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FXN Blocking Peptide (N-term)

Synthetic peptide
Catalog # BP19965A

Specification

FXN Blocking Peptide (N-term) - Product Information

Primary Accession	Q16595
Other Accession	NP_000135.2

FXN Blocking Peptide (N-term) - 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 is selected from aa 51-64 of HUMAN FXN

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 Blocking Peptide (N-term) - 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:24971490, PubMed: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). 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.

FXN Blocking Peptide (N-term) - Protocols

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

- Blocking Peptides

FXN Blocking Peptide (N-term) - Images

FXN Blocking Peptide (N-term) - Background

This nuclear gene encodes a mitochondrial protein which belongs to FRATAXIN family. The protein functions in regulating mitochondrial iron transport and respiration. The expansion of intronic trinucleotide repeat GAA results in Friedreich ataxia. Alternative splicing results in multiple transcript variants.

FXN Blocking Peptide (N-term) - References

Tsai, C.L., et al. Biochemistry 49(43):9132-9139(2010)
Thierbach, R., et al. Biochem. J. 432(1):165-172(2010)
Bailey, S.D., et al. Diabetes Care 33(10):2250-2253(2010)
Marino, T.C., et al. Clin. Genet. 77(6):598-600(2010)
Li, K., et al. PLoS ONE 5 (8), E12286 (2010) :