abcam

Product datasheet

Anti-ALAS2/ASB antibody ab136799

1 References 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	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

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 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

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab136799 in the following tested applications.

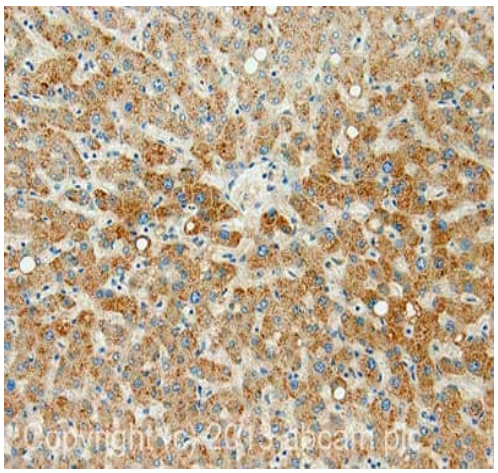
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

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
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.

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