PAEDIATRIC ACUTE CARE GUIDELINE	
Hypercyanotic Spells	
Scope (Staff):	All Emergency Department Clinicians
Scope (Area):	Emergency Department

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http://kidshealthwa.com/about/disclaimer/

Hypercyanotic Spells

Background

- Infants with Tetralogy of Fallot or other congenital cardiac defects causing a right ventricular outflow tract obstruction (RVOTO) are at risk of hypercyanotic spells.
- Hypercyanotic spells are rare in the neonatal period, but may occur in the first few months of life (usually 2 – 6 months of age) while the infant is awaiting surgical intervention.
- The episode is caused by an acute reduction in pulmonary blood flow associated with an increase in the magnitude of the right-to-left shunt.

Assessment

Spells are usually characterised by a paroxysmal event involving:

- Irritability and crying
- Rapid and deep respiration
- Worsening cyanosis (may be very intense)
- Decreased intensity (even disappearance) of the systolic murmur
- Severe or untreated episodes may progress to loss of consciousness, seizures or even death

Management

- Place child in the **knee-chest position**, often in carer's arms (hips flexed, knees brought up onto chest). This helps to increase systemic vascular resistance, thereby

reducing the right-to-left shunt and resulting in more blood flow through the lungs.
- **Oxygen**, mask CPAP has proved useful anecdotally in resistant cases.
- **Calm the child**, avoid painful interventions if possible. Spells will often settle with simple manoeuvres.

If hypercyanotic spells continue:

- Give intranasal **fentanyl 1 microgram/kg**

If hypercyanotic spells continue:

- Give **morphine sulphate 0.1 mg/kg** – administer this subcutaneously unless IV access is already available (don't delay).

Fentanyl and morphine suppresses the respiratory centre, thereby reducing the hyperpnoea and the resultant systemic venous overload which contributes to the right-to-left shunt. Sedation also reduces the body's oxygen requirement.

If further intervention is required:

- IV access – can be sited in hand, foot or scalp vessel (while the infant is held chest to chest).

These measures are generally sufficient to terminate the hypercyanotic spell. The child's saturation should improve and the systolic murmur becomes louder. If the child fails to respond quickly or fully, specialist management in ICU is necessary.

Any underlying condition e.g. arrhythmia should be corrected after discussion with the Cardiologist on call.

Further Possible Management

- Volume expansion: modest IV bolus 10mL/kg of 0.9% saline initially
- Treat metabolic acidosis (if base excess < -12) using sodium bicarbonate. Give **half** the mmol deficit initially and then review **(mmol deficit = 0.3 x base excess x weight in kg)**
- **If further intervention is required** consult with PICU and Cardiology

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