# abcam

# Product datasheet

# Anti-Frataxin antibody [EPR6107] - BSA and Azide free ab244232



RabMAb

# 5 Images

#### Overview

Product name Anti-Frataxin antibody [EPR6107] - BSA and Azide free

**Description** Rabbit monoclonal [EPR6107] to Frataxin - BSA and Azide free

Host species Rabbit

Specificity We suggest optimizing experimental protocols (increasing lysate amount, using lower dilution or

higher sensitivity ECL substrate) to improve results.

Tested applications Suitable for: WB

Unsuitable for: IHC-P

Species reactivity Reacts with: Human

**Immunogen** Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.

Positive control WB: Human fetal heart tissue, human skin, His tagged Human FXN recombinant protein (aa 82-

210), Human liver and hippocampus tissues, HCT116, Huh7 and SH-SY5Y whole cell lysates.

**General notes** ab244232 is the carrier-free version of <u>ab124680</u>.

Our <u>carrier-free</u> antibodies are typically supplied in a PBS-only formulation, purified and free of BSA, sodium azide and glycerol. The carrier-free buffer and high concentration allow for

increased conjugation efficiency.

This conjugation-ready format is designed for use with fluorochromes, metal isotopes, oligonucleotides, and enzymes, which makes them ideal for antibody labelling, functional and cell-based assays, flow-based assays (e.g. mass cytometry) and Multiplex Imaging applications.

Use our <u>conjugation kits</u> for antibody conjugates that are ready-to-use in as little as 20 minutes with <1 minute hands-on-time and 100% antibody recovery: available for fluorescent dyes, HRP, biotin and gold.

This product is compatible with the Maxpar<sup>®</sup> Antibody Labeling Kit from Fluidigm, without the need for antibody preparation. Maxpar<sup>®</sup> is a trademark of Fluidigm Canada Inc.

This product is a recombinant monoclonal antibody, which offers several advantages including:

- High batch-to-batch consistency and reproducibility
- Improved sensitivity and specificity
- Long-term security of supply
- Animal-free production

1

For more information see here.

Our RabMAb<sup>®</sup> technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to **RabMAb**<sup>®</sup> **patents**.

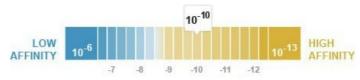
Mouse, Rat: We have preliminary internal testing data to indicate this antibody may not react with these species. Please contact us for more information.

#### **Properties**

Form Liquid

**Storage instructions** Shipped at 4°C. Store at +4°C. Do Not Freeze.

**Dissociation constant (K<sub>D</sub>)**  $K_D = 3.19 \times 10^{-10} M$ 



Learn more about K<sub>D</sub>

**Storage buffer** pH: 7.2

Constituent: PBS

Carrier free Yes

Purity Protein A purified

ClonalityMonoclonalClone numberEPR6107

**Isotype** IgG

# **Applications**

#### The Abpromise guarantee

Our **Abpromise guarantee** covers the use of ab244232 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          |           | Use at an assay dependent concentration. Detects a band of approximately 17, 30 kDa (predicted molecular weight: 19, 23 kDa). |

**Application notes** Is unsuitable for IHC-P.

# **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

# Tissue specificity

#### Involvement in disease

using heterologous overexpression systems. Modulates the RNA-binding activity of ACO1.

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

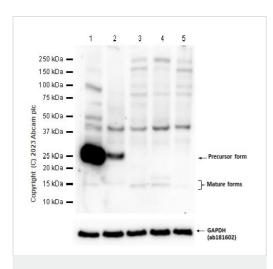
Belongs to the frataxin family.

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**



Western blot - Anti-Frataxin antibody [EPR6107] - BSA and Azide free (ab244232)

**All lanes :** Anti-Frataxin antibody [EPR6107] (<u>ab124680</u>) at 1/1000 dilution

Lane 1: Human liver tissue lysate

Lane 2: Human hippocampus tissue lysate

Lane 3: HCT116 (human colorectal carcinoma epithelial cell)

whole cell lysate

Lane 4: Huh7 (human hepatocellular carcinoma epithelial cell)

whole cell lysate

Lane 5: SH-SY5Y (human neuroblastoma epithelial cell) whole cell

lysate

Lysates/proteins at 20 µg per lane.

### Secondary

All lanes: Goat Anti-Rabbit lgG H&L (HRP) (ab97051) at 1/20000

dilution

**Predicted band size:** 19, 23 kDa **Observed band size:** 14,17 kDa

Exposure time: 60 seconds

Blocking buffer and concentration: 5% NFDM/TBST.

Diluting buffer and concentration: 5% NFDM/TBST.

This blot was developed using a high sensitivity ECL substrate.

For different forms of frataxin, you can refer to PMID: 17468497,

PMID: 31279523, PMID: 17468497 etc.

ab181602 was used as a loading control.

This data was developed using <u>ab124680</u>, the same antibody clone in a different buffer formulation.

Anti-Frataxin antibody [EPR6107] (ab124680) at 1/1000 dilution + His tagged Human FXN recombinant protein (aa 82-210), 10ng

# Secondary

Goat Anti-Rabbit IgG H&L (HRP) (ab97051) at 1/20000 dilution

**Predicted band size:** 19, 23 kDa **Observed band size:** 16 kDa

Exposure time: 60 seconds

This data was developed using <u>ab124680</u>, the same antibody clone in a different buffer formulation.

Blocking and diluting buffer and concentration: 5% NFDM/TBST.

ab213204 was used as a loading control.

**All lanes :** Anti-Frataxin antibody [EPR6107] (ab124680) at 1/5000 dilution (purified)

Lane 1: Human fetal heart tissue lysate

Lane 2: Human skin lysate

Lysates/proteins at 20 µg per lane.

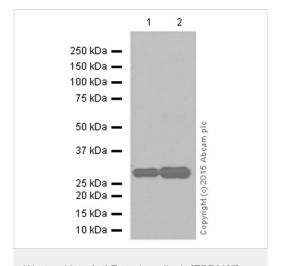
#### Secondary

**All lanes :** Anti-Rabbit IgG (HRP), specific to the non-reduced form of IgG at 1/2000 dilution

**Predicted band size:** 19, 23 kDa **Observed band size:** 30 kDa



Western blot - Anti-Frataxin antibody [EPR6107] - BSA and Azide free (ab244232)



Western blot - Anti-Frataxin antibody [EPR6107] - BSA and Azide free (ab244232)

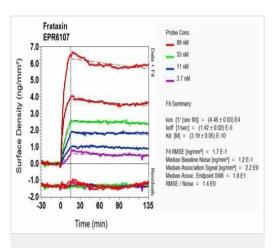
This data was developed using <u>ab124680</u>, the same antibody clone in a different buffer formulation.

Blocking and dilution buffer: 5% NFDM/TBST.

This data was developed using  $\underline{ab124680}$ , the same antibody clone in a different buffer formulation. Equilibrium disassociation constant ( $K_D$ )

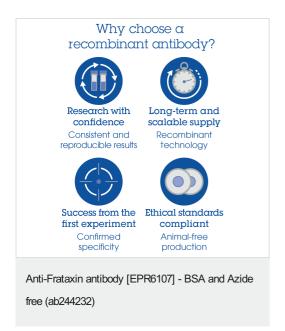
Learn more about KD

Click here to learn more about K<sub>D</sub>



OI-RD Scanning - Anti-Frataxin antibody [EPR6107]

- BSA and Azide free (ab244232)



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 <a href="https://www.abcam.com/abpromise">https://www.abcam.com/abpromise</a> or contact our technical team.

#### Terms and conditions

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