# abcam

#### Product datasheet

## Anti-Frataxin antibody [17A11] ab113691

★★★★★ 3 Abreviews 16 References 4 图像

#### 概述

产**品名称** Anti-Frataxin抗体[17A11]

**小**鼠单**克隆抗体**[17A11] to Frataxin

**宿主** Mouse

经测试应用 适用于: WB, ICC/IF, Flow Cyt, IHC-P

种属反应性 与反应: Mouse, Rat, Cow, Human, Recombinant fragment

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

Database link: Q16595

阳性对照 WB: heart, liver tissue lysate. IHC-P: heart muscle, liver, kidney tissue (FFPE).

常规说明

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.

#### 性能

形式 Liquid

**存放说明** Shipped at 4°C. Store at +4°C.

**存储溶液** pH: 7.5

Preservative: 0.02% Sodium azide Constituent: HEPES buffered saline

纯**化说明** The antibody was produced in vitro using hybridomas grown in serum-free medium, and then

purified by affinity purification.

 克隆
 单克隆

 克隆编号
 17A11

1

同种型 lgG1

轻链类型 kappa

#### 应用

The Abpromise guarantee Abpromise™承诺保证使用ab113691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	* * * * * <u>(2)</u>	Use a concentration of 4 µg/ml. Detects a band of approximately 14 kDa (predicted molecular weight: 14 kDa).
ICC/IF	*** <u>*</u>	Use a concentration of 1 µg/ml.
Flow Cyt		Use a concentration of 1 $\mu$ g/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.

#### 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

疾病相关

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.

序列相似性

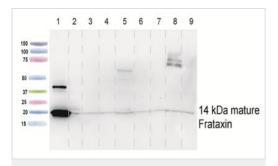
Belongs to the frataxin family.

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.



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

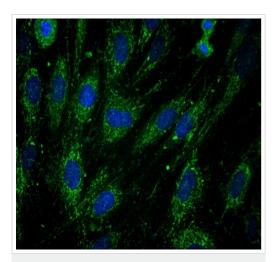
Lane 8: Mouse liver mitochondria

at 15 µg

Lane 9: Rat liver mitochondria

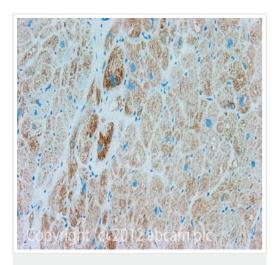
at 15 µg

Predicted band size: 14 kDa



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

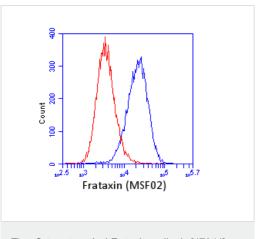
ab113691 stained fibroblast cells. The cells were paraformaldehyde fixed (4%, 20 min) and Triton X-100 permeabilized (0.1%, 15min). The cells were incubated with the antibody (ab113691, 1µg/ml) for 2h at room temperature or over night at 4°C. The secondary antibody was (green) Alexa Fluor® 488 goat anti-mouse 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.



Flow Cytometry - Anti-Frataxin antibody [17A11] (ab113691)

Normal human lymphoblast cells were stained with 1  $\mu$ 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"

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