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Rabbit Anti-AASS antibody

SL11391R

Product Name:	AASS
Chinese Name:	赖氨酸酮戊二酸还原酶抗体
Alias:	Alpha aminoadipic semialdehyde synthase mitochondrial; LKR/SDH; LKRSDH; LORSDH; Lysine ketoglutarate reductase; Saccharopine dehydrogenase; AASS_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,Sheep,
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:	99kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human LKRSDH:878-926/926
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:	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.

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.

SWISS:
Q9UDR5

Gene ID:
10157

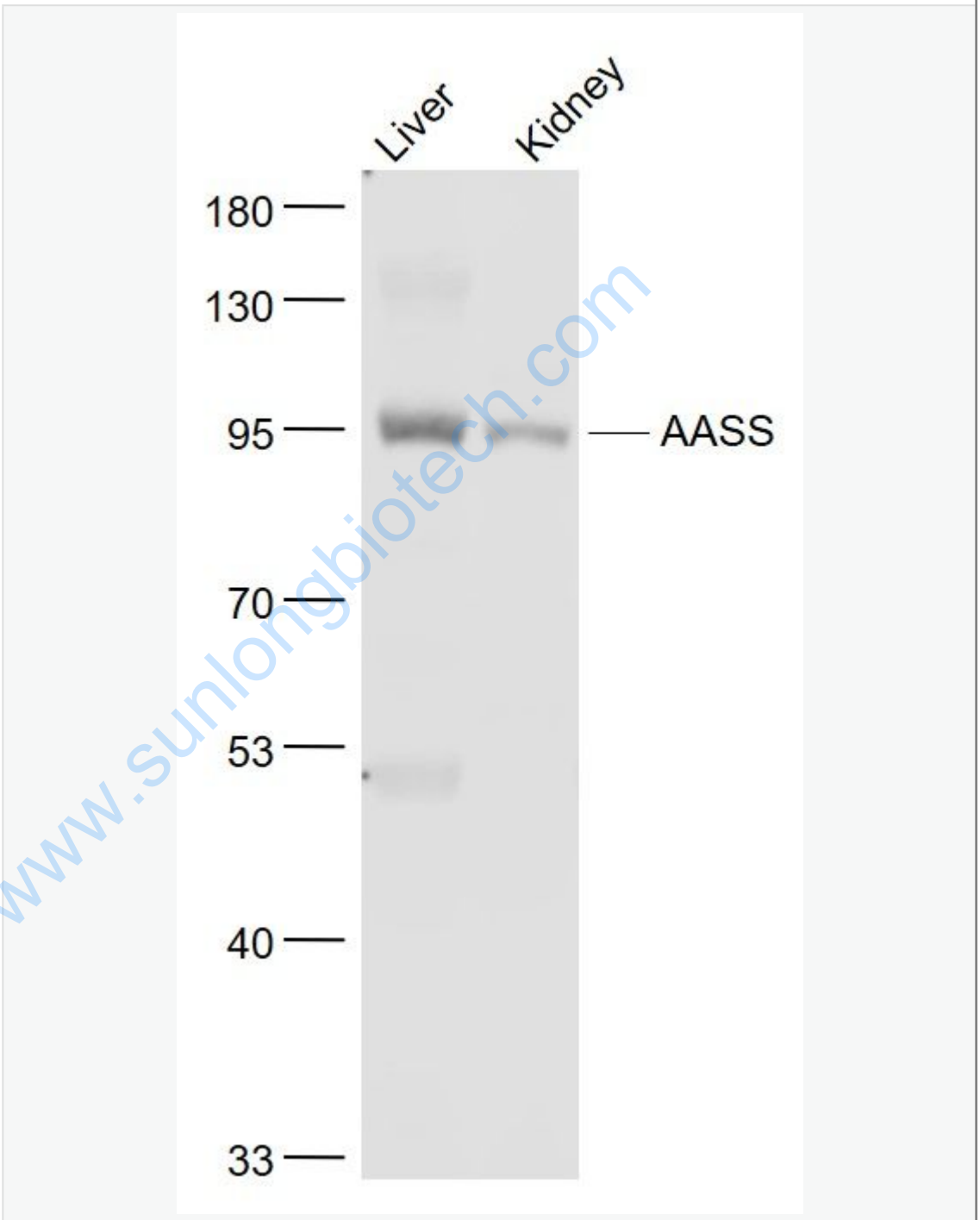
Database links:

Entrez Gene: 10157 Human

Omim: 605113 Human

SwissProt: Q9UDR5 Human

Unigene: 156738 Human

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