

AAAS Antibody (monoclonal) (M02)

Mouse monoclonal antibody raised against a partial recombinant AAAS.

Catalog # AT1001a

Product Information

Application	WB, IHC, E
Primary Accession	Q9NRG9
Other Accession	NM_015665
Reactivity	Human, Mouse, Rat
Host	mouse
Clonality	monoclonal
Isotype	IgG2a Kappa
Clone Names	5A1
Calculated MW	59574

Additional Information

Gene ID	8086
Other Names	Aladin, Adracalin, AAAS, ADRACALA
Target/Specificity	AAAS (NP_056480, 1 a.a. ~ 100 a.a) partial recombinant protein with GST tag. MW of the GST tag alone is 26 KDa.
Dilution	WB~~1:500~1000 IHC~~1:100~500 E~~N/A
Format	Clear, colorless solution in phosphate buffered saline, pH 7.2 .
Storage	Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.
Precautions	AAAS Antibody (monoclonal) (M02) is for research use only and not for use in diagnostic or therapeutic procedures.

Background

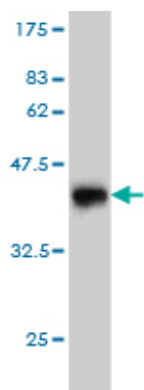
The protein encoded by this gene is a member of the WD-repeat family of regulatory proteins and may be involved in normal development of the peripheral and central nervous system. The encoded protein is part of the nuclear pore complex and is anchored there by NDC1. Defects in this gene are a cause of achalasia-addisonianism-alacrima syndrome (AAAS), also called triple-A syndrome or Allgrove syndrome. Two transcript variants encoding different isoforms have been found for this gene.

References

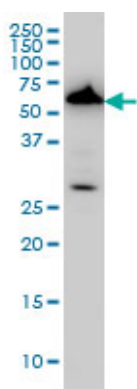
Two Italian patients with novel AAAS gene mutation expand allelic and phenotypic spectrum of triple A (Allgrove) syndrome. Palka C, et al. Clin Genet, 2010 Mar. PMID 20447142. Deficiency of ferritin heavy-chain

nuclear import in triple a syndrome implies nuclear oxidative damage as the primary disease mechanism. Storr HL, et al. Mol Endocrinol, 2009 Dec. PMID 19855093. The nuclear pore complex protein ALADIN is anchored via NDC1 but not via POM121 and GP210 in the nuclear envelope. Kind B, et al. Biochem Biophys Res Commun, 2009 Dec 11. PMID 19782045. The transmembrane nucleoporin NDC1 is required for targeting of ALADIN to nuclear pore complexes. Yamazumi Y, et al. Biochem Biophys Res Commun, 2009 Nov 6. PMID 19703420. Tissue-specific expression and subcellular localization of ALADIN, the absence of which causes human triple A syndrome. Cho AR, et al. Exp Mol Med, 2009 Jun 30. PMID 19322026.

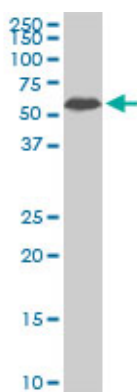
Images



Antibody Reactive Against Recombinant Protein. Western Blot detection against Immunogen (36.74 KDa) .

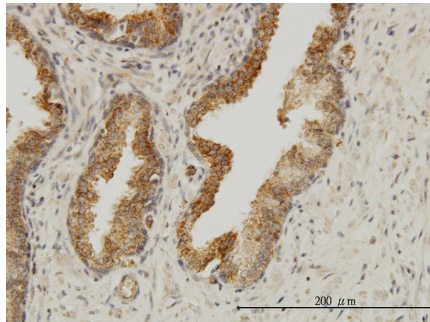
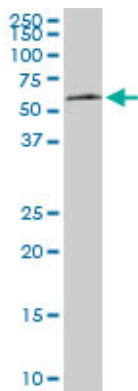


AAAS monoclonal antibody (M02), clone 5A1 Western Blot analysis of AAAS expression in HeLa ((Cat # AT1001a)

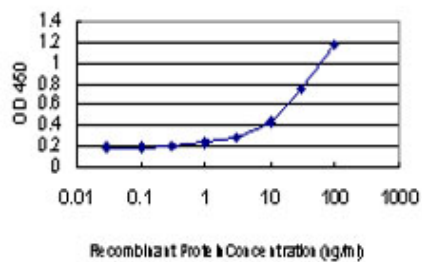


AAAS monoclonal antibody (M02), clone 5A1. Western Blot analysis of AAAS expression in PC-12((Cat # AT1001a)

AAAS monoclonal antibody (M02), clone 5A1. Western Blot analysis of AAAS expression in NIH/3T3((Cat # AT1001a)



Immunoperoxidase of monoclonal antibody to AAAS on formalin-fixed paraffin-embedded human prostate. [antibody concentration 3 ug/ml]



Detection limit for recombinant GST tagged AAAS is approximately 1ng/ml as a capture antibody.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.