

Human Frataxin Matched Antibody Pair Kit ab212062

1 Image

Overview

Product name	Human Frataxin Matched Antibody Pair Kit
Detection method	Colorimetric
Assay type	ELISA set
Sensitivity	8.4 pg/ml
Range	39 pg/ml - 2500 pg/ml
Species reactivity	Reacts with: Human
Product overview	<p>Human Frataxin Matched Antibody Pair Kits include a capture and a biotinylated detector antibody pair, along with a calibrated protein standard, suitable for sandwich ELISA. The Matched Antibody Pair Kit can be used to quantify native and recombinant human Frataxin.</p> <p>Optimization of the kit reagents to sample type, immunoassay format or instrumentation may be required. Guidelines for use of this kit in a standard 96-well microplate sandwich ELISA using HRP/TMB system of colorimetric detection is described in this assay procedure for the purposes of quantification.</p> <p>Protocol information and tips on the use of the Matched Antibody Pair kits for sandwich ELISA can be found on our website. An accessory pack can be purchased which includes buffer reagents required to perform 10 x 96-well plate sandwich ELISAs (ab210905).</p> <p>For additional information on the performance of the antibody pair used in this kit, please see our equivalent SimpleStep ELISA kit ab176112. Please note that while the antibody pair is the same provided in the corresponding SimpleStep ELISA Kit, due to differences in their formulation, this antibody pair cannot be used with the consumables provided with our SimpleStep ELISA Kits.</p>
Tested applications	Suitable for: ELISA, IA
Platform	Reagents

Properties

Storage instructions	Store at -20°C. Please refer to protocols.
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Components	5 x 96 tests
Human Frataxin Capture Antibody	1 x 50µg
Human Frataxin Detector Antibody	1 x 12.5µg
Human Frataxin Lyophilized Protein	1 vial

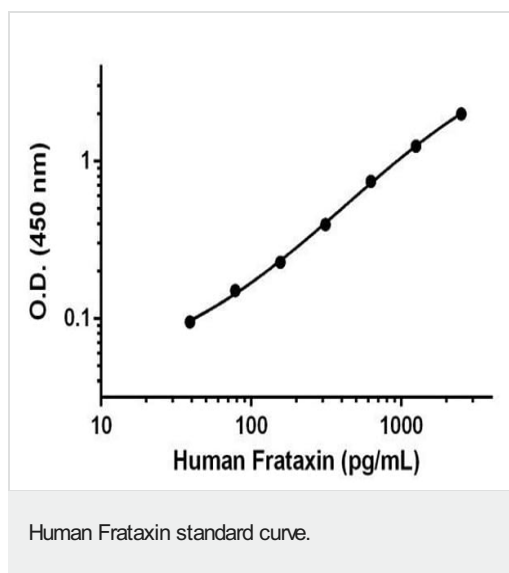
Function	Promotes the biosynthesis of heme and assembly and repair of iron-sulfur clusters by delivering Fe(2+) to proteins involved in these pathways. May play a role in the protection against iron-catalyzed oxidative stress through its ability to catalyze the oxidation of Fe(2+) to Fe(3+); the oligomeric 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.

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab212062 in the following tested applications. The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
ELISA		Use at an assay dependent concentration.
IA		Use at an assay dependent concentration.

Images



Standard calibration curve. Background subtracted values are graphed

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