生物素酶抗体

产品货号： mlR11813

英文名称： BTD

中文名称： 生物素酶抗体

别　　名： Biotinase; Biotinidase; Btd; Sp8; BTD_HUMAN; EC 3.5.1.12.

研究领域： 细胞生物　神经生物学　信号转导

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Chicken, Dog, Cow, Horse, Rabbit, Sheep,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 57kDa

细胞定位： 分泌型蛋白

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human Biotinidase:401-500/543

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： Biotin, also known as vitamin B7, is an essential water-soluble vitamin that is a cofactor in glucogenesis and in the metabolism of fatty acids and leucine. Biotinidase is a 523 amino acid enzyme that catalyzes the hydrolysis of biocytin to biotin and lysine. Secreted into extracellular space, biotinidase is expressed in liver, heart, placenta, brain, skeletal muscle, pancreas and kidney. Biotinidase contains one carbon-nitrogen hydrolase domain, which is involved in the reduction of organic nitrogen compounds and ammonia production. Defects in the gene encoding biotinidase are the cause of biotinidase deficiency, which is characterized by skin rash, ataxia, seizures, hearing loss, hypotonia and optic atrophy. These symptoms are due to the individual’s inability to reutilize biotin and can, therefore, typically be treated with the addition of free biotin.

Function:

Catalytic release of biotin from biocytin, the product of biotin-dependent carboxylases degradation.

Subcellular Location:

Secreted.

DISEASE:

Defects in BTD are the cause of biotinidase deficiency (BTD deficiency) [MIM:253260]; also called late-onset multiple carboxylase deficiency. BTD deficiency is a juvenile form of multiple carboxylase deficiency, an autosomal recessive disorder of biotin metabolism, characterized by ketoacidosis, hyperammonemia, excretion of abnormal organic acid metabolites, and dermatitis.

BTD deficiency is characterized by seizures, hypotonia, skin rash, alopecia, ataxia, hearing loss, and optic atrophy. If untreated, symptoms usually become progressively worse, and coma and death may occur.

Similarity:

Belongs to the CN hydrolase family. BTD/VNN subfamily.

Contains 1 CN hydrolase domain.

SWISS:

P43251

Gene ID:

686

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