MONOGRAPH

ELOSULFASE ALFA

Scope (Staff):	Medical, Pharmacy, Nursing
Scope (Area):	All Clinical Areas

Child Safe Organisation Statement of Commitment

CAHS commits to being a child safe organisation by applying the National Principles for Child Safe Organisations. This is a commitment to a strong culture supported by robust policies and procedures to reduce the likelihood of harm to children and young people.

This document should be read in conjunction with this DISCLAIMER

QUICKLINKS			
Dosage/Dosage Adjustments	Administration	Compatibility	Monitoring

DRUG CLASS

Elosulfase alfa is an enzyme produced by recombinant DNA technology that provides replacement therapy in conditions caused by N-acetylgalactosamine-6-sulfatase (GALNS) deficiency.[1, 2]

INDICATIONS AND RESTRICTIONS

- Used as an enzyme replacement therapy in the treatment of mucopolysaccharidosis type IVA (Morquio A syndrome).
- This drug is available on the Pharmaceutical Benefits Scheme Life Saving Drugs Program (LSDP). The prescriber and patient must be registered with LSDP to access this medication under this scheme.
- Individual Patient Application to the CAHS Drug and Therapeutics Committee (DTC) is required for patients who are ineligible for elosulfase treatment under the LSDP access.
- **See Formulary 1**.

CONTRAINDICATIONS

- Hypersensitivity to elosulfase alfa or any component of the formulation.[1]
- Patients with fructose intolerance should not take this medicine.[1]

PRECAUTIONS

- Anaphylaxis and severe allergic reactions are possible as well as infusion reactions. Appropriate medical support should be readily available if required.[1]
- Patients with acute febrile illness or respiratory infection at the time of elosulfase alfa infusion are at a higher risk of complications from hypersensitivity reactions. Give careful consideration to the patient's clinical status and consider delaying infusion.[1]
- Sleep apnoea is common in patients with Morquio A syndrome; consider evaluating airway patency prior to initiation of treatment. Patients using supplemental oxygen or continuous positive airway pressure (CPAP) should have these treatments readily available during the infusion in case of an acute reaction or extreme drowsiness/sleep from antihistamines.[1]
- Spinal/cervical cord compression is a known serious complication in Morquio A syndrome and was also observed in those receiving treatment; monitor for symptoms e.g. back pain, limb paralysis, urinary and faecal incontinence.[3]
- Sodium content should be taken into consideration as each vial contains 8 mg of sodium and it is administered diluted in sodium chloride 0.9%.[1]

FORMULATIONS

Listed below are products available at PCH. Other formulations may be available, check with pharmacy if required:

- Elosulfase alfa 1 mg/mL solution. Each vial of approximately 5 mL extractable solution contains 5 mg elosulfase alfa.

DOSAGE & DOSAGE ADJUSTMENTS

Mucopolysaccharidosis type IVA

≥ 9 months: 2 mg/kg administered once a week intravenously.[1, 3]

Renal impairment:

- No data available.[1-3]

Hepatic impairment:

- No data available.[1-3]

RECONSTITUTION & ADMINISTRATION

IV infusion only (Do not administer as an IV push or bolus)

- Administer premedication (e.g. antihistamine, paracetamol, corticosteroids) as prescribed 30 - 60 minutes prior to commencing infusion.
- Chart elosulfase alfa on a Parenteral Fluid Therapy Order Chart (MR828) along with a 29 mL sodium chloride 0.9% flush.
- **Prepared in Pharmacy Compounding Services (PCS).**
- Diluted in sodium chloride 0.9% to a volume of 100 mL or 250 mL depending on patient body weight.[1]
    - ≥ 5 kg to < 25 kg in final volume 100 mL
    - ≥ 25 kg in final volume 250 mL
- **Do not shake the bag.**
- **Do not use** if opaque particles or discolouration are visible.
    - A diluted solution with slight flocculation (e.g. thin translucent fibres) is acceptable for administration.[1]

Infusion rate[1, 4] Refer also to Appendix A: Recommended Infusion Volumes and Rates

For 100 mL

- The initial rate will be 3 mL/hour for the first 15 minutes.
- If well tolerated, increase the rate to 6 mL/hour for the next 15 minutes.
- If well tolerated increase rate every 15 minutes by 6 mL/hour increments until a maximum rate of 36 mL/hour is reached.

Refer to Appendix A.[1]

For 250 mL

- The initial rate will be 6 mL/hour for the first 15 minutes.
- If well tolerated increase to 12 mL/hour for the next 15 minutes.
- If well tolerated increase rate every 15 minutes by 12 mL/hour increments until a maximum rate of 72 mL/hour is reached.

Refer to Appendix A.[1]

- The total volume of the infusion should be delivered over approximately 4 hours using a 0.2 micrometre in-line membrane filter.[1]
- Infusion rate may be slowed or temporarily interrupted if required to minimise infusion related reaction risk.[1]
- Flush the remaining medication through the intravenous set using 29 mL of sodium chloride 0.9% flush as per standard practice.

COMPATIBILITY *(LIST IS NOT EXHAUSTIVE)*

Compatible fluids: Sodium Chloride 0.9%.[1]

Compatible at Y-site: No information.[1]

Compatibilities of IV drugs must be checked when two or more drugs are given concurrently.

INCOMPATIBLE drugs: No information.[1]

MONITORING

Before and during infusion

- Medications for treatment of severe hypersensitivity reactions, including anaphylaxis, should be available for immediate use if needed.
- Monitor temperature, blood pressure and pulse rate every 30 minutes for the first hour then hourly until the end of the infusion including sodium chloride 0.9% flush, and for 30 minutes post infusion. Monitoring 30 minutes post infusion is not required if Consultant agrees to discharge immediately post flush.

After infusion

- Parent/patient to wait in the department for 30 minutes after administration, ***or earlier depending on Consultant approval for regular patients,*** to facilitate monitoring of any allergic or adverse reaction to elosulfase alfa.[1]
- Monitor for signs and symptoms of spinal or cervical compression.[1]

ADVERSE EFFECTS

Common: anaphylactic reactions, infusion related reactions (headache, nausea, vomiting, pyrexia, fatigue, chills and abdominal pain).[1]

STORAGE

- Elosulfase vials must be refrigerated between 2 and 8°Celsius. Do not freeze. Store in original packaging to protect from light.
- Once reconstituted and diluted, it must be refrigerated between 2 and 8°Celsius and is stable for up to 24 hours refrigerated followed by 24 hours at room temperature.[1, 4]

INTERACTIONS

This medication may interact with other medications; consult PCH approved references (e.g. Clinical Pharmacology), a clinical pharmacist or PCH Medicines Information Service on extension 63546 for more information.

Please note: The information contained in this guideline is to assist with the preparation and administration of **Elosulfase alfa**. Any variations to the doses recommended should be clarified with the prescriber prior to administration

Related external legislation, policies and guidelines
Guiding Principles for Medicines Access Programs in Western Australian Public Hospitals Western Australia Therapeutics Advisory Group publication
Life Saving Drugs Program Australian Government; Department of Health and Aged Care

References

1. AusDI *[full product information]*. 2025,Health Communication Network Pty Limited. Available online from : Vimizim - AusDI
2. *Clinical Pharmacology powered by ClinicalKey*. 2025, Elsvier: Philadelphia (PA).
3. Zand JM and Sterling A (ed), *Up To Date - Paediatric Drug information*, in *Up To Date*. 2025, Lexicomp.
4. Symons K. Ermer J. (editors), *Australian injectable drugs handbook*. 2025, The Society of Hospital Pharmacists of Australia: Collingwood.

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Appendix A: Recommended Infusion Volumes and Rates[1]

	Step 1 Initial Infusion Rate		Step 2	Step 3	Step 4	Step 5	Step 6	Step 7
Patient Weight	Total Infusion Volume	0 – 15 minutes	15 – 30 minutes	30 – 45 minutes	45 – 60 minutes	60 – 75 minutes	75 – 90 minutes	90+ minutes
(kg)	(mL)	(mL/hr)	(mL/hr)	(mL/hr)	(mL/hr)	(mL/hr)	(mL/hr)	(mL/hr)
≥ 5 and < 25	100	3	6	12	18	24	30	36
≥ 25	250	6	12	24	36	48	60	72