

Adenylosuccinate Lyase Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP58422

Product Information

Application WB, IHC-P, IHC-F, IF, E

Primary Accession <u>P30566</u>

Reactivity Rat, Pig, Dog, Bovine

Host Rabbit
Clonality Polyclonal
Calculated MW 54889
Physical State Liquid

Immunogen KLH conjugated synthetic peptide derived from human Adenylosuccinate

Lyase

Epitope Specificity 185-280/484

Isotype IgG

Purity affinity purified by Protein A

Buffer 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.

SIMILARITY Belongs to the lyase 1 family. Adenylosuccinate lyase subfamily.

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.

Important Note This product as supplied is intended for research use only, not for use in

human, therapeutic or diagnostic applications.

Background Descriptions 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 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].

Additional Information

Gene ID 158

Other Names Adenylosuccinate lyase, ADSL, ASL, 4.3.2.2, Adenylosuccinase, ASase, ADSL,

AMPS

Target/Specificity Ubiquitously expressed. Both isoforms are produced by all tissues. Isoform 2

is 10-fold less abundant than isoform 1.

Dilution WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000

-10000

Format 0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce

Storage Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name ADSL

Synonyms AMPS

Function Catalyzes two non-sequential steps in de novo AMP synthesis: converts

(S)-2-(5-amino-1-(5-phospho-D-ribosyl)imidazole-4- carboxamido)succinate

(SAICAR) to fumarate plus 5-amino-1-(5-phospho-D-

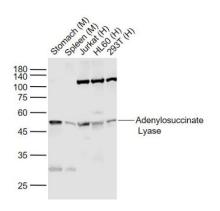
ribosyl)imidazole-4-carboxamide, and thereby also contributes to de novo IMP synthesis, and converts succinyladenosine monophosphate (SAMP) to

AMP and fumarate.

Tissue Location Ubiquitously expressed. Both isoforms are produced by all tissues. Isoform 2

is 10-fold less abundant than isoform 1

Images



Sample:

Lane 1: Stomach (Mouse) Lysate at 40 ug Lane 2: Spleen (Mouse) Lysate at 40 ug Lane 3: Jurkat (Human) Cell Lysate at 30 ug Lane 4: HL60 (Human) Cell Lysate at 30 ug Lane 5: 293T (Human) Cell Lysate at 30 ug

Primary: Anti-Adenylosuccinate Lyase (AP58422) at

1/1000 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at

1/20000 dilution

Predicted band size: 55/48 kD Observed band size: 50 kD

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