

Product datasheet

Recombinant Human PRPS1 protein ab88987

概述

产品名称	重组人PRPS1蛋白
蛋白长度	Full length protein

描述

性质	Recombinant
来源	Saccharomyces cerevisiae

氨基酸序列

种属	Human
序列	<p>MPNIKIFSGSSHQDLSQKIADRLGLELGKVVTKKFSNQETCVEIGESVRG EDVYIVQSGCGEINDNLM ELLIMINACKIASASRVTAVIPCFPYARQD KKDKSRAPISAKLVANMLSVAGADHIITMDLHASQIQG FFDIPVDNLY AEPAVLKWIRENISEWRNCTIVSPDAGGAKRVTSIADRLNVDFALIHKER KKANEVDR MVLVGDVKDRVAILVDDMADTCGTICHAADKLLSAGATRV YAILTHGIFSGPAISRINNACFEAVVVT NTIPQEDKMKHCSKIQVIDI SMILAEAIRRTHNGESVSYLFSHVPL</p>

技术指标

Our [Abpromise guarantee](#) covers the use of **ab88987** in the following tested applications.

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

应用	SDS-PAGE
纯度	> 90 % SDS-PAGE. ab88987 was purified using affinity chromatography
形式	Liquid

制备和贮存

稳定性和存储	Shipped at 4°C. Store at -20°C. Stable for 12 months at -20°C.
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Preservative: None

Constituents: 30% Glycerol, 0.5% Triton-X-100, 50mM HEPES, 30mM Glutathione, 100mM Sodium chloride, 1mM DTT, pH 7.5

常规信息

功能	Catalyzes the synthesis of phosphoribosylpyrophosphate (PRPP) that is essential for nucleotide synthesis.
通路	Metabolic intermediate biosynthesis; 5-phospho-alpha-D-ribose 1-diphosphate biosynthesis; 5-phospho-alpha-D-ribose 1-diphosphate from D-ribose 5-phosphate (route I); step 1/1.
疾病相关	<p>Defects in PRPS1 are the cause of phosphoribosylpyrophosphate synthetase superactivity (PRPS1 superactivity) [MIM:300661]; also known as PRPS-related gout. It is a familial disorder characterized by excessive purine production, gout and uric acid urolithiasis.</p> <p>Defects in PRPS1 are the cause of Charcot-Marie-Tooth disease X-linked recessive type 5 (CMTX5) [MIM:311070]; also known as optic atrophy-polyneuropathy-deafness or Rosenberg-Chutorian syndrome. CMTX5 is a form of Charcot-Marie-Tooth disease, the most common inherited disorder of the peripheral nervous system. Charcot-Marie-Tooth disease is classified in two main groups on the basis of electrophysiologic properties and histopathology: primary peripheral demyelinating neuropathies characterized by severely reduced motor nerve conduction velocities (NCVs) (less than 38m/s) and segmental demyelination and remyelination, and primary peripheral axonal neuropathies characterized by normal or mildly reduced NCVs and chronic axonal degeneration and regeneration on nerve biopsy.</p> <p>Defects in PRPS1 are the cause of ARTS syndrome (ARTS) [MIM:301835]; also known as fatal ataxia X-linked with deafness and loss of vision. ARTS is a disorder characterized by mental retardation, early-onset hypotonia, ataxia, delayed motor development, hearing impairment, and optic atrophy. Susceptibility to infections, especially of the upper respiratory tract, can result in early death.</p> <p>Defects in PRPS1 are the cause of deafness X-linked type 1 (DFNX1) [MIM:304500]; also known as congenital sensorineural deafness X-linked 2 (DFN2). It is a form of deafness characterized by progressive, severe-to-profound sensorineural hearing loss in males. Females manifest mild to moderate hearing loss.</p>
序列相似性	Belongs to the ribose-phosphate pyrophosphokinase family.

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