FMR1 (PT1226R) PT™ Rabbit mAb

CatalogNo: YM9068 Recombinant

Key Features

Host Species
- Rabbit

Reactivity
- Human,Mouse,Rat

Applications
- WB,IHC,IF,ELISA

MW
- 71kD (Calculated)

70-77kD (Observed)

Isotype
- IgG,Kappa

Storage

Storage*	-15°C to -25°C/1 year(Do not lower than -25°C)
Formulation	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%BSA

Recommended Dilution Ratios

IHC 1:200-1:1000
WB 1:2000-1:10000
IF 1:200-1:1000
ELISA 1:5000-1:20000

Basic Information

Clonality	Monoclonal
Clone Number	PT1226R

Immunogen Information

Specificity	Endogenous

Target Information

Gene name FMR1

Protein Name Fragile X mental retardation 1 protein

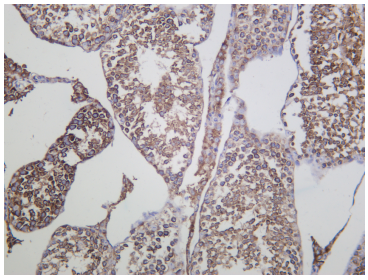
Organism	Gene ID	UniProt ID
Human	2332;	Q06787;
Mouse		P35922;

Cellular Localization Nucleus . Nucleus, nucleolus . Chromosome, centromere . Chromosome . Cytoplasm . Cytoplasm, perinuclear region . Cytoplasm, Cytoplasmic ribonucleoprotein granule . Perikaryon . Cell projection, neuron projection . Cell projection, axon . Cell projection, dendrite . Cell projection, dendritic spine . Cell junction, synapse, synaptosome . Cell projection, growth cone . Cell projection, filopodium tip . Cell junction, synapse . Cell junction, synapse, postsynaptic cell membrane . Cell junction, synapse, presynaptic cell membrane . Cell membrane . Cytoplasm, Stress granule . 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). .; [Isoform 6]: Cytoplasm . Cytoplasm, perinuclear region .; [Isoform 9]: Cytoplasm .; [Isoform 10]: Nucleus . Nucleus, Cajal body . Colocalizes with Colin and SMN in Cajal bodies (PubMed:24204304).; [Isoform 11]: Nucleus . Nucleus, Cajal body .

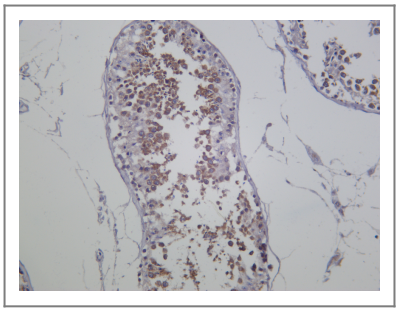
Tissue specificity Expressed in the brain, cerebellum and testis (PubMed:8401578). Also expressed in epithelial tissues (PubMed:8401578). Expressed in mature oligodendrocytes (OLGs) (PubMed:23891804). Expressed in fibroblast (PubMed:24204304). Expressed in neurons, Purkinje cells and spermatogonias (at protein level) (PubMed:8401578). Expressed in brain, testis and placenta (PubMed:8504300). Expressed in neurons and lymphocytes (PubMed:8504300).

Function	Alternative products:At least 12 different isoforms are produced,Disease:Defects in FMR1 are the cause of fragile X syndrome. [MIM:300624]. It is a common genetic disease (has a prevalence of one in every 2000 children) which is characterized by moderate to severe mental retardation, macroorchidism (enlargement of the testicles), large ears, prominent jaw, and high-pitched, jocular speech. The defect in most fragile X syndrome patients results from an amplification of a CGG repeat region which is directly in front of the coding region.,Disease:Defects in FMR1 are the cause of fragile X tremor/ataxia syndrome (FXTAS) [MIM:300623]. In FXTAS, the expanded repeats range in size from 55 to 200 repeats and are referred to as 'premutations'. Full repeat expansions with greater than 200 repeats results in fragile X mental retardation syndrome [MIM:300624]. Carriers of the premutation typically do not show the full fragile X syndrome phenotype, but comprise a subgroup that may have some physical features of fragile X syndrome or mild cognitive and emotional problems.,Function:RNA-binding protein that plays a role in intracellular RNA transport and in the regulation of translation of target mRNAs. Associated with polysomes. May play a role in the transport of mRNA from the nucleus to the cytoplasm. Binds strongly to poly(G), binds moderately to poly(U) but shows very little binding to poly(A) or poly(C).,miscellaneous:RNA-binding activity is inhibited by RANBP9.,miscellaneous:The mechanism of the severe phenotype in the Asn-304 patient lies in the sequestration of bound mRNAs in nontranslatable mRNP particles. In the absence of FMRP, these same mRNAs may be partially translated via alternate mRNPs, although perhaps abnormally localized or regulated, resulting in typical fragile X syndrome. Asn-304 mutation maps to a position within the second KH domain of FMRP that is critical for stabilizing sequence-specific RNA-protein interactions. Asn-304 mutation abrogates the association of the FMRP KH 2 domain with its target, kissing complex RNA.,PTM:Phosphorylated on several serine residues.,similarity:Belongs to the FMR1 family.,similarity:Contains 1 KH domain.,similarity:Contains 2 KH domains.,subunit:Homooligomer. Found in a RNP granule complex with IGF2BP1. Directly interacts with SMN and TDRD3. Interacts with the SMN core complex that contains SMN1, SIP1/GEMIN2, DDX20/GEMIN3, GEMIN4, GEMIN5, GEMIN6, GEMIN7, GEMIN8 and STRAP/UNRIP. Interacts with FXR1, FXR2, IGF2BP1, NUFIP1, NUFIP2, MCRS1 and RANBP9. Interacts with CYFIP1 and CYFIP2.,tissue specificity:Highest levels found in neurons, brain, testis, placenta and lymphocytes. Also expressed in epithelial tissues and at very low levels in glial cells.,

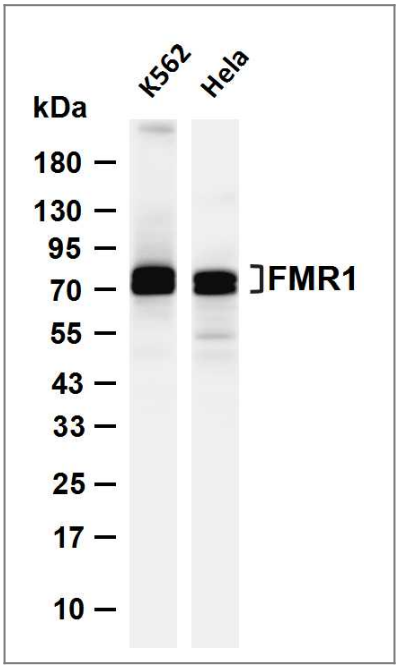
Validation Data



Mouse testis was stained with anti-FMR1 (PT1226R) Rabbit antibody



Human testis was stained with anti-FMR1 (PT1226R) Rabbit antibody



Various whole cell lysates were separated by 4-20% SDS-PAGE, and the membrane was blotted with anti-FMR1 (PT1226R) antibody. The HRP-conjugated Goat anti-Rabbit IgG (H + L) antibody was used to detect the antibody. Lane 1: K562 Lane 2: Hela Predicted band size: 71kDa Observed band size: 70-77kDa

Contact information

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Please scan the QR code to access additional product information:
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