

**TECHNICAL DATA SHEET****Catalog # MSF41****Frataxin Protein Quantity Microplate Assay Kit**

Rev.1

LOT #:

APPLICATIONS: Analysis of frataxin quantity

SPECIES CROSS-  
REACTIVITY: Human

KIT COMPONENTS:

Item	MSF41
20X Buffer (Tube 1)	20 mL
10X Blocking Solution (Tube 2)	8 mL
Development Solution (Tube 3)	20 mL
Detergent	1 mL
20X Detector Antibody (Tube A)	1 mL
20X HRP Label (Tube B)	1 mL
96-well pre-coated microplate (12 strips)	1

STORAGE CONDITIONS: Store all tubes and the covered microplate at 4°C. The kit is stable for 6 months.

COUNTRY OF ORIGIN: USA

**BACKGROUND:**

Frataxin (Q16595) is encoded by a gene FXN on chromosome 9. Expression has been shown in many tissues where it is imported into the mitochondrial matrix and processed to a mature 17.3 kDa protein. The role of Frataxin is unclear however it has been proposed to function as an iron chaperone, an iron storage protein, as a repair enzyme of damaged FeS clusters in aconitase, as a defense against oxidative stress and activator of mitochondrial oxidative phosphorylation.

Partial frataxin deficiency in humans results in Friedrich ataxia (FRDA) an autosomal recessive disease characterized mainly by progressive neurodegeneration in the spinal cord, and frequently by hypertrophic cardiomyopathy and diabetes. FRDA is the most common form of ataxia (1:30,000-1:50,000 in US and Europe). In greater than 95% of cases the genetic basis of the deficiency is a triplet-nucleotide repeat of GAA in intron 1 of both Frataxin alleles. Normal individuals carry 'small number of repeat' (6-12) while less than 20% of normal individuals contain 'large normal repeats' (14-34). The role of these repeats, if any, is unknown. Individuals with large number repeats are at increased risk of producing offspring with expanded repeats (66-1500) which result in disease. Expanded repeats interfere with transcription of frataxin mRNA and result in a decrease in frataxin protein causing impairment of energy metabolism, increased oxidative stress, reduced heme biosynthesis, impaired iron metabolism and iron accumulation in the heart and nervous system.

The Frataxin Protein Quantity Microplate Assay Kit (Catalog # MSF41) is used to determine the quantity of this protein in a sample. Frataxin is immunocaptured within the wells. The quantity of protein is measured by adding a second frataxin specific antibody that is labeled with horseradish peroxidase. This peroxidase changes the substrate from colorless to blue. The rate of color development is proportional to the amount of protein captured in the well and can be monitored at 600 nm. Alternatively the assay can be terminated, at a user defined time, by the addition of 1N HCl (not supplied) and the assay performed as an end point measurement at 450 nm.

**Note: This product is for research purposes only. It is not to be used in humans or for diagnostic purposes.**

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