


Product datasheet

Anti-HLCS antibody ab100925

2 Images

Overview

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

Properties

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

Applications

Our [Abpromise guarantee](#) covers the use of **ab100925** in the following tested applications.

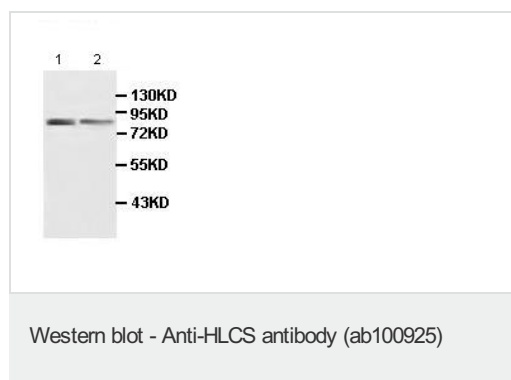
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

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

## Target

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

## Images

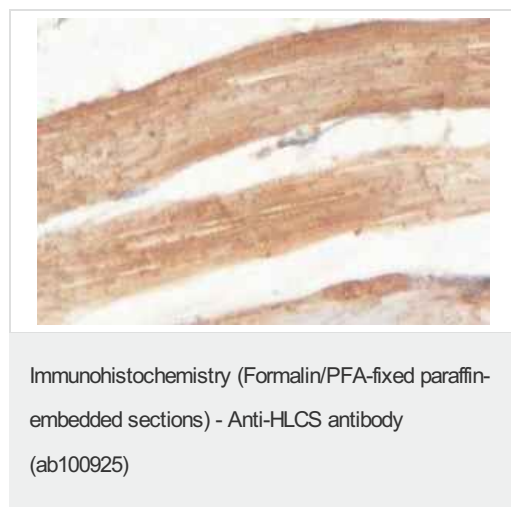


**All lanes :** Anti-HLCS antibody (ab100925) at 1/500 dilution

**Lane 1 :** Human fetal lung lysate

**Lane 2 :** Human fetal kidney lysate

**Predicted band size:** 81 kDa



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 AND ARE NOT INTENDED FOR DIAGNOSTIC OR THERAPEUTIC USE"

## **Our Abpromise to you: Quality guaranteed and expert technical support**

---

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
  
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit <http://www.abcam.com/abpromise> or contact our technical team.

## **Terms and conditions**

---

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors