

HAH1 (22-B): sc-100557

BACKGROUND

Delivery of copper to a specific P-type ATPase in the Golgi apparatus (Ccc2 in yeast, ATP7A and ATP7B in humans) is an important step in copper homeostasis that is accomplished by a small copper chaperone protein. HAH1 (also designated ATOX1), a metal transport protein that belongs to the ATX1 family, is involved in cellular antioxidant defense and can bind and deliver cytosolic copper to the copper ATPase proteins. Both HAH1 (the human homolog of Atx1) and Atx1 preferentially interact with domains 2 and 4 of ATP7B. Atx1 also interacts with both Ccc2 domains.

REFERENCES

1. Klomp, L.W., Lin, S.J., Yuan, D.S., Klausner, R.D., Culotta, V.C. and Gitlin, J.D. 1997. Identification and functional expression of HAH1, a novel human gene involved in copper homeostasis. *J. Biol. Chem.* 272: 9221-9226.
2. Harrison, M.D., Jones, C.E., Solioz, M. and Dameron, C.T. 2000. Intracellular copper routing: the role of copper chaperones. *Trends Biochem. Sci.* 25: 29-32.
3. Boultonwood, J., Strickson, A.J., Jabs, E.W., Cheng, J.F., Fidler, C. and Wainscoat, J.S. 2000. Physical mapping of the human ATX1 homologue (HAH1) to the critical region of the 5q- syndrome within 5q32, and immediately adjacent to the SPARC gene. *Hum. Genet.* 106: 127-129.
4. van Dongen, E.M., Klomp, L.W. and Merks, M. 2004. Copper-dependent protein-protein interactions studied by yeast two-hybrid analysis. *Biochem. Biophys. Res. Commun.* 323: 789-795.

CHROMOSOMAL LOCATION

Genetic locus: ATOX1 (human) mapping to 5q33.1.

SOURCE

HAH1 (22-B) is a mouse monoclonal antibody raised against recombinant HAH1 of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

HAH1 (22-B) is recommended for detection of HAH1 of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for HAH1 siRNA (h): sc-45252, HAH1 shRNA Plasmid (h): sc-45252-SH and HAH1 shRNA (h) Lentiviral Particles: sc-45252-V.

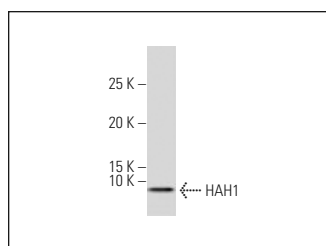
Molecular Weight of HAH1: 8 kDa.

Positive Controls: HeLa nuclear extract: sc-2120.

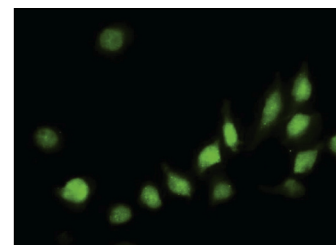
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



HAH1 (22-B): sc-100557. Western blot analysis of HAH1 expression in HeLa nuclear extract.



HAH1 (22-B): sc-100557. Immunofluorescence staining of paraformaldehyde-fixed HeLa cells showing nuclear localization.

SELECT PRODUCT CITATIONS

1. Charbonnier, P., Chovelon, B., Ravelet, C., Ngo, T.D., Chevallet, M. and Deniaud, A. 2022. ATP7B-deficient hepatocytes reveal the importance of protein misfolding induced at low copper concentration. *Cells* 11: 3400.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.