

Product datasheet

Recombinant Human MeCP2 protein ab114182

1 图像

概述

产品名称	重组人MeCP2蛋白
蛋白长度	Full length protein

描述

性质	Recombinant
来源	Wheat germ

氨基酸序列

Accession [P51608](#)

种属 Human

序列
 MVAGMLGLREEKSEDQLQGLKDKPLKFKKVKKDKKKEEKGKHEPVQPSA
 HHSAPAEAGKAETSESGSAPAVPEASASPKQRRSIIRDRGPMYDDPTL
 PEGWTRKLRKSGRSAGKYDVYLINPQGKAFRSKVELIAYFEKVGDTSL
 DPNDFDFTVTGRGSPSRREQPPKKPKSPKAPGTGRGRGRPKGSGTTRPK
 AATSEGVQVQRVLEKSPGKLLVKMPFQTSPGGKAEGGGATTSTQVMVIKR
 PGRKRKAEADPQAIPKKRGRKPGSVVAAAAAEAKKAVKESSIRSVQETV
 LPIKKRKTRETVSIEVKEVVKPLLVSTLGEKSGKGLKTKSPGRKSKESS
 PKGRSSASSPPKKEHHHHHHHSESPKAPVPLLPLPPPPPEPESSEDPT
 SPPEPQDLSSVCKEEKMPRGGSLSDGCPKEPAKTQPAVATAATAAEKY
 KHRGEGERKDIVSSMPRPNREEPVDSRTPVTERVS

分子量	80 kDa including tags
氨基酸	1 to 486
标签	GST tag N-Terminus

技术指标

Our [Abpromise guarantee](#) covers the use of **ab114182** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

应用	SDS-PAGE
	Western blot
	ELISA

形式	Liquid
补充说明	Protein concentration is above or equal to 0.05 mg/ml. Best used within three months from the date of receipt.
制备和贮存	
稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl
常规信息	
功能	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.
组织特异性	Present in all adult somatic tissues tested.
疾病相关	<p>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.</p> <p>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 adaptive 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>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.</p> <p>Defects in MECP2 may be the cause of susceptibility autism X-linked type 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.</p> <p>Defects in MECP2 are the cause of encephalopathy neonatal severe due to MECP2 mutations (ENS-MECP2) [MIM:300673]. Note=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.</p> <p>Defects in MECP2 are the cause of mental retardation syndromic X-linked Lubs type (MRXSL) [MIM:300260]. Mental retardation is characterized by significantly below average general intellectual functioning associated with impairments in adaptive behavior and manifested during the developmental period. MRXSL patients manifest mental retardation associated with</p>

variable features. They include swallowing dysfunction and gastroesophageal reflux with secondary recurrent respiratory infections, hypotonia, mild myopathy and characteristic facies such as downslanting palpebral fissures, hypertelorism and a short nose with a low nasal bridge. Note=Increased dosage of MECP2 due to gene duplication appears to be responsible for the mental retardation phenotype.

序列相似性

Contains 2 A.T hook DNA-binding domains.
Contains 1 MBD (methyl-CpG-binding) domain.

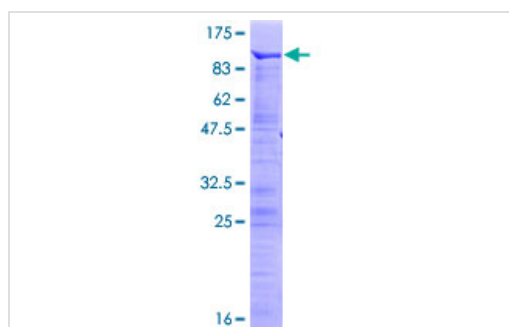
翻译后修饰

Phosphorylated on Ser-423 in brain upon synaptic activity, which attenuates its repressor activity and seems to regulate dendritic growth and spine maturation.

细胞定位

Nucleus. Colocalized with methyl-CpG in the genome.

图片



ab114182 on a 12.5% SDS-PAGE Stained with Coomassie Blue.

SDS-PAGE - MeCP2 protein (ab114182)

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