


Product datasheet

Anti-ALAS2/ASB antibody ab136799

2 Images

Overview

Product name	Anti-ALAS2/ASB antibody
Description	Rabbit polyclonal to ALAS2/ASB
Host species	Rabbit
Tested applications	Suitable for: WB, IHC-P
Species reactivity	Reacts with: Human Predicted to work with: Mouse, Rat, Rabbit, Horse, Cow, Dog, Pig, Macaque monkey, Gorilla, Orangutan 
Immunogen	Synthetic peptide corresponding to Human ALAS2/ASB aa 550 to the C-terminus conjugated to keyhole limpet haemocyanin. (Peptide available as ab166589)
Positive control	This antibody gave a positive signal in Human Heart tissue and Human Heart Mitochondrial lysates. This antibody gave a positive result in IHC in the following FFPE tissue: Human normal liver.
General notes	This product was previously labelled as ALAS2

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 or -80°C. Avoid freeze / thaw cycle.
Storage buffer	pH: 7.40 Preservative: 0.02% Sodium azide Constituent: PBS Note: Batches of this product that have a concentration < 1mg/ml may have BSA added as a stabilising agent. If you would like information about the formulation of a specific lot, please contact our scientific support team who will be happy to help.
Purity	Immunogen affinity purified
Clonality	Polyclonal
Isotype	IgG

Applications

Our [Abpromise guarantee](#) covers the use of **ab136799** 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		Use a concentration of 1 µg/ml. Detects a band of approximately 58 kDa (predicted molecular weight: 64 kDa).
IHC-P		Use a concentration of 5 µg/ml. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.

Target

Tissue specificity

Erythroid specific.

Pathway

Porphyrin metabolism; protoporphyrin-IX biosynthesis; 5-aminolevulinate from glycine: step 1/1.

Involvement in disease

Defects in ALAS2 are a cause of anemia sideroblastic X-linked (XLSA) [MIM:300751]. Sideroblastic anemia is characterized by anemia of varying severity, hypochromic peripheral erythrocytes, systemic iron overload secondary to chronic ineffective erythropoiesis, and the presence of bone marrow ringed sideroblasts. Sideroblasts are characterized by iron-loaded mitochondria clustered around the nucleus. XLSA shows a variable hematologic response to pharmacologic doses of pyridoxine.

Defects in ALAS2 are the cause of erythropoietic protoporphyria X-linked dominant (XLDPT) [MIM:300752]. Porphyrias are inherited defects in the biosynthesis of heme, resulting in the accumulation and increased excretion of porphyrins or porphyrin precursors. They are classified as erythropoietic or hepatic, depending on whether the enzyme deficiency occurs in red blood cells or in the liver. XLDPT is a form of porphyria characterized biochemically by a high proportion of zinc-protoporphyrin in erythrocytes, in which a mismatch between protoporphyrin production and the heme requirement of differentiating erythroid cells leads to overproduction of protoporphyrin in amounts sufficient to cause photosensitivity and liver disease. Note=Gain of function mutations in ALS2 are responsible for XLDPT, but they can also be a possible aggravating factor in congenital erythropoietic porphyria and other erythropoietic disorders caused by mutations in other genes (PubMed:21309041).

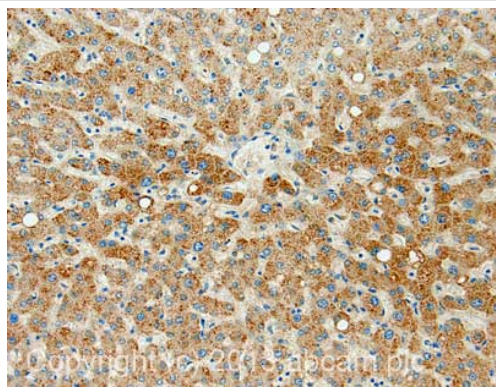
Sequence similarities

Belongs to the class-II pyridoxal-phosphate-dependent aminotransferase family.

Cellular localization

Mitochondrion matrix.

Images



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-ALAS2/ASB antibody (ab136799)

IHC image of ALAS2/ASB staining in Human normal liver formalin fixed paraffin embedded tissue section, performed on a Leica Bond™ system using the standard protocol F. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH6, epitope retrieval solution 1) for 20 mins. The section was then incubated with ab136799, 5µg/ml, for 15 mins at room temperature and detected using an HRP conjugated compact polymer system. DAB was used as the chromogen. The section was then counterstained with haematoxylin and mounted with DPX.

For other IHC staining systems (automated and non-automated) customers should optimize variable parameters such as antigen retrieval conditions, primary antibody concentration and antibody incubation times.



Western blot - Anti-ALAS2/ASB antibody (ab136799)

All lanes : Anti-ALAS2/ASB antibody (ab136799) at 1 µg/ml

Lane 1 : Human heart tissue lysate - total protein (ab29431)

Lane 2 : Human Heart Mitochondrial Lysate

Lysates/proteins at 10 µg per lane.

Secondary

All lanes : Goat Anti-Rabbit IgG H&L (HRP) (ab97051) at 1/10000 dilution

Developed using the ECL technique.

Performed under reducing conditions.

Predicted band size: 64 kDa

Observed band size: 58 kDa

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

Additional bands at: 43 kDa. We are unsure as to the identity of these extra bands.

Exposure time: 10 seconds

The band observed at 58 kDa could potentially be a cleaved form of ALAS2/ASB due to the presence of a 49 amino acid transit peptide. This blot was produced using a 4-12% Bis-tris gel under the MOPS buffer system. The gel was run at 200V for 50 minutes before being transferred onto a Nitrocellulose membrane at 30V for 70 minutes. The membrane was then blocked for an hour using 5% Bovine Serum Albumin before being incubated with ab136799 overnight at 4°C. Antibody binding was detected using an anti-rabbit antibody conjugated to HRP, and visualised using ECL development solution.

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

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 <https://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