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Argininosuccinate Lyase Rabbit pAb

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Catalog # AP54872

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P04424
Predicted	Rat, Pig, Dog, Human, Mouse, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	51658
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ASL
Epitope Specificity	301-400/464
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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]

Additional Information

Gene ID	435
Other Names	Argininosuccinate lyase, ASAL, 4.3.2.1, Arginosuccinase, ASL

Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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	ASL
Function	Catalyzes the reversible cleavage of L-argininosuccinate to fumarate and L-arginine, an intermediate step reaction in the urea cycle mostly providing for hepatic nitrogen detoxification into excretable urea as well as de novo L-arginine synthesis in nonhepatic tissues (PubMed:11747432, PubMed:11747433, PubMed:22081021, PubMed:2263616, PubMed:9045711). Essential regulator of intracellular and extracellular L-arginine pools. As part of citrulline-nitric oxide cycle, forms tissue-specific multiprotein complexes with argininosuccinate synthase ASS1, transport protein SLC7A1 and nitric oxide synthase NOS1, NOS2 or NOS3, allowing for cell-autonomous L-arginine synthesis while channeling extracellular L-arginine to nitric oxide synthesis pathway (PubMed:22081021).

Background

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