

UROS (G-9): sc-365120

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 porphyrin 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

1. Tsai, S.F., et al. 1987. Purification and properties of uroporphyrinogen III synthase from human erythrocytes. *J. Biol. Chem.* 262: 1268-1273.
2. Tsai, S.F., et al. 1988. Human uroporphyrinogen III synthase: molecular cloning, nucleotide sequence, and expression of a full-length cDNA. *Proc. Natl. Acad. Sci. USA* 85: 7049-7053.
3. Astrin, K.H., et al. 1991. Regional assignment of the to chromosome 10q25.2-q26.3. *Hum. Genet.* 87: 18-22.
4. Xu, W., et al. 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.
5. Geronimi, F., et al. 2003. Lentivirus-mediated gene transfer of uroporphyrinogen III synthase fully corrects the porphyric phenotype in human cells. *J. Mol. Med.* 81: 310-320.

CHROMOSOMAL LOCATION

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

SOURCE

UROS (G-9) is a mouse monoclonal antibody raised against amino acids 1-265 representing full length UROS 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.

UROS (G-9) is available conjugated to agarose (sc-365120 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-365120 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-365120 PE), fluorescein (sc-365120 FITC), Alexa Fluor[®] 488 (sc-365120 AF488), Alexa Fluor[®] 546 (sc-365120 AF546), Alexa Fluor[®] 594 (sc-365120 AF594) or Alexa Fluor[®] 647 (sc-365120 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-365120 AF680) or Alexa Fluor[®] 790 (sc-365120 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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

APPLICATIONS

UROS (G-9) is recommended for detection of Uroporphyrinogen-III synthase (UROS) 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 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.

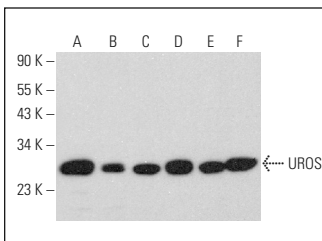
Molecular Weight of UROS: 30 kDa.

Positive Controls: K-562 whole cell lysate: sc-2203, ZR-75-1 cell lysate: sc-2241 or Sol8 cell lysate: sc-2249.

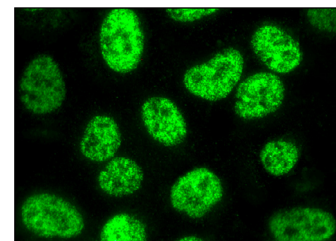
RECOMMENDED SUPPORT REAGENTS

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



UROS (G-9): sc-365120. Western blot analysis of UROS expression in K-562 (A), ZR-75-1 (B), Sol8 (C), A-673 (D), L6 (E) and C2C12 (F) whole cell lysates.



UROS (G-9): sc-365120. Immunofluorescence staining of formalin-fixed Hep G2 cells showing nuclear localization.

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.