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Rabbit Anti-Argininosuccinate Lyase antibody

SL12515R

Product Name:	Argininosuccinate Lyase
Chinese Name:	琥珀酸裂解酶抗体
Alias:	Argininosuccinase; Argininosuccinate lyase; Arginosuccinase; ARLY_HUMAN; ASAL; ASL; EC 4.3.2.1.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Horse,
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:	52kDa
Cellular localization:	cytoplasmicExtracellular matrix
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ASL/Argininosuccinate Lyase:301-400/464
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:	This gene encodes a member of the lyase 1 family. The encoded protein forms a cytosolic homotetramer and primarily catalyzes the reversible hydrolytic cleavage of argininosuccinate into arginine and fumarate, an essential step in the liver in detoxifying ammonia via the urea cycle. Mutations in this gene result in the autosomal recessive disorder argininosuccinic aciduria, or argininosuccinic acid lyase deficiency.

A nontranscribed pseudogene is also located on the long arm of chromosome 22. Alternatively spliced transcript variants encoding different isoforms have been described. [provided by RefSeq, Jul 2008]

Function:
Amino-acid biosynthesis; L-arginine biosynthesis; L-arginine from L-ornithine and carbamoyl phosphate: step 3/3.
Nitrogen metabolism; urea cycle; L-arginine and fumarate from (N(omega)-L-arginino)succinate: step 1/1.

Subcellular Location:
Acetylation modifies enzyme activity in response to alterations of extracellular nutrient availability. Acetylation increased with trichostin A (TSA) or with nicotinamide (NAM). Glucose increases acetylation by about a factor of 3 with decreasing enzyme activity. Acetylation on Lys-288 is decreased on the addition of extra amino acids resulting in activation of enzyme activity.

DISEASE:
Defects in ASL are the cause of arginosuccinicaciduria (ARGINSA) [MIM:207900]. Arginosuccinicaciduria is an autosomal recessive disorder of the urea cycle. The disease is characterized by mental and physical retardation, liver enlargement, skin lesions, dry and brittle hair showing trichorrhexis nodosa microscopically and fluorescing red, convulsions, and episodic unconsciousness.

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

SWISS:
P04424

Gene ID:
435

Database links:

Entrez Gene: 435Human

Entrez Gene: 109900Mouse

Entrez Gene: 59085Rat

Entrez Gene: 512771Cow

Omim: 608310Human

SwissProt: Q3SZJ0Cow

SwissProt: P04424Human

	SwissProt: Q91YI0Mouse SwissProt: P20673Rat Unigene: 632015Human Unigene: 23869Mouse Unigene: 64591Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.