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CPS1 Recombinant Rabbit mAb

CPS1 Recombinant Rabbit mAb
Catalog # AP94494

Product Information

Application	WB, IHC-P, IHC-F, IF
Host	Rabbit
Clonality	Recombinant
Physical State	Liquid
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	Mitochondrion.
SIMILARITY	Contains 2 ATP-grasp domains. Contains 1 glutamine amidotransferase type-1 domain.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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]

Additional Information

Target/Specificity	Primarily in the liver and small intestine.
Dilution	WB=1:500-1:1000,IHC-P=1:100-500,IHC-F=1:100-500,IF=0

Format	0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce
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.

Background

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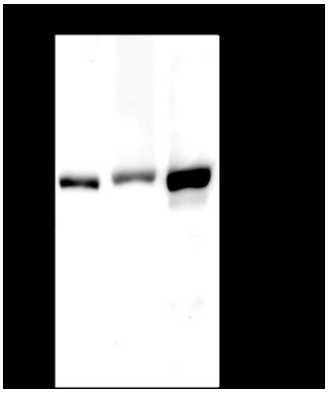
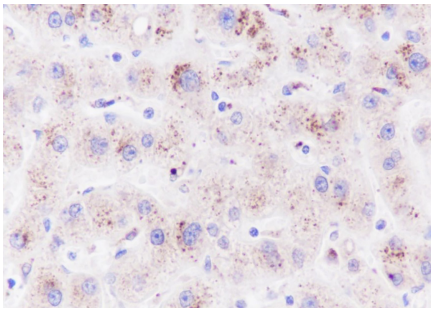
Images



Blocking buffer: 5% NFDM/TBST Primary ab dilution: 1:1000 Primary ab incubation condition: 2 hours at room temperature Secondary ab: Goat Anti-Rabbit IgG H&L (HRP) Lysate: 1: Human liver, 2: Rat liver, 3: Mouse liver Protein loading quantity: 20 µg Exposure time: 60 s Predicted MW: 165 kDa Observed MW: 165 kDa



Tissue: Human liver cancer Section type: Formalin fixed & Paraffin -embedded section Retrieval method: High temperature and high pressure Retrieval buffer: Tris/EDTA buffer, pH 9.0 Primary ab dilution: 1:100 Primary ab incubation condition: 1 hour at room temperature Secondary ab: SP Kit(Rabbit) (sp-0023) Counter stain: Hematoxylin (Blue) Comment: Color brown is the positive signal for AP94494