Worcestershire Acute Hospitals NHS Trust

Cyanotic Congenital Heart Disease

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The following guidance is taken from the Partners In Paediatrics (PIP)

CYANOTIC CONGENITAL HEART DISEASE

RECOGNITION AND ASSESSMENT

Symptoms and signs

- Central cyanosis may be respiratory or cardiac in origin
- Respiratory illness producing cyanosis will usually have signs of respiratory distress (e.g. cough, tachypnoea, recession and added respiratory sounds)
- Cardiac decompensation may occur with a respiratory infection; they may co-exist
- Cyanosis more likely due to cardiac disease if:
- SpO_2 responds poorly to high flow oxygen (15 L/min) via face mask with reservoir bag
- marked tachycardia
- enlarged heart (clinically or on CXR)
- gallop rhythm/murmur
- enlarged liver/raised JVP
- basal crackles
- absent femoral pulses
- finger clubbing occurs after a few months (also consider endocarditis)

Causes of cardiac cyanosis

Significant right-to-left shunt

- Transposition with inadequate mixing, pulmonary or tricuspid atresia
- Fallot's tetralogy: hypercyanotic episodes follow emotional or painful upset

Duct-dependent pulmonary circulation

- Commonly presents in first 10–14 days of life
- severely blue, breathless or shocked
- pulmonary atresia
- critical pulmonary valve stenosis
- tricuspid atresia
- severe Fallot's tetralogy
- transposition of the great arteries without septal defect
- single ventricle anatomy

Acute pulmonary outflow obstruction (cyanotic episodes)

Fallot's tetralogy or other complex congenital cyanotic heart disease
severe pallor
loss of consciousness
convulsions

Physical examination

- Remember to check femoral pulses
- If coarctation of the aorta suspected: check BP in upper and lower limbs – normal difference <15 mmHg

Investigations

If infant cyanosed or in heart failure, discuss urgency of investigations with consultant

SpO_2

- Check pre (right arm) and postductal (lower limbs)
- when breathing air before oxygen given
- after giving 15 L/min oxygen by mask with a reservoir bag for 10 min

Chest X-ray

- For cardiac conditions, specifically record:
- cardiac situs (normal or right side of chest)

- aortic arch left or right-sided
- bronchial situs (is right main bronchus on the right?)
- cardiac size and configuration
- size of pulmonary vessels and pulmonary vascular markings

Electrocardiogram
See **ECG interpretation** guideline

Nitrogen washout in cyanosed babies
- Monitor SpO_2 in air then in headbox after breathing 100% oxygen for 10 min
- in cyanotic congenital heart disease, PaO_2 will remain below 20 kPa with SpO_2 unchanged
- not as reliable as echocardiogram

Echocardiogram
Locally, if available, or refer to regional paediatric cardiac centre

IMMEDIATE TREATMENT

If infant cyanosed or in heart failure, discuss urgency of referral to local paediatric cardiac surgical centre with consultant

Duct-dependent congenital heart disease
- Immediate treatment before transfer to paediatric cardiac centre:
- open duct with alprostadil (prostaglandin E_1) or dinoprostone (E_2): see **Prostaglandin infusion**

Acute pulmonary outflow obstruction (cyanotic episodes)
Immediate treatment before transfer to paediatric cardiac centre:
- do not upset child
- give morphine 50–100 microgram/kg IV over 5 min or IM
- provide high concentration face mask oxygen (15 L/min with reservoir bag)
- if Fallot's tetralogy has been diagnosed by echocardiography, discuss use of IV beta-blocker with cardiologist

SUBSEQUENT MANAGEMENT
- On advice of consultant and paediatric cardiac centre

PROSTAGLANDIN INFUSION
Dosage
- Ranges from 5–50 nanogram/kg/min (higher doses may be advised by cardiologist)
- Antenatal diagnosis of duct dependent lesion:
- start at 5 nanogram/kg/min
- Cyanotic baby or with poorly palpable pulses who is otherwise well and non-acidotic:
- start at 5–15 nanogram/kg/min
- Acidotic or unwell baby with suspected duct dependent lesion:
- start at 10–20 nanogram/kg/min. If no response within first hour, consider an increase of up to 50 nanogram/kg/min

Desired response
- Suspected left-sided obstruction:
- aim for palpable pulses, normal pH and normal lactate
- Suspected right-sided obstruction:
- aim for SpO_2 75–85% and normal lactate
- Suspected or known transposition of the great arteries (TGA) or hypoplastic left or right heart syndrome with SpO_2 <70% or worsening lactate
- liaise urgently with cardiology and/or intensive care/retrieval team as rapid assessment and atrial septostomy may be necessary

Preparations

Dinoprostone (prostaglandin E_2) is the recommended prostaglandin*

Dinoprostone infusion	Other information
• Standard dinoprostone infusion • Make a solution of 500 microgram in 500 mL by adding 0.5 mL of dinoprostone 1 mg in 1 mL to a 500 mL bag of suitable diluent (glucose 5% or 10% or sodium chloride 0.45% and 0.9%) • Transfer 50 mL of this solution into a 50 mL Luer lock syringe and label • Discard the 500 mL bag immediately into clinical waste – single patient and single dose use only • Infusion rate: 0.3 mL/kg/hr = 5 nanogram/kg/min	• **Stability:** ◦ syringe stable for 24 hr • **Compatibility:** ◦ infuse dinoprostone via separate line • **Flush:** ◦ sodium chloride 0.9% at same rate as infusion • **Administration:** ◦ continuously (short half-life). Ensure 2 working points of IV access at all times ◦ infusions can be given via long line, UVC or peripherally ◦ extravasation can cause necrosis – use central access if available

*If dinoprostone IV not available, use alprostadil (prostaglandin E_1) IV as alternative (see **BNFc**)

Oral dinoprostone (see BNFc)

- Used temporarily on very rare occasions when IV access is extremely difficult
- Discuss with cardiac centre before using
- Use dinoprostone injection orally
- May not be as effective as prostaglandin IV

Side effects

Common

- Apnoea – tends to occur in first hour after starting prostaglandin or when dose increased. Consider ventilation
- Hypotension – due to systemic vasodilatation. Consider sodium chloride 0.9% 10 mL/kg bolus
- Fever
- Tachycardia
- Hypoglycaemia

Uncommon

- Hypothermia
- Bradycardia
- Convulsions
- Cardiac arrest
- Diarrhoea
- Disseminated intravascular coagulation (DIC)
- Gastric outlet obstruction
- Cortical hyperostosis
- Gastric hyperplasia (prolonged use)

Monitor

- Heart rate
- Blood pressure
- Respiratory rate
- Temperature
- Oxygen saturations
- Blood gases
- Blood glucose and lactate