SunLong Biotech Co.,LTD
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[Primary Antibody]

Rabbit Anti-AASS antibody

Catalog Number: SL11391R

FOR RESEARCH USE ONLY

[DATASHEET]

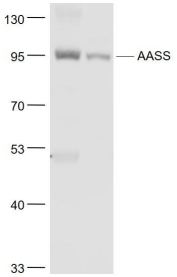
Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	AASS
Immunogen:	KLH conjugated synthetic peptide derived from human LKRSDH: 878-926/926
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	WB=1:500-2000 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Mouse, (predicted: Human, Rat, Dog, Pig, Cow, Horse, Rabbit, Sheep)
Predicted MW.:	99kDa
Subcellular Location:	cytoplasmic
Background:	Alpha-aminoadipic semialdehyde synthase (AASS), also designated lysine ketoglutarate reductase (LKR) or saccharopine dehydrogenase (SDH), is a 926 amino acid protein that exists as a homodimer in the mitochondria. AASS acts as a bifunctional enzyme containing the lysine alpha-ketoglutarate reductase (LKR) and saccharopine dehydrogenase activities that catalyzes the first two steps in lysine degradation. It is widely expressed with highest expression in liver and transcription of the AASS gene is induced upon starvation. Mutations in the gene encoding AASS result in various forms familial hyperlysinemias (FH), autosomal recessive disorders characterized by hyperlysinemia, lysinuria, and variable saccharopinuria. However, no adverse mental or physical effects have been found in patients with hyperlysinemia. **Function:** Bifunctional enzyme that catalyzes the first two steps in lysine degradation. The N-terminal and the C-terminal contain lysine-ketoglutarate reductase and saccharopine dehydrogenase activity, respectively.

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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	Subunit: Homodimer **Subcellular Location:** Mitochondrial **Tissue Specificity:** Expressed in all 16 tissues examined with highest expression in the liver. DISEASE: Defects in AASS are the cause of hyperlysinemia (HYPLYS) [MIM:238700]. Hyperlysinemia is an autosomal recessive condition characterized by hyperlysinemia lysinuria and variable saccharopinuria. **Similarity:** In the N-terminal section; belongs to the AlaDH/PNT family. In the C-terminal section; belongs to the saccharopine dehydrogenase family. **Database links:** **Entrez Gene:** 10157 Human Omim: 605113 Human **SwissProt:** Q9UDR5 Human **Unigene:** 156738 Human

[VALIDATION IMAGES]



Sample:
Liver(Mouse) Lysate at 40 ug
Kidney (Mouse) Lysate at 40 ug
Primary: Anti- AASS (SL11391R) at 1/1000 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 99 kD
Observed band size: 95 kD

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