abcam

Product datasheet

HRP Anti-Frataxin antibody [17A11] ab197964

1 Image

Overview

Product name HRP Anti-Frataxin antibody [17A11]

Description HRP Mouse monoclonal [17A11] to Frataxin

Host species Mouse
Conjugation HRP

Tested applications Suitable for: WB

Species reactivity Reacts with: Human

Predicted to work with: Mouse, Rat, Cow

Immunogen Recombinant full length protein corresponding to Frataxin.

Positive control WB: Human heart mitochondrial lysate and HepG2 whole cell lysate.

General notes

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

Properties

Form Liquid

Storage instructions Shipped at 4°C. Upon delivery aliquot. Store at +4°C. Avoid freeze / thaw cycle. Store In the Dark.

Storage buffer pH: 7.40

Preservative: 0.1% Proclin 300 Solution

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

Purity Affinity purified

Clonality Monoclonal

Clone number 17A11

Isotype IgG1

Light chain type kappa

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Applications

The Abpromise guarantee

Our Abpromise guarantee covers the use of ab197964 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

| Application | Abreviews | Notes |
|-------------|-----------|--|
| WB | | 1/5000. Detects a band of approximately 14 kDa (predicted molecular weight: 14 kDa). |

Target

Function

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.

Tissue specificity

Involvement in disease

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.

Sequence similarities

Post-translational modifications

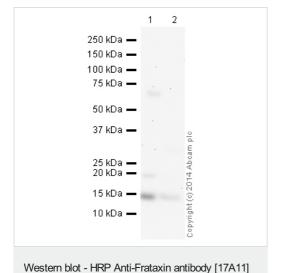
Belongs to the frataxin family.

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.

Cellular localization

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

Images



(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**.

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

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