abcam

Product datasheet

Recombinant Human ATP7b protein ab152216

1 Image

Description

Product name Recombinant Human ATP7b protein

Expression system Wheat germ
Accession P35670

Protein length Protein fragment

Animal free No

Nature Recombinant

Species Human

Sequence QLKCYKKPDLERYEAQAHGHMKPLTASQVSVHIGMDDR

WRDSPRATPWDQ

VSYVSQVSLSSLTSDKPSRHSAAADDDGDKWSLLLNGR

DEEQY

Predicted molecular weight 36 kDa including tags

Amino acids 1372 to 1465

Specifications

Our Abpromise guarantee covers the use of ab152216 in the following tested applications.

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

Applications SDS-PAGE

ELISA

Western blot

Form Liquid

Additional notes

Preparation and Storage

Stability and Storage Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.

pH: 8.00

Constituents: 0.31% Glutathione, 0.79% Tris HCI

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General Info

Function Involved in the export of copper out of the cells, such as the efflux of hepatic copper into the bile.

Tissue specificity Most abundant in liver and kidney and also found in brain. Isoform 2 is expressed in brain but not

in liver. The cleaved form WND/140 kDa is found in liver cell lines and other tissues.

Involvement in disease Defects in ATP7B are the cause of Wilson disease (WD) [MIM:277900]. WD is an autosomal

recessive disorder of copper metabolism in which copper cannot be incorporated into

ceruloplasmin in liver, and cannot be excreted from the liver into the bile. Copper accumulates in the liver and subsequently in the brain and kidney. The disease is characterized by neurologic

manifestations and signs of cirrhosis.

Sequence similarities Belongs to the cation transport ATPase (P-type) (TC 3.A.3) family. Type IB subfamily.

Contains 6 HMA domains.

Post-translational

modifications

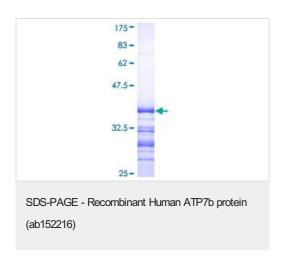
Isoform 1 may be proteolytically cleaved at the N-terminus to produce the WND/140 kDa form.

Cellular localization Cytoplasm; Mitochondrion and Golgi apparatus > trans-Golgi network membrane. Predominantly

found in the trans-Golgi network (TGN). Not redistributed to the plasma membrane in response to

elevated copper levels.

Images



12.5% SDS-PAGE analysis of ab152216 stained with Coomassie Blue.

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