FXN Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	FXN, Polyclonal Antibody
Full Product Name	FXN, CT (Frataxin, Mitochondrial, Friedreich Ataxia Protein, Frataxin Intermediate Form, Frataxin(56-210), Frataxin(81-210), FRDA, X25) (MaxLight 650)
Product Gene Name	anti-FXN antibody
Clonality	Polyclonal
Isotype	IgG
Host	Rabbit
Species Reactivity	Human
Form/Format	Supplied as a liquid in PBS, pH 7.2. No preservative added. Labeled with MaxLight650.
Purity/Purification	Purified by Ammonium Sulfate Precipitation.
Specificity	Recognizes human FXN.
Preparation and Storage	Store product at 4 degree C in the dark. DO NOT FREEZE! Stable at 4 degree C for 12 months after receipt as an undiluted liquid. Dilute required amount only prior to immediate use. Further dilutions can be made in assay buffer. Caution: MaxLight650 conjugates are sensitive to light. For maximum recovery of product, centrifuge the original vial prior to removing the cap.
Molecular Weight	21,416 Da
OMIM	phenotype 606829
3D Structure	ModBase 3D Structure for Q16595
Immunogen	KLH-conjugated synthetic peptide mapping to a fragment of residues within amino acids 143-172 in the C-terminal region of human FXN, UniProt Accession #Q16595.
Conjugate	MaxLight650
Note	Preservative Free
Product Categories/Family for anti-FXN antibody	Antibodies; Mitochondrial Proteins
Applications Tested/Suitable for anti-FXN antibody	Immunohistochemistry (IHC), Western Blot (WB), FLISA
Application Notes for anti-FXN antibody	IHC: 1:10-1:50 WB: 1:1000 Applications are based on unconjugated antibody.
Related Product Information for anti-FXN antibody	MBS6476893 is an antibody targeting FXN. Also known as FXN, CT (Frataxin, Mitochondrial, Friedreich Ataxia Protein, Frataxin Intermediate Form, Frataxin(56-210), Frataxin(81-210), FRDA, X25) (MaxLight 650). Reactive in Human. Host: Rabbit.
Other Notes	Small volumes of anti-FXN antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for FXN. It may not necessarily be applicable to this product.

NCBI GI #	31077081
NCBI GeneID	2395
NCBI Accession #	NP_000135
UniProt Primary Accession #	Q16595
UniProt Secondary Accession #	O15545; O95656; Q15294; Q5VZ01; A8MXJ6; C9JJ89
UniProt Related Accession #	Q16595
GenBank Nucleotide #	NM_000144.4
NCBI Official Full Name	frataxin, mitochondrial isoform 1 preproprotein
NCBI Official Synonym Full Names	frataxin
NCBI Official Symbol	FXN
NCBI Official Synonym Symbols	FA; X25; CyaY; FARR; FRDA
UniProt Protein Name	Frataxin, mitochondrial
UniProt Synonym Protein Names	Friedreich ataxia protein; Fxn
Protein Family	Frataxin
UniProt Gene Name	FXN
UniProt Synonym Gene Names	FRDA; X25; Fxn; i-FXN
UniProt Entry Name	FRDA_HUMAN
NCBI Protein Information	frataxin, mitochondrial
NCBI Summary for FXN	This nuclear gene encodes 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 from 8-33 repeats to >90 repeats results in Friedreich ataxia. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Jul 2016]

UniProt Comments for FXN	FXN: 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. Defects in FXN are the cause of Friedreich ataxia (FRDA). FRDA is an autosomal recessive, progressive degenerative disease characterized by neurodegeneration and cardiomyopathy it is the most common inherited ataxia. The disorder is usually manifest before adolescence and is generally characterized by incoordination of limb movements, dysarthria, nystagmus, diminished or absent tendon reflexes, Babinski sign, impairment of position and vibratory senses, scoliosis, pes cavus, and hammer toe. In most patients, FRDA is due to GAA triplet repeat expansions in the first intron of the frataxin gene. But in some cases the disease is due to mutations in the coding region. Belongs to the frataxin family. 2 isoforms of the human protein are produced by alternative splicing. Protein type: Motility/polarity/chemotaxis; Mitochondrial; EC 1.16.3.1 Chromosomal Location of Human Ortholog: 9q21.11 Cellular Component: mitochondrion; mitochondrial matrix; cytosol Molecular Function: 2 iron, 2 sulfur cluster binding; ferroxidase activity; protein binding; ferric iron binding; ferrous iron binding; iron-sulfur cluster binding Biological Process: mitochondrion organization and biogenesis; negative regulation of multicellular organism growth; cellular iron ion homeostasis; positive regulation of metalloenzyme activity; positive regulation of transferase activity; proprioception; negative regulation of organ growth; positive regulation of cell growth; adult walking behavior; protein autoprocessing; embryonic development ending in birth or egg hatching; iron incorporation into metallo-sulfur cluster; positive regulation of lyase activity; positive regulation of cell proliferation; aerobic respiration; ion transport; response to iron ion; negative regulation of apoptosis; positive regulation of oxidoreductase activity; oxidative phosphorylation; heme biosynthetic process Disease: Friedreich Ataxia 1
Research Articles on FXN	**1.** In this study, the authors demonstrate that frataxin regulates the expression of genes involved in interferon induced apoptosis, DNA damage and blood clotting regulation.
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