

# Product Datasheet

## PMPCA Antibody - BSA Free

### NBP1-89126

Unit Size: 0.1 ml

Store at 4C short term. Aliquot and store at -20C long term. Avoid freeze-thaw cycles.

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**NBP1-89126**

PMPCA Antibody - BSA Free

**Product Information**

<b>Unit Size</b>	0.1 ml
<b>Concentration</b>	Concentrations vary lot to lot. See vial label for concentration. If unlisted please contact technical services.
<b>Storage</b>	Store at 4C short term. Aliquot and store at -20C long term. Avoid freeze-thaw cycles.
<b>Clonality</b>	Polyclonal
<b>Preservative</b>	0.02% Sodium Azide
<b>Isotype</b>	IgG
<b>Purity</b>	Immunogen affinity purified
<b>Buffer</b>	PBS (pH 7.2) and 40% Glycerol

**Product Description**

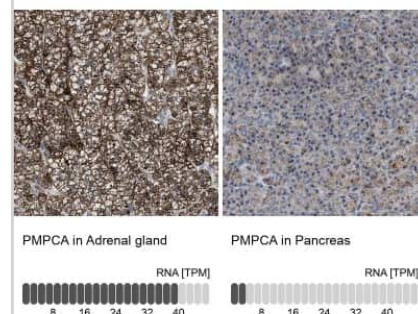
<b>Host</b>	Rabbit
<b>Gene ID</b>	23203
<b>Gene Symbol</b>	PMPCA
<b>Species</b>	Human
<b>Reactivity Notes</b>	Immunogen displays the following percentage of sequence identity for non-tested species: Mouse (89%)
<b>Immunogen</b>	This antibody was developed against Recombinant Protein corresponding to amino acids: VEHEHLVDCARKYLLGVQPAWGSAEAVDIDRSVAQYTGGIAKLERDMSNVSLG PTPIPELTHIMVGLSCSFLEEDFIPFAVLNMMMGGGGSFSAGGPGKGMFSR

**Product Application Details**

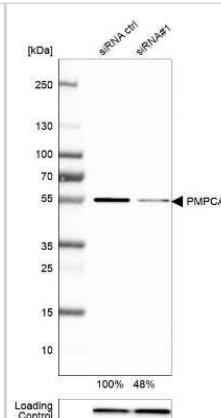
<b>Applications</b>	Western Blot, Immunocytochemistry/ Immunofluorescence, Immunohistochemistry, Immunohistochemistry-Paraffin, Knockdown Validated
<b>Recommended Dilutions</b>	Western Blot 0.04-0.4 ug/ml, Immunohistochemistry 1:50 - 1:200, Immunocytochemistry/ Immunofluorescence 0.25-2 ug/ml, Immunohistochemistry-Paraffin 1:50 - 1:200, Knockdown Validated
<b>Application Notes</b>	For IHC-Paraffin, HIER pH 6 retrieval is recommended. ICC/IF Fixation Permeabilization: Use PFA/Triton X-100.

**Images**

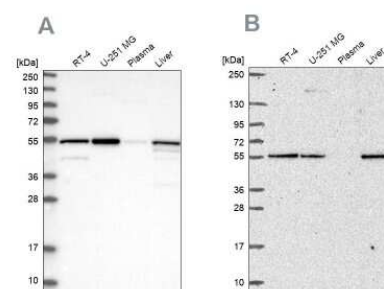
Immunohistochemistry-Paraffin: PMPCA Antibody [NBP1-89126] - Staining in human adrenal gland and pancreas tissues using anti-PMPCA antibody. Corresponding PMPCA RNA-seq data are presented for the same tissues.



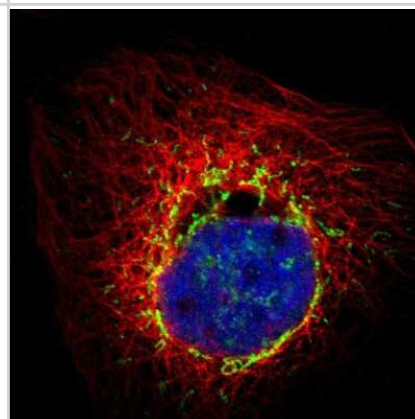
Western Blot: PMPCA Antibody [NBP1-89126] - Analysis in HEK293 cells transfected with control siRNA, target specific siRNA probe #1, using Anti-PMPCA antibody. Remaining relative intensity is presented. Loading control: Anti-PPIB.



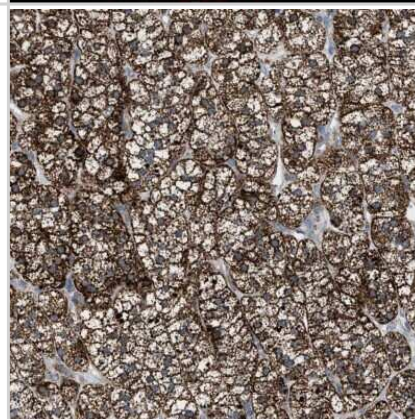
Western Blot: PMPCA Antibody [NBP1-89126] - Analysis using Anti-PMPCA antibody NBP1-89126 (A) shows similar pattern to independent antibody NBP2-57169 (B).



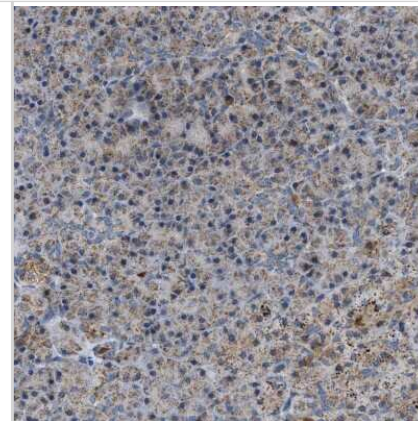
Immunocytochemistry/Immunofluorescence: PMPCA Antibody [NBP1-89126] - Immunofluorescent staining of human cell line A-431 shows localization to mitochondria. Antibody staining is shown in green.



Immunohistochemistry-Paraffin: PMPCA Antibody [NBP1-89126] - Staining of human adrenal gland shows high expression.



Immunohistochemistry-Paraffin: PMPCA Antibody [NBP1-89126] - Staining of human pancreas shows low expression as expected.



## Publications

Vibhuti Rambani, Miriam Kolnikova, Michal Cagalinec, Martina Skopkova, Daniela Gasperikova PMPCA-Related Encephalopathy Neurology: Genetics 2023-12-01 [PMID: 38235041]

Charif M, Chevrollier A, Gueguen N et al. Next-Generation Sequencing Identifies Novel PMPCA Variants in Patients with Late-Onset Dominant Optic Atrophy Genes 2022-07-05 [PMID: 35885985]

Joshi M, Anselm I, Shi J, Bale T et al. Mutations in the substrate binding glycine-rich loop of the mitochondrial processing peptidase-alpha protein (PMPCA) cause a severe mitochondrial disease Molecular Case Studies 1905-07-08 [PMID: 27148589] (WB, ICC/IF, Human)

Jobling RK, Assoum M, Gakh O et al. PMPCA mutations cause abnormal mitochondrial protein processing in patients with non-progressive cerebellar ataxia. Brain. 2015-03-25 [PMID: 25808372] (ICC/IF, Human)



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### **Products Related to NBP1-89126**

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NBP1-89126PEP	PMPCA Recombinant Protein Antigen
HAF008	Goat anti-Rabbit IgG Secondary Antibody [HRP]
NB7160	Goat anti-Rabbit IgG (H+L) Secondary Antibody [HRP]
NBP2-24891	Rabbit IgG Isotype Control

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### **Limitations**

This product is for research use only and is not approved for use in humans or in clinical diagnosis. Primary Antibodies are guaranteed for 1 year from date of receipt.

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