10320 Camino Santa Fe, Suite G
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Tel: 858.875.1900 Fax: 858.875.1999

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

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 scaffoding 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.

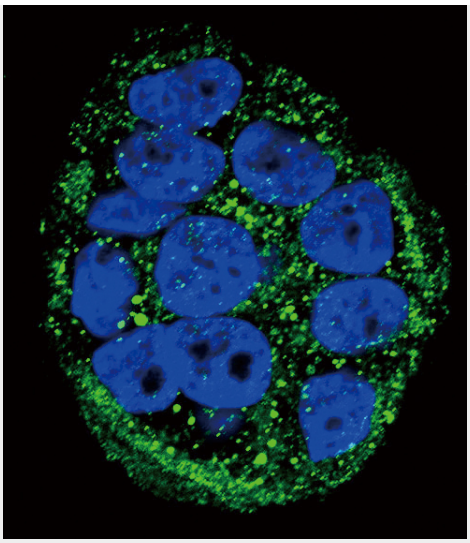
FXN Antibody (Center) - Protocols

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

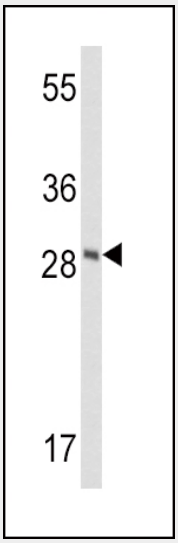
- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

FXN Antibody (Center) - Images

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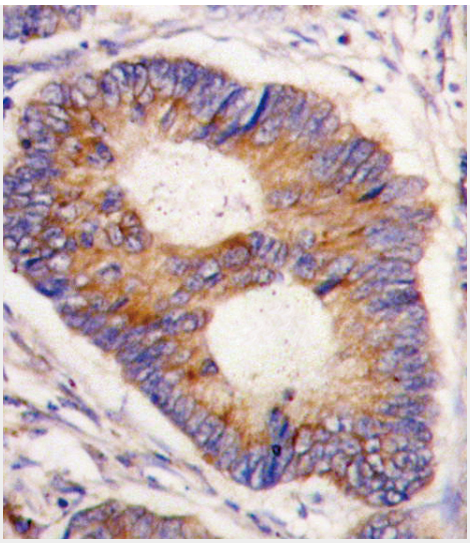


Confocal immunofluorescent analysis of FXN Antibody (Center)(Cat#AP6409c) with WiDr cell followed by Alexa Fluor 488-conjugated goat anti-rabbit lgG (green). DAPI was used to stain the cell nuclear (blue).



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

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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.