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## FXN Antibody Blocking Peptide

Catalog Number: bs-9601P

Activity: Not tested

Purification: HPLC

Storage: Shipped at 4°C. Stored at -20°C for one year. Avoid repeated freeze/thaw cycles.

Background: Friedreich ataxia is a progressive neurodegenerative disorder caused by loss of function mutations in the frataxin gene. The human frataxin gene maps to chromosome 9q13. The frataxin gene encodes a mitochondrial protein of the same name. Frataxin assembles into a stable homopolymer with iron-binding capabilities. When expressed in E. Coli human frataxin binds iron atoms at a rate of 10 iron atoms per 1 molecule of the frataxin polymer. Thus, frataxin appears to function in some capacity for iron-storage for the mitochondria. Frataxin may also function as an activator of oxidative phosphorylation to increase mitochondrial membrane potential and elevate cellular ATP. Frataxin is expressed in tissues with high metabolic activity including heart, liver and brown fat.