abcam

Product datasheet

Recombinant Human Frataxin protein ab95502

1 References　画像数 1

製品の詳細

製品名	Recombinant Human Frataxin protein
精製度	> 95 % SDS-PAGE. ab95502 is purified using conventional chromatography techniques.
発現系	Escherichia coli
アクセッション番号	Q16595
タンパク質長	Full length protein
Animal free	No
由来	Recombinant
生物種	Human
配列	MGSSHHHHHH SSGLVPRGSH MLRTDIDATC TPRRASSNQR GLNQIWNVKK QSVYLMNLRK SGTLGHPGSL DETTYERLAE ETLDSLAEFF EDLADKPYTF EDYDVSFGSG VLTVKLGGDL GTYVINKQTP NKQIWLSSPS SGPKRYDWTG KNWVYSHDGV SLHELLAAEL TKALKTKLDL SSLAYSGKDA
予測される分子量	21 kDa including tags
実際の分子量	21 kDa including tags
領域	42 to 210
タグ	His tag N-Terminus

特性

Our **Abpromise guarantee** covers the use of **ab95502** in the following tested applications.

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

アプリケーション	SDS-PAGE Mass Spectrometry
製品の状態	Liquid

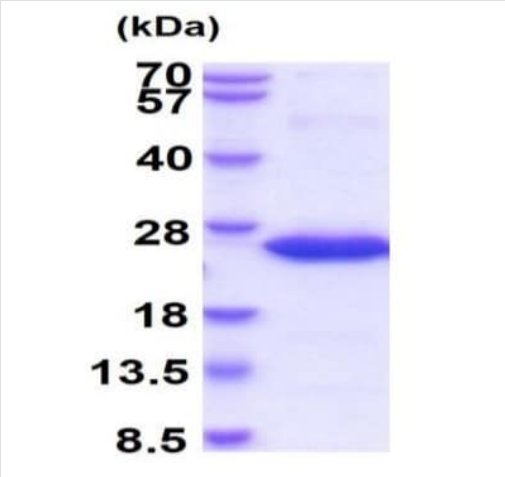
前処理および保存

保存方法および安定性	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles. pH: 8.5 Constituents: 0.0154% DTT, 0.316% Tris HCl, 10% Glycerol (glycerin, glycerine)

関連情報

機能	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.
組織特異性	Expressed in the heart, peripheral blood lymphocytes and dermal fibroblasts.
関連疾患	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.
配列類似性	Belongs to the frataxin family.
翻訳後修飾	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.
細胞内局在	Cytoplasm. Mitochondrion. PubMed:18725397 reports localization exclusively in mitochondria.

画像



SDS-PAGE - Recombinant Human Frataxin protein (ab95502)

15% SDS-PAGE analysis of 3µg ab95502.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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