abcam

ab110328

Anti-Frataxin antibody [18A5DB1]

Anti-Frataxin antibody [18A5DB1] (ab110328) is a mouse monoclonal antibody detecting Frataxin in **Western Blot, Flow Cytometry, IHC-P, ICC/IF**. Suitable for **Human**.

- Over 30 publications

Key facts

Isotype	IgG1
Host species	Mouse
Storage buffer	pH: 7.5 Preservative: 0.02% Sodium azide Constituents: HEPES buffered saline
Form	Liquid
Clonality	Monoclonal
Immunogen	The exact immunogen used to generate this antibody is proprietary information.
Clone number	18A5DB1
Purity	IgG fraction
Light chain type	kappa
Concentration	1 mg/mL The concentration of this product may be batch-dependent Batch concentration finder →
Purification notes	ab110328 was produced in vitro using hybridomas grown in serum-free medium, and then purified by biochemical fractionation.

Reactivity data

Flow Cyt

Tested

Species	Human
Dilution info	1 µg/mL
Notes	ab170190 - Mouse monoclonal IgG1, is suitable for use as an isotype control with this antibody.

WB

Tested

Species	Human
Dilution info	3-5 µg/mL
Notes	(A chemiluminescent detection system is highly recommended)

ICC/IF

Tested

Species	Human
Dilution info	0.5 µg/mL
Notes	-

IHC-P

Tested

Species	Human
Dilution info	10 µg/mL
Notes	-

Target data

See full target information FXN

Function	Frataxin mature form. Functions as an activator of persulfide transfer to the scaffoding protein ISCU as component of the core iron-sulfur cluster (ISC) assembly complex and participates to the [2Fe-2S] cluster assembly (PubMed:12785837, PubMed:24971490). Accelerates sulfur transfer from NFS1 persulfide intermediate to ISCU and to small thiols such as L-cysteine and glutathione leading to persulfuration of these thiols and ultimately sulfide release (PubMed:24971490). Binds ferrous ion and is released from FXN upon the addition

of both L-cysteine and reduced FDX2 during [2Fe-2S] cluster assembly (PubMed:29576242). The core iron-sulfur cluster (ISC) assembly complex is involved in the de novo synthesis of a [2Fe-2S] cluster, the first step of the mitochondrial iron-sulfur protein biogenesis. This process is initiated by the cysteine desulfurase complex (NFS1:LYRM4:NDUFAB1) that produces persulfide which is delivered on the scaffold protein ISCU in a FXN-dependent manner. Then this complex is stabilized by FDX2 which provides reducing equivalents to accomplish the [2Fe-2S] cluster assembly. Finally, the [2Fe-2S] cluster is transferred from ISCU to chaperone proteins, including HSCB, HSPA9 and GLRX5 (By similarity). 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 (PubMed:15641778). 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 (PubMed:11823441, PubMed:12755598). May function as an iron chaperone protein that protects the aconitase [4Fe-4S]2+ cluster from disassembly and promotes enzyme reactivation (PubMed:15247478). May play a role as a high affinity iron binding partner for FECH that is capable of both delivering iron to ferrochelatase and mediating the terminal step in mitochondrial heme biosynthesis (PubMed:15123683, PubMed:16239244). Extramitochondrial frataxin. Modulates the RNA-binding activity of ACO1 (PubMed:20053667). May be involved in the cytoplasmic iron-sulfur protein biogenesis (PubMed:16091420). May contribute to oxidative stress resistance and overall cell survival (PubMed:16608849).

Storage

Shipped at conditions	Blue Ice
Appropriate short-term storage conditions	+4°C
Appropriate long-term storage conditions	+4°C
Storage information	Do Not Freeze

Notes

Although this antibody does work in Western blot, we have found that ab113691 typically gives a more robust signal. Therefore, if you plan on using this antibody in WB only, we recommend ab113691 as an alternative.

Product Specifications

Anti-Frataxin antibody [18A5DB1] (ab110328) is a mouse monoclonal antibody and is validated for use in Flow Cyt, ICC/IF, IHC-P, WB in human samples.

Anti-Frataxin antibody [18A5DB1] (ab110328) specifically detects Frataxin (UniProt ID: Q16595; Molecular weight: 19kDa) and is sold in 100 µg and 1 mg selling sizes.

Quality and Validation

Abcam's high quality validation processes ensure Anti-Frataxin antibody [18A5DB1] (ab110328) has high sensitivity and specificity.
Anti-Frataxin antibody [18A5DB1] (ab110328) has been cited over 36 times in peer reviewed journals and is trusted by the scientific community.
Anti-Frataxin antibody [18A5DB1] (ab110328) has 5 independent reviews from customers.

Related Products
Antibody clone 18A5DB1 is also available pre-conjugated to a variety of labels for your convenience - Alexa Fluor® 488 (ab156033).

Want a custom formulation?
This antibody clone is manufactured by Abcam. If you require a custom buffer formulation or conjugation for your experiments, please contact orders@abcam.com

Supplementary info

This supplementary information is collated from multiple sources and compiled automatically.

Activity summary	Frataxin often known by the alternate name FXN is a mitochondrial protein with a mass of approximately 21000 Dalton. It is expressed mainly in tissues with high energy demands like the heart liver and pancreas. Frataxin plays an important role in iron-sulfur cluster assembly which is essential for various cellular processes. The protein is a part of mitochondria where it regulates iron homeostasis and prevents oxidative damage by minimizing iron-induced free radical generation.
Biological function summary	Several cellular processes depend on the correct function of this protein. Frataxin assists in forming iron-sulfur clusters acting within a multiprotein complex in the mitochondria. The complex includes proteins such as ISCU which are involved in the assembly and repair of iron-sulfur clusters. These clusters are necessary for supporting mitochondrial electron transport and other fundamental metabolic pathways that require iron-sulfur dependencies.
Pathways	Frataxin's involvement extensively affects the mitochondrial respiratory chain and the mitochondrial biogenesis process. It plays a role in the electron transport chain by stabilizing iron-sulfur-containing complexes. NAB is one associated protein that interacts closely within these pathways sharing a connection through iron-sulfur cluster transportation and assembly systems. Efficient function of these pathways ensures a proper energetic output of cells.
Associated diseases and disorders	Frataxin mutations are directly linked to Friedreich's ataxia a neurodegenerative disease causing progressive damage to the nervous system. The deficiency or dysfunction in frataxin causes accumulation of iron in mitochondria leading to increased oxidative stress. Another related disorder includes heart disease which emerges due to the same oxidative stress pathway. Proteins such as Nfs1 are also involved sharing the responsibility with frataxin in scavenging excess iron protecting against related tissue damage.

Product promise

Tested

We have tested this species and application combination and it works. It is covered by our product promise.

Expected

We have not tested this specific species and application combination in-house, but expect it will work. It is covered by our product promise.

Predicted

This species and application combination has not been tested, but we predict it will work based on strong homology. However, this combination is not covered by our product promise.

Not recommended

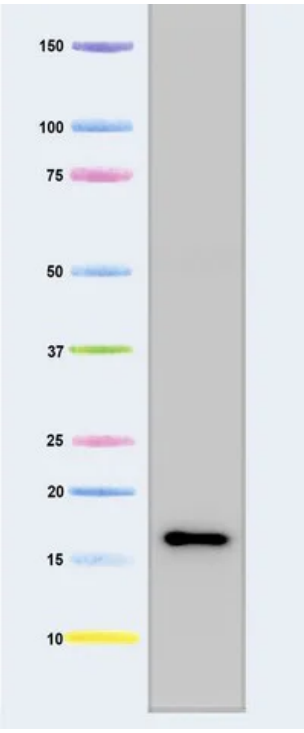
We do not recommend this combination. It is not covered by our product promise.

We are dedicated to supporting your work with high quality reagents and we are here for you every step of the way should you need us.

In the unlikely event of one of our products not working as expected, you are covered by our product promise.

Full details and terms and conditions can be found here:
Terms & Conditions.

10 product images



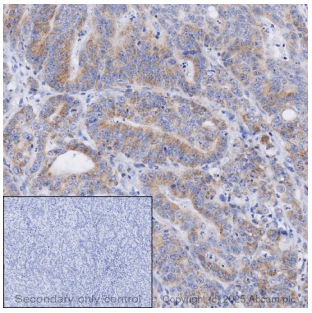
Western blot - Anti-Frataxin antibody [18A5DB1] (ab110328)

Frataxin Western blot staining of Recombinant Human Frataxin using mouse Anti-Frataxin antibody

All lanes:
Western blot - Anti-Frataxin antibody [18A5DB1] (ab110328) at 5 µg/mL

All lanes:
Recombinant Human Frataxin at 0.0005 µg

Predicted band size: 23 kDa



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Frataxin antibody [18A5DB1] (ab110328)

Frataxin Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) staining of Human colon adenocarcinoma using mouse Anti-Frataxin antibody

Immunohistochemical analysis of formalin fixed paraffin embedded human colon adenocarcinoma labelling frataxin with ab110328 at a concentration of 0.5µg/ml. The immunostaining was performed on a Ventana DISCOVERY ULTRA (Roche Tissue Diagnostics) instrument with a OptiView DAB IHC Detection Kit. Heat mediated antigen retrieval was performed with DISCOVERY cell conditioning solution (CC1) 100°C, pH8.5 for 32 mins. ab110328 anti-Frataxin antibody [18A5DB1] was incubated for 16 mins at 37°C. Sections were counterstained with Hematoxylin II. Image inset shows absence of staining in secondary antibody only control. Customers are encouraged to optimise antigen retrieval conditions, antibody concentration, incubation times and temperature for best results in their own IHC assay workflow (automated and manual).

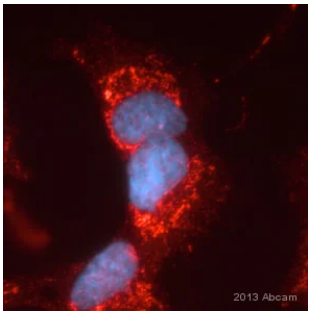
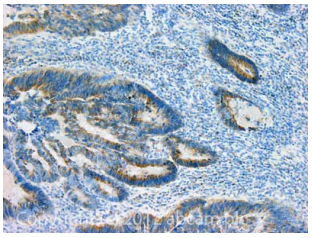


Image courtesy of Dr George Allen

Immunocytochemistry/ Immunofluorescence - Anti-Frataxin antibody [18A5DB1] (ab110328)

Frataxin Immunocytochemistry/ Immunofluorescence staining using mouse Anti-Frataxin antibody

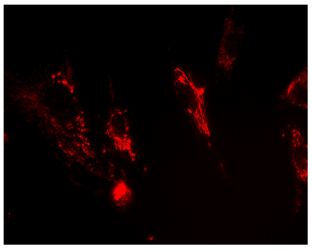
Immunofluorescent analysis of paraformaldehyde-fixed, 0.2% NP-40 permeabilized SH-SY5Y (human neuroblastoma cell line from bone marrow) cells labeling Frataxin with ab110328 at 1/500 dilution, followed by Goat anti-MouseAlexaFluor® 594 conjugated secondary antibody at 1/500 dilution (red). The nuclear counter stain is DAPI (blue).



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Frataxin antibody [18A5DB1] (ab110328)

Frataxin Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) staining using mouse Anti-Frataxin antibody

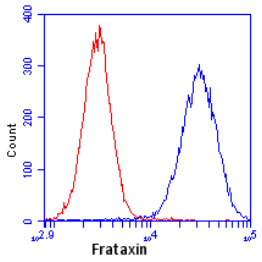
IHC image of Frataxin staining in Human colon adenocarcinoma formalin fixed paraffin embedded tissue section, performed on a Leica BondTM system using the standard protocol F. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH6, epitope retrieval solution 1) for 20 mins. The section was then incubated with ab110328, 10µg/ml, for 15 mins at room temperature and detected using an HRP conjugated compact polymer system. DAB was used as the chromogen. The section was then counterstained with haematoxylin and mounted with DPX.
For other IHC staining systems (automated and non-automated) customers should optimize variable parameters such as antigen retrieval conditions, primary antibody concentration and antibody incubation times.



Immunocytochemistry/ Immunofluorescence - Anti-Frataxin antibody [18A5DB1] (ab110328)

Frataxin Immunocytochemistry/ Immunofluorescence staining using mouse Anti-Frataxin antibody

Immunocytochemistry analysis using ab110328 at 0.5µg/ml staining Frataxin in normal Human fibroblasts (MRC5) (fixed and permeabilized) followed by an AlexaFluor® 594-conjugated-goat-anti-mouse IgG1 isotype specific secondary antibody (2 µg/ml). .



Flow Cytometry - Anti-Frataxin antibody [18A5DB1] (ab110328)

Frataxin Flow Cytometry staining using mouse Anti-Frataxin antibody

Flow cytometric analysis using ab110328 at 1µg/ml staining Frataxin in HL60 cells (blue). Isotype control antibody (red).

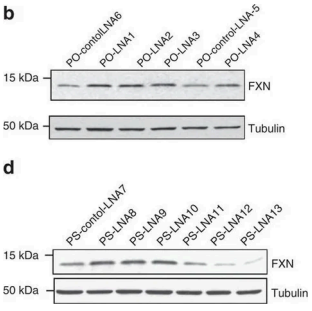


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Western blot - Anti-Frataxin antibody [18A5DB1] (ab110328)

Frataxin Western blot staining using mouse Anti-Frataxin antibody

Western Blotting using Anti-Frataxin antibody [18A5DB1], ab110328. Publication image from Corey, D. R. et al., 2016, Nat Commun, 26842135. Legend direct from paper.

LNA-mediated activation of FXN expression.(a) Structure of LNA. (b) Western analysis of the effect of LNAs with phosphodiester (PO) backbone on FXN protein expression. (c) Quantitation of western analysis (n=2). (d) Western analysis of the effect of LNAs with phosphorothioate (PS) backbone on FXN protein expression. (e) Quantitation of quadruplicate western analysis. (f,g) qPCR showing effect on FXN mRNA expression of (f) PO LNAs (n=5) or (g) PS LNAs (n=3). PO-control-LNA5 is a negative control LNA with PO backbone that is not complementary to FXN RNA. PO-control-LNA6 has five mismatches relative to the repeat region within FXN RNA target. PS-control-LNA7 is a negative control LNA similar to PO-control-LNA5 but with PS backbone. FRDA patient fibroblast cells (GM03816) were treated with 12.5 nM duplex LNAs. Cells were collected at day 3 for RNA extraction and day 4 for protein extraction. All data are presented as mean±STDEV. NT, no treatment; **P<0.01, by Student t-test.

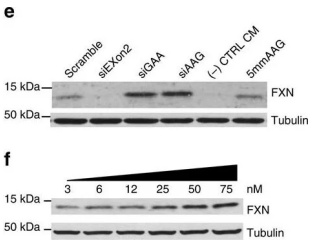


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Western blot - Anti-Frataxin antibody [18A5DB1] (ab110328)

Frataxin Western blot staining using mouse Anti-Frataxin antibody

Western Blotting using Anti-Frataxin antibody [18A5DB1], ab110328. Publication image from Corey, D. R. et al., 2016, Nat Commun, 26842135. Legend direct from paper.

RNA-mediated activation of FXN expression.(a) Schematic of repeat expansion within intronic FXN mRNA and binding of AGO:RNA complexes. The longer mutant repeat is predicted to bind more AGO:RNA complexes than the shorter wild-type repeat. (b) Schematic of R-loop formation at FXN locus and potential to influence histone modification and gene expression (Adapted from Groh et al.19). (c) Complementarity of guide strand RNAs (25 nM) to FXN RNA. (d,e) Effect of anti-GAA duplex RNAs on d, FXN mRNA (n=3) and e, protein expression. (f) A dose-response profile of upregulation of FXN protein expression by siGAA. FRDA patient-derived fibroblast cells (GM03816) were used. siExon2 is a duplex siRNA that targets FXN exon and is expected to decrease FXN expression. CM is a negative control RNA that is not complementary to FXN RNA. RNA scramble is a duplex RNA in which the sequences of siGAA and siAAG are mixed to preserve nucleotide composition by alter their order. si5mmAAG is similar to siAAG but has five mismatches relative to the repeat region within FXN mRNA target. Cells were collected at day 3 for RNA extraction and day 4 for protein extraction. Error bars:±STDEV. **P<0.01, by Student t-test.

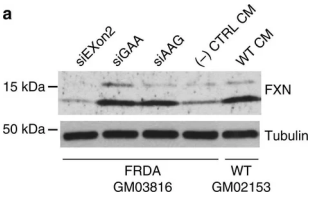


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Western blot - Anti-Frataxin antibody [18A5DB1] (ab110328)

Frataxin Western blot staining using mouse Anti-Frataxin antibody

Western Blotting using Anti-Frataxin antibody [18A5DB1], ab110328. Publication image from Corey, D. R. et al., 2016, Nat Commun, 26842135. Legend direct from paper.

Comparison of RNA-mediated activation of FXN expression in patient cells to FXN expression in normal cells and FXN activation by a histone deacetylase inhibitor.(a) Activation of FXN protein expression in FRDA cells (GM03816) and wild-type fibroblast cells (GM02153). (b,c) Effect of HDAC inhibitor BML210 (5 µM) treatment on expression of bFXN mRNA (n=3) and (c) protein (inset, western analysis, n=4) expression in FRDA patient fibroblast cells (GM03816). All data are presented as mean±STDEV.

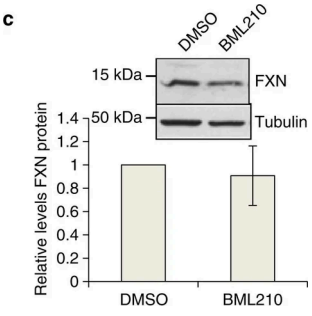


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