

Frataxin (V-21): sc-130757

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 ten iron atoms per one 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. Gen.* 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.

CHROMOSOMAL LOCATION

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

SOURCE

Frataxin (V-21) is a purified rabbit polyclonal antibody raised against a peptide mapping near the C-terminus of Frataxin of human origin.

PRODUCT

Each vial contains 100 µg IgG in 1.0 ml PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

Frataxin (V-21) is recommended for detection of Frataxin of human origin by Western Blotting (starting dilution 1:200, 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), immunohistochemistry (including paraffin-embedded sections) (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.

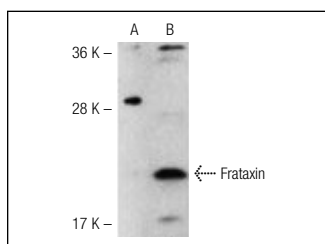
Molecular Weight of Frataxin: 18 kDa.

Positive Controls: Frataxin transfected 293 whole cell lysate.

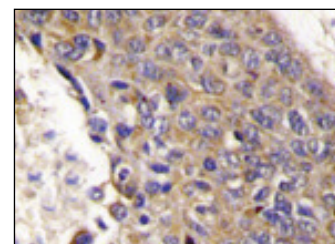
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941. 4) Immunohistochemistry: use ImmunoCruz™: sc-2051 or ABC: sc-2018 rabbit IgG Staining Systems.

DATA



Frataxin (V-21): sc-130757. Western blot analysis of Frataxin expression in non-transfected (A) and human Frataxin transfected (B) 293 whole cell lysates.



Frataxin (V-21): sc-130757. Immunoperoxidase staining of formalin-fixed, paraffin-embedded human lung carcinoma tissue showing cytoplasmic localization.

SELECT PRODUCT CITATIONS

1. Jones, J., et al. 2012. Human adipose stem cell-conditioned medium increases survival of Friedreich's ataxia cells submitted to oxidative stress. *Stem Cells Dev.* 21: 2817-2826.

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 or our catalog for detailed protocols and support products.



Try **Frataxin (1D9): sc-293431**, our highly recommended monoclonal alternative to Frataxin (V-21).