赖氨酸酮戊二酸还原酶抗体

产品货号： mlR11391

英文名称： AASS

中文名称： 赖氨酸酮戊二酸还原酶抗体

别　名： Alpha aminoadipic semialdehyde synthase mitochondrial; LKR/SDH; LKRSDH; LORSDH; Lysine ketoglutarate reductase; Saccharopine dehydrogenase; AASS_HUMAN.

研究领域： 肿瘤　细胞生物　信号转导

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Dog, Pig, Cow, Horse, Rabbit, Sheep,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500（石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 99kDa

细胞定位： 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human LKRSDH:878-926/926

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍： 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

Important Note:

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

产品图片