

Anti-Frataxin antibody ab175402

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Overview

Product name	Anti-Frataxin antibody
Description	Rabbit polyclonal to Frataxin
Host species	Rabbit
Tested applications	Suitable for: ICC/IF, IHC-P, WB
Species reactivity	Reacts with: Mouse, Rat, Human
Immunogen	Recombinant full length protein corresponding to Human Frataxin. Database link: Q16595
Positive control	WB: Jurkat, K562, Raji, DU 145 and HL-60 cell lysates; Mouse heart and liver tissue lysates. IHC-P: Mouse and rat heart tissues; ICC/IF: L929 cells.
General notes	<p>The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing.</p> <p>If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As</p>

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.
Storage buffer	pH: 7.30 Preservative: 0.02% Sodium azide Constituents: 50% Glycerol, 49% PBS
Purity	Immunogen affinity purified
Clonality	Polyclonal
Isotype	IgG

Applications

The Abpromise guarantee

Our **Abpromise guarantee** covers the use of ab175402 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
ICC/IF		Use at an assay dependent concentration.
IHC-P		1/50 - 1/200. ab171870 - Rabbit polyclonal IgG, is suitable for use as an isotype control with this antibody.
WB		1/500 - 1/2000. Detects a band of approximately 14 kDa (predicted molecular weight: 23 kDa).

Target

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.

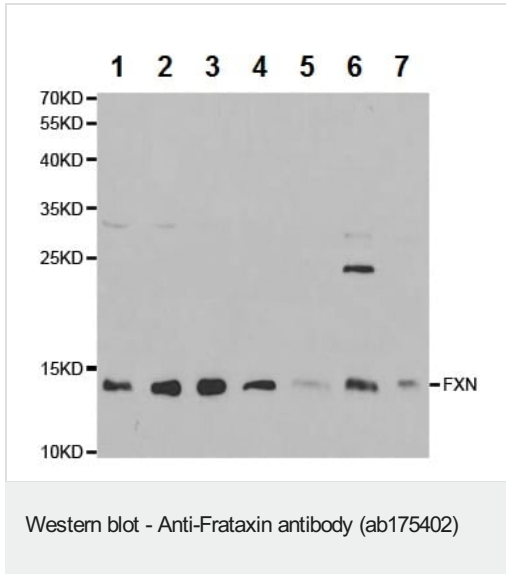
Post-translational modifications

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



All lanes : Anti-Frataxin antibody (ab175402) at 1/1000 dilution

Lane 1 : Jurkat (human T cell leukemia cell line from peripheral blood) cell lysate

Lane 2 : K562 (human chronic myelogenous leukemia cell line from bone marrow) cell lysate

Lane 3 : Raji (human Burkitt's lymphoma cell line) cell lysate

Lane 4 : DU 145 (human prostate carcinoma cell line) cell lysate

Lane 5 : HL-60 (human promyelocytic leukemia cell line) cell lysate

Lane 6 : Mouse heart lysate

Lane 7 : Mouse liver lysate

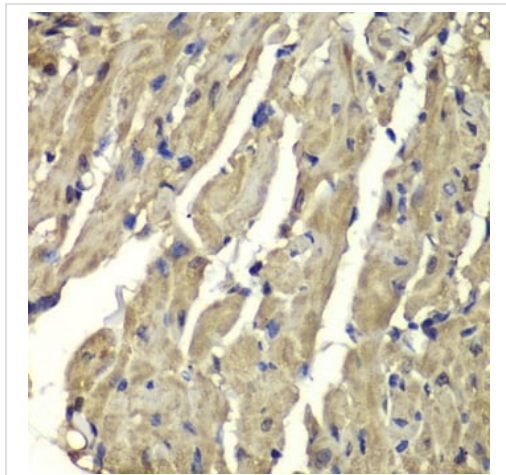
Lysates/proteins at 25 µg per lane.

Secondary

All lanes : HRP Goat Anti-Rabbit IgG

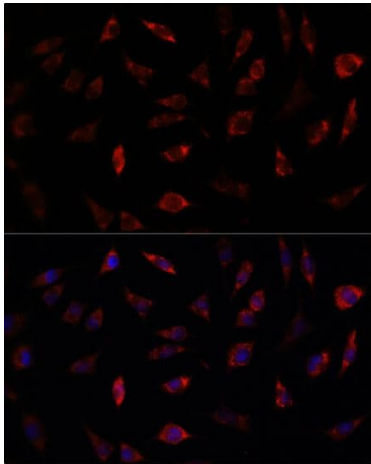
Predicted band size: 23 kDa

Observed band size: 14 kDa



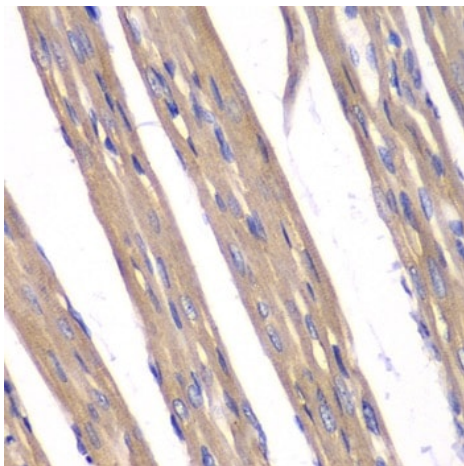
Paraffin-embedded mouse heart tissue labelling Frataxin using ab175402 at 1/100 dilution in immunohistochemical analysis.

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Frataxin antibody (ab175402)



Immunofluorescence staining of L929 cells stained for Frataxin with ab175402 at 1/100 dilution. DAPI was used as a nuclear counterstain.

Immunocytochemistry/ Immunofluorescence - Anti-Frataxin antibody (ab175402)



Paraffin-embedded rat heart tissue labelling Frataxin using ab175402 at 1/100 dilution in immunohistochemical analysis.

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Frataxin antibody (ab175402)

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