

TECHNICAL DATA SHEET**ab110353
(MSF42)****Recombinant Human Frataxin**

Rev. 6

LOT #:

APPLICATIONS: To generate a standard curve for use with Frataxin Assay Kits (MSF31, MSF41, and MS956).**SPECIES CROSS-REACTIVITY:** Human**KIT COMPONENTS:**

Part Number	Item	Amount
MSF42	Recombinant Human Frataxin (56-210)	12 ng lyophilized

STORAGE CONDITIONS: Ships at 4°C. Store lyophilized powder at 4°C. Reconstitute with 0.2 mL of H₂O to a final concentration of 60ng/mL. Reconstituted solution should be stored at -80°C.**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.

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

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