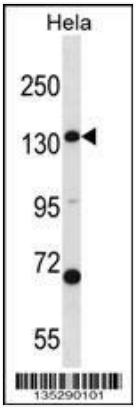
CPS1 Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	CPS1, Polyclonal Antibody
Full Product Name	CPS1, NT (CPS1, Carbamoyl-phosphate synthase [ammonia], mitochondrial, Carbamoyl-phosphate synthetase I) (AP)
Product Gene Name	anti-CPS1 antibody
Product Synonym Names	CPS1; Carbamoyl-phosphate synthase [ammonia], mitochondrial; Carbamoyl-phosphate synthetase I
Clonality	Polyclonal
Isotype	IgG
Host	Rabbit
Species Reactivity	Human
Form/Format	Supplied as a liquid in PBS, pH 7.2. No preservative added. Labeled with Alkaline Phosphatase (AP).
Purity/Purification	Purified by Protein A Affinity Chromatography.
Preparation and Storage	Store product at 4 degree C. DO NOT FREEZE! Stable at 4 degree C for 12 months after receipt as an undiluted liquid. Dilute required amount only prior to immediate use. Further dilutions can be made in assay buffer. For maximum recovery of product, centrifuge the original vial prior to removing the cap.
Molecular Weight	~165kDa
OMIM	gene 615371
3D Structure	ModBase 3D Structure for P31327
Immunogen	CPS1 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 261-291 amino acids from the N-terminal region of human CPS1.
Conjugate	AP
Note	Preservative Free
Product Categories/Family for anti-CPS1 antibody	Antibodies; Mitochondrial Proteins
Applications Tested/Suitable for anti-CPS1 antibody	Western Blot (WB), ELISA (EIA)
Application Notes for anti-CPS1 antibody	WB: 1:100-500 ELISA: 1:1,000 Applications are based on unconjugated antibody.
Related Product Information for anti-CPS1 antibody	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.
Product References and Citations for anti-CPS1 antibody	Bailey, S.D., et al. Diabetes Care 33(10):2250-2253(2010). Jia, P., et al. Schizophr. Res. 122 (1-3), 38-42 (2010): Pekkala, S., et al. Hum. Mutat. 31(7):801-808(2010). Huo, R., et al. J. Biochem. Mol. Biol. 38(1):28-33(2005). Hoshide, R., et al. Genomics 28(1):124-125(1995).

Other Notes	Small volumes of anti-CPS1 antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.
Searchable Terms for CPS1 purchase	MBS6288395 is an antibody targeting CPS1. Also known as CPS1, NT (CPS1, Carbamoyl-phosphate synthase [ammonia], mitochondrial, Carbamoyl-phosphate synthetase I) (AP). Reactive in Human. Host: Rabbit.



Western Blot (WB) of anti-CPS1 antibody
CPS1 Antibody (N-term) western blot analysis in Hela cell line lysates (35ug/lane).This demonstrates the CPS1 antibody detected the CPS1 protein (arrow).

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for CPS1. It may not necessarily be applicable to this product.

NCBI GI #	169790915
NCBI GeneID	1373
NCBI Accession #	NP_001116105.1
NCBI Related Accession #	NP_001866.2; NP_001116106.1
UniProt Primary Accession #	P31327
UniProt Secondary Accession #	O43774; Q53TL5; Q59HF8; Q7Z5I5; B7Z818; J3KQL0
UniProt Related Accession #	P31327
GenBank Nucleotide #	NM_001122633.2
NCBI Official Full Name	carbamoyl-phosphate synthase
NCBI Official Synonym Full Names	carbamoyl-phosphate synthase 1
NCBI Official Symbol	CPS1
NCBI Official Synonym Symbols	PHN; CPSASE1
UniProt Protein Name	Carbamoyl-phosphate synthase [ammonia], mitochondrial
UniProt Synonym Protein Names	Carbamoyl-phosphate synthetase I; CPSase I
Protein Family	Carboxypeptidase
UniProt Gene Name	CPS1
UniProt Synonym Gene Names	CPSase I
UniProt Entry Name	CPSM_HUMAN
NCBI Protein Information	carbamoyl-phosphate synthase [ammonia], mitochondrial

UniProt Comments for CPS1	CPS1: Involved in the urea cycle of ureotelic animals where the enzyme plays an important role in removing excess ammonia from the cell. Defects in CPS1 are the cause of carbamoyl phosphate synthetase 1 deficiency (CPS1D). 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. 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. 2 isoforms of the human protein are produced by alternative splicing. Protein type: Amino Acid Metabolism - arginine and proline; Mitochondrial; Nucleolus; Ligase; EC 6.3.4.16; Energy Metabolism - nitrogen; Amino Acid Metabolism - alanine, aspartate and glutamate Chromosomal Location of Human Ortholog: 2q35 Cellular Component: protein complex; mitochondrial matrix; mitochondrial inner membrane; nucleolus Molecular Function: carbamoyl-phosphate synthase (ammonia) activity; protein binding; glutamate binding; carbamoyl-phosphate synthase (glutamine-hydrolyzing) activity; endopeptidase activity; protein complex binding; phospholipid binding; calcium ion binding; ATP binding Biological Process: response to food; response to drug; glycogen catabolic process; arginine biosynthetic process; response to toxin; response to lipopolysaccharide; homocysteine metabolic process; positive regulation of vasodilation; proteolysis; response to amino acid stimulus; nitric oxide metabolic process; response to starvation; citrulline biosynthetic process; response to zinc ion; triacylglycerol catabolic process; glutamine catabolic process; midgut development; anion homeostasis; response to amine stimulus; urea cycle Disease: Pulmonary Hypertension, Neonatal, Susceptibility To; Carbamoyl Phosphate Synthetase I Deficiency, Hyperammonemia Due To
Research Articles on CPS1	**1.** These results suggest that glycine metabolism and/or the urea cycle represent potentially novel sex-specific mechanisms for the development of atherosclerosis.
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