SANTA CRUZ BIOTECHNOLOGY, INC.

FMR1 (148.1): sc-101048

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Fragile X syndrome is the most frequent form of inherited mental retardation and is the result of transcriptional silencing of the FMR1 gene on the X chromosome. The FMR1 gene contains a distinct CpG dinucleotide repeat located in the 5'-untranslated region of the gene, and in the fragile X syndrome this tandem repeat is substantially amplified, and subjected to extensive methylation and enhanced transcriptional silencing. The FMR1 protein (or FMRP) is an RNA-binding protein that associates with polyribosomes and is a likely component of a messenger ribonuclear protein (mRNP) particle. FMR1 contains several features that are characteristics of RNA-binding proteins, including two hnRNPK homology (KH) domains and an RGG amino acid motif (RGG box). FMR1 can also interact with two fragile X syndrome related factors, FXR1 and FXR2, and these proteins form heterodimers through their N-terminal coiled-coil domains. FMR1 localizes to both the nucleus and the cytoplasm, and since it contains both a nuclear localization signal and a nuclear export signal it is also implicated in the nucleocytoplasmic transport of mRNAs.

REFERENCES

1. Verkerk, A.J., et al. 1991. Identification of a gene (FMR1) containing a CGG repeat coincident with a breakpoint cluster region exhibiting length variation in fragile X syndrome. Cell 65: 905-914.
2. Pieretti, M., et al. 1991. Absence of expression of the FMR1 gene in fragile X syndrome. Cell 66: 817-822.

CHROMOSOMAL LOCATION

Genetic locus: FMR1 (human) mapping to Xq27.3; Fmr1 (mouse) mapping to X A7.1.

SOURCE

FMR1 (148.1) is a mouse monoclonal antibody raised against a partial recombinant FMR1of human origin.

PRODUCT

Each vial contains 100 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

FMR1 (148.1) is recommended for detection of FMR1 of mouse, human and, to a lesser extent, rat 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)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for FMR1 siRNA (h): sc-36870, FMR1 siRNA (m): sc-36871, FMR1 shRNA Plasmid (h): sc-36870-SH, FMR1 shRNA Plasmid (m): sc-36871-SH, FMR1 shRNA (h) Lentiviral Particles: sc-36870-V and FMR1 shRNA (m) Lentiviral Particles: sc-36871-V.

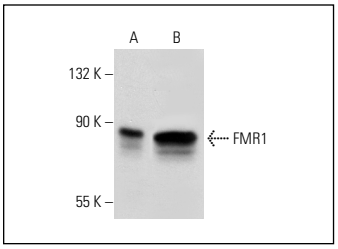
Molecular Weight of FMR1: 85 kDa.

Positive Controls: FMR1 (h): 293T Lysate: sc-115468, T98G cell lysate: sc-2294 or Jurkat whole cell lysate: sc-2204.

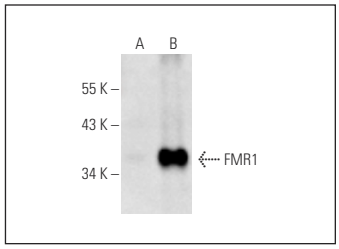
STORAGE

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

DATA



FMR1 (148.1): sc-101048. Western blot analysis of FMR1 expression in 293T (**A**) and Jurkat (**B**) whole cell lysates.



FMR1 (148.1): sc-101048. Western blot analysis of FMR1 expression in non-transfected: sc-117752 (**A**) and human FMR1 transfected: sc-115468 (**B**) 293T whole cell lysates. Detection reagent used: m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM.

SELECT PRODUCT CITATIONS

1. Simpson-Holley, M., et al. 2011. Formation of antiviral cytoplasmic granules during orthopoxvirus infection. J. Virol. 85: 1581-1593.
2. Fujimura, K., et al. 2012. Selenite targets eIF4E-binding protein-1 to inhibit translation initiation and induce the assembly of non-canonical stress granules. Nucleic Acids Res. 40: 8099-8110.
3. Panas, M.D., et al. 2015. Methods for the characterization of stress granules in virus infected cells. Methods 90: 57-64.
4. Brykczynska, U., et al. 2016. CGG repeat-induced FMR1 silencing depends on the expansion size in human iPSCs and neurons carrying unmethylated full mutations. Stem Cell Reports 7: 1059-1071.
5. Zhu, T., et al. 2017. Cancer and cancer therapy and their association with ventricular arrhythmia. Can. J. Cardiol. 33: 293.e11.
6. Sanders, D.W., et al. 2020. Competing protein-RNA interaction networks control multiphase intracellular organization. Cell 181: 306-324.e28.
7. Fay, M.M., et al. 2021. Bisphenol A promotes stress granule assembly and modulates the integrated stress response. Biol. Open 10: bio057539.
8. Cascella, R., et al. 2022. A quantitative biology approach correlates neuronal toxicity with the largest inclusions of TDP-43. Sci. Adv. 8: eabm6376.
9. Sirois, C.L., et al. 2024. CGG repeats in the human FMR1 gene regulate mRNA localization and cellular stress in developing neurons. Cell Rep. 43: 114330.

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.