abcam

Product datasheet

Recombinant Human Frataxin protein ab110353

4 References 2 Images

Description

Product name	Recombinant Human Frataxin protein
Expression system	Escherichia coli
Accession	Q16595
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	SSNQRGLNQI WNVKKQSVYL MNLRKSGTLG HPGSLDETTY ERLAEETLDS LAEFFEDLAD KPYTFEDYDV SFGSGVLTVK LGGDLGTYVI NKQTPNKQIW LSSPSSGPKR YDWTGKNWVY SHDGVSLHEL LAAELTKALK TKLDLSSLAY SGKDA
Predicted molecular weight	17 kDa
Amino acids	56 to 210

Specifications

Our Abpromise guarantee covers the use of **ab110353** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	Sandwich ELISA
Form	Lyophilized

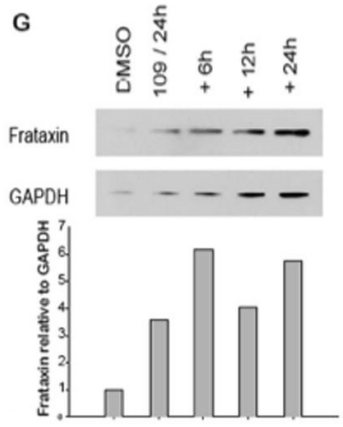
Preparation and Storage

Stability and Storage	Shipped on Dry Ice. Store at -80°C. Avoid freeze / thaw cycle. Constituent: 1% BSA
Reconstitution	Ships at 4°C. Store lyophilized powder at 4°C. Reconstitute with 0.2 mL of H_2O to a final concentration of 60ng/mL. Reconstituted solution should be stored at -80°C. Concentration: 0.00006 mg/mL after reconstitution with 0.2 mL H_2O

General Info

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.
Tissue specificity	Expressed in the heart, peripheral blood lymphocytes and dermal fibroblasts.
Involvement in disease	Defects in FXN are the cause of Friedreich ataxia (FRDA) [MIM:229300]. 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.
Sequence similarities	Belongs to the frataxin family.
Post-translational modifications	Processed in two steps by mitochondrial processing peptidase (MPP). MPP first cleaves the precursor to intermediate form and subsequently converts the intermediate to yield frataxin mature form (frataxin(81-210)) which is the predominant form. The additional forms, frataxin(56-210) and frataxin(78-210), seem to be produced when the normal maturation process is impaired; their physiological relevance is unsure.
Cellular localization	Cytoplasm. Mitochondrion. PubMed:18725397 reports localization exclusively in mitochondria.

Images



Western blot - Recombinant Human Frataxin protein (ab110353)

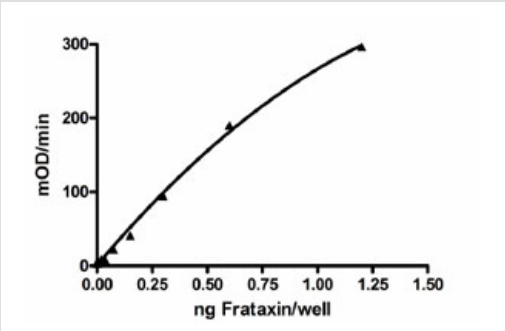
Rai et al PLoS One. 2010 Jan 21;5(1):e8825. doi: 10.1371/journal.pone.0008825. Fig 2.

Effect of compounds 136 and 109 on FRDA patients' primary lymphocytes.

Another extraction of PBMC from patient P13 were treated with either 109 at 10 µM or DMSO for 48 hours and harvested after a 6, 12 or 24 hours wash out period. *FXN* mRNA was determined by real-time RT-PCR and in Panel G., frataxin protein was assessed by western blotting followed by densitometry quantification after the same time points for wash out.

Tissues were homogenized in T-PER tissue protein extraction reagent for total proteins extraction. Histones were purified by acid extraction. Primary antibodies were diluted in Odyssey blocking buffer for frataxin and actin or in PBS for total and acetylated histones antibodies and for human frataxin (ab110353). Infrared dye conjugated secondary antibodies (anti-rabbit IRdye800Cw and anti-mouse IRdye680) were used to detect and quantify the signal of mouse frataxin / actin using a Li-Cor Odyssey imaging system. Horseradish Peroxidase (HRP) - conjugated secondary antibodies

were used to detect the signal of total and acetylated histones and for the human frataxin / Gapdh signal by chemiluminescence.



Sandwich ELISA - Recombinant Human Frataxin protein (ab110353)

Sandwich ELISA standard curve using ab110353 within the range 0.00-1.25 ng/well.

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