

### Anti-CPS1 antibody [2E4AE11] ab110303

★★★★★ [1 Abreviews](#) [2 References](#) [4 Images](#)

#### Overview

<b>Product name</b>	Anti-CPS1 antibody [2E4AE11]
<b>Description</b>	Mouse monoclonal [2E4AE11] to CPS1
<b>Host species</b>	Mouse
<b>Tested applications</b>	<b>Suitable for:</b> IHC-P, Flow Cyt, ICC, IP
<b>Species reactivity</b>	<b>Reacts with:</b> Human
<b>Immunogen</b>	Tissue, cells or virus. This information is proprietary to Abcam and/or its suppliers.
<b>Positive control</b>	HDFn and HeLa cells; Human liver mitochondria lysate.
<b>General notes</b>	<p>This antibody clone is manufactured by Abcam. If you require a custom buffer formulation or conjugation for your experiments, please contact <a href="mailto:orders@abcam.com">orders@abcam.com</a>.</p> <p>The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing.</p> <p>If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&amp;As</p> <p>Product was previously marketed under the MitoSciences sub-brand.</p>

#### Properties

<b>Form</b>	Liquid
<b>Storage instructions</b>	Shipped at 4°C. Store at +4°C. Do Not Freeze.
<b>Storage buffer</b>	<p>pH: 7.5</p> <p>Preservative: 0.02% Sodium azide</p> <p>Constituent: HEPES buffered saline</p>
<b>Purity</b>	Affinity purified
<b>Purification notes</b>	ab110303 was produced in vitro using hybridomas grown in serum-free medium. Purity: >95% by SDS-PAGE.
<b>Clonality</b>	Monoclonal

Clone number	2E4AE11
Isotype	IgG1
Light chain type	kappa

## Applications

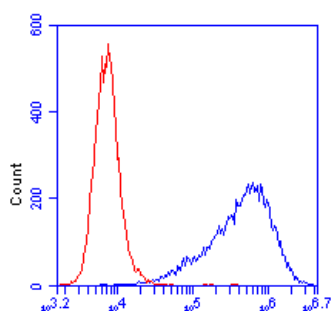
**The Abpromise guarantee** Our **Abpromise guarantee** covers the use of ab110303 in the following tested applications. The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
IHC-P		Use a concentration of 10 µg/ml. Perform heat mediated antigen retrieval before commencing with IHC staining protocol.
Flow Cyt		Use a concentration of 1 µg/ml. <b>ab170190</b> - Mouse monoclonal IgG1, is suitable for use as an isotype control with this antibody.
ICC		Use a concentration of 10 µg/ml.
IP		Use at an assay dependent concentration.

## Target

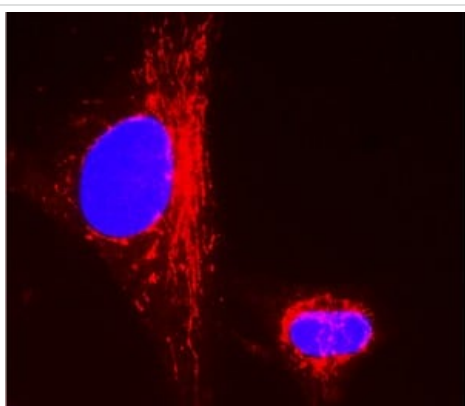
<b>Function</b>	Involved in the urea cycle of ureotelic animals where the enzyme plays an important role in removing excess ammonia from the cell.
<b>Tissue specificity</b>	Primarily in the liver and small intestine.
<b>Involvement in disease</b>	Defects in CPS1 are the cause of carbamoyl phosphate synthetase 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.
<b>Sequence similarities</b>	Contains 2 ATP-grasp domains. Contains 1 glutamine amidotransferase type-1 domain.
<b>Domain</b>	The type-1 glutamine amidotransferase domain is defective.
<b>Cellular localization</b>	Mitochondrion.

## Images



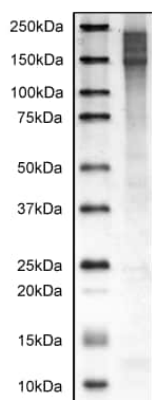
Flow Cytometry - Anti-CPS1 antibody [2E4AE11]  
(ab110303)

Flow cytometric analysis using ab110303 at 1µg/ml staining CPS1 in HeLa cells (blue). Isotype control antibody (red).



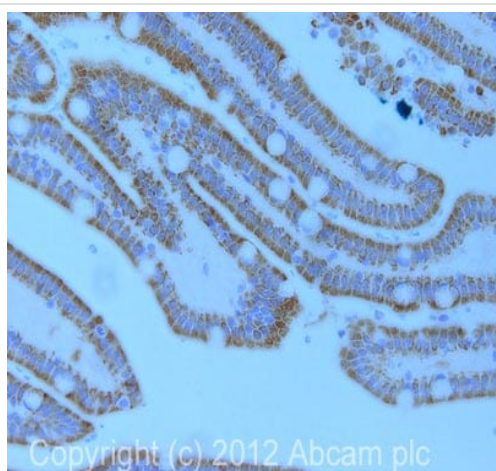
Immunocytochemistry - Anti-CPS1 antibody  
[2E4AE11] (ab110303)

Immunocytochemistry analysis using ab110303 at 10µg/ml staining CPS1 in HDFn cells (paraformaldehyde fixed and Triton X-100 permeabilized). The secondary antibody was (red) Alexa Fluor® 594 goat anti-mouse IgG (H+L) used at a 1/1000 dilution for 1h. 10% Goat serum was used as the blocking agent for all blocking steps. DAPI was used to stain the cell nuclei (blue). Target protein locates mainly in mitochondria.



Immunoprecipitation - Anti-CPS1 antibody  
[2E4AE11] (ab110303)

Detection of ab110303 by immunoprecipitation staining of 164kDa CPS1, a liver mitochondrial marker in human liver mitochondrial lysate.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-CPS1 antibody [2E4AE11] (ab110303)

IHC image of ab110303 staining in human duodenum formalin fixed paraffin embedded tissue section, performed on a Leica Bond™ system using the standard protocol F. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH6, epitope retrieval solution 1) for 20 mins. The section was then incubated with ab110303, 10µg/ml, for 15 mins at room temperature and detected using an HRP conjugated compact polymer system. DAB was used as the chromogen. The section was then counterstained with haematoxylin and mounted with DPX.

For other IHC staining systems (automated and non-automated) customers should optimize variable parameters such as antigen retrieval conditions, primary antibody concentration and antibody incubation times.

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