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Rabbit Anti-Adenylosuccinate Lyase antibody

SL6352R

Product Name:	Adenylosuccinate Lyase
Chinese Name:	腺苷酸琥珀酸裂解酶抗体
Alias:	Adenylosuccinase; Adenylosuccinate lyase; ADSL; AMPS; ASase; ASL; PUR8_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=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:	55kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Adenylosuccinate Lyase:185-280/484
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:	Adenylsuccinate lyase is involved in both de novo synthesis of purines and formation of adenosine monophosphate from inosine monophosphate. It catalyzes two reactions in AMP biosynthesis: the removal of a fumarate from succinylaminoimidazole carboxamide (SAICA) ribotide to give aminoimidazole carboxamide ribotide (AICA) and removal of fumarate from adenylosuccinate to give AMP. Adenylosuccinase

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deficiency results in succinylpurinemic autism, psychomotor retardation, and , in some cases, growth retardation associated with muscle wasting and epilepsy. Two transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Jul 2008].

Tissue Specificity:
Ubiquitously expressed. Both isoforms are produced by all tissues. Isoform 2 is 10-fold less abundant than isoform 1.

DISEASE:
Defects in ADSL are the cause of adenylosuccinase deficiency (ADSL deficiency) . ADSL deficiency is an autosomal recessive disorder characterized by the accumulation in the body fluids of succinylaminoimidazole-carboxamide riboside (SAICA-riboside) and succinyladenosine (S-Ado). Most children display marked psychomotor delay, often accompanied by epilepsy or autistic features, or both, although some patients may be less profoundly retarded. Occasionally, growth retardation and muscular wasting are also present.

Similarity:
Belongs to the lyase 1 family. Adenylosuccinate lyase subfamily.

SWISS:
P30566

Gene ID:
158

Database links:

Entrez Gene: 158Human

Entrez Gene: 11564Mouse

Entrez Gene: 315150Rat

Omim: 608222Human

SwissProt: P30566Human

SwissProt: P54822Mouse

Unigene: 75527Human

Unigene: 38151Mouse

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

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