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SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

linkedin.com/company/szaboscandic

SANTA CRUZ BIOTECHNOLOGY, INC.

Frataxin (C-2): sc-518078

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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

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4. Ristow, M., Pfister, M.F., Yee, A.J., Schubert, M., Michael, L., Zhang, C.Y., Ueki, K., Michael, M.D., 2nd., Lowell, B.B. and Kahn, C.R. 2000. Frataxin activates mitochondrial energy conversion and oxidative phosphorylation. Proc. Natl. Acad. Sci. USA 97: 12239-12243.
5. Cavadini, P., O'Neill, H.A., Benada, O. and Isaya, G. 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 (C-2) 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 IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

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

APPLICATIONS

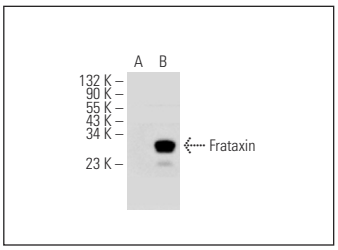
Frataxin (C-2) is recommended for detection of Frataxin of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) 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.

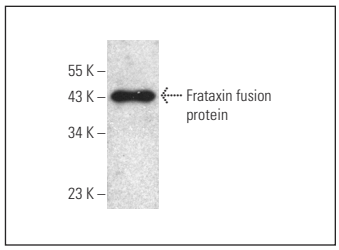
Molecular Weight of Frataxin: 18 kDa.

Positive Controls: human Frataxin transfected HEK293T whole cell lysate.

DATA



Frataxin (C-2): sc-518078. Western blot analysis of Frataxin expression in non-transfected (**A**) and human Frataxin transfected (**B**) HEK293T whole cell lysates.



Frataxin (C-2): sc-518078. Western blot analysis of human recombinant Frataxin fusion protein. Detection reagent used: m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM.

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.