

Key points	To be prescribed only in consultation with paediatric metabolic specialist. Also known as ubiquinone, ubiquinone, or CoQ10.
Indication	Inborn errors of co-enzyme Q10 synthesis. Defects of mitochondrial respiratory chain.
Action	Coenzyme Q10 (CoQ10) is the predominant human form of endogenous ubiquinone. It is synthesised in the mitochondrial inner membrane. It is an electron carrier in mitochondrial respiratory chain and an important antioxidant. ^{1,2}
Contraindications	Hypersensitivity to CoQ10 or any component of the formulation.
Precautions	May reduce insulin requirement in diabetes mellitus; monitor blood glucose where relevant. ³ Consider dose reduction in moderate and severe hepatic impairment. ³
Dose	10-30 mg/kg/day in 1-2 divided doses to a maximum of 200 mg/day in neonate and a maximum of 300 mg in older child. ³ Note: Tablets are available as 100 mg. Dose is approximated to convenient preparation.
Route	ORAL
Presentation	Ubidecarenone (Coenzyme Q10) 100 mg, 150 mg chewable tablets ⁴ , 50 mg, 75 mg, 100 mg, 150 mg, and 300 mg soft capsules
Preparation	Due to poor solubility, it is recommended to round dose to the nearest quarter, half or whole tablet. May be crushed and mixed with feeds. (ANMF consensus) Soft capsule: Pierce the capsule and squeeze the contents.
IV Compatibility	Not applicable.
IV Incompatibility	Not applicable.
Administration	Give with feeds to maximise absorption.
Monitoring	No specific monitoring has been reported.
Drug Interactions	Warfarin: CoQ10 may reduce the anticoagulant effect; monitor INR if co-administered. ³ Insulin/glucose-lowering therapy: requirements may be reduced; monitor blood glucose. ^{3,6}
Adverse Reactions	Uncommon – diarrhoea, dyspepsia, nausea, wakefulness, irritability
Overdose	No reports. AUSTRALIA: Contact the Poisons Information Centre on 13 11 26 for information on the 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	To check the product information.
Evidence	<p>Overview</p> <p>Coenzyme Q10 (CoQ10) is also known as ubiquinone. It is a naturally occurring substance, and is located within intracellular membranes, principally in mitochondria, but also within the membranes of other subcellular organelles such as lysosomes, peroxisomes, and Golgi apparatus. CoQ10 has a key role in many aspects of cellular metabolism: an important role in the generation of ATP within mitochondria, an important antioxidant, and roles in the metabolism of fatty acids, amino acids, sulphides, and cholesterol.⁵</p> <p>CoQ10 deficiency</p> <p>CoQ10 deficiencies are clinically and genetically heterogeneous diseases that are potentially treatable and therefore important to diagnose early. Deficiency of CoQ10 is broadly divided into primary CoQ10 deficiency and secondary CoQ10 deficiency. Primary CoQ10 deficiency results from mutations in genes involved in the CoQ10 biosynthetic pathway.^{2,5} Secondary CoQ10 deficiency can occur in other genetic and mitochondrial disorders.</p> <p>Primary CoQ10 deficiency</p> <p>Primary CoQ10 deficiency is a very rare disorder, the prevalence and incidence of which have been estimated to be less than 1 per 100,000 population.⁵ They are generally inherited via an autosomal recessive pattern with a clinical spectrum that encompasses at least five major clinical phenotypes: (1) encephalomyopathy, (2) severe infantile multisystemic disease, (3) cerebellar ataxia, (4) isolated myopathy, and (5) nephrotic syndrome.^{1,2}</p> <p>Treatment of primary CoQ10 deficiency</p>

	<p>It is suggested that early diagnosis and treatment with CoQ10 is critical in CoQ10 deficient patients to maximize the efficacy of CoQ10 supplementation.^{1,2} Clinical studies supplementing CoQ10 in neonates are restricted to subjects with primary CoQ10 deficiency or acyl-CoA dehydrogenase deficiency, respectively.⁵ Doses used in clinical studies are typically in the range of 50–50 mg/kg/day.^{7,8}</p> <p><u>Treatment of Mitochondrial disorder</u></p> <p>Dose of ubiquinone used is 10-30 nmg/kg/day in two divided doses.⁹</p> <p><u>Safety of CoQ10 supplementation</u></p> <p>It is generally a safe supplementation with no major adverse effects reported.⁵</p>
References	<ol style="list-style-type: none"> 1. Quinzii CM, Hirano M. Coenzyme Q and mitochondrial disease. Dev Disabil Res Rev. 2010;16(2):183–8. 2. Quinzii CM, DiMauro S, Hirano M. Human coenzyme Q10 deficiency. Neurochem Res. 2007;32(4-5):723–7. 3. British Inherited Metabolic Diseases Group (BIMDG). https://bimdg.org.uk/emergency/paediatric-guidelines/. Accessed on 11 August 2026. 4. Coenzyme Q10 chewable tablets. https://www.devanutrition.com/vegan-coenzyme-q10-100mg-chewable.html. 5. Mantle D, Hargreaves IP. Efficacy and safety of coenzyme Q10 supplementation in Neonates, infants and children: an overview. Antioxidants. 2024;13(5):530. 6. National Center for Complementary and Integrative Health. Coenzyme Q10. Available from: https://www.nccih.nih.gov/health/coenzyme-q10. Accessed 20 September 2026. 7. Salvati L, Trevisson E, Agosto C, Doimo M, Navas P. Primary Coenzyme Q10 Deficiency Overview. In: GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2026. Updated 8 June 2023. 8. Sue CM, Balasubramaniam S, Bratkovic D, et al. Patient care standards for primary mitochondrial disease in Australia: an Australian adaptation of the Mitochondrial Medicine Society recommendations. Intern Med J. 2022;52(1):110-120. doi:10.1111/imj.15505. 9. Parikh S, Saneto R, et al. A Modern Approach to the Treatment of Mitochondrial Disease. Curr Treat Options Neurol. 2009 November; 11(6): 414–430.

VERSION/NUMBER	DATE
Original 1.0	29/10/2026
REVIEW (5 years)	29/10/2031

Authors Contribution of the current version

Author/s	Dr Srinivas Bolisetty, Dr Shanti Balasubramaniam, Dr Kate Lichkus, Dr Gregory Woodhead
Expert Review	Drs Shanti Balasubramaniam, Kate Lichkus, Gregory Woodhead (Paediatric metabolic specialists, Sydney Children's Hospital Network)
Pharmacy review	Mohammad Irfan Azeem, Simarjit Kaur
Nursing review	Benjamin Emerson-Parker
ANMF group contributors	Nilkant Phad, Bhavesh Mehta, Jutta van den Boom, Amber Seigel, Rebecca Barzegar, Kerrie Knox, Cindy Chen, Thao Tran, Celia Cunha Brites, Gloria Yoo, Dianne Lee, Michelle Jenkins, Benjamin Emerson-Parker, Renae Gengaroli, Bryony Malloy, Marvin Dangnui, Marguerite Mason-Elliott
Final editing	Srinivas Bolisetty
Facilitator	Srinivas Bolisetty

Citation

Australasian Neonatal Medicines Formulary (ANMF). Coenzyme Q10. Version number: 1. Date of publication 29/10/2026. <https://www.anmfonline.org/>