



Anti-Frataxin Monoclonal Antibody

CATALOG #:	456300
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% sodium 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 <i>in vitro</i> 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.

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Related Products

<i>Product</i>	<i>Conjugate</i>	<i>Cat. No.</i>
Protein A	Sepharose 4B	10-1041
rec-Protein G	Sepharose 4B	10-1241
ZyMAX™ Goat anti-rabbit IgG	Unconjugated	81-6100
ZyMAX™ Goat anti-mouse IgG	Unconjugated	81-6500

Secondary Antibody Conjugates

<i>Conjugate</i>	<i>Goat anti-rabbit IgG (H+L)</i>	<i>Goat anti-mouse IgG (H+L)</i>	<i>Ex/Em*</i>	<i>Fluorescence similar to--</i>
Alexa Fluor® 488	A11008	A11001	495/519	FITC
Alexa Fluor® 555	A21428	A21422	555/565	Cy3
Alexa Fluor® 594	A11012	A11005	590/617	Texas Red
Alexa Fluor® 647	A21244	A21235	650/668	Cy5
HRP	81-6120	81-6520	NA**	NA
AP	81-6122	81-6522	NA	NA
Biotin	B2770	B2763	NA	NA

*Excitation/emission (nm); **Not applicable

For additional secondary antibody conjugates, visit www.invitrogen.com/antibodies

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