10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

UROD Rabbit pAb

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Catalog # AP58277

Product Information

Application	WB
Primary Accession	P06132
Reactivity	Human
Predicted	Rat, Dog, Mouse, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	40787
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human UROD
Epitope Specificity	121-220/367
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Cytoplasm.
DISEASE	Defects in UROD are the cause of familial porphyria cutanea tarda (FPCT) [MIM:176100]; also known as porphyria cutanea tarda type II. FPCT is an autosomal dominant disorder characterized by light-sensitive dermatitis, with onset in later life. It is associated with the excretion of large amounts of uroporphyrin in the urine. Iron overload is often present in association with varying degrees of liver damage. Besides the familial form of PCT, a relatively common idiosyncratic form is known in which only the liver enzyme is reduced. This form is referred to as porphyria cutanea tarda "sporadic" type or type I [MIM:176090]. PCT type I occurs sporadically as an unusual accompaniment of common hepatic disorders such as alcohol-associated liver disease. Defects in UROD are the cause of hepatoerythropoietic porphyria (HEP) [MIM:176100]. HEP is a rare autosomal recessive disorder. It is the severe form of cutaneous porphyria, and presents in infancy. The level of UROD is very low in erythrocytes and cultured skin fibroblasts, suggesting that HEP is the homozygous state for porphyria cutanea tarda.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	UROD is the fifth enzyme of the heme biosynthetic pathway. This enzyme is responsible for catalyzing the conversion of uroporphyrinogen to coproporphyrinogen through the removal of four carboxymethyl side chains. Mutations and deficiency in this enzyme are known to cause familial porphyria cutanea tarda and hepatoerythropoetic porphyria. Porphyria cutanea tarda is an autosomal dominant disorder characterized by light-sensitive dermatitis and associated with the excretion of large amounts of uroporphyrin in urine. Hepatoerythropoetic porphyria is a form of porphyria cutanae tarda that may also be a manifestation of benign or malignant hepatic tumors.

Additional Information

Gene ID	7389
Other Names	Uroporphyrinogen decarboxylase, UPD, URO-D, 4.1.1.37, UROD (HGNC:12591)
Dilution	WB=1:500-2000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	UROD (HGNC:12591)
Function	Catalyzes the sequential decarboxylation of the four acetate side chains of uroporphyrinogen to form coproporphyrinogen and participates in the fifth step in the heme biosynthetic pathway (PubMed:11069625, PubMed:11719352, PubMed:14633982, PubMed:18004775, PubMed:21668429). Isomer I or isomer III of uroporphyrinogen may serve as substrate, but only coproporphyrinogen III can ultimately be converted to heme (PubMed:11069625, PubMed:11719352, PubMed:14633982, PubMed:21668429). In vitro also decarboxylates pentacarboxylate porphyrinogen I (PubMed:12071824).
Cellular Location	Cytoplasm, cytosol {ECO:0000250|UniProtKB:P70697}

Background

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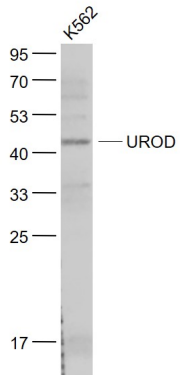
Images



Sample:
K562(Human) Cell Lysate at 30 ug
Primary: Anti- UROD (AP58277) at 1/1000 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 41 kD
Observed band size: 41 kD