SunLong Biotech Co.,LTD
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[Primary Antibody]

Rabbit Anti-CPS1 antibody

Catalog Number: SL14034R

FOR RESEARCH USE ONLY

[DATASHEET]

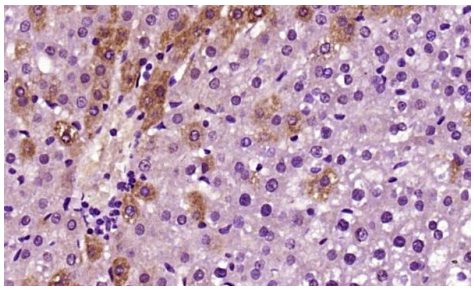
Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	CPS1
Immunogen:	KLH conjugated synthetic peptide derived from human CPS1: 1351-1500/1500
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	IHC-P=1:100-500 IHC-F=1:100-500 IF=1:100-500 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Rat(predicted:Human,Mouse,Dog,Sheep)
Predicted MW.:	160kDa
Subcellular Location:	The nucleus cytoplasmic Mitochondrion
Background:	The mitochondrial enzyme encoded by this gene catalyzes synthesis of carbamoyl phosphate from ammonia and bicarbonate. This reaction is the first committed step of the urea cycle, which is important in the removal of excess urea from cells. The encoded protein may also represent a core mitochondrial nucleoid protein. Three transcript variants encoding different isoforms have been found for this gene. The shortest isoform may not be localized to the mitochondrion. Mutations in this gene have been associated with carbamoyl phosphate synthetase deficiency, susceptibility to persistent pulmonary hypertension, and susceptibility to venoocclusive disease after bone marrow transplantation.[provided by RefSeq, May 2010] **Function:** Involved in the urea cycle of ureotelic animals where the enzyme plays an important role in removing excess ammonia from the cell.

Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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	Subcellular Location: Mitochondrion. **Tissue Specificity:** Primarily in the liver and small intestine. DISEASE: Defects in CPS1 are the cause of carbamoyl phosphate synthetase 1 deficiency (CPS1D) [MIM:237300]. CPS1D is an autosomal recessive disorder of the urea cycle causing hyperammonemia. Clinical features include protein intolerance, intermittent ataxia, seizures, lethargy, developmental delay and mental retardation. Note=Genetic variations in CPS1 influence the availability of precursors for nitric oxide (NO) synthesis and play a role in clinical situations where endogenous NO production is critically important, such as neonatal pulmonary hypertension, increased pulmonary artery pressure following surgical repair of congenital heart defects or hepatovenocclusive disease following bone marrow transplantation. Infants with neonatal pulmonary hypertension homozygous for Thr-1406 have lower L-arginine concentrations than neonates homozygous for Asn-1406. **Similarity:** Contains 2 ATP-grasp domains. Contains 1 glutamine amidotransferase type-1 domain. **Database links:** **Entrez Gene:** 1373 Human Omim: 608307 Human **SwissProt:** P31327 Human **Unigene:** 149252 Human

[VALIDATION IMAGES]



Paraformaldehyde-fixed, paraffin embedded (Rat liver); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (CPS1) Polyclonal Antibody, Unconjugated (SL14034R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

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