

## Frataxin (N-19): sc-25106

### 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.

### REFERENCES

1. Montermini, L., et al. 1995. The Friedreich ataxia critical region spans a 150-kb interval on chromosome 9q13. *Am. J. Hum. Genet.* 57: 1061-1067.
2. Koutnikova, H., et al. 1997. Studies of human, mouse and yeast homologues indicate a mitochondrial function for Frataxin. *Nat. Genet.* 16: 345-351.
3. Campuzano, V., et al. 1997. Frataxin is reduced in Friedreich ataxia patients and is associated with mitochondrial membranes. *Hum. Mol. Genet.* 6: 1771-1780.
4. Ristow, M., et al. 2000. Frataxin activates mitochondrial energy conversion and oxidative phosphorylation. *Proc. Natl. Acad. Sci. USA* 97: 12239-12243.
5. Cavadini, P., et al. 2002. Assembly and iron-binding properties of human Frataxin, the protein deficient in Friedreich ataxia. *Hum. Mol. Genet.* 11: 217-227.

### CHROMOSOMAL LOCATION

Genetic locus: FXN (human) mapping to 9q21.11.

### SOURCE

Frataxin (N-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of Frataxin of human origin.

### PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-25106 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

### STORAGE

Store at 4° C, **\*\*DO NOT FREEZE\*\***. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

### RESEARCH USE

For research use only, not for use in diagnostic procedures.

### APPLICATIONS

Frataxin (N-19) is recommended for detection of Frataxin of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Frataxin siRNA (h): sc-40580, Frataxin shRNA Plasmid (h): sc-40580-SH and Frataxin shRNA (h) Lentiviral Particles: sc-40580-V.

Molecular Weight of Frataxin: 18 kDa.

### RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

### PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.



Try **Frataxin (1D9): sc-293431**, our highly recommended monoclonal alternative to Frataxin (N-19).