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Anti-Frataxin Antibody

Catalog # AP61627

Product Information

Application	WB, IHC
Primary Accession	Q16595
Reactivity	Human, Mouse, Rat
Host	Rabbit
Clonality	Polyclonal
Calculated MW	23135

Additional Information

Gene ID	2395
Other Names	FRDA; X25; Frataxin mitochondrial; Friedreich ataxia protein; Fxn
Target/Specificity	Recombinant fusion protein of human Frataxin
Dilution	WB~~WB (1/500 - 1/1000) IHC~~1:100~500
Format	Liquid in 0.42% Potassium phosphate, 0.87% Sodium chloride, pH 7.3, 30% glycerol, and 0.01% sodium azide.
Storage	Store at -20 °C.Stable for 12 months from date of receipt

Protein Information

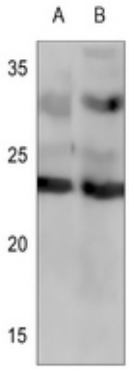
Name	FXN (HGNC:3951)
Synonyms	FRDA, X25
Function	[Frataxin mature form]: Functions as an activator of persulfide transfer to the scaffoding protein ISCU as component of the core iron-sulfur cluster (ISC) assembly complex and participates to the [2Fe-2S] cluster assembly (PubMed:12785837, PubMed:24971490).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

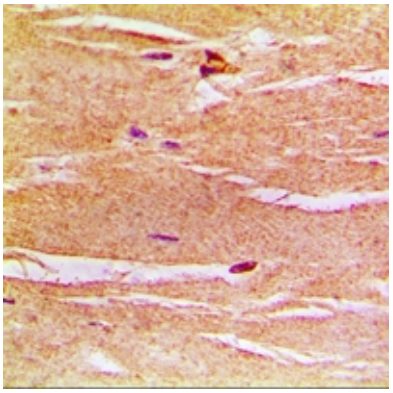
Cellular Location [Frataxin mature form]: Mitochondrion

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

Images



Western blot analysis of Frataxin expression in mouse liver (A), rat kidney (B) whole cell lysates. (Predicted band size: 23 kD; Observed band size: 23 kD)



Immunohistochemical analysis of Frataxin staining in rat heart formalin fixed paraffin embedded tissue section. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH 6.0). The section was then incubated with the antibody at room temperature and detected using an HRP conjugated compact polymer system. DAB was used as the chromogen. The section was then counterstained with haematoxylin and mounted with DPX.

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