Frataxin, mitochondrial Antibody

Mouse Monoclonal Antibody [Clone FXN/2124]

Catalog No	Format	Size
2395-MSM1-P0	Purified Ab with BSA and Azide at 200ug/ml	20 ug
2395-MSM1-P1	Purified Ab with BSA and Azide at 200ug/ml	100 ug
2395-MSM1-P1ABX	Purified Ab WITHOUT BSA or Azide at 1.0mg/ml	100 ug

Applications	Tested Dillution	Note
Immunohistochemistry (IHC)	1-2ug/ml	30 min at RT. Staining of formalin-fixed tissues requires heating tissue sections in 10mM Tris with 1mM EDTA, pH 9.0, for 45 min at 95°C followed by cooling at RT for 20 minutes
Western Blot (WB)	2-4ug/ml	

Product Details

Clone	FXN/2124
Immunogen	Recombinant fragment (around aa 57-210) of human FXN protein
Host	Mouse
Clonality	Monoclonal
Isotype / Light Chain	IgG2 / Kappa
Mol. Weight of Antigen	23.13kDa
Cellular Localization	Cytoplasm, Cytosol, Mitochondrion
Species Reactivity	Human
Positive Control	Expressed in the heart, peripheral blood lymphocytes and dermal fibroblasts

**Optimal dilution for a specific application should be determined.*

Product Images for Frataxin, mitochondrial Antibody

Specificity & Comments

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)., 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).

Supplied As

200ug/ml of Ab purified from Bioreactor Concentrate by Protein A. Prepared in 10mM PBS with 0.05% BSA & 0.05% azide. Also available WITHOUT BSA & azide at 1.0mg/ml.

Storage and Stability

Antibody with azide - store at 2 to 8 °C. Antibody without azide - store at -20 to -80 °C. Antibody is stable for 24 months. Non-hazardous. No MSDS required.

Limitations and Warranty

This antibody is available for research use only and is not approved for use in diagnosis.There are no warranties, expressed or implied, which extend beyond this description. Company is not liable for any personal injury or economic loss resulting from this product.