

# Rabbit Anti-Adracalin antibody

# SL11807R

Product Name:	Adracalin
Chinese Name:	Allgrove综合征相关蛋白抗体
Alias:	AAA; AAAS; AAASb; Achalasia adrenocortical insufficiency alacrimia (Allgrove triple A); Achalasia adrenocortical insufficiency alacrimia; ADRACALA; Aladin; Allgrove triple A; DKFZp586G1624; GL003; AAAS_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat, Cow,
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:	60kDa
Cellular localization:	The nucleus
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Adracalin:51-150/546
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:	<u>PubMed</u>
Product Detail:	Aladin (Adracalin) belongs to a family of WD repeat-containing proteins. These proteins have a wide variety of functions, including signal transduction regulation, RNA processing and transcription. Aladin plays a role in peripheral and central nervous system development. It is widely expressed, with the highest expression seen in pituitary gland, corpus callosum, cerebellum, adrenal gland and gastrointestinal

structures. Defects in Aladin cause the autosomal recessive disorder achalasia-addisonianism-alacrima (triple A) syndrome. Triple A syndrome is characterized by achalasia, alacrima and adrenocortico-tropin-resistant adrenal insufficiency. Robust expression in neural systems associated with cognitive, motor and sensory functions is consistent with the myriad of symptoms experienced by patients with triple A syndrome.

#### **Function:**

Adracalin (AAAS) is expressed in both neuroendocrine and cerebral structures and may function in the normal development of the peripheral and central nervous system. It localizes to nuclear pore complexes (NPCs), large multiprotein assemblies that are the sole sites of nucleocytoplasmic transport. Defects in AAAS are the cause of achalasia-addisonianism-alacrima syndrome (AAA syndrome); also known as triple-A syndrome or Allgrove syndrome.

#### **Subcellular Location:**

nuclear pore

### Tissue Specificity:

Widely expressed. Particularly abundant expression is found in cerebellum, corpus callosum, adrenal gland, pituitary gland, gastrointestinal structures and fetal lung.

#### **DISEASE:**

Defects in AAAS are the cause of achalasia-addisonianism-alacrima syndrome (AAAS) [MIM:231550]; also known as triple-A syndrome or Allgrove syndrome. AAAS is an autosomal recessive disorder characterized by adreno-corticotropic hormone (ACTH)-resistant adrenal failure, achalasia of the esophageal cardia and alacrima. The syndrome is associated with variable and progressive neurological impairment involving the central, peripheral, and autonomic nervous system. Other features such as palmoplantar hyperkeratosis, short stature, facial dysmorphy and osteoporosis may also be present.

#### Similarity:

Contains 4 WD repeats.

#### SWISS:

O9NRG9

#### Gene ID:

8086

#### Database links:

Entrez Gene: 8086Human

Entrez Gene: 223921Mouse

Entrez Gene: 300259Rat

Omim: 605378Human

SwissProt: Q9NRG9Human

SwissProt: P58742Mouse

Unigene: 369144Human

Unigene: 352946 Mouse

Unigene: 104730Rat

## **Important Note:**

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.