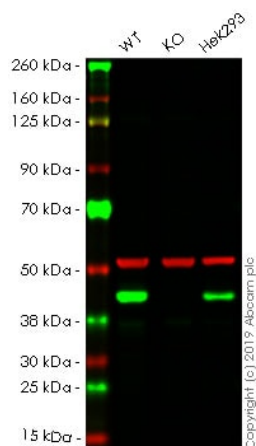
**The Abpromise guarantee** **Abpromise保証は、次のテスト済みアプリケーションにおけるab110330の使用に適用されます**  
 アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
WB	★★★★★ (6)	Use a concentration of 1 µg/ml. Predicted molecular weight: 43 kDa.
ICC/IF	★★★★★ (1)	Use a concentration of 5 µg/ml. (heat-induced antigen-retrieval improves signal)
Flow Cyt		Use a concentration of 1 µg/ml. <b>ab170190</b> - Mouse monoclonal IgG1, is suitable for use as an isotype control with this antibody.

## ターゲット情報

機能	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).
組織特異性	Ubiquitous.
関連疾患	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.
細胞内局在	Mitochondrion matrix.

## 画像



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

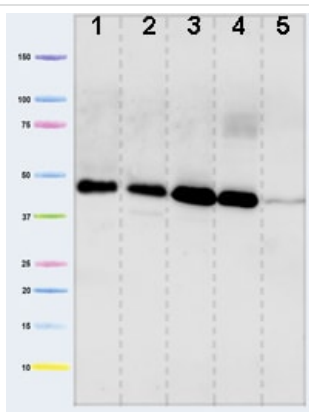
Lysates/proteins at 20 µg per lane.

**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 [ab52866](#) (Rabbit anti alpha Tubulin loading control) were incubated overnight at 4°C at 1 µg/ml and 1/1000 dilution respectively. Blots were developed with Goat anti-Mouse IgG H&L (IRDye® 800CW) preabsorbed [ab216772](#) and Goat anti-Rabbit IgG H&L (IRDye® 680RD) preabsorbed [ab216777](#) 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 µg/ml

**Lane 1 :** Isolated mitochondria from Human heart at 10 µg

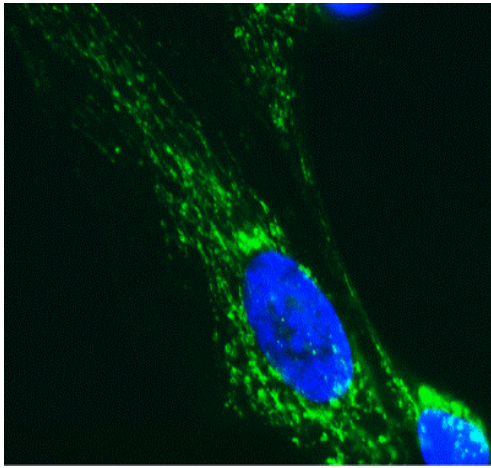
**Lane 2 :** Isolated mitochondria from Bovine heart at 4 µg

**Lane 3 :** Isolated mitochondria from Rat heart at 10 µg

**Lane 4 :** Isolated mitochondria from Mouse heart at 10 µg

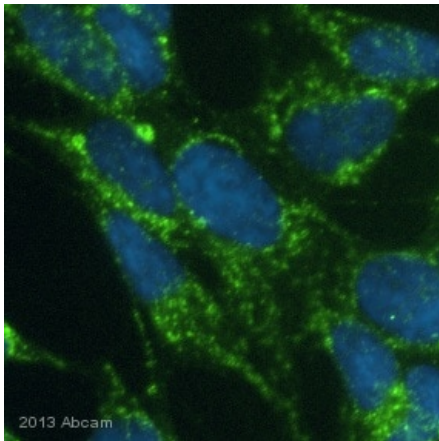
**Lane 5 :** hepG2 cell lysate at 20 µg

**Predicted band size:** 43 kDa



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

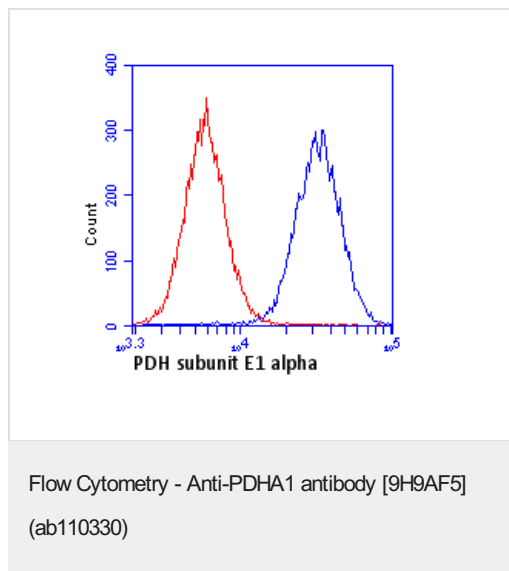
Immunocytochemistry/ Immunofluorescence - Anti-PDHA1 antibody [9H9AF5] (ab110330)



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

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

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



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

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