


PYRIDOXAL-5-PHOSPHATE

Trade Name	P-5-P Enteric Coated tablets. (KAL USA)
Class	Water soluble vitamin (active metabolite of pyridoxine Vitamin B6)
Mechanism of Action	Co-factor in metabolism of amino acids, carbohydrate and fats and the release of glucose from glycogen
Indications	Pyridoxal -5-phosphate oxidase (PNPO) deficiency
Contraindications	Hypersensitivity to pyridoxal
Supplied As	50 mg enteric coated tablets
Dilution	Crush the tablet/part tablet with mortar and pestle and disperse in 5-10mL water (see below)
Dosage	<p>30 mg/kg/DAY</p> <p>Round up the dose (we do not want to under dose) to equate to a $\frac{1}{4}$, $\frac{1}{2}$, $\frac{3}{4}$ or a whole tablet and cut the tablet using a tablet cutter as best you can</p> <p>Dose can be increased to 60mg/kg/DAY on the advice of the Metabolic Team</p> <p>Tablets can be quite difficult to dissolve so it is recommended to reduce inaccuracies in measuring an aliquot to crush the tablets as indicated and give the full 5mL volume .</p> <p>However, if the dose required is >50mg then the tablets will need to be dissolved in up to 10ml water.</p>
Interval	12 hourly
Administration	May be given directly into the mouth or via NG or NJ tube Give dose immediately
Compatible With	Do not mix with any other medicines
Incompatible With	Do not mix with any other medicines
Monitoring	Monitor BP, heart rate, respiratory rate, and oxygen saturation every 15 minutes for the first 3 hours following the first dose Monitor LFTs
Stability	Tablets: manufacturer's expiry. Solution: single use. Discard any unused solution immediately after use
Storage	Tablets; room temperature protected from moisture and light

Adverse Reactions	Sudden respiratory arrest, profound hypotension,
Metabolism	Excreted by the kidneys
Comments	<p>Section 29 medication unregistered in NZ, will require application for NPPA funding if prescribed on discharge.</p> <p>Pyridoxal - 5 – phosphate supply can be difficult to source. Check pharmacy supply.</p> <p>An oral liquid 25mg/mL extemporaneous product from Optimus is available with a 45 day shelf life and turn around of about a week from ordering, so this is only an option for ongoing management if pyridoxal-5-phosphate deficiency is confirmed</p>
References	<ol style="list-style-type: none"> 1. National Formulary for Inherited Metabolic Diseases (IMDs) 2nd Edition October 2020  BIMDG_Metabolic_Formulary_Second_E 2. www.uptodate.com 3. https://pubchem.ncbi.nlm.nih.gov/compound/Pyridoxal-phosphate 4. https://go.drugbank.com/drugs/DB00114 5. Amenable Treatable Severe Pediatric Epilepsies. <i>Seminars in Ped Neurol.</i> 23:158-166.
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