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FMR1 Antibody

Catalog # ASC11766

Product Information

Application	WB, IF, E, IHC-P
Primary Accession	Q06787
Other Accession	NP_002015, 4503765
Reactivity	Human, Mouse, Rat
Host	Rabbit
Clonality	Polyclonal
Isotype	IgG
Calculated MW	71174
Concentration (mg/ml)	1 mg/mL
Conjugate	Unconjugated
Application Notes	FMR1 antibody can be used for detection of FMR1 by Western blot at 1 - 2 g/ml. Antibody can also be used for Immunohistochemistry starting at 5 g/mL. For immunofluorescence start at 20 g/mL.

Additional Information

Gene ID	2332
Other Names	Fragile X mental retardation protein 1, FMRP, Protein FMR-1, FMR1
Target/Specificity	FMR1; FMR1 antibody is human, mouse and rat reactive. Multiple isoforms of FMR1 are known to exist.
Reconstitution & Storage	FMR1 antibody can be stored at 4℃ for three months and -20℃, stable for up to one year.
Precautions	FMR1 Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	FMR1 {ECO:0000303|PubMed:8504300, ECO:0000312|HGNC:HGNC:3775}
Function	Multifunctional polyribosome-associated RNA-binding protein that plays a central role in neuronal development and synaptic plasticity through the regulation of alternative mRNA splicing, mRNA stability, mRNA dendritic transport and postsynaptic local protein synthesis of target mRNAs (PubMed:12417522, PubMed:16631377, PubMed:18653529, PubMed:19166269, PubMed:23235829, PubMed:25464849). Acts as an mRNA regulator by mediating formation of some phase- separated membraneless compartment: undergoes liquid-liquid phase separation upon binding to target mRNAs, leading to assemble mRNAs into cytoplasmic ribonucleoprotein granules that concentrate mRNAs with associated regulatory factors (PubMed:12417522, PubMed:30765518,

PubMed:31439799). Plays a role in the alternative splicing of its own mRNA (PubMed:18653529). Stabilizes the scaffolding postsynaptic density protein DLG4/PSD-95 and the myelin basic protein (MBP) mRNAs in hippocampal neurons and glial cells, respectively; this stabilization is further increased in response to metabotropic glutamate receptor (mGluR) stimulation (By similarity). Plays a role in selective delivery of a subset of dendritic mRNAs to synaptic sites in response to mGluR activation in a kinesin-dependent manner (By similarity). Undergoes liquid-liquid phase separation following phosphorylation and interaction with CAPRIN1, promoting formation of cytoplasmic ribonucleoprotein granules that concentrate mRNAs with factors that inhibit translation and mediate deadenylation of target mRNAs (PubMed:31439799). Acts as a repressor of mRNA translation in synaptic regions by mediating formation of neuronal ribonucleoprotein granules and promoting recruitmtent of EIF4EBP2 (PubMed:30765518). Plays a role as a repressor of mRNA translation during the transport of dendritic mRNAs to postsynaptic dendritic spines (PubMed:11157796, PubMed:11532944, PubMed:12594214, PubMed:23235829). Component of the CYFIP1-EIF4E-FMR1 complex which blocks cap-dependent mRNA translation initiation (By similarity). Represses mRNA translation by stalling ribosomal translocation during elongation (By similarity). Reports are contradictory with regards to its ability to mediate translation inhibition of MBP mRNA in oligodendrocytes (PubMed:23891804). Also involved in the recruitment of the RNA helicase MOV10 to a subset of mRNAs and hence regulates microRNA (miRNA)-mediated translational repression by AGO2 (PubMed:14703574, PubMed:17057366, PubMed:25464849). Facilitates the assembly of miRNAs on specific target mRNAs (PubMed:17057366). Also plays a role as an activator of mRNA translation of a subset of dendritic mRNAs at synapses (PubMed:19097999, PubMed:19166269). In response to mGluR stimulation, FMR1-target mRNAs are rapidly derepressed, allowing for local translation at synapses (By similarity). Binds to a large subset of dendritic mRNAs that encode a myriad of proteins involved in pre- and postsynaptic functions (PubMed:11157796, PubMed:11719189, PubMed:12594214, PubMed:17417632, PubMed:23235829, PubMed:24448548, PubMed:7692601). Binds to 5'-ACU[GU]-3' and/or 5'-[AU]GGA-3' RNA consensus sequences within mRNA targets, mainly at coding sequence (CDS) and 3'-untranslated region (UTR) and less frequently at 5'-UTR (PubMed:23235829). Binds to intramolecular G-quadruplex structures in the 5'- or 3'-UTRs of mRNA targets (PubMed:11719189, PubMed:18579868, PubMed:25464849, PubMed:25692235). Binds to G-quadruplex structures in the 3'-UTR of its own mRNA (PubMed:11532944, PubMed:12594214, PubMed:15282548, PubMed:18653529, PubMed:7692601). Also binds to RNA ligands harboring a kissing complex (kc) structure; this binding may mediate the association of FMR1 with polyribosomes (PubMed:15805463). Binds mRNAs containing U-rich target sequences (PubMed:12927206). Binds to a triple stem-loop RNA structure, called Sod1 stem loop interacting with FMRP (SoSLIP), in the 5'-UTR region of superoxide dismutase SOD1 mRNA (PubMed:19166269). Binds to the dendritic, small non-coding brain cytoplasmic RNA 1 (BC1); which may increase the association of the CYFIP1-EIF4E-FMR1 complex to FMR1 target mRNAs at synapses (By similarity). Plays a role in mRNA nuclear export (PubMed:31753916). Specifically recognizes and binds a subset of N6-methyladenosine (m6A)-containing mRNAs, promoting their nuclear export in a XPO1/CRM1-dependent manner (PubMed:31753916). Together with export factor NXF2, is involved in the regulation of the NXF1 mRNA stability in neurons (By similarity). Associates with export factor NXF1 mRNA-containing ribonucleoprotein particles (mRNPs) in a NXF2-dependent manner (By similarity). Binds to a subset of miRNAs in the brain (PubMed:14703574, PubMed:17057366). May associate with nascent transcripts in a nuclear protein NXF1-dependent manner (PubMed:18936162). In vitro, binds to RNA homomer; preferentially on poly(G) and to a lesser extent on poly(U), but not

on poly(A) or poly(C) (PubMed:12950170, PubMed:15381419, PubMed:7688265, PubMed:7781595, PubMed:8156595). Moreover, plays a role in the modulation of the sodium-activated potassium channel KCNT1 gating activity (PubMed:20512134). Negatively regulates the voltage-dependent calcium channel current density in soma and presynaptic terminals of dorsal root ganglion (DRG) neurons, and hence regulates synaptic vesicle exocytosis (By similarity). Modulates the voltage- dependent calcium channel CACNA1B expression at the plasma membrane by targeting the channels for proteasomal degradation (By similarity). Plays a role in regulation of MAP1B-dependent microtubule dynamics during neuronal development (By similarity). Has been shown to play a translation-independent role in the modulation of presynaptic action potential (AP) duration and neurotransmitter release via large- conductance calcium-activated potassium (BK) channels in hippocampal and cortical excitatory neurons (PubMed:25561520). May be involved in the control of DNA damage response (DDR) mechanisms through the regulation of ATR-dependent signaling pathways such as histone H2AX/H2A.x and BRCA1 phosphorylations (PubMed:24813610). Forms a cytoplasmic messenger ribonucleoprotein (mRNP) network by packaging long mRNAs, serving as a scaffold that recruits proteins and signaling molecules. This network facilitates signaling reactions by maintaining proximity between kinases and substrates (PubMed:39106863).

Cellular Location

Cytoplasm, Cytoplasmic ribonucleoprotein granule. Cytoplasm, Stress granule. Cytoplasm. Perikaryon. Cytoplasm, perinuclear region. Cell projection, neuron projection. Cell projection, axon {ECO:0000250|UniProtKB:P35922}. Cell projection, dendrite {ECO:0000250|UniProtKB:P35922}. Cell projection, dendritic spine {ECO:0000250|UniProtKB:P35922}. Synapse, synaptosome {ECO:0000250|UniProtKB:P35922}. Cell projection, growth cone. Cell projection, filopodium tip {ECO:0000250|UniProtKB:P35922}. Synapse {ECO:0000250|UniProtKB:P35922} Postsynaptic cell membrane {ECO:0000250|UniProtKB:P35922}. Presynaptic cell membrane {ECO:0000250|UniProtKB:P35922}. Nucleus. Nucleus, nucleolus. Chromosome, centromere {ECO:0000250|UniProtKB:P35922}. Chromosome {ECO:0000250|UniProtKB:P35922}. Cell membrane {ECO:0000250|UniProtKB:P35922}. Note=Mediates formation and localizes to cytoplasmic ribonucleoprotein membraneless compartments (PubMed:30765518, PubMed:31439799). Localizes to cytoplasmic ribonucleoprotein granules, also referred to as messenger ribonucleoprotein particles or mRNPs, along dendrites and dendritic spines (PubMed:14532325, PubMed:9659908). FMR1-containing cytoplasmic granules colocalize to F-actin-rich structures, including filopodium, spines and growth cone during the development of hippocampal neurons (By similarity). FMR1-containing cytoplasmic granules are transported out of the soma along axon and dendrite to synaptic contacts in a microtubule- and kinesin-dependent manner (PubMed:12417734, PubMed:15380484). Colocalizes with FXR1 and FXR2 in discrete granules, called fragile X granules (FXGs), along axon and presynaptic compartments (By similarity). Colocalizes with TDRD3 in cytoplasmic stress granules (SGs) in response to various cellular stress (PubMed:16636078, PubMed:18632687, PubMed:18664458). Colocalizes with FXR1, kinesin, 60S acidic ribosomal protein RPLP0 and SMN in cytoplasmic granules in the soma and neurite cell processes (PubMed:12417734, PubMed:16636078, PubMed:18093976). Colocalizes with H2AX/H2A.x in pericentromeric heterochromatin in response to DNA damaging agents (By similarity). Localizes on meiotic pachytene-stage chromosomes (By similarity). Forms nuclear foci representing sites of ongoing DNA replication in response to DNA damaging agents (By similarity). Shuttles between nucleus and cytoplasm in a XPO1/CRM1- dependent manner (PubMed:10196376). Colocalizes with CACNA1B in the cytoplasm and at the cell membrane of

	neurons (By similarity) Colocalizes with CYFIP1, CYFIP2, NXF2 and ribosomes in the perinuclear region (By similarity). Colocalizes with CYFIP1 and EIF4E in dendrites and probably at synapses (By similarity) {ECO:0000250|UniProtKB:P35922, ECO:0000250|UniProtKB:Q80WE1, ECO:0000269|PubMed:10196376, ECO:0000269|PubMed:12417734, ECO:0000269|PubMed:14532325, ECO:0000269|PubMed:15380484, ECO:0000269|PubMed:16636078, ECO:0000269|PubMed:18093976, ECO:0000269|PubMed:18632687, ECO:0000269|PubMed:18664458, ECO:0000269|PubMed:30765518, ECO:0000269|PubMed:31439799, ECO:0000269|PubMed:9659908} [Isoform 9]: Cytoplasm [Isoform 11]: Nucleus. Nucleus, Cajal body
Tissue Location	Expressed in the brain, cerebellum and testis (PubMed:8401578, PubMed:9259278). Also expressed in epithelial tissues (PubMed:8401578). Expressed in mature oligodendrocytes (OLGs) (PubMed:23891804). Expressed in fibroblast (PubMed:24204304). Expressed in neurons, Purkinje cells and spermatogonias (at protein level) (PubMed:8401578, PubMed:9259278). Expressed in brain, testis and placenta (PubMed:8504300, PubMed:9259278). Expressed in neurons and lymphocytes (PubMed:8504300).

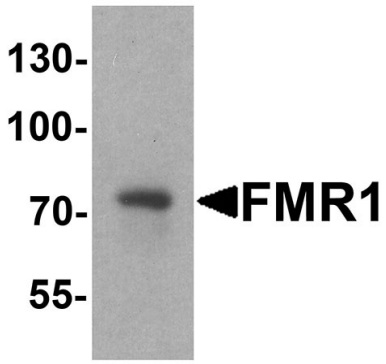
Background

Fragile X syndrome is a frequent form of inherited mental retardation caused by functional loss of the fragile X mental retardation protein, FMR1, also known as FMRP (1). FMR1 binds RNA and is associated with polysomes. The encoded protein may be involved in mRNA trafficking from the nucleus to the cytoplasm (2). A trinucleotide repeat (CGG) in the 5' UTR is normally found at 6-53 copies, but an expansion to 55-230 repeats is the cause of fragile X syndrome (1). Expansion of the trinucleotide repeat may also cause one form of premature ovarian failure (POF1) (3).

References

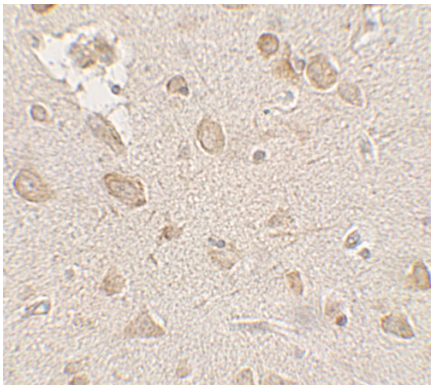
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Corbin F, Bouillon M, Fortin A, et al. The fragile X mental retardation protein is associated with poly(A)+ mRNA in actively translating polyribosomes. Hum. Mol. Genet. 1997; 1465-72.
Lu C, Lin L, Tan H, et al. Fragile X premutation RNA is sufficient to cause primary ovarian insufficiency in mice. Hum. Mol. Genet. 2012; 5039-47.

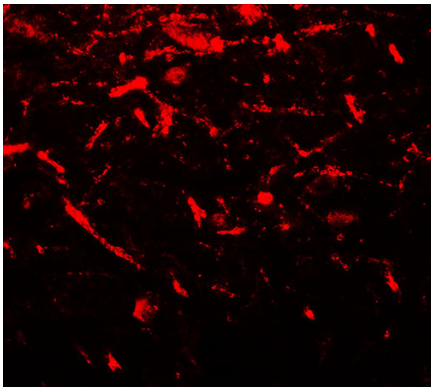
Images



Western blot analysis of FMR1 in rat brain tissue lysate with FMR1 antibody at 1 µg/ml.

Immunohistochemistry of FMR1 in human brain tissue with FMR1 antibody at 2.5 µg/mL.





Immunofluorescence of FMR1 in human brain tissue with FMR1 antibody at 20 µg/mL.

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