

Rabbit Anti-phospho-FMRP (Ser500) antibody

SL13188R

Product Name:	phospho-FMRP (Ser500)
Chinese Name:	磷酸化脆性X综合征相关蛋白AFF1抗体
Alias:	FMRP (phospho S500); FMRP (phospho Ser500); p-FMRP (phospho S500); p-FMR1 (phospho S500); FMR1; FMR1_HUMAN; FMRP; FMRP phospho S499; Fragile X mental retardation 1; Fragile X mental retardation 1 protein; Fragile X mental retardation protein 1; FRAXA; MGC87458; POF; POF1; Protein FMR-1; Protein FMR1.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Rabbit,Zebrafish,Sheep,Guinea Pig,Danio rerio
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100- 500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	75kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthesised phosphopeptide derived from human FMRP around the phosphorylation site of Ser500:NA(p-S)ET
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	?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.

Function:

Translation repressor. Component of the CYFIP1-EIF4E-FMR1 complex which binds to the mRNA cap and mediates translational repression. In the CYFIP1-EIF4E-FMR1 complex this subunit mediates translation repression (By similarity). 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).

Subunit:

Component of the CYFIP1-EIF4E-FMR1 complex which is composed of CYFIP, EIF4E and FMR1. Interacts with CYFIP1 and CYFIP2. The interaction with brain cytoplasmic RNA 1 (BC1) increases binding affinity for the CYFIP1-EIF4E complex in the brain (By similarity). Homooligomer. Found in a RNP granule complex with IGF2BP1. Directly interacts with SMN and TDRD3. Interacts with the SMN core complex that contains SMN1, GEMIN2/SIP1, DDX20/GEMIN3, GEMIN4, GEMIN5, GEMIN6, GEMIN7, GEMIN8 and STRAP/UNRIP. Interacts with FXR1, FXR2, IGF2BP1, NUFIP1, NUFIP2, MCRS1 and RANBP9.

Subcellular Location:

Cytoplasm. Nucleus, nucleolus.

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.

Post-translational modifications:

Phosphorylated on several serine residues.

DISEASE:

Defects in FMR1 are the cause of fragile X syndrome (FRAX) [MIM:300624]. Fragile X syndrome 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. 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. Defects in FMR1 are the cause of premature ovarian failure syndrome type 1 (POF1) [MIM:311360]. An ovarian disorder defined as the cessation of ovarian function under the age of 40 years. It is characterized by oligomenorrhea or amenorrhea, in the presence of elevated levels of serum gonadotropins and low estradiol. Similarity: Belongs to the FMR1 family. Contains 2 Agenet-like domains. Contains 2 KH domains. SWISS: O06787 Gene ID: 2332 Database links: UniProtKB/Swiss-Prot: Q06787.1 Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.





Tissue/cell: rat brain tissue; 4% Paraformaldehyde-fixed and paraffinembedded;

Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking

buffer (normal goat serum,C-0005) at 37°C for 20 min;

Incubation: Anti-phospho-FMRP (Ser500) Polyclonal Antibody,

Unconjugated(SL13188R) 1:200, overnight at 4°C, followed by conjugation to

the secondary antibody(SP-0023) and DAB(C-0010) staining





Paraformaldehyde-fixed, paraffin embedded (rat brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (phospho-FMRP (Ser500)) Polyclonal Antibody, Unconjugated (SL13188R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.