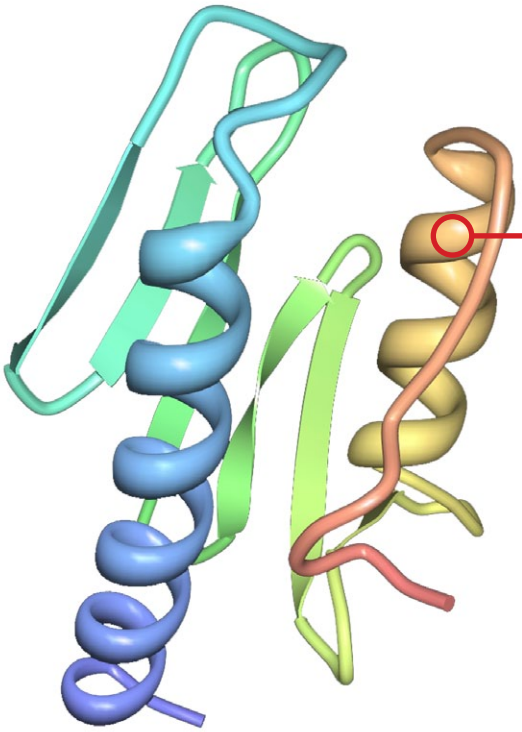
MITOCHONDRIAL DISEASE TOOLKIT

Friedreich's Ataxia



Solutions for Monitoring

Friedreich's ataxia patients or cell models, including measurement of frataxin levels and downstream effects of frataxin dysregulation.

All products are for Research or Investigational Use Only.
Not for use as diagnostic tests or for any other clinical applications.

MitoSciences®
Advancing vital discoveries in mitochondrial research

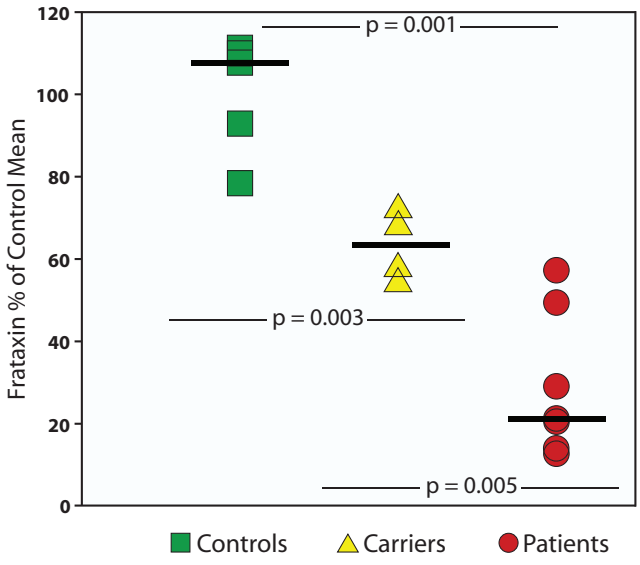
MitoSciences Products for Research into Friedreich's Ataxia

Frataxin

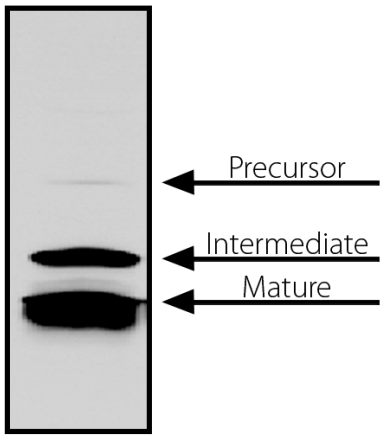
Frataxin plays an essential role in iron metabolism and is required for assembly and activation of iron-sulfur clusters, heme biosynthesis, iron detoxification and anti-oxidant protection. Therefore, frataxin deficiency results in widespread pleiotropic effects, including impaired energy metabolism, increased oxidative stress, neurodegeneration, and cardiotoxicity. Inherited deficiency of frataxin is the cause of Friedreich's Ataxia, the most common inherited human ataxia.

Catalog #	Product Name	Applications	Amount	Species Reactivity
Monoclonal Antibodies				
MSF01	Frataxin Antibody	WB, ICC, IP	100 µg	H
Protein Quantity Assays				
MSF31	Frataxin Protein Quantity Dipstick Assay Kit	ELISA	30 or 90 tests	H
MSF41	Frataxin Protein Quantity Microplate Assay Kit	ELISA	96 tests	H
Reference Protein				
MSF42	Frataxin Recombinant Protein (Human)		12 ng	

Frataxin levels measured with MSF31 in cultured lymphoblasts identify Friedreich's ataxia patients and carriers.



Western blots performed with anti-frataxin mAb MSF01 reveal precursor, intermediate, and mature forms of frataxin in whole cell extracts of cultured lymphoblasts.

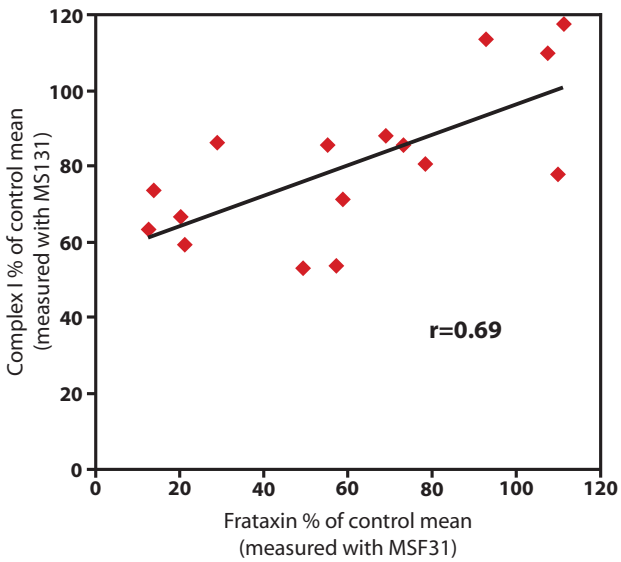


Downstream Effects of Frataxin Dysregulation

MitoSciences also has a wide range of reagents and assays that can be used to assess the downstream consequences of altered frataxin expression on enzyme levels, enzymatic activities, and expression pattern of proteins that depend on iron-containing prosthetic groups for activity and assembly, including:

- FeS cluster-containing enzymes such as OXPHOS complexes I, II, III, and aconitase.
- Heme-containing OXPHOS complex IV.
- Oxidative stress markers, including nitration levels on individual proteins and expression of the oxidative stress response protein SOD2.

Complex I protein levels correlate with frataxin protein levels in controls, carriers, and Friedreich's ataxia patients.



	Protein Quantification	Enzyme Activity	Immuno-precipitation	Immuno-cytochemistry	Western Blotting
OXPHOS Complex I	+	+	+	+	+
OXPHOS Complex II	+	+	+	+	+
OXPHOS Complex III	+		+	+	+
OXPHOS Complex IV	+	+	+	+	+
Aconitase		+			
SOD2	+				
Nitrotyrosine	+		+	+	+

Citations of peer-reviewed research in which MitoSciences anti-frataxin monoclonal antibodies and/or immunoassays have been used:

Western Blotting of frataxin with MSF01: citations 1, 2, 3, 4

Immunocytochemistry of frataxin with MSF01: citation 4

Immunoprecipitation of frataxin with MSF01: citations 3, 5

Quantitative assays of frataxin with MSF31 and MSF42: citations 7, 8, 9

Oxidative stress modifications of frataxin monitored with MSF31 and anti-nitrotyrosine MS703: citation 6

CITATIONS

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4. Long S., Jirku M., Ayala F.J., and Lukes J. (2008). Mitochondrial localization of human frataxin is necessary but processing is not for rescuing frataxin deficiency in Trypanosoma brucei. PNAS. 105(36):13468-13473. Epub 2008 Sep 3.

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MitoSciences®
1850 Millrace Drive, Suite 3A
Eugene, Oregon 97403
541.284.1800
www.mitosciences.com
sales@mitosciences.com