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Product datasheet

Anti-MTCO1 antibody [11D8B7] ab110270

★★★★★ 1 Abreviews 16 References 1 图像

概述

产品名称	Anti-MTCO1抗体[11D8B7]
描述	**小鼠单克隆抗体**[11D8B7] to MTCO1
宿主	Mouse
经测试应用	**适用于:** WB
种属反应性	**与反应:** Saccharomyces cerevisiae **不与反应:** Mouse, Rat, Human
免疫原	Full length protein. This information is considered to be commercially sensitive.
阳性对照	Mitochondria from yeast membrane extract
常规说明	This antibody clone is manufactured by Abcam. If you require a custom buffer formulation or conjugation for your experiments, please contact **orders@abcam.com**. 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 Product was previously marketed under the MitoSciences sub-brand.

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at +4°C. Do Not Freeze.
存储溶液	pH: 7.5 Preservative: 0.02% Sodium azide Constituent: HEPES buffered saline
纯度	IgG fraction
纯化说明	Near homogeneity as judged by SDS-PAGE. ab110270 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	11D8B7
同种型	IgG2b
轻链类型	kappa

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab110270于以下的经测试应用**

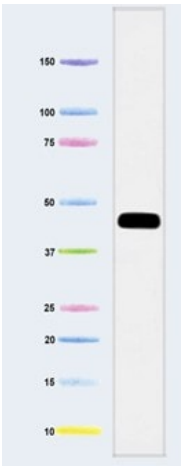
"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
WB	★★★★★ (1)	Use a concentration of 3 µg/ml. Predicted molecular weight: 57 kDa.

靶标

功能	Cytochrome c oxidase is the component of the respiratory chain that catalyzes the reduction of oxygen to water. Subunits 1-3 form the functional core of the enzyme complex. CO I is the catalytic subunit of the enzyme. Electrons originating in cytochrome c are transferred via the copper A center of subunit 2 and heme A of subunit 1 to the bimetallic center formed by heme A3 and copper B.
通路	Energy metabolism; oxidative phosphorylation.
疾病相关	Defects in MT-CO1 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-CO1 are a cause of anemia sideroblastic acquired idiopathic (AISA) [MIM:516030]; a disease characterized by inadequate formation of heme and excessive accumulation of iron in mitochondria. Defects in MT-CO1 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, excercise intolerance, developmental delay, delayed motor development and mental retardation. A subset of patients manifest Leigh syndrome. Defects in MT-CO1 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. Defects in MT-CO1 are a cause of deafness sensorineural mitochondrial (DFNM) [MIM:500008]. DFNM is a form of non-syndromic deafness with maternal inheritance. Affected individuals manifest progressive, postlingual, sensorineural hearing loss involving high frequencies. Defects in MT-CO1 are a cause of colorectal cancer (CRC) [MIM:114500].
序列相似性	Belongs to the heme-copper respiratory oxidase family.
细胞定位	Mitochondrion inner membrane.

图片



Western blot - Anti-MTCO1 antibody [11D8B7] (ab110270)

Anti-MTCO1 antibody [11D8B7] (ab110270) at 3 µg/ml +
Mitochondria from yeast membrane extract

Predicted band size: 57 kDa

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