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FXN Antibody (Center)

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP6409C

Product Information

Application	WB, IHC-P, IF, E
Primary Accession	Q16595
Reactivity	Human
Predicted	Mouse, Rat, Bovine, Canine, Rabbit, Chicken
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB12541
Calculated MW	23135
Antigen Region	72-102

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	WB~~1:1000 IHC-P~~1:100~500 IF~~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.

Protein Information

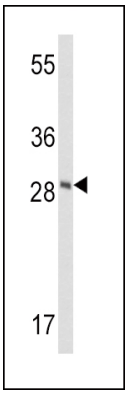
Name	FXN (HGNC:3951)
Synonyms	FRDA, X25

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: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.

Background

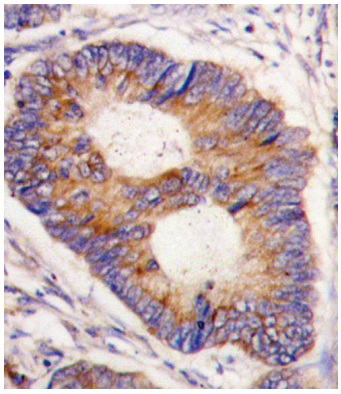
FXN is a mitochondrial protein which belongs to the FRATAXIN family. The protein functions in regulating mitochondrial iron transport and respiration. The expansion of intronic trinucleotide repeat GAA results in Friedreich ataxia.

Images

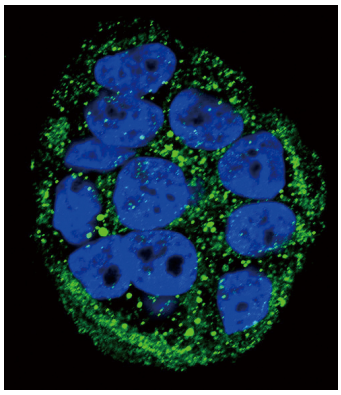


Western blot analysis of FXN Antibody (Center) (Cat.#AP6409c) in CEM cell line lysates (35ug/lane). FXN (arrow) was detected using the purified Pab.

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.



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).

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.