

# Human Frataxin ELISA Kit (10 x 96 well plate) ab201121

SimpleStep ELISA

## 5 图像

### 概述

**产品名称** 人Frataxin ELISA试剂盒(10 x 96 well plate)

**检测方法** Colorimetric

**精确度** 批次内

样品	n	Mean	SD	CV%
HeLa	9			2.3%

批次间

样品	n	Mean	SD	CV%
HeLa	3			6.3%

**样品类型** Cell culture extracts, Tissue Extracts

**检测类型** Sandwich (quantitative)

**灵敏度** 2.1 pg/ml

**范围** 9.88 pg/ml - 800 pg/ml

**回收率** 特定样本回收率

样品类型	平均%	范围
Serum	92	86% - 98%
Cell culture media	97	93% - 101%
EDTA Plasma	89	88% - 92%

**检测时间** 1h 30m

**实验步骤** One step assay

**种属反应性** 与反应: Human

**产品概述** Abcam's Frataxin in vitro SimpleStep ELISA® (Enzyme-Linked Immunosorbent Assay) kit is designed for the quantitative measurement of Frataxin protein in in human cell and tissue extracts.

The 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. TMB substrate is added and during incubation is catalyzed by HRP, generating blue coloration. This reaction is then stopped by addition of Stop Solution completing any color change from blue to yellow. Signal is generated proportionally to the amount of bound analyte and the intensity is measured at 450 nm. Optionally, instead of the endpoint reading, development of TMB can be recorded kinetically at 600 nm.

**说明**

Frataxin is a 17 kDa nuclear-encoded mitochondrial protein. In humans 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 human 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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**平台**

Microplate

**性能**

**存放说明**

Please refer to protocols.

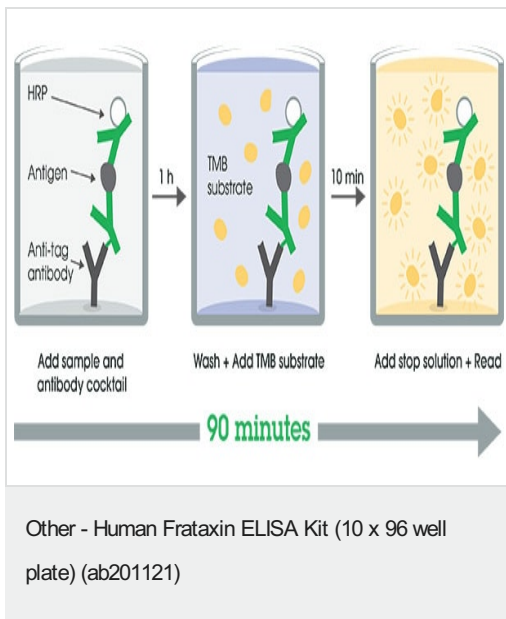
组件	10 x 96 tests
10X Human Frataxin Capture Antibody	1 x 6ml
10X Human Frataxin Detector Antibody	1 x 6ml
10X Wash Buffer PT (ab206977)	1 x 200ml
50X Cell Extraction Enhancer Solution (ab193971)	1 x 5ml
5X Cell Extraction Buffer PTR (ab193970)	1 x 50ml
Antibody Diluent 4BI	1 x 60ml
Human Frataxin Recombinant Protein	1 x 2ml
Plate Seals	10 units

组件	10 x 96 tests
Sample Diluent NS (ab193972)	1 x 120ml
SimpleStep Pre-Coated 96-Well Microplate (ab206978)	10 units
Stop Solution	1 x 120ml
TMB Development Solution	1 x 120ml

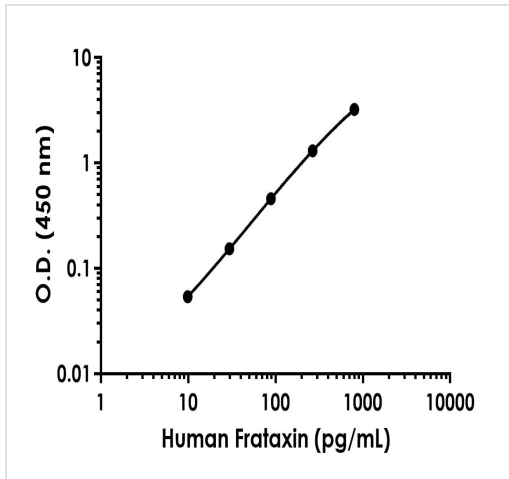
**相关性** Function: Promotes the biosynthesis of heme and assembly and repair of iron-sulfur clusters by delivering Fe<sup>2+</sup> to proteins involved in these pathways. May play a role in the protection against iron-catalyzed oxidative stress through its ability to catalyze the oxidation of Fe<sup>2+</sup> to Fe<sup>3+</sup>; 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.

**细胞定位** Cytoplasmic and Mitochondrial

**图片**

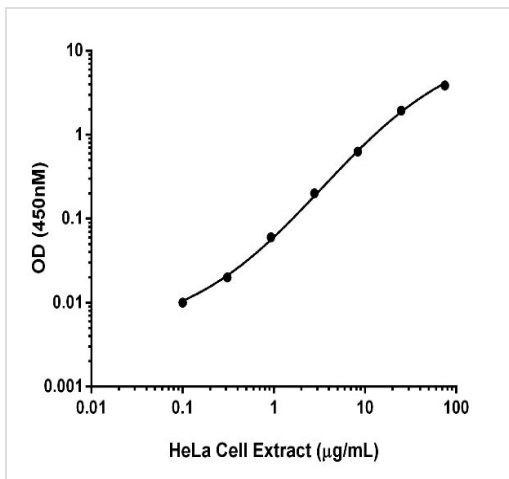


ELISA Protocol Summary



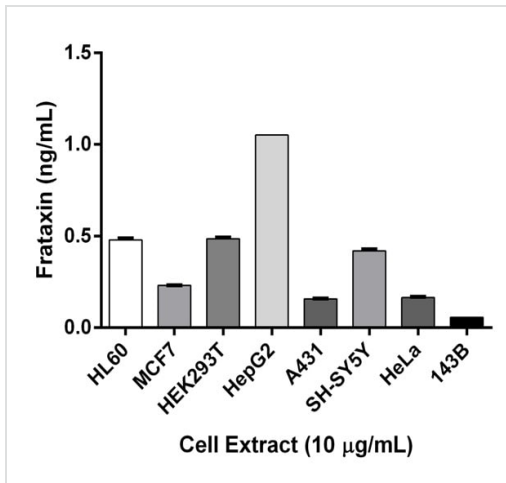
Background-subtracted data values (mean +/- SD) are graphed.

Example of Frataxin standard curve



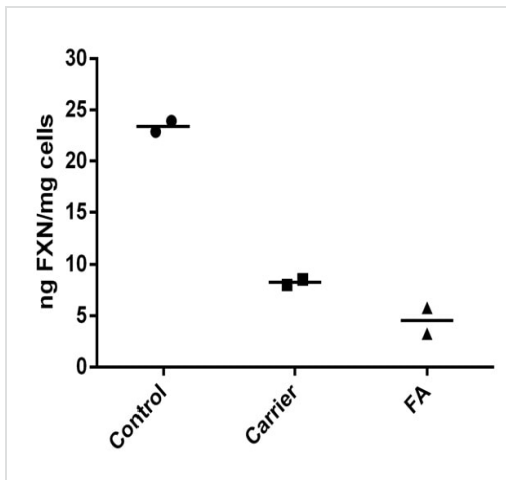
Background subtracted data from duplicate measurements are plotted.

Titration of HeLa cell extract within the working range of the assay



Interpolated values of Frataxin are plotted for the indicated cell lines based on a extract load of 10 µg/mL.

Quantification of Frataxin expression in different cell lines



Transformed B lymphocyte cells from Friedreich's Ataxia (FRDA) samples, due to a stable homozygous GAA repeat insertion, are compared to heterozygous carrier B lymphocyte cells and control B lymphocyte cells. B lymphocyte cell extracts were analyzed across a 7-point titration (0.1||100 µg/mL) and frataxin levels were interpolated from the standard curve. Average interpolated values of Frataxin are plotted.

Quantification of Frataxin expression in different human patient samples

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