



MECP2 Rabbit mAb

Catalog No	YP-rAb-17594
Isotype	IgG
Reactivity	Human,Mouse,Rat
Applications	WB,IHC,IF,IP,ELISA
Gene Name	MECP2
Protein Name	Methyl-CpG-binding protein 2
Purification Process	Protein A
Specificity	Endogenous
Formulation	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%BSA
Source	Monoclonal, Rabbit,IgG
Dilution	IHC 1:200-1:1000; WB 1:2000-1:10000; IF 1:200-1:1000; ELISA 1:5000-1:20000; IP 1:50-1:200; Note: For IHC, we suggest antigen retrieval with TE buffer pH 9.0
Concentration	0.5 mg/ml
Purity	≥90%
Storage Stability	-15° C to -25° C/1 year(Do not lower than -25° C)
Synonyms	Methyl-CpG-binding protein 2 ; MeCp-2 protein ; MeCp2 ;
Observed Band	75kD
Calculated Molecular Weight	52kD
Cell Pathway	Nucleus
Tissue Specificity	Present in all adult somatic tissues tested.
Function	<p>Disease:A chromosomal duplication involving MECP2 is the cause of mental retardation syndromic X-linked Lubs type (MRXSL) [MIM:300260]. Increased dosage of MECP2 appears to be responsible for the mental retardation phenotype. The main features present in affected males are severe to profound mental retardation with onset at birth, axial and facial hypotonia, progressive spasticity predominantly at the lower limbs, seizures and recurrent infections.</p> <p>Disease:Defects in MECP2 are the cause of mental retardation syndromic X-linked type 13 (MRXS13) [MIM:300055]. Mental retardation is a mental disorder characterized by significantly sub-average general intellectual functioning associated with impairments in adaptative behavior and manifested during the developmental period. MRXS13 patients manifest mental retardation associated with other variable features such as spasticity, episodes of manic depressive psychosis, increased tone and macroorchidism.</p> <p>Disease:Defects in</p>





MECP2 are the cause of neonatal severe encephalopathy due to MECP2 mutations [MIM:300673]. The MECP2 gene is mutated in Rett syndrome, a severe neurodevelopmental disorder that almost always occurs in females. Although it was first thought that MECP2 mutations causing Rett syndrome were lethal in males, later reports identified a severe neonatal encephalopathy in surviving male sibs of patients with Rett syndrome. Additional reports have confirmed a severe phenotype in males with Rett syndrome-associated MECP2 mutations. Disease: Defects in MECP2 are the cause of Rett syndrome (RTT) [MIM:312750]. RTT is an X-linked dominant disease, it is a progressive neurologic developmental disorder and one of the most common causes of mental retardation in females. Patients appear to develop normally until 6 to 18 months of age, then gradually lose speech and purposeful hand movements and develop microcephaly, seizures, autism, ataxia, intermittent hyperventilation, and stereotypic hand movements. After initial regression, the condition stabilizes and patients usually survive into adulthood. Disease: Defects in MECP2 may be a cause of Angelman syndrome (AS) [MIM:105830]; also known as happy puppet syndrome. AS is a neurodevelopmental disorder characterized by severe mental retardation, absent speech, ataxia, sociable affect and dysmorphic facial features. AS and Rett syndrome have overlapping clinical features. Disease: Defects in MECP2 may be the cause of susceptibility to X-linked autism 3 (AUTSX3) [MIM:300496]. AUTSX3 is a pervasive developmental disorder (PDD), prototypically characterized by impairments in reciprocal social interaction and communication, restricted and stereotyped patterns of interests and activities, and the presence of developmental abnormalities by 3 years of age. Function: Chromosomal protein that binds to methylated DNA. It can bind specifically to a single methyl-CpG pair. It is not influenced by sequences flanking the methyl-CpGs. Mediates transcriptional repression through interaction with histone deacetylase and the corepressor SIN3A. online information: IRSA MECP2 variation database, online information: Rett syndrome website, PTM: Phosphorylated on Ser-423 in brain upon synaptic activity, which attenuates its repressor activity and seems to regulate dendritic growth and spine maturation. similarity: Contains 1 MBD (methyl-CpG-binding) domain. similarity: Contains 2 A.T hook DNA-binding domains. subcellular location: Colocalized with methyl-CpG in the genome. subunit: Interacts with FNBP3. tissue specificity: Present in all adult somatic tissues tested.

Background

DNA methylation is the major modification of eukaryotic genomes and plays an essential role in mammalian development. Human proteins MECP2, MBD1, MBD2, MBD3, and MBD4 comprise a family of nuclear proteins related by the presence in each of a methyl-CpG binding domain (MBD). Each of these proteins, with the exception of MBD3, is capable of binding specifically to methylated DNA. MECP2, MBD1 and MBD2 can also repress transcription from methylated gene promoters. In contrast to other MBD family members, MECP2 is X-linked and subject to X inactivation. MECP2 is dispensible in stem cells, but is essential for embryonic development. MECP2 gene mutations are the cause of most cases of Rett syndrome, a progressive neurologic developmental disorder and one of the most common causes of mental retardation in females. Alternative splicing results in multiple transcript variants encoding different isoforms.

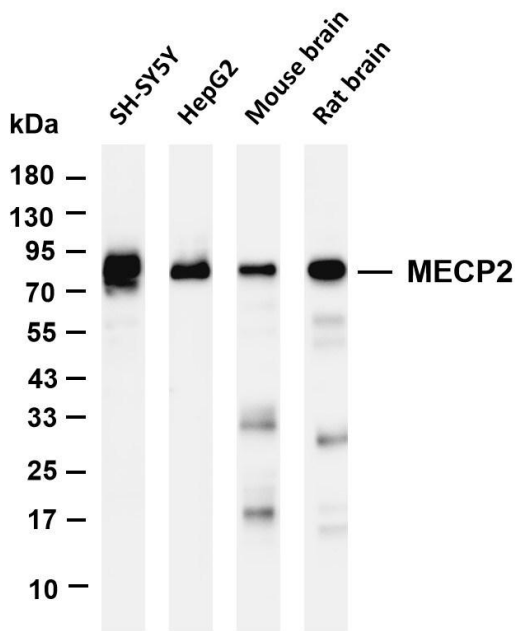
matters needing attention

Avoid repeated freezing and thawing!

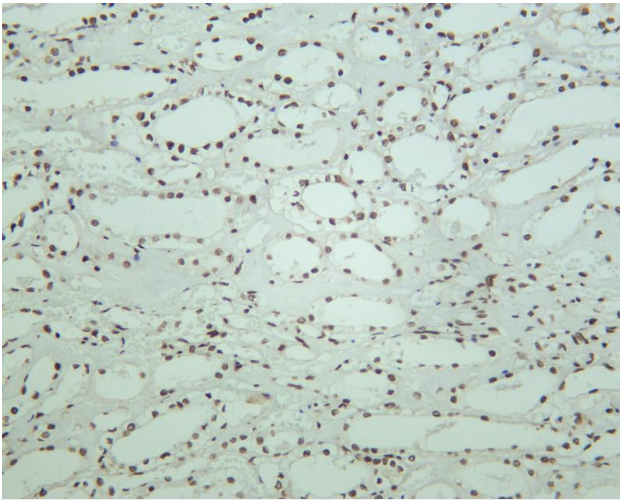
Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

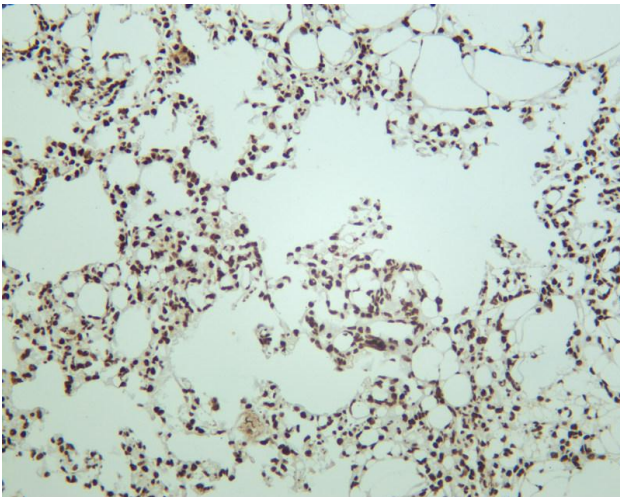




Various whole cell lysates were separated by 4-20% SDS-PAGE, and the membrane was blotted with anti-MECP2 antibody. The HRP-conjugated Goat anti-Rabbit IgG(H + L) antibody was used to detect the antibody. Lane 1: SH-SY5Y Lane 2: HepG2 Lane 3: Mouse brain Lane 4: Rat brain Predicted band size: 52kDa Observed band size: 75kDa



Human kidney was stained with anti-MECP2 rabbit antibody



Mouse lung was stained with anti-MECP2 rabbit antibody

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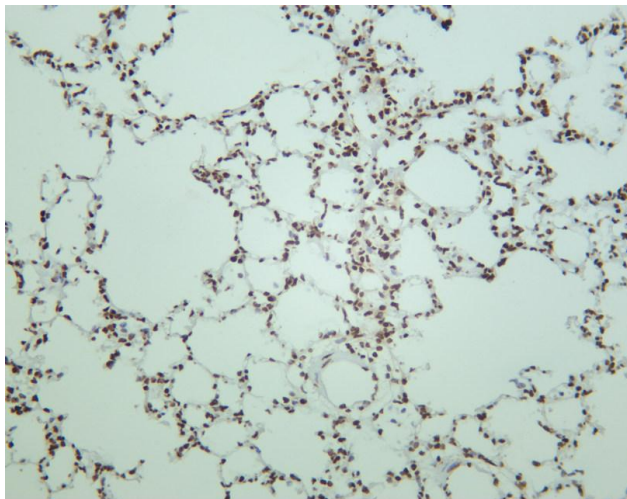
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Rat lung was stained with anti-MECP2 rabbit antibody

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