血影蛋白 A 链红细胞型抗体

产品货号： mlR10190

英文名称： Spectrin (alpha + beta)

中文名称： 血影蛋白 A 链红细胞型抗体

别　名： Spectrin alpha chain, erythrocytic 1; Alpha I spectrin; EL 2; EL2; Elliptocytosis 2; Elliptocytosis2; Erythrocyte alpha spectrin; erythrocyte; Erythroid alpha spectrin; Erythroid alpha-spectrin; Erythroid spectrin alpha; HPP; HS3; Spectrin alpha chain; Spectrin alpha chain erythrocyte; Spectrin alpha erythrocytic 1; SPH3; SPTA 1; SPTA; SPTA1; SPTA1_HUMAN; Spectrin alpha chain, erythrocytic 1.

研究领域： 心血管　信号转导

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Pig, Horse, Rabbit,

产品应用： WB=1:500-2000 ELISA=1:500-1000
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 280kDa

细胞定位： 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from Human Spectrin alpha chain, erythrocytic 1:2151-2350/2419

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

PubMed： PubMed

产品介绍： Spectrin is an actin crosslinking and molecular scaffold protein that links the plasma membrane to the actin cytoskeleton, and functions in the determination of cell shape, arrangement of transmembrane proteins, and organization of organelles. It is a tetramer made up of alpha-beta dimers linked in a head-to-head arrangement. This gene is one member of a family of alpha-spectrin genes. The encoded protein is primarily composed of 22 spectrin repeats which are involved in dimer formation. It forms weaker tetramer interactions than non-erythrocytic alpha spectrin, which may increase the plasma membrane elasticity and deformability of red blood cells. Mutations in this gene result in a variety of hereditary red blood cell disorders, including elliptocytosis type 2, pyropoikilocytosis, and spherocytic hemolytic anemia. [provided by RefSeq, Jul 2008].

Function:

Spectrin is the major constituent of the cytoskeletal network underlying the erythrocyte plasma membrane. It associates with band 4.1 and actin to form the cytoskeletal superstructure of the erythrocyte plasma membrane.

Subunit:

Composed of non-homologous chains, alpha and beta, which aggregate side-to-side in an antiparallel fashion to form dimers, tetramers, and higher polymers. Interacts with FASLG.

Subcellular Location:

Cytoplasm, cytoskeleton. Cytoplasm, cell cortex.

DISEASE:

Defects in SPTA1 are the cause of elliptocytosis type 2 (EL2) [MIM:130600]. EL2 is a Rhesus-unlinked form of hereditary elliptocytosis, a genetically heterogeneous, autosomal dominant hematologic disorder. It is characterized by variable hemolytic anemia and elliptical or oval red cell shape.

Defects in SPTA1 are a cause of hereditary pyropoikilocytosis (HPP) [MIM:266140]. HPP is an autosomal recessive disorder characterized by hemolytic anemia, microspherocytosis, poikilocytosis, and an unusual thermal sensitivity of red cells.

Defects in SPTA1 are the cause of spherocytosis type 3 (SPH3) [MIM:270970]; also known as hereditary spherocytosis type 3 (HS3). Spherocytosis is a hematologic disorder leading to chronic hemolytic anemia and characterized by numerous abnormally shaped erythrocytes which are generally spheroidal. SPH3 is characterized by severe hemolytic anemia. Inheritance is autosomal recessive.

Similarity:

Belongs to the spectrin family.

Contains 3 EF-hand domains.

Contains 1 SH3 domain.

Contains 21 spectrin repeats.

SWISS:

P02549

Gene ID:

6708

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

产品图片