



## biotinidase (S-12): sc-102346

### BACKGROUND

Biotin, also known as vitamin B7, is an essential water-soluble vitamin that is a cofactor in glucogenesis and in the metabolism of both 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.

### REFERENCES

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4. Norrgard, K.J., Pomponio, R.J., Swango, K.L., Hymes, J., Reynolds, T., Buck, G.A. and Wolf, B. 1998. Double mutation (A171T and D444H) is a common cause of profound biotinidase deficiency in children ascertained by newborn screening the the United States. *Mutations in brief no. 128. Online Hum. Mutat.* 11: 410.
5. Knight, H.C., Reynolds, T.R., Meyers, G.A., Pomponio, R.J., Buck, G.A. and Wolf, B. 1998. Structure of the human biotinidase gene. *Mamm. Genome* 9: 327-330.
6. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 253260. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>.
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### CHROMOSOMAL LOCATION

Genetic locus: BTB (human) mapping to 3p24.3; Btd (mouse) mapping to 14 B.

### SOURCE

biotinidase (S-12) is an affinity purified rabbit polyclonal antibody raised against a peptide mapping within an internal region of biotinidase of human origin.

### PRODUCT

Each vial contains 100 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-102346 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

### APPLICATIONS

biotinidase (S-12) is recommended for detection of biotinidase of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for biotinidase siRNA (h): sc-78477, biotinidase siRNA (m): sc-141707, biotinidase shRNA Plasmid (h): sc-78477-SH, biotinidase shRNA Plasmid (m): sc-141707-SH, biotinidase shRNA (h) Lentiviral Particles: sc-78477-V and biotinidase shRNA (m) Lentiviral Particles: sc-141707-V.

Molecular Weight of biotinidase: 57 kDa.

### RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

### STORAGE

Store at 4° C, **\*\*DO NOT FREEZE\*\***. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

### RESEARCH USE

For research use only, not for use in diagnostic procedures.

### PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.