

Anti-Frataxin Antibody

Rabbit polyclonal antibody to Frataxin
Catalog # AP61627

Product Information

Application	WB, IF/IC, IHC
Primary Accession	Q16595
Other Accession	Q35943
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	Recognizes endogenous levels of Frataxin protein.
Dilution	WB~~1:1000 IF/IC~~N/A IHC~~1:100~500
Format	Liquid in 0.42% Potassium phosphate, 0.87% Sodium chloride, pH 7.3, 30% glycerol, and 0.09% (W/V) 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 scaffolding 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

Recombinant full length protein of human Frataxin

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