

Fxn antibody - middle region Rabbit Polyclonal Antibody Catalog # Al12861

Specification

Fxn antibody - middle region - Product Information

Application Primary Accession Other Accession Reactivity

Predicted

Host Clonality Calculated MW WB <u>O35943</u> <u>NM_008044</u>, <u>NP_032070</u> Human, Mouse, Rat, Rabbit, Horse, Bovine, Guinea Pig, Dog Human, Mouse, Rabbit, Pig, Chicken, Horse, Guinea Pig, Dog Rabbit Polyclonal 23kDa KDa

Fxn antibody - middle region - 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.

Fxn antibody - middle region - Protein Information

Name Fxn {ECO:0000312|MGI:MGI:1096879}

Synonyms Frda

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:25597503, PubMed:19805308). 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 oligometric 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

Fxn antibody - middle region - Protocols

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

- <u>Western Blot</u>
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- <u>Cell Culture</u>

Fxn antibody - middle region - Images



