# abcam

## Product datasheet

## Recombinant human AK2 protein ab167993

## 1 Image

**Description** 

Product name Recombinant human AK2 protein

Biological activity: > 1.5 units/ml. One unit will convert 2.0 µmoles of ADP to ATP + AMP per minute

at pH 7.5 at 25°C.

**Purity** > 95 % SDS-PAGE.

**Endotoxin level** < 1.000 Eu/µg

Expression system Escherichia coli

Accession P54819

Protein length Full length protein

Animal free No

**Nature** Recombinant

**Species** Human

Sequence MGSSHHHHHH SSGLVPRGSH MAPSVPAAEP

EYPKGIRAVL LGPPGAGKGT QAPRLAENFC VCHLATGDML RAMVASGSEL GKKLKATMDA GKLVSDEMVV ELIEKNLETP LCKNGFLLDG

FPRTVRQAEM LDDLMEKRKE KLDSVIEFSI PDSLLIRRIT

GRLIHPKSGR SYHEEFNPPK EPMKDDITGE

PLIRRSDDNE KALKIRLQAY HTQTTPLIEY YRKRGIHSAI

DASQTPDVVF ASILAAFSKA TCKDLVMFI

Predicted molecular weight 29 kDa including tags

Amino acids 1 to 239

Tags His tag N-Terminus

## **Specifications**

Our **Abpromise guarantee** covers the use of **ab167993** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

**Applications** SDS-PAGE

**Functional Studies** 

Form Liquid

1

#### **Additional notes**

This product is manufactured by BioVision, an Abcam company and was previously called 6386 Human Recombinant AK2. 6386-100 is the same size as the 100 µg size of ab167993.

#### **Preparation and Storage**

## Stability and Storage

Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.

pH: 7.50

Constituents: 0.08% DTT, 0.32% Tris HCI, 20% Glycerol

This product is an active protein and may elicit a biological response in vivo, handle with caution.

#### **General Info**

#### **Function**

Catalyzes the reversible transfer of the terminal phosphate group between ATP and AMP. This small ubiquitous enzyme involved in energy metabolism and nucleotide synthesis that is essential for maintenance and cell growth. Plays a key role in hematopoiesis.

#### **Tissue specificity**

Present in most tissues. Present at high level in heart, liver and kidney, and at low level in brain, skeletal muscle and skin. Present in thrombocytes but not in erythrocytes, which lack mitochondria. Present in all nucleated cell populations from blood, while AK1 is mostly absent. In spleen and lymph nodes, mononuclear cells lack AK1, whereas AK2 is readily detectable. These results indicate that leukocytes may be susceptible to defects caused by the lack of AK2, as they do not express AK1 in sufficient amounts to compensate for the AK2 functional deficits (at protein level).

### Involvement in disease

Defects in AK2 are the cause of reticular dysgenesis (RDYS) [MIM:267500]; also known as aleukocytosis. RDYS is the most severe form of inborn severe combined immunodeficiencies (SCID) and is characterized by absence of granulocytes and almost complete deficiency of lymphocytes in peripheral blood, hypoplasia of the thymus and secondary lymphoid organs, and lack of innate and adaptive humoral and cellular immune functions, leading to fatal septicemia within days after birth. In bone marrow of individuals with reticular dysgenesis, myeloid differentiation is blocked at the promyelocytic stage, whereas erythro- and megakaryocytic maturation is generally normal.In addition, affected newborns have bilateral sensorineural deafness. Defects may be due to its absence in leukocytes and inner ear, in which its absence can not be compensated by AK1.

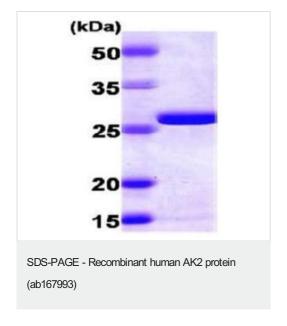
#### Sequence similarities

Belongs to the adenylate kinase family. AK2 subfamily.

#### **Cellular localization**

Mitochondrion intermembrane space.

## **Images**



15% SDS-PAGE analysis of ab167993 (3 µg).

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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