

## Product datasheet

### Anti-NSDHL antibody [EPR14490] ab190353

KO VALIDATED

Recombinant

RabMAb

[2 References](#) [6 Images](#)

#### Overview

<b>Product name</b>	Anti-NSDHL antibody [EPR14490]
<b>Description</b>	Rabbit monoclonal [EPR14490] to NSDHL
<b>Host species</b>	Rabbit
<b>Tested applications</b>	<b>Suitable for:</b> WB, IHC-P, ICC/IF
<b>Species reactivity</b>	<b>Reacts with:</b> Human
<b>Immunogen</b>	Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.
<b>Positive control</b>	WB: HEK293T, A431, HeLa and Human fetal brain lysates. IHC-P: Human liver and Human adenocarcinoma of endometrium tissue. ICC: U87-MG cells.
<b>General notes</b>	<p>This product is a recombinant monoclonal antibody, which offers several advantages including:</p> <ul style="list-style-type: none"> <li>- High batch-to-batch consistency and reproducibility</li> <li>- Improved sensitivity and specificity</li> <li>- Long-term security of supply</li> <li>- Animal-free production</li> </ul> <p>For more information <a href="#">see here</a>.</p> <p>Our RabMAb<sup>®</sup> technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to <a href="#">RabMAb<sup>®</sup> patents</a>.</p>

#### Properties

<b>Form</b>	Liquid
<b>Storage instructions</b>	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.
<b>Storage buffer</b>	<p>pH: 7.2</p> <p>Preservative: 0.01% Sodium azide</p> <p>Constituents: 59% PBS, 40% Glycerol, 0.05% BSA</p>
<b>Purity</b>	Protein A purified
<b>Clonality</b>	Monoclonal
<b>Clone number</b>	EPR14490
<b>Isotype</b>	IgG

## Applications

### The Abpromise guarantee

Our **Abpromise guarantee** covers the use of ab190353 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		1/1000 - 1/10000. Detects a band of approximately 38 kDa (predicted molecular weight: 42 kDa).
IHC-P		1/50 - 1/100. Perform heat mediated antigen retrieval with Tris/EDTA buffer pH 9.0 before commencing with IHC staining protocol.
ICC/IF		1/50.

## Target

### Tissue specificity

Brain, heart, liver, lung, kidney, skin and placenta.

### Pathway

Steroid biosynthesis; zymosterol biosynthesis; zymosterol from lanosterol: step 4/6.

### Involvement in disease

Defects in NSDHL are the cause of congenital hemidysplasia with ichthyosiform erythroderma and limb defects (CHILD) [MIM:308050]. CHILD is an X-linked dominant disorder of lipid metabolism with disturbed cholesterol biosynthesis, which typically results in male lethality. Clinically, it is characterized by congenital, unilateral, ichthyosiform erythroderma with striking lateralization, sharp midline demarcation, and ipsilateral limb defects and hypoplasia of the body. Limbs defects range from hypoplasia of digits or ribs to complete amelia, often including scoliosis.

Defects in NSDHL are the cause of CK syndrome (CKS) [MIM:300831]. CKS is a disorder characterized by mild to severe cognitive impairment, seizures, microcephaly, cerebral cortical malformations, dysmorphic facial features, and thin body habitus.

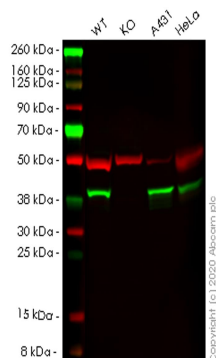
### Sequence similarities

Belongs to the 3-beta-HSD family.

### Cellular localization

Membrane.

## Images



Western blot - Anti-NSDHL antibody [EPR14490] (ab190353)

**All lanes :** Anti-NSDHL antibody [EPR14490] (ab190353) at 1/1000 dilution

**Lane 1 :** Wild-type HEK293T cell lysate

**Lane 2 :** NSDHL knockout HEK293T cell lysate

**Lane 3 :** A431 cell lysate

**Lane 4 :** HeLa cell lysate

Lysates/proteins at 20 µg per lane.

### Secondary

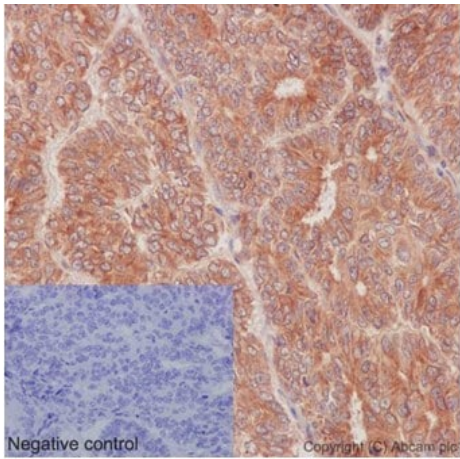
**All lanes :** Goat anti-Rabbit IgG H&L (IRDye® 800CW) preadsorbed ([ab216773](#)) at 1/10000 dilution

**Predicted band size:** 42 kDa

**Observed band size:** 38 kDa

**Lanes 1-4:** Merged signal (red and green). Green - ab190353 observed at 38 kDa. Red - loading control [ab7291](#) observed at 50 kDa.

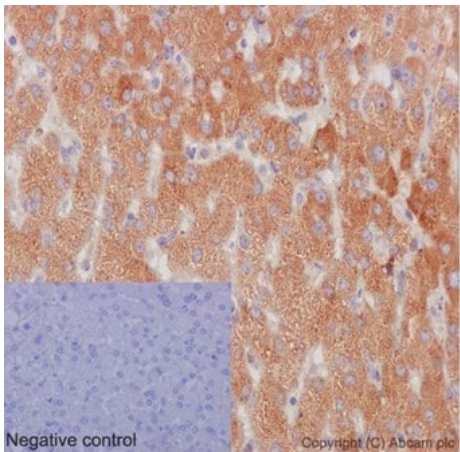
ab190353 Anti-NSDHL antibody [EPR14490] was shown to specifically react with NSDHL in wild-type HEK293T cells. Loss of signal was observed when knockout cell line [ab266682](#) (knockout cell lysate [ab258082](#)) was used. Wild-type and NSDHL knockout samples were subjected to SDS-PAGE. ab190353 and Anti-alpha Tubulin antibody [DM1A] - Loading Control ([ab7291](#)) were incubated overnight at 4°C at 1 in 1000 dilution and 1 in 20000 dilution respectively. Blots were developed with Goat anti-Rabbit IgG H&L (IRDye® 800CW) preadsorbed ([ab216773](#)) and Goat anti-Mouse IgG H&L (IRDye® 680RD) preadsorbed ([ab216776](#)) secondary antibodies at 1 in 20000 dilution for 1 hour at room temperature before imaging.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-NSDHL antibody [EPR14490] (ab190353)

Immunohistochemical analysis of paraffin-embedded Human adenocarcinoma of endometrium tissue labeling NSDHL with ab190353 at 1/100 dilution. Secondary antibody HRP Polymer for Rabbit IgG prediluted. Counter stain Hematoxylin. Negative control also shown.

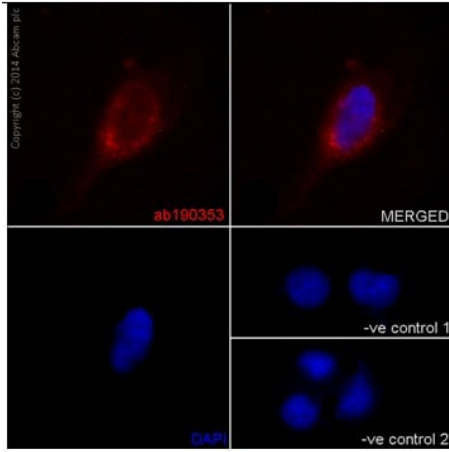
Perform heat mediated antigen retrieval with Tris/EDTA buffer pH 9.0 before commencing with IHC staining protocol.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-NSDHL antibody [EPR14490] (ab190353)

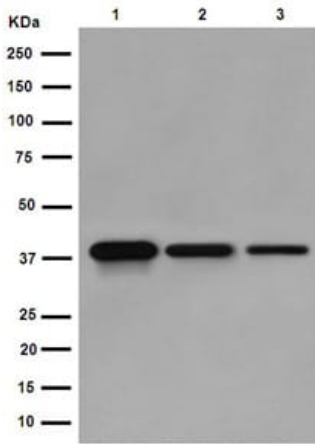
Immunohistochemical analysis of paraffin-embedded Human liver tissue labeling NSDHL with ab190353 at 1/100 dilution. Secondary antibody HRP Polymer for Rabbit IgG prediluted. Counter stain Hematoxylin. Negative control also shown.

Perform heat mediated antigen retrieval with Tris/EDTA buffer pH 9.0 before commencing with IHC staining protocol.



Immunocytochemistry/ Immunofluorescence - Anti-NSDHL antibody [EPR14490] (ab190353)

Immunofluorescence analysis of U87-MG cells, labeling NSDHL (red) with ab190353 at 1/50 dilution. Alexa Fluor®555-conjugated goat anti-rabbit IgG was used as a secondary antibody at 1/200 dilution. Nuclei were counterstained with DAPI (blue).



Western blot - Anti-NSDHL antibody [EPR14490] (ab190353)

**All lanes :** Anti-NSDHL antibody [EPR14490] (ab190353) at 1/10000 dilution

**Lane 1 :** A431 cell lysate

**Lane 2 :** HeLa cell lysate

**Lane 3 :** Human fetal brain tissue lysate

Lysates/proteins at 20 µg per lane.

#### Secondary

**All lanes :** Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/1000 dilution

**Predicted band size:** 42 kDa

**Observed band size:** 38 kDa

### Why choose a recombinant antibody?



**Research with confidence**  
Consistent and reproducible results



**Long-term and scalable supply**  
Recombinant technology



**Success from the first experiment**  
Confirmed specificity



**Ethical standards compliant**  
Animal-free production

Anti-NSDHL antibody [EPR14490] (ab190353)

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

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- 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

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