SANTA CRUZ BIOTECHNOLOGY, INC.

AUH (C-14): sc-82518

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 homohexamer. 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.
7. Illsinger, S., et al. 2004. 3-methylglutaconic aciduria type I in a boy with fever-associated seizures. Pediatr. Neurol. 30: 213-215.
8. Mack, M., et al. 2006. Biochemical characterization of human 3-methylglutaconyl-CoA hydratase and its role in leucine metabolism. FEBS J. 273: 2012-2022.
9. Kurimoto, K., et al. 2009. AU-rich RNA-binding induces changes in the quaternary structure of AUH. Proteins 75: 360-372.

CHROMOSOMAL LOCATION

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

SOURCE

AUH (C-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of AUH of human origin.

RESEARCH USE

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

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-82518 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

AUH (C-14) is recommended for detection of AUH of mouse, rat and 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).

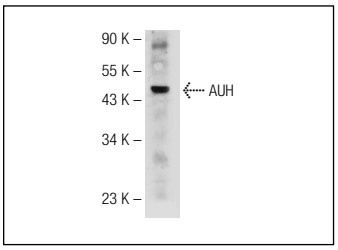
AUH (C-14) is also recommended for detection of AUH in additional species, including equine, canine, bovine and porcine.

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: mouse brain extract: sc-2253.

DATA



AUH (C-14): sc-82518. Western blot analysis of AUH expression in mouse brain tissue extract.

SELECT PRODUCT CITATIONS

1. Ness, G.C., et al. 2012. Involvement of tristetraprolin in transcriptional activation of hepatic 3-hydroxy-3-methylglutaryl coenzyme A reductase by Insulin. Biochem. Biophys. Res. Commun. 420: 178-182.

STORAGE

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

PROTOCOLS

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