

FMR1 (D-6): sc-518105

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: 817-822.
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.

CHROMOSOMAL LOCATION

Genetic locus: FMR1 (human) mapping to Xq27.3.

SOURCE

FMR1 (D-6) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 497-516 near the C-terminus of FMR1 of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

FMR1 (D-6) is available conjugated to agarose (sc-518105 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-518105 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-518105 PE), fluorescein (sc-518105 FITC), Alexa Fluor® 488 (sc-518105 AF488), Alexa Fluor® 546 (sc-518105 AF546), Alexa Fluor® 594 (sc-518105 AF594) or Alexa Fluor® 647 (sc-518105 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-518105 AF680) or Alexa Fluor® 790 (sc-518105 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

FMR1 (D-6) is recommended for detection of FMR1 of human origin by Western Blotting (starting dilution 1:100, 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).

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

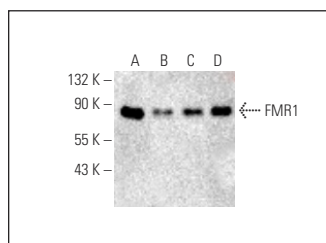
Molecular Weight of FMR1: 85 kDa.

Positive Controls: MCF7 whole cell lysate: sc-2206, T98G cell lysate: sc-2294 or K-562 whole cell lysate: sc-2203.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



FMR1 (D-6): sc-518105. Western blot analysis of FMR1 expression in MCF7 (A), T98G (B), K-562 (C) and SK-OV-3 (D) whole cell lysates. Detection reagent used: m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM.

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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