abcam

Product datasheet

HRP Anti-Frataxin antibody [17A11] ab197964

1 图像

概述

产品名称	HRP Anti-Frataxin抗体[17A11]
描述	HRP小鼠单克隆抗体[17A11] to Frataxin
宿主	Mouse
偶联物	HRP
经测试应用	**适用于:** WB
种属反应性	**与反应:** Human **预测可用于:** Mouse, Rat, Cow
免疫原	Recombinant full length protein corresponding to Frataxin.
阳性对照	WB: Human heart mitochondrial lysate and HepG2 whole cell lysate.
常规说明	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

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot. Store at +4°C. Avoid freeze / thaw cycle. Store In the Dark.
存储溶液	pH: 7.40 Preservative: 0.1% Proclin 300 Solution Constituents: 30% Glycerol (glycerin, glycerine), 1% BSA, PBS
纯度	Affinity purified
克隆	单克隆
克隆编号	17A11
同种型	IgG1
轻链类型	kappa

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab197964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		1/5000. Detects a band of approximately 14 kDa (predicted molecular weight: 14 kDa).

靶标

功能 Promotes the biosynthesis of heme and assembly and repair of iron-sulfur clusters by delivering Fe(2+) to proteins involved in these pathways. May play a role in the protection against iron-catalyzed oxidative stress through its ability to catalyze the oxidation of Fe(2+) to Fe(3+); the oligomeric form but not the monomeric form has in vitro ferroxidase activity. May be able to store large amounts of iron in the form of a ferrihydrite mineral by oligomerization; however, the physiological relevance is unsure as reports are conflicting and the function has only been shown using heterologous overexpression systems. Modulates the RNA-binding activity of ACO1.

组织特异性 Expressed in the heart, peripheral blood lymphocytes and dermal fibroblasts.

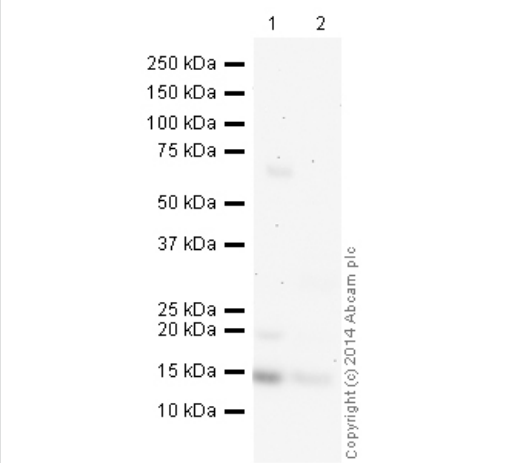
疾病相关 Defects in FXN are the cause of Friedreich ataxia (FRDA) [MIM:229300]. FRDA is an autosomal recessive, progressive degenerative disease characterized by neurodegeneration and cardiomyopathy it is the most common inherited ataxia. The disorder is usually manifest before adolescence and is generally characterized by incoordination of limb movements, dysarthria, nystagmus, diminished or absent tendon reflexes, Babinski sign, impairment of position and vibratory senses, scoliosis, pes cavus, and hammer toe. In most patients, FRDA is due to GAA triplet repeat expansions in the first intron of the frataxin gene. But in some cases the disease is due to mutations in the coding region.

序列相似性 Belongs to the frataxin family.

翻译后修饰 Processed in two steps by mitochondrial processing peptidase (MPP). MPP first cleaves the precursor to intermediate form and subsequently converts the intermediate to yield frataxin mature form (frataxin(81-210)) which is the predominant form. The additional forms, frataxin(56-210) and frataxin(78-210), seem to be produced when the normal maturation process is impaired; their physiological relevance is unsure.

细胞定位 Cytoplasm. Mitochondrion. PubMed:18725397 reports localization exclusively in mitochondria.

图片



Western blot - HRP Anti-Frataxin antibody [17A11] (ab197964)

All lanes : HRP Anti-Frataxin antibody [17A11] (ab197964) at 1/5000 dilution

Lane 1 : Human heart tissue lysate - mitochondrial extract (**ab110337**)
Lane 2 : HepG2 (Human hepatocellular liver carcinoma cell line) Whole Cell Lysate

Lysates/proteins at 20 µg per lane.

Developed using the ECL technique.

Performed under reducing conditions.

Predicted band size: 14 kDa
Observed band size: 14 kDa

Exposure time: 20 minutes

This blot was produced using a 4-12% Bis-tris gel under the MES buffer system. The gel was run at 200V for 35 minutes before being transferred onto a Nitrocellulose membrane at 30V for 70 minutes. The membrane was then blocked for an hour using 3% milk before being incubated with ab197964 overnight at 4°C. Antibody binding was visualised using ECL development solution **ab133406**.

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If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit **https://www.abcam.cn/abpromise** or contact our technical team.

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