

FXN Antibody / Frataxin (FY13347)

Catalog No.	Formulation	Size
FY13347	Adding 0.2 ml of distilled water will yield a concentration of 500 ug/ml	100 ug

Bulk quote request

Availability	1-2 days
Species Reactivity	Human
Format	Lyophilized
Clonality	Polyclonal (rabbit origin)
Isotype	Rabbit IgG
Purity	Immunogen affinity purified
Buffer	Each vial contains 4 mg Trehalose, 0.9 mg NaCl, 0.2 mg Na2HPO4.
UniProt	Q16595
Applications	Western Blot: 0.25-0.5ug/ml Immunohistochemistry: 2-5ug/ml Flow Cytometry: 1-3ug/million cells ELISA: 0.1-0.5ug/ml
Limitations	This FXN antibody is available for research use only.

Description

FXN antibody detects Frataxin, a mitochondrial matrix protein encoded by the FXN gene on chromosome 9q21.11. Frataxin plays a fundamental role in iron-sulfur (Fe-S) cluster biogenesis, iron homeostasis, and oxidative stress protection. It is ubiquitously expressed but highly abundant in metabolically active tissues such as heart, skeletal muscle, pancreas, and neurons, where mitochondrial energy metabolism is critical. FXN belongs to the frataxin family of mitochondrial proteins and is essential for the proper function of enzymes involved in oxidative phosphorylation and the tricarboxylic acid (TCA) cycle.

FXN acts as an iron chaperone that delivers ferrous iron (Fe²?) to scaffold proteins such as ISCU for the assembly of FeS clusters. These clusters serve as cofactors for numerous mitochondrial enzymes, including aconitase and complex I–III components of the respiratory chain. By regulating mitochondrial iron utilization, FXN prevents toxic iron accumulation and the generation of reactive oxygen species (ROS). Co-localization studies demonstrate FXN interaction with ISCU and NFS1 within mitochondrial nucleoids, coordinating Fe-S cluster synthesis and incorporation into target proteins.

Structurally, Frataxin is a small alphaÂ-beta fold protein with a conserved acidic ridge that binds iron ions. It belongs to the mitochondrial Fe-S cluster assembly machinery family. FXN also associates with chaperones and proteases such as HSP60 and Lon protease, maintaining protein stability and turnover in the mitochondrial matrix. It undergoes processing from a cytosolic precursor into a mature mitochondrial form via the mitochondrial targeting sequence at its N-terminus.

Functionally, FXN is crucial for cellular respiration, antioxidant defense, and metabolic balance. It supports the activity of Fe-S-dependent enzymes in the electron transport chain and modulates mitochondrial redox status. In neurons, FXN ensures energy supply and protects against oxidative stress, while in cardiac tissue, it maintains contractile efficiency by preserving mitochondrial integrity. Developmentally, FXN expression peaks during embryogenesis and early postnatal stages, coinciding with rapid mitochondrial biogenesis in growing tissues.

Loss-of-function mutations in FXN cause FriedreichÂ's ataxia, an autosomal recessive neurodegenerative disease characterized by impaired motor coordination, cardiomyopathy, and diabetes. The deficiency leads to mitochondrial iron overload, defective Fe-S cluster assembly, and increased oxidative stress. Pathway involvement includes Fe-S cluster biosynthesis, oxidative phosphorylation, and cellular stress response. In cancer research, FXN downregulation is associated with altered metabolic reprogramming and mitochondrial dysfunction.

Immunohistochemical staining using FXN antibody shows mitochondrial localization in neurons, cardiac myocytes, and hepatocytes. The FXN antibody from NSJ Bioreagents is a reliable reagent for studying mitochondrial metabolism, iron homeostasis, and neurodegenerative disease mechanisms such as FriedreichÂ's ataxia.

Application Notes

Optimal dilution of the FXN antibody should be determined by the researcher.

Immunogen

E.coli-derived human FXN recombinant protein (Position: Q20-A210) was used as the immunogen for the FXN antibody.

Storage

After reconstitution, the FXN antibody can be stored for up to one month at 4oC. For long-term, aliquot and store at -20oC. Avoid repeated freezing and thawing.