

Frataxin Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP59300

Specification

Frataxin Polyclonal Antibody - Product Information

Application WB, IHC-P, IHC-F, IF, ICC, E

Primary Accession <u>Q16595</u>

Reactivity Rat, Pig, Dog, Bovine

Host Rabbit
Clonality Polyclonal
Calculated MW 23135

Frataxin Polyclonal Antibody - Additional Information

Gene ID 2395

Other Names

Frataxin, mitochondrial, 1.16.3.1, 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

Dilution

WB~~1:1000<br \><span class
="dilution_IHC-P">IHC-P~~N/A<br \><span class
="dilution_IHC-F">IHC-F~~N/A<br \><span class
="dilution_IF">IF~~1:50~200<br \>ICC~~N/A<br \>ICC~~N/A<br \>ICC~~N/A

Format

0.01M TBS(pH7.4), 0.09% (W/V) sodium azide and 50% Glyce

Storage

Store at -20 $^{\circ}$ C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 $^{\circ}$ C.

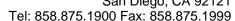
Frataxin Polyclonal Antibody - Protein Information

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.

Frataxin Polyclonal Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

Frataxin Polyclonal Antibody - Images