

FXN Antibody (Center)
Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP6409C**Specification**

FXN Antibody (Center) - Product Information

Application	IF, WB, IHC-P,E
Primary Accession	Q16595
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Calculated MW	23135
Antigen Region	72-102

FXN Antibody (Center) - 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 72-102 amino acids from the Central region of human FXN.

Dilution

IF~~1:10~50
WB~~1:1000
IHC-P~~1:10~50
E~~Use at an assay dependent concentration.

Format

Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is prepared by Saturated Ammonium Sulfate (SAS) precipitation followed by dialysis against PBS.

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 (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

FXN Antibody (Center) - 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

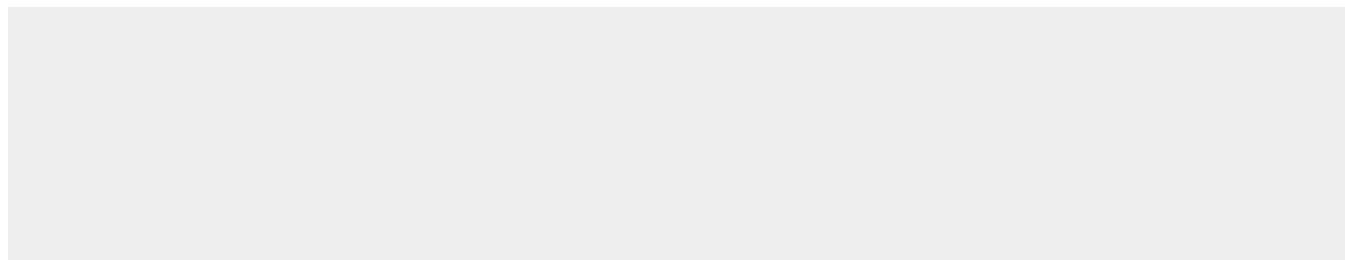
Expressed in the heart, peripheral blood lymphocytes and dermal fibroblasts.

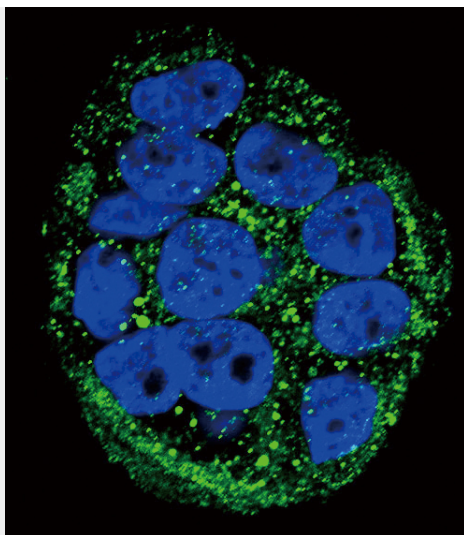
FXN Antibody (Center) - Protocols

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

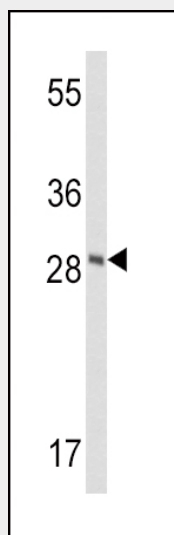
- [Western Blot](#)
- [Blocking Peptides](#)
- [Dot Blot](#)
- [Immunohistochemistry](#)
- [Immunofluorescence](#)
- [Immunoprecipitation](#)
- [Flow Cytometry](#)
- [Cell Culture](#)

FXN Antibody (Center) - Images

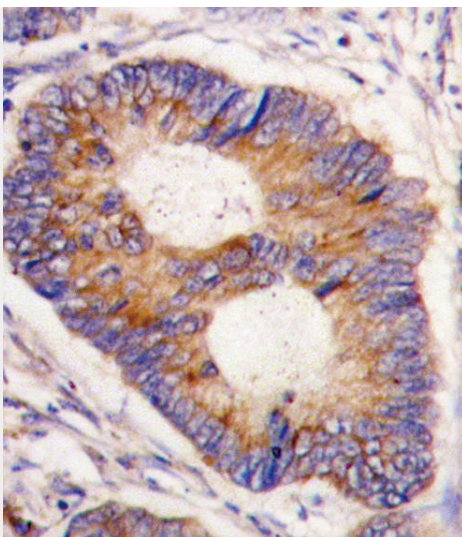




Confocal immunofluorescent analysis of FXN Antibody (Center)(Cat#AP6409c) with WiDr cell followed by Alexa Fluor 488-conjugated goat anti-rabbit IgG (green). DAPI was used to stain the cell nuclear (blue).



Western blot analysis of FXN Antibody (Center) (Cat.#AP6409c) in CEM cell line lysates (35ug/lane). FXN (arrow) was detected using the purified Pab.



Formalin-fixed and paraffin-embedded human colon carcinoma tissue reacted with FXN antibody (Center)(Cat.#AP6409c), which was peroxidase-conjugated to the secondary antibody, followed by DAB staining. This data demonstrates the use of this antibody for immunohistochemistry; clinical relevance has not been evaluated.

FXN Antibody (Center) - Background

FXN is a mitochondrial protein which belongs to the FRATAXIN family. The protein functions in regulating mitochondrial iron transport and respiration. The expansion of intronic trinucleotide repeat GAA results in Friedreich ataxia.