

Frataxin (H-155): sc-25820

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 9q21.11. *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.

CHROMOSOMAL LOCATION

Genetic locus: FXN (human) mapping to 9q21.11; Fxn (mouse) mapping to 19 B.

SOURCE

Frataxin (H-155) is a rabbit polyclonal antibody raised against amino acids 56-210 mapping at the C-terminus 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.

APPLICATIONS

Frataxin (H-155) is recommended for detection of Frataxin of mouse, rat and 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Frataxin (H-155) is also recommended for detection of Frataxin in additional species, including porcine.

Suitable for use as control antibody for Frataxin siRNA (h): sc-40580, Frataxin siRNA (m): sc-40581, Frataxin shRNA Plasmid (h): sc-40580-SH, Frataxin shRNA Plasmid (m): sc-40581-SH, Frataxin shRNA (h) Lentiviral Particles: sc-40580-V and Frataxin shRNA (m) Lentiviral Particles: sc-40581-V.

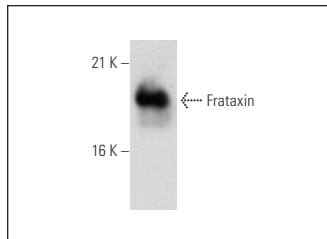
Molecular Weight of Frataxin: 18 kDa.

Positive Controls: rat brain extract: sc-2392.

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.

DATA



Frataxin (H-155): sc-25820. Western blot analysis of Frataxin expression in rat brain tissue extract.

SELECT PRODUCT CITATIONS

1. Grant, L., et al. 2006. Rational selection of small molecules that increase transcription through the GAA repeats found in Friedreich's ataxia. *FEBS Lett.* 580: 5399-5405.
2. Paupe, V., et al. 2009. Impaired nuclear Nrf2 translocation undermines the oxidative stress response in Friedreich ataxia. *PLoS ONE* 4: e4253.
3. Steinkellner, H., et al. 2010. A high throughput electrochemiluminescence assay for the quantification of frataxin protein levels. *Anal. Chim. Acta* 659: 129-132.
4. Kirches, E., et al. 2011. Dual role of the mitochondrial protein frataxin in astrocytic tumors. *Lab. Invest.* 91: 1766-1776.

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.


 MONOS
Satisfaction
Guaranteed

Try **Frataxin (1D9): sc-293431**, our highly recommended monoclonal alternative to Frataxin (H-155).