

Fxn antibody - middle region

Rabbit Polyclonal Antibody

Catalog # AI12861

Product Information

Application	WB
Primary Accession	O35943
Other Accession	NM_008044 , NP_032070
Reactivity	Human, Mouse, Rat, Rabbit, Dog, Guinea Pig, Horse, Bovine
Predicted	Human, Mouse, Rabbit, Pig, Chicken, Dog, Guinea Pig, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	22924

Additional Information

Gene ID	14297
Alias Symbol	FA, FARR, Frda, X25
Other Names	Frataxin, mitochondrial, Fxn, 1.16.3.1, Frataxin intermediate form, Frataxin mature form, Fxn, Frda
Format	Liquid. Purified antibody supplied in 1x PBS buffer with 0.09% (w/v) sodium azide and 2% sucrose.
Reconstitution & Storage	Add 50 ul of distilled water. Final anti-Fxn antibody concentration is 1 mg/ml in PBS buffer with 2% sucrose. For longer periods of storage, store at 20°C. Avoid repeat freeze-thaw cycles.
Precautions	Fxn antibody - middle region is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	Fxn {ECO:0000312 MGI:MGI:1096879}
Synonyms	Frda
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: 19805308 , PubMed: 25597503). 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: 25597503). Binds ferrous ion and is released from FXN upon the addition of both L-cysteine and reduced FDX2 during [2Fe-2S] cluster

assembly (By similarity). 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. 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. May function as an iron chaperone protein that protects the aconitase [4Fe-4S]₂⁺ cluster from disassembly and promotes enzyme reactivation. 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 (By similarity).

Cellular Location

[Frataxin mature form]: Mitochondrion

Tissue Location

Heart, liver, skeletal muscle, kidney, spleen and thymus. Weakly expressed in the brain and lung

Images



WB Suggested Anti-Fxn Antibody Titration: 1.0 µg/ml
Positive Control: Mouse Heart

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