

FMR1 (4G9): sc-293156

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. In 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. It 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 localizes to both the nucleus and the cytoplasm and can also interact with two fragile X syndrome related factors, FXR1 and FXR2, which form heterodimers through their N-terminal coiled-coil domains. Since FMR1 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: 817-22.
3. Matunis, M.J., et al. 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., et al. 1993. A point mutation in the FMR1 gene associated with fragile X mental retardation. *Nat. Genet.* 3: 31-35.
5. Zhang, Y., et al. 1995. The fragile X mental retardation syndrome protein interacts with novel homologs FXR1 and FXR2. *EMBO J.* 14: 5358-5366.
6. Eberhart, D.E., et al. 1996. The fragile X mental retardation protein is a ribonucleoprotein containing both nuclear localization and nuclear export signals. *Hum. Mol. Genet.* 5: 1083-1091.
7. 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., et al. 1999. Oligomerization properties of fragile X mental-retardation protein (FMRP) and the fragile X-related proteins FXR1P and FXR2P. *Biochem. J.* 343: 517-523.

CHROMOSOMAL LOCATION

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

SOURCE

FMR1 (4G9) is a mouse monoclonal antibody raised against recombinant protein fragment corresponding to FMR1 of human origin.

PRODUCT

Each vial contains 50 μ l ascites containing IgG₁ with < 0.1% sodium azide.

APPLICATIONS

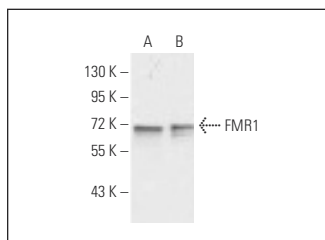
FMR1 (4G9) is recommended for detection of FMR1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), 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 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: Jurkat whole cell lysate: sc-2204 or K-562 whole cell lysate: sc-2203.

DATA



FMR1 (4G9): sc-293156. Western blot analysis of FMR1 expression in Jurkat (A) and K-562 (B) whole cell lysates.

STORAGE

For immediate and continuous use, store at 4° C for up to one month. For sporadic use, freeze in working aliquots in order to avoid repeated freeze/thaw cycles. If turbidity is evident upon prolonged storage, clarify solution by centrifugation.

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.