


Anti-HLCS antibody ab100925

2 图像

概述	
产品名称	Anti-HLCS抗体
描述	兔多克隆抗体to HLCS
宿主	Rabbit
经测试应用	适用于: WB, IHC-P
种属反应性	与反应: Human 预测可用于: Mouse, Rat 
免疫原	Recombinant fragment, corresponding to amino acids 544-725 of Human HLCS (BC060787).
阳性对照	Human Fetal Lung, Human Fetal Kidney, Human Fetal Skeletal Muscle
常规说明	<p>The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing.</p> <p>If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As</p>
性能	
形式	Lyophilized:Reconstitute with 200ul distilled sterile water. Please note that if you receive this product in liquid form it has already been reconstituted as described and no further reconstitution is necessary.
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
存储溶液	Constituent: PBS
纯度	Protein A purified
克隆	多克隆
同种型	IgG
应用	

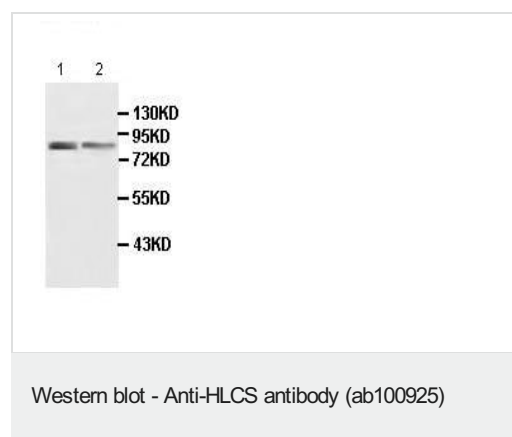
“应用说明”部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
WB		1/500 - 1/1000. Predicted molecular weight: 81 kDa.
IHC-P		1/100 - 1/500.

靶标

功能	Post-translational modification of specific protein by attachment of biotin. Acts on various carboxylases such as acetyl-CoA-carboxylase, pyruvate carboxylase, propionyl CoA carboxylase, and 3-methylcrotonyl CoA carboxylase.
组织特异性	Mostly expressed in muscle, placenta, in lesser extent in the brain, kidney, pancreas, liver and lung.
疾病相关	Defects in HLCS are the cause of holocarboxylase synthetase deficiency (HLCS deficiency) [MIM:253270]; also known as biotin-responsive multiple carboxylase deficiency. HLCS deficiency is a neonatal form of multiple carboxylase deficiency, an autosomal recessive disorder characterized by metabolic ketoacidosis, hyperammonemia, excretion of abnormal organic acid metabolites and dermatitis. Clinical and biochemical symptoms improve dramatically with administration of biotin.
序列相似性	Belongs to the biotin--protein ligase family.
细胞定位	Cytoplasm. Mitochondrion.

图片



All lanes : Anti-HLCS antibody (ab100925) at 1/500 dilution

Lane 1 : Human fetal lung lysate

Lane 2 : Human fetal kidney lysate

Predicted band size: 81 kDa



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-HLCS antibody (ab100925)

Cytoplasmic staining of HLCS in a Formalin/PFA-fixed paraffin-embedded section of Human Fetal Skeletal Muscle using ab100925 at a dilution of 1/100.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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