



Rabbit Anti-BTD/FITC Conjugated antibody

SL11813R-FITC

Product Name:	Anti-BTD/FITC
Chinese Name:	FITC标记的生物素酶抗体
Alias:	Biotinase; Biotinidase; Btd; Sp8; BTD HUMAN; EC 3.5.1.12.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Cow,Horse,Rabbit,Sheep,
Applications:	ICC=1:50-200IF=1:50-200 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	57kDa
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Biotinidase
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	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.
Product Detail:	background: 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 BTBD9 are the cause of biotinidase deficiency (BTBD9 deficiency) [MIM:253260]; also called late-onset multiple carboxylase deficiency. BTBD9 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.

BTBD9 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. BTBD/VNN subfamily.
Contains 1 CN hydrolase domain.

Database links:

[Entrez Gene: 686](#) Human

[Omicron: 609019](#) Human

[SwissProt: P43251](#) Human

[Unigene: 517830](#) Human

Important Note:

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