

FXN Blocking Peptide (N-term)

Synthetic peptide Catalog # BP19965A

Specification

FXN Blocking Peptide (N-term) - Product Information

Primary Accession <u>Q16595</u> Other Accession <u>NP 000135.2</u>

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

Synonyms FRDA, X25

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) :



Function

Promotes the biosynthesis of heme and assembly and repair of iron-sulfur clusters by delivering Fe(2+) to proteins involved in these pathways. 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. Modulates the RNA-binding activity of ACO1.

Cellular Location

Mitochondrion. Cytoplasm, cytosol. Note=PubMed:18725397 reports localization exclusively in mitochondria.

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