# abcam

### **Product datasheet**

## Alexa Fluor® 647 Anti-PDHA1 antibody [9H9AF5] ab197653

画像数 1

製品の概要		
製品名	Alexa Fluor® 647 Anti-PDHA1 antibody [9H9AF5]	
製品の詳細	Alexa Fluor® 647 Mouse monoclonal [9H9AF5] to PDHA1	
由来種	Mouse	
標識	Alexa Fluor® 647. Ex: 652nm, Em: 668nm	
アプリケーション	適用あり: ICC/IF	
種交差性	交差種: Human	
免疫原	Recombinant full length protein corresponding to Human PDHA1.	
ポジティブ・コントロール	ICC/IF: HeLa cells	
特記事項	Alexa Fluor <sup>®</sup> is a registered trademark of Molecular Probes, Inc, a Thermo Fisher Scientific Company. The Alexa Fluor <sup>®</sup> dye included in this product is provided under an intellectual property license from Life Technologies Corporation. As this product contains the Alexa Fluor <sup>®</sup> dye, the purchase of this product conveys to the buyer the non-transferable right to use the purchased product and components of the product only in research conducted by the buyer (whether the buyer is an academic or for-profit entity). As this product contains the Alexa Fluor <sup>®</sup> dye the sale of this product is expressly conditioned on the buyer not using the product or its components, or any materials made using the product or its components, in any activity to generate revenue, which may include, but is not limited to use of the product or its components: in manufacturing; (ii) to provide a service, information, or data in return for payment (iii) for therapeutic, diagnostic or prophylactic purposes; or (iv) for resale, regardless of whether they are sold for use in research. For information on purchasing a license to this product for purposes other than research, contact Life Technologies Corporation, 5781 Van Allen Way, Carlsbad, CA 92008 USA or <b>outlicensing@thermofisher.com</b> .	
	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	

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C. Avoid freeze / thaw cycle. Store In the Dark.
バッファー	pH: 7.40 Preservative: 0.02% Sodium azide Constituents: 1% BSA, PBS, 30% Glycerol (glycerin, glycerine)
精製度	lgG fraction
特記事項(精製)	<b>ab110330</b> 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	lgG1
軽鎖の種類	kappa

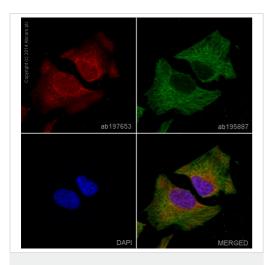
### アプリケーション

### 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ab197653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
ICC/IF		1/100.

ターゲット情報	
機能	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).
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.



ab197653 staining PDHA1 in HeLa cells. The cells were fixed with 100% methanol (5 min), permeabilised in 0.1% Triton X-100 for 5 minutes and then blocked in 1% BSA/10% normal goat serum/0.3M glycine in 0.1%PBS-Tween for 1h. The cells were then incubated with ab197653 at 1/100 dilution (shown in red) and **ab195887**, Mouse monoclonal [DM1A] to alpha Tubulin (Alexa Fluor<sup>®</sup> 488, shown in green) at 1/167 dilution overnight at +4°C. Nuclear DNA was labelled in blue with DAPI.

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

Immunocytochemistry/ Immunofluorescence - Alexa Fluor® 647 Anti-PDHA1 antibody [9H9AF5] (ab197653)

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