

Anti-Frataxin Antibody
Rabbit polyclonal antibody to Frataxin
Catalog # AP61627**Specification**

Anti-Frataxin Antibody - Product Information

Application	WB, IF/IC, IHC
Primary Accession	Q16595
Other Accession	O35943
Reactivity	Human, Mouse, Rat
Host	Rabbit
Clonality	Polyclonal
Calculated MW	23135

Anti-Frataxin Antibody - Additional Information**Gene ID** 2395**Other Names**

FRDA; X25; Frataxin mitochondrial; Friedreich ataxia protein; Fxn

Target/Specificity

Recognizes endogenous levels of Frataxin protein.

Dilution

WB~~1:1000

IF/IC~~N/A

IHC~~1:100~500

Format

Liquid in 0.42% Potassium phosphate, 0.87% Sodium chloride, pH 7.3, 30% glycerol, and 0.09% (W/V) sodium azide.

Storage

Store at -20 °C. Stable for 12 months from date of receipt

Anti-Frataxin Antibody - Protein Information**Name** FXN ([HGNC:3951](#))**Synonyms** FRDA, X25**Function**

[Frataxin mature form]: Functions as an activator of persulfide transfer to the scaffolding protein ISCU as component of the core iron-sulfur cluster (ISC) assembly complex and participates to the [2Fe-2S] cluster assembly (PubMed:12785837, PubMed:24971490). Accelerates sulfur transfer from NFS1 persulfide intermediate to

ISCU and to small thiols such as L-cysteine and glutathione leading to persulfuration of these thiols and ultimately sulfide release (PubMed:24971490). Binds ferrous ion and is released from FXN upon the addition of both L-cysteine and reduced FDX2 during [2Fe-2S] cluster assembly (PubMed:29576242). The core iron-sulfur cluster (ISC) assembly complex is involved in the de novo synthesis of a [2Fe-2S] cluster, the first step of the mitochondrial iron-sulfur protein biogenesis. This process is initiated by the cysteine desulfurase complex (NFS1:LYRM4:NDUFAB1) that produces persulfide which is delivered on the scaffold protein ISCU in a FXN-dependent manner. Then this complex is stabilized by FDX2 which provides reducing equivalents to accomplish the [2Fe-2S] cluster assembly. Finally, the [2Fe-2S] cluster is transferred from ISCU to chaperone proteins, including HSCB, HSPA9 and GLRX5 (By similarity). 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 (PubMed:15641778). 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 (PubMed:11823441, PubMed:12755598). May function as an iron chaperone protein that protects the aconitase [4Fe-4S]₂ cluster from disassembly and promotes enzyme reactivation (PubMed:15247478). May play a role as a high affinity iron binding partner for FECH that is capable of both delivering iron to ferrochelatase and mediating the terminal step in mitochondrial heme biosynthesis (PubMed:15123683, PubMed:16239244).

Cellular Location

[Frataxin mature form]: Mitochondrion

Tissue Location

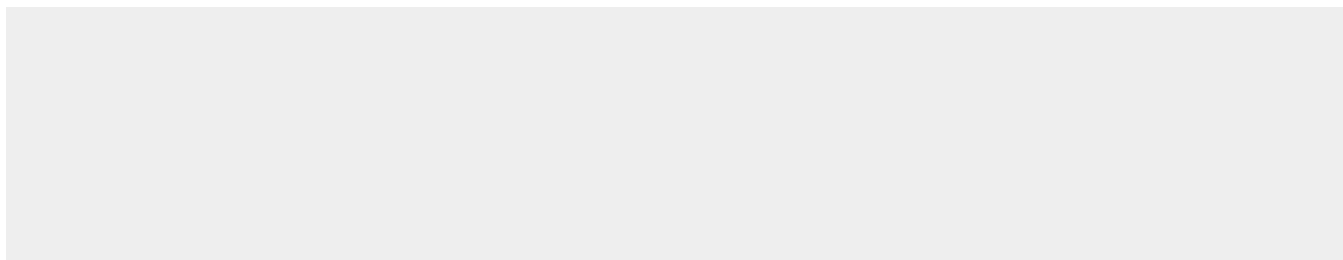
Expressed in the heart, peripheral blood lymphocytes and dermal fibroblasts.

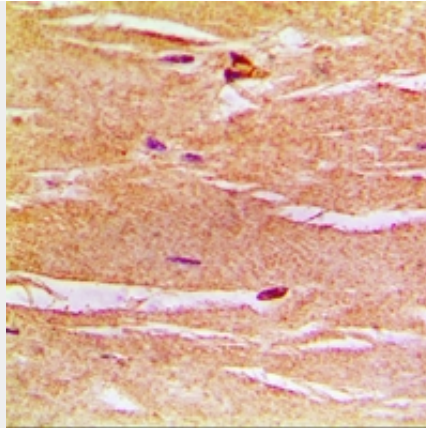
Anti-Frataxin Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

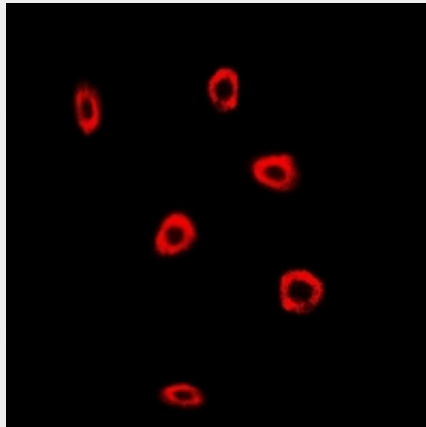
- [Western Blot](#)
- [Blocking Peptides](#)
- [Dot Blot](#)
- [Immunohistochemistry](#)
- [Immunofluorescence](#)
- [Immunoprecipitation](#)
- [Flow Cytometry](#)
- [Cell Culture](#)

Anti-Frataxin Antibody - Images





Immunohistochemical analysis of Frataxin staining in rat heart formalin fixed paraffin embedded tissue section. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH 6.0). The section was then incubated with the antibody 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.



Immunofluorescent analysis of Frataxin staining in MCF7 cells. Formalin-fixed cells were permeabilized with 0.1% Triton X-100 in TBS for 5-10 minutes and blocked with 3% BSA-PBS for 30 minutes at room temperature. Cells were probed with the primary antibody in 3% BSA-PBS and incubated overnight at 4 °C in a humidified chamber. Cells were washed with PBST and incubated with a DyLight 594-conjugated secondary antibody (red) in PBS at room temperature in the dark.

Anti-Frataxin Antibody - Background

Recombinant full length protein of human Frataxin