

### Anti-Frataxin antibody [18A5DB1] ab110328

★★★★★ [5 Abreviews](#) [36 References](#) [5 图像](#)

#### 概述

产品名称	Anti-Frataxin抗体[18A5DB1]
描述	小鼠单克隆抗体[18A5DB1] 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: Human
免疫原	Recombinant full length protein. This information is considered to be commercially sensitive.
阳性对照	Recombinant Human Frataxin; normal Human fibroblasts (MRC5); HL60 cells. This antibody gave a positive result in IHC in the following FFPE tissue: Human colon adenocarcinoma.
常规说明	<p>Although this antibody does work in Western blot, we have found that <a href="#">ab113691</a> typically gives a more robust signal. Therefore, if you plan on using this antibody in WB only, we recommend <a href="#">ab113691</a> as an alternative.</p> <p>This antibody clone is manufactured by Abcam. If you require a custom buffer formulation or conjugation for your experiments, please contact <a href="mailto:orders@abcam.com">orders@abcam.com</a>.</p> <p>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.</p> <p>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&amp;As</p> <p>Product was previously marketed under the MitoSciences sub-brand.</p>

#### 性能

形式	Liquid
存放说明	Shipped at 4°C. Store at +4°C. Do Not Freeze.
存储溶液	<p>pH: 7.5</p> <p>Preservative: 0.02% Sodium azide</p> <p>Constituent: HEPES buffered saline</p>
纯度	IgG fraction

纯化说明	ab110328 was produced in vitro using hybridomas grown in serum-free medium, and then purified by biochemical fractionation.
克隆	单克隆
克隆编号	18A5DB1
同种型	IgG1
轻链类型	kappa

应用

**The Abpromise guarantee**      **Abpromise™**承诺保证使用ab110328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	★★★★☆ (4)	Use a concentration of 3 - 5 µg/ml. Predicted molecular weight: 23 kDa. (A chemiluminescent detection system is highly recommended)
ICC/IF	★★★★☆ (1)	Use a concentration of 0.5 µg/ml.
Flow Cyt		Use a concentration of 1 µg/ml. <b>ab170190</b> - Mouse monoclonal IgG1, is suitable for use as an isotype control with this antibody.
IHC-P		Use a concentration of 10 µg/ml.

靶标

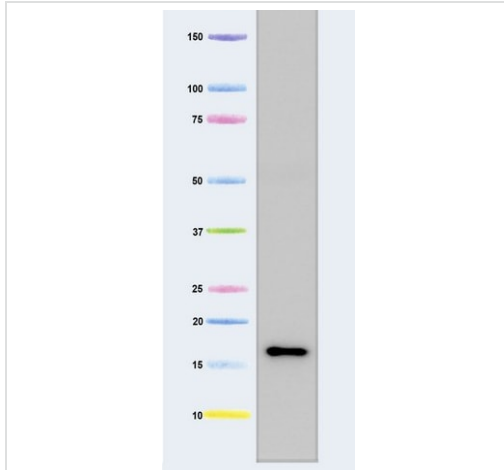
功能	Promotes the biosynthesis of heme and assembly and repair of iron-sulfur clusters by delivering Fe(2+) to proteins involved in these pathways. May play a role in the protection against iron-catalyzed oxidative stress through its ability to catalyze the oxidation of Fe(2+) to Fe(3+); the oligomeric 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	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.

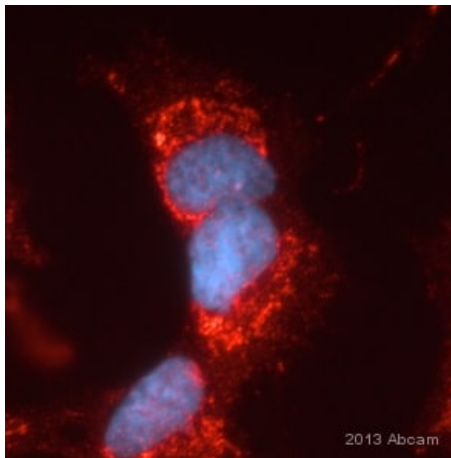
## 图片



Western blot - Anti-Frataxin antibody [18A5DB1]  
(ab110328)

Anti-Frataxin antibody [18A5DB1] (ab110328) at 5 µg/ml +  
Recombinant Human Frataxin at 0.0005 µg

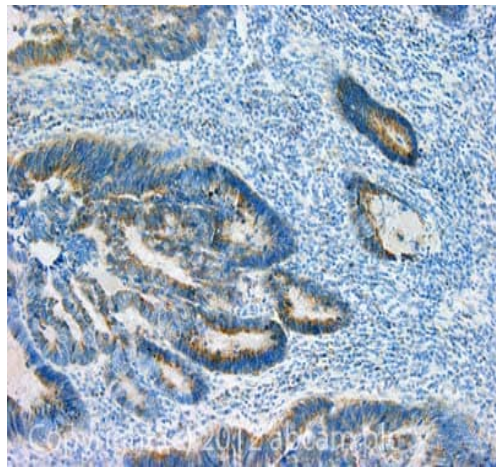
**Predicted band size:** 23 kDa



Immunocytochemistry/ Immunofluorescence - Anti-  
Frataxin antibody [18A5DB1] (ab110328)

Image courtesy of Dr George Allen

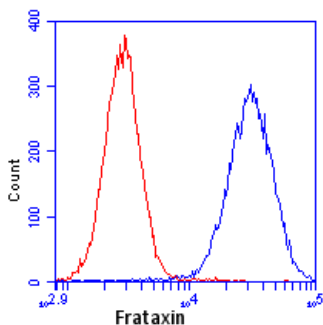
Immunofluorescent analysis of paraformaldehyde-fixed, 0.2% NP-40 permeabilized SH-SY5Y (human neuroblastoma cell line from bone marrow) cells labeling Frataxin with ab110328 at 1/500 dilution, followed by Goat anti-MouseAlexaFluor® 594 conjugated secondary antibody at 1/500 dilution (red). The nuclear counter stain is DAPI (blue).



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Frataxin antibody [18A5DB1] (ab110328)

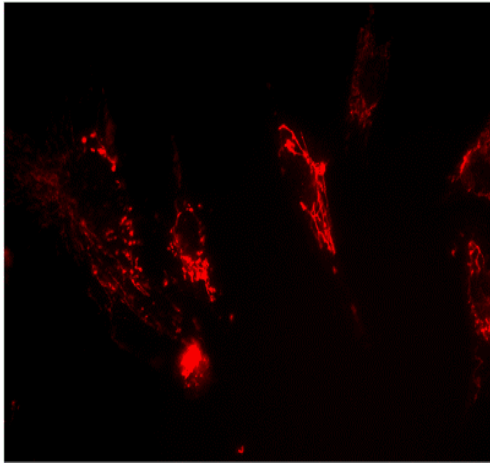
IHC image of Frataxin staining in Human colon adenocarcinoma formalin fixed paraffin embedded tissue section, performed on a Leica Bond™ system using the standard protocol F. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH6, epitope retrieval solution 1) for 20 mins. The section was then incubated with ab110328, 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 [18A5DB1] (ab110328)

Flow cytometric analysis using ab110328 at 1µg/ml staining Frataxin in HL60 cells (blue). Isotype control antibody (red).



Immunocytochemistry/ Immunofluorescence - Anti-Frataxin antibody [18A5DB1] (ab110328)

Immunocytochemistry analysis using ab110328 at 0.5µg/ml staining Frataxin in normal Human fibroblasts (MRC5) (fixed and permeabilized) followed by an AlexaFluor® 594-conjugated-goat-anti-mouse IgG1 isotype specific secondary antibody (2 µg/ml). .

**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.cn/abpromise> or contact our technical team.

### Terms and conditions

---

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