LEVOCARNITINE

<table>
<tr><td>Trade Name</td><td>Oral: L-Carnitine 1500 Oral Liquid (Prosupps)
IV: Levocarnitine Injection (Sigma-Tau)</td></tr>
<tr><td>Class</td><td>Quaternary Ammonium Compound</td></tr>
<tr><td>Mechanism of Action</td><td>Carnitine is an amino acid essential for transportation of long-chain fatty acids into mitochondria. Carnitine is mostly found in skeletal and cardiac muscle. Deficiency presents with hypotonia, weakness, poor feeding and developmental delay.
For the indications below levocarnitine works by either replacing the missing amino acid or acts as a scavenger to remove accumulated toxins</td></tr>
<tr><td>Indications</td><td>Indication 1: Organic aciduria eg: methylmalonic, propionic or isovaleric acidaemia
Indication 2: Carnitine deficiency – rare in neonates</td></tr>
<tr><td>Contraindications</td><td>Hypersensitivity to carnitine
Avoid use in patients with cardiomyopathy or cardiac arrhythmias unless on advice of the metabolic specialist
Use with caution in patients with renal impairment.
Use with caution in patients with seizures – both new onset seizure activity and increased frequency of seizures have been reported.</td></tr>
<tr><td>Supplied As</td><td>Oral: 1.5 g /15 mL (= 100 mg/mL)
IV: 1g /5 mL 5 mL vial (= 200mg/mL)</td></tr>
<tr><td>Dilution</td><td>Oral: No dilution required

IV infusion: Take 5mL (= 1 g) and add 45mL sodium chloride to give a final volume of 50mL final concentration = 20 mg/mL

<table>
<tr><th>Drug</th><th>Sodium Chloride 0.9%</th><th>Final Volume</th><th>Final concentration</th></tr>
<tr><td>5mL (1g)</td><td>45 mL</td><td>50 mL</td><td>20 mg/mL</td></tr>
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IV bolus: no dilution required (note IV infusion is preferred)</td></tr>
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Dosage ***Must chart guardrail and use Alaris pump ***	**Oral:** 100-200mg/kg/day **IV:** **Loading dose:** 100mg/kg infused over 30 minutes **Maintenance:** 4mg/kg/hr continuous infusion (see infusion sheet) Higher doses on the advice of the Metabolic team **Bolus**: 100mg/kg/DAY Maximum dose= 3 grams/DAY
Guardrails **ALARIS PUMP**	Min Concentration: 20mg/mL Max Concentration: 200 mg/mL Soft Alert Min: 1mg/kg/hr Hard Alert Max: 200mg/kg/hr Soft Alert Max: 8mg/kg/hr Default Setting: 4mg/kg/hr
Interval	**Oral:** 6 hourly **IV:** Continuous infusion is the preferred method **IV Bolus:** 6 hourly, slow injection over 2-3 minutes
Administration	**Oral:** may be mixed with flavoured drink to mask taste **IV:** IV infusion is the preferred method of IV administration
Compatible With	**Solution:** sodium chloride 0.9%, lactated Ringer's **Terminal Y-site:** fat emulsion, meropenem, naloxone IV compatibility of levocarnitine with IV solutions is very limited There is no information available on compatibility with TPN or amino acid solutions. **Note:** CDHB Paediatrics report compatibility with Glucose 5% and 10 % and successful Y-site infusion of Actrapid insulin, sodium benzoate, sodium phenyl butyrate and sodium bicarbonate in combination with levocarnitine.
Incompatible With	No information available
Monitoring	Plasma carnitine concentrations, serum triglycerides, fatty acids, electrolytes, blood pressure, heart rate.
Stability	**Oral:** 6 months after opening or manufacturers expiry - whichever is shorter **IV:** Single use ampoules. Discard any unused solution immediately after use.
Storage	**Oral solution:** Room temperature, protect from light **IV:** Room temperature, protect from light
Adverse Reactions	Nausea, vomiting, abdominal pain, diarrhoea, anorexia, fishy body odour, hypertension, tachycardia and rash
Metabolism	Oral bioavailability is poor (approx. 15%), Half life (in adults) =18 hrs Extensive hepatic metabolism, 4-9% excreted as unchanged drug

Comments	Section 29 medication. Application for NPPA funding will be needed for supply on discharge. Babies who require levocarnitine usually require transfer to Auckland PICU for haemofiltration so treatment would be short-term
References	1. www.uptodate.com 2. www.nzf.org.nz 3. Medicines for Children RCPCH 2005, NPPG UK 4. Taketomo et al Pediatric and Neonatal Dosage Handbook 19th edition. Lexicomp 5. National Formulary for Inherited Metabolic Diseases (IMDs) 2nd Edition October 2020  BIMDG_Metabolic_ Formulary_Second_E
Updated By	A Lynn, B Robertshawe, C Wilson July 2022