

Frataxin (G-12): sc-518079

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 9q21.11. 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. Campuzano, V., et al. 1997. Frataxin is reduced in Friedreich ataxia patients and is associated with mitochondrial membranes. *Hum. Mol. Genet.* 6: 1771-1780.
3. Koutnikova, H., et al. 1997. Studies of human, mouse and yeast homologues indicate a mitochondrial function for Frataxin. *Nat. Genet.* 16: 345-351.
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 (G-12) is a mouse monoclonal antibody raised against amino acids 56-210 mapping at the C-terminus of Frataxin of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Frataxin (G-12) is available conjugated to agarose (sc-518079 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-518079 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-518079 PE), fluorescein (sc-518079 FITC), Alexa Fluor® 488 (sc-518079 AF488), Alexa Fluor® 546 (sc-518079 AF546), Alexa Fluor® 594 (sc-518079 AF594) or Alexa Fluor® 647 (sc-518079 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-518079 AF680) or Alexa Fluor® 790 (sc-518079 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

Frataxin (G-12) is recommended for detection of Frataxin of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], 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.

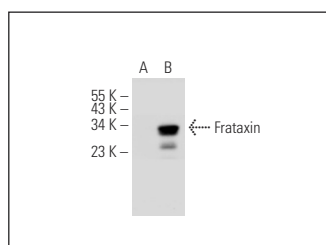
Molecular Weight of Frataxin: 18 kDa.

Positive Controls: human Frataxin transfected HEK293T whole cell lysate.

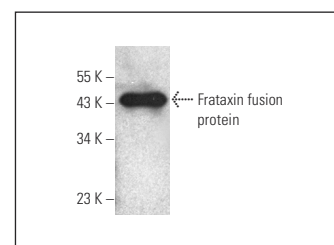
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BPHRP: sc-516102 or m-IgGκ BPHRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



Frataxin (G-12): sc-518079. Western blot analysis of Frataxin expression in non-transfected (A) and human Frataxin transfected (B) HEK293T whole cell lysates.



Frataxin (G-12): sc-518079. Western blot analysis of human recombinant Frataxin fusion protein. Detection reagent used: m-IgGκ BPHRP (Cruz Marker): sc-516102-CM.

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.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.