abcam

Product datasheet

Anti-Frataxin antibody [17A11] ab113691

* ★ ★ ★ ★ ★ 3 Abreviews 16 References 4 Images

Overview

Product name Anti-Frataxin antibody [17A11]

Description Mouse monoclonal [17A11] to Frataxin

Host species Mouse

Tested applications Suitable for: WB, ICC/IF, Flow Cyt, IHC-P

Species reactivity Reacts with: Mouse, Rat, Cow, Human, Recombinant fragment

Immunogen Recombinant fragment within Human Frataxin aa 1-120. The exact sequence is proprietary.

Database link: Q16595

Positive control WB: heart, liver tissue lysate. IHC-P: heart muscle, liver, kidney tissue (FFPE).

General notes

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.

Properties

Form Liquid

Storage instructions Shipped at 4°C. Store at +4°C.

Storage buffer pH: 7.5

Preservative: 0.02% Sodium azide Constituent: HEPES buffered saline

Purification notes The antibody was produced in vitro using hybridomas grown in serum-free medium, and then

purified by affinity purification.

Clonality Monoclonal

Clone number 17A11

1

lsotype lgG1 **Light chain type** kappa

Applications

The Abpromise guarantee

Our Abpromise quarantee covers the use of ab113691 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	★★★★☆ (2)	Use a concentration of 4 µg/ml. Detects a band of approximately 14 kDa (predicted molecular weight: 14 kDa).
ICC/IF	**** <u>(1)</u>	Use a concentration of 1 µg/ml.
Flow Cyt		Use a concentration of 1 µg/ml. <u>ab170190</u> - Mouse monoclonal lgG1, is suitable for use as an isotype control with this antibody.
IHC-P		Use a concentration of 10 µg/ml.

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

Belongs to the frataxin family.

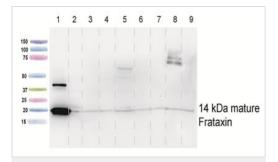
Post-translational modifications

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



Western blot - Anti-Frataxin antibody [17A11] (ab113691)

All lanes: Anti-Frataxin antibody [17A11] (ab113691)

Lane 1: Recombinant frataxin

at 0.1 µg

Lane 2: Human heart mitochondria

at 15 µg

Lane 3: Human liver mitochondria

at 15 µg

Lane 4: HepG2 whole cell lysate

at 15 µg

Lane 5: Bovine heart mitochondria

at 15 µg

Lane 6: H9C2 rat whole cell lysate

at 15 µg

Lane 7: MEF mouse embryonic fibroblast

at 15 µq

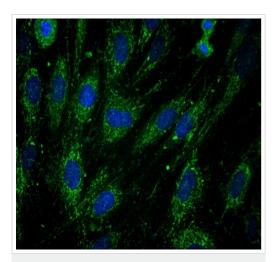
Lane 8: Mouse liver mitochondria

at 15 µg

Lane 9: Rat liver mitochondria

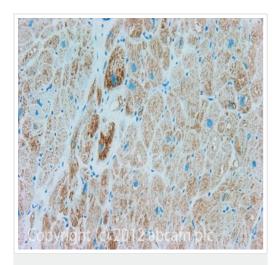
at 15 µg

Predicted band size: 14 kDa



Immunocytochemistry/ Immunofluorescence - Anti-Frataxin antibody [17A11] (ab113691)

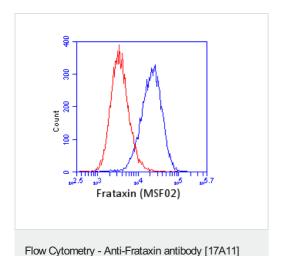
ab113691 stained fibroblast cells. The cells were paraformaldehyde fixed (4%, 20 min) and Triton X-100 permeabilized (0.1%, 15min). The cells were incubated with the antibody (ab113691, 1µg/ml) for 2h at room temperature or over night at 4°C. The secondary antibody was (green) Alexa Fluor® 488 goat anti-mouse lgG (H+L) used at a 1/1000 dilution for 1h. 10% Goat serum was used as the blocking agent for all blocking steps. The target protein locates to the mitochondrial matrix.



Immunohistochemistry (Formalin/PFA-fixed paraffinembedded sections) - Anti-Frataxin antibody
[17A11] (ab113691)

IHC image of Frataxin staining in Human normal heart muscle formalin fixed paraffin embedded tissue section, performed on a Leica BondTM system using the standard protocol F. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH6, epitope retrieval solution 1) for 20 mins. The section was then incubated with ab113691, 10µg/ml, for 15 mins at room temperature and detected using an HRP conjugated compact polymer system. DAB was used as the chromogen. The section was then counterstained with haematoxylin and mounted with DPX.

For other IHC staining systems (automated and non-automated) customers should optimize variable parameters such as antigen retrieval conditions, primary antibody concentration and antibody incubation times.



(ab113691)

Normal human lymphoblast cells were stained with 1 μ g/mL ab113691 (blue) or an equal amount of an isotype control antibody (red) and analyzed by flow cytometry.

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

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- · Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

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.com/abpromise or contact our technical team.

Terms and conditions

Guarantee only valid for products bought direct from Abcam or one of our authorized distributors