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BTD Rabbit pAb

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Catalog # AP54631

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P43251
Reactivity	Rat
Predicted	Dog, Human, Mouse, Rabbit, Chicken, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	58913
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Biotinidase
Epitope Specificity	401-500/543
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	686

Other Names	Biotinidase, Biotinase, 3.5.1.12, BTD (HGNC:1122)
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	BTD (HGNC:1122)
Function	Catalytic release of biotin from biocytin, the product of biotin-dependent carboxylases degradation.
Cellular Location	Secreted, extracellular space

Background

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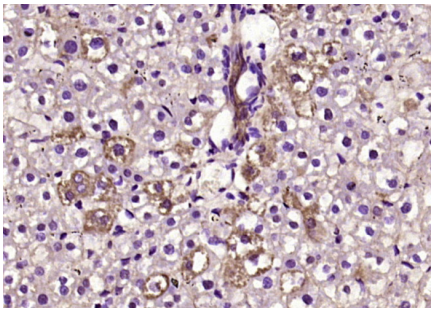
Images



Paraformaldehyde-fixed, paraffin embedded (rat liver); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (BTD) Polyclonal Antibody, Unconjugated (AP54631) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.