

FMR1 Antibody

Catalog # ASC11766

Specification

FMR1 Antibody - Product Information

WB, IHC, IF Application Primary Accession Q06787 Other Accession NP 002015, 4503765

Reactivity Human, Mouse,

Rat Rabbit

Host Clonality **Polyclonal** Isotype laG

Calculated MW Predicted: 70 kDa

Observed: 74 kDa

KDa

Application Notes FMR1 antibody

can be used for detection of FMR1 **bv Western blot** at $1 - 2 \mu g/ml$. Antibody can also be used for Immu nohistochemistry starting at 5 μg/mL. For immun ofluorescence start at 20 µg/mL.

FMR1 Antibody - Additional Information

Gene ID 2332

Target/Specificity

FMR1; FMR1 antibody is human, mouse and rat reactive. Multiple isoforms of FMR1 are known to exist.

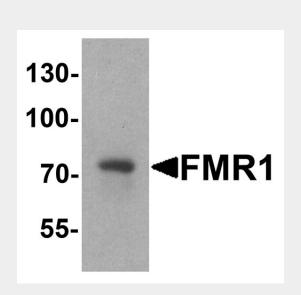
Reconstitution & Storage

FMR1 antibody can be stored at 4°C for three months and -20°C, stable for up to one year.

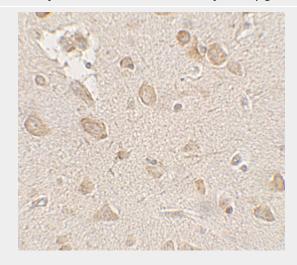
Precautions

FMR1 Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

FMR1 Antibody - Protein Information



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.



Name FMR1 (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 a subset of mRNAs (PubMed:<a href="http://www.uniprot.org/citations/16631377"</code>

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PubMed: <a href="http://www.uniprot.org/ci tations/18653529"

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target="_blank">25464849). Plays a role in the alternative splicing of its own mRNA (PubMed:<a href="http://www.uniprot.org/citations/18653529"

target=" blank">18653529). Plays a role in mRNA nuclear export (By similarity). Together with export factor NXF2, is involved in the regulation of the NXF1 mRNA stability in neurons (By similarity). 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). Plays a role as a repressor of mRNA translation during the transport of dendritic mRNAs to postsynaptic dendritic spines (PubMed:<a href="http://www.uniprot.org/c

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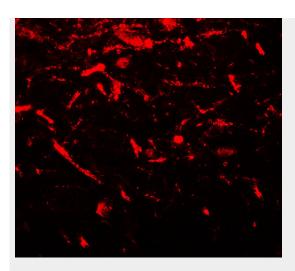
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Immunofluorescence of FMR1 in human brain tissue with FMR1 antibody at 20 µg/mL.

FMR1 Antibody - 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).

FMR1 Antibody - References

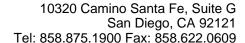
Jin P and Warren ST. Understanding the molecular basis of fragile X syndrome. Hum. Mol. Genet. 2000; 9:901-8.

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.



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). Plays also 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:7692601, PubMed:11719189, PubMed: tations/11157796" target=" blank">11157796, PubMed:12594214,





PubMed:<a href="http://www.uniprot.org/ci tations/17417632"

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PubMed:<a href="http://www.uniprot.org/ci tations/24448548"

target="_blank">24448548). 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" target="_blank">23235829). Binds to intramolecular G-quadruplex structures in the 5'- or 3'-UTRs of mRNA targets

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PubMed: <a href="http://www.uniprot.org/ci tations/18579868"

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PubMed:<a href="http://www.uniprot.org/ci tations/25692235"

target="_blank">25692235). Binds to G-quadruplex structures in the 3'-UTR of its own mRNA (PubMed:<a href="http://www.u niprot.org/citations/7692601"

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PubMed:<a href="http://www.uniprot.org/ci tations/11532944"

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PubMed:<a href="http://www.uniprot.org/ci tations/12594214"

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PubMed:<a href="http://www.uniprot.org/ci tations/15282548"

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PubMed: <a href="http://www.uniprot.org/ci tations/18653529"

target="_blank">18653529). Binds also to RNA ligands harboring a kissing complex (kc) structure; this binding may mediate the association of FMR1 with polyribosomes (PubMed:15805463" target="_blank">15805463). Binds mRNAs containing U-rich target sequences (PubMed:<a href="http://www.uniprot.org/citations/12927206"

target="_blank">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). 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:<a hr ef="http://www.uniprot.org/citations/76882 65" target=" blank">7688265, PubMed:7781595, PubMed:12950170, PubMed:15381419, PubMed:8156595). Moreover, plays a role in the modulation of the sodium-activated potassium channel KCNT1 gating activity (PubMed:20512134). Negatively regulates the voltagedependent 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 voltagedependent calcium channel CACNA1B expression at the plasma membrane by targeting the channels for proteosomal degradation (By similarity). Plays a role in



regulation of MAP1B-dependent microtubule dynamics during neuronal development (By similarity). Recently, 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). Finally, FMR1 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).

Cellular Location

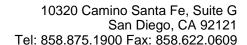
Nucleus, Nucleus, nucleolus. Chromosome, centromere {ECO:0000250|UniProtKB:P35922}. Chromosome {ECO:0000250|UniProtKB:P35922}. Cytoplasm. Cytoplasm, perinuclear region. Cytoplasm, Cytoplasmic ribonucleoprotein granule. Perikaryon. Cell projection, neuron projection. Cell projection, axon {ECO:0000250|UniProtKB:P35922}. Cell projection, dendrite {ECO:0000250|UniProtKB:P35922}. Cell projection, dendritic spine {ECO:0000250|UniProtKB:P35922}. Cell junction, synapse, synaptosome {ECO:0000250|UniProtKB:P35922}. Cell projection, growth cone. Cell projection, filopodium tip {ECO:0000250|UniProtKB:P35922}. Cell junction, synapse {ECO:0000250|UniProtKB:P35922}. Cell junction, synapse, postsynaptic cell membrane {ECO:0000250|UniProtKB:P35922}. Cell junction, synapse, presynaptic cell membrane {ECO:0000250|UniProtKB:P35922}. Cell membrane {ECO:0000250|UniProtKB:P35922}. Cytoplasm, Stress granule. Note=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). Localizes to cytoplasmic ribonucleoprotein granules, also referred to as messenger ribonucleoprotein particles or mRNPs, along dendrites and dendritic spines (PubMed:9659908, PubMed:14532325) 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 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). Colocalizes with FXR1, kinesin, 60S acidic ribosomal protein RPLP0 and SMN in cytoplasmic granules in the soma and neurite cell processes (PubMed:12417734, PubMed:18093976, PubMed:16636078). 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:18632687, PubMed:18664458, PubMed:16636078) {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:9659908} [Isoform 9]: Cytoplasm [Isoform 11]: Nucleus. Nucleus, Cajal body

Tissue Location

Expressed in the brain, cerebellum and testis (PubMed:8401578). 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). Expressed in brain, testis and placenta (PubMed:8504300). Expressed in neurons and lymphocytes (PubMed:8504300).

FMR1 Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- <u>Immunofluorescence</u>
- <u>Immunoprecipitation</u>
- Flow Cytomety
- Cell Culture