

Product datasheet

Recombinant Human AK2 protein ab78832

画像数 1

製品の概要

製品名	Recombinant Human AK2 protein
タンパク質長	Full length protein

製品の詳細

由来	Recombinant
由来	Escherichia coli

アミノ酸配列

生物種	Human
配列	MGSSHHHHHH SSGLVPRGSH MAPSVAAEP EYPKGIRAVL LGPPGAGKGT QAPRLAENFC VCHLATGDML RAMVASGSEL GKCLKATMDA GKLVSDMVV ELIEKNLETP LCKNGFLLDG FPRTVRQAEM LDDLMEKRKE KLDSVIEFSI PDSLLIRRIT GRLIHPKSGR SYHEEFNPPK EPMKDDITGE PLIRRSDDNE KALKIRLQAY HTQTTPLIEY YRKRGIHSAI DASQTPDVVF ASILAAFSKA TCKDLVMFI

特性

Our [Abpromise guarantee](#) covers the use of **ab78832** in the following tested applications.

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

アプリケーション	SDS-PAGE
精製度	> 95 % SDS-PAGE. ab78832 is purified using conventional chromatography techniques. Endotoxin Level: < 1.0 EU per 1ug of protein (determined by LAL method)
製品の状態	Liquid

前処理および保存

保存方法および安定性	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
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Preservative: None

Constituents: 20% Glycerol, 5mM DTT, 20mM Tris, pH 7.5

関連情報

機能

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.

組織特異性

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).

関連疾患

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.

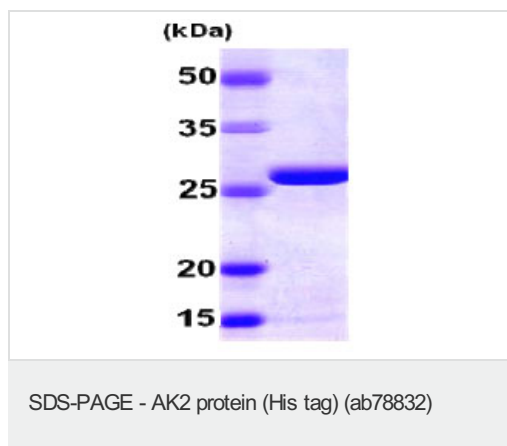
配列類似性

Belongs to the adenylate kinase family. AK2 subfamily.

細胞内局在

Mitochondrion intermembrane space.

画像



15% SDS-PAGE showing ab78832 at approximately 29kDa (3µg).

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