

Key points	Vitamin B2.
Indication	Riboflavin responsive inherited metabolic disease, such as multiple acyl-CoA dehydrogenase deficiency (MADD), acyl-CoA dehydrogenase 9 (ACAD9) deficiency.
Action	Water soluble vitamin B2 of which the co-factors flavin mononucleotide (FMN) and flavin adenine dinucleotide (FAD) are critical to numerous cellular processes, including mitochondrial energy metabolism and the function of various flavoenzymes. ¹
Contraindications	
Precautions	May interfere with lab analysis of some tests, particularly fluorescence-based assays; inform the laboratory if the patient is receiving high-dose riboflavin.
Dose	MADD (including neonatal MADD): 50 mg (NOT /kg) three times a day. ² For other riboflavin-responsive inherited metabolic disorders, dosing varies; use as directed by the paediatric metabolic specialist.
Route	ORAL
Presentation	Riboflavin 100 mg, 200 mg tablets (various brands)
Preparation	Crush and disperse a tablet in water and give required portion.
IV Compatibility	Not applicable.
IV Incompatibility	Not applicable.
Administration	May administer with feeds
Monitoring	No specific monitoring has been reported.
Drug Interactions	No specific interactions are reported.
Adverse Reactions	Generally well tolerated. Mild gastrointestinal symptoms may occur. Yellow-orange discoloration of urine is expected and harmless. ¹
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	Not applicable.
Storage	Store below 25°C
Excipients	Refer to product information.
Evidence	<p>Overview</p> <p>Riboflavin deficiency can be acquired from prolonged vitamin free parenteral nutrition or inherited due to inborn error of metabolism.³⁻⁸</p> <p>Riboflavin is a water-soluble B vitamin. It is a cofactor required for numerous flavoprotein-catalysed reactions. Riboflavin has important antioxidant properties, essential for cell functioning. It is required for the conversion of oxidised glutathione to the reduced form and for the mitochondrial respiratory chain.⁹</p> <p>Riboflavin transporter deficiency</p> <p>Riboflavin transporter deficiencies (RTD) are autosomal recessive diseases caused by biallelic mutations of SLC52A2 and SLC52A3 genes that encode riboflavin transporters. RTD is formerly known as Brown-Vialetto-VanLaere (BVVL) or Fazio Londe syndrome. RTD affects one in 1,000,000 people in the general population. It causes multifocal and progressive neurological deficits. Most cases present at a young age and the disease course in infancy is rapidly progressive and lethal when left untreated. Patients present with multiple cranial neuropathies, with hearing loss as the most frequent symptom. Visual loss, facial weakness, and marked bulbar dysfunction with difficulties speaking and swallowing may all develop over time. In addition, a peripheral neuropathy leads to limb muscle weakness, sensory ataxia, and respiratory insufficiency due to weakness of the diaphragm.⁷ High doses of riboflavin represents the mainstay of therapy for these diseases and should be commenced promptly when the diagnosis is suspected and continued lifelong.⁹</p> <p>Riboflavin therapy</p> <p>Riboflavin has been administered in a range of inborn errors of metabolism, including Riboflavin transporter deficiency type 2 and 3, as well as a substantial number of mitochondrial IMDs, such as multiple acyl-CoA dehydrogenase deficiency (MADD) and acyl-CoA dehydrogenase 9 (ACAD9) deficiency.¹ The level of evidence supporting the effectiveness of riboflavin therapy in these disorders differs. In a systematic review, riboflavin treatment was considered effective (improvement in >75% of patients) in 4</p>

	<p>metabolic diseases: MADD type 3, RTD type 2 and 3, ACAD9, and flavin adenine dinucleotide (FAD) transporter deficiency.¹</p> <p>Systematic review reported following dose ranges of riboflavin: (1) In MADD type 3 deficiency – ranged: 9-800 mg/day from 1 month to 80 years of age; (2) in RTD type 2 and 3 – ranged: 1.7-80 mg/kg/day and 20–1500 mg/day; (3) in ACAD 9 deficiency – ranged: 15–20 mg/kg/day to 40–300 mg/day, and (4) in FAD transporter deficiency – ranged: 30–200 mg/day.¹</p> <p>Safety</p> <p>Generally well tolerated. May experience mild gastrointestinal symptoms.¹</p>
References	<ol style="list-style-type: none"> Jaeger B, Stolwijk NN, Aaldering F, Ruys CA, Smits M, Wolf NI, et al. Effectiveness of riboflavin in inherited metabolic diseases: a systematic review. <i>Journal of Inherited Metabolic Disease</i>. 2026;49(3):e70190. British Inherited Metabolic Diseases Group (BIMDG). https://bimdg.org.uk/emergency/paediatric-guidelines/. Accessed on 11 August 2026. Chalmers RA, Bain MD, Zschocke J. Riboflavin-responsive glutaryl CoA dehydrogenase deficiency. <i>Mol Genet Metab</i>. 2006;88(1):29–37. Vieira P, Myllynen P, Perhomaa M, Tuominen H, Keski-Filppula R, Rytty S, et al. Riboflavin-Responsive Multiple Acyl-CoA Dehydrogenase Deficiency Associated with Hepatoencephalomyopathy and White Matter Signal Abnormalities on Brain MRI. <i>Neuropediatrics</i>. 2017;48(3):194–8. Adie MA, Martes Gomez M, Yom J, Durand M, Wertheimer F, McGowan R, et al. Twin Premature Infants With Riboflavin and Biotin Deficiency Presenting With Refractory Lactic Acidosis, Rash, and Multiorgan Failure During Prolonged Parenteral Nutrition. <i>Journal of Investigative Medicine High Impact Case Reports</i>. 2023;11:23247096231168111. Grunert SC, Ziagaki A, Heinen A, Schumann A, Tucci S, Spiekerkoetter U, et al. Riboflavin 1 Transporter Deficiency: Novel SLC52A1 Variants and Expansion of the Phenotypic Spectrum. <i>Genes (Basel)</i>. 2023;14(7):07. Jaeger B, Hoytema van Konijnenburg E, Groenveld MA, Langeveld M, Wolf NI, Bosch AM. Riboflavin transporter deficiency, the search for the undiagnosed: a retrospective data mining study. <i>Orphanet Journal Of Rare Diseases</i>. 2024;19(1):410. Panichsillaphakit E, Laphathai P, Dhachpramuk D, Vatanavicharn N. Transient neonatal multiple acyl-CoA dehydrogenase deficiency due to riboflavin deficiency in an infant on total parenteral nutrition. <i>BMJ Case Rep</i>. 2025;18(5):05. Plantone D, Pardini M, Rinaldi G. Riboflavin in Neurological Diseases: A Narrative Review. <i>Clinical Drug Investigation</i>. 2021;41(6):513–27.

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