

Anti-MTCO3 antibody [DA5BC4] ab110259

★★★★★ **3 Abreviews** **42 References** **1 图像**

概述

产品名称	Anti-MTCO3抗体[DA5BC4]
描述	小鼠单克隆抗体[DA5BC4] to MTCO3
宿主	Mouse
经测试应用	适用于: WB
种属反应性	与反应: Mouse, Rat, Human, Saccharomyces cerevisiae
免疫原	Full length native protein (purified). This information is considered to be commercially sensitive.
阳性对照	Isolated mitochondria from Human , Rat and Mouse hearts.
常规说明	<p>This antibody clone is manufactured by Abcam. If you require a custom buffer formulation or conjugation for your experiments, please contact orders@abcam.com.</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&As</p> <p>Product was previously marketed under the MitoSciences sub-brand.</p>

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at +4°C. Do Not Freeze.
存储溶液	<p>pH: 7.5</p> <p>Preservative: 0.02% Sodium azide</p> <p>Constituent: HEPES buffered saline</p>
纯度	IgG fraction
纯化说明	ab110259 was produced in vitro using hybridomas grown in serum-free medium, and then purified by biochemical fractionation. Near homogeneity ab110259 was judged as near homogeneity by SDS-PAGE.
克隆	单克隆

克隆编号	DA5BC4
同种型	IgG2a
轻链类型	kappa

应用

The Abpromise guarantee **Abpromise™**承诺保证使用ab110259于以下的经测试应用

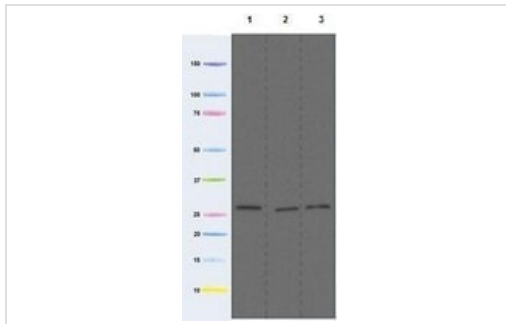
“应用说明”部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
WB	★★★★☆ (3)	Use a concentration of 2 - 4 µg/ml. Predicted molecular weight: 30 kDa. Due to the hydrophobic nature of this protein, it is recommended that the WB is performed on a PVDF membrane with a 90 minute CAPS-buffer transfer.

靶标

功能	Subunits I, II and III form the functional core of the enzyme complex.
疾病相关	Defects in MT-CO3 are a cause of Leber hereditary optic neuropathy (LHON) [MIM:535000]. LHON is a maternally inherited disease resulting in acute or subacute loss of central vision, due to optic nerve dysfunction. Cardiac conduction defects and neurological defects have also been described in some patients. LHON results from primary mitochondrial DNA mutations affecting the respiratory chain complexes. Defects in MT-CO3 are a cause of mitochondrial complex IV deficiency (MT-C4D) [MIM:220110]; also known as cytochrome c oxidase deficiency. A disorder of the mitochondrial respiratory chain with heterogeneous clinical manifestations, ranging from isolated myopathy to severe multisystem disease affecting several tissues and organs. Features include hypertrophic cardiomyopathy, hepatomegaly and liver dysfunction, hypotonia, muscle weakness, exercise intolerance, developmental delay, delayed motor development and mental retardation. A subset of patients manifest Leigh syndrome. Defects in MT-CO3 are associated with recurrent myoglobinuria mitochondrial (RM-MT) [MIM:550500]. Recurrent myoglobinuria is characterized by recurrent attacks of rhabdomyolysis (necrosis or disintegration of skeletal muscle) associated with muscle pain and weakness, and followed by excretion of myoglobin in the urine.
序列相似性	Belongs to the cytochrome c oxidase subunit 3 family.
细胞定位	Mitochondrion inner membrane.

图片



Western blot - Anti-MTCO3 antibody [DA5BC4]
(ab110259)

All lanes : Anti-MTCO3 antibody [DA5BC4] (ab110259) at 4 µg/ml

Lane 1 : Isolated mitochondria from Human heart at 5 µg

Lane 2 : Isolated mitochondria Rat heart at 10 µg

Lane 3 : Isolated mitochondria from Mouse heart at 10 µg

Predicted band size: 30 kDa

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

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