# FMR1 (H-120): sc-28739



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.

#### CHROMOSOMAL LOCATION

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

#### **SOURCE**

FMR1 (H-120) is a rabbit polyclonal antibody raised against amino acids 513-632 mapping at the C-terminus of FMR1 of human origin.

## **PRODUCT**

Each vial contains 200  $\mu g$  lgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

# **APPLICATIONS**

FMR1 (H-120) is recommended for detection of all FMR1 splice variants 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), 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).

FMR1 (H-120) is also recommended for detection of all FMR1 splice variants in additional species, including canine, bovine and porcine.

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: T98G cell lysate: sc-2294 or Jurkat whole cell lysate: sc-2204.

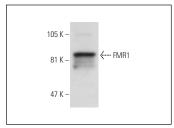
## **PROTOCOLS**

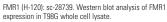
See our web site at www.scbt.com or our catalog for detailed protocols and support products.

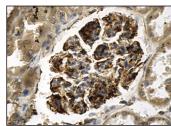
#### **STORAGE**

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

#### **DATA**







FMR1 (H-120): sc-28739. Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic staining of cells in glomerulus and tubules.

#### **SELECT PRODUCT CITATIONS**

- Dobson, T., et al. 2008. Identifying intrinsic and extrinsic determinants that regulate internal initiation of translation mediated by the FMR1 5' leader. BMC Mol. Biol. 9: 89.
- Dictenberg, J.B., et al. 2008. A direct role for FMRP in activity-dependent dendritic mRNA transport links filopodial-spine morphogenesis to fragile X syndrome. Dev. Cell 14: 926-939.
- 3. lacoangeli, A., et al. 2008. Reply to bagni: on BC1 RNA and the fragile X mental retardation protein. Proc. Natl. Acad. Sci. USA 105: E29.
- 4. lacoangeli, A., et al. 2008. On BC1 RNA and the fragile X mental retardation protein. Proc. Natl. Acad. Sci. USA 105: 734-739.
- 5. Fähling, M., et al. 2009. Translational regulation of the human achaete-scute homologue-1 by fragile X mental retardation protein. J. Biol. Chem. 284: 4255-4266.
- 6. Perlewitz, A., et al. 2010. Aldosterone and vasopressin affect  $\alpha$  and  $\gamma$ -ENaC mRNA translation. Nucleic Acids Res. 38: 5746-5760.
- 7. Dziembowska, M., et al. 2012. Activity-dependent local translation of matrix metalloproteinase-9. J. Neurosci. 32: 14538-14547.
- Janusz, A., et al. 2013. The fragile X mental retardation protein regulates matrix metalloproteinase 9 mRNA at synapses. J. Neurosci. 33: 18234-18241.

## **RESEARCH USE**

For research use only, not for use in diagnostic procedures.



Try FMR1 (148.1): sc-101048 or FMR1 (4G9): sc-293156, our highly recommended monoclonal aternatives to FMR1 (H-120).