

FXN Antibody (N-term)
Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP19965A**Specification**

FXN Antibody (N-term) - Product Information

Application	WB,E
Primary Accession	Q16595
Other Accession	NP_000135.2
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Calculated MW	23135
Antigen Region	36-64

FXN Antibody (N-term) - Additional Information**Gene ID** 2395**Other Names**

Frataxin, mitochondrial, Friedreich ataxia protein, Fxn, Frataxin intermediate form, i-FXN, Frataxin(56-210), m56-FXN, Frataxin(78-210), d-FXN, m78-FXN, Frataxin mature form, Frataxin(81-210), m81-FXN, FXN, FRDA, X25

Target/Specificity

This FXN antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 36-64 amino acids from the N-terminal region of human FXN.

Dilution

WB~~1:1000

Format

Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.

Storage

Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions

FXN Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures.

FXN Antibody (N-term) - 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:[24971490](#), PubMed:[12785837](#)). 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

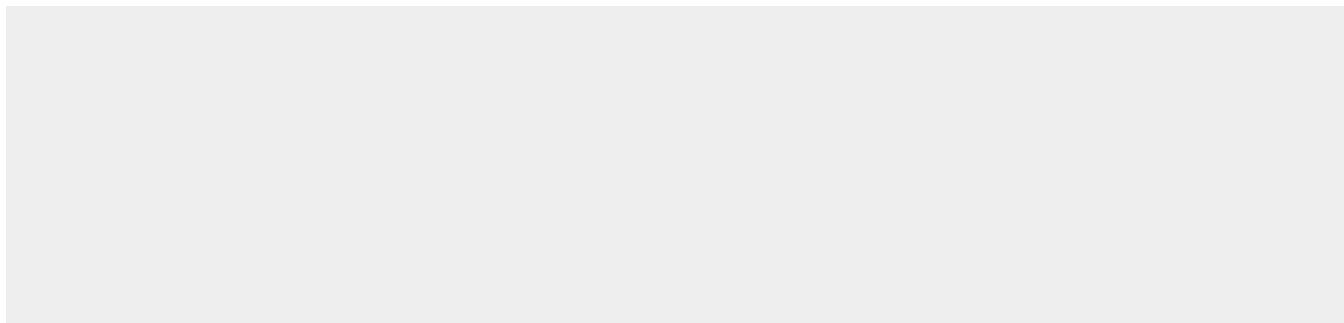
Tissue Location

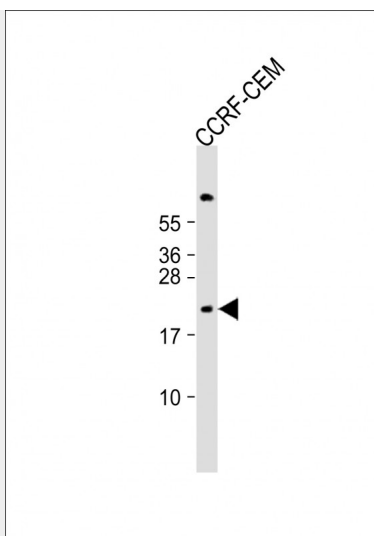
Expressed in the heart, peripheral blood lymphocytes and dermal fibroblasts.

FXN Antibody (N-term) - 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](#)

FXN Antibody (N-term) - Images



Anti-FXN Antibody (N-term) at 1:1000 dilution + CCRF-CEM whole cell lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 23 kDa Blocking/Dilution buffer: 5% NFDm/TBST.

FXN Antibody (N-term) - Background

This nuclear gene encodes a mitochondrial protein which belongs to FRATAXIN family. The protein functions in regulating mitochondrial iron transport and respiration. The expansion of intronic trinucleotide repeat GAA results in Friedreich ataxia. Alternative splicing results in multiple transcript variants.

FXN Antibody (N-term) - References

Tsai, C.L., et al. Biochemistry 49(43):9132-9139(2010)
Thierbach, R., et al. Biochem. J. 432(1):165-172(2010)
Bailey, S.D., et al. Diabetes Care 33(10):2250-2253(2010)
Marino, T.C., et al. Clin. Genet. 77(6):598-600(2010)
Li, K., et al. PLoS ONE 5 (8), E12286 (2010) :