

Product datasheet

Anti-NSDHL antibody [EPR14490] ab190353

Recombinant RabMAb

[4 Images](#)

Overview

Product name	Anti-NSDHL antibody [EPR14490]
Description	Rabbit monoclonal [EPR14490] to NSDHL
Host species	Rabbit
Tested applications	Suitable for: WB, IHC-P, ICC/IF
Species reactivity	Reacts with: Mouse, Rat, Human
Immunogen	Synthetic peptide within Human NSDHL aa 150-250. The exact sequence is proprietary. Database link: Q15738
Positive control	A431, HeLa and Human fetal brain lysates. Human liver and Human adenocarcinoma of endometrium tissue. U87-MG cells.

General notes

Our RabMAb[®] technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to [RabMAb[®] patents](#).

This product is a [recombinant rabbit monoclonal antibody](#).

Properties

Form	Liquid
Storage instructions	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.
Storage buffer	Preservative: 0.01% Sodium azide Constituents: 59% PBS, 40% Glycerol, 0.05% BSA
Purity	Protein A purified
Clonality	Monoclonal
Clone number	EPR14490
Isotype	IgG

Applications

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.

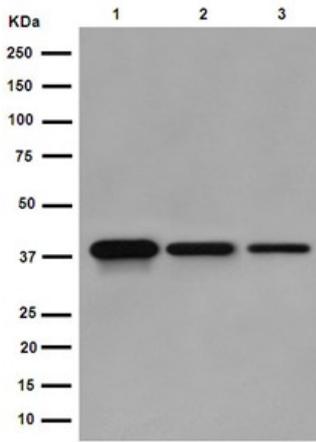
Sequence similarities

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 1/10000 dilution

Lane 1 : A431 cell lysate

Lane 2 : HeLa cell lysate

Lane 3 : Human fetal brain lysate

Lysates/proteins at 20 µg per lane.

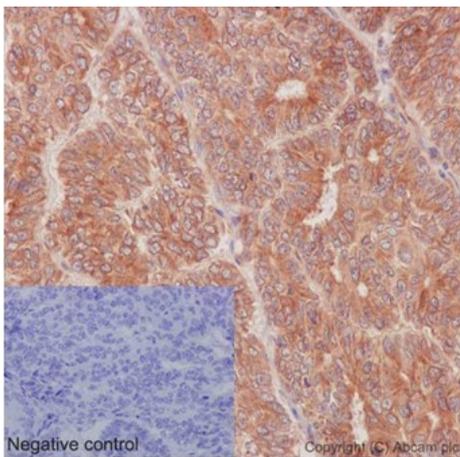
Secondary

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

Predicted band size: 42 kDa

Observed band size: 38 kDa

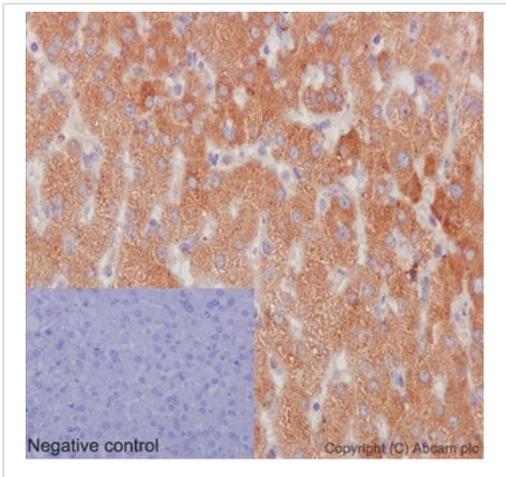
[why is the actual band size different from the predicted?](#)



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.

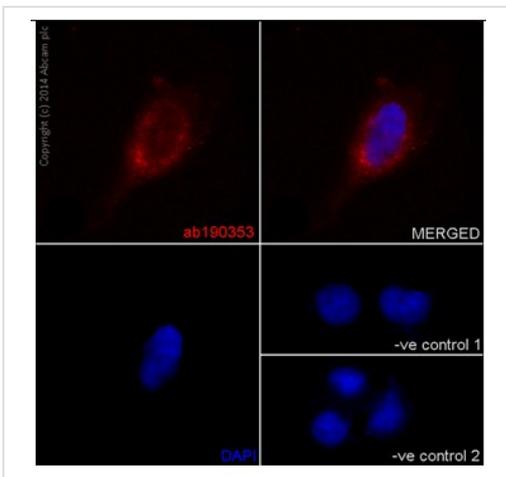
Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-NSDHL antibody [EPR14490] (ab190353)



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.

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - 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).

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

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