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Product datasheet

Anti-MTCO1 antibody [11D8B7] ab110270

★★★★★ 1 Abreviews 16 References 画像数 1

製品の概要

製品名 Anti-MTCO1 antibody [11D8B7]

製品の詳細 Mouse monoclonal [11D8B7] to MTCO1

由来種 Mouse

アプリケーション **適用あり**: WB

種交差性 交差種: Saccharomyces cerevisiae

非交差種: Mouse, Rat, Human

免疫原 Full length protein. This information is considered to be commercially sensitive.

ポジティブ・コントロール Mitochondria from yeast membrane extract

特記事項

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.

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) 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

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アプリケーション

The Abpromise guarantee <u>Abpromise保証は、</u>次のテスト済みアプリケーションにおけるab110270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	**** <u>(1)</u>	Use a concentration of 3 µg/ml. Predicted molecular weight: 57 kDa.

ターゲット情報

機能

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.

パスウェイ

関連疾患

Energy metabolism; oxidative phosphorylation.

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.

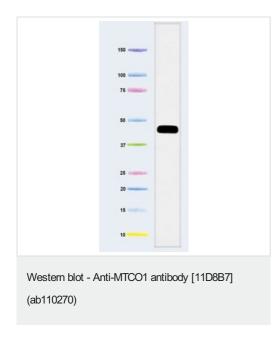
Defects in MT-CO1 are a cause of deafness sensorineural mitochondrial (DFNM) [MIM:500008]. DFNM is a form of non-syndromic deafness with maternal inheritance. Affected individuals manifest progressive, postlingual, sensorineural hearing loss involving high frequencies. Defects in MT-CO1 are a cause of colorectal cancer (CRC) [MIM:114500].

配列類似性

Belongs to the heme-copper respiratory oxidase family.

細胞内局在

Mitochondrion inner membrane.



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

Predicted band size: 57 kDa

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

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