

# FXR1/2 (C-5): sc-166433

## 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.
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.

## CHROMOSOMAL LOCATION

Genetic locus: FXR2 (human) mapping to 3q26.33, FXR1 (human) mapping to 17p13.1; Fxr1 (mouse) mapping to 3 A3, Fxr2 (mouse) mapping to 11 B3.

## SOURCE

FXR1/2 (C-5) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 526-557 within an internal region of FXR1 of human origin.

## PRODUCT

Each vial contains 200 µg IgG<sub>3</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-166433 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

## APPLICATIONS

FXR1/2 (C-5) is recommended for detection of FXR1 and FXR2 of mouse, rat and 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).

FXR1/2 (C-5) is also recommended for detection of FXR1 and FXR2 in additional species, including equine, canine, bovine and porcine.

Molecular Weight of FXR1: 78 kDa.

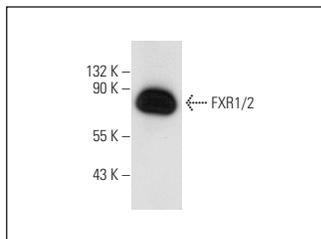
Molecular Weight of FXR2: 90 kDa.

Positive Controls: F9 cell lysate: sc-2245, Jurkat whole cell lysate: sc-2204 or WI-38 whole cell lysate: sc-364260.

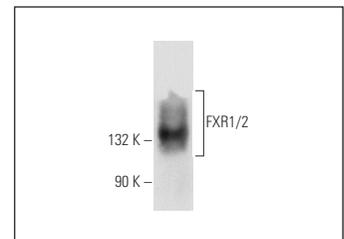
## RECOMMENDED SUPPORT REAGENTS

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



FXR1/2 (C-5): sc-166433. Western blot analysis of FXR1/2 expression in F9 whole cell lysate.



FXR1/2 (C-5): sc-166433. Western blot analysis of FXR1/2 expression in WI-38 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.



See **FXR1 (B-2): sc-374148** for FXR1 antibody conjugates, including AC, HRP, FITC, PE, and Alexa Fluor® 488, 546, 594, 647, 680 and 790.