FMR1 (6A15): sc-57005



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-coiled 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 nucleo-cytoplasmic transport of mRNAs.

REFERENCES

- Pieretti, M., Zhang, F.P., Fu, Y.H., Warren, S.T., Oostra, B.A., Caskey, C.T. and Nelson, D.L. 1991. Absence of expression of the FMR-1 gene in fragile X syndrome. Cell 66: 817-22.
- Verkerk, A.J., Pieretti, M., Sutcliffe, J.S., Fu, Y.H., Kuhl, D.P., Pizzuti, A., Reiner, O., Richards, S., Victoria, M.F., Zhang, F.P., et al. 1991. Identification of a gene (FMR-1) containing a CGG repeat coincident with a breakpoint cluster region exhibiting length variation in fragile X syndrome. Cell 65: 905-914.
- Matunis, M.J., Michael, W.M. and Dreyfuss, G. 1992. Characterization and primary structure of the poly(C)-binding heterogeneous nuclear ribonucleoprotein complex K protein. Mol. Cell. Biol. 12: 164-171.
- 4. De Boulle, K., Verkerk, A.J., Reyniers, E., Vits, L., Hendrickx, J., Van Roy, B., Van den Bos, F., de Graaff, E.,Oostra, B.A. and Willems, P.J. 1993. A point mutation in the FMR-1 gene associated with fragile X mental retardation. Nat. Genet. 3: 31-35.
- Zhang, Y., O'Connor, J.P., Siomi, M.C., Srinivasan, S., Dutra, A., Nussbaum, R.L. and Dreyfuss, G. 1995. The fragile X mental retardation syndrome protein interacts with novel homologs FXR1 and FXR2. EMBO J. 14: 5358-5366.
- Eberhart, D.E., Malter, H.E., Feng, Y. and Warren, S.T. 1996. The fragile X mental retardation protein is a ribonucleoprotein containing both nuclear localization and nuclear export signals. Hum. Mol. Genet. 5: 1083-1091.
- Ceman, S., et al. 1999. Isolation of an FMRP-associated messenger ribonucleoprotein particle and identification of nucleolin and the fragile X-related proteins as components of the complex. Mol. Cell. Biol. 19: 7925-7932.
- 8. Tamanini, F., Van Unen, L., Bakker, C., Sacchi, N., Galjaard, H., Oostra, B.A. and Hoogeveen, A.T. 1999. Oligomerization properties of fragile X mental-retardation protein (FMRP) and the fragile X-related proteins FXR1P and FXR2P. Biochem. J. 343: 517-523.

SOURCE

FMR1 (6A15) is a mouse monoclonal antibody raised against amino-terminal 580 amino acids of His-FMR1 fusion protein of *Drosophila melanogaster* origin.

PRODUCT

Each vial contains 200 μg lgG_1 in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

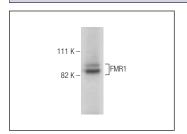
APPLICATIONS

FMR1 (6A15) is recommended for detection of FMR1 of *Drosophila melanogaster* origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)] and immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

Molecular Weight of FMR1: 85 kDa.

Positive Controls: Drosophila whole cell lysate.

DATA



FMR1 (6A15): sc-57005. Western blot analysis of FMR1 expression in *Drosophila* whole cell lysate.

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.

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