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FXN (11J13) Rabbit Monoclonal Antibody

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Catalog # AP93739

Product Information

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

Additional Information

Gene ID	2395
Dilution	WB~~1:1000 IHC~~1:100~500 IF~~1:50~200 FC~~1:10~50 ICC~~N/A IP~~N/A
Storage Conditions	-20℃

Protein Information

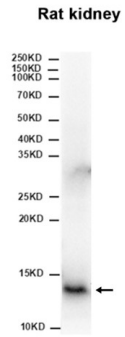
Name	FXN (HGNC:3951)
Synonyms	FRDA, X25
Function	[Frataxin mature form]: Functions as an activator of persulfide transfer to the scaffoding protein ISCU as component of the core iron-sulfur cluster (ISC) assembly complex and participates to the [2Fe-2S] cluster assembly (PubMed:12785837, PubMed:24971490).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