

Frataxin Antibody / Friedreich Ataxia Protein Antibody [clone FDAX-1] (V3997)

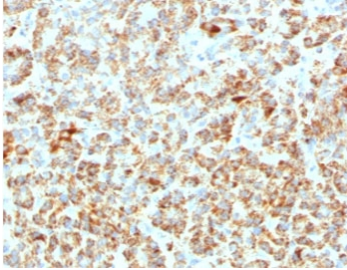
Catalog No.	Formulation	Size
V3997-100UG	0.2 mg/ml in 1X PBS with 0.1 mg/ml BSA (US sourced) and 0.05% sodium azide	100 ug
V3997-20UG	0.2 mg/ml in 1X PBS with 0.1 mg/ml BSA (US sourced) and 0.05% sodium azide	20 ug
V3997SAF-100UG	1 mg/ml in 1X PBS; BSA free, sodium azide free	100 ug
V3997IHC-7ML	Prediluted in 1X PBS with 0.1 mg/ml BSA (US sourced) and 0.05% sodium azide; *For IHC use only*	7 ml

Bulk quote request

Availability	1-3 business days
Species Reactivity	Human
Format	Purified
Host	Mouse
Clonality	Monoclonal (mouse origin)
Isotype	Mouse IgG2b, kappa
Clone Name	FDAX-1
Purity	Protein G affinity chromatography
UniProt	Q16595
Localization	Cytoplasmic
Applications	Western Blot : 1-2ug/ml Immunohistochemistry (FFPE) : 1-2ug/ml for 30 min at RT Prediluted IHC Only Format : incubate for 30 min at RT (1)
Limitations	This Frataxin Antibody / Friedreich Ataxia Protein Antibody is available for research use only.



Frataxin Antibody HepG2 Mitochondrial WB. Western blot analysis of human HepG2 cell lysate using monoclonal clone FDAX-1 demonstrates a distinct band near 14-15 kDa, consistent with the mature processed form of Frataxin / FXN following mitochondrial import-associated cleavage. The observed signal supports expression of this mitochondrial iron homeostasis protein in hepatic-derived cells and aligns with the established role of FXN in iron-sulfur cluster assembly and oxidative metabolic regulation.



Frataxin Antibody Pancreatic Mitochondrial IHC. Immunohistochemistry staining of FFPE human pancreas tissue using monoclonal clone FDAX-1 demonstrates widespread granular cytoplasmic HRP-DAB brown staining within pancreatic epithelial and acinar-associated cellular populations, consistent with mitochondrial localization of Frataxin / FXN in metabolically active tissue compartments. The observed staining pattern supports the established role of FXN in mitochondrial iron regulation and oxidative phosphorylation-associated energy metabolism. Required HIER: boil tissue sections in pH 9 10mM Tris with 1mM EDTA for 10-20 min and allow to cool before testing.

Description

Frataxin (FXN) is a mitochondrial matrix-associated protein involved in iron homeostasis, iron-sulfur cluster biogenesis, oxidative phosphorylation, and mitochondrial metabolic regulation. Frataxin Antibody / Friedreich Ataxia Protein Antibody recognizes a highly conserved mitochondrial protein that contributes to cellular energy production pathways and protection against mitochondrial oxidative stress-associated damage.

Frataxin antibody, also referred to as FXN antibody and Friedreich ataxia protein antibody in the literature, is widely used in mitochondrial biology, neurodegeneration, iron metabolism, and oxidative stress research applications. Monoclonal clone FDAX-1 supports investigation of mitochondrial iron handling pathways and Frataxin-associated metabolic signaling mechanisms linked to neurologic and mitochondrial disease biology.

FXN is synthesized as a precursor protein that undergoes mitochondrial import and proteolytic maturation to generate the active mature Frataxin protein localized within the mitochondrial matrix. Within mitochondria, Frataxin interacts with iron-sulfur cluster assembly machinery including ISCU scaffold proteins and cysteine desulfurase-associated complexes involved in generation of iron-sulfur cofactors required for oxidative metabolism and respiratory chain enzyme activity.

Reduced Frataxin expression is strongly associated with Friedreich ataxia, an inherited neurodegenerative disorder characterized by progressive ataxia, cardiomyopathy, mitochondrial dysfunction, and abnormal iron accumulation within affected tissues. Defective FXN-associated signaling contributes to impaired iron-sulfur cluster formation, oxidative stress accumulation, mitochondrial injury, and altered energy metabolism.

Because Frataxin functions at the intersection of mitochondrial metabolism and iron regulation, FXN has become increasingly important in studies examining oxidative stress adaptation, mitochondrial quality control pathways, neurodegenerative signaling, and metabolic homeostasis. Altered mitochondrial iron handling may additionally influence aging-associated pathways and cellular stress response signaling networks.

Western blot analysis with Frataxin antibodies commonly identifies mature FXN near approximately 14-15 kDa, while precursor forms may migrate at higher molecular weights depending on processing state and mitochondrial import-associated cleavage. Immunohistochemistry and immunofluorescence staining frequently demonstrate granular cytoplasmic patterns consistent with mitochondrial localization in metabolically active cellular populations.

Clone FDAX-1 supports investigation of Frataxin-associated mitochondrial pathways and contributes to studies examining iron-sulfur cluster assembly, oxidative phosphorylation regulation, and mitochondrial dysfunction-associated disease mechanisms. The established role of FXN in Friedreich ataxia and mitochondrial metabolism further supports the value of

Frataxin antibodies in neurologic and metabolic research applications.

Together, the available biologic characterization supports the use of Frataxin antibody clone FDAX-1 for investigating mitochondrial iron homeostasis, oxidative metabolism-associated pathways, and Frataxin-linked mitochondrial signaling biology.

For additional Frataxin and mitochondrial metabolism research antibodies targeting iron-sulfur cluster assembly, oxidative phosphorylation pathways, and mitochondrial iron homeostasis proteins, explore the broader [FXN Antibody](#) page featuring monoclonal clone FXN/2124.

Application Notes

Optimal dilution of the Frataxin Antibody / Friedreich Ataxia Protein Antibody should be determined by the researcher.

1. The prediluted format is supplied in a dropper bottle and is optimized for use in IHC. After epitope retrieval step (if required), drip mAb solution onto the tissue section and incubate at RT for 30 min.

Immunogen

Amino acids 57-210 from the human protein were used as the immunogen for this Frataxin antibody.

Storage

Store the Frataxin antibody at 2-8oC (with azide) or aliquot and store at -20oC or colder (without azide).

Alternate Names

Frataxin antibody, FXN antibody, Friedreich ataxia protein antibody, mitochondrial iron homeostasis protein antibody, iron-sulfur cluster assembly protein antibody