abcam

Product datasheet

Anti-HLCS antibody ab100925

2 Images

Overview

Product name	Anti-HLCS antibody
Description	Rabbit polyclonal to HLCS
Tested applications	**Suitable for:** WB, IHC-P
Species reactivity	**Reacts with:** Human **Predicted to work with:** Mouse, Rat
Immunogen	Recombinant fragment, corresponding to amino acids 544-725 of Human HLCS (BC060787).
Positive control	Human Fetal Lung, Human Fetal Kidney, Human Fetal Skeletal Muscle

Properties

Form	Lyophilised: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.
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
Storage buffer	Preservative: 0.02% Sodium Azide Constituents: PBS, pH 7.2
Purity	Protein A purified
Clonality	Polyclonal
Isotype	IgG

Applications

Our Abpromise guarantee covers the use of **ab100925** in the following tested applications.

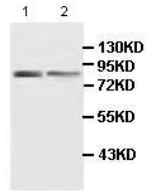
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		1/500 - 1/1000. Predicted molecular weight: 81 kDa.
IHC-P		1/100 - 1/500.

Target

Function	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.
Tissue specificity	Mostly expressed in muscle, placenta, in lesser extent in the brain, kidney, pancreas, liver and lung.
Involvement in disease	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.
Sequence similarities	Belongs to the biotin--protein ligase family.
Cellular localization	Cytoplasm. Mitochondrion.

Images

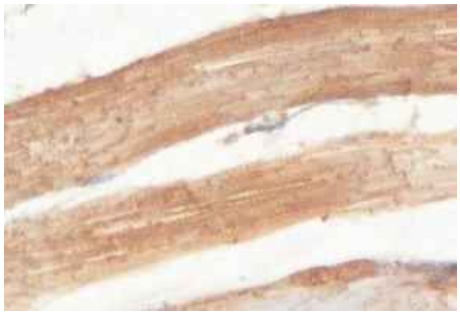


Western blot - HLCS antibody (ab100925)

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) - 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.

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