SANTA CRUZ BIOTECHNOLOGY, INC.

UROD (G-7): sc-374318

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Uroporphyrinogen decarboxylase, also known as UROD or UPD, is a 367 amino acid protein that exists as a homodimer. UROD is the fifth enzyme in the human heme biosynthetic pathway and is responsible for the conversion of uroporphyrinogen to coproporphyrinogen through the removal of four carboxymethyl side chains. Mutations in the UROD gene are responsible for three autosomal disorders in humans: familial porphyria cutanea tarda (F-PCT), sporadic porphyria cutanea tarda (S-PCT) and hepatoerythropoietic porphyria (HEP). F-PCT is an autosomal dominant disorder characterized by late-onset light-sensitive dermititis. High levels of uroporphyrin excretion in the urine and varying degrees of liver damage are associated with this disease. S-PCT is an idiosyncratic form of PCT that is characterized by a reduction of liver enzymes. HEP is an autosomal recessive disorder that affects infants. It is characterized by excessive excretion of acetate-substituted porphyrins and accumulation of protoporphyrin in erythrocytes.

REFERENCES

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2. Phillips, J.D., et al. 1997. Characterization and crystallization of human uroporphyrinogen decarboxylase. Protein Sci. 6: 1343-1346.
3. Akhtar, R.A., et al. 1998. Chromosomal linkage analysis of porphyria in mice induced by hexachlorobenzene-iron synergism: a model of sporadic porphyria cutanea tarda. Pharmacogenetics 8: 485-494.
4. Christiansen, L., et al. 1999. Screening for mutations in the uroporphyrinogen decarboxylase gene using denaturing gradient gel electrophoresis. Identification and characterization of six novel mutations associated with familial PCT. Hum. Mutat. 14: 222-232.
5. Phillips, J.D., et al. 2001. Functional consequences of naturally occurring mutations in human uroporphyrinogen decarboxylase. Blood 98: 3179-3185.
6. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 176100. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
7. Brancaleoni, V., et al. 2007. Novel human pathological mutations. Gene symbol: UROD. Disease: porphyria, cutaneous. Hum. Genet. 122: 415.
8. Phillips, J.D., et al. 2007. Two novel uroporphyrinogen decarboxylase (UROD) mutations causing hepatoerythropoietic porphyria (HEP). Transl. Res. 149: 85-91.

CHROMOSOMAL LOCATION

Genetic locus: UROD (human) mapping to 1p34.1; Urod (mouse) mapping to 4 D1.

SOURCE

UROD (G-7) is a mouse monoclonal antibody raised against amino acids 81-133 mapping within an internal region of UROD of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

UROD (G-7) is recommended for detection of UROD 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 UROD siRNA (h): sc-88548, UROD siRNA (m): sc-154937, UROD shRNA Plasmid (h): sc-88548-SH, UROD shRNA Plasmid (m): sc-154937-SH, UROD shRNA (h) Lentiviral Particles: sc-88548-V and UROD shRNA (m) Lentiviral Particles: sc-154937-V.

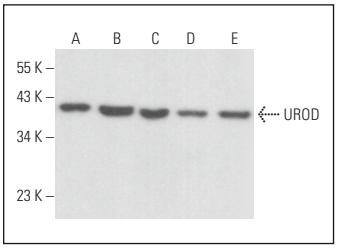
Molecular Weight of UROD: 41 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, M1 whole cell lysate: sc-364782 or U-87 MG cell lysate: sc-2411.

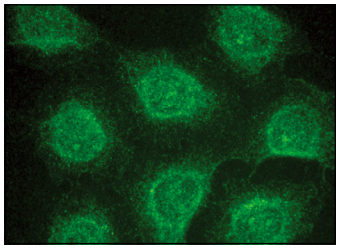
RECOMMENDED SUPPORT REAGENTS

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



UROD (G-7): sc-374318. Western blot analysis of UROD expression in Hep G2 (**A**), U-87 MG (**B**), NIH/3T3 (**C**), M1 (**D**) and L6 (**E**) whole cell lysates.



UROD (G-7): sc-374318. Immunofluorescence staining of methanol-fixed HeLa cells showing nuclear and cytoplasmic 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.