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FXN (11J13) Rabbit Monoclonal Antibody

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Catalog # AP93739

Product Information

Application	WB, IHC-P, ICC, FC, IP, IF
Primary Accession	Q16595, O35943, D3ZYW7
Reactivity	Human, Mouse, Rat
Clonality	Monoclonal
Calculated MW	23135

Additional Information

Gene ID	2395
Dilution	WB~~1:1000 IHC-P~~N/A ICC~~N/A FC~~1:10~50 IP~~N/A IF~~1:50~200
Storage Conditions	-20℃

Protein Information

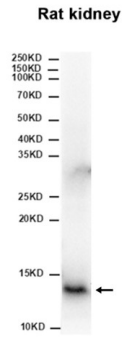
Name	FXN (HGNC:3951)
Synonyms	FRDA, X25
Function	[Frataxin mature form]: Functions as an activator of persulfide transfer to the scaffoding protein ISCU as component of the core iron-sulfur cluster (ISC) assembly complex and participates to the [2Fe-2S] cluster assembly (PubMed:12785837, PubMed:24971490). 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

This nuclear gene encodes 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 from 8-33 repeats to >90 repeats results in Friedreich ataxia. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Jul 2016]

Images



Western blot analysis of extracts from Rat kidney tissue using AP93739 at 1:1000.

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