Scottish Paediatric & Adolescent Rheumatology Network (SPARN)

Guidance for Iloprost Use

NOTE
This guidance is not intended to be construed or to serve as a standard of care. Standards of care are determined based on all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guidance recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guidance or any local guidance derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

This guidance has been prepared by NHS National Services Scotland (NSS) National Networks. Accountable to Scottish Government, NSS works at the heart of the health service providing national strategic services to the rest of NHSScotland and other public sector organisations to help them deliver their services more efficiently and effectively. Working across professional and organisational boundaries, National Networks support the delivery of safe, effective healthcare that's designed around patients, carers and families.

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Contents

Authors	**C Anderson**, Consultant Paediatric Rheumatologist, NHS Lothian **M Dunbar**, Senior Pharmacist, NHS Highland
Stakeholders involved	Consultant Paediatric Rheumatologists, Consultant Paediatricians, Rheumatology Nurse Specialists, Occupational Therapist, Lead Pharmacist, Highly Specialist Physiotherapist
Methodology used	• Literature search • Review of evidence • Review of available national and international guidance including UK Paediatric Rheumatology guidance (BSPAR) • Engagement with key stakeholders (see above) • Guidance drafted with review date • Submitted to Steering Group (see appendix 1) for comment then approval
Rationale	This guidance is helpful and useful as a practical guide to the use of iloprost[1] to treat digital ulceration and severe Raynaud's. No appropriate national alternative exists.
Scope	For rheumatology specialists including hospital staff involved in management of children and young people with digital ulceration and severe Raynaud's.
Approval process	The guidance was approved by the SPARN Steering Group on 20/02/2026. See appendix 2 for list of Steering Group members.

[1] See Appendix 1 for more information

Introduction

Iloprost is a synthetic analogue of prostacyclin, a naturally occurring prostaglandin in the body. It is a vasodilator and a platelet aggregation inhibitor. Iloprost is not licensed in the UK for use in children and treatment must be initiated by a consultant who retains responsibility for the treatment.

Indications for use

- Severe Raynaud's disease unresponsive to other therapies or critical ischaemia.

Contraindications

- History of hypersensitivity to iloprost or any of its excipients
- Severe heart failure, heart disease or arrhythmias
- Pulmonary veno-occlusive disease
- Conditions which may increase the risk of haemorrhage e.g. active peptic ulcers, trauma
- Pregnancy

Cautions

- Renal and hepatic impairment
- Hypotension
- Severe asthma or acute pulmonary infections
- Concurrent use of medicines that have anticoagulant or antiplatelet effects may increase the risk of bleeding. See BNFC to check for drug interactions before starting.

Initiation of therapy and baseline assessment

- Full clinical history.
- Height and weight
- Bloods including FBC, ESR, U&E, LFT and CRP.
- Pregnancy test if appropriate.
- Responsible consultant to discuss risks and benefits of treatment and document discussion in patient's medical notes.

Dosage

- The dose is adjusted according to individual tolerability within the range of 0.5 nanograms/kg/min to 2 nanograms/kg/min.
- Infusion rates are calculated as per Table 1. Refer to Appendix 3 to double check calculation.
- Dose is administered by intravenous infusion daily for 5 to 10 days and is given over 6 hours.

Table 1: Calculation of iloprost infusion rates			
Step	**Dose**	**Calculation**	**Infusion rate of 200 nanogram/ml solution**
1	0.5 nanograms/kg/min	Dose (nanograms/kg/min) x Weight (kg) x 60 ---------- 200	……..ml/hr
2	1 nanogram/kg/min		……..ml/hr
3	1.5 nanogram/kg/min		……..ml/hr
4	2 nanogram/kg/min (maximum rate)		……..ml/hr

Example calculation

Initial infusion rate of 0.5 nanograms/kg/min of a 200nanogram/ml solution for a 20kg patient would be:

$$\frac{0.5 \text{ (nanograms/kg/min)} \times 20 \text{ (kg)} \times 60}{200} = 3\text{ml/hr}$$

Maximum infusion rate of 2 nanograms/kg/min of a 200nanogram/ml solution for a 20kg patient would be:

$$\frac{2 \text{ (nanograms/kg/min)} \times 20 \text{ (kg)} \times 60}{200} = 12\text{ml/hr}$$

Possible side effects

- Headache, flushing, nausea, vomiting and sweating
- Abdominal pain, diarrhoea, cramping and constipation
- Agitation, sedation, drowsiness, dizziness, vertigo, restlessness and confusion
- Changes in blood pressure, tachycardia, arrhythmias,
- Dyspnoea, asthma, cough and pulmonary oedema
- Myalgia, arthralgia, jaw pain, malaise.
- Local infusion site reactions – redness, site pain and erythema
- See BNFC or Iloprost summary of product characteristics, available online at Iloprost 100 micrograms/ml concentrate for solution for infusion - Summary of Product Characteristics (SmPC) - (emc) | 10034 for full list of side effects.

Prescribing

- Iloprost is prescribed on a fluid administration chart. Example infusion rates are shown in Appendix 3.
- At any point during the patients' infusion if they are experiencing unacceptable side effects the clinical team should be contacted for review and consider reduction of the rate.
- Note not all side effects require a rate change; this medication will induce mild symptoms, and is a sign of sufficient treatment, patient should be warned to expect mild headache". Paracetamol could be considered for symptomatic relief.
- From day 4, the infusion should be prescribed from the start at the highest tolerated infusion rate determined on days 1 to 3.
- Iloprost can be pre-prescribed for a number of days at a time at the discretion of the prescribing doctor but they should be contacted at any stage if there are any concerns.

Preparation

- Add the contents of one ampoule (50 micrograms in 0.5ml) to a 250ml bag of 0.9% sodium chloride. This will provide a solution with a final concentration of 200 nanograms/ml.
- Visually inspect the infusion for particulate matter or discolouration or foreign particles.
- Ensure batch numbers of the Iloprost and the sodium chloride 0.9% used are documented on the administration chart.
- Any Iloprost solution left at the end of the patient's infusion should be discarded and a new Iloprost infusion solution prepared each day.

Monitoring prior to infusion

- Prior to the infusion carry out a clinical evaluation to ensure the child is well. If the child is unwell or febrile on presentation, ward staff to evaluate for possible underlying infection, and discuss with responsible consultant prior to commencing infusion.
- If applicable (suggest females >12 years), check that the patient is not pregnant and perform a pregnancy test if necessary.
- Site an intravenous cannula in an appropriate vein.
- Take bloods to check FBC, ESR, U&E, LFT and CRP.
- Record baseline temperature, pulse, respiratory rate, blood pressure, immediately prior to commencing the infusion.

During and post infusion

- Monitor temperature, pulse, respiratory rate and blood pressure every 30 minutes until the patient is at their maximum infusion rate and then monitor hourly until the end of the infusion and for 1 hour post infusion.
- Some side effects are expected and patients may be able to tolerate them for the length of the infusion. If the patient experiences unacceptable side effects the rate can be reduced until a tolerable dose is reached. If side effects are

severe administration should be stopped. Advise patients and carers to report any signs of infection or possible side effects.
- Advise patients and carers to seek immediate medical advice if hypersensitivity symptoms occur.

Administration

- Prior to starting the infusion ensure emergency drugs are available in case of any adverse reaction.
- Check all prescribed rates of Iloprost are correct for the patient.
- On day 1 to 3 the infusion rate is started at 0.5 nanograms/kg/min and then gradually increased every 30 minutes until the patient experiences unacceptable side effects or the maximum dose is achieved (maximum dose 2 nanograms/kg/min).
- The infusion should be stopped after 6 hours, regardless of total dose administered.
- From day 4, the infusion can be administered at the highest tolerated infusion rate determined on days 1 to 3, from the start of the infusion.

Appendix 1 – Off label drugs statement

Prescribing of medicines outwith their marketing authorisation
Recommendations within this pathway are based on the best clinical evidence. Some recommendations may be for medicines prescribed outwith the marketing authorisation (MA) also known as product licence. This is known as 'off-label' use. Medicines may be prescribed 'off-label' in the following circumstances:

- for an indication not specified within the marketing authorisation
- for administration, via a different route
- for administration
- for a different dose for a different patient population.

An unlicensed medicine is a medicine which does not have MA for medicinal use in humans. Generally, 'off-label' prescribing of medicines becomes necessary if the clinical need cannot be met by licensed medicines within the marketing authorisation. Such use should be supported by appropriate evidence and experience.

"Prescribing medicines outside the conditions of their marketing authorisation alters (and probably increases) the prescribers' professional responsibility and potential liability".

The General Medical Council (GMC) recommends that when prescribing a medicine 'off-label', doctors should:

- be satisfied that there is no suitably licensed medicine that will meet the patient's need
- be satisfied that there is sufficient evidence or experience of using the medicine to show its safety and efficacy
- take responsibility for prescribing the medicine and for overseeing the patient's care, including monitoring the effects of the medicine, and any follow-up treatment, or ensure that arrangements are made for another suitable doctor to do so.

Make a clear, accurate and legible record of all medicines prescribed and when not following common practice, the reasons for prescribing an unlicensed medicine. Non-medical prescribers should ensure that they are familiar with the legislative framework and Royal Pharmaceutical Society's Competency Framework for all Prescribers.

Prior to any prescribing, the licensing status of a medication should be checked in the summary of product characteristics (www.medicines.org.uk). The prescriber must be competent, operate within the professional code of ethics of their statutory bodies and the prescribing practices of their employers.

Appendix 2 – Steering Group membership

Name	Designation	Role	Area representing
Andrew Fell	Paediatric Rheumatology Nurse Specialist	Data Lead	NHS Greater Glasgow & Clyde
Angela Cruickshank	Paediatric Rheumatology Nurse Specialist	Former Nurse Lead	NHS Fife
Catriona Anderson	Consultant Paediatric Rheumatologist	Education Lead	NHS Lothian
Elaine Wallace	Senior Child Health Physiotherapist	Physio Lead	NHS Tayside
Emma Carson	Paediatric Rheumatology Nurse Specialist	Working with Families Lead	NHS GGC
Imogen Kelly	Rheumatology Nurse Specialist	Working with Families Lead	NHS Lothian
Jane Adam	Paediatric Rheumatology Nurse Specialist	Nurse Lead	NHS Grampian
Julie Duncan	Consultant Paediatrician	Clinical Guidance Lead	NHS Lothian
Karen Lapsley	Highly Specialist Physiotherapist	Physio Lead	NHS Forth Valley
Kirsten Healy	Consultant Paediatrician	Paediatrician with an interest	NHS Fife
Kirsty McLellan	Paediatric Rheumatology Consultant	Clinical Guidance Lead	NHS Greater Glasgow & Clyde
Klaire Connor	Young People and Families Manager Scotland	Third Sector Representative	Versus Arthritis
Lindsay Robertson	Consultant Rheumatologist	Transition Lead	NHS Grampian
Lois Freeland	SNAC Chair	Third Sector Representative	SNAC
Mairi Dunbar	Lead Pharmacist – Paediatrics	Pharmacy Lead	NHS Tayside
Mandy Fanning	Occupational Therapist	Occupational Therapy Lead	NHS GGC
Mary Brennan	Consultant Paediatric Rheumatologist	Chair	NHS Lothian
Neil Martin	Consultant Paediatric Rheumatologist	Lead Clinician	NHS Greater Glasgow & Clyde

Appendix 3 - Example infusion rates using diluted iloprost 200 nanograms/ml infusion

Table 2				
Patients weight	**Step 1 (initial rate)**	**Step 2**	**Step 3**	**Step 4 (max rate)**
	0.5 nanograms/kg/min	1 nanograms/kg/min	1.5 nanograms/kg/min	2 nanograms/kg/min
10kg	1.5ml/hr	3ml/hr	4.5ml/hr	6ml/hr
15kg	2.3ml/hr	4.5ml/hr	6.8ml/hr	9ml/hr
20kg	3ml/hr	6ml/hr	9ml/hr	12ml/hr
25kg	3.8ml/hr	7.5ml/hr	11.3ml/hr	15ml/hr
30kg	4.5ml/hr	9ml/hr	13.5ml/hr	18ml/hr
35kg	5.3ml/hr	10.5ml/hr	15.8ml/hr	21ml/hr
40kg	6ml/hr	12ml/hr	18ml/hr	24ml/hr
45kg	6.8ml/hr	13.5ml/hr	20.3ml/hr	27ml/hr
50kg	7.5ml/hr	15ml/hr	22.5ml/hr	30ml/hr
55kg	8.3ml/hr	16.5ml/hr	24.8ml/hr	33ml/hr
60kg	9ml/hr	18ml/hr	27ml/hr	36ml/hr
65kg	9.8ml/hr	19.5ml/hr	29.3ml/hr	39ml/hr
70kg	10.5ml/hr	21ml/hr	31.5ml/hr	42ml/hr
75kg	11.3ml/hr	22.5ml/hr	33.8ml/hr	45ml/hr
80kg	12ml/hr	24ml/hr	36ml/hr	48ml/hr
All patients are started at step 1 (initial rate) and the infusion rate may be increased every 30minutes up to step 4 (max rate). Infusion rates should only be increased if patient tolerates current rate and is not experiencing unacceptable side effects.				

References

- Callaghan C, Clifford D. Guidance for the Preparation and Administration of iloprost (50micrograms/0.5ml). NHS Lothian Pharmacy Service/Rheumatic Disease Unit. Version 7.
- Iloprost 100 micrograms/ml concentrate for solution for infusion - Summary of Product Characteristics (SmPC) - (emc) | 10034
- Foster H, Brogan PA. Oxford Specialist Handbook in Paediatrics: Paediatric Rheumatology. Second edition.
- British National Formulary: MedicinesComplete — CONTENT > BNF > Drug: Iloprost
- Kettering General Hospital NHS Foundation Trust. Intravenous iloprost guideline. MMG31. Medicines Management Committee. Review date August 2020.

Contact email address: nss.sparn@nhs.scot