# **PRODUCT INFORMATION**

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**

### **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.



#### 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

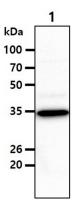
PRPS1 is an enzyme that catalyzes the phosphoribosylation of ribose 5-phosphate to 5-phosphoribosyl-1pyrophosphate, which is necessary for purine metabolism and nucleotide biosynthesis. A mutation in PRPS1 may result in PRPS superactivity, a disease characterized by gout and the overproduction of purine 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 Biophsy Acta 241130(2): 139-48. Kim HJ, et al. (2007) Arn J Hum Gernet 81(3):552-8.

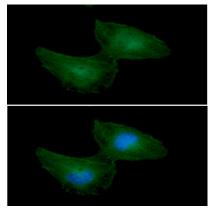
### DATA

#### 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).

