Levocarnitine (L-Carnitine) 2026

Newborn Use only

Key points	Oral doses may be titrated gradually to improve gastrointestinal tolerance. Acute IV therapy should follow the relevant metabolic emergency protocol and metabolic specialist advice.
Indication	Primary carnitine deficiency (SLC22A5/OCTN2 deficiency) Secondary carnitine deficiency Organic acidaemia
Action	An essential carrier molecule that helps transport long-chain fatty acids into the mitochondria for beta-oxidation which produces energy in the form of adenosine triphosphate (ATP). Carnitine also helps transport some toxic compounds (acylcarnitine esters) out of the mitochondria.[1,2]
Contraindications	None known. Hypersensitivity reactions including rash, urticaria, and facial oedema have been reported with oral levocarnitine. With intravenous levocarnitine, more serious reactions—including anaphylaxis, laryngeal oedema, and bronchospasm—have been reported, sometimes within minutes of administration. Most severe IV reactions have occurred in patients with end-stage renal disease receiving dialysis. Discontinue if a serious hypersensitivity reaction occurs.[8]
Precautions	In patients with cardiomyopathy or cardiac arrhythmia, particularly where a long-chain fatty-acid oxidation disorder (VLCAD deficiency and LCHAD/trifunctional protein deficiency) is suspected, use only on advice of a paediatric metabolic specialist.
Dose	**ORAL** <u>Primary carnitine deficiency</u> 100-200 mg/kg/day in 2-4 divided doses. Higher doses may be required according to plasma carnitine concentrations and clinical response; discuss with paediatric metabolic specialist. <u>Secondary carnitine deficiency and organic acidaemias</u> Dose depends on the underlying disorder and clinical context; discuss with a paediatric metabolic specialist. **IV** Intermittent bolus: 25 mg/kg/dose IV 6 hourly (100 mg/kg/day) Continuous infusion: 100 mg/kg IV loading over 30 minutes, followed by 4 mg/kg/hour as maintenance.[6]
Route	ORAL and IV
Presentation	ORAL Levocarnitine Oral Solution (USP) 1g/10mL (100 mg/mL) Levocarnitine 300mg/mL solution IV Levocarnitine (SAS) (L-Carn) 1g/5mL injection – Available under Special Access Scheme in Australia.
Preparation	Oral: No preparation is required. IV bolus: Can be given undiluted. IV infusion: Draw up 2 mL (400 mg) of levocarnitine injection and dilute with 48 mL of glucose 5% or 10%, or sodium chloride 0.9% to make a volume of 50 mL with a final concentration of 8 mg/mL.
IV Compatibility	Fluids: No data PN at Y site: No data. SMOFlipid: Compatible Y site: Dexmedetomidine, Meropenem, Naloxone
IV Incompatibility	No information.
Administration	Oral: Administer during or following meals to reduce GI reactions and taste fatigue. IV: For intermittent dosing, administer over 2-3 minutes. For continuous infusion, prepare and administer according to the relevant metabolic emergency protocol and local pharmacy guidance.
Monitoring	Monitor plasma free and total carnitine concentrations and adjust dose according to the underlying disorder, biochemical results, and clinical response. Monitor clinical status and gastrointestinal tolerance.
Drug Interactions	Warfarin: increased INR has been reported; monitor INR if co-administered. No other clinically important interactions are routinely reported.
Adverse Reactions	Abdominal cramps, diarrhoea, reduced appetite, vomiting, and a fish-like body odour has also been reported
Overdose	No specific human toxicity has been established. Large doses may cause diarrhoea. Provide supportive and symptomatic treatment. AUSTRALIA: Contact the Poisons Information Centre on 13 11 26 for information on management.

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	NEW ZEALAND: Contact the National Poisons Centre on 0800 764 766 for information on management.
Stability	Not applicable.
Storage	Store at room temperature (below 25^0C).
Excipients	Refer to product information.
Evidence	**Overview** Carnitine is derived from amino acid and is naturally present in many foods. The majority of carnitine in the body is supplied from diet with small amounts produced endogenously. Carnitine plays a critical role in energy production. It is an essential cofactor that helps transport long-chain fatty acids into the mitochondria so that they can be oxidized to produce energy in the form of adenosine triphosphate (ATP). Carnitine also helps transport some toxic compounds (acylcarnitine esters) out of the mitochondria.[1,2] L-Carnitine is also known as β-hydroxy-y-trimethylaminobutyric acid. It is not the same as beta-hydroxybutyric acid.[2] **L- carnitine treatment** Primary systemic carnitine deficiency is caused by biallelic pathogenic variants in SLC22A5, resulting in defective OCTN2-mediated carnitine transport. Levocarnitine is established treatment and is usually given at 100-200 mg/kg/day orally in divided doses, with dose adjustment according to plasma carnitine concentrations and clinical response. ORAL Levocarnitine is also used in selected secondary carnitine deficiencies and organic acidaemias. The indication and dose depend on the disorder and clinical context.[2,6,7] Carnitine supplementation should not be presented as routine treatment for all fatty-acid oxidation disorders. In MCAD deficiency, treatment of secondary carnitine deficiency remains controversial; when used, lower oral doses such as 25-50 mg/kg/day are commonly described. In long-chain fatty-acid oxidation disorders, use should be individualised and guided by a metabolic specialist because long-chain acylcarnitine accumulation may be clinically important.[9,10] IV L-carnitine (100-200 mg/kg/day) is recommended in acute and ongoing management of many primary and secondary carnitine deficiencies, and some organic acidaemias.[2-7] Dose ranges recommended in this formulary are based on the consensus within Sydney Children's Hospital network paediatric metabolic specialist group and largely adapted from the British Inherited Metabolic Diseases Group. **Safety** L-carnitine is generally well tolerated with no major side effects. Diarrhoea and an unpleasant fishy body odour are dose-related adverse effects and may improve with dose reduction.[2]
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Authors Contribution of the current version

Author/s	Srinivas Bolisetty, Shanti Balasubramaniam, Kate Lichkus, Gregory Woodhead
Expert Review	Drs Shanti Balasubramaniam, Kate Lichkus, Gregory Woodhead (Paediatric metabolic specialists, Sydney Children's Hospital Network)
Pharmacy review	Simarjit Kaur, Carmen Burman, Mohammad Irfan Azeem
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, Renae Gengaroli, Bryony Malloy, Marvin Dangnui, Marguerite Mason-Elliott
Final editing	Srinivas Bolisetty
Facilitator	Srinivas Bolisetty

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