abcam

Product datasheet

Anti-BTD antibody ab126140

1 Image

Overview

Product name	Anti-BTD antibody
Description	Rabbit polyclonal to BTD
Host species	Rabbit
Tested applications	**Suitable for:** WB
Species reactivity	**Reacts with:** Human **Predicted to work with:** Mouse
Immunogen	Recombinant fragment, corresponding to a region within amino acids 206-449 of Human BTD (P43251).
Positive control	293T, A431, Jurkat and Raji whole cell lysates.
General notes	Store as concentrated solution.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.
Storage buffer	pH: 7.00 Preservative: 0.025% Proclin Constituents: PBS, 1% BSA, 20% Glycerol
Purity	Immunogen affinity purified
Clonality	Polyclonal
Isotype	IgG

Applications

Our Abpromise guarantee covers the use of **ab126140** in the following tested applications.

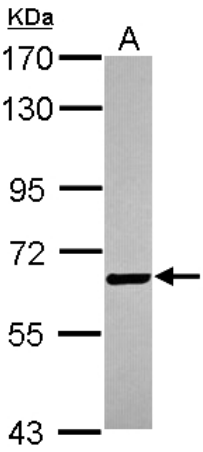
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		1/500 - 1/3000. Predicted molecular weight: 61 kDa.

Target

Function	Catalytic release of biotin from biocytin, the product of biotin-dependent carboxylases degradation.
Involvement in disease	Defects in BTD are the cause of biotinidase deficiency (BTD deficiency) [MIM:253260]; also called late-onset multiple carboxylase deficiency. BTD deficiency is a juvenile form of multiple carboxylase deficiency, an autosomal recessive disorder of biotin metabolism, characterized by ketoacidosis, hyperammonemia, excretion of abnormal organic acid metabolites, and dermatitis. BTD deficiency is characterized by seizures, hypotonia, skin rash, alopecia, ataxia, hearing loss, and optic atrophy. If untreated, symptoms usually become progressively worse, and coma and death may occur.
Sequence similarities	Belongs to the CN hydrolase family. BTD/VNN subfamily. Contains 1 CN hydrolase domain.
Cellular localization	Secreted > extracellular space.

Images



Western blot - Anti-BTD antibody (ab126140)

Anti-BTD antibody (ab126140) at 1/1000 dilution + Jurkat lysate at 30 µg

Predicted band size: 61 kDa

7.5% SDS PAGE

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