

# Fxn antibody - middle region

Rabbit Polyclonal Antibody Catalog # Al12861

# **Specification**

# Fxn antibody - middle region - Product Information

Application WB
Primary Accession 035943

Other Accession NM 008044, NP 032070

Reactivity Human, Mouse, Rat, Rabbit, Horse, Bovine,

Guinea Pig, Dog

Predicted Human, Mouse, Rabbit, Pig, Chicken,

Horse, Guinea Pig, Dog

Host Rabbit
Clonality Polyclonal
Calculated MW 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:<a href="http://www.uniprot.org/citations/25597503" target="\_blank">25597503</a>, PubMed:<a href="http://www.uniprot.org/citations/19805308" target="\_blank">19805308</a>). 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:<a href="http://www.uniprot.org/citations/25597503" target=" blank">25597503</a>). 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**

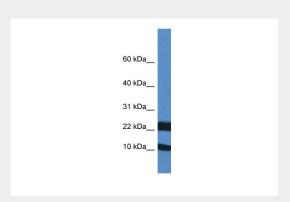
Heart, liver, skeletal muscle, kidney, spleen and thymus. Weakly expressed in the brain and lung

### Fxn antibody - middle region - 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

### Fxn antibody - middle region - Images



WB Suggested Anti-Fxn Antibody Titration: 1.0 μg/ml

Positive Control: Mouse Heart

