

UROS (A-8): sc-365877

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 heme 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. Géronimi, 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.
6. Torres, G., et al. 2003. HLA-DQA1 and polymarker in the Uros population from Peruvian Highland. *J. Forensic Sci.* 48: 1426-1427.

CHROMOSOMAL LOCATION

Genetic locus: Uros (mouse) mapping to 7 F3.

SOURCE

UROS (A-8) is a mouse monoclonal antibody raised against amino acids 1-265 representing full length UROS of mouse 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 (A-8) is available conjugated to agarose (sc-365877 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-365877 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-365877 PE), fluorescein (sc-365877 FITC), Alexa Fluor® 488 (sc-365877 AF488), Alexa Fluor® 546 (sc-365877 AF546), Alexa Fluor® 594 (sc-365877 AF594) or Alexa Fluor® 647 (sc-365877 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-365877 AF680) or Alexa Fluor® 790 (sc-365877 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 (A-8) is recommended for detection of UROS of mouse 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 (m): sc-61761, UROS shRNA Plasmid (m): sc-61761-SH and UROS shRNA (m) Lentiviral Particles: sc-61761-V.

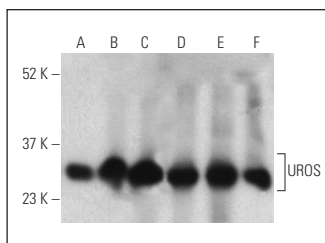
Molecular Weight of UROS: 30 kDa.

Positive Controls: c4 whole cell lysate: sc-364186, mouse liver extract: sc-2256 or mouse thyroid extract: sc-2407.

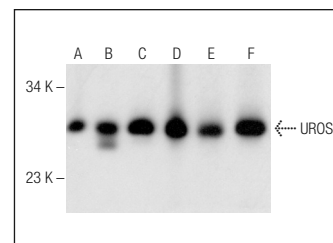
RECOMMENDED SUPPORT REAGENTS

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 (A-8): sc-365877. Western blot analysis of UROS expression in c4 (A) and EOC 20 (B) whole cell lysates and mouse brain (C), mouse liver (D), mouse kidney (E) and mouse thyroid (F) tissue extracts. Detection reagent used: m-IgG₁ BP-HRP: sc-525408.



UROS (A-8): sc-365877. Western blot analysis of UROS expression in EOC 20 whole cell lysate (A) and mouse colon (B), mouse brain (C), mouse kidney (D), mouse liver (E) and mouse thyroid (F) tissue extracts.

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.