



MitoProfile[®] Dipstick Kit for Frataxin Quantity

INFORMATION SHEET

Catalog #: KHM1171

Description: Frataxin Quantity

Lot: See Product Label

Intended Use: Quantitative analysis of Frataxin

Monoclonal Antibody Clone ID: 17A11AC7 (capture antibody on dipstick)
18A5DB1 (dried gold-conjugated frataxin)

Species Cross Reactivity: human, mouse

Kit Component:

Item	Quantity
Dipsticks	30
Extraction Buffer	15 mL
Blocking Solution	1 mL
Wash Buffer	1 mL
Gold-Conjugated Antibody (dried in microplate wells)	1

Storage Condition: Store dipsticks at room temperature out of direct sunlight in their provided storage containers. Avoid high humidity conditions. Store Extraction Buffer, Blocking Solution, Wash Buffer, and microplate at 4°C.

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.

This product is for research use only. Not for use in diagnostic procedures.



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