abcam

Product datasheet

Anti-Frataxin antibody ab55021

1 图像

概述

产品名称	Anti-Frataxin抗体
描述	**小鼠单克隆抗体**to Frataxin
宿主	Mouse
经测试应用	**适用于:** WB
种属反应性	**与反应:** Recombinant fragment **预测可用于:** Human
免疫原	Recombinant fragment, corresponding to amino acids 91-201 of Human Frataxin

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	Preservative: None PBS, pH 7.2
纯度	Protein G purified
克隆	单克隆
同种型	IgG1
轻链类型	kappa

应用

Our Abpromise guarantee covers the use of **ab55021** in the following tested applications.

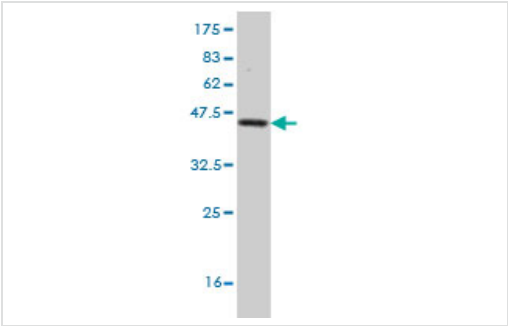
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

应用	Ab评论	说明
WB		Use a concentration of 1 - 5 µg/ml. This antibody has only been tested in WB against the recombinant fragment used as immunogen. We have no data on the detection of endogenous protein.

靶标

功能	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.
组织特异性	Expressed in the heart, peripheral blood lymphocytes and dermal fibroblasts.
疾病相关	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.
序列相似性	Belongs to the frataxin family.
翻译后修饰	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.
细胞定位	Cytoplasm. Mitochondrion. PubMed:18725397 reports localization exclusively in mitochondria.

图片



Western blot - Anti-Frataxin antibody (ab55021)

Western blot against tagged recombinant protein immunogen using ab55021 Frataxin antibody at 1ug/ml. Predicted band size of immunogen is 37 kDa.

This antibody has only been tested in WB against the recombinant fragment used as immunogen. We have no data on the detection of endogenous protein.

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