

- **TREATMENT CESSATION** Avoid abrupt withdrawal (risk of rebound pulmonary hypertension and pulmonary hypertensive crisis).
- **DIRECTIONS FOR ADMINISTRATION** Directions for administration vary depending on the preparation used—for instructions in *adults*, consult product literature. For *neonatal intensive care*—consult local protocols.

- **MEDICINAL FORMS** There can be variation in the licensing of different medicines containing the same drug.

Powder for solution for infusion

- ▶ **Veletri** (Actelion Pharmaceuticals UK Ltd)
Epoprostenol (as Epoprostenol sodium) 500 microgram Veletri 500microgram powder for solution for infusion vials | 1 vial [PoM] £24.44
- ▶ **Epoprostenol (as Epoprostenol sodium) 1.5 mg** Veletri 1.5mg powder for solution for infusion vials | 1 vial [PoM] £49.24

Powder and solvent for solution for infusion

- ELECTROLYTES: May contain Sodium
- ▶ **Flofan** (GlaxoSmithKline UK Ltd)
Epoprostenol (as Epoprostenol sodium) 500 microgram Flofan 500microgram powder and solvent (pH12) for solution for infusion vials | 1 vial [PoM] £22.22
 - ▶ **Epoprostenol (as Epoprostenol sodium) 1.5 mg** Flofan 1.5mg powder and solvent (pH12) for solution for infusion vials | 1 vial [PoM] £44.76

Iloprost

20-Feb-2019

● INDICATIONS AND DOSE

Idiopathic or familial pulmonary arterial hypertension (initiated under specialist supervision)

- ▶ BY INHALATION OF NEBULISED SOLUTION
- ▶ Child 8–17 years: Initially 2.5 micrograms for 1 dose, increased to 5 micrograms for 1 dose, increased if tolerated to 5 micrograms 6–9 times a day, adjusted according to response; reduced if not tolerated to 2.5 micrograms 6–9 times a day, reduce to lower maintenance dose if high dose not tolerated

Raynaud's syndrome

- ▶ BY INTRAVENOUS INFUSION
- ▶ Child 12–17 years: Initially 30 nanograms/kg/hour, increased to 60–120 nanograms/kg/hour daily for 3–5 days, dose to be given over 6 hours, dose increase should be performed gradually

- **UNLICENSED USE** Not licensed for use in children.
- **CONTRA-INDICATIONS** Conditions which increase risk of haemorrhage · congenital or acquired valvular defects of the myocardium · decompensated cardiac failure (unless under close medical supervision) · pulmonary veno-occlusive disease · severe arrhythmias · severe coronary heart disease

● CAUTIONS

GENERAL CAUTIONS Hypotension (do not initiate if systolic blood pressure below 85 mmHg) · unstable pulmonary hypertension with advanced right heart failure

SPECIFIC CAUTIONS

- ▶ When used by inhalation Acute pulmonary infection · severe asthma
- **INTERACTIONS** → Appendix 1: iloprost
- **SIDE-EFFECTS**
- ▶ **Common or very common** Chest discomfort · cough · diarrhoea · dizziness · dyspnoea · haemorrhage · headache · hypotension · nausea · oral disorders · pain · palpitations · rash · syncope · tachycardia · throat complaints · vasodilation · vomiting
- ▶ **Frequency not known** Respiratory disorders · taste altered · thrombocytopenia

● PREGNANCY

- ▶ When used by inhalation Use if potential benefit outweighs risk.

● BREAST FEEDING

- ▶ When used by inhalation Manufacturer advises avoid—no information available.

● HEPATIC IMPAIRMENT

- ▶ **Dose adjustments** ▶ When used by inhalation Initially 2.5 micrograms at intervals of 3–4 hours (max. 6 times daily), adjusted according to response (consult product literature).
Dose may need to be halved in liver cirrhosis.

● DIRECTIONS FOR ADMINISTRATION

- ▶ With intravenous use For *intravenous infusion* dilute to a concentration of 200 nanograms/mL with Glucose 5% or Sodium Chloride 0.9%; alternatively, may be diluted to a concentration of 2 micrograms/mL and given via syringe driver.
- ▶ When used by inhalation For *inhaled treatment*, to minimise accidental exposure use only with nebulisers listed in *Ventavis*® product literature in a well ventilated room.
- **PRESCRIBING AND DISPENSING INFORMATION**
- ▶ When used by inhalation Delivery characteristics of nebuliser devices may vary—only switch devices under medical supervision.
- ▶ With intravenous use Concentrate for infusion available on a named patient basis from Bayer Schering in 0.5 mL and 1 mL ampoules.

- **MEDICINAL FORMS** There can be variation in the licensing of different medicines containing the same drug.

Solution for infusion

▶ Iloprost (Non-proprietary)

- Iloprost (as Iloprost trometamol) 100 microgram per 1 ml Iloprost 100micrograms/1ml solution for infusion ampoules | 1 ampoule [PoM] [X]
- Iloprost 50micrograms/0.5ml solution for infusion ampoules | 1 ampoule [PoM] [X]
- Iloprost 100micrograms/1ml concentrate for solution for infusion ampoules | 1 ampoule [PoM] £75.00 (Hospital only) | 5 ampoule [PoM] £300.00 (Hospital only)

Nebuliser liquid

- ▶ **Ventavis** (Bayer Plc)
Iloprost (as Iloprost trometamol) 10 microgram per 1 ml Ventavis 10micrograms/ml nebuliser solution 1ml ampoules | 42 ampoule [PoM] £560.27 | 168 ampoule [PoM] £2,241.08
- Iloprost (as Iloprost trometamol) 20 microgram per 1 ml Ventavis 20micrograms/ml nebuliser solution 1ml ampoules | 42 ampoule [PoM] £560.27 | 168 ampoule [PoM] £2,241.08

ENDOTHELIN RECEPTOR ANTAGONISTS

Bosentan

07-Jun-2018

● INDICATIONS AND DOSE

Pulmonary arterial hypertension (initiated under specialist supervision)

▶ BY MOUTH

- ▶ Child 2–17 years (body-weight 10–20 kg): Initially 31.25 mg once daily for 4 weeks, then increased to 31.25 mg twice daily
- ▶ Child 2–17 years (body-weight 20–40 kg): Initially 31.25 mg twice daily for 4 weeks, then increased to 62.5 mg twice daily
- ▶ Child 12–17 years (body-weight 40 kg and above): Initially 62.5 mg twice daily for 4 weeks, then increased to 125 mg twice daily (max. per dose 250 mg)

- **CONTRA-INDICATIONS** Acute porphyrias p. 624

- **CAUTIONS** Not to be initiated if systemic systolic blood pressure is below 85 mmHg

- **INTERACTIONS** → Appendix 1: bosentan