

**FXN Antibody (C-term) Blocking Peptide**  
**Synthetic peptide**  
**Catalog # BP6409b****Specification**

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**FXN Antibody (C-term) Blocking Peptide - Product Information**Primary Accession [Q16595](#)**FXN Antibody (C-term) Blocking Peptide - Additional Information**

Gene ID 2395

**Other Names**

Frataxin, mitochondrial, 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

**Target/Specificity**

The synthetic peptide sequence used to generate the antibody [AP6409b](/product/products/AP6409b) was selected from the C-term region of human FXN. A 10 to 100 fold molar excess to antibody is recommended. Precise conditions should be optimized for a particular assay.

**Format**

Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

**Storage**

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

**Precautions**

This product is for research use only. Not for use in diagnostic or therapeutic procedures.

**FXN Antibody (C-term) Blocking Peptide - 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: [24971490](http://www.uniprot.org/citations/24971490), PubMed: [12785837](http://www.uniprot.org/citations/12785837)). 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](http://www.uniprot.org/citations/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:<a href="http://www.uniprot.org/citations/29576242" target="\_blank">29576242</a>). 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:<a href="http://www.uniprot.org/citations/15641778" target="\_blank">15641778</a>). 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:<a href="http://www.uniprot.org/citations/11823441" target="\_blank">11823441</a>, PubMed:<a href="http://www.uniprot.org/citations/12755598" target="\_blank">12755598</a>). May function as an iron chaperone protein that protects the aconitase [4Fe-4S]<sub>2</sub> cluster from disassembly and promotes enzyme reactivation (PubMed:<a href="http://www.uniprot.org/citations/15247478" target="\_blank">15247478</a>). 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:<a href="http://www.uniprot.org/citations/15123683" target="\_blank">15123683</a>, PubMed:<a href="http://www.uniprot.org/citations/16239244" target="\_blank">16239244</a>).

#### Cellular Location

[Frataxin mature form]: Mitochondrion

#### Tissue Location

Expressed in the heart, peripheral blood lymphocytes and dermal fibroblasts.

### FXN Antibody (C-term) Blocking Peptide - Protocols

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

- [Blocking Peptides](#)

### FXN Antibody (C-term) Blocking Peptide - Images

### FXN Antibody (C-term) Blocking Peptide - Background

FXN is a mitochondrial protein which belongs to the FRATAXIN family. The protein functions in regulating mitochondrial iron transport and respiration. The expansion of intronic trinucleotide repeat GAA results in Friedreich ataxia.