

Biotin

Newborn Use only

2026

Key points	To be prescribed only in consultation with paediatric metabolic specialist.
Indication	Defects of biotin metabolism including biotinidase deficiency and holocarboxylase synthetase (HLCS) deficiency (multiple carboxylase deficiency).
Action	Water-soluble B-vitamin. Also known as vitamin B7. Biotin functions as a cofactor for enzymes involved in carboxylation reactions essential for gluconeogenesis, lipid metabolism, and amino acid catabolism.
Contraindications	None reported.
Precautions	High-dose biotin may interfere with laboratory immunoassays that use biotin-streptavidin technology, potentially causing falsely high or falsely low results. Inform the laboratory that the infant is receiving biotin before testing and follow assay-specific advice. ⁶
Dose	Starting dose: 5-10 mg ONCE DAILY IV or oral. Usual maintenance dose: 5-20 mg once DAILY. ⁶ Higher doses may be required as per the advice of the paediatric metabolic specialist for specific conditions, for example, Biotin and Thiamine Basal Ganglia Disease and Holocarboxylase Synthetase Deficiency. ^{1,2}
Route	ORAL IV
Presentation	5 mg/mL, 10 mg/mL, 20 mg/mL oral suspension (prepared by hospital pharmacy), 2 mg/mL, 5 mg/mL solution for injection
Preparation	ORAL – Crush and disperse in water. ²
IV Compatibility	Not applicable.
IV Incompatibility	Not applicable.
Administration	IV – Administer over 3-5 minutes. ² ORAL
Monitoring	No specific monitoring has been reported.
Drug Interactions	No known clinically significant drug interactions. See Precautions regarding laboratory assay interference.
Adverse Reactions	Generally well tolerated; no known adverse effects from pharmacological doses have been established. ⁶
Overdose	No reports. AUSTRALIA: Contact the Poisons Information Centre on 13 11 26 for information on management. NEW ZEALAND: Contact the National Poisons Centre on 0800 764 766 for information on management.
Stability	Oral suspension prepared in hospital pharmacy: 5 mg/mL is 90 days refrigerated. 20 mg/mL is 60 days refrigerated.
Storage	Oral suspension: 2 to 8 °C (refrigeration) Tablet and injection: Below 25 °C
Excipients	Oral suspension: Methylhydroxybenzoate, sorbates and sorbitol Injection: methyl propyl parahydroxybenzoate
Evidence	<p>Overview</p> <p>Biotin is a water-soluble B-vitamin. Biotin functions as a cofactor for enzymes involved in carboxylation reactions essential for gluconeogenesis, lipid metabolism, and amino acid catabolism. Main enzymes that require biotin are acetyl CoA carboxylase (ACC) 1 and 2 (ACC2), methylcrotonyl CoA carboxylase (MCC), propionyl CoA carboxylase (PCC), and pyruvate carboxylase. Holocarboxylase synthetase (HLCS) covalently attaches biotin to these apo-carboxylases, enabling enzyme activity.³ The adequate intake for biotin is 5 µg/day for infants aged 0–6 months.³</p> <p>Inherited disorders of biotin metabolism</p> <p>Biotinidase deficiency impairs recycling of biotin, whereas HLCS deficiency impairs attachment of biotin to biotin-dependent carboxylases. Both disorders can cause functional intracellular biotin deficiency and multiple carboxylase deficiency.^{1,4} Holocarboxylase synthetase deficiency (HLCS deficiency) is an exceedingly rare metabolic disorder resulting in multiple carboxylase deficiencies owing to impaired biotin cycle. Clinical manifestations include severe metabolic acidosis, hyperammonemia, tachypnoea, skin rash, alopecia, feeding problems, hypotonia, developmental delay, seizures, and – in severe cases – death.¹</p>

	<p>HLCS deficiency can be treated with oral pharmacological doses of biotin (10–50 mg/day), particularly in patients with biotin-responsive variants, leading to significant improvements in clinical outcomes. Higher doses may be required.^{1,3,5}</p> <p>Sodium-dependent multivitamin transporter (SMVT) mimics biotinidase deficiency. A multivitamin cocktail has been successfully used in a 4-month-old infant. In this case, biotin 10-15 mg/day was used along with other multivitamins.⁶</p> <p>Safety</p> <p>Well tolerated. No adverse effects have been reported.</p>
References	<ol style="list-style-type: none"> Kim SW, Lee HJ, Choi N, Kim EK, Ko JM. Dramatic Clinical Improvement With Biotin Mega-Dose Therapy in a Neonate With Holocarboxylase Synthetase Deficiency. <i>Mol Genet Genomic Med.</i> 2024;12(8):e70002. British Inherited Metabolic Diseases Group (BIMDG). https://bimdg.org.uk/emergency/paediatric-guidelines/. Accessed on 11 August 2026. Perry CA, Butterick TA. Biotin. <i>Adv Nutr (Bethesda).</i> 2024;15(7):100251. Wolf B. Biotinidase Deficiency. In: Adam MP, Bick S, Mirzaa GM, et al., editors. <i>GeneReviews®</i> [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2026. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1322/. Accessed 20 September 2026. Said HM. Biotin: biochemical, physiological and clinical aspects. <i>Water soluble vitamins: clinical research and future application.</i> 2011:1–19. Van Vyve FX, Mercier N, Papadopoulos J, Heijmans C, Dessy H, Monestier O, et al. A new case of sodium-dependent multivitamin transporter defect occurring as a life-threatening condition responsive to early vitamin supplementation and literature review. <i>Mol Genet Genomic Med.</i> 2024;12(2):e2388.

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