UROD (C-4): sc-365297

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 porphyrinuria cutanea tarda (f-PCT), sporadic porphyrinuria cutanea tarda (s-PCT) and hepatoerythropoietic porphyria (HEP). F-PCT is an autosomal dominant disorder characterized by late-onset light-sensitive dermatitis. High levels of uroporphyrin excretion in the urine and varying degrees of liver damage are associated with this disease. S-PCT is an idiopathic 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


CHROMOSOMAL LOCATION

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

SOURCE

UROD (C-4) is a mouse monoclonal antibody raised against a peptide mapping near the N-terminus of UROD of human origin.

PRODUCT

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

UROD (C-4) is available conjugated to agarose (sc-365297 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-365297 HRP), 200 µg/ml for WB, HRP and ELISA; to either phycoerythrin (sc-365297 PE) or Fluorescein (sc-365297 FITC), Alexa Fluor® 488 or Alexa Fluor® 546; to either Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml); to either m-IgG or m-IgG (sc-516140 or sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-359850. 4) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgG BP-FITC: sc-516140 or m-IgG BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgG BP-HRP: sc-516102 or m-IgG BP-PE: sc-516102 (dilution range 1:50-1:500), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:300).

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.