

Anti-Frataxin antibody [EPR6107] ab124680

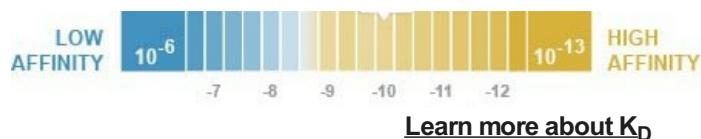
重组 RabMAb

1 References 5 图像

概述	
产品名称	Anti-Frataxin抗体[EPR6107]
描述	兔单克隆抗体[EPR6107] to Frataxin
宿主	Rabbit
特异性	We suggest optimizing experimental protocols (increasing lysate amount, using lower dilution or higher sensitivity ECL substrate) to improve results.
经测试应用	适用于: WB 不适用于: IHC-P
种属反应性	与反应: Human
免疫原	Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.
阳性对照	WB: Human fetal heart tissue, human skin, His tagged Human FXN recombinant protein (aa 82-210), Human liver and hippocampus tissues, HCT116, Huh7 and SH-SY5Y whole cell lysates.
常规说明	<p>This product is a recombinant monoclonal antibody, which offers several advantages including:</p> <ul style="list-style-type: none">- High batch-to-batch consistency and reproducibility- Improved sensitivity and specificity- Long-term security of supply- Animal-free production <p>For more information see here.</p> <p>Our RabMAb[®] technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to RabMAb[®] patents.</p> <p>Mouse, Rat: We have preliminary internal testing data to indicate this antibody may not react with these species. Please contact us for more information.</p>

性能	
形式	Liquid
存放说明	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C. Stable for 12 months at -20°C.
解离常数 (K _D)	K _D = 3.19 x 10 ⁻¹⁰ M





存储溶液	pH: 7.20 Preservative: 0.01% Sodium azide Constituents: 40% Glycerol (glycerin, glycerine), 0.05% BSA, 59% PBS
纯度	Protein A purified
克隆	单克隆
克隆编号	EPR6107
同种型	IgG

应用

The Abpromise guarantee **Abpromise™** 承诺保证使用 ab124680 于以下的经测试应用

“应用说明”部分 下显示的仅为推荐的起始稀释度；实际最佳的稀释度/浓度应由使用者检定。

应用	Ab 评论	说明
WB		1/1000 - 1/10000. Detects a band of approximately 17, 30 kDa (predicted molecular weight: 19, 23 kDa).

应用说明 Is unsuitable for IHC-P.

靶标

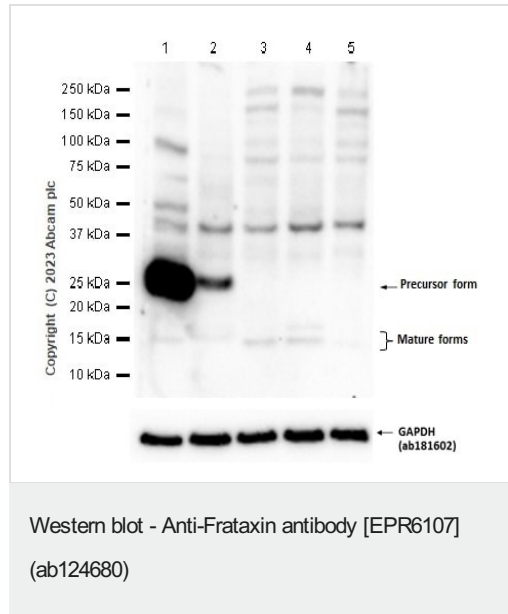
功能	Promotes the biosynthesis of heme and assembly and repair of iron-sulfur clusters by delivering Fe(2+) to proteins involved in these pathways. May play a role in the protection against iron-catalyzed oxidative stress through its ability to catalyze the oxidation of Fe(2+) to Fe(3+); the oligomeric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



All lanes : Anti-Frataxin antibody [EPR6107] (ab124680) at 1/1000 dilution

Lane 1 : Human liver tissue lysate

Lane 2 : Human hippocampus tissue

Lane 3 : HCT116 (human colorectal carcinoma epithelial cell)

whole cell lysate

Lane 4 : Huh7 (human hepatocellular carcinoma epithelial cell)

whole cell lysate

Lane 5 : SH-SY5Y (human neuroblastoma epithelial cell) whole cell lysate

Lysates/proteins at 20 µg per lane.

Secondary

All lanes : Goat Anti-Rabbit IgG H&L (HRP) ([ab97051](#)) at 1/20000 dilution

Predicted band size: 19, 23 kDa

Observed band size: 14,17 kDa

Exposure time: 60 seconds

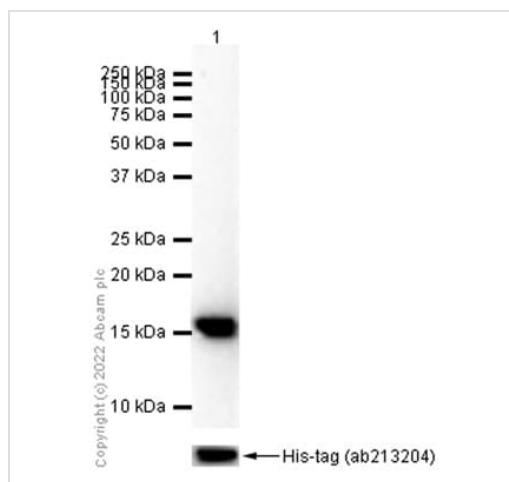
Blocking buffer and concentration: 5% NFDM/TBST.

Diluting buffer and concentration: 5% NFDM/TBST.

This blot was developed using a high sensitivity ECL substrate.

For different forms of frataxin, you can refer to PMID: 17468497, PMID: 31279523, PMID: 17468497 etc.

[ab181602](#) was used as a loading control.



Western blot - Anti-Frataxin antibody [EPR6107]
(ab124680)

Anti-Frataxin antibody [EPR6107] (ab124680) at 1/1000 dilution +
His tagged Human FXN recombinant protein (aa 82-210), 10ng

Secondary

Goat Anti-Rabbit IgG H&L (HRP) ([ab97051](#)) at 1/20000 dilution

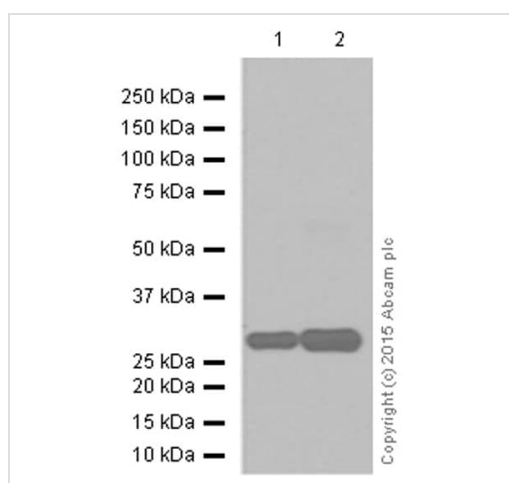
Predicted band size: 19, 23 kDa

Observed band size: 16 kDa

Exposure time: 60 seconds

Blocking and diluting buffer and concentration: 5% NFDm/TBST.

[ab213204](#) was used as a loading control.



Western blot - Anti-Frataxin antibody [EPR6107]
(ab124680)

All lanes : Anti-Frataxin antibody [EPR6107] (ab124680) at 1/5000
dilution (purified)

Lane 1 : Human fetal heart tissue lysate

Lane 2 : Human skin lysate

Lysates/proteins at 20 µg per lane.

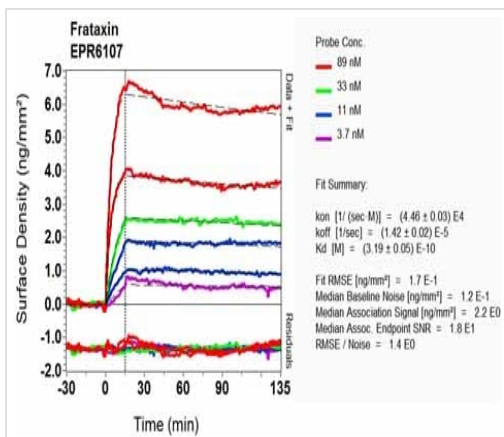
Secondary

All lanes : Anti-Rabbit IgG (HRP), specific to the non-reduced form
of IgG at 1/2000 dilution

Predicted band size: 19, 23 kDa

Observed band size: 30 kDa

Blocking and diluting buffer 5% NFDm/TBST



SPR Scanning - Anti-Frataxin antibody [EPR6107]
(ab124680)

Equilibrium dissociation constant (K_D)

Learn more about K_D

[Click here to learn more about \$K_D\$](#)

Why choose a recombinant antibody?



Research with confidence
Consistent and reproducible results



Long-term and scalable supply
Recombinant technology



Success from the first experiment
Confirmed specificity



Ethical standards compliant
Animal-free production

Anti-Frataxin antibody [EPR6107] (ab124680)

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

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