SANTA CRUZ BIOTECHNOLOGY, INC.

UROS (M-265): sc-134760



BACKGROUND

Uroporphyrinogen-III synthase (UROS), also designated hydroxymethylbilane hydrolyase, is a 265 amino acid ubiquitously expressed protein that is predominantly found in skeletal muscle, heart muscle, the caudate nucleus, and the amygdala. UROS is the fourth enzyme in the human heme biosynthetic pathway and is responsible for the conversion of hydroxymethylbilane to the cyclic tetrapyrrole, uroporphyrinogen III. UROS is a monomer involved in pophyrin biosynthesis by the Shemin pathway. Defects in the UROS gene can cause molecular lesions that lead to the autosomal recessive Gunther disease, also known as congenital erythropoietic porphyria (CEP). CEP is characterized by accumulation of uroporphyrin I in the bone marrow, peripheral blood, and other organs, often causing death early in adult life.

REFERENCES

- Tsai, S.F., Bishop, D.F. and Desnick, R.J. 1987. Purification and properties of uroporphyrinogen III synthase from human erythrocytes. J. Biol. Chem. 262: 1268-1273.
- Tsai, S.F., Bishop, D.F. and Desnick, R.J. 1988. Human uroporphyrinogen III synthase: molecular cloning, nucleotide sequence, and expression of a fulllength cDNA. Proc. Natl. Acad. Sci. USA 85: 7049-7053.
- Astrin, K.H., Warner, C.A., Yoo, H.W., Goodfellow, P.J., Tsai, S.F. and Desnick, R.J. 1991. Regional assignment of the to chromosome 10q25.2-q26.3. Hum. Genet. 87: 18-22.
- Xu, W., Kozak, C.A. and Desnick, R.J. 1995. Uroporphyrinogen-III synthase: molecular cloning, nucleotide sequence, expression of a mouse full-length cDNA, and its localization on mouse chromosome 7. Genomics 26: 556-562.
- Geronimi, F., Richard, E., Lamrissi-Garcia, I., Lalanne, M., Ged, C., Redonnet-Vernhet, I., Moreau-Gaudry, F. and de Verneuil, H. 2003. Lentivirus-mediated gene transfer of uroporphyrinogen III synthase fully corrects the porphyric phenotype in human cells. J. Mol. Med. 81: 310-320.
- Torres, G., Quiñones, M., Hau, J., Perez, L. and García, O. 2003. HLA-DQA1 and polymarker in the UROS population from Peruvian Highland. J. Forensic Sci. 48: 1426-1427.
- Omata, Y., Sakamoto, H., Higashimoto, Y., Hayashi, S. and Noguchi, M. 2004. Purification and characterization of human uroporphyrinogen III synthase expressed in *Escherichia coli*. J. Biochem. 136: 211-220.
- de Villiers, J.N., Kotze, M.J., van Heerden, C.J., Sadie, A., Gardner, H.F., Liebenberg, J., van Zyl, R., du Plessis, L., Kimberg, M., Frank, J. and Warnich, L. 2005. Overrepresentation of the founder PPOX gene mutation R59W in a South African patient with severe clinical manifestation of porphyria. Exp. Dermatol. 14: 50-55.

CHROMOSOMAL LOCATION

Genetic locus: UROS (human) mapping to 10q26.13; Uros (mouse) mapping to 7 F3.

RESEARCH USE

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

SOURCE

UROS (M-265) is a rabbit polyclonal antibody raised against amino acids 1-265 representing full length UROS of mouse origin.

PRODUCT

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

APPLICATIONS

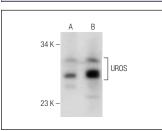
UROS (M-265) is recommended for detection of uroporphyrinogen-III synthase (UROS) of mouse, rat and, to a lesser extent, human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (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 UROS siRNA (h): sc-61760, UROS siRNA (m): sc-61761, UROS shRNA Plasmid (h): sc-61760-SH, UROS shRNA Plasmid (m): sc-61761-SH, UROS shRNA (h) Lentiviral Particles: sc-61760-V and UROS shRNA (m) Lentiviral Particles: sc-61761-V.

Molecular Weight of UROS: 30 kDa.

Positive Controls: mouse thyroid extract: sc-2407, mouse colon extract: sc-364238 or mouse brain extract: sc-2253.

DATA



UROS (M-265): sc-134760. Western blot analysis of UROS expression in mouse colon (**A**) and mouse brain (**B**) tissue extracts.

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.