Note:
This guideline provides advice of a general nature. This statewide guideline has been prepared to promote and facilitate standardisation and consistency of practice, using a multidisciplinary approach. The guideline is based on a review of published evidence and expert opinion.

Information in this statewide guideline is current at the time of publication.

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Health practitioners in the South Australian public health sector are expected to review specific details of each patient and professionally assess the applicability of the relevant guideline to that clinical situation.

If for good clinical reasons, a decision is made to depart from the guideline, the responsible clinician must document in the patient’s medical record, the decision made, by whom, and detailed reasons for the departure from the guideline.

This statewide guideline does not address all the elements of clinical practice and assumes that the individual clinicians are responsible for discussing care with consumers in an environment that is culturally appropriate and which enables respectful confidential discussion. This includes:

- The use of interpreter services where necessary,
- Advising consumers of their choice and ensuring informed consent is obtained,
- Providing care within scope of practice, meeting all legislative requirements and maintaining standards of professional conduct, and
- Documenting all care in accordance with mandatory and local requirements

Dose and Indications

Diagnosis and treatment of Pyridoxine dependant seizures

Intravenous and oral

Initial diagnostic dose: 50 to 100mg intravenously stops seizures within minutes

Maintenance dose: 50 to 100mg orally daily

Electroencephalography (EEG) monitoring with initial diagnostic dose is recommended.

Management of Homocystinuria

Intravenous and oral

Begin at a dose of 100mg daily, and progressively increase the dose whilst monitoring total plasma homocysteine and methionine.

Maintenance doses can range between 100mg to 1000mg daily.

Prophylaxis for peripheral neuropathy

Oral

12.5mg daily

Used in conjunction with other drugs e.g. Isoniazid.
Preparation and Administration

**Intravenous**

<table>
<thead>
<tr>
<th>Dose</th>
<th>50mg</th>
<th>100mg</th>
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</thead>
<tbody>
<tr>
<td>Volume</td>
<td>1mL</td>
<td>2mL</td>
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</table>

Administer as an Intravenous push over 5 minutes.

**Oral**

For a 12.5mg dose: Crush HALF of a 25mg tablet (12.5 mg) and disperse in a small amount of water and administer the entire mixture.

For a 100mg dose: Crush a 100mg tablet and disperse in small amount of water and administer the entire mixture.

Administer with feeds to minimise GI irritation.

**Compatible Fluids**

Glucose 5%, Sodium chloride 0.9%

**Adverse Effects**

**Common**

Profound sedation, hypotonia, hypotension, respiratory depression and apnoea have been reported following the first dose of IV or oral pyridoxine (first-dose effect).

**Infrequent**

Neurotoxicity

**Monitoring**

> In view of the potential for a first dose effect, cardiorespiratory monitoring is required for at least the first dose and may be appropriate for subsequent doses.

**Practice Points**

> IV Pyridoxine is a Special access Scheme (SAS) product and appropriate paperwork should be completed prior to use.

> Some patients with pyridoxine deficiency present when greater than four weeks old. All infants with infantile spasms or drug resistant seizures merit a trial of pyridoxine for two weeks.

> When using for homocystinuria, pyridoxine responsiveness should be assessed by measuring plasma methionine and homocysteine under basal conditions, and during a two to three week trial of pyridoxine, while ensuring a constant protein intake.
Long term management should be overseen by a paediatric neurologist or metabolic specialist.

Pyridoxine is converted to pyridoxal phosphate by pyridox(amine) phosphate oxidase, and patients with the rare recessive defect of this enzyme present with neonatal seizures that respond to pyridoxal phosphate, but not to pyridoxine. Consult the metabolic team in such cases.

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