

Title: Resuscitation of blue baby and use of prostaglandins 2010	
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Related Policies :PICU Drug Infusion Policies	Replaces/updates: Congenital cardiac disease 2006

1. Aims and scope of this policy

Early management of the blue baby admitted to the Neonatal unit or Paediatrics. This guideline was written for the Central South Coast Neonatal Network. It is now updated and incorporated for use within the PICF network.

2. Key points

This document relates to the acute management of infants presenting acutely with collapse and/or cyanosis suggestive of CHD. Significant proportions of infants with congenital cardiac disease go undetected antenatally and may have a normal first day check. It can present in several ways, but tends to be in one of 3 groups:

- 1) Heart murmur in an otherwise healthy child
- 2) CHD presenting as a "blue baby"
 - a. Transposition of the great arteries (duct dependent)
 - b. Severe right sided obstructive lesions (duct dependent)
 - c. Defects causing mixing of blood from the pulmonary and systemic circulation in the heart
 - d. Pulmonary venous drainage problems
- 3) CHD presenting as a collapsing, but not necessarily cyanosed baby
 - a. Severe left sided obstructive lesions (duct dependent)

b. Cardiomyopathy

Infants with duct dependent lesions present as the duct closes, often in extremis. Awareness of congenital heart disease is crucial to allow recognition and appropriate resuscitation. Good communication with the PICU Consultant and Paediatric Cardiologists is essential.

All unwell infants with congenital heart disease must be discussed with PICU as well as cardiology particularly if a transfer is required.

Please Note: Alprostadil (PGE1) has been replaced by Dinoprostone (PGE2) as the prostaglandin of choice. It has the same characteristics and dose range as alprostadil. However please refer to the PICU drug infusion guidance (available on the intranet at SRH) for preparation details.

GUIDANCE

1. Indications

1.1 Background

Infants with profound cyanosis are a medical emergency requiring senior help they should be considered for prostaglandin infusion and commenced on parenteral antibiotics.

1.2 Aim/purpose

This protocol gives guidance on when to start prostaglandins for suspected congenital cardiac defects.

1.4 Exceptions/ contraindications

This protocol is intended for use in facilities where accurate neonatal cardiac echo is not readily available. Those centres where it is available may wish to adapt the guidelines.

1.5 Options

At all times these infants should be discussed at Consultant level.

2. Clinical Management

Differential diagnosis of “blue baby”

- Primary lung disease
- Cardiac malformations (may present at any stage perinatally)
- Persistent pulmonary hypertension
- Sepsis

- Asphyxia

General approach to the cyanotic infant

These infants are often complex and should be managed in cooperation with the paediatric intensivist and Consultant cardiologist at Southampton.

- Manage ABC and call immediately for **senior help**.
- Parenteral antibiotics (cefotaxime).
- Confirm central cyanosis with oximetry (right arm) and arterial gas (record oxygen administered).
- Take detailed history and thorough examination looking especially for dysmorphic features. Note relevant family history.
- Measure 4 limb BP, (upper/lower discrepancy >10 mmHG is significant and suggests a coarctation). Request ECG and CXR. Do not be reassured by a normal 4 limb BP, palpate femoral pulses.
- Obtain good access ideally UVC and UAC or Peripheral arterial line.
- Correct metabolic acidosis and systemic hypoperfusion with fluid and inotropes +/- bicarbonate (initially dopamine).
 - **Hypotension in cyanotic infant is a serious finding.**
- Hyperoxia test (100% Oxygen for 10 minutes and monitor SaO₂).
- A saturation increased of 15%, would make cardiac disease unlikely.
Caution total anomalous pulmonary venous drainage and hypoplastic left heart may respond; lung disease with large intrapulmonary shunt may not.

If unclear discuss with Paediatric Cardiologist on call, the combination of clinical signs, ECG and hyperoxia test should help differentiate lung pathology from cardiac disease.

- Consider prostaglandin.

Who should be given Prostaglandin (Dinoprostone)?

This is a clinical decision usually made without formal diagnosis. Dinoprostone is used to reopen or maintain a patent duct and hence support duct dependent lesions.

1. **Presence of cyanosis** is the most useful discriminator for Dinoprostone sensitive lesions. This is further increased by presence of murmur.
2. Reduced or **absent femoral pulses** in an unwell infant.

The more clinically unwell the infant the lower threshold for starting Dinoprostone.

If in doubt there is no harm in trying a 30 to 60 minute trial of Dinoprostone followed by repeat arterial gas. Using Dinoprostone is not in its own an indication for intubation.

Dose

See PICU formulary.

Start Dinoprostone on 5 to 10 nanograms /kg/min if baby stable and saturations above 80%. In this situation it would be unusual to need more than 10 nanograms/kg/min and risk of apnoea is low at this dose. If baby is deeply desaturated or with circulatory collapse (left heart obstruction) start at 20 nanograms/kg/min. If no response can try 50 – 100 nanogrammes/kg/min but discuss with Cardiologist. Aim to decrease to 10 nanogrammes/kg/min after improvement.

Risk of apnoea, need for volume expansion and inotrope support increases with dose.

Who to intubate?

- Presence of apnoea on prostaglandin.
- Clinical state of the infant (acidotic, shock, severe distress and tachypnoea).
- Requiring 30 nanograms/kg/min or more of Dinoprostone. (but always discuss first with PICU).

Therefore, in a stable infant with an Dinoprostone responsive ductal lesion, transport without intubation may be appropriate.

Ventilator management

- Aim for pCO₂, 5 – 6 kPa and right arm oxygen saturations 75 – 85%.
- Sedate well with morphine and consider paralysis.

Troubleshooting

Always discuss with PICU.

1) *Continuing problems or a deterioration in systemic perfusion, (oliguria, shock, metabolic acidosis and myocardial dysfunction).*

This may be a critically obstructed systemic circulation where increases in pulmonary blood flow have reduced systemic flow. Need to balance pulmonary and systemic circulations by:

- Dinoprostone at rate sufficient to maintain ductal patency.
- Reduce pulmonary circulation by ventilating with a modest PEEP (4 – 6 cm H₂O) and in room air in the first instance to maintain systemic saturations 75% – 85%, avoiding respiratory alkalosis with pCO₂ 5 – 6 kPa.
- If signs persist, review dinoprostone dose, need for volume expansion and correction of anaemia. Discuss with PICU management and need for low

dose inotrope or nitroprusside (if BP will allow). High dose inotrope in this situation may increase systemic vascular resistance and worsen systemic blood flow.

An example of this is **hypoplastic left heart syndrome**. This situation can sometimes be difficult to differentiate from severe sepsis.

2) No improvement in saturations despite Dinoprostone.

It can be very difficult to differentiate **persistent pulmonary hypertension** (PPH) from cyanotic heart disease. These infants would normally respond to nitric oxide (available on PICU transfer) and benefit from good oxygenation, alkalosis (bicarbonate or brief hyperventilation to raise pH to >7.55) and raised systemic pressure with use of inotropes. Discuss management with PICU; safe transfer for echocardiogram is needed.

In **complete transposition**, mixings prevented by having a virtually intact atrial septum; an emergency balloon septostomy can be lifesaving.

3) Cyanotic heart disease masquerading as lung pathology

Total anomalous pulmonary venous drainage (TAPVD) can present with an abnormal CXR suggesting lung disease with pulmonary hypertension. Although a classical X Ray appearance is described, more frequently, it is indistinguishable from common lung diseases such as pneumonia and meconium aspiration. Therefore these infants need safe transfer for echocardiogram.

Potential Complications

Transport

It is sometimes difficult to decide whether an infant should be transferred to either a PICU or NICU. In particular if persistent pulmonary hypertension is a likely differential. If the infant is transferred to a neonatal unit a formal cardiac echo should be arranged as soon as possible.

Drug errors can be caused by

1. Miscalculation of Dinoprostone dose, therefore use only the Southampton PICU formulary.
2. To avoid confusion between the different prostaglandins some recommend the use of the name Dinoprostone (rather than E2 etc). In this document "Dinoprostone" has been used in preference to "Prostaglandin E2".

3. Audit

Transfer of cyanotic infants within the network. Use and timing of prostaglandin.

4. Evidence Base

5.1 Sources of information and others involved in preparation

Peter Wilson, PICU Consultant, Southampton General Hospital
James Gnanapragasam, Consultant Paediatric Cardiologist (and again in 2010)
Ian Rodd, Consultant Paediatrician, RHCH
Members of CSCNN

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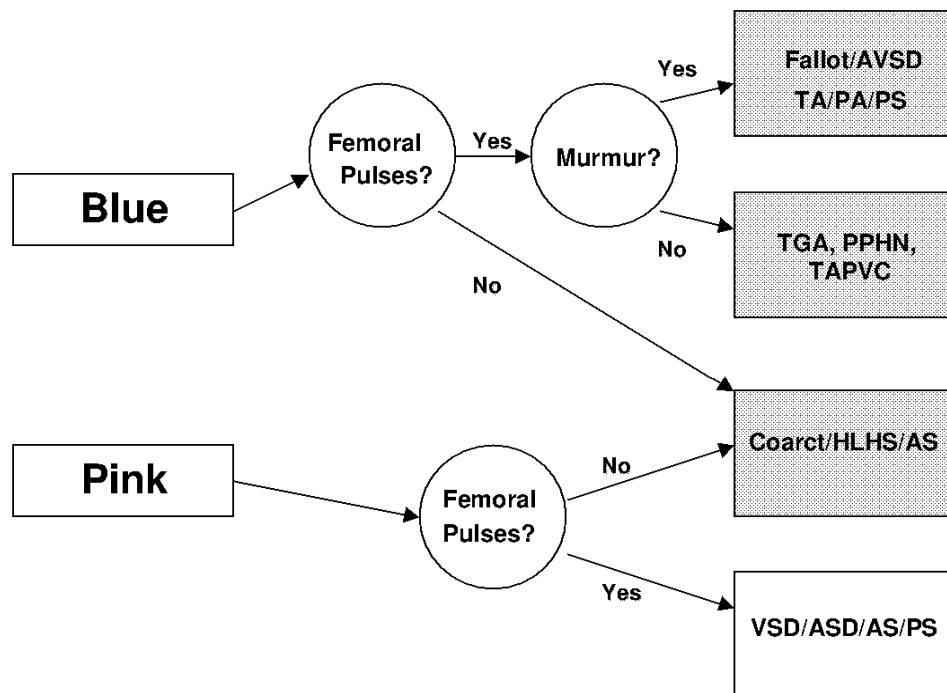
CSCNN Guidelines Group involved in preparation of 2006 version

References

Penny DJ. Management of the neonate with symptomatic heart disease. Arch Dis Child Fetal Neonatal Ed 2001;84:F14-145

Appendix

Quick differential diagnosis of cardiac lesions (CATS WEBSITE 2009)



Grey box = potential duct dependent lesion: START PROSTIN
Consider iNO if pulmonary hypertension likely

3. Main policy

[Refer to any legislative, regulatory framework or professional guidance within which the policy operates.

List any penalties or other outcomes linked to non-compliance.

Give its distribution and publicity schedules and implementation procedure as an appendix.]

[End of template].

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[Use Arial typeface. Titles in 16pt bold, headings in 14pt bold, text in 12pt.]

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