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Product datasheet

HRP Anti-Frataxin antibody [17A11] ab197964

画像数1

製品の概要

製品名 HRP Anti-Frataxin antibody [17A11]

製品の詳細 HRP Mouse monoclonal [17A11] to Frataxin

由来種 Mouse 標識 HRP

アプリケーション **適用あり:** WB 種交差性 交差種: Human

交差が予測される動物種: Mouse, Rat, Cow 4

免疫原 Recombinant full length protein corresponding to Frataxin.

ポジティブ・コントロール WB: Human heart mitochondrial lysate and HepG2 whole cell lysate.

特記事項 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

製品の特性

製品の状態

保存方法 Shipped at 4°C. Upon delivery aliquot. Store at +4°C. Avoid freeze / thaw cycle. Store In the Dark.

バッファー pH: 7.40

Preservative: 0.1% Proclin 300 Solution

Constituents: 30% Glycerol (glycerin, glycerine), 1% BSA, PBS

精製度 Affinity purified ポリ/モノ モノクローナル

クローン名 17A11 アイソタイプ lgG1 軽鎖の種類 kappa

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ab197964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		1/5000. Detects a band of approximately 14 kDa (predicted molecular weight: 14 kDa).

ターゲット情報

機能

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.

組織特異性

関連疾患

Expressed in the heart, peripheral blood lymphocytes and dermal fibroblasts.

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.

配列類似性

Belongs to the frataxin family.

翻訳後修飾

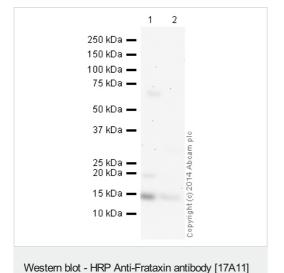
Processed in two steps by mitochondrial processing peptidase (MPP). MPP first cleaves the precursor to intermediate form and subsequently converts the intermediate to yield frataxin mature form (frataxin(81-210)) which is the predominant form. The additional forms, frataxin(56-210) and frataxin(78-210),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

physiological relevance is unsure.

細胞内局在

Cytoplasm. Mitochondrion. PubMed:18725397 reports localization exclusively in mitochondria.

画像



(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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