

# Human PRPS1 antibody

Catalog Number: ATGA0279

## PRODUCT INFORMATION

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**Catalog number**

ATGA0279

**Clone No.**

AT1E11

**Product type**

Monoclonal Antibody

**UnitProt No.**

P60891

**NCBI Accession No.**

NP\_002755

**Alternative Names**

Ribose-phosphate pyrophosphokinase 1, ARTS, CMTX5, PPRibP, PRSI, DFN2, DFNX1, Ribose-phosphate pyrophosphokinase 1 EC 2.7.6.1, KIAA0967, Phosphoribosyl pyrophosphate synthetase I, PRS I, PRS-I, Ribose phosphate pyrophosphokinase I

## PRODUCT SPECIFICATION

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**Antibody Host**

Mouse

**Reacts With**

Human

**Concentration**

1mg/ml (determined by BCA assay)

**Formulation**

Liquid in. Phosphate-Buffered Saline (pH 7.4) with 0.02% Sodium Azide, 10% glycerol

**Immunogen**

Recombinant human PRPS1 (1-318aa) purified from E. coli

**Isotype**

IgG2a kappa

**Purification Note**

By protein-A affinity chromatography

**Application**

ELISA, WB, ICC/IF

**Usage**

The antibody has been tested by ELISA, Western blot and ICC/IF analysis to assure specificity and reactivity. Since application varies, however, each investigation should be titrated by the reagent to obtain optimal results.

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## Storage

Can be stored at +2C to +8C for 1 week. For long term storage, aliquot and store at -20C to -80C. Avoid repeated freezing and thawing cycles.

## BACKGROUND

### Description

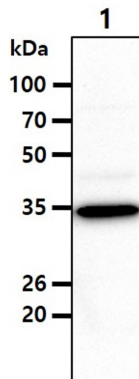
PRPS1 is an enzyme that catalyzes the phosphoribosylation of ribose 5-phosphate to 5-phosphoribosyl-1-pyrophosphate, which is necessary for purine metabolism and nucleotide biosynthesis. A mutation in PRPS1 may result in PRPS superactivity, a disease characterized by gout and the overproduction of purine nucleotides, uric acid and PRPP. PRPS1 mutations can also lead to a reduction in PRPS1 activity resulting in ARTS syndrome or CMTX5 (Charcot-Marie-Tooth disease X-linked recessive type 5).

### General References

Ishizuka T, et al. (1992) *Biochim Biophys Acta* 241:130(2): 139-48.  
Kim HJ, et al. (2007) *Am J Hum Genet* 81(3):552-8.

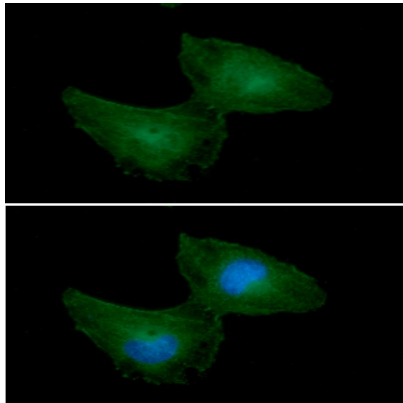
## DATA

### Western blot analysis (WB)



The cell lysate (40ug) was resolved by SDS-PAGE, transferred to PVDF membrane and probed with anti-human PRPS1 antibody (1:1000). Proteins were visualized using a goat anti-mouse secondary antibody conjugated to HRP and an ECL detection system.  
Lane 1.: 293T cell lysate

### Immunocytochemistry/Immunofluorescence (ICC/IF)



ICC/IF analysis of PRPS1 in HeLa cells. The cell was stained with ATGA0279 (1:100). The secondary antibody (green) was used Alexa Fluor 488. DAPI was stained the cell nucleus (blue).