

AUH (B-8): sc-518216

BACKGROUND

AUH (AU-binding protein/enoyl-CoA hydratase), also known as mitochondrial methylglutaconyl-CoA hydratase, is a 339 amino acid member of the enoyl-CoA hydratase/isomerase family. AUH is involved in the amino acid degradation pathway by catalyzing the conversion of 3-methylglutaconyl-CoA to 3-hydroxy-3-methylglutaryl-CoA and water. Localized to the mitochondria, AUH has been found to have very low enoyl-CoA hydratase activity. AUH is expressed as two isoforms produced by alternative splicing and forms a homo-hexamer. Defects in AUH result in 3-methylglutaconic aciduria type 1 (MGA1), an inborn error of leucine metabolism. MGA1 has a varied clinical phenotype, including coma, severe psychomotor retardation, delayed speech development, failure to thrive, metabolic acidosis and dystonia.

REFERENCES

1. Nakagawa, J., et al. 1995. AUH, a gene encoding an AU-specific RNA binding protein with intrinsic enoyl-CoA hydratase activity. *Proc. Natl. Acad. Sci. USA* 92: 2051-2055.
2. Nakagawa, J. and Moroni, C. 1997. A 20-amino-acid autonomous RNA-binding domain contained in an enoyl-CoA hydratase. *Eur. J. Biochem.* 244: 890-899.
3. Brennan, L.E., et al. 1999. Characterisation and mitochondrial localisation of AUH, an AU-specific RNA-binding enoyl-CoA hydratase. *Gene* 228: 85-91.
4. Kurimoto, K., et al. 2001. Crystal structure of human AUH protein, a single-stranded RNA binding homolog of enoyl-CoA hydratase. *Structure* 9: 1253-1263.
5. IJlst, L., et al. 2002. 3-methylglutaconic aciduria type I is caused by mutations in AUH. *Am. J. Hum. Genet.* 71: 1463-1466.
6. Ly, T.B., et al. 2003. Mutations in the AUH gene cause 3-methylglutaconic aciduria type I. *Hum. Mutat.* 21: 401-407.

CHROMOSOMAL LOCATION

Genetic locus: AUH (human) mapping to 9q22.31; Auh (mouse) mapping to 13 B1.

SOURCE

AUH (B-8) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 321-339 of AUH of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

AUH (B-8) is available conjugated to agarose (sc-518216 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-518216 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-518216 PE), fluorescein (sc-518216 FITC), Alexa Fluor® 488 (sc-518216 AF488), Alexa Fluor® 546 (sc-518216 AF546), Alexa Fluor® 594 (sc-518216 AF594) or Alexa Fluor® 647 (sc-518216 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-518216 AF680) or Alexa Fluor® 790 (sc-518216 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

AUH (B-8) is recommended for detection of AUH of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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 AUH siRNA (h): sc-72593, AUH siRNA (m): sc-72594, AUH shRNA Plasmid (h): sc-72593-SH, AUH shRNA Plasmid (m): sc-72594-SH, AUH shRNA (h) Lentiviral Particles: sc-72593-V and AUH shRNA (m) Lentiviral Particles: sc-72594-V.

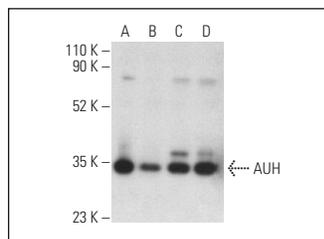
Molecular Weight of AUH: 32 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, SH-SY5Y cell lysate: sc-3812 or SW480 cell lysate: sc-2219.

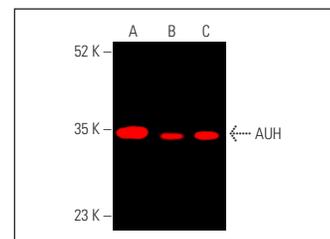
RECOMMENDED SUPPORT REAGENTS

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



AUH (B-8): sc-518216. Western blot analysis of AUH expression in Hep G2 (A), SH-SY5Y (B), A549 (C) and SW480 (D) whole cell lysates. Detection reagent used: m-IgG₁ BP-HRP: sc-525408.



AUH (B-8): sc-518216. Near-Infrared western blot analysis of AUH expression in Hep G2 (A), A549 (B) and SW480 (C) whole cell lysates. Blocked with UltraCruz® Blocking Reagent: sc-516214. Detection reagent used: m-IgG₁ BP-CFL 790: sc-533666.

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.

Alexa Fluor® is a trademark of Molecular Probes, Inc., Oregon, USA