PRODUCT INFORMATION

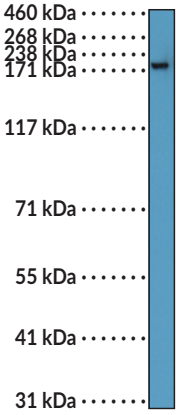
CPS1 (N-Term) Rabbit Monoclonal Antibody (RM395)

Item No. 32320

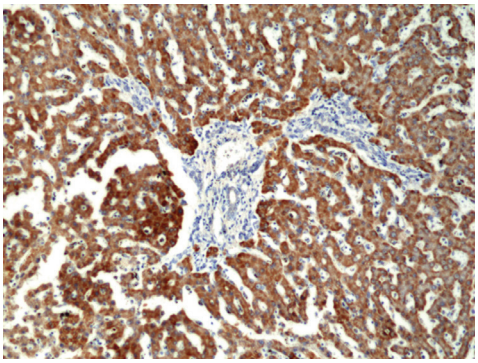
Overview and Properties

Contents:	This vial contains 100 µl of protein A-affinity purified monoclonal antibody.
Synonyms:	Carbamoyl Phosphate Synthase, mitochondrial, CPSase I
Immunogen:	Peptide from the N-terminal region of CPS1
Cross Reactivity:	(+) CPS1
Species Reactivity:	(+) Human
Form:	Liquid
Storage:	-20°C (as supplied)
Stability:	≥1 year
Storage Buffer:	PBS, with 50% glycerol, 1% BSA, and 0.09% sodium azide
Clone:	RM395
Host:	Rabbit
Isotype:	IgG
Applications:	Immunohistochemistry (IHC) and Western blot (WB); the recommended starting dilution is 1:100-1:200 for IHC and 1:5,000-1:10,000 for WB. Other applications were not tested, therefore optimal working concentration/dilution should be determined empirically.

Images



WB of human T cell lysate using CPS1 (N-Term) Rabbit Monoclonal Antibody (RM395) at a dilution of 1:10,000.



Immunohistochemical staining of formalin-fixed and paraffin-embedded human liver tissue using CPS1 (N-Term) Rabbit Monoclonal Antibody (RM395) at a dilution of 1:100.

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Description

Carbamoyl phosphate synthetase I (CPS1) is a ligase involved in the urea cycle.[1] It is composed of an N-terminal domain containing an ancestral inactive glutaminase, a bicarbonate phosphorylation site, and a C-terminal domain containing a carbamate phosphorylation site and the binding site for the CPS1 allosteric activator N-acetyl-L-glutamate (NAG). It is primarily expressed in the liver and intestinal epithelial cells and is localized to the mitochondrial matrix.[1,2] CPS1, with NAG as a cofactor, catalyzes the formation of carbamoyl phosphate from ammonia, bicarbonate, and ATP as the first, rate-limiting step in the urea cycle. Mutations in *CPS1* result in CPS1 deficiency, a disorder characterized by low levels of active CPS1, arginine, and citrulline and high levels of ammonia and glutamine, leading to neurological deficits and, potentially, death.[3,4] Protein levels of CPS1 are increased in patient-derived rectal cancer tumors and associated with shorter disease-specific survival and metastasis-free survival.[5] Cayman's CPS1 (N-Term) Rabbit Monoclonal Antibody (RM395) can be used for immunohistochemistry (IHC) and Western blot (WB) applications.

References

1. Martínez, A.I., Pérez-Arellano, I., Pekkala, S., *et al*. Genetic, structural and biochemical basis of carbamoyl phosphate synthetase 1 deficiency. *Mol. Genet. Metab*. **101(4)**, 311-323 (2010).
2. Ryall, J.G., Nguyen, M., Bendayan, M., *et al*. Expression of nuclear genes encoding the urea cycle enzymes, carbamoyl-phosphate synthetase I and ornithine carbamoyl transferase, in rat liver and intestinal mucosa. *Eur. J. Biochem*. **152(2)**, 287-292 (1985).
3. Nitzahn, M. and Lipshutz, G.S. CPS1: Looking at an ancient enzyme in a modern light. *Mol. Genet. Metab*. **131(3)**, 289-298 (2020).
4. Pekkala, S., Martínez, A.I., Barcelona, B., *et al*. Understanding carbamoyl-phosphate synthetase I (CPS1) deficiency by using expression studies and structure-based analysis. *Hum. Mutat*. **31(7)**, 801-808 (2010).
5. Lee, Y.-Y., Li, C.-F., Lin, C.-Y., *et al*. Overexpression of CPS1 is an independent negative prognosticator in rectal cancers receiving concurrent chemoradiotherapy. *Tumour Biol.* **35(11)**, 11097-11105 (2014).

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