FXR1 (h): 293T Lysate: sc-114030



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. 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

- 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 (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., 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 FMR1 gene in fragile X syndrome. Cell 66: 817-822.
- 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.
- 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 FMR1 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.
- 6. 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.
- 7. Ceman, S., Brown, V. and Warren, S.T. 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.

CHROMOSOMAL LOCATION

Genetic locus: FXR1 (human) mapping to 3q26.33.

PRODUCT

FXR1 (h): 293T Lysate represents a lysate of human FXR1 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

FXR1 (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive FXR1 antibodies. Recommended use: 10-20 μ l per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-tranfected 293T cells.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. 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.

Santa Cruz Biotechnology, Inc. 1.800.457.3801 831.457.3800 fax 831.457.3801 Europe +00800 4573 8000 49 6221 4503 0 www.scbt.com