

TECHNICAL DATA SHEET**Catalog # MSF01****Frataxin monoclonal antibody**

Rev.0

LOT #:**COMPONENTS:** 100 µg monoclonal antibody**APPLICATIONS:** Western blotting and Immunocytochemistry**CLONE ID OF MONOCLONAL ANTIBODYV(mAb):** 18A5DB1**SPECIES CROSS-REACTIVITY:** human**HOST SPECIES AND ISOTYPE:** Mouse IgG1, k**IMMUNOGEN:** Human frataxin (full length mature form, residues 56-210)**CONCENTRATION:** 1 mg/mL in HEPES-Buffered Saline (HBS) with 0.02% azide as a preservative.**SUGGESTED WORKING CONCENTRATION:** 3-5 µg/mL for Western blotting (a chemiluminescent detection system is highly recommended), 0.5 µg/mL for Immunocytochemistry**mAb PURITY:** Near homogeneity as judged by SDS-PAGE. The antibody was produced *in vitro* using hybridomas grown in serum-free medium, and then purified by biochemical fractionation.**STORAGE CONDITIONS:** Store at 4°C. Do not freeze.**COUNTRY OF ORIGIN:** USA**BACKGROUND:**

Frataxin is a nuclear-encoded mitochondrial protein. In humans the gene is localized on chromosome 9 and is highly conserved during evolution. Frataxin is ubiquitously expressed, although levels of expression may vary in different tissues and during development.

The specific function of frataxin is still unknown, but it has been shown to play a role in iron metabolism. Studies have demonstrated that deletion of the frataxin gene in yeast results in iron accumulation in mitochondria and loss of respiration. Recombinant human frataxin has been shown to bind iron *in vitro*, and increased mitochondrial iron levels have been observed in patients with Friedreich's Ataxia (FRDA).

FRDA is an inherited recessive neurodegenerative disorder caused by decreased expression levels of frataxin. The most common mutation causing FRDA is the expansion of the GAA triplet repeat. Disease-associated repeats have 100 to 1700 triplets within the first intron of both copies of the gene. FRDA is the most common inherited ataxia and it causes progressive damage to the 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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