# abcam

# Product datasheet

# Mouse Frataxin ELISA Kit, Fluorescent ab229424

CatchPoint SimpleStep ELISA

4 Images

Overview

**Precision** 

Recovery

Product name Mouse Frataxin ELISA Kit, Fluorescent

**Detection method** Fluorescent

Detection metriod indicacer

Sample	n	Mean	SD	CV%
cell extract	3			1.3%

Inter-assay

Sample specific recovery

Intra-assay

Sample	n	Mean	SD	CV%	
cell extract	3			2%	

Sample type Cell culture extracts, Tissue Extracts

Assay type Sandwich (quantitative)

Sensitivity 4.8 pg/ml

**Range** 7.8 pg/ml - 16000 pg/ml

Sample type	Average %	Range
Serum	100	95% - 109%
Fetal Bovine Serum	108	95% - 126%

Assay time 1h 30m

**Assay duration** One step assay

Species reactivity Reacts with: Mouse

Product overview Frataxin in vitro CatchPoint SimpleStep ELISA (Enzyme-Linked Immunosorbent Assay) kit is

designed for the quantitative measurement of Frataxin protein in mouse cell and tissue extracts.

This CatchPoint SimpleStep ELISA kit has been optimized for Molecular Devices Microplate

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**Readers**. Click **here** for a list of recommended Microplate Readers.

If using a Molecular Devices' plate reader supported by SoftMax® Pro software, a preconfigured protocol for these CatchPoint SimpleStep ELISA Kits is available with all the protocol and analysis settings at www.softmaxpro.org.

The CatchPoint SimpleStep ELISA employs an affinity tag labeled capture antibody and a reporter conjugated detector antibody which immunocapture the sample analyte in solution. This entire complex (capture antibody/analyte/detector antibody) is in turn immobilized via immunoaffinity of an anti-tag antibody coating the well. To perform the assay, samples or standards are added to the wells, followed by the antibody mix. After incubation, the wells are washed to remove unbound material. CatchPoint HRP Development Solution containing the Stoplight Red Substrate is added. During incubation, the substrate is catalyzed by HRP generating a fluorescent product. Signal is generated proportionally to the amount of bound analyte and the intensity is measured in a fluorescence plater reader at 530/570/590 nm Excitation/Cutoff/Emission.

Mouse Frataxin is a 23 kDa nuclear-encoded mitochondrial protein. The gene is localized on chromosome 9 and is highly conserved during evolution. The gene is expressed in every cell, although in varying levels in different tissues and during development. The specific function of frataxin is still unknown, but it has been shown to play a role in iron metabolism. Studies have demonstrated that the deletion of the frataxin gene in yeast results in iron accumulation in mitochondria and loss of respiration. Recombinant frataxin has been shown to bind iron in vitro, and increased mitochondrial iron levels have been observed in patients with Friedreich's Ataxia (FRDA). FRDA is an autosomal recessive, progressive degenerative disease characterized by neurodegeneration and cardiomyopathy it is the most common inherited ataxia.

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**Platform** 

**Notes** 

Pre-coated microplate (12 x 8 well strips)

#### **Properties**

#### Storage instructions

Store at +4°C. Please refer to protocols.

Components	1 x 96 tests
100X Stoplight Red Substrate	1 x 120µl
10X Mouse Frataxin Capture Antibody	1 x 600µl
10X Mouse Frataxin Detector Antibody	1 x 600µl
10X Wash Buffer PT (ab206977)	1 x 20ml
500X Hydrogen Peroxide (H2O2, 3%)	1 x 50µl
50X Cell Extraction Enhancer Solution (ab193971)	1 x 1ml
5X Cell Extraction Buffer PTR (ab193970)	1 x 10ml

Components	1 x 96 tests
Antibody Diluent 5BI	1 x 6ml
Mouse Frataxin Lyophilized Recombinant Protein	2 vials
Plate Seals	1 unit
Sample Diluent NS (ab193972)	1 x 50ml
SimpleStep Pre-Coated Black 96-Well Microplate	1 unit
Stoplight Red Substrate Buffer	1 x 12ml

#### **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

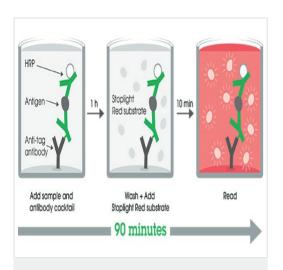
Belongs to the frataxin family.

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**

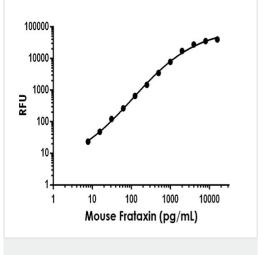
Cytoplasm. Mitochondrion. PubMed:18725397 reports localization exclusively in mitochondria.

## **Images**



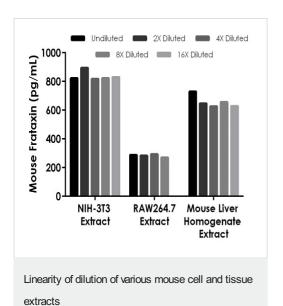
SimpleStep ELISA technology allows the formation of the antibodyantigen complex in one single step, reducing assay time to 90 minutes. Add samples or standards and antibody mix to wells all at once, incubate, wash, and add your final substrate. See protocol for a detailed step-by-step guide.



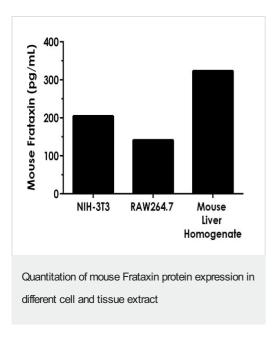


Example of mouse Frataxin standard curve in 1X Cell Extraction Buffer PTR

The Frataxin standard curve was prepared as described in Section 10.



NIHI3T3, RAW264.7 and, mouse liver extracts were prepared as described previously and interpolated values corrected by dilution factor (mean +/- SD) are graphed.



Interpolated values of mouse Frataxin protein are plotted for the indicated cell and tissue extract based on an extract load of 1,250 ng/mL.

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