

## Product Specification Sheet

### Frataxin (FXN) Antibodies

<b>Cat. # FXN11-S</b>	Rabbit Anti-Mouse FXN antiserum	<b>SIZE:</b> 100 ul
<b>Cat. # FXN11-A</b>	Rabbit Anti-Mouse FXN IgG (aff pure)	<b>SIZE:</b> 100 ug
<b>Cat. # FXN11-P</b>	Mouse FXN Control/blocking peptide	<b>SIZE:</b> 100 ug

Elemental iron is required for a variety of normal cellular functions and vital for proper growth and development. However, natural iron is quite insoluble and excess iron is harmful, since it can catalyze the formation of potentially damaging reactive oxygen species. Humans also have very limited capacity to excrete iron. Therefore, cells have developed mechanisms to improve solubility of iron and to control intracellular iron levels at the point of absorption in the intestine and other tissue. Several proteins including **Ferritin**, **transferrin (Tf)**, **transferrin receptors (TfRs)**, and **iron regulatory proteins (IRPs)**, iron transporter (**NRMAMP2/DMT1/DCT1**) etc play a key role in iron metabolism. Some genes involved in iron-metabolism are associated with genetic disorders such as Friedreich's Ataxia (**Frataxin**), genetic hemochromatosis (**HFE**), and Sex-linked anemia (**Hephaestin**).

Friedrich ataxia (FA), a degenerative disease characterized by progressive ataxia and hypertrophic cardiomyopathy leading to premature death, is caused by a deficiency in **frataxin (Fxn)**. Fxn (mouse 207 aa, human 210 aa major isoform A; alternatively transcribed minor forms A1, 196 aa, and 171-aa isoform B) is a nuclear-encoded and highly conserved mitochondrial membrane protein. Fxn is expressed in tissues with highly metabolic activity, such as heart, liver, and brown fat. In FA, Fxn levels are greatly reduced due to inhibition of its transcription by expansion of an intronic GAA repeat. Defects in yeast Fxn produce mitochondrial iron-overload. It is suggested that Fxn could play a role in mitochondrial iron storage.

### Source of Antigen and Antibodies

<b>Antigen</b>	A 20-aa peptide sequence ( <b>designated FXN11-P or control peptide</b> ) within the C-terminus of <b>mouse FXN (1)</b> was synthesized, coupled to KLH
<b>Ab Host/type</b>	Rabbit, Polyclonal antiserum # <b>FXN11-S</b> and IgG, purified over antigen-agarose (Cat # <b>FXN11-A</b> )
<b>2-Ab</b>	Cat # 20320, goat anti-rabbit IgG-HRP (AP, biotin, FITC conjugates also available).
<b>-ve control IgG</b>	Cat # 20009-1, Rabbit (non-immune) Serum IgG, purified, suitable for ELISA, Western, IHC as -ve control

### Form & Storage of Antibodies/Peptide Control

**Antiserum (unpurified)**  
100ul solution lyophilized powder  
Supplied 0.05% azide, **Reconstitute** powder in 100 ul PBS

### Affinity pure IgG

100 ug/100ul solution lyophilized powder  
Supplied in **Buffer:** PBS+0.1% BSA  
**Reconstitute powder** in PBS at 1 mg/ml

### Control/blocking peptide

100 ug/100 ul solution lyophilized powder  
Supplied in **Buffer:** PBS pH 7.5,  
**Reconstitute powder** in PBS at 1 mg/ml.

### Storage

**Short-term:** unopened, undiluted liquid vials at -200C and powder at 4oC or -20oC..

**Long-term:** at -20C or below in suitable aliquots after reconstitution. Do not freeze and thaw and store working, diluted solutions.

**Stability:** 6-12 months at -20oC or below.

**Shipping:** 4oC for solutions and room temp for powder

### Recommended Usage

**Western Blotting** (1:1K-5K for antiserum and 1-10 ug/ml for aff. pure IgG using Chemiluminescence technique).

**ELISA** (1:100K; using 50-100 ng control peptide/well).

**Histochemistry & Immunofluorescence:** Not tested

### Specificity & Cross-reactivity

The FXN11-P sequence is 94% conserved in human FXN. Antibody cross-reactivity in various species has not been studied. Control peptides, because of its small size (2-3 kDa), is not recommended for Western. It should be used in ELISA or antibody blocking (use 5-10 ug control peptide per 1 ug of IgG or 1 ul of antiserum) Control peptide, because of its low mol. Wt (<3 kDa), is not suitable for Western. It should be used for ELISA or antibody blocking experiments (use 5-10 ug control peptide per 1 ug of aff pure IgG or 1 ul antiserum) to confirm antibody specificity (see detailed protocol at the web site).

**General References:** (1). Koutnikova H et al (1997) Nat. genet. 16, 345-351; Campuzano V et al (1996) Science 271, 1423-1427; Campuzano V et al (1997) Hum. Mol. Genet. 6, 1771-1780; Gordon DM et al (1999) Humn. Mol. Genet. 8, 2255-2262; Branda SS et al (1999) JBC 274, 22763-22769; Cavadini P et al (2000) JBC 275, 41469-41475

\*This product is for in vitro research use only.

### Some New Antibodies

IRP1-2, HFE, Frataxin, Hepcidin, Hephaestin, NRAMPs, USF2, Ferritin, Light and heavy chains, ferritin and B2-M ELISA, Tfr1-2, ceruloplasmin, B2-Micro globulin,

FXN11-S-A-P

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